MCQs in Pediatrics

PART IV

ZUHAIR M. ALMUSAWI

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ZUHAIR M. ALMUSAWI

CABP, IBCLC, FRCP Edin

PROFESSOR OF PEDIATRICS

COLLEGE OF MEDICINE – UNIVERSITY OF KERBALA

CONSULTANT PEDIATRICIAN

KARBALA TEACHING HOSPITAL FOR CHILDREN

KARBALA – IRAQ

CONTRIBUTORS

AHMED TAWFIQ; CABP, MRCPCH
Consultant Pediatrician
Karbala Teaching Hospital for Children/Karbala

USAMA AL-JUMAILY; FICMS, ABMS, JBMS
Assistant Professor of Pediatrics
College of Medicine/University of Kerbala
Consultant Pediatric Hemato-Oncologist
Imam Hussein Teaching Hospital/Karbala

KHALID KHALIL ALAARAJI; CABP, FICMS, DCH
Assistant Professor of Pediatrics
College of Medicine/University of Kerbala
Consultant Pediatrician
Karbala Teaching Hospital for Children/Karbala

QAHTAN ALOBAIDAY; CABP, FICMS, FICMS (nephro)
Consultant Pediatric Nephrologist
Karbala Teaching Hospital for Children/Karbala

HASANEIN H. GHALI; FICMS, FICMS (hem/onc)
Assistant Professor of Pediatrics
College of Medicine/University of Baghdad
Consultant Pediatric Hemato-Oncologist
Children Welfare Teaching Hospital
Medical City/Baghdad

HAYDER ALMUSAWI; CABP, DCH
Pediatrician
ALKhidhir General Hospital/ ALMuthanna

AQEEL MAHDI; CABP, FICMS
Consultant Pediatrician
Karbala Teaching Hospital for Children /Karbala

HAIDAR A N ABOOD; MSc, PhD
Assistant Professor of Clinical Pharmacology
College of Medicine/University of Kerbala
Karbala Teaching Hospital for Children/Karbala

MARYAM ZUHAIR ALMUSAWI; FICMS
Pediatrician
Obstetric and Gynecology Teaching Hospital/Karbala

MUSTAFA SHIHAB AL-ANBAKI; CABP, DCH
Pediatrician
Children Welfare Teaching Hospital
Medical City/Baghdad

DEDICATION

In the time of COVID-19 pandemic, when the silence and distancing in the globe were accepted as norms; when the great armies became worthless; and when the governments became perturbed; some people accepted the challenge and sacrifice.

Those who wear the white coats; started a momentous pivotal battle, and showed great heroism towards this ever-worst crisis in life-time with no much choices.

This book was written during the time of social and physical distancing amidst COVID-19 time, where uncertainties lie ahead, waiting for the life to come back to normalcy.

To all our colleagues over the world; all medical staff; all those who are working to find a solution, to the souls of our colleagues who sacrificed, to all people who lost their lives; to all families and kids who lost their beloved ones, we dedicate this humble work.

A special greeting to the participating authors Dr. Khalid and Dr. Akeel who survived the hard times of COVID19 infection.

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PREFACE

William Osler (1849-1919)

''To study the phenomena of disease without books is to sail an uncharted sea, while to study books without patients is not to go to sea at all''

This book contains a wealth of data and a myriad of information. It is divided into chapters; each one has two sections; questions and answers. The questions are made with different levels (easy to difficult). All are prepared to test the depth of knowledge and accuracy of practice. The information on most of the questions is made according to the standards of Nelson's textbook of Pediatrics 21st edition.

The concept of questions is that; there is one or more keys included in the given question, according to the difficulty of each one. The reader needs to recognize it and choose the answer accordingly. Sometimes more that one correct answer is possible which could be deceptive; the candidate needs to choose the most appropriate one that fits the given scenario. The section of answers usually highlights the important key and the related explained information.

It is highly recommended to keep a notebook and a marker pen to dictate and highlight the keys of difficult questions to save time to review them all again. Discussion with colleagues and peer groups is the benchmark for success in reading this work. By the end of reading this book, the author is sure that every student will be assiduous in pointing out every important point in this book.

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Chapter 1

The Field of Pediatrics Questions

ZUHAIR ALMUSAWI

1. What is the leading cause of worldwide under-five mortality rate (U5MR)?
   A. Preterm birth complications
   B. Pneumonia
   C. Perinatal asphyxia
   D. Diarrheal diseases
   E. Malaria

2. Which of the following is the **MOST** common cause of under-5 mortality in developing countries?
   A. Diarrheal disease
   B. Pneumonia
   C. Malaria
   D. AIDS
   E. Measles

3. Which of the following countries has the lowest rank of child mortality rate in the world?
   A. U.S.A.
   B. United Kingdom
   C. Cuba
   D. Czech Republic
   E. Japan

4. In 2002 an estimated 1.5 million under-5 deaths were caused by vaccine preventable diseases.
Which of the following is the top contributor?
   A. Haemophilus influenzae B (Hib)
   B. Measles
   C. Rotavirus
   D. Pertussis

E. Tetanus

5. For which of the following conditions curative treatment is possible but may not succeed?

A. Cystic fibrosis
B. Severe immunodeficiency
C. Chronic respiratory failure
D. Muscular dystrophy
E. Advanced cancer

6. For which of the following conditions, there is intensive long-term treatment, but premature death is still possible?

A. Severe cerebral palsy
B. Cystic fibrosis
C. Tay-Sachs disease
D. Batten disease
E. Severe forms of osteogenesis imperfect

7. What is the first line drug used in moderate pain management?

A. Short-acting opioid
B. Acetaminophen
C. Ibuprofen
D. Salicylates
E. Codeine

8. Which of the following can be used as adjuvant drugs for bone pain?

A. Amitriptyline
B. Carbamazepine
C. Steroids
D. Codeine
E. Ketamine

9. Which of the following can be used in a 10-old child with life-threatening illness and excessive respiratory secretions?

A. Intramuscular hyoscyamine sulfate
B. Scopolamine patch
C. Intramuscular atropine
D. Oral dexamethasone

E. Intravenous lorazepam

10. What is the **MOST** effective antidepressant for children with life-threatening illness and depression?

A. Clonidine
B. Methylphenidate
C. Benzodiazepines
D. Nortriptyline
E. Methadone

11. What is the **MOST** common cause of global injury deaths to children, adolescents, and young adults?

A. Suicide
B. Road traffic injuries
C. Drowning
D. Fire and burn
E. Falls

12. Which of the following age groups is most vulnerable to poisonings?

A. Infants
B. Toddlers
C. Preschool children
D. School children
E. Adolescents

13. Which of the following is an important precursor of serious school violence?

A. Mental health problems
B. Racial tensions
C. Alcohol use
D. poverty
E. Bullying

14. Which of the following is a common manifestation of bullying?

A. School avoidance
B. Academic failure
C. Social problems
D. Lack of friends
E. Suicidal ideation

15. A 14-year-old boy may need careful screening for bullying if he is
    A. quiet
    B. overly confident
    C. depressed
    D. disabled
    E. obese

16. Pediatricians can counsel parents to help their children avoid exposure to any form of media violence under age of
    A. 4 years
    B. 6 years
    C. 8 years
    D. 10 years
    E. 12 years

17. Which of the following is a manifestation of stress reactions in children ≤6 years exposed to war or terrorism?
    A. Terrified affect
    B. Truancy
    C. Somatization
    D. Depressive affect
    E. Irrational fear

18. Which type of child abuse is difficult to prove inspite of its extreme harm?
    A. Physical abuse
    B. Sexual abuse
    C. Psychological abuse
    D. Inadequate healthcare
    E. Inadequate education

19. What is the most common manifestation of child physical abuse?
    A. Hair pulling
    B. Bites
    C. Burns
    D. Bruises
    E. Fractures

20. Which of the following features is suggestive of inflicted bruises?

A. Bruising in a toddler
B. Bruising of shin
C. Bruising of nose
D. Patterned bruising
E. Multiple bruises of same age

21. Which of the following fractures is **MOSTLY** inflicted?
A. Clavicular
B. Femoral
C. Supracondylar humeral
D. Posterior rib fractures
E. Distal extremity

22. A single young mother brought her 8-year-old daughter complaining from genital redness and bleeding for the last 3 days. The mother gives history that her daughter has social withdrawal, fearfulness, and learning difficulties with new-onset bed-wetting.
Of the following, the **MOST** likely cause of the genital complaint is
A. urethral prolapse
B. vaginal foreign body
C. accidental trauma
D. sexual abuse
E. vaginal tumor

Chapter 1

The Field of Pediatrics
Answers

ZUHAIR ALMUSAWI

1.(A) The leading causes of worldwide U5MR are preterm birth complications, pneumonia, perinatal asphyxia, diarrheal diseases, and malaria. Many of these causes are linked to malnutrition.

2.(A) Causes of under-5 mortality differ greatly between developed and developing nations. In developing countries, 66% of deaths in children <5 yr old resulted from infectious and parasitic diseases. Among the 42 countries having 90% of childhood deaths, diarrheal disease accounted for 22% of deaths, pneumonia 21%, malaria 9%, AIDS 3%, and measles 1%.

3.(E) Although the wealth of the United States places it in the 8th position with respect to gross domestic product (GDP) per capita (2016) in the world, the U.S. child mortality rate is ranked 56th in the world, at 5.8 deaths per 1,000 live births, which is higher than the United Kingdom (4.3), Cuba (4.4), Canada (4.5), Czech Republic (2.6), and Japan (2.0).

4.(C) Top contributors were pneumococcus and rotavirus, followed by Haemophilus influenzae B (Hib), measles, pertussis, and tetanus.

5.(E) Conditions for Which Curative Treatment Is Possible but May Not Succeed

- ✓ Advanced or progressive cancer or cancer with a poor prognosis
- ✓ Complex and severe congenital or acquired heart disease

6.(B) Conditions for Which There Is Intensive Long-Term Treatment Aimed at Prolonging Life and Maintaining Quality of Life, but Premature Death Is Still Possible

- ✓ Cystic fibrosis
- ✓ Severe immunodeficiency
- ✓ High-risk solid-organ transplant candidates and/or recipients (e.g., lung,
- ✓ multivisceral)
- ✓ Chronic or severe respiratory failure
- ✓ Muscular dystrophy
- ✓ Complex multiple congenital malformation syndromes
- ✓ Primary pulmonary hypertension
- ✓ Severe chromosomal disorders (aneuploidy, deletions, duplications)

7.(A) For moderate or severe pain, start with a short-acting opioid at regular intervals. When dose requirements have stabilized, consider converting opioid to a long-acting formulation with doses available for breakthrough or uncontrolled pain, as needed.

8.(C)

- ✓ Antidepressants (e.g., amitriptyline, nortriptyline) and anticonvulsants (e.g., gabapentin, carbamazepine, topiramate) for neuropathic pain
- ✓ Steroids or NSAIDs for bone pain
- ✓ Sedatives and hypnotics for anxiety and muscle spasm

9.(B) Scopolamine patch:1.5 mg patch, change q72h (for children >8-12 yr old).

10.(B) Because of its immediate and positive effect on mood, methylphenidate may be an effective antidepressant for children at end of life, when there may not be time for a traditional antidepressant to take effect.

11.(B)

12.(B) Toddlers do not have the judgment to know that medications can be poisonous or that some houseplants are not to be eaten.

13.(E) Bullying and weapon carrying may be important precursors to more serious school violence.

14.(A) Signs of a child being involved in bullying or exposed to school violence include physical complaints such as insomnia, stomachaches, headaches, and new-onset enuresis. Psychological symptoms, such as depression, loneliness, anxiety, and suicidal ideation, may occur. Behavioral changes, such as irritability, poor concentration, school avoidance, and substance abuse, are common. School problems, such as academic failure, social problems, and lack of friends, can also occur.

15.(B) Children who are aggressive, overly confident, lacking in empathy, or having persistent conduct problems may need careful screening.

16.(C) These younger children do not have the capacity to distinguish fantasy from reality.

17.(A) Manifestations of Stress Reactions in Children and Adolescents Exposed to War, Terrorism, and Urban Violence

Children ≤6 Yr

- ✓ Excessive fear of separation
- ✓ Clinging behavior
- ✓ Uncontrollable crying or screaming
- ✓ Freezing (persistent immobility)
- ✓ Sleep disorders
- ✓ Terrified affect

- ✓ Regressive behavior
- ✓ Expressions of helplessness and passivity

Children 7-11 Yr

- ✓ Decline in school performance
- ✓ Truancy
- ✓ Sleep disorders
- ✓ Somatization
- ✓ Depressive affect
- ✓ Abnormally aggressive or violent behavior
- ✓ Irrational fears
- ✓ Regressive and childish behavior
- ✓ Expressions of fearfulness, withdrawal, and worry

18.(C) Psychological abuse includes verbal abuse and humiliation and acts that scare or terrorize a child. Although this form of abuse may be extremely harmful to children, resulting in depression, anxiety, poor self-esteem, or lack of empathy, child protective services seldom becomes involved because of the difficulty in proving such allegations.

19.(D)

20.(D) Bruises are the most common manifestation of physical abuse. Features suggestive of inflicted bruises include (1) bruising in a preambulatory infant (occurring in just 2% of infants), (2) bruising of padded and less exposed areas (buttocks, cheeks, ears, genitalia), (3) patterned bruising or burns conforming to shape of an object or ligatures around the wrists, and (4) multiple bruises, especially if clearly of different ages.

21.(D) Clavicular, femoral, supracondylar humeral, and distal extremity fractures in children older than 2 yr are most likely noninflicted unless they are multiple or accompanied by other signs of abuse.

22.(D)

Chapter 2

Growth, Development, and Behavior Questions

ZUHAIR ALMUSAWI

1. What is the fetal age by which external genitals are distinguishable?
   A. 8 weeks
   B. 10 weeks
   C. 12 weeks
   D. 14 weeks
   E. 16 weeks

2. What is the approximate average term newborn weight?
   A. 3 kg
   B. 3.2 kg
   C. 3.4 kg
   D. 3.5 kg
   E. 3.75 kg

3. At what age, the baby starts to cruise?
   A. 4 months
   B. 7 months
   C. 10 months
   D. 12 months
   E. 15 months

4. What is the approximate age of humerus head appearance?
   A. At birth
   B. 3 weeks
   C. 6 weeks
   D. 12 weeks
   E. 6 months

5. At what age, Infants can discriminate rhythmic patterns in native vs non-native language?

A. 2 months
B. 4 months
C. 6 months
D. 8 months
E. 10 months

6. What is the age of achievement of object permanence (constancy)?
A. 4 months
B. 7 months
C. 9 months
D. 12 months
E. 15 months

7. Which of the following is of benefit for colic management in a 2-month-old crying infant?
A. Simethicone
B. Anticholinergic medications
C. Fennel extract
D. Chiropractic manipulation
E. Continuous carrying

8. Ali can sit on small chair; walks up stairs with 1 hand held; makes tower of 4 cubes; and can name pictures.
What is the expected age of Ali?
A. 12 months
B. 15 months
C. 18 months
D. 24 months
E. 30 months

9. Salem runs well, opens doors; climbs on furniture; makes tower of 7 cubes, puts 3 words together; handles spoon well; and helps to undress.
What is the expected age of Salem?
A. 15 months
B. 18 months
C. 24 months
D. 30 months
E. 36months

10. Mohammed rides tricycle; stands momentarily on 1 foot; makes tower of 10 cubes; copies circle; knows age and sex; helps in dressing; and washes hands. What is the expected age of Mohammed?

A. 18months
B. 24 months
C. 30 months
D. 36 months
E. 48months

11. Ali can hop on 1 foot; throws ball overhand; uses scissors to cut out pictures; copies cross and square; draws man with 2-4 parts besides head; tells story; plays with several children; and goes to toilet alone.
What is the expected age of Ali?

A. 30 months
B. 36 months
C. 42 months
D. 48months
E. 60 months

12. Salma can skip; draws triangle from copy; names heavier of 2 weights; names 4 colors; counts 10 pennies correctly; dresses and undresses; and asks questions about meaning of words.
What is the expected age of Salma?

A. 36 months
B. 42 months
C. 48months
D. 60 months
E. 72 months

13. A normal height mother brought her 2-year-old son whose height is 90 cm asking you about the expected adult height of her son, you answer that the approximate adult height is

A. 170 cm
B. 175 cm
C. 180 cm
D. 185 cm
E. 190 cm

14. A 4-year-old child should put in a typical sentence at least
    A. 2 words
    B. 3 words
    C. 4 words
    D. 5 words
    E. 6 words

15. Although 5% of preschool children will stutter, it will resolve in 80% of those children by age of
    A. 6 years
    B. 7 years
    C. 8 years
    D. 9 years
    E. 10 years

16. The upper-to-lower body segment ratio equals approximately 1.7 at birth, and 1.0 after
    A. 5 yr
    B. 7yr
    C. 9 yr
    D. 11yr
    E. 13 yr

17. What is the **MOST** common cause of delayed teeth eruption?
    A. Hypothyroidism,
    B. Hypoparathyroidism
    C. Familial
    D. Idiopathic
    E. Rickets

18. A couple came to you asking about the expected adult height of their 2-year-old daughter, the mother is 63 inches tall and the father is 70 inches tall. Your proper answer will be, the daughter's sex-adjusted midparental height will be
    A. 56-64 inches
    B. 58-66 inches
    C. 60-68 inches
    D. 62-70 inches
    E. 64-72 inches

19. A couple came to you asking about the expected adult height of their 3-year-old son, the mother is 63 inches tall and the father is 70 inches tall.
Your proper answer will be, the son's sex-adjusted midparental height will be

A. 62-70 inches
B. 63-71 inches
C. 64-72 inches
D. 65-73 inches
E. 66-74 inches

20. A 1-year-old child presents with repetitive, stereotyped, and rhythmic movements in the form of head banging and body rocking. These behaviors typically occur with the transition to sleep at bedtime, but also at nap times and after nighttime arousals.
Of the following, the **MOST** important aspect in management is

A. ordering EEG
B. ordering MRI brain
C. referral to pediatric neurology
D. reassurance to the family
E. starting antiepileptic drug

21. Which of the following is suggested by poor linear growth in the context of good BMI?

A. Malnutrition
B. Celiac disease
C. Inflammatory bowel disease
D. Hypothyroidism
E. Renal tubular acidosis

22. In which of the following conditions with short stature, bone age is normal?

A. Familial short stature
B. Constitutional delay
C. Hypothyroidism
D. Undernutrition
E. Celiac disease

23. Which of the following provide an accurate clinical index of adiposity?

A. BMI
B. Triceps skinfold thickness

C. Hydrodensitometry
D. Bioelectrical impedance
E. Total body water measurement

24. Which of the following is a cause of early exfoliation of teeth?
A. Hypothyroidism
B. Hypoparathyroidism
C. Gum fibrosis
D. Hypophosphatasia
E. Crowding of teeth

25. At what age, children do understand that death is irreversible and that it may involve them or their families?
A. Children approximately 3 yr old
B. Preschool-age children
C. Early school-age children
D. Children approximately 9 yr
E. Pubertal age

26. Which of the following is **TRUE** regarding sleep duration and sleep patterns in the first 2 months of life?
A. Average total sleep 15-18 hr
B. Prematures sleep less than full-term infants
C. Bottle-fed babies sleep longer than breastfed babies
D. Established nocturnal diurnal pattern
E. Sleep periods are separated by 2-4 hr awake

27. There is substantial overlap between the clinical impairments associated with obstructive sleep apnea syndrome (OSAS) and the diagnostic criteria for ADHD, including
A. Poor concentration
B. Increased irritability
C. Mood instability
D. Low frustration tolerance
E. Depression

28. Which of the following can accurately predict which children with snoring have obstructive sleep apnea syndrome?

A. Clinical history
B. Physical findings
C. Overnight polysomnogram
D. X-ray of post-nasal space
E. MRI of post-nasal space

29. A 12-year-old obese boy presents with loud, frequent, and disruptive snoring; restless sleep; with nocturnal diaphoresis. He is mouth breather, with hyponasal speech, and morning headach. Oropharyngeal examination revealed enlarged tonsils.
Of the following, the first-line treatment of this boy is

A. watchful waiting
B. weight reduction
C. positional therapy
D. continuous positive airway pressure
E. adenotonsillectomy

30. Which of the following parasomnias occur during rapid eye movement sleep?

A. Confusional arousals
B. Sleep terrors
C. Sleep walking
D. Night mares
E. All the above

31. Which of the following is **MOST** successful in the management of partial arousal episodes occurring on a nightly basis?

A. Parental education
B. Reassurance
C. Avoidance of caffeine
D. Scheduled awakenings
E. Benzodiazepines

32. A 13-year-boy has uncomfortable and unpleasant sensations in the legs, in addition to the urge to move the legs on lying in bed to sleep at night or riding in a car for prolonged periods.
His parents report that their child is a restless sleeper, moves around, and even falls out of bed during the night.

Of the following, the **MOST** likely diagnosis is

A. Leg cramps
B. Neuropathy
C. Arthritis
D. Restless legs syndrome
E. Nerve compression (“leg fell asleep”)

Chapter 2

Growth, Development, and Behavior Answers

ZUHAIR ALMUSAWI

1.(C) By wk 12, the gender of the external genitals becomes clearly distinguishable.

2.(C) The average term newborn weighs approximately 3.4 kg (7.5 lb); boys are slightly heavier than girls. Average weight does vary by ethnicity and socioeconomic status. The average length and head circumference are about 50 cm (20 in) and 35 cm (14 in), respectively, in term infants.

3.(C) At 10 months, baby can pulls to standing position; “cruises” or walks holding on to furniture.

4.(B)

5.(A) Infants at 2 mo of age can discriminate rhythmic patterns in native vs non-native language.

6.(C) A major milestone is the achievement by 9 mo of object permanence (constancy), the understanding that objects continue to exist, even when not seen. At 4-7 mo of age, infants look down for a yarn ball that has been dropped but quickly give up if it is not seen. With object constancy, older infants persist in searching. They will find objects hidden under a cloth or behind the examiner's back.

7.(C) Gripe water (containing herbal supplements), and fennel extract may have benefit, but the evidence is weak.

8.(C) 18 Months

Motor: Runs stiffly; sits on small chair; walks up stairs with 1 hand held; explores drawers and wastebaskets

Adaptive: Makes tower of 4 cubes; imitates scribbling; imitates vertical stroke; dumps raisin from bottle

Language: 10 words (average); names pictures; identifies 1 or more parts of body

Social: Feeds self; seeks help when in trouble; may complain when wet or soiled; kisses parent with pucker

9.(C) 24 Months

Motor: Runs well, walks up and down stairs, 1 step at a time; opens doors; climbs on furniture; jumps
Adaptive: Makes tower of 7 cubes (6 at 21 mo); scribbles in circular pattern; imitates horizontal stroke; folds paper once imitatively
Language: Puts 3 words together (subject, verb, object)
Social: Handles spoon well; often tells about immediate experiences; helps to undress; listens to stories when shown pictures.

10.(D) 36 Months
Motor: Rides tricycle; stands momentarily on 1 foot
Adaptive: Makes tower of 10 cubes; imitates construction of “bridge” of 3 cubes; copies circle; imitates cross
Language: Knows age and sex; counts 3 objects correctly; repeats 3 numbers or a sentence of 6 syllables; most of speech intelligible to strangers
Social: Plays simple games (in “parallel” with other children); helps in dressing (unbuttons clothing and puts on shoes); washes hands.

11.(D) 48 Months
Motor: Hops on 1 foot; throws ball overhand; uses scissors to cut out pictures; climbs well
Adaptive: Copies bridge from model; imitates construction of “gate” of 5 cubes; copies cross and square; draws man with 2-4 parts besides head; identifies longer of 2 lines
Language: Counts 4 pennies accurately; tells story
Social: Plays with several children, with beginning of social interaction and role-playing; goes to toilet alone

12.(D) 60 Months
Motor: Skips
Adaptive: Draws triangle from copy; names heavier of 2 weights
Language: Names 4 colors; repeats sentence of 10 syllables; counts 10 pennies correctly
Social: Dresses and undresses; asks questions about meaning of words; engages in domestic role-playing

13.(C) By 24 mo, children are about half their ultimate adult height. Head growth slows slightly, with 85% of adult head circumference achieved by age 2 yr, leaving only an additional 5 cm (2 in) gain over the next few years.

14.(C) As a rule of thumb, between ages 2 and 5 yr, the number of words the child puts in a typical sentence should, at a minimum, equal the child's age (2 by age 2 yr, 3 by age 3 yr, and so on).

15.(C) Children with stuttering should be referred for evaluation if it is severe, persistent, or associated with anxiety, or if parental concern is elicited.

16.(B) The U/L ratio equals approximately 1.7 at birth, 1.3 at 3 year, and 1.0 after 7 year. Higher U/L ratios are characteristic of short-limb dwarfism, as occurs with Turner syndrome or bone disorders, whereas lower ratios suggest hypogonadism or Marfan syndrome.

17.(D) Delayed eruption is usually considered when no teeth have erupted by approximately 13 month of age (mean + 3 SD). Common causes include congenital or genetic disorders, endocrine disorders (e.g., hypothyroidism, hypoparathyroidism), familial conditions, and (the most common) idiopathic conditions. Individual teeth may fail to erupt because of mechanical blockage (crowding, gum fibrosis).

18.(C) Boys: [(Maternal height + 5 inches) + Paternal height]/2

Girls: [Maternal height + (Paternal height – 5 inches)]/2

Furthermore, generally 4 inches (2 SD) is applied above and below this value to provide a genetic target height range . For example, if the mother is 63 inches tall and the father 70 inches tall, the daughter's sex-adjusted midparental height is 64 inches ± 4 inches, for a target height range of 60-68 inches.

19.(D) The son of these parents would have a sex-adjusted midparental height of 69 inches, with a range of 65-73 inches.

20.(D) Sleep-related rhythmic movements, including head banging, body rocking, and head rolling, are characterized by repetitive, stereotyped, and rhythmic movements or behaviors that involve large muscle groups. These behaviors typically occur with the transition to sleep at bedtime, but also at nap times and after nighttime arousals. Children typically engage in these behaviors as a means of soothing themselves to (or back to) sleep; these are much more common in the 1st yr of life and usually disappear by preschool age.

These behaviors typically occur in normally developing children and in the majority of cases do not indicate some underlying neurologic or psychologic problem. Usually, the most important aspect in management of sleep-related rhythmic movements is reassurance to the family that this behavior is normal, common, benign, and self-limited.

21.(D) Poor linear growth in the setting of decreasing BMI suggests a nutritional or gastrointestinal issue, whereas poor linear growth in the context of good or robust BMI suggests a hormonal condition (hypothyroidism, growth hormone deficiency, cortisol excess).

22.(A) In familial short stature the bone age is normal (comparable to chronological age), whereas constitutional delay, endocrinologic short stature,

and undernutrition may be associated with delay in bone age comparable to the height age.

23.(B) Although widely accepted as the best clinical measure of underweight and overweight, BMI may not provide an accurate index of adiposity because it does not differentiate lean tissue and bone from fat.

In otherwise a healthy individual, lean body mass is largely represented by BMI at lower percentiles. BMI >80–85% largely reflects increased body fat with a nonlinear relationship between BMI and adiposity. In the setting of chronic illness, increased body fat may be present at low BMI, whereas in athletes, high BMI may reflect increased muscle mass. Measurement of the triceps, subscapular, and suprailiac skinfold thickness have been used to estimate adiposity. Other methods of measuring fat, such as hydrodensitometry, bioelectrical impedance, and total body water measurement, are used in research, but not in clinical evaluation, but whole body dual-energy x-ray absorptiometry (DXA) is beginning to emerge as a tool for measuring body fat and lean body mass.

24.(D) Causes of early exfoliation include hypophosphatasia, histiocytosis X, cyclic neutropenia, leukemia, trauma, and idiopathic factors.

25.(D) Children approximately 9 yr and older do understand that death is irreversible and that it may involve them or their families. These children tend to experience more anxiety, overt symptoms of depression, and somatic complaints than do younger children.

26.(C) Total sleep: 10-19 hr per 24 hr (average, 13-14.5 hr), may be higher in premature babies

Bottle-fed babies generally sleep for longer periods (2-5 hr bouts) than breastfed babies (1-3hr).

Sleep periods are separated by 1-2 hr awake.

No established nocturnal diurnal pattern in 1st few wk; sleep is evenly distributed throughout the day and night, averaging 8.5 hr at night and 5.75 hr during day.

27.(A) There is substantial overlap between the clinical impairments associated with OSAS and the diagnostic criteria for ADHD, including inattention, poor concentration, and distractibility.

28.(C) Because no combination of clinical history and physical findings can accurately predict which children with snoring have OSAS, the gold standard for diagnosing OSAS remains an in-lab overnight polysomnogram (PSG).

29.(E) In the majority of cases of pediatric OSAS, adenotonsillectomy is the first-line treatment in any child with significant adenotonsillar hypertrophy, even in the presence of additional risk factors such as obesity.

30.(D) Nightmares, which are much more common than partial arousal parasomnias but are often confused with them, tend to be concentrated in the last third of the night, when REM sleep is most prominent.

31.(D) Scheduled awakenings is a behavioral intervention that involves having the parent wake the child 15-30 min before the time of night that the 1st parasomnia episode occurs and is most likely to be successful in situations where partial arousal episodes occur on a nightly basis.

32.(D) Diagnostic Criteria for Restless Legs Syndrome

A. An urge to move legs, usually accompanied by or in response to uncomfortable and unpleasant sensations in the legs, characterized by the following:

1. The urge to move the legs begins or worsens during periods of rest or inactivity.
2. The urge to move the legs is partially or totally relieved by movement.
3. The urge to move the legs is worse in the evening or at night than during the day, or occurs only in the evening or at night.

B. The symptoms in Criterion A occur at least three times per week and have persisted for at least 3 months.

C. The symptoms in Criterion A are accompanied by significant distress or impairment in social, occupational, educational, academic, behavioral, or other important areas of functioning.

D. The symptoms in criterion A are not attributable to another mental disorder or medical condition (e.g., arthritis, leg edema, peripheral ischemia, leg cramps) and are not better explained by a behavioral condition (e.g., positional discomfort, habitual foot tapping).

E. The symptoms are not attributable to the physiological effects of a drug or abuse or medication (e.g., akathisia).

Chapter 3

Behavioral and Psychiatric Disorders Questions

AHMED TAWFIQ

1. You are evaluating a 10-month-old girl with frequent regurgitation, rechewing of regurgitated material, arching of back, inattentiveness, developmental delay, and failure to gain weight. Taking deep social history reveals baby separation from biological mother because of divorce 3 months ago, and the new care giver is the step mother. Baby was completely well in the first 7 months of life.
Of the following, the **MOST** likely diagnosis is

A. GERD
B. rumination disorder
C. Sandifer syndrome
D. pyloric stenosis
E. diencephalic tumor

2. A 4-year-old boy presents with history eating of clay, paper, and soap. He is quite intelligent boy with normal anthropometric parameters, he is eating balanced diet. During interview you noticed that the mother is not paying attention to the child bad behaviors.
Of the following the **MOST** likely cause is

A. iron deficiency
B. autism
C. child neglect
D. family disorganization
E. zinc deficiency

3. Which of the following is representing a tic disorder in childhood?

A. thumb sucking
B. nail biting
C. trichotillomania
D. bruxism (teeth grinding)
E. head jerking

4. What are the **MOST** common psychiatric disorders of childhood?

A. Anxiety disorders
B. Tic disorders
C. Major depressive disorders
D. Eating disorders
E. Psychotic disorders

5. A completely well and lacking any history of drug intoxication or trauma, a 4-year-old girl presents with frightening episodes for the last 2 days. She is complaining that a snake is crawling over her and she is trying to remove it. Each episode lasts about 30 minutes.
Of the following the **MOST** likely cause is

A. posttraumatic stress disorder
B. acute phobic hallucinations
C. attention-deficit hyperactivity disorder
D. Tourette syndrome
E. psychosis

6. An 8-year-old boy referred to psychologist for evaluation because of frequent running away from school.
The **BEST** way for avoidance of such behavior in future is to

A. transfer the child to new environment and new school
B. teach him in special school
C. find a way to make father or mother present during school time
D. address the pressure factors and make a plan with parents and school staff
E. refer to pediatric psychiatrist

7. A 12-year-old adolescent girl with established diagnosis of anorexia nervosa has started successful inpatient management by interdisciplinary team (dietician, mental health and physician), she showed increased weight of about 1.5 Kg/week. She starts to develop episodes of tachycardia, hypertension, confusion, and seizures.
Of the following, the **MOST** appropriate investigation that helps in management is

A. thyroid function test
B. cranial CT
C. serum phosphorus

D. renal function test
E. random blood sugar

8. A 15-year-old girl referred from primary health care center for progressive weight loss. During interview, you find her highly obsessed about fatness especially in stomach and thigh areas; she has lanugo hair on her face and upper body, dry skin, some bruising, gooseflesh orange-yellow skin hands. BMI 18 kg/m^2, HR 60 bpm, B.P 90/60, and the investigational work up were normal.
Of the following, the **MOST** appropriate plan of management is

A. hospital admission with interdisciplinary team management
B. referral to mental health service
C. reassurance with follow up visits
D. primary health care management with interdisciplinary team
E. scheduled focused discussion visits to improve body image

9. What is the **MOST** common cause for complete suicide?

A. Family conflicts
B. Substance abuse
C. Hormonal changes
D. Social and media violence
E. Preexisting psychiatric illness

10. What is the antidepressant of choice in the treatment of depression in preadolescent period?

A. Fluoxetine
B. Escitalopram
C. Sertraline
D. Paroxetine
E. Citalopram

11. Separation anxiety disorder SAD can vary in presentation according to the affected age; those between the age of 13-16 years often have

A. excessive fear that harm will come to a parent
B. physical complaints
C. nightmares
D. reluctance to sleep alone
E. persistent avoidance of being alone

12. An 8-year-old girl referred to cardiology clinic because of recurrent attacks of chest pain which occurs almost always at night. During the interview you find a well-dressed and quite intelligent girl who describes the episode as palpitations, sweating, shaking, shortness of breath, dizziness, chest pain, and nausea. She has concern about recurrence of these symptoms and she is afraid from impending death.
Of the following, the **MOST** likely diagnosis is

A. panic disorder
B. obsessive compulsive disorder
C. ischemic heart disease
D. separation anxiety disorder
E. hyperthyroidism

13. A 3-year-old girl with epilepsy on valproate 30mg/kg/day. She is under treatment for severe empyema with linezolid. She has responded very well initially but on fifth day of antibiotic treatment, she develops severe attack of hyperthermia, agitation, tachycardia, diaphoresis and tremor.
Of the following, the **MOST** likely diagnosis is

A. malignant hyperthermia
B. central hyperthermia
C. neuroleptic malignant syndrome
D. serotonin syndrome
E. evolving bacterial resistance

14. What is the **MOST** common dose-dependent side effect of stimulant medications used in ADHD?

A. Irritability
B. Appetite suppression
C. Aggression
D. Social withdrawal
E. Visual hallucinations

15. Which of the following antidepressants is a selective serotonin reuptake inhibitor SSRI?

A. Fluoxetine
B. Mirtazapine
C. Venlafaxine
D. Duloxetine

E. Bupropion

16. Which of the following is classified as first generation antipsychotic medication?

A. Risperidone
B. Olanzapine
C. Haloperidol
D. Lithium
E. Lurasidone

Chapter 3

Behavioral and Psychiatric Disorders Answers

AHMED TAWFIQ

1.(B) Risk factors for rumination disorder in infants and young children include a disturbed relationship with primary caregivers, lack of an appropriately stimulating environment, neglect, stressful life situations, learned behavior reinforced by pleasurable sensations, distraction from negative emotions, and inadvertent reinforcement (attention) from primary caregivers. Treatment is focused on resolving the social issue and behavioral treatment is to reinforce correct eating behavior while minimizing attention to rumination. A and C can have similar presentation but in absence of rechewing and developmental delay. D can present earlier with completely different scenario. In E usually the child mood is very cheerful.

2.(D) Pica involves the persistent eating of nonnutritive, nonfood substances (e.g., paper, soap, plaster, charcoal, clay, wool, ashes, paint, earth) over a period of at least 1 mo. The eating behavior should be inappropriate to the developmental level therefore minimum age of 2 yr is suggested. Numerous etiologies have been proposed but not proved, ranging from psychosocial causes to physical ones. They include nutritional deficiencies (e.g., iron, zinc, and calcium), low socioeconomic factors (e.g., lead paint exposure), child abuse and neglect, family disorganization (e.g., poor supervision), mental disorder, learned behavior, underlying (but undetermined) biochemical disorder, and cultural and familial factors. Regarding distracters; nutritional deficiencies are less likely because of balance diet, child neglect is also less likely because of normal growth and development. Pica and autism can be a co-morbidities but not in such scenario.

3.(E) Habits involve an action or pattern of behavior that is repeated often. Habits are common in childhood and range from usually benign and transient behavior (e.g., thumb sucking, nail biting) to more problematic (e.g., trichotillomania, bruxism). The last distracter is representing a simple motor tics, other examples: eye blinking, shoulder shrugging, extension of the extremities, all are fast, brief movements involving one or a few muscle groups.

4.(A) Anxiety disorders are the most common psychiatric disorders of childhood, occurring in 5–18% of all children and adolescents, prevalence rates comparable to physical disorders such as asthma and diabetes.

5.(B) Acute phobic hallucinations are benign and common and occur in previously healthy preschool children. The hallucinations are often visual or tactile, last 10-60 min, and occur at any time but most often at night. The child is quite frightened and might complain that bugs or snakes are crawling over him or her and attempt to remove them. The cause is unknown. The differential diagnosis includes drug overdose or poisoning, high fever, encephalitis, and psychosis. The child's fear is not alleviated by reassurance by the parents or physician, and the child is not amenable to reason. Physical and mental status examination is otherwise normal. Symptoms can persist for 1-3 days, slowly abating over 1-2 wk.

6.(D) Truancy is more common in older children and can be a function of multiple factors, including but not limited to learning difficulties, social anxiety, depression, traumatic exposure, bullying, peer pressure, and substance use. In any of these cases, the child should be referred for further evaluation to assess the barriers to returning to school. Best practices for dealing with truancy resulting from school avoidance and anxiety include addressing the underlying psychological symptoms causing the school avoidance and empowering parents, children, and school staff to work on a consistent plan for a return to school.

7.(C) For patients with anorexia nervosa and low weight, the nutrition prescription should work toward gradually increasing weight at the rate of about 0.5-1 lb/wk, by increasing energy intake by 100-200 kcal increments every few days, toward a target of approximately 90% of average body weight for sex, height, and age. In addition, with extremely low weight, refeeding syndrome (a result of the rapid drop in serum phosphorus, magnesium, and potassium with excessive reintroduction of calories, especially carbohydrates), is associated with acute tachycardia and heart failure and neurologic symptoms.

8.(D) Classification of anorexia nervosa AN, Mild: BMI ≥ 17 kg/m2, Moderate: BMI 16-16.99 kg/m2, Severe: BMI 15-15.99 kg/m2, Extreme: BMI < 15 kg/m2. Indication for admission AN, Heart rate <50 beats/min, other cardiac rhythm disturbances, blood pressure <80/50 mm Hg, postural hypotension resulting in >10 mm Hg decrease or >25 beats/min increase, hypokalemia, hypoglycemia hypophosphatemia, dehydration, body temperature <36.1°C, <80% healthy body weight, hepatic, cardiac, or renal compromise, suicidal intent and plan, very poor motivation to recover (in family and patient), preoccupation with ego-syntonic thoughts, coexisting psychiatric disorders, requires supervision after

meals and while using the restroom, failed day treatment. The treatment of a child or adolescent diagnosed with an ED is ideally provided by an interdisciplinary team (physician, nurse, dietitian, mental health provider) with expertise treating pediatric patients.

9.(E) Approximately 90% of youths who completed suicide have a preexisting psychiatric illness, most often major depression. Among females, chronic anxiety, especially panic disorder, also is associated with suicide attempts and completion. Among males, conduct disorder and substance use convey increased risk.

10.(A) Two selective serotonin reuptake inhibitors (SSRIs), fluoxetine and escitalopram, are the only antidepressants approved by the U.S. Food and Drug Administration (FDA) for the treatment of depression in youth, and fluoxetine alone is approved for preadolescents.

11.(B) SAD is more common in prepubertal children, with an average age of onset of 7.5 yr. Girls are more frequently affected than boys. SAD is characterized by unrealistic and persistent worries about separation from the home or a major attachment figure. Concerns include possible harm befalling the affected child or the child's primary caregivers, reluctance to go to school or to sleep without being near the parents, persistent avoidance of being alone, and nightmares involving themes of separation, numerous somatic symptoms, and complaints of subjective distress. Symptoms vary depending on the child's age: Children <8 yr often have associated school refusal and excessive fear that harm will come to a parent; children 9-12 yr have excessive distress when separated from a parent; and those 13-16 yr often have school refusal and physical complaints.

12.(A) Panic disorder is a syndrome of recurrent, discrete episodes of marked fear or discomfort in which patients experience abrupt onset of physical and psychological symptoms called panic attacks. Physical symptoms can include palpitations, sweating, shaking, and shortness of breath, dizziness, chest pain, and nausea. Children can present with acute respiratory distress but without fever, wheezing, or stridor, ruling out organic causes of the distress. The associated psychological symptoms include fear of death, impending doom, loss of control, persistent concerns about having future attacks, and avoidance of settings where attacks have occurred.

13.(D) Patients with severe serotonin syndrome require to control agitation, autonomic instability, and hyperthermia as well as administration of 5-hydroxytryptamine (5-HT2A , serotonin) antagonists (e.g., cyproheptadine). Serotonin syndrome is generally self-limited and can resolve spontaneously

after the serotonergic agents are discontinued in mild cases. It is characterized by the triad of mental status changes (e.g., agitation, delirium, hallucinations, and coma), autonomic instability (e.g., tachycardia, labile BP, dizziness, diaphoresis, flushing, and hyperthermia), and neuromuscular symptoms (e.g., tremor, rigidity, myoclonus, hyperreflexia, and incoordination). Serotonin syndrome results from excessive agonism of the CNS and peripheral nervous system serotonergic receptors and can be caused by a range of drugs, including SSRIs, valproate, and lithium. Interactions that can cause serotonin syndrome include SSRIs with linezolid (antibiotic with monoamine oxidase inhibitor properties) and with antimigraine preparations, as well as with amphetamine preparations, trazodone, buspirone, and venlafaxine.

14.(B) The most common (generally dose-dependent) side effects of stimulants include headache, stomachache, appetite suppression, weight loss, blood pressure (BP) and heart rate increases, and delayed sleep onset. Less common side effects include irritability (particularly prominent in younger children), aggression, social withdrawal, and hallucinations (visual or tactile). Amphetamine preparations prescribed concurrently with serotonergic antidepressants can be associated with the development of serotonin syndrome.

15.(A) Other distracters are atypical antidepressants (non SSRI). The non-SSRI antidepressants include bupropion, duloxetine, venlafaxine, and mirtazapine. These medications all lack rigorous evidence to support their effectiveness in children and adolescents, and as such should not be considered first-line options.

16.(C) Based on their mechanism of action, antipsychotic medications can be divided into first-generation (typical antipsychotics) act by blocking dopamine D2 receptors; may be used in psychosis, Tourette disorder, severe behavioral disorders and agitation. Second-generation antipsychotics (atypical antipsychotics, A, B and E) which act as dopaminergic and serotonergic antagonists can be used in addition to above in mania, schizophrenia and bipolar disorders. Distracter D is mood stabilizer and used mainly in mania and bipolar disorders.

Chapter 4

Learning and Developmental Disorders Questions

USAMA A. AL JUMAILY

1. Which of the following manifestations is seen in pre-school children with neurodevelopmental and executive dysfunctions?
   A. Difficulty with learning colors and shapes
   B. Problems of syntactic and semantic skills
   C. Struggling with root words, prefixes, and suffixes
   D. Deficiency in reading comprehension
   E. Difficulty in answering open-ended questions

2. A 7-year-old boy acts before thinking, Interrupts olders during conversations, and has poor behavioral and emotional control. The intervention that is helpful for this child is to
   A. repeat instructions as needed
   B. break tasks into smaller, manageable steps
   C. ask child to verbalize plan before beginning work
   D. teach response-delay techniques
   E. place child with partner for modeling and cuing

3. Which of the following is classified as a problem related to poor attention in a child with attention deficit hyperactivity disorder (ADHD)?
   A. Runs about excessively in situations in which it is inappropriate
   B. Overlooks or misses details, with inaccurate work
   C. Difficulty playing or engaging in leisure activities quietly
   D. Leaves seat in classroom or in other situations in which remaining seated is expected
   E. Fidgets with hands or feet or squirms in seat

4. Which of the following is classified as a problem related to hyperactivity/impulsivity in a child with attention deficit hyperactivity disorder (ADHD)?
   A. Acts as if “driven by a motor"

B. Does not seem to listen when spoken to directly
C. Fails to finish schoolwork, chores, or duties in the workplace
D. Dislikes engaging in tasks that require sustained mental effort
E. Loses things necessary for tasks or activities (e.g., pencils, books, tools)

5. An 8-year-old boy has poor academic performance. His mother describes that he does not seem to listen when spoken to him directly; he has often fails to finish homework and assigned chores; he always loses pencils, books, and school tools. His teacher describes that he often leaves seat in his classroom, and has difficulty playing or engaging in leisure activities quietly.
Of the following, the MRI finding that is **MOST** likely seen in this boy is

A. large volume prefrontal cortex
B. loss of the normal hemispheric asymmetry
C. small volume occipital cortex
D. large volume basal ganglia
E. loss of cerebellar vermis

6. Which of the following clinical manifestations of ADHD is **MOST** commonly seen in pre-school children?

A. Disorganization
B. Distractibility
C. Inattention
D. Fidgeting
E. Disruptive behavior

7. Which of the following is the **MOST** appropriate initial drug for the treatment of children with ADHD?

A. Guanfacine
B. Clonidine
C. Atomoxetine
D. Ritalin
E. Lithium

8. Which of the following developmental impacts is usually encountered in school aged children with ADHD?

A. Unintentional injuries
B. Academic impairment
C. Low self-esteem

D. Alcohol and substance misuse
E. Occupational difficulties

9. Which of the following is the **MOST** common cause of dyslexia in children?
A. Low birth weight
B. Social stress
C. Familial
D. Birth injury
E. Substance abuse

10. A 7-year-old girl is brought by her parents complaining of failure to speak in school and during social meetings. She speaks normally with her parents in their home. She is described by the parents that she has excessive shyness, withdrawal, and parental dependency. The mother has a history of social anxiety during childhood.
Of the following, the **MOST** likely diagnosis is
A. autism
B. anxiety disorder
C. specific language impairment
D. selective mutism
E. Isolated expressive language disorder

11. A 3-year-old boy is brought to you by his parents because of poor social communications. He communicates by reaching partner`s hand and placing it on the desired objects. Protests are demonstrated through pushing hands. He plays functionally with toys when seated and used eye gaze appropriately during cause-and-effect play, but otherwise eye gaze is absent. He appears to be non-engaged and responds inconsistently to his name. He has repetitive finger movements, body rocking and lunging, and echoing words immediately after parents are said. He has a pattern of repeated turning light switches on and off, and opening and closing doors.
Of the following, the **MOST** likely diagnosis is
A. social communication disorders
B. language disorders
C. autism spectrum disorders
D. hearing loss
E. social anxiety
12. At which of the following ages onset of stuttering is typically occurring?

A. Below 2 yr
B. 2-4 yr
C. 5-6 yr
D. 7-8 yr
E. Above 12 yr

13. Which of the following syndromes is associated with mild intellectual disability (IQ 50-70)?
A. Down
B. Wolf-Hirschhorn
C. Fragile X
D. Prader-Willi
E. Noonan

14. Which of the following congenital infections is associated with progressive encephalopathy and subsequent regression of intellectual disabilities that is started after the age of 2 year?
A. Cytomegalovirus
B. Toxoplasmosis
C. Rubella
D. Syphilis
E. Herpes

15. Which of the following features encountered in infancy is a strong predictor for risk of autism spectrum disorders (ASD)?
A. Reduced response to name
B. Trouble picking up on the nuances of social interactions
C. Difficulty engaging in group interactions
D. Repetitive behaviors of body rocking and lunging
E. Repetitive speech such as echoing words

16. Which of the following factors is a strong contributor to autism spectrum disorders (ASD)?
A. Younger maternal age
B. Maternal underweight
C. Long interval from prior pregnancy
D. Post term baby
E. Prenatal cytomegalovirus infection

Chapter 4

Learning and Developmental Disorders Answers

USAMA A. AL JUMAILY

1.(A) Preschool-age children might present with delayed language development, including problems with articulation, vocabulary development, word finding, and rhyming. They often experience early challenges with learning colors, shapes, letters, and numbers; the alphabet; and days of the week.

2.(D) This child mostly has disinhibition/impulsivity, so teach response-delay techniques (e.g., counting to 10 before acting) is helpful. Choice "A" can be applied when there is working memory dysfunction. Choice "B" is helpful when there is a problem of initiation. Choice "C" and "E" are helpful in planning dysfunction.

3.(B) A child with ADHD often fails to give close attention to details or makes careless mistakes in schoolwork, at work, or during other activities (e.g., overlooks or misses details, work is inaccurate). All other distracters are related to problems of hyperactivity/impulsivity.

4.(A) All other distracters are related to problems of poor attention.

5.(B) Brain MRI studies in children with ADHD indicate a reduction or even loss of the normal hemispheric asymmetry in the brain, as well as smaller brain volumes of specific structures, such as the prefrontal cortex and basal ganglia. Children with ADHD have approximately a 5–10% reduction in the volume of these brain structures. Functional MRI data suggest deficits in dispersed functional networks for selective and sustained attention in ADHD that include the striatum, prefrontal regions, parietal lobe, and temporal lobe.

6.(E) The symptoms may vary from motor restlessness, aggressive, and disruptive behavior which are common in preschool children, to disorganized, distractible, and inattentive symptoms, which are more typical in older adolescents and adults. ADHD is often difficult to diagnose in preschoolers because distractibility and inattention are often considered developmental norms during this period.

7.(D) The most widely used medications for the treatment of ADHD are the presynaptic dopaminergic agonists, commonly called psychostimulant medications, including methylphenidate, dexmethylphenidate, amphetamine,

and various amphetamine and dextroamphetamine preparations. If a methylphenidate compound is unsuccessful, the clinician should switch to an amphetamine product. If satisfactory treatment results are not obtained with the 2nd stimulant, clinicians may choose to prescribe atomoxetine, a noradrenergic reuptake inhibitor that has been approved by the U.S. Food and Drug Administration (FDA) for the treatment of ADHD in children, adolescents, and adults. Long-acting guanfacine and clonidine are also FDA approved for the treatment of ADHD. These medications can also treat motor and vocal tics and so may be a reasonable choice in a child with a comorbid tic disorder.

8.(B) Unintentional injuries are seen in preschool children with ADHD. Low self-esteem and alcohol/drug abuse are encountered in adolescents with ADHD. During adulthood, occupational difficulties are encountered.

9.(C) Dyslexia is familial, occurring in 50% of children who have a parent with dyslexia, in 50% of the siblings of dyslexic persons, and in 50% of the parents of dyslexic persons.

10.(D) Specific language impairment (SLI) is characterized by a significant discrepancy between the child's overall cognitive level (typically nonverbal measures of intelligence) and functional language level. These children also follow an atypical pattern of language acquisition and use. Closer examination of the child's skills might reveal deficits in understanding and use of word meaning (semantics) and grammar (syntax). Often, children are delayed in starting to talk. Children with isolated expressive language disorder (“late talker syndrome”) have age- appropriate receptive language and social ability. Once they start talking, their speech is clear. There is no increased risk for language or learning disability as they progress through school. A family history of other males with a similar developmental pattern is often reported.

11.(C) Children with language disorders do not have associated restricted and repetitive behavior or atypical use of language. The diagnosis of social communication disorder is also distinguished from ASD by the lack of restrictive and repetitive behaviors. Children with hearing loss may present with some “red flags” for ASD, such as poor response to name. However, they typically develop nonverbal communication and play skills as expected and do not have stereotyped or restricted behavior patterns. Children with social anxiety may present with some symptoms suggestive of ASD. Shy children may have reduced eye contact and social initiation. However, they typically have preserved social interest and insight and will not exhibit high levels of stereotyped behaviors.

12.(B) Seldom does a child begin stuttering before 2 yr of age or after 12 yr; in fact, the mean age of onset is 2-4 yr, and most children stop stuttering within 4 yr of onset.

13.(E) The most common biologic causes of mild ID include genetic or chromosomal syndromes (e.g., velocardiofacial, Williams, and Noonan syndromes)**.** In children with severe ID, a biologic cause (usually prenatal) can be identified in about three fourths of all cases. Causes include chromosomal (e.g., Down, Wolf-Hirschhorn, and deletion 1p36 syndromes) and other genetic and epigenetic disorders (e.g., fragile X, Rett, Angelman, and Prader-Willi syndromes).

14.(D)

15.(A) Symptoms can present early in infancy, with reduced response to name and unusual use of objects being strong predictors for risk of ASD. All other features are seen in toddlers and older children.

16.(E) Older maternal or paternal age may increase the risk of ASD. In addition, factors influencing the intrauterine environment, such as maternal obesity or overweight, short interval from prior pregnancy, premature birth, and certain prenatal infections (e.g., rubella, cytomegalovirus) are associated with ASD.

Chapter 5

Nutrition Questions

AQEEL MAHDI

1. Which of the following age groups have the highest energy and nutrient requirements relative to body size?
   A. Infancy
   B. Early childhood
   C. School age
   D. Early adolescent
   E. Late adolescent

2. Which of the following statements about successful breast feeding is recommended by American Academy of Pediatrics (AAP) rather than by WHO?
   A. Help woman initiate breastfeeding within1hour of birth
   B. Give newborns no food or drink other than breast milk unless medically indicated
   C. Encourage breastfeeding on demand
   D. Uses of pacifiers to breastfeeding infants
   E. Age of starting complementary feeding

3. What is the indication of IV fluid in acute severe malnutrition?
   A. Severe anorexia
   B. Severe dehydration
   C. Hypoglycemia
   D. Hyponatremia
   E. Shock

4. Which of the following foods is very rich in vitamin A?
   A. Lean meat
   B. Organ meat
   C. Milk
   D. Cheese
   E. Fruits

5. What is the **MOST** common micronutrient deficiency in the world?

A. Iron
B. Magnesium
C. Selenium
D. Zinc
E. Copper

6. Which food stuff is considered as thiamine antagonist that may contribute to thiamine deficiency?

A. Milk
B. Red meat
C. Oat
D. Coffee
E. Fish

7. What is the usual cause of death in patients with thiamine deficiency?

A. Cardiac
B. Renal
C. CNS
D. Infection
E. Hepatic

8. A 6-year-old girl lives in refugee camp brought to you with tingling and paresthesia of feet; she has decreased deep tendon reflexes, loss of vibration sense, and hoarseness of voice, with ptosis of eye lids. Chest X-ray shows cardiomegaly. They were eating a polished rice-based monotonous diet for last 3 months.

Of the following, the **MOST** likely cause of these findings is

A. B12 deficiency
B. thiamine deficiency
C. Horner syndrome
D. degenerative brain disease
E. Guillain-Barre syndrome

9. Among B complex vitamins deficiency, the one that specifically causes cheilosis and glossitis is

A. thiamine
B. niacin

C. riboflavin
D. pyridoxine
E. cobalamin

10. Which of the following vitamin B complex is destroyed by phototherapy?
A. B1
B. B2
C. B3
D. B6
E. B12

11. Malnutrition is defined as acute when its duration is below
A. 1 month
B. 2 months
C. 3 months
D. 4 months
E. 5 months

12. A 10-year-old boy complains of headache for last 4 weeks associated with vomiting, anorexia, and dry itchy desquamating seborrheic skin lesion with no history of fever or trauma. The family mentions that he was well previously and only he took some tonics capsules for several weeks prior to this event.
Of the following, the **MOST** likely diagnosis is
A. thiamine toxicity
B. niacin toxicity
C. hypervitaminosis A
D. histiocytosis
E. riboflavin toxicity

13. A 14-year-old girl diagnosed with anorexia nervosa presents with glossitis and erythematosus skin rash involving both hands and feet sharply demarcated from the surrounding healthy skin giving the appearance of gloves and stockings. The girl has also diarrhea, anorexia, and depression with insomnia and delirium.
Of the following, the **MOST** likely diagnosis is
A. pellagra
B. biotin-responsive basal ganglia disease
C. beriberi

D. riboflavinosis
E. contact dermatitis

14. Which of the following clinical features of pyridoxine(B6) deficiency IS **NOT** a usual finding in infants?

A. Irritability
B. Seizure
C. Peripheral neuritis
D. Microcytic anemia
E. Vomiting

15. Which vitamin deficiency may be attributed to oxalic acid bladder stones formation?

A. Niacin
B. Thiamine
C. Riboflavin
D. Pyridoxine
E. Cobalamin

16. Consumptions of large amount of raw egg whites over long periods results in deficiency of which of the following vitamins?

A. C
B. D
C. E
D. B7
E. B2

17. Which of the following vitamins is almost exclusively come from animal food?

A. Cobalamin
B. Vitamin D
C. Pyridoxine
D. Folic acid
E. Thiamine

18. A 3-year-old boy diagnosed with autism presents with difficulty in walking and bone pain. His diet is devoid of most fruits and vegetables as part of his management for the last 3 months.

Which of the following vitamins deficiency should be evaluated for?

A. A
B. B
C. C
D. D
E. K

19. In a patient with biliary atresia; the expected age of onset of neurological symptoms due to vitamin E deficiency is usually after

A. 2 months
B. 4 months
C. 6 months
D. 8 months
E. 12 months

20. The typical age of onset of hemolysis due to vitamin E deficiency in premature babies is during

A. 1st month of life
B. 2nd month of life
C. 3rd month of life
D. 4^{th} month of life
E. 5^{th} month of life

21. What is an initial neurological finding usually seen in child with symptomatic vitamin E deficiency?

A. Loss of deep tendon reflexes
B. Limb ataxia
C. Unsteady gait
D. Dysarthria
E. Ophthalmoplegia

22. Which of the following vitamins has limited body store and the symptoms of deficiency can develop within weeks of limited supply?

A. E
B. D
C. A
D. K
E. B

23. What is the **MOST** common site of bleeding in late onset vitamin K deficiency bleeding (VKDB)?

A. CNS
B. GIT
C. Skin
D. Muscles
E. Genito-urinary

24. What is the **FIRST** clotting factor affected by vitamin K deficiency?

A. VII
B. VIII
C. IX
D. X
E. XIII

25. What is the **MOST** common cause of nipple pain in early breastfeeding mothers?

A. Nipple candidiasis
B. Breast engorgement
C. Lack of breast massage during pregnancy
D. Hormonal changes
E. Improper latch on

26. What is the **MOST** common reason for using infant formula?

A. Parental preference
B. Medical condition affecting the baby
C. Medical condition affecting the mother
D. Inadequate weight gain
E. Commercial factors

27. Micronutrients include vitamins and trace elements. By definition, a trace element is

A. <0.01% of the body weight
B. <0.02% of the body weight
C. <0.03% of the body weight
D. <0.04% of the body weight
E. <0.05% of the body weight

28. Which of the following micronutrients deficiency is associated with cardiomyopathy?

A. Chromium
B. Copper
C. Molybdenum
D. Selenium
E. Iron

29. Which of the following micronutrients deficiency is associated with microcytic anemia?

A. Chromium
B. Copper
C. Molybdenum
D. Selenium
E. Iodine

30. At which age bottle weaning should begin?

A. 12-15 mo.
B. 16- 18 mo.
C. 19- 21 mo.
D. 22-24 mo.
E. 25-27 mo.

31. Which of following micronutrients should not be given in the stabilization phase of acute severe malnutrition?

A. Zinc
B. Vitamin A
C. Iron
D. Copper
E. Folic acid

32. A 17-year-old girl suffering from non-alcoholic fatty liver disease because of obesity and BMI of 42, she had failed a 6 months multidisciplinary weight management program. The girl is highly concerned about her weight and asks you about **NEXT** step in management; your advice is

A. behaviour changes with caloric reduction
B. behaviour changes and pharmacotherapy
C. hormone replacement therapy

D. hormone replacement therapy plus behaviour changes with caloric reduction
E. bariatric surgery

33. Which of the following genetic causes of rickets is mainly found in Middle East?
A. Vitamin D–dependent rickets type 1A
B. Autosomal dominant hypophosphatemic rickets
C. Autosomal recessive hypophosphatemic rickets
D. X-linked hypophosphatemic rickets
E. Hereditary hypophosphatemic rickets with hypercalciuria

34. A breast feeding mother consult you regarding feeding of her 3-month-old baby. She has recently diagnosed with primary Herpes simplex infection.
Which of the following is the **BEST** advice for her?
A. She can breastfeed her baby with no contraindications
B. She can start breastfeeding only after receiving treatment
C. Breast feeding is contraindicated
D. Breastfeeding is contraindicated with active herpetic lesions of the breast
E. Breastfeeding is contraindicated only with genital herpes infection

35. Which of the following infections is absolute contraindication in developed countries to breastfeeding?
A. Hepatitis C infection
B. CMV infection
C. Hepatitis B infection
D. Hepatitis A
E. HIV infection

36. A nursing school teacher who works 8 hours per day asks you if she can use milk expression technique and keep her milk in refrigerator when she is in duty. This expressed breast milk can be used within
A. 12 hours
B. 24 hours
C. 36 hours
D. 48 hours

E. 60 hours

37. Which of the following is the recommended duration of using the milk formula after removing the aluminium foil or plastic wrap?

A. 1 wk
B. 2 wk
C. 3 wk
D. 4 wk
E. 5 wk

38. Which of the following is an indication for soy based formula in infancy?

A. Infantile colic
B. Cow's milk protein–induced enteropathy
C. Celiac disease
D. Acute gastroenteritis
E. Galactosemia

39. You are invited to a nutritional medical conference where the lecturer starts to speak about children who are vegetarian but occasionally eat meat.
What is the **BEST** term used to describe these children?

A. Veganism
B. Ovovegetarianism
C. Lactovegetarianism
D. Flexitarian
E. Pescatarian

40. Which of the following plants have a high concentration of iron?

A. Leafy green vegetables
B. Whole grains
C. Lentils
D. Dark-green vegetables
E. bright-orange vegetables

41. Which of the following is the cutoff value to define anaemia in a 9-month-old infant?

A. 95 g/L

B. 10 g/L
C. 105 g/L
D. 110 g/L
E. 115 g/L

42. You are called by a nurse to calculate the fluid to be given to a severely dehydrated malnourished 2-year-old boy with weight of 5 kg.
Which of the following is the **BEST** treatment option?

A. ReSoMal 25 mL every 30 min for 1st 2 hr
B. ORS 25 mL every 30 min for 1st 2 hr
C. ReSoMal 50mL every 30 min for 1st 2 hr
D. ORS 50 mL every 30 min for 1st 2 hr
E. IV normal saline 100 mL over 30 min.

43. Which of the following **BEST** describes the composition of ReSoMal?

A. Water 1 L, WHO ORS One sachet, Sucrose 50 g, electrolyte/mineral solution 40 mL
B. Water 2 L, WHO ORS One sachet, Sucrose 50 g, electrolyte/mineral solution 40 mL
C. Water 2 L, WHO ORS One sachet, Sucrose 25g, electrolyte/mineral solution 40 mL
D. Water 2 L, WHO ORS One sachet, Sucrose 50 g, electrolyte/mineral solution 20 mL
E. Water 2 L, WHO ORS two sachet, Sucrose 50 g, electrolyte/mineral solution 40 mL

44. You are asked to prescribe antibiotics for a 1-year-old malnourished boy in the ward. His mother tells you that he has diarrhoea for the last 3 weeks, and on examination he is hypothermic.
Which of the following is the **BEST** antibiotics regime?

A. No regime is indicated
B. Amoxicillin oral for 5 days
C. Ampicillin IV or IM for 2 days, then amoxicillin oral for 5 days
D. Gentamicin IV or IM once daily for 7 days and Ampicillin IV or IM for 2 days oral for 5 days
E. Gentamicin IV or IM once daily for 7 days and Ampicillin IV or IM for 2 days, then amoxicillin oral for 5 days plus metronidazole oral for 7 days

45. Which of the following is considered the **MOST** common genetic cause of obesity?

A. Down syndrome
B. Leptin or leptin receptor gene deficiency
C. Prader-Willi syndrome
D. Melanocortin 4 receptor gene mutation
E. Bardet-Biedl syndrome

46. Which of the following vitamins used in preterm infants to improve respiratory function and prevent development of chronic lung disease?

A. A
B. D
C. E
D. C
E. K

47. Which of the following drugs is the only Food and Drug Administration (FDA)–approved medication for obesity in children<16?

A. Orlistat
B. Phentermine
C. Liraglutide
D. Lorcaserin
E. Phentermine/ topiramate

48. Which of the following is the recommended screen time by the American Academy of Pediatrics?

A. No more than 2 hr/day for children at any age.
B. No more than 2 hr/day for children >2 yr old and 1hr/day for children <2 yr old
C. No more than 2 hr/day for children >2 yr old and that children <2 yr old not watch television
D. No more than 3 hr/day for children >2 yr old and that children <2 yr old not watch television
E. No more than 4 hr/day for children >2 yr old and that children <2 yr old not watch television

49. Which of the following is considered as an earlier clinical feature of vitamin A deficiency?

A. Xerophthalmia
B. Bitôt spots
C. Delayed dark adaptation
D. Keratomalacia
E. Corneal ulceration

50. Which of the following is the mainstay of initial treatment of hypervitaminosis D?

A. Eliminate the source of excess vitamin D
B. Calcitonin
C. Bisphosphonates
D. Glucocorticoids
E. Normal saline

51. Which of the following vitamins B complex will be lost when the rice is repeatedly washed and the cooking water is discarded?

A. Thiamine (B1)
B. Riboflavin (B2)
C. Niacin (B3)
D. Pyridoxine (B6)
E. Biotin

52. Children with unexplained ataxia should be screened for which of the following vitamins deficiency?

A. A
B. D
C. K
D. E
E. C

53. Which of the following food items has the highest vitamin D content?

A. Fish liver oils
B. Red meat
C. Green vegetable
D. Soya milk
E. Chicken

54. How much is the approximate content of vitamin D in breast milk?

A. 12-60 IU/L
B. 60-100 IU/L
C. 100-200 IU/L
D. 200-300 IU/L
E. 300-400 IU/L

55. Which of the following age groups is most frequently affected by vitamin D deficiency?

A. Newborn
B. Infant
C. Toddler
D. Preschool
E. School age

56. Transplacental transport of vitamin D can provide enough vitamin D for the first

A. 1 month
B. 2 months
C. 3 months
D. 4 months
E. 6 months

57. Which of the following causes of vitamin D deficiency is associated with a high serum phosphate level?

A. Vitamin D–dependent rickets
B. Nutritional vitamin D deficiency
C. Chronic kidney disease
D. Congenital rickets
E. Dent disease

58. An 8-year-old girl with precocious puberty had a history of multiple pathological fractures. Examination shows hyperpigmented macules and her investigations reveal hyperthyroidism, low levels of 1,25-D, and elevated ALP levels.

Of the following, the **MOST** likely diagnosis is

A. ovarian cyst
B. McCune Albright syndrome

C. ovarian tumour
D. epidermal nevus syndrome
E. Raine syndrome

59. Which of the following is the **MOST** likely presentation of rickets of prematurity?

A. Asymptomatic
B. Respiratory distress
C. Poor linear growth
D. Fractures and softening of the ribs
E. Classic rachitic findings

60. Which of the following is the **MOST** common cause of hypervitaminosis D?

A. Misuse of vitamin D supplements
B. Overfortification of milk
C. Contamination of table sugar
D. Use of vitamin D supplements as cooking oil
E. Over exposure to sunlight

61. What is the recommended daily upper limit for long-term vitamin D intake for infants?

A. 500 IU
B. 1000 IU
C. 1500 IU
D. 2000 IU
E. 2500 IU

62. Which of the following laboratory finding will be normal in hypervitaminosis D?

A. Serum calcium
B. 25-D level
C. 1,25-D level
D. PTH level
E. Phosphate level

Chapter 5

Nutrition
Answers

AQEEL MAHDI

1.(A) Growth during infancy is rapid, critical for neurocognitive development, and has the highest energy and nutrient requirements relative to body size than any other period of growth.

2.(D) WHO recommendation: Give no pacifiers or artificial nipples to breastfeeding infants.

AAP recommendation: The American Academy of Pediatrics endorsed the UNICEF-WHO Ten Steps to Successful Breastfeeding, but does not support a categorical ban on pacifiers because of their role in reducing the risk of sudden infant death syndrome and their analgesic benefit during painful procedures when breastfeeding cannot provide the analgesia.

3.(E) In acute severe malnutrition do not give IV fluids except in shock.

4.(B) Organ meats (especially liver, kidney) are very rich in vitamin A, whereas other meats, milk, and cheese contain moderate levels.

5.(A) Iron deficiency is the most common micronutrients deficiency in the world and is associated with iron deficiency anemia and neurocognitive deficit in some children.

6.(D) Coffee, tea, fermented fish are thiamine antagonist and may contribute to thiamine deficiency.

7.(A) Death from thiamine deficiency usually is secondary to cardiac involvement. The initial signs are cyanosis and dyspnea, but tachycardia, enlargement of the liver, loss of consciousness, and convulsions can develop rapidly.

8.(B) Thiamine deficiency is classically associated with a diet consisting largely of polished rice. It develops within 2-3 months of deficient intake.

9.(C) It begins with pallor at the angles of the mouth and progresses to thinning and maceration of the epithelium.

10.(B) The side chain of the B 2 vitamin is photochemically destroyed during phototherapy for hyperbilirubinemia.

11.(C) Malnutrition is defined as acute when its duration is below 3 months and chronic when its equal or more than 3 months.

12.(C) Hypervitaminosis A is most often caused by excessive ingestion of vitamin A containing supplements for several weeks or months .
13.(A) Because of lack of a good functional test to evaluate niacin status, the diagnosis of deficiency is usually made from the physical signs of glossitis, GI symptoms, and a symmetric dermatitis.
14.(C) peripheral neuritis is a feature of deficiency in adult but is not usual in children, the other clinical features may occur in infants.
15.(D) Oxaluria, oxalic acid bladder stones formation, hyperglycinemia, and lymphopenia are associated with vitamin B6 deficiency.
16.(D) Avidin found in raw egg whites acts as a biotin antagonist. Signs of biotin deficiency have been demonstrated in persons who consume large amounts of raw egg whites over long periods. Deficiency also occurs with parenteral and enteral nutritional formula that lack biotin.
17.(A) Dietary sources of vitamin B12 are almost exclusively from animal foods. Organ meats, muscle meats, seafood (mollusks, oysters, fish), poultry, and egg yolk are rich sources.
18.(C) Infants and children on highly restrictive diets, devoid of most fruits and vegetables, are at risk of acquiring severe vitamin C deficiency. Such diets are occasionally promoted with unsubstantiated claims of benefit in autism and other developmental disorders. High index of suspicion is required in children on restrictive diets, particularly those with autism and other developmental disorders, and they should be evaluated for scurvy whenever they present with difficulty in walking or bone pains.
19.(E) Clinical manifestations do not appear until after 1-year of age, even in children with cholestasis since birth. Patients may have cerebellar disease, posterior column dysfunction, and retinal disease. Loss of deep tendon reflexes is usually the initial finding.
20.(B) In premature infants, hemolysis as a result of vitamin E deficiency typically develops during the 2nd month of life. Edema may also be present.
21.(A) Loss of deep tendon reflexes is usually the initial finding. Subsequent manifestations include limb ataxia (intention tremor, dysdiadochokinesia), truncal ataxia (wide-based, unsteady gait), dysarthria, ophthalmoplegia (limited upward gaze), nystagmus, decreased proprioception (positive Romberg test), decreased vibratory sensation, and dysarthria.
22.(D) Unlike other fat-soluble vitamins, there are limited body stores of vitamin K. In addition, there is high turnover of vitamin K, and the vitamin K–dependent clotting factors have a short half-life. Hence, symptomatic vitamin K deficiency

can develop within weeks when there is inadequate supply because of low intake or malabsorption.

23.(A) The most common site of bleeding in late VKDB is intracranial, although cutaneous and GI bleeding may be the initial manifestation. Intracranial bleeding can cause convulsions, permanent neurologic sequelae, or death.

24.(A) Factor VII has the shortest half-life of the coagulation factors and is the first to be affected by vitamin K deficiency, but isolated factor VII deficiency does not affect the partial thromboplastin time.

25.(E) Poor infant positioning and improper latch are the most common reasons for nipple pain beyond the mild discomfort felt early in breastfeeding. If the problem persists and the infant refuses to feed, evaluation for nipple candidiasis is indicated.

26.(A) Parental preference is the most common reason for using infant formula. However, infant formula is also indicated for infants whose intake of breast milk is contraindicated for infant factors (e.g., inborn errors of metabolism) and maternal factors. In addition, infant formula is used as a supplement to support inadequate weight gain in breastfed infants.

27.(A)

28.(D) Selenium is an enzyme cofactor (prevents oxidative damage), its deficiency causes cardiomyopathy (Keshan disease), myopathy. Its available in meat, seafood, whole grains, and garlic.

29.(B) Copper deficiency causes Microcytic anemia, osteoporosis, neutropenia, neurologic symptoms, and depigmentation of hair and skin.

30.(A) Around 12 mo. of age, the child learns to drink from a cup and may still breastfeed or desire formula bottle feeding. Bottle weaning should begin around 12-15mo, and bedtime bottles should be discouraged because of the association with dental carries.

31.(C) Iron is given in the maintenance phase of treatment.

32.(E) The American Paediatric Surgical Association guidelines recommends that surgery be considered only in children with complete or near-complete skeletal maturity, a BMI ≥40, and a medical complication resulting from obesity, after they have failed 6 mo. of a multidisciplinary weight management program. Hormone replacement therapy is available for patients with leptin deficiency.

33.(E) Hereditary hypophosphatemic rickets with hypercalciuria (HHRH) is a rare disorder that is mainly found in the Middle East.

34.(D) Breastfeeding has documented short- and long-term medical and neurodevelopmental advantages and rare contraindications of which is active herpetic lesions of the breast.

35.(E)
- HIV infection in United states, breastfeeding is contraindicated.
- Hepatitis C infection, breastfeeding is not contraindicated.
- CMV infection may be found in milk of mothers who are CMV seropositive. Transmission through human milk causing symptomatic illness in term infants is uncommon
- Hepatitis B infection, infants routinely receives hepatitis B immune globulin and hepatitis B vaccine if mother is HBsAg positive. No delay in initiation of breastfeeding is required.

36.(D) The pumping of breast milk is a common practice when the mother and baby are separated. Glass or plastic containers should be used to collect the milk, and milk should be refrigerated and then used within 48 hr. Expressed breast milk can be frozen and used for up to 6 mo. Milk should be thawed rapidly by holding under running tepid water and used completely within 24 hr after thawing. Milk should never be microwaved.

37.(D) cans should be covered with the original plastic cap or aluminum foil, and the powdered product can be used within 4 wk. Once prepared, all bottles, regardless of type of formula, should be used within 24 hr. Formula should be used within 2 hr of removal from the refrigerator, and once a feeding has started, that formula should be used within 1 hr or be discarded. Prepared formula stored in the refrigerator should be warmed by placing the container in warm water for about 5 min. Formula should not be heated in a microwave because it can heat unevenly and result in burns, despite appearing to be at the right temperature when tested.

38.(E) Indications for soy formula include galactosemia, preference for a vegetarian diet, and hereditary lactase deficiency, because soy-based formulas are lactose free. The routine use of soy protein–based formula has no proven value in the prevention or management of infantile colic, fussiness, or atopic disease. Infants with documented cow's milk protein–induced enteropathy or enterocolitis often are also sensitive to soy protein. They should be provided formula derived from extensively hydrolyzed protein or synthetic amino acids.

39.(D) It is important to understand different variations in vegetarianism, as follows:

- ✓ Veganism: excludes all animal products. It may be part of a larger practice of abstaining from the use of animal products for any purpose.
- ✓ Ovovegetarianism: includes eggs but not dairy products.
- ✓ Lactovegetarianism: includes dairy products but excludes eggs.
- ✓ Lactoovovegetarianism: includes eggs and dairy products.

- ✓ Flexitarian: a vegetarian who will occasionally eat meat.
- ✓ Pescatarian: consumes fish, but often self-labeled a vegetarian.

40.(C) Iron absorption may be inhibited by dietary constituents, such as phytate (found in leafy green vegetables and whole grains). Foods rich in iron include iron-fortified cereals, black beans, cashews, kidney beans, lentils, oatmeal, raisins, black-eyed peas, soybeans, sunflower seeds, chickpeas, molasses, chocolate, and tempeh. Iron absorption can be increased by eating food containing ascorbic acid (vitamin C) along with foods containing iron.

41.(D) Hemoglobin cutoffs to define anemia are 110 g/L for children 6-59 mo, 115 g/L for children 5-11 year, and 120 g/L for children 12-14 yr.

42.(A) Give ReSoMal 5 mL/kg every 30 min for 1st 2 hr orally or NG tube. Then give 5-10 mL/kg in alternate hours for up to 10 hr. Amount depends on stool loss and eagerness to drink. Feed in the other alternate hour. Do not give IV fluid unless the child is in shock.

43.(B) Water 2 L, WHO ORS One sachet, Sucrose 50 g, electrolyte/mineral solution 40 mL. ReSoMal contains 37.5 mmol sodium and 40 mmol potassium/L. WHO ORS sachet contains 2.6 sodium chloride, 2.9 g trisodium citrate dihydrate, 1.5 g potassium chloride, and 13.5 g glucose.

44.(E) If no complications give Amoxicillin, 25 mg/kg PO twice daily for 5 days.If complications (shock, hypoglycemia, hypothermia, skin lesions, respiratory or urinary tract infections, or lethargy/sickly give Gentamicin, 7.5 mg/kg IV or IM once daily for 7 days and Ampicillin, 50 mg/kg IV or IM every 6 hr for 2 days, then amoxicillin, 25-40 mg/kg PO every 8 hr for 5 days. For persistent diarrhea or small bowel overgrowth, add metronidazole, 7.5 mg/kg PO every 8 hr for 7 days.

45.(D) Melanocortin 4 receptor gene mutation causes early-onset severe obesity, increased linear growth, hyperphagia, hyperinsulinemia. Its most common known genetic cause of obesity. Homozygous worse than heterozygous.

46.(A) An analysis of 9 randomized controlled trials found that vitamin A appears to be beneficial in reducing death or oxygen requirement with no difference in neurodevelopmental outcomes.

47.(A) Orlistat, which decreases absorption of fat, resulting in modest weight loss. Complications include flatulence, oily stools, and spotting.

48.(C) TV watching is often associated with eating, and many highly caloric food products are marketed directly to children during child-oriented television programs.

49.(C) An earlier symptom of vitamin A deficiency is delayed dark adaptation, as a result of reduced resynthesis of rhodopsin; this may progress to night blindness.

50.(E) The mainstay of the initial treatment is aggressive therapy with normal saline, often in conjunction with a loop diuretic to further increase calcium excretion; this is often adequate for treating mild or moderate hypercalcemia.

51.(A) Thiamine is water soluble and heat labile; most of the vitamin is lost when the rice is repeatedly washed and the cooking water is discarded.

52.(D) Ataxia with isolated vitamin E deficiency (AVED), a rare autosomal recessive disorder, there are mutations in the gene for α-tocopherol transfer protein (TTPA). Because children with AVED do not have symptoms of malabsorption, a correct diagnosis requires a high index of suspicion. Patients with this disorder are unable to incorporate vitamin E into lipoproteins before their release from the liver, leading to reduced serum levels of vitamin E. There is no associated fat malabsorption, and absorption of vitamin E from the intestine occurs normally.

53.(A) There are few natural dietary sources of vitamin D. Fish liver oils have a high vitamin D content. Other good dietary sources include fatty fish and egg yolks. Most children in industrialized countries receive vitamin D via fortified foods, especially formula and milk (both of which contain 400 IU/L) and some breakfast cereals and breads.

54.(A) Breast milk has a low vitamin D content, approximately 12-60 IU/L.

55.(B) Vitamin D deficiency most frequently occurs in infancy because of a combination of poor intake and inadequate cutaneous synthesis.

56.(B) Transplacental transport of vitamin D, mostly 25-D, typically provides enough vitamin D for the 1st 2 mo of life unless there is severe maternal vitamin D deficiency.

57.(C) In chronic kidney disease, unlike the other causes of vitamin D deficiency, patients have hyperphosphatemia as a result of decreased renal excretion.

58.(B) McCune Albright syndrome, an entity that includes the triad of polyostotic fibrous dysplasia, hyperpigmented macules, and polyendocrinopathy. Affected patients have inappropriately low levels of 1,25-D and elevated ALP levels.

59.(A) Most infants with rickets of prematurity have no clinical manifestations, and the diagnosis is based on radiographic and laboratory findings. Rickets of prematurity occurs 1-4 mo after birth. Infants can have nontraumatic fractures. Some infants have respiratory distress from atelectasis and poor ventilation. These infants have poor linear growth. There may be classic rachitic findings,

such as frontal bossing, rachitic rosary, craniotabes, and widened wrists and ankles

60.(A) Hypervitaminosis D is caused by excessive intake of vitamin D. It can occur with long-term high intake or with a substantial, acute ingestion. Most cases are secondary to misuse of prescribed or nonprescription vitamin D supplements, but other cases have been secondary to accidental overfortification of milk, contamination of table sugar, and inadvertent use of vitamin D supplements as cooking oil. Vitamin D intoxication is never secondary to excessive exposure to sunlight.

61.(B) The recommended upper limits for long-term vitamin D intake are 1,000 IU for children <1 yr old and 2,000 IU for older children and adults.

62.(C) The classic findings in vitamin D intoxication are hypercalcemia, elevated levels of 25-D (>100 ng/mL), hypercalciuria, and suppressed PTH. Hyperphosphatemia is also common. Hypercalciuria can lead to nephrocalcinosis, which is visible on renal ultrasound. Surprisingly, levels of 1,25-D are usually normal. Anemia is sometimes present; the mechanism is unknown.

Chapter 6

Fluid and Electrolyte Disorders Questions

KHALID ALAARJI

1. A 6-month-old infant waiting for surgery, should begin receiving maintenance IV fluids within
   A. 4 hr of the last feeding
   B. 6 hr of the last feeding
   C. 8 hr of the last feeding
   D. 10 hr of the last feeding
   E. 12 hr of the last feeding

2. What is the percentile of body weight for height to calculate the maintenance IV fluid in an overweight child?
   A. 10^{th}
   B. 25^{th}
   C. 50^{th}
   D. 75^{th}
   E. 90^{th}

3. The upper limit of maintenance IV fluid in an overweight child (adult-sized patients) is
   A. 1L/24 hr
   B. 1.6L/24 hr
   C. 2L/24 hr
   D. 2.4L/24 hr
   E. 3L/24 hr

4. Calculating daily maintenance fluid volume for children weighing 11-20 kg is
   A. 100 mL/kg
   B. 150 mL/kg
   C. 1,000 mL + 50 mL/kg for each kg >10 kg
   D. 1,000 mL + 20 mL/kg for each kg >10 kg
   E. 1,500 mL + 20 mL/kg for each kg >10 kg

5. Which of the following intravenous solutions is composed from Na^+ (77 meq/L) and Cl^- (77 meq/L)?

A. Normal saline (0.9% NaCl)
B. ½ normal saline (0.45% NaCl)
C. 0.2 normal saline (0.2% NaCl)
D. ½ Glucose saline
E. Ringer lactate

6. Administration of 0.2 NS (osmolality=68) peripherally as maintenance therapy is contraindicated due to risk of

A. hemolysis
B. hyponatremia
C. hypernatremia
D. brain edema
E. central pontine myelinolysis

7. As maintenance IV fluid, surgical patients during surgery and in the recovery room for 6-8 hr postoperatively typically should receive

A. normal saline (0.9% NaCl)
B. ½ normal saline (0.45% NaCl)
C. 0.2 normal saline (0.2% NaCl)
D. ½ glucose saline
E. 5% dextrose

8. The **BEST** solution for replacement of ongoing losses in a 3-year-old child presented with diarrhea and a limited ability to take oral fluid is

A. Ringer lactate
B. normal saline + 10 mEq/L KCl
C. ½ normal saline + 10 mEq/L KCl
D. NS + 30 mEq/L sodium bicarbonate + 20 mEq/L KCl
E. D5 ½ NS + 30 mEq/L sodium bicarbonate + 20 mEq/L KCl

9. A 15-year-old girl admitted to the hospital for evaluation; investigation reveals serum Na+ (131 mEq/L), serum K+ (2.6 mEq/L), serum Cl^- (82 mEq/L), BUN (42 mg/dl), and serum creatinine (0.9 mg/dl), pH (7.52), $HCO3^-$ (41 mEq/L), PCO2 (51 mm Hg) and urine for [Cl^-] (25 mEq/L)
Of the following, the **MOST** likely diagnosis is

A. frequent emesis

B. chloride-losing diarrhea
C. low chloride formula
D. Gitelman syndrome
E. cystic fibrosis

10. Which of the following is a feature of mild dehydration?
A. Thirst
B. Tachycardia
C. Lethargy
D. Sunken fontanel
E. Decreased tears

11. Which of the following is a clue to severe dehydration?
A. Cold and mottled skin
B. Oliguria
C. Sunken eyes and fontanel
D. Dry mucous membranes
E. Irritability

12. An 18-hour-old newborn with normal vaginal delivery without complication admitted to NICU with repeated apneic attacks and hypotonia with history of severe maternal preeclampsia treated with IV magnesium and hydralazine; O/E floppy, decrease deep tendon reflexes, and hypotension. ECG shows prolonged PR interval, QRS complex, and QT interval.
Of the following, the **MOST** likely diagnosis is
A. perinatal asphyxia
B. early onset sepsis
C. hypermagnesemia
D. hypocalcemia
E. Werdnig-Hoffmann disease

13. A 15-year-old girl presents to emergency unit with breathlessness and chest pain; ECG and chest radiograph are normal; yesterday she was notified with school failure.
Of the following, the **MOST** likely arterial blood gases in this condition is
A. pH (7.09), $HCO3^-$ (28 mEq/L), PCO2 (68 mm Hg)
B. pH (7.12), $HCO3^-$ (14 mEq/L), PCO2 (65 mm Hg)
C. pH (7.35), $HCO3^-$ (20 mEq/L), PCO2 (45 mm Hg)

D. pH (7.55), $HCO3^-$ (16 mEq/L), PCO2 (18 mm Hg)
E. pH (7.52), $HCO3^-$ (40 mEq/L), PCO2 (48 mm Hg)

14. An 11-month-old baby, who has small bowel resection at age of 2 months brought to emergency unit with seizure; Examination shows failure to thrive and seatorrhea; serum calcium (6.5 mg/dl); treated with IV infusion of 10% calcium gluconate 10 ml but still no response. ECG shows flattening of the T wave and lengthening of the ST segment.
Of the following, the **MOST** likely explanation is
A. incorrect dose
B. hypoparathyroidism
C. hypomagnesemia
D. hyperphosphatemia
E. hyponatremia

15. An 18-month-old child presents with history of vomiting, diarrhea, and decreased urine output. Examination shows irritable child with capillary refill of 2 sec, dry mucous membranes, and tachycardia.
Of the following, the **MOST** likely percent of dehydration is
A. <3%
B. 3-6 %
C. 7-9%
D. 10–12%
E. >12%

16. A 12-month-old infant presents with vomiting, diarrhea, and no urine output. Examination shows depressed consciousness with capillary refill of 4 sec and very sunken eyes.
Of the following, the **BEST** initial fluid therapy is
A. 0.9% NS
B. ½ NS
C. ¼ NS
D. 0.2 NS
E. ½ GS

17. Which of the following should be avoided in a child with isolated vomiting?
A. Isotonic NS
B. Ringer lactate

C. ½ NS
D. ½ GS
E. 0.2 NS

18. A 7-year-old girl admitted to intensive care unit for management of Guillain-Barré syndrome, at morning of 7th day, the patient develops severe headache and repeated vomiting; O/E well hydrated, afebrile, no neck stiffness, and blood pressure 100/60 mm Hg; serum Na^+ (120 mEq/L), serum K^+ (4.1 mEq/L), serum Chloride (75 mEq/L), blood glucose (98 mg/dl), BUN 20 mg/dl), serum creatinine (0.5 mg/dl), and urine sodium (80 mEq/L).
Of the following, the **MOST** likely explanation of this condition is

A. meningitis
B. side effect of IVIG
C. syndrome of inappropriate antidiuretic hormone secretion
D. cerebral salt wasting
E. acute tubular necrosis

19. A 4-month-old girl present with sudden onset of rapid deep breathing and mild dehydration; serum Na+ (145 mEq/L), serum K+ (6 mEq/L), serum Cl^- (90 mEq/L), pH (7.11), $HCO3^-$ (10 mEq/L), lactate 4 mmol/L and PCO2 (23 mm Hg)
Of the following, the **MOST** likely cause is

A. diarrhea
B. distal (type I) RTA
C. proximal (type II) RTA
D. urinary tract diversion
E. isovelaric acidemia

20. Which of the following is a risk factor for hyponatremic dehydration in children with diarrhea?

A. Consumption of diluted formula
B. Inability to take oral fluid
C. Poor thirst mechanism
D. Intractable emesis
E. Hyperpyrexia

21. Hyponatremia is defined as serum sodium less than

A. 125 mEq/L
B. 130 mEq/L

C. 135 mEq/L
D. 140 mEq/L
E. 145 mEq/L

22. In hyponatremic dehydration, the serum sodium **SHOULD NOT** be corrected more than

A. 10 mEq/L/24 hr
B. 12 mEq/L/24 hr
C. 14 mEq/L/24 hr
D. 16 mEq/L/24 hr
E. 18 mEq/L/24 hr

23. A bottle fed 18-month-old boy presents with history of vomiting and diarrhea. Examination shows severe degree of dehydration; serum Na+ (126 mEq/L), serum K+ (4.5mEq/L), pH (7.25), HCO3 (14 mEq/L), and PCO2 30 mm Hg; he received IV fluid therapy and after 12 hours, the patient became confused and agitated. Serum electrolytes were done revealing serum Na+ (145 mEq/L), serum K+ (3.7mEq/L), serum Ca+ (8.7 mg/dl), and blood glucose (98 mg/dl).
Of the following, the **MOST** likely cause of his deterioration is

A. cerebral edema
B. central pontine myelinolysis
C. arrhythmias
D. brain herniation
E. intracranial hemorrhage

24. An 11-month-old bottle fed infant was brought to emergency unit with tonic-clonic seizure. He complains from vomiting and diarrhea with excessive water intake for the last 3 days. Examination shows moderate degree of dehydration; serum Na^+ (116 mEq/L), serum K^+ (3.8mEq/L), serum Ca+ (8.7 mg/dl), and blood glucose (98 mg/dl).
Of the following, the **FIRST** line of treatment is

A. 0.9% NS
B. 3% NS
C. Ringer lactate
D. ½ NS
E. 0.2 NS

25. A 10-month-old infant presents with frequent loose bowel motions and excessive thirst over the past 24 hours. Examination shows restlessness, sunken eyes, and skin pinch goes back slowly.
Of the following, the **MOST** appropriate therapy is

A. IV normal saline (0.9% NaCl)
B. IV ½ normal saline + 10 mEq/L KCl
C. IV D5 ½ NS + 30 mEq/L sodium bicarbonate
D. Standard oral rehydration solution (ORS)
E. Low osmolarity ORS

26. An 18-month-old bottle fed girl was presented with vomiting and diarrhea. Examination showed moderate degree of dehydration; serum Na+ (172 mEq/L), serum K+ (4.5mEq/L), pH (7.25), HCO3 (14 mEq/L), and PCO2 30mm Hg; she received IV fluid therapy and after 12 hours, the patient develops convulsion with decerebrate posture, repeat serum electrolyte reveals; serum Na+ (156 mEq/L), serum K+ (3.7 mEq/L), serum Ca+ (8.7 mg/dl), and blood glucose (98 mg/dl).
Of the following, the **MOST** likely cause of his deterioration is

A. cerebral edema
B. central pontine myelinolysis
C. arrhythmia
D. renal failure
E. intracranial hemorrhage

27. A 14-month-old bottle fed girl presents with vomiting and diarrhea for the last 24 hours. Examination shows an irritable child with doughy skin and capillary refill of 3 seconds; serum Na^+ (160 mEq/L), serum K^+ (4.1 mEq/L), BUN (110 mg/dl), and serum creatinine (1.3 mg/dl).
Of the following, the **BEST** initial fluid therapy is

A. Isotonic N S
B. ½ N S
C. ¼ N S
D. 0.2 N S
E. Ringer lactate

28. A 5-year-old child admitted to emergency unit with rapid deep breathing, polyuria, polydipsia, and dehydration for the last 2 weeks; serum Na^+ (110 mEq/L) and serum osmolality 320 mOsm/kg.

Of the following, the **MOST** likely cause of hyponatremia is

A. leukocytosis
B. increased blood urea
C. acidosis
D. increased blood glucose
E. water depletion

29. A 9-month-old girl presents with vomiting and diarrhea for the last 24 hours. Examination shows an irritable child with doughy skin and capillary refill of 3 seconds; serum Na^+ (168 mEq/L), serum K^+ (4.1 mEq/L), BUN (110 mg/dl), and serum creatinine (1.3 mg/dl); the patient shows improvement on two shots of isotonic N S (each 20 mL/kg over 20 min) and has passed urine.
Of the following, the **NEXT** step in management is administration of

A. oral rehydration solution
B. 5% dextrose + ½ normal saline over 24 hr
C. 5% dextrose + ½ normal saline over 48 hr
D. 5% dextrose + ¼ normal saline over 24 hr
E. 5% dextrose + ¼ normal saline over 48 hr

30. A 9-month-old girl presents with rapid deep breathing and mild dehydration; serum Na+ (132 mEq/L), serum K+ (3 mEq/L), serum Cl^- (114 mEq/L), pH (7.05), $HCO3^-$ (12 mEq/L), and PCO2 (25 mm Hg).
Of the following, the **MOST** likely cause is

A. lactic acidosis
B. diabetic ketoacidosis
C. renal tubular acidosis
D. renal failure
E. proprionic acidemia

31. In acute respiratory acidosis; each 10 mm Hg increase in PCO2 is compensated by increase plasma bicarbonate of

A. 1
B. 1.5
C. 2
D. 2.5
E. 3

32. A 6-month-old infant is evaluated for failure to thrive, has past history of frequent hospital admissions because of repeated vomiting, dehydration, and increased urine output with constipation; abdominal ultrasound reveals nephrocalcinosis; serum K+ (2mmol/L).
Of the following, the **EXPECTED** arterial blood gases of this patient is

A. pH (7.09), $HCO3^-$ (28 mEq/L), PCO2 (68 mm Hg)
B. pH (7.12), $HCO3^-$ (14 mEq/L), PCO2 (65 mm Hg)
C. pH (7.35), $HCO3^-$ (20 mEq/L), PCO2 (45 mm Hg)
D. pH (7.50), $HCO3^-$ (16 mEq/L), PCO2 (25 mm Hg)
E. pH (7.52), $HCO3^-$ (40 mEq/L), PCO2 (40 mm Hg)

33. Evaluation of a 6-month-old infant with failure to thrive reveals serum Na+ (128 mEq/L), serum K+ (2.3 mEq/L), serum Cl^- (82 mEq/L), BUN (32 mg/dl), and serum creatinine (0.6 mg/dl). Blood gas analysis showed pH (7.52), $HCO3^-$ (36 mEq/L), and PCO2 (46 mm Hg).
Of the following, the **MOST** helpful test for the diagnosis of this condition is

A. urine for Na^+
B. urine for Cl^-
C. urine for K^+
D. serum chloride
E. serum calcium

34. What is the **FIRST** electrocardiogram (ECG) finding in hyperkalemia?

A. Peaked T wave
B. ST-segment depression
C. Increased PR interval
D. P wave flattening
E. QRS complex widening

35. A 14-year-old boy with sickle cell disease presents with generalized muscle weakness; examination shows irregular heart rate with decreased muscle strength; ECG shows peaked T wave, ST-segment depression, and increased PR interval.
Of the following, the **FIRST** line of therapy is

A. dialysis
B. insulin and glucose
C. kayexalate
D. I V calcium

E. albuterol nebulizer

36. What is the **MOST** likely cause of hypokalemia and metabolic acidosis?

A. Emesis
B. Diarrhea
C. Aldosterone excess
D. Use of diuretics
E. Bartter syndrome

37. A 6-year-old girl presents to emergency unit with deep rapid breathing and dehydration; blood glucose (560 mg/dl).
Of the following, the **EXPECTED** arterial blood gas analysis is

A. pH (7.05), $HCO3^-$ (12 mEq/L), PCO2 (25 mm Hg)
B. pH (7.09), $HCO3^-$ (28 mEq/L), PCO2 (68 mm Hg)
C. pH (7.12), $HCO3^-$ (14 mEq/L), PCO2 (65 mm Hg)
D. pH (7.35), $HCO3^-$ (20 mEq/L), PCO2 (45 mm Hg)
E. pH (7.45), $HCO3^-$ (32 mEq/L), PCO2 (65 mm Hg)

Chapter 6

Fluid and Electrolyte Disorders Answers

KHALID ALAARJI

1.(C) A normal teenager who is given nothing by mouth (NPO) overnight for a morning procedure does not require maintenance fluids because a healthy adolescent can easily tolerate 12 or 18 hr without oral intake. In contrast, a 6 mo old child waiting for surgery should begin receiving IV fluids within 8 hr of the last feeding. Infants become dehydrated more quickly than older patients.

2.(C) Calculations of maintenance IV fluid based on weight do overestimate the water needs of an overweight child, in whom it is better to base the calculations on the lean body weight, which can be estimated by using the 50^{th} percentile of body weight for the child's height.

3.(D)

4.(C)

5.(B)

6.(A) A normal plasma osmolality is 285-295 mOsm/kg. Infusing an IV solution peripherally with a much lower osmolality can cause water to move into red blood cells, leading to hemolysis. Thus, 0.2 NS (osmolality = 68) should not be administered peripherally, but D5 0.2NS (osmolality = 346) or D5 ½ NS + 20 mEq/L potassium chloride (KCl) with an osmolality of 472 can be administered.

7.(A) Surgical patients typically receive isotonic fluids (NS, LR) during surgery and in the recovery room for 6-8 hr postoperatively; the rate is typically approximately two-thirds the calculated maintenance rate, with dextrose added if clinically indicated.

8.(E) Average composition of diarrhea; sodium: 55 mEq/L, potassium: 25 mEq/L, bicarbonate: 15 mEq/L; Approach to replacement of ongoing losses; Solution (D5 ½ NS + 30 mEq/L sodium bicarbonate + 20 mEq/L KCl). Average composition of gastric fluid; sodium: 60 mEq/L, potassium: 10 mEq/L, chloride: 90 mEq/L; Approach to replacement of ongoing losses; Solution (normal saline + 10 mEq/L KCl).

9.(D)

Causes of Chloride-Responsive Metabolic Alkalosis (urinary chloride <15 mEq/L)	
Gastric losses (Emesis or Nasogastric suction) Diuretics (loop or thiazide) Chloride-losing diarrhea Low chloride formula Cystic fibrosis Posthypercapnia	
Causes of Chloride-Resistant Metabolic Alkalosis (urinary chloride >20 mEq/L)	
Normal Blood Pressure	***High Blood Pressure***
Adrenal adenoma or hyperplasia Glucocorticoid-remediable aldosteronism Renovascular disease Renin-secreting tumor 17α-Hydroxylase deficiency Cushing syndrome Liddle syndrome	Gitelman syndrome Bartter syndrome AD hypoparathyroidism EAST syndrome Base administration

10.(A) Mild dehydration (<5% in an infant; <3% in an older child or adult): Normal or increased pulse; decreased urine output; thirsty; normal physical findings

11.(A) Severe dehydration (>10% in an infant; >6% in an older child or adult): Peripheral pulses either rapid and weak or absent; decreased blood pressure; no urine output; very sunken eyes and fontanel; no tears; parched mucous membranes; delayed elasticity (poor skin turgor); very delayed capillary refill (>3 sec); cold and mottled; limp, depressed consciousness.

12.(C) Clinically significant hypermagnesemia is almost always secondary to excessive intake. It is unusual, except in neonates born to mothers who are receiving IV magnesium for preeclampsia or eclampsia. Symptoms usually do not appear until the plasma magnesium level is >4.5 mg/dL. Hypermagnesemia inhibits acetylcholine release at the neuromuscular junction, producing hypotonia, hyporeflexia, and weakness; paralysis occurs at high concentrations. The neuromuscular effects may be exacerbated by aminoglycoside antibiotics. Direct CNS depression causes lethargy and sleepiness; infants have a poor suck. Elevated magnesium values are associated with hypotension because of vascular dilation, which also causes flushing.

13.(D) Respiratory alkalosis due to hyperventilation and washout of CO_2. Hyperventilation may be secondary to an underlying disease that causes pain, stress, or anxiety. In psychogenic hyperventilation or in panic attacks, there is no disease process accounting for the hyperventilation. This disorder may occur in a child who has had an emotionally stressful experience. Alternatively, it may be part of a panic disorder, especially if there are repeated episodes of hyperventilation.

14.(C) Hypomagnesemia causes secondary hypocalcemia by impairing the release of PTH by the parathyroid gland and through blunting of the tissue response to PTH. Thus, hypomagnesemia is part of the differential diagnosis of hypocalcemia. It usually occurs only at magnesium levels < 0.7 mg/dL. The dominant manifestations of hypomagnesemia are caused by hypocalcemia: tetany, presence of Chvostek and Trousseau signs, and seizures. However, with severe hypomagnesemia, these same signs and symptoms may be present despite normocalcemia.

ECG changes with hypomagnesemia include flattening of the T wave and lengthening of the ST segment. Arrhythmias may occur, almost always in the setting of underlying heart disease. Gastrointestinal and renal losses are the major causes of hypomagnesemia. Diarrheal fluid contains up to 200 mg/L of magnesium; gastric contents have only approximately 15 mg/L, but high losses can cause depletion. Steatorrhea causes magnesium loss as a result of the formation of magnesium-lipid salts; restriction of dietary fat can decrease losses.

15.(B)

- ✓ **Mild dehydration** (<5% in an infant; <3% in an older child or adult): Normal or increased pulse; decreased urine output; thirsty; normal physical findings
- ✓ **Moderate dehydration** (5–10% in an infant; 3–6% in an older child or adult): Tachycardia; little or no urine output; irritable/lethargic; sunken eyes and fontanel; decreased tears; dry mucous membranes; mild delay in elasticity (skin turgor); delayed capillary refill (>1.5 sec); cool and pale
- ✓ **Severe dehydration** (>10% in an infant; >6% in an older child or adult):Peripheral pulses either rapid and weak or absent; decreased blood pressure; no urine output; very sunken eyes and fontanel; no tears; parched mucous membranes; delayed elasticity (poor skin turgor); very delayed capillary refill (>3 sec); cold and mottled; limp, depressed consciousness

16.(A)

- ✓ Restore intravascular volume: Isotonic fluid (NS or LR): 20 mL/kg over 20 min and repeat as needed
- ✓ Calculate 24 hr fluid needs: maintenance +deficit volume
- ✓ Subtract isotonic fluid already administered from 24 hr fluid needs
- ✓ Administer remaining volume over 24 hr using 5% dextrose NS +20 mEq/L KCl
- ✓ Replace ongoing losses as they occur

17.(B) In a child with a known or probable metabolic alkalosis (e.g., child with isolated vomiting), LR or PlasmaLyte should not be used because the lactate or acetate would worsen the alkalosis. However, LR or PlasmaLyte may be preferable to NS in shock since it is a balanced solution; NS may cause a hyperchloremic metabolic acidosis.

18.(C) SIADH typically occurs with disorders of the CNS (infection, hemorrhage, trauma, tumor, thrombosis, Guillain-Barré syndrome), but lung disease (infection, asthma, positive pressure ventilation) and malignant tumors (producing ADH) are other potential causes.

Diagnostic Criteria for Syndrome of Inappropriate Antidiuretic Hormone Secretion

- ✓ Absence of: Renal, adrenal, or thyroid insufficiency - Heart failure, nephrotic syndrome, or cirrhosis - Diuretic ingestion - Dehydration
- ✓ Urine osmolality >100 mOsm/kg (usually >plasma)
- ✓ Serum osmolality <280 mOsm/kg and serum sodium <135 mEq/L
- ✓ Urine sodium >30 mEq/L
- ✓ Reversal of "sodium wasting" and correction of Hyponatremia with water restriction

19.(E) Metabolic Acidosis with Increased Anion Gap

- ✓ ***Lactic Acidosis***
- Tissue hypoxia (Shock, Hypoxemia, Severe anemia)
- Liver failure
- Malignancy
- Intestinal bacterial overgrowth
- Inborn errors of metabolism
- ✓ Medications (Metformin, Propofol, Linezolid)
- ✓ ***Ketoacidosis*** (Diabetic ketoacidosis, Starvation ketoacidosis, Alcoholic ketoacidosis)
- ✓ ***Kidney Failure***
- ✓ ***Poisoning*** (Ethylene glycol, Methanol, Salicylate, Toluene, Paraldehyde)

20.(A)

21.(C)

22.(A) Even though central pontine myelinolysis (CPM) is rare in pediatric patients, it is advisable to avoid correcting the serum [Na+] by >10 mEq/L/24 hr or >18 mEq/L/48 hr.

23.(B) With all causes of hyponatremia, it is important to avoid overly rapid correction, which may cause central pontine myelinolysis (CPM). This syndrome, which occurs within several days of rapid correction of hyponatremia, produces neurologic symptoms, including confusion, agitation, flaccid or spastic quadriparesis, and death. There are usually characteristic pathologic and radiologic changes in the brain, especially in the pons, but extrapontine lesions are quite common and may cause additional symptoms. Despite severe symptoms, full recovery does occur in some patients. Even though CPM is rare in pediatric patients, it is advisable to avoid correcting the serum [Na+] by >10 mEq/L/24 hr or >18 mEq/L/48 hr.

24.(B) Patients with hyponatremia can have severe neurologic symptoms, such as seizures and coma. The seizures associated with hyponatremia generally are poorly responsive to anticonvulsants. The child with hyponatremia and severe symptoms needs treatment that will quickly reduce cerebral edema. This goal is best accomplished by increasing the extracellular osmolality so that water moves down its osmolar gradient from the ICS to the ECS. Intravenous hypertonic saline rapidly increases serum [Na^+], and the effect on serum osmolality leads to a decrease in brain edema. Each mL/kg of 3% NaCl increases the serum [Na+] by approximately 1 mEq/L. A child with active symptoms often improves after receiving 4-6 mL/kg of 3% NaCl.

25.(E) The World Health Organization defines some dehydration as the presence of two or more of the following signs: restlessness/irritability, sunken eyes, drinks eagerly, thirsty, and skin pinch goes back slowly. Infants and children with some dehydration need rehydration therapy with ORS: ORS, 50-100 mL/kg over 3-4 hr. Continue breast feeding. After 4 hr, give food every 3-4 hr for children who normally receive solid foods. Item D is old ORS recommended in 1976 and item E is new ORS recommended in July, 2001c,

26.(A) The goal is to decrease the serum [Na^+] by <10 mEq/L every 24 hr. Idiogenic osmoles are generated within the brain during the development of hypernatremia; they increase the osmolality within the cells of the brain, providing protection against brain cell shrinkage caused by movement of water out of the cells and into the hypertonic ECF. Idiogenic osmoles dissipate slowly during the correction of hypernatremia. With overly rapid lowering of the

extracellular osmolality during the correction of hypernatremia, an osmotic gradient may be created that causes water movement from the ECS into the cells of the brain, producing cerebral edema. Symptoms of the resultant cerebral edema can range from seizures to brain herniation and death.

27.(A) The initial resuscitation of hypernatremic dehydration requires restoration of the intravascular volume 20 mL/kg over 20 min with isotonic N S. Ringer lactate should not be used because it is more hypotonic than NS and may cause to rapid a decrease in the serum [Na^+], especially if multiple fluid boluses are necessary.

28.(D) Hyperosmolality, as may occur with hyperglycemia, causes a low [Na^+] because water moves down its osmotic gradient from the ICS into the ECS, diluting the [Na^+]. However, because the manifestations of hyponatremia are a result of the low plasma osmolality, patients with hyponatremia resulting from hyperosmolality do not have symptoms of hyponatremia. When the etiology of the hyperosmolality resolves, such as hyperglycemia in diabetes mellitus, water moves back into the cells, and the [Na^+] rises to its "true" value. Mannitol or sucrose, a component of intravenous immune globulin (IVIG) preparations, may cause hyponatremia because of hyperosmolality.

29.(C) Treatment of Hypernatremic Dehydration

- ✓ Restore intravascular volume: Normal saline: 20 mL/kg over 20 min (repeat until intravascular volume restored)
- ✓ Determine time for correction on basis of initial sodium concentration:
  - [Na] 145-157mEq/L: 24 hr
  - [Na] 158-170 mEq/L: 48 hr
  - [Na] 171-183 mEq/L: 72 hr
  - [Na] 184-196 mEq/L: 84 hr
- ✓ Administer fluid at constant rate over time for correction: Typical fluid: 5% dextrose + half-normal saline (with 20 mEq/L KCl unless contraindicated)
- ✓ Typical rate: 1.25-1.5 times maintenance

30.(C) Metabolic Acidosis with Normal Anion Gap

- ✓ Diarrhea
- ✓ Renal tubular acidosis (RTA)
- Distal (type I) RTA
- Proximal (type II) RTA
- Mixed (type III) RTA
- Hyperkalemic (type IV) RTA
- Urinary tract diversions

- Posthypocapnia
- Ammonium chloride intake

31.(A) In chronic respiratory acidosis; plasma bicarbonate increases by 3.5 for each 10 mm Hg increase in the PCO2 (chronic compensation).

32.(E) In Bartter syndrome there are severe hypokalemia (usually <2.5 mmol/L), metabolic alkalosis, hypercalciuria, and increased urinary chloride with normal blood pressure; hypomagnesemia is seen in a minority of patients but is more common in Gitelman syndrome.

33.(B) Measurement of the urine [Cl−] is the most helpful test in differentiating among the causes of a metabolic alkalosis. The urine [Cl−] is low in patients with a metabolic alkalosis resulting from volume depletion, unless there is a defect in renal handling of Cl−. The urine [Cl−] is superior to the urine [Na+] in assessment of volume status in patients with a metabolic alkalosis, because the normal renal response to a metabolic alkalosis is to excrete bicarbonate. Because bicarbonate is negatively charged, it can be excreted only with a cation, usually Na+ and K+. Therefore, a patient with a metabolic alkalosis may excrete Na+ in the urine despite the presence of volume depletion, which normally causes avid Na+ retention. The urine [Cl−] is usually a good indicator of volume status, and it differentiates Cl−-resistant and Cl−-responsive causes of a metabolic alkalosis.

34.(A)

35.(D) The goal of treatment of hyperkalemia is to stabilize the heart to prevent life-threatening arrhythmias with **calcium** which stabilizes the cell membrane of heart cells, preventing arrhythmias; it is given intravenously over a few minutes, and its action is almost immediate. **Bicarbonate** causes potassium to move intracellularly, lowering the plasma [K^+]; it is most efficacious in a patient with a metabolic acidosis. **Insulin** causes $K^+$to move intracellularly but must be given with glucose to avoid hypoglycemia. The combination of insulin and glucose works within 30 min. Nebulized **albuterol**, by stimulation of β1-adrenergic receptors, leads to rapid intracellular movement of K^+. While other measures (dialysis and kayexalate) are to remove K^+ from the body.

36.(B) The combination of hypokalemia and metabolic acidosis is characteristic of diarrhea and distal and proximal RTA. A concurrent metabolic alkalosis is characteristic of emesis or nasogastric losses, aldosterone excess, use of diuretics, and Bartter and Gitelman syndromes.

37.(A) In insulin-dependent diabetes mellitus, inadequate insulin leads to hyperglycemia and DKA. Production of acetoacetic acid and β-hydroxybutyric acid causes the metabolic acidosis.

Chapter 7

Pediatric Drug Therapy Questions

HAIDAR A. N. ABOOD

1. Which of the following terms is **BEST** to describe the study of genetic variations that give rise to interindividual differences in response to drugs?
   A. Pharmacoproteomics
   B. Pharmacokinetics
   C. Pharmacogenetics
   D. Pharmacodynamic
   E. Pharmacogenomics

2. Which of the following drug responses is due to the effect of gene polymorphism?
   A. Hyperpyrexia following aspirin overdose
   B. Prolonged paralysis after succinylcholine administration
   C. Antipyretic effect of paracetamol
   D. Bradycardia after administration of small dose of atropine
   E. Hypoglycemia associated with insulin therapy

3. Drug biotransformation reactions are conveniently classified into 2 main types, which occur sequentially and serve to terminate biologic activity and enhance elimination, phase I and phase II reactions.
Which of the following is **phase I** reaction?
   A. Glucuronidation reaction
   B. Conjugation with glutathione
   C. Sulphation reaction
   D. Oxidation reaction
   E. Conjugation with glycine

4. Which of the following is the major consequence of pharmacogenetic polymorphisms in drug metabolizing enzymes?
   A. Increased drug toxicity
   B. Increased drug metabolism

C. Increased drug clearance
D. Decreased drug efficacy
E. Decreased drug accumulation

5. A case of acute lymphoblastic leukemia, on maintenance treatment regime, has recurrent attacks of severe myelosuppression that required stopping of chemotherapy for a long time. On screening, the patient was found genetically having low TPMT (Thiopurine S-methyltransferase) activity. TMPT is cytosolic enzyme that catalyses the S-methylation of sulfur-containing drugs, hence, reducing their cytotoxic effects on bone marrow. For this reason, which of the following anticancer drugs should be removed from the regime?

A. Methotrexate
B. Vincristine
C. Prednisolone
D. L-asparaginase
E. 6-thioguanine

6. Polymorphisms of the β2-adrenergic receptor gene (ADRB2) are associated with variable responses to bronchodilator drugs.
Of the following, the **MOST** likely claimed drug is

A. ipratropium
B. salbutamol
C. theophylline
D. aminophylline
E. tiotropium

7. Drug bioavailability represents the fraction of the drugs that reach the systemic circulation after administration. It is always 100% if the drug is given by IV route.
Which age group has the **LEAST** bioavailability of drugs given by IM route?

A. Neonates
B. Infants
C. Toddlers
D. Preschool children
E. School children

8. Aminoglycoside antibiotics have a higher volume of distribution in neonates and infants (0.4-0.7 L/kg) than in adults (0.2-0.3 L/kg) because

A. the percentage of total body water is lower in neonates and infants
B. adults have higher percentage of intracellular water
C. the percentage of intracellular water is higher in neonates and infants
D. adults have higher percentage of extracellular water
E. the percentage of extracellular water is higher in neonates and infants

9. Which of the following age groups has the **HIGHEST** drug absorption after administration through rectal route?

A. Neonates
B. Infants
C. Toddlers
D. Preschool children
E. School children

10. Which of the following is the primary organ responsible for the elimination of drugs and their metabolites in pediatric age group?

A. Liver
B. Kidney
C. GIT
D. Skin
E. Lung

11. Which of the following dosing interval is recommended for gentamicin in young children who have serious infections?

A. 4 hours interval
B. 6 hours interval
C. 8 hours interval
D. 12 hours interval
E. 24 hours interval

12. In therapeutic drug monitoring (TDM), plasma drug concentration is used for dose individualization for a given patient. Which of the following drugs are frequently requiring TDM?

A. NSAIDs like aspirin
B. Beta-blockers like atenolol
C. Anticonvulsants like phenytoin
D. Antibiotics like penicillins
E. Anticoagulants like warfarin

13. Which of the following administration devices is **MOST** convenient to ensure accurate peroral administration of liquid drug formulations?

A. Kitchen teaspoon
B. 5 ml syringe
C. Hypodermic syringe
D. 5 mL dosing spoon
E. Medication bottle cap

14. Regarding drug administration by inhalation, which of the following factors has the **LEAST** effect on pulmonary drug absorption?

A. Drug particle size
B. Chemical stability of drug molecule in the lung
C. Internal airway diameter
D. Active versus passive drug delivery to tracheobronchial tree
E. Pulmonary surface area

15. A 24-month-old girl presents to the outpatient clinic with poor asthma control, she received montelukast (4 mg chewable tablets) daily for the last 3 months without satisfactory control of symptoms, if you decided to start inhaled corticosteroids for her, which of the following devices you will choose?

A. Metered-dose inhaler (MDI) alone
B. Ultrasonic nebulizer
C. MDI + spacer with mask
D. MDI + spacer without mask
E. Jet nebulizer

16. Drug-drug interactions can be defined as alteration of pharmacokinetic and/or pharmacodynamic properties of drugs when ≥2 drugs are administered to a patient at the same time.
Which of the following drug properties **MOSTLY** altered by drug interactions?

A. Absorption
B. Distribution
C. Metabolism
D. Elimination
E. Receptor binding

17. Drug interactions due to physicochemical incompatibility of 2 medications when combined are potentially dangerous. In this regard, which of the following

cephalosporin antibiotics should be avoided in neonates receiving IV calcium-containing products?

A. Cefixime
B. Ceftriaxone
C. Cefotaxime
D. Cefuroxime
E. Cefaclor

18. Which of the following drug-drug interactions is of antagonism type?

A. Fentanyl and midazolam
B. Penicillins and aminoglycosides
C. Paracetamol and codeine
D. Naloxone and pethidine
E. Penicillins and probenecid

19. Which of the following drugs can increase the hemorrhagic risk in patients taking warfarin by displacement of warfarin from plasma protein binding?

A. Ibuprofen
B. Phenobarbital
C. Erythromycin
D. Cimetidine
E. Ketoconazole

20. Regarding the safety of dietary supplements, which of the following agents can exacerbate gastroesophageal reflux?

A. Ginger
B. Magnesium
C. Peppermint
D. Chamomile
E. Zinc

21. Specific adverse drug reactions (ADRs) occur at a much greater frequency in infants and children than in adults. In this regard, which of the following ADRs is specific for infants <2 years of age?

A. Aspirin-associated Reye syndrome
B. Cefaclor-associated serum sickness-like reactions
C. Lamotrigine induced cutaneous toxicity.
D. Valproic acid-induced hepatotoxicity

E. Tetracycline induced dental discoloration

22. Which of the following is the **BEST** approach that can be used to facilitate therapeutic compliance and adherence in pediatric patients of all age groups?

A. Using positive reinforcement
B. Improving drug formulation quality coupled with positive reinforcement
C. Ensuring that single caregiver is responsible for the medication of a single child
D. Repetitive education of caregivers and older children
E. Increasing caregiver vigilance and repetitive education with positive reinforcement

23. A 3-year-old boy presents with fever, mucopurulent nasal discharge, productive cough, and wheezing for the last 48 hours without history of chronic respiratory problem. The boy is a candidate for elective surgery to correct his left inguinal hernia and hydrocele next week.
What would be your decision regarding safety of general anesthesia?

A. Treating chest infection and advice for using IV hydrocortisone before induction of GA
B. Postpone the surgery for 6 wks
C. Adding prednisolone 1mg/kg for 5 days to the treatment regime of chest infection
D. Postponed the surgery till symptoms subside
E. Treating chest infection and reevaluate the patient after 1 wk for fitness to GA

24. A newborn presents with imperforated anus 36 hours after normal vaginal delivery at home, following stabilization the pediatric surgeon was consulted who prepared him for surgery.
Of the following, the **MOST** critical preanesthetic medication is

A. vitamin K
B. prophylactic antibiotics
C. atropine
D. H_2-blockers
E. benzodiazepines

25. A 4-year-old girl presents to the outpatient clinic with frequent enuresis, temper tantrums, and nighttime crying as well as fear of strangers 3 days after her skin graft surgery to release contractures affecting her right elbow. She was

exposed to burn before 2 years and now undergoing scheduled operations to improve her joints mobility. A diagnosis of postoperative behavioral changes was made.
What is the **BEST** measure to be taken in order to reduce postoperative behavioral changes in the next surgery?

A. Using IV midazolam as preanesthetic medication
B. Parental presence during induction
C. Preoperative psychological preparation program
D. Parental presence during induction and using oral midazolam
E. Preoperative psychological preparation program and using oral midazolam

26. Regarding preoperative fasting guidelines, breast milk may be given to infants up to

A. 2 hours before surgery
B. 4 hours before surgery
C. 6 hours before surgery
D. 8 hours before surgery
E. 10 hours before surgery

27. Benzodiazepines are sedative-anxiolytic agents which are very effective as premedications, but they can severely depress respiration, especially when administered with

A. propofol
B. barbiturates
C. opioids
D. chloral hydrate
E. ketamine

28. A 12-year-old boy presents to the E/D with severe abdominal pain mostly localized over a sutured midline incision, he underwent laparotomy before 18 hours for removal of liver hydatid cyst which necessitated the use of skeletal muscle relaxant. The boy was received the usual dose of morphine after surgery which controlled his pain initially, in addition to abdominal pain he is also suffering from moderate headache and generalized pain in all extremities.
Of the following, the skeletal muscles relaxant responsible for his condition is

A. atracurium
B. tubocurarine

C. pancuronium
D. succinylcholine
E. vecuronium

29. Which of the following general anesthetic agents represent the gold standard for providing analgesia?
A. Nitrous oxide
B. Midazolam
C. Ketamine
D. Thiopental
E. Fentanyl

30. A 5-year-old boy presents with severe pain, swelling, and obvious deformity in the left arm following fall on the ground while he is playing. X-ray showed supracondylar fracture of the humorous, he received opioid analgesia as a part of his initial management, after 30 minutes, the boy developed swelling in his lips, generalized urticarial rash, and inching all over the body.
Of the following, the **MOST** likely blamed opioid is
A. morphine
B. codeine
C. pethidine
D. methadone
E. tramadol

31. Malignant hyperthermia is critical condition that may develop during general anesthesia, particularly when inhalational agents combined with certain skeletal muscle relaxants.
Which combination has greater risk of triggering malignant hyperthermia in susceptible individuals?
A. Nitrous oxide plus vecuronium
B. Halothane plus vecuronium
C. Halothane plus succinylcholine
D. Nitrous oxide plus succinylcholine
E. Sevoflurane plus vecuronium

32. Which of the following is the **MOST** commonly used inhalational agent for induction and maintenance of general anesthesia in children?
A. Nitrous oxide

B. Isoflurane
C. Halothane
D. Sevoflurane
E. Desflurane

33. For children with vascular access, IV induction should be routine during general anesthesia.
Of the following, the **MOST** commonly administered IV induction agent is
A. Fentanyl
B. Propofol
C. Etomidate
D. Ketamine
E. Sodium thiopental

34. A 5-year-old asthmatic boy presents to the E/D with severe shortness of breath, agitation, interrupted speech, and cyanosis. On auscultation the chest was silent, SpO_2 65%. He was nebulized 5 times at home with alternating salbutamol and ipratropium every 20 minutes and received oral dose of prednisolone (2mg/kg) in addition to his regular medications. In the E/D he was kept on continues nebulized salbutamol driven by oxygen and received IV hydrocortisone, aminophylline and magnesium sulfate without significant clinical improvement that necessitate ICU admission.
Of the following, the **MOST** appropriate drug for sedating this patient before intubation is
A. diazepam
B. etomidate
C. ketamine
D. fentanyl
E. midazolam

35. Which of the following is the **MOST** commonly used benzodiazepine in pediatric anesthesia?
A. Lorazepam
B. Clonazepam
C. Diazepam
D. Midazolam
E. Oxazepam

36. During recovery from general anesthesia, opioid reversal with naloxone may increase the postoperative

A. pain
B. nausea and vomiting
C. hypoventilation
D. hypotension
E. hypothermia

37. Which of the following dietary supplements is recommended to prevent antibiotic-associated diarrhea?

A. Zinc
B. Probiotics
C. Peppermint
D. Chamomile
E. Omega-3 fatty acids

38. Which of the following is the **MOST** likely complication of spinal anesthesia?

A. Infection
B. Epidural hematoma
C. Nausea and vomiting
D. Pruritus
E. Cephalad spread of blockade

39. Regarding house-hold products, ingestion of which of the following cleaning agents is considered as caustic ingestion?

A. Soap solution
B. Electric dishwasher detergent
C. Single-use detergent sac
D. Powdered detergent
E. Glass cleaner

40. A 14-month-old girl frequently admitted to the hospital because of recurrent chest infections. She always expresses fears from hospital environment and resists physical examination since her first admission. Her CXR shows slightly enlarged cardiac shadow; accordingly she was referred for cardiac assessment.
Of the following, the **MOST** convenient sedating agent to perform echocardiography for her is

A. chloral hydrate
B. morphine
C. methohexital
D. nitrous oxide
E. phenobarbital

41. In the presence of an anesthesiologist and/or specially trained pediatrician, which of the following drugs is the **MOST** effective sedating agent for painful procedures like bone marrow aspiration in children?

A. Midazolam
B. Propofol
C. Diazepam
D. Fentanyl
E. Dexmedetomidine

42. Regarding pain categories, a sharp, pulsatile, and well localized pain is characteristic for

A. deep somatic pain
B. visceral pain
C. superficial somatic pain
D. dysfunctional neuropathic pain
E. inflammatory neuropathic pain

43. Which of the following pain measurement tools is suitable for infants and nonverbal children?

A. Visual analog scale
B. Likert scale
C. Wong-Baker face scale
D. FLACC scale
E. FACES-R scale

44. In the treatment of pediatric pain, which of the following NSAIDs has extensive pediatric safety experience?

A. Aspirin
B. Ibuprofen
C. Diclofenac sodium
D. Ketorolac
E. Naproxen

45. Using sucrose solution via pacifier or gloved finger before starting procedure; can provide significant analgesia that may last up to

A. 2 minutes
B. 4 minutes
C. 6 minutes
D. 8 minutes
E. 10 minutes

46. Which of the following NSAIDs is useful in treating pediatric patients with moderate to severe acute somatic pain?

A. Aspirin
B. Ibuprofen
C. Diclofenac sodium
D. Naproxen
E. Ketorolac

47. Which of the following is the preferred antidote for cyanide poisoning?

A. amyl nitrite
B. sodium nitrite
C. sodium thiosulfate
D. pyridoxine
E. hydroxocobalamin

48. A 6-year-old boy, known case of sickle cell anemia, presents to the E/D with severe intractable back pain for the last 6 hours.
Of the following, the drug of **CHOICE** to control his pain is

A. diclofenac sodium
B. ketorolac
C. morphine
D. naproxen
E. gabapentin

49. Which of the following drugs is useful in treating pediatric patients with neuropathic pain?

A. Diclofenac sodium
B. Codeine
C. Acetaminophen
D. Ketorolac

E. Amitriptyline

50. In the treatment of chronic severe pain with opioid, what is the most common, troubling, but treatable side effect?

A. Nausea and vomiting
B. Respiratory depression
C. Constipation
D. Pruritus
E. Addiction

51. A 5-year-old boy, previously healthy, exposed to trauma while playing football with his peers. On ER arrival, a 3 cm long lacerated wound is observed in this left arm.
Of the following, the **MOST** appropriate measure to control his pain during suturing is

A. IV Morphine
B. IV acetaminophen
C. topical lidocaine
D. topical phenylephrine-tetracaine
E. IM tramadol

52. The diagnosis of carbon monoxide poisoning is confirmed by

A. determination of HbCO level by CO-oximetry
B. determination of O2 saturation by pulse-oximetry
C. measuring partial pressure of O2 by blood gas analysis
D. measuring partial pressure of CO2 by blood gas analysis
E. applying oxygen-hemoglobin dissociation curve

53. A 3-year-old boy presents to the E/D with agitation and delirium for the last 2 hours. He was completely healthy before accompanying his older brother in a forest trip. Physical examination shows plethoric face, clear chest, dilated pupils, and dry hot skin. HR 150 bpm, RR 24 bpm, and temp 38 ˚C. The boy was witnessed holding a wild plant during the trip.
Of the following, the **MOST** likely plant poisoning is

A. foxglove
B. rosary pea
C. datura stramonium
D. rhododendron

E. conium maculatum

54. Pharmaceutical preparations account for approximately 60% of pediatric poisoning cases.
Of the following, the **MOST** commonly reported category is

A. antibiotics
B. analgesics
C. anticonvulsants
D. antihypertensives
E. antidepressants

55. In children <6 years old, the majority of poisoning exposure can be managed through

A. outpatient visit
B. emergency department visit
C. hospital admission
D. phone call to the poison control center
E. self-directed fist aids by caregivers

56. Which of the following agents is potentially toxic to young children (<6 years) in small doses?

A. Aspirin
B. Soap solution
C. Diphenoxylate and atropine
D. Diazepam
E. Paracetamol and chlorpheniramine

57. Some poisons are associated with characteristic odors that can be smelt in exposed children.
Of the following, the agent that produces garlic odor is

A. cyanide
B. organophosphate
C. methyl salicylate
D. hydrogen sulfide
E. methanol

58. What is the **MOST** appropriate action for potentially toxic plant ingestions present within 1-2 hours after ingestion?

A. Supportive care and symptomatic treatment
B. Decontamination with activated charcoal
C. Decontamination with whole-bowel irrigation
D. Gastric lavage and toxicologic analysis of gastric content
E. Identification of the plant and toxicologic consultation

59. A 3-year-old girl presents to the E/D with diarrhea, vomiting, and drooling from the mouth. Physical examination shows wheezy chest, bradycardia, excessive sweating, and miosis in both eyes. The girl was found alone in the house garden few minutes before the development of symptoms and poisoning is suspected.
Of the following, the **MOST** likely recognizable poison syndrome (toxidrome) is

A. sympathomimetic
B. cholinergic
C. anticholinergic
D. sympatholytic
E. serotoninergic

60. Massive ingestion of which of the following drugs is associated with high anion gap metabolic acidosis?

A. Acetazolamide
B. Ethanol
C. Salicylates
D. Barbiturates
E. Benzodiazepines

61. Regarding laboratory evaluation of poisoned children, liver transaminases, as well as an international normalized ratio (INR) should be measured for every patients has had overdose of

A. acetaminophen
B. aspirin
C. diazepam
D. ibuprofen
E. loratadine

62. For which of the following toxicants quantitative blood concentrations are integral to confirming the diagnosis and formulating a treatment plan?

A. Organophosphate insecticide

B. Diclofenac sodium
C. Kerosene
D. Desloratadine
E. Iron

63. Physostigmine is effective antidote for poisoning with
A. morphine
B. atropine
C. organophosphorous compounds
D. sulfonylureas
E. heavy metals

64. A 16-year-old girl found unconscious in her bedroom and brought to the hospital. On examination she is in deep coma with hypertonia, tachycardia and both pupils were slightly dilated. CT-scan is normal and ECG shows widening of QRS complex (150 milliseconds).
Which of the following drug overdose is highly suspected?
A. Barbiturates
B. Opioids
C. Tricyclic antidepressants
D. Benzodiazepines
E. Ethanol

65. Glucagon is the preferred antidote for poisoning with
A. acetaminophen
B. nifedipine
C. diazepam
D. atropine
E. propranolol

66. A 15-year-old girl is brought to the emergency department by her family two hours after ingestion of many ferrous sulphate tablets as a suicidal attempt. After stabilization, the **MOST** likely effective method of GI decontamination for this patient is
A. whole-bowel irrigation
B. induction of emesis by syrup of ipecac
C. administration of activated charcoal
D. administration of magnesium sulphate as cathartic agent

E. gastric lavage

67. In the management of poisoning, gastrointestinal decontamination strategies are most likely to be effective in the first or second hour after an acute ingestion, but it may be considered more than 2 hours after ingestion of

A. cholinergic medications
B. massive amounts of liquid medications
C. enteric-coated salicylates
D. sympathomimetic agents
E. lipid soluble B-blockers

68. Which of the following poisonings is effectively adsorbed by activated charcoal?

A. Ethanol
B. Cyanide
C. Lead
D. Carbamazepine
E. Lithium

69. Which of the following methods is no longer recommended in the decontamination of most cases of pediatric poisoning?

A. Whole-bowel irrigation
B. Single-dose activated charcoal
C. Gastric lavage
D. Dermal decontamination
E. Ocular decontamination

70. A 10-year-old boy underwent removal of skin lesion under local anesthesia using subcutaneous bupivacaine without adrenaline. Few minutes after surgery, he develops severe postoperative headache, confusion and perioral paresthesias. Physical examination shows hypotension and bradycardia, while ECG demonstrates widening of QRS complex.
Of the following, the **MOST** likely treatment is

A. hemodialysis
B. intralipid emulsion therapy
C. peritoneal dialysis
D. multidose activated charcoal
E. forced diuresis with alkalinization of urine

71. Alkalinization of the urine is **MOST** useful in managing poisoning with
    A. acetaminophen
    B. ibuprofen
    C. carbamazepine
    D. methotrexate
    E. amphetamine

72. Which of the following toxic agents is effectively removed by hemodialysis?
    A. Valproic acid
    B. Cyanide
    C. Lead
    D. Amitriptyline
    E. Warfarin

73. A 2-year-old girl presents to the emergency department with severe cyanosis 4 hours following ingestion of two tablets of dapsone 50 mg previously prescribed for her dermatitis herpetiformis. The recommended dose for her was 25 mg once daily. Physical examination shows HR 140 bpm, RR 30 bpm, SpO_2 85%. ECG and CXR are normal. She partially responded to O_2 therapy, but returned back to previous condition once O_2 stopped.
Of the following, the **MOST** appropriate treatment is
    A. continuous oxygen therapy and wait for spontaneous drug clearance
    B. multidose activated charcoal
    C. hemodialysis
    D. forced diuresis with alkalinization of urine
    E. exchange transfusion

74. Which of the following antidotes is useful in the diagnosis of coma due to benzodiazepines overdose?
    A. Physostigmine
    B. Dimercaprol
    C. Flumazenil
    D. Pyridoxine
    E. Naloxone.

75. In the general management of poisoning, gastrointestinal dialysis is the term that **BEST** describes which of the following methods?
    A. Whole-bowel irrigation

B. Oral administration of magnesium sulphate
C. Gastric lavage
D. Repeated administration of oral sorbitol
E. Multidose activated charcoal

76. Which of the following doses of acetaminophen can be considered as a lowest toxic after single ingestion?

A. 60 mg/kg
B. 120 mg/kg
C. 160 mg/kg
D. 220 mg/kg
E. 260 mg/kg

77. A 2-year-old boy weighing 11 kg presents to the emergency department, 12 hours after ingestion of 50 ml of acetaminophen syrup 250mg/5ml. The expected clinical manifestations at this stage is

A. anorexia, vomiting, and malaise with normal hepatic transaminases
B. liver failure with peak transaminase elevation
C. right upper quadrant abdominal pain with normal hepatic transaminases
D. anorexia, vomiting, and malaise with elevated hepatic transaminases
E. right upper quadrant abdominal pain with elevated hepatic transaminases

78. At which of the following durations after ingestion, a serum acetaminophen level should be measured if a toxic ingestion is suspected?

A. 2 hours
B. 4 hours
C. 6 hours
D. 8 hours
E. 10 hours

79. A 15-year-old boy brought by his father to the emergency department 10 hours after ingestion of 2 sheets (8 tablets each) of paracetamol 500 mg as a suicidal attempt. On examination the boy looks healthy with normal vital signs. Of the following, the **MOST** appropriate management is

A. measurement of acetaminophen level and applying Rumack-Matthew nomogram
B. forced diuresis with alkalinization of urine

C. emergency hemodialysis
D. starting empiric N-acetylcysteine
E. observation for 24 hours followed by assessment of liver function

80. A 3-year-old girl has had high grade fever associated with URTI for the past 3 days. She received several doses of oral and rectal acetaminophen, collectively about 80mg/kg/day for the last 2 days. You have been counseled about the safety of the antipyretic regimen.
Of the following, the **MOST** appropriate advice is
A. measurement of acetaminophen level
B. no intervention is required
C. starting empiric N-acetylcysteine
D. requesting liver ultrasound
E. requesting AST, ALT, and INR

81. A 2-year-old boy presents to E/D with history of ingestion of large number of unknown tablets before 3 hours. He vomits twice at home. Physical examination shows altered mental state, HR 140 bpm, RR 40 bpm, temp 38 C°. Random blood sugar 250 mg/dL and arterial blood gas reveals a primary respiratory alkalosis. During his stay in the E/D, the kid developed seizures twice that necessitate diazepam administration.
Of the following, the **MOST** likely culprit medication is
A. acetaminophen
B. carbamazepine
C. salicylate
D. tricyclic antidepressant
E. metformin

82. In acute salicylism, salicylate pills (especially enteric-coated forms) occasionally form bezoars, which should be suspected if
A. the patient develops seizures
B. serum salicylate concentrations continue to rise
C. the patient develops abdominal cramps
D. respiratory alkalosis shifted to high anion gap metabolic acidosis
E. hyperglycemia shifted to hypoglycemia

83. What is the primary mode of therapy for salicylate toxicity?
A. Multidose activated charcoal

B. Hemodialysis
C. Use of n-acetylcysteine
D. Urinary alkalinization
E. Rehydration with ringer solution

84. A 2-year-old boy weighing 12 kg presents to the emergency department 3 hours after ingestion of about the half of a 100 ml bottle containing Ibuprofen suspension (100 mg/5ml). He is asymptomatic with normal vital signs.
Of the following, the **MOST** appropriate measure is
A. administration of activated charcoal
B. performing gastric lavage
C. urinary alkalinization
D. measurement of ibuprofen level
E. supportive care with observation

85. A 3-year-old girl presents to the emergency department, 2 hours after ingestion of one tablet of a medication prescribed for her brother to control his drug craving. She is in coma with pupils reacting to light but moderately constricted, HR 70 bpm, RR 8 bpm, temp 35 C°, SpO2
89 %.
Of the following, the **MOST** appropriate treatment is
A. Administration of activated charcoal
B. IV flumazenil and treating according to response
C. Starting IV naloxone therapy
D. Supportive care and request toxicological screen
E. Hemodialysis

86. Which of the following is considered to be the **MOST** toxic β-blocker in overdose situations?
A. Propranolol
B. Atenolol
C. Bisoprolol
D. Carvedilol
E. Metoprolol

87. A 2-year-old boy presents to E/D with history of ingestion of large number of unknown tablets before 6 hours. Physical examination shows altered mental state, HR 60 bpm, Bp 65/45, RR 24 bpm, temp 37.2 °C, SpO2 96%. Chest

auscultation reveals diffuse expiratory rhonchi. Random blood sugar is 50 mg/dL and blood gas is normal.
Of the following, the **MOST** likely culprit medication is

A. oral antidiabetic
B. tricyclic antidepressant
C. calcium channel blocker
D. β-blocker
E. α- blocker

88. Which of the following is the **MOST** dangerous calcium channel blocker in overdose situations?

A. Nimodipine
B. Diltiazem
C. Nifedipine
D. Amlodipine
E. Nicardipine

89. Which of the following is considered as the antidote of **CHOICE** in the treatment of calcium channel blockers overdose?

A. Calcium salts
B. Dextrose
C. Adrenaline
D. Lipid emulsion therapy
E. Insulin

90. Which of the following is the antidote of **CHOICE** for both methanol and ethylene glycol poisoning?

A. Ethanol
B. Fomepizole
C. Sodium bicarbonate
D. Folic acid
E. Pyridoxine

91. A 4-year-old girl presents to the E/D with nausea and vomiting for the last 6 hours. She is a known case of heart failure on digoxin therapy. Before one day the girl was treated with a macrolide antibiotic for tonsillitis, because she is allergic to penicillins. On examination she looks pale, lethargic, HR 50 bpm, with irregular pulse. Investigations reveal serum digoxin level 16 ng/mL, serum K^+ 5.6

mEq/L, creatinine 0.7 mg/dL, and ECG shows widening of QRS complex (110 milliseconds).
Of the following, the **BEST** predictor of poor outcome is

A. QRS widening
B. serum digoxin level
C. serum potassium
D. serum creatinine
E. heart rate

92. What is the expected time for development of GIT strictures following massive ingestion of iron in children?

A. 5-10 days
B. 2-3 weeks
C. 4-6 weeks
D. 7-8 weeks
E. 9-10 weeks

93. A 2-year-old boy presents to the E/D with altered mental state for the last 6 hours. His mother noticed that her pills organizer is empty for the next two days; she is a diabetic patient on oral antidiabetic therapy (glibenclamide 5mg tablets twice daily). On examination the boy looks drowsy with profuse sweating, HR 160 bpm, RBS 35 mg/dL, and serum electrolytes are normal. The patient was treated with two bolus doses of IV dextrose that raises his blood sugar to 50 mg/dL and partially improved his mental state.
Of the following, the **MOST** appropriate next step is

A. hemodialysis
B. continuous dextrose infusion
C. administration of glucagon
D. administration of a third bolus dose of IV dextrose
E. administration of octreotide

94. What is the expected toxicity following large overdose of metformin?

A. hypoglycemia
B. nephrotoxicity
C. neurotoxicity
D. lactic acidosis
E. hepatotoxicity

95. A 14-year-old boy found unconscious in his bedroom and brought to the hospital. Before six months, he survived a suicidal attempt and kept on antidepressant medications (imipramine 25 mg at bed time). The medication container was found empty beside the victim. On examination he is in deep coma with hypertonia, mydriasis, HR 130 bpm, Bp 70/50, temp 37.8 °C. ECG shows widening of QRS complex (150 milliseconds) with multiple premature ventricular contractions.
Of the following, the **MOST** appropriate initial treatment is

A. esmolol
B. sodium bicarbonate
C. hypertonic saline (3%)
D. lipid emulsion therapy
E. amiodarone

96. A 10-year-old boy presents to the E/D with visual disturbances and rapid deep breathing 12 hours after accidental ingestion of a paint remover stored in a carbonated beverages bottle. Visual disturbances include cloudy vision, decreased acuity, and the "feeling of being in a snowstorm". Ophthalmic examination reveals dilated pupils, retinal edema, and optic disc hyperemia.
Of the following, the **MOST** likely ingested poison is

A. acetone
B. ethanol
C. methanol
D. ethylene glycol
E. benzene

97. A 2-year-old boy presents to the E/D one hour after ingestion of unknown quantity of sewage cleaner. The boy has excessive drooling and he refuse to swallow anything. Examination of the chest is normal.
Of the following, the **MOST** appropriate management is

A. performing gastric lavage
B. administration of activated charcoal
C. nothing by mouth and proton pump inhibitor therapy
D. corticosteroids therapy and prophylactic antibiotics
E. immediate endoscopy

98. Which of the following pesticides is **MOST** commonly used in home?

A. Pyrethrins

B. Organophosphates
C. Carbamates
D. Organochlorines
E. Coumarins

99. A 2-year-old boy presents to the E/D with coughing and drooling 3 hours following ingestion of unknown quantity of kerosene. On examination the boy is drowsy with odor of kerosene clearly smelt in his breath, HR 100 bpm, RR 35 bpm, temp 37.5 ˚C, SpO2 95%, with clear chest. The boy vomited twice at home and his cloths were found contaminated with kerosene.
Of the following, the **MOST** appropriate management is

A. supportive care and observation for 24 hours
B. start prophylactic antibiotics
C. start systemic corticosteroids
D. immediate CXR and treat accordingly
E. supportive care and request CXR after 6 hours

100. A 2-year-old girl presents to the E/D one hour after accidental ingestion of kerosene which was stored in a domestic container. She is asymptomatic with odor of kerosene clearly smelt on her cloths, otherwise normal physical examination.
Of the following, the **MOST** appropriate decontamination method is

A. Emesis
B. Gastric lavage
C. Activated charcoal
D. removing contaminated clothing
E. whole-bowel irrigation

101. A 5-year-old boy presents to the E/D with difficulty in breathing few minutes after spraying an insecticide in the garden by his father. He is conscious with oral frothy discharge, diffuse expiratory wheeze, excessive sweating, and slightly constricted pupils.
Of the following, the **MOST** appropriate way for decontaminating the victim is

A. activated charcoal
B. gastric lavage
C. whole-bowel irrigation
D. washing all exposed skin with soap and water
E. induction of emesis

102. What is the **MOST** appropriate measure to establish a primary diagnosis of organophosphorous or carbamate poisoning?

A. Recommending history and physical exam findings
B. Monitoring signs and symptoms after administration of atropine
C. Qualitative assay of the pesticide in the blood
D. Measuring pseudocholinesterase activity level
E. Measuring RBC cholinesterase activity level

103. A 14-year-old boy presents to the E/D with recurrent seizures after ingesting unknown quantity of an insecticide as suicidal attempt. He is in deep coma and shallow breathing; physical examination shows drooling from the mouth, wheezy chest, excessive sweating, miosis in both eyes, generalized fasciculation, HR 50 bpm, RR 12 bpm, and SpO2 80% with oxygen. After suction of oral secretions rapid sequence intubation is followed.
Which of the following skeletal muscle relaxant should be **AVOIDED** during intubation?

A. Rocuronium
B. Tubocurarine
C. Pancuronium
D. Vecuronium
E. Succinylcholine

104. Which of the following antidotes is **SPECIFIC** for organophosphates poisoning?

A. Atropine
B. Activated charcoal
C. Pralidoxime
D. Clonidine
E. Glycopyrrolate

105. A 15-year-old boy presents to the E/D with recurrent seizures after ingesting an insecticide available at home as suicidal attempt. He is confused with generalized choreoathetosis and tremor. Both eyes are normal on examination, clear chest, HR 80 bpm, RR 18 bpm, Bp 100/60 and SpO2 95%. A generalized urticarial rash with excessive pruritus is also observed.
Of the following, the **MOST** likely culprit insecticides is

A. organophosphates
B. pyrethroids

C. carbamates
D. organochlorines
E. ryanoids

106. Which of the following quantities of kerosene is considered as a lowest toxic after aspiration?

A. 1 mL
B. 2 mL
C. 3 mL
D. 4 mL
E. 5 mL

Chapter 7

Pediatric Drug Therapy Answers

HAIDAR A. N. ABOOD

1.(C) The finding that drug responses can be influenced by the patient's genetic profile has offered great hope for realizing individualized pharmacotherapy. In this regard, pharmacogenetics classically is defined as the study or clinical testing of genetic variations that give rise to interindividual response to drugs. Pharmacogenomics defined as the broader application of genome-wide technologies and strategies to identify both disease processes that represent new targets for drug development and factors predictive of efficacy and risk of adverse drug reaction.

2.(B) Genetic polymorphism in butyrylcholinesterase enzyme will reduce the ability of the body to hydrolyze succinylcholine which lead to prolonged respiratory muscles paralysis after administration.

3.(D) Phase I reactions introduce or reveal (through oxidation, reduction, or hydrolysis) a functional group within the substrate drug molecule that serves as a site for a phase II conjugation reaction. Phase II reactions involve conjugation with endogenous substrates, such as acetate, glucuronic acid, glutathione, glycine, and sulfate. These reactions further increase the polarity of an intermediate metabolite, make the compound more water soluble, and thereby enhance its renal excretion.

4.(A) The major consequence of pharmacogenetic polymorphisms in drug metabolizing enzymes is concentration-dependent toxicity caused by impaired drug clearance. In certain cases, reduced conversion of prodrug to therapeutically active compounds is also of clinical importance.

5.(E) Thiopurine S-methyltransferase (TPMT) is a cytosolic enzyme that catalyses the S-methylation of aromatic and heterocyclic sulfur-containing compounds, such as 6-mercaptopurine (6MP), azathioprine, and 6-thioguanine, used in the treatment of acute lymphoblastic leukemia (ALL). To exert its cytotoxic effects, 6MP requires metabolism to thioguanine nucleotides by a multistep process. TPMT prevents thioguanine nucleotide production by methylating 6MP. The relatively few patients with low to absent TPMT activity are at increased risk for severe myelosuppression if treated with routine doses of thiopurines.

6.(B) Salbutamol acts as agonist drug on β2 adrenergic receptors, while other bronchodilator drugs are not (ipratropium and tiotropium are muscarinic antagonist, while theophylline and aminophylline are phosphodiesterase inhibitors)

7.(A) Reduced muscular blood flow in the 1st few days of life, the relative inefficiency of muscular contractions (useful in dispersing an IM drug dose), and an increased percentage of water per unit of muscle mass may delay the rate and extent of drugs given intramuscularly to the neonate. Muscular blood flow increases into infancy, and thus the bioavailability of drugs given by the IM route is comparable to that seen in children and adolescents.

8.(E) Newborns have a much higher proportion of body mass in the form of water (total body water (TBW) approximately 75%) than older infants and children. In addition, the percentage of extracellular water (ECW) changes (decreases) from the newborn stage (approximately 45%) into adulthood (20–30%). In fact, the increase of TBW in the neonate is attributable to ECW. In contrast ,the percentage of intracellular water (ICW) as a function of body mass remains stable from the 1st months of life through adulthood. Water spaces are exemplified by drugs such as the aminoglycoside antibiotics; compounds that distribute predominantly throughout the extracellular fluid space and have a higher VD (0.4-0.7 L/kg) in neonates and infants than in adults (0.2-0.3 L/kg).

9.(A) Mucosal permeability (rectal and buccal) in the neonate is increased and thus result in enhanced absorption by this route.

10.(B) The kidney is the primary organ responsible for the elimination of drugs and their metabolites. The development of renal function begins during early fetal development and is complete by early childhood

11.(B) Altered renal drug clearance in the newborn and infants result in the different dosing recommendations seen in pediatrics. The aminoglycoside antibiotic gentamicin provides an illustrative example. In adolescents and young adults with normal values for GFR (85-130 mL/min/1.73 m2), the recommended dosing interval for gentamicin is 8 hours. In young children who may have a GFR >130 mL/min/1.73 m2, a gentamicin dosing interval of every 6 hours may be necessary in selected patients who have serious infections that require maintaining steady-state peak and trough plasma concentrations near the upper boundary of the recommended therapeutic range. In contrast, to maintain “therapeutic” gentamicin plasma concentrations in neonates during the 1st few weeks of life, a dosing interval of 18-24 hours is required.

12.(C) Therapeutic drug monitoring (TDM) largely entails measurement of drug concentrations in plasma at some point during a drug’s dosing interval. These

levels are then compared with those that are “desired” for a given drug based on published information and used to adjust the dose/dosing regimen. TDM approach is used for many drugs which are therapeutically monitored in the clinical setting (e.g., aminoglycoside antibiotics, vancomycin, phenytoin, phenobarbital, cyclosporine, tacrolimus, mycophenolate mofetil, selected antiretroviral drugs, and acyclovir).

13.(D) Administration of liquid medications for children can be associated with risk if the device for administering the medication is not appropriate (e.g., use of a kitchen teaspoon versus 5.0 mL dosing spoon) or is used improperly to insure the drug dose. The low cost and convenience of hypodermic syringes has prompted many physicians and pharmacists to dispense them with liquid medications in order to improve accuracy. While this approach would seemingly be associated with greater accuracy in dosing, parents/caregivers can have difficulty in reading the graduations on a syringe, and the plastic caps on the plunger of syringes can produce a choking hazard for infants and young children.

14.(E) The pulmonary surface area in pediatric patients of all ages is a very effective, easily traversable barrier for drug absorption. Rate-limiting factors for pulmonary drug absorption include physicochemical factors associated with the drug and delivery system (e.g., particle size, diffusion coefficient, chemical stability of drug molecule in the lung) and physical factors that influence intrapulmonary drug disposition (e.g., active versus passive drug delivery to tracheobronchial tree, respiratory minute volume ,internal airway diameter), many of which are developmentally determined.

15.(C) For drugs formulated for delivery using a metered-dose inhaler, developmental factors (e.g., incoordination of device actuation with inhalation, inability to follow instructions for clearing of airway, passive inhalation with actuation of delivery device) either prevent their use (as in infants and small children) or limit the bioavailability of the drug to be administered. In these instances, specific devices (e.g., spacer chambers with mask for children less than 5 years or without mask for older children in case of long-term use) and methods of delivery (e.g., continuous aerosolization by mask for short-term use) can be used to improve the efficiency of drug delivery and thus drug efficacy.

16.(C) Drug-drug interactions largely occur at the level of drug metabolism but may occur at the level of drug absorption, distribution, elimination and receptor binding.

17.(B) Ceftriaxone should be avoided in infants <28 days of age if they are receiving or expected to receive IV calcium-containing products, due to reports of neonatal deaths resulting from crystalline deposits in the lungs and kidneys.
18.(D) Naloxone is competitive antagonist that blocks all opioid receptors.
19.(A) Drug interactions largely occur at the level of drug metabolism (e.g., phenobarbital is CYP450 enzyme inducer that increase the metabolism of warfarin and increases thrombotic risk, while erythromycin, cimetidine and ketoconazole are CYP450 enzyme inhibitors that decrease the metabolism of warfarin and increase patient hemorrhagic risk) but may occur at the level of drug distribution (e.g., displacement of warfarin plasma protein binding by ibuprofen with consequent increased hemorrhagic risk).
20.(C) Even when a dietary supplement is safe when used correctly, it can cause mild or severe toxicity when used incorrectly. For example, although peppermint is a commonly used and usually benign gastrointestinal spasmolytic included in after-dinner mints, it can exacerbate gastroesophageal reflux.
21.(D) Valproic acid causes hepatotoxicity in infants <2 years of age. For other drugs, ADRs can occur in older children.
22.(E) The only approach that can be used to facilitate therapeutic compliance and adherence in the pediatric patient is the combination of vigilance (on behalf of all caregivers) and repetitive education coupled with positive reinforcement.
23.(B) Respiratory illnesses associated with fever, mucopurulent nasal discharge, productive cough, or lower respiratory symptoms (wheezing, rales) are associated with increased airway reactivity and anesthetic complications for up to 6 wk thereafter. There may also be increased risk of perioperative laryngospasm and bronchospasm, reduced mucociliary clearance, atelectasis, and hypoxemia. It is recommended that elective procedures requiring general anesthesia be postponed 4-6 wk in this setting.
24.(A) In neonates, assurance of vitamin K prophylaxis and adequate coagulation status is critical before any major surgery.
25.(E) Surgery and painful medical procedures are psychologically traumatic events for children and families. Many children undergoing surgery have new-onset negative behavioral changes postoperatively. These maladaptive behavioral responses may include enuresis, separation anxiety, temper tantrums, and nighttime crying, as well as fear of strangers, doctors, and hospitals. Risk factors for postoperative behavioral changes include preoperative anxiety and emergence excitation. Need for recurrent procedures is another risk factor. Preoperative psychological preparation programs decrease the incidence of postoperative behavioral changes. Parental presence

during induction (PPI) has not been shown to improve postoperative behavior. Oral midazolam (0.5 mg/kg) may decrease negative behavioral changes after surgery. Midazolam has the benefit of providing rapid-onset anxiolysis and amnesia.

26.(B) Preoperative fasting guidelines have been developed to reduce the incidence of aspiration of gastric contents during anesthesia. Clear, sweet liquids (e.g., Pedialyte, 5% dextrose in water) facilitate gastric emptying, prevent hypoglycemia, and may be given up to 2 hours before anesthesia. Breast milk may be given to infants up to 4 hours before surgery. Infant formula, fruit juices and gelatin may be given up to 6 hours before surgery. Solids should be avoided for 6-8 hours before surgery.

27.(C) Benzodiazepines are effective premedication drugs with good safety profile, but can depress respiration when administered with opioids (known potent respiratory depressant agents). Combination with barbiturates can also increase the incidence of respiratory depression but to lesser extent than that observed with opioids.

28.(D) Succinylcholine is a depolarizing neuromuscular blocking agent that produces muscle fasciculations which may be associated with immediate increases in intracranial and intraocular pressures as well as postoperative muscle pain.

29.(E) Opioids represent the gold standard for providing analgesia.

30.(A) Among opioids, morphine and, to a lesser extent, hydromorphone may cause histamine release after administration.

31.(C) Succinylcholine is a depolarizing skeletal muscle relaxant agent that can trigger malignant hyperthermia in susceptible individuals, this risk is greatly increased when succinylcholine combined with volatile general anesthetic agents like halothane.

32.(D) Sevoflurane is the most commonly used inhalational agent for induction and maintenance of general anesthesia in children.

33.(B) Propofol is the most commonly administered IV induction agent.

34.(C) Ketamine is a bronchodilator and is a useful agent for sedating asthmatic patients in the ICU. Etomidate is associated with increased mortality when used as a sedative in the ICU (for which it is now contraindicated). Benzodiazepines and opioids can depress respiration.

35.(D) Midazolam is the most commonly used benzodiazepine in pediatric anesthesia. It is short acting and water soluble; it can be injected without pain. It is a potent hypnotic-anxiolytic-anticonvulsant and is approximately 4 times more potent than diazepam. Midazolam may be administered orally, nasally,

rectally, intravenously, or intramuscularly. Midazolam (0.10-0.15 mg/kg IV) has minimal effect on respiratory rate, heart rate, or blood pressure and provides excellent preoperative anxiolysis and amnesia. Premedication with oral midazolam (0.5-1.0 mg/kg) mixed in sweet-flavored syrup induces anxiolysis in approximately 90% of children without hemodynamic or respiratory depressant effects.

36.(A) Opioid reversal with naloxone may be indicated in rare instances when excessive opioid effect is suspected. However, naloxone reverses both the respiratory depressant and the analgesic effects of opioids. Following naloxone reversal, children may experience increased pain.

37.(B) Some uses of dietary supplements are common and recommended, such as vitamin D supplements for breastfed infants and probiotics to prevent antibiotic-associated diarrhea, whereas other uses are more controversial, such as using herbal products to treat otitis media.

38.(E) Complications of neuraxial (epidural, spinal) anesthesia include cephalad spread of blockade with respiratory depression, paralysis of respiratory muscles, and brainstem depression. Common complications of neuraxial analgesia are paresthesias and, if opioids are used, pruritus, nausea, and vomiting. Neuraxial opioid use necessitates antipruritic and antiemetic therapy. Infection and epidural hematoma are extremely rare.

39.(B) Electric dishwasher detergent especially when in the form of crystals are highly alkaline (pH >13), and exposure by ingestion can cause significant burns to the vocal cords and GI tract. Single-use detergent sacs resemble candy to many children. When bitten into, a relatively large dose of concentrated detergent is expelled under pressure onto the posterior pharynx and vocal cords. This can lead to stridor and other signs of respiratory distress. Importantly, these are not considered caustic ingestions; the pH of these products is in the neutral zone.

40.(A) Light sedation with chloral hydrate, benzodiazepines, and dexmedetomidine is often sufficient for non-painful procedures.

41.(B) For painful procedures (e.g., bone marrow aspiration), the combination of hypnosis and analgesia is required. The addition of opioids to sedation regimens increases the risk of respiratory insufficiency. Short-acting anesthetics (e.g., propofol, methohexital, remifentanil) provide effective procedural sedation, but their use carries a higher likelihood of inadvertent induction of general anesthesia. Use of these medications requires the presence of an anesthesiologist and/or specially trained, experienced, and qualified physicians.

42.(C)

Pain categories

- ✓ **Somatic** (pain resulting from injury to or inflammation of tissues)
  Characteristics:
  In skin and superficial structures: sharp, pulsatile, well localized
  In deep somatic structures: dull, aching, pulsatile, not well localized
- ✓ **Visceral** (pain resulting from injury to or inflammation of viscera)
  Characteristics:
  Aching and cramping; nonpulsatile; poorly localized (e.g., appendiceal pain perceived around umbilicus) or referred to distant locations (e.g., angina perceived in shoulder)
- ✓ **Neuropathic** (pain resulting from injury to, inflammation of, or dysfunction of the peripheral or central nervous system)
  Characteristics:
  Spontaneous; burning; lancinating or shooting; dysesthesias (pins and needles, electrical sensations); hyperalgesia (amplification of noxious stimuli); hyperpathia (widespread pain in response to a discrete noxious stimulus); allodynia (pain in response to nonpainful stimulation); pain may be perceived distal or proximal to site of injury, usually corresponding to innervation pathways (e.g., sciatica)

43.(D) FLACC (Face, Leg, Activity, Cry, and Consolability) pain scoring system may be used in preverbal, mechanically ventilated, or cognitively impaired patients. It is an acronym that includes five indicators, each scored as a 0, 1, or 2 that forms a ten-point composite scale with a range from "0" (no pain) to "10" (worst pain).

44.(B) Compared to other NSAIDs, ibuprofen has extensive pediatric safety experience.

45.(D) Using sucrose solution via pacifier or gloved finger over 2 minutes before starting procedure; can provide significant analgesia that may last up to 8 minutes and the dose may be repeated once.

46.(E) Ketorolac, an IV NSAID, is useful in treating moderate to severe acute pain.

47.(E) The FDA has approved hydroxocobalamin for use in known or suspected cyanide poisoning. This antidote reacts with cyanide to form the nontoxic cyanocobalamin (vitamin B12), which is then excreted in urine. Side effects of hydroxocobalamin include red discoloration of the skin and urine, transient hypertension, and interference with colorimetric lab assays. Hydroxocobalamin has an overall safety profile that appears superior to that of the cyanide

antidote kit (amyl nitrite, sodium nitrite, and sodium thiosulfate) and thus is the preferred antidote for cyanide poisoning.

48.(C) Opioids are administered for moderate and severe pain, such as acute postoperative pain, sickle cell crisis pain, and cancer pain.

49.(E) Antidepressants like amitriptyline and anticonvulsants like gabapentin are useful in treating pediatric patients with neuropathic pain.

50.(C) The most common, troubling, but treatable side effect of opioids use is constipation. Patients who take opioids for chronic pain for long periods predictably develop tolerance to the sedative and analgesic effects of opioids over time, but tolerance to constipation does not occur, and constipation remains a troublesome and distressing problem in almost all patients with long-term opioid administration. Stool softeners and stimulant laxatives should be administered to most patients receiving opioids for more than a few days. Osmotic and bulk laxatives are less effective, usually producing more distention and discomfort. A peripherally acting opiate μ-receptor antagonist, methylnaltrexone, promptly and effectively reverses opioid-induced constipation in patients with chronic pain who are receiving opioids daily.

51.(D) Topical local anesthetic preparations can reduce pain in diverse circumstances: suturing of lacerations, placement of peripheral IV catheters, lumbar punctures, and accessing indwelling central venous ports. The application of tetracaine, epinephrine, and cocaine results in good anesthesia for suturing wounds but should not be used on mucous membranes. Combinations of tetracaine with phenylephrine and lidocaine-epinephrine-tetracaine are equally as effective, eliminating the need to use a controlled substance (cocaine).

52.(A) CO binds to hemoglobin with an affinity >200 times that of oxygen, forming carboxyhemoglobin (HbCO). HbCO level is determined by CO-oximetry.

53.(C) Datura stramonium or jimson weed belong to belladonna alkaloids that contain scopolamine and atropine. Ingestion results in typical anticholinergic toxidrome.

54.(B) Approximately 40% of poisoning cases involve nondrug substances, such as cosmetics, personal care items, cleaning solutions, plants, and foreign bodies. Pharmaceutical preparations account for the remainder of exposures, and analgesics, topical preparations, vitamins, and antihistamines are the most commonly reported categories.

55.(D) The majority of poisoning exposures in children <6 years old can be managed without direct medical intervention beyond a call to the regional poison control center (PCC). This is because the product involved is not

inherently toxic or the quantity of the material is not sufficient to produce clinically relevant toxic effects. However, a number of substances can be highly toxic to toddlers in small doses.

56.(C) "Small dose" typically implies 1 or 2 pills or 5 mL.

57.(B) Cyanide (bitter almond odor), organophosphate (garlic), methyl salicylate (wintergreen), hydrogen sulfide (rotten eggs), and methanol (acetone).

58.(B) For potentially toxic plant ingestions, consider decontamination with activated charcoal in patients who present within 1-2 hr of ingestion; otherwise, treatment is primarily supportive and based on symptoms. The most common manifestation of toxicity after plant ingestion is GI upset, which can be managed with antiemetics and fluid and electrolyte support.

59.(B) Cholinergic toxidrome is characterized by parasympathetic manifestations (diarrhea, urination, miosis, bradycardia, bronchorrhea, bronchospasm, emesis, lacrimation, and salivation) in addition to CNS manifestations (confusion, coma, and fasciculations) and excessive sweating.

60.(C) Any patient with acidosis (low serum bicarbonate level on serum chemistry panel) must have an anion gap calculated because of the more specific differential diagnoses associated with an elevated anion gap metabolic acidosis which include (methanol, metformin, uremia, diabetic ketoacidosis, propylene glycol, isoniazid, iron, massive ibuprofen, lactic acidosis, ethylene glycol, salicylates, cellular asphyxiants (cyanide, carbon monoxide, hydrogen sulfide), alcoholic ketoacidosis, tylenol (clinical significance depends upon presence or absence of liver injury).

61.(A) Patients with a known overdose of acetaminophen should have liver transaminases (ALT, AST) assessed, as well as an international normalized ratio (INR) to evaluate hepatic injury (specific toxicity of acetaminophen).

62.(E) For select intoxications (e.g., salicylates, some anticonvulsants, acetaminophen, iron, digoxin, methanol, ethanol, lithium, ethylene glycol, theophylline, CO, lead), quantitative blood concentrations are integral to confirming the diagnosis and formulating a treatment plan. However, for most exposures, quantitative measurement is not readily available and is not likely to alter management. All intoxicant levels must be interpreted in conjunction with the history. For example, a methanol level of 20 mg/dL 1 hour after ingestion may be nontoxic, whereas a similar level 24 hours after ingestion implies a significant poisoning.

63.(B) Consider physostigmine in the treatment of anticholinergic poisoning if patient is a threat to self or others; only use if no conduction delays on ECG.

64.(C) Cardiovascular and CNS symptoms dominate the clinical presentation of tricyclic antidepressants toxicity. Symptoms typically develop within 1-2 hours of ingestion, and serious toxicity usually manifests within 6 hours of ingestion. Patients can have an extremely rapid progression from mild symptoms to life-threatening dysrhythmias. Patients often develop features of the anticholinergic toxidrome, including delirium, mydriasis, dry mucous membranes, tachycardia, hyperthermia, urinary retention, and slow GI motility. CNS toxicity can include lethargy, coma, myoclonic jerks, and seizures. Sinus tachycardia is the most common cardiovascular manifestation of toxicity; however, patients can also develop widening of the QRS complex.

65.(E) Glucagon is theoretically the preferred antidote of choice for β-blocker toxicity. Glucagon stimulates adenyl cyclase and increases levels of cAMP independent of the β-receptor. Glucagon is typically given as a bolus and, if this is effective, followed by a continuous infusion.

66.(A) Described methods of gastrointestinal decontamination include induced emesis with ipecac, gastric lavage, cathartics, activated charcoal, and whole-bowel irrigation (WBI). Of these, only activated charcoal and WBI are of potential benefit. WBI involves instilling large volumes (35 mL/kg/hr in children or 1-2 L/hr in adolescents) of a polyethylene glycol electrolyte solution to "wash out" the entire GI tract. This technique may have some success for the ingestion of SR preparations, substances not well adsorbed by charcoal (e.g., lithium, iron), transdermal patches, foreign bodies (e.g., lead chips), and drug packets.

67.(C) Gastrointestinal (GI) decontamination strategies are most likely to be effective in the 1 or 2 hours after an acute ingestion. GI absorption may be delayed after ingestion of agents that slow GI motility (anticholinergic medications, opioids), massive amounts of pills, sustained-release (SR) preparations, and agents that can form pharmacologic bezoars (e.g., enteric-coated salicylates). GI decontamination more than 2 hours after ingestion may be considered in patients who ingest toxic substances with these properties.

68.(D) Substances poorly adsorbed by activated charcoal are:

- ✓ Alcohols
- ✓ Caustics: alkalis and acids
- ✓ Cyanide
- ✓ Heavy metals (e.g., lead)
- ✓ Hydrocarbons
- ✓ Iron
- ✓ Lithium

69.(C) Although gastric lavage was used routinely for many years, objective data do not document or support clinically relevant efficacy. This is particularly true in children, in whom only small-bore tubes can be used. Lavage is time-consuming and painful and can induce bradycardia through a vagal response to tube placement. It can delay administration of more definitive treatment (activated charcoal) and under the best circumstances, only removes a fraction of gastric contents. Thus, in most clinical scenarios, the use of gastric lavage is no longer recommended.

70.(B) Intralipid emulsion (ILE) therapy is a potentially lifesaving intervention. ILE therapy sequesters fat-soluble drugs, decreasing their impact at target organs. It also enhances cardiac function by supplying an alternative energy source to a depressed myocardium and acting on calcium channels in the heart, increasing myocardial calcium and thus cardiac function. Intralipid is most effective as a reversal agent for toxicity from inadvertent intravenous (IV) injection of bupivacaine. Using the same 20% Intralipid used for total parenteral nutrition (TPN), a bolus dose of 1.5 mL/kg is given over 3 min, followed by an infusion of 0.25 mL/kg/min until recovery or until a total of 10 mL/kg has been infused.

71.(D) Urinary alkalinization enhances the elimination of drugs that are weak acids by forming charged molecules, which then become trapped in the renal tubules. Charged molecules, being polar and hydrophilic, do not easily cross cellular membranes, thus they remain in the renal tubules and are excreted. Urinary alkalinization is accomplished by a continuous infusion of sodium bicarbonate–containing IV fluids, with a goal urine pH of 7.5-8. Alkalinization of the urine is most useful in managing salicylate and methotrexate toxicity.

72.(A) Few drugs or toxins are removed by dialysis in amounts sufficient to justify the risks and difficulty of dialysis. Toxins amenable to dialysis have the following properties: low volume of distribution (<1 L/kg) with a high degree of water solubility, low molecular weight, and low degree of protein binding. Hemodialysis may be useful for toxicity from methanol, ethylene glycol, salicylates, theophylline, bromide, lithium, and valproic acid. Hemodialysis is also used to correct severe electrolyte disturbances and acid-base derangements resulting from the ingestion (e.g., severe metformin-associated lactic acidosis).

73.(B) Whereas single-dose activated charcoal is used as a method of decontamination, multidose activated charcoal (MDAC) can help to enhance the elimination of certain toxins. MDAC is recommended in managing significant

ingestions of carbamazepine, dapsone, phenobarbital, quinine, and theophylline. MDAC is typically given as 0.5 g/kg every 4-6 hr (for 4 doses).

74.(C) Flumazenil 0.2 mg IV over 30 seconds can reverse the CNS effects of benzodiazepines; if response is inadequate, repeat every 1min to 1 mg maximum.

75.(E) Multidose activated charcoal enhances elimination by 2 proposed mechanisms: interruption of enterohepatic recirculation and "GI dialysis." The concept of GI dialysis involves using the intestinal mucosa as a dialysis membrane and pulling toxins from the bloodstream back into the intraluminal space, where they are adsorbed to the charcoal.

76.(D) The single acute toxic dose of acetaminophen is generally considered to be >200 mg/kg in children and >7.5-10 g in adolescents and adults.

77.(A) Classically, 4 general stages of acetaminophen toxicity have been described:

- ✓ **Stage I:** 0.5-24 hr after ingestion; characterized by (anorexia, vomiting, malaise. Lab tests typically normal, except for acetaminophen level)
- ✓ **Stage II:** 24-48 hr after ingestion; characterized by (resolution of earlier symptoms; right upper quadrant abdominal pain and tenderness; elevated hepatic transaminases (AST >ALT) and INR)
- ✓ **Stage III:** 3-5 days after ingestion; characterized by (peak transaminase elevations; development of liver failure, multi organ-system failure, death or recovery begins)
- ✓ **Stage IV:** 4 days to 2 wk after ingestion; characterized by (resolution of liver function abnormalities, clinical recovery precedes histologic recovery)

78.(B) If a toxic ingestion is suspected, a serum acetaminophen (APAP) level should be measured 4 hr after the reported time of ingestion. If the APAP level is known and the ingestion is within 24 hr of the level being drawn, treatment decisions are based on where the level falls on the Rumack-Matthew nomogram.

79.(D) Any patient presenting close to or beyond 8 hours after an acetaminophen overdose should be empirically started on N-acetylcysteine pending laboratory results.

80.(B) Repeated administration of acetaminophen at supratherapeutic doses (>90 mg/kg/day for consecutive days) can lead to hepatic injury or failure in some children, especially in the setting of fever, dehydration, poor nutrition, and other conditions that serve to reduce glutathione stores.

81.(C) Early signs of acute salicylism include nausea, vomiting, diaphoresis, and tinnitus. Moderate salicylate toxicity can manifest as tachypnea and hyperpnea, tachycardia, and altered mental status. The tachycardia largely results from marked insensible losses from vomiting, tachypnea, diaphoresis, and uncoupling of oxidative phosphorylation. Signs of severe salicylate toxicity include mild hyperthermia, coma, and seizures. Classically, laboratory values from a patient poisoned with salicylates reveal a primary respiratory alkalosis and a primary, elevated anion gap metabolic acidosis. Early in the course of acute salicylism, respiratory alkalosis dominates and the patient is alkalemic. As the respiratory stimulation diminishes, the patient will move toward acidemia. Hyperglycemia (early) and hypoglycemia (late) have been described. Abnormal coagulation studies and acute kidney injury may be seen but are not common.

82.(B) Salicylate pills occasionally form bezoars (especially enteric-coated forms), which should be suspected if serum salicylate concentrations continue to rise many hours after ingestion or are persistently elevated despite appropriate management.

83.(D) The primary mode of therapy for salicylate toxicity is urinary alkalinization. Urinary alkalinization enhances the elimination of salicylates by converting salicylate to its ionized form, “trapping” it in the renal tubules, thus enhancing elimination. In addition, maintaining an alkalemic serum pH decreases CNS penetration of salicylates because charged particles are less able to cross the blood-brain barrier. Alkalinization is achieved by administration of a sodium bicarbonate infusion at approximately 2 times maintenance fluid rates. The goals of therapy include a urine pH of 7.5-8, a serum pH of 7.45-7.55, and decreasing serum salicylate levels.

84.(E) Ibuprofen, the primary NSAID used in pediatrics, is well tolerated, even in overdose. In children, acute doses of <200 mg/kg rarely cause toxicity, but ingestions of >400 mg/kg can produce more serious effects, including altered mental status and metabolic acidosis. Supportive care, including use of antiemetics and acid blockade as indicated, is the primary therapy for NSAID toxicity.

85.(C) Two specific oral opioids, buprenorphine and methadone, can cause life-threatening toxicity in toddlers with ingestion of even 1 pill. Both agents are used in managing opioid dependence. Ingestions can manifest with the classic opioid toxidrome of respiratory depression, sedation, and miosis. Signs of more severe toxicity can include bradycardia, hypotension, and hypothermia. Patients with significant respiratory depression or CNS depression should be treated with the opioid antidote naloxone. In pediatric patients who are not chronically

taking opioids, the full reversal dose of 1-2 mg should be used. In contrast, opioid dependent patients should be treated with smaller initial doses (0.04-0.4 mg), which can then be repeated as needed to achieve the desired clinical response, avoiding abrupt induction of withdrawal. Because the half-life of methadone and buprenorphine is much longer than that of naloxone, patients can require multiple doses of naloxone. These patients may benefit from a continuous infusion of naloxone.

86.(A) Because of its lipophilicity and blockade of fast sodium channels, propranolol is considered to be the most toxic member of the β-blocker class. Overdoses of water-soluble β-blockers (e.g., atenolol) are associated with milder symptoms.

87.(D) Toxicity of β-blockers typically develops within 6 hr of ingestion, although it may be delayed after ingestion of slow-release (SR) preparations. Clinically, these effects are manifested as bradycardia, hypotension, and heart block. Patients with reactive airways disease can experience bronchospasm as a result of blockade of β2-mediated bronchodilation. B2-blockers interfere with glycogenolysis and gluconeogenesis, which can sometimes lead to hypoglycemia, especially in patients with poor glycogen stores (e.g., toddlers). Lipophilic agents, including propranolol, can enter the CNS and cause altered mental status, coma, and seizures. Evaluation after β-blocker overdose should include an ECG, frequent reassessments of hemodynamic status, and blood glucose. Serum levels of β-blockers are not readily available for routine clinical use and are not useful in management of the poisoned patient.

88.(B) Calcium channel blockers are used for a variety of therapeutic indications and have the potential to cause severe toxicity. Specific agents include verapamil, diltiazem, and the dihydropyridines (e.g., amlodipine, nifedipine). Of these, diltiazem and verapamil are the most dangerous in overdose because of their higher lipophilicity and direct cardiac suppressant effects.

89.(E) Calcium salts, administered through a peripheral IV line as calcium gluconate or a central line as calcium chloride, help to overcome blocked calcium channels. High-dose insulin euglycemia therapy is considered the antidote of choice for calcium channel blockers (CCBs) toxicity. An initial bolus of 1 unit/kg of regular insulin is followed by an infusion at 0.5-1 unit/kg/hr. The main mechanism of high-dose insulin euglycemia is to improve the metabolic efficiency of a poisoned heart that is in need of carbohydrates for energy (instead of the usual free fatty acids), but has minimal circulating insulin. Blood glucose levels should be closely monitored, and supplemental glucose may be given to maintain euglycemia, although this is rarely necessary in the severely

poisoned patient. Additional therapies include judicious IV fluid boluses and vasopressors (often in very high doses). Lipid emulsion therapy is a potentially lifesaving intervention, especially for patients poisoned with the more lipid-soluble CCBs, verapamil and diltiazem.

90.(B) The classic antidote for methanol and ethylene glycol poisoning was ethanol, a preferential substrate for alcohol dehydrogenase, thus preventing the metabolism of parent compounds to toxic metabolites. Fomepizole, a potent competitive inhibitor of alcohol dehydrogenase, has almost entirely replaced ethanol because of its ease of administration, lack of CNS and metabolic effects, and overall excellent patient tolerability profile. Classic indications for fomepizole include ethylene glycol or methanol level >20 mg/dL (assuming no ethanol is present), history of potentially toxic ingestion (e.g., any intentional overdose), or history of ingestion with evidence of acidosis. Adjunctive therapy includes folate (methanol toxicity), pyridoxine (ethylene glycol toxicity), and sodium bicarbonate infusion for both (if acidemic).

91.(C) In acute digoxin toxicity, initial assessment should include an ECG, serum digoxin level, serum potassium, and kidney function tests. The serum digoxin level should be assessed at least 6 hr after ingestion and carefully interpreted in the setting of clinical symptoms, because the digoxin level alone does not entirely reflect the severity of intoxication. In acute ingestions, serum potassium is an independent marker of morbidity and mortality, with levels >5.5 mEq/L predicting poor outcomes.

92.(C) Iron toxicity is described in 5 often-overlapping stages:

- ✓ **The 1st stage**, 30 min to 6 hr after ingestion, consists of profuse vomiting and diarrhea (often bloody), abdominal pain, and significant volume losses leading to potential hypovolemic shock. Patients who do not develop GI symptoms within 6 hr of ingestion are unlikely to develop serious toxicity.
- ✓ **The 2nd stage**, 6-24 hr after ingestion, is often referred to as the "quiescent phase" since the GI symptoms typically have resolved. However, careful clinical examination can reveal subtle signs of hypoperfusion, including tachycardia, pallor, and fatigue.
- ✓ **The 3rd stage**, 12-36 hr after ingestion, patients develop multisystem organ failure, shock, hepatic and cardiac dysfunction, acute lung injury, and profound metabolic acidosis. Death usually occurs during the 3rd stage.
- ✓ **The 4th stage** (hepatic) results in fulminant liver failure and coagulopathy about 2-5 days after ingestion.

- ✓ **The 5th stage**, 4-6 wk after ingestion, is marked by formation of strictures and signs of GI obstruction.

93.(E) In patients with sulfonylurea overdose symptomatic hypoglycemia should be promptly treated with dextrose. In patients with mild symptoms, oral dextrose may be sufficient. However, patients with severe symptoms or profound hypoglycemia should be treated with a bolus of IV dextrose. Continuous dextrose infusions and repeated IV dextrose boluses should be avoided if possible, because this can stimulate further insulin release and lead to recurrent and prolonged hypoglycemia. Instead, the preferred antidote for persistent (i.e., requiring ≥2 doses of IV dextrose) sulfonylurea toxicity is octreotide; a somatostatin analog that inhibits insulin release. Octreotide is given intravenously (IV) or subcutaneously (SC), typically in doses of 1-2 µg/kg (50-100 µg in teens or adults) every 6-8 hr.

94.(D) Although metformin does not cause hypoglycemia, its association with lactic acidosis is well documented (metformin-associated lactic acidosis, MALA). This state typically arises after a large overdose in which the agent interferes with the liver's ability to clear lactic acid. Dangerously high serum lactate levels can result, leading to hemodynamic instability. Hemodialysis is usually the best option for patients with severe MALA.

95.(B) In the treatment of tricyclic antidepressants (TCAs) overdose, initial attention should be directed to supporting vital functions, including airway and ventilation as needed. TCAs will block the fast Na+ channels on the myocardial cells, slowing the upstroke of the QRS complex. Because the effect on Na+ channels is greatest within the 1st 6 hr, frequent ECGs (i.e., every 20-30 min) during this period are important. As the QRS approaches 160 msec, the risk of the patient developing monomorphic ventricular tachycardia rises to 30%. Sodium, usually in the form of sodium bicarbonate, is the antidote of choice. Indications for sodium bicarbonate include a QRS duration ≥110 msec, ventricular dysrhythmias, and hypotension. Multiple bolus doses of sodium bicarbonate, 1-2 mEq/kg each, may be needed to narrow the QRS to <110 msec. Some prefer then to place the patient on an infusion of sodium bicarbonate, but this may not be necessary if the QRS is carefully monitored after the initial doses and repeat bolus dosing is provided as needed during the 1st 6-12 hr. Hypertonic (3%) saline and/or lipid emulsion therapy may be beneficial in refractory cases.

96.(C) Drowsiness, mild inebriation, nausea, and vomiting develop early after methanol ingestion. The onset of serious effects, including profound metabolic acidosis and visual disturbances, is often delayed up to 12-24 hr as the parent

methanol undergoes metabolism to its toxic metabolites. This metabolism is further slowed if ethanol has also been ingested, since the liver will preferentially metabolize ethanol. Visual disturbances include blurred or cloudy vision, constricted visual fields, decreased acuity, and the "feeling of being in a snowstorm" and appear only after acidosis is well established. These visual defects may be reversible if treated early, but untreated can lead to permanent blindness. On examination, dilated pupils, retinal edema, and optic disc hyperemia may be noted. Initially, patients have an elevated osmolar gap, and then develop an anion gap metabolic acidosis as the parent compound is metabolized to formic acid.

97.(C) Regarding treatment of children who ingest caustic materials, emesis and lavage are contraindicated. Activated charcoal should not be used because it does not bind these agents and can predispose the patient to vomiting and subsequent aspiration. Stridor or other signs of respiratory distress should alert the provider to the need for a thorough evaluation of the airway for potential intubation or surgical airway management. Endoscopy can be performed within 12-24 hr of ingestion for prognostic and diagnostic purposes in symptomatic patients or those with suspected injury on the basis of history and known characteristics of the ingested product. Endoscopy's role is purely diagnostic. Whether the risks of the procedure are justified is debatable. Expectant management with a period of nothing by mouth (NPO) and proton pump inhibitor therapy is likely appropriate for the majority of patients without airway burns or signs of mediastinitis or peritonitis. Endoscopy is contraindicated in such patients, who instead require immediate surgical consultation. Corticosteroids or prophylactic antibiotics are not beneficial.

98.(A) Pyrethrins are derived from the chrysanthemum flower and along with pyrethroids, synthetic derivatives, are the most commonly used pesticides in the home, while the most commonly used insecticides in agriculture are organophosphates and carbamates.

99.(D) The most important manifestation of hydrocarbon toxicity is aspiration pneumonitis through inactivation of the type II pneumocytes and resulting in surfactant deficiency. Aspiration usually occurs during coughing and gagging at the time of ingestion or vomiting after the attempted ingestion of an aliphatic hydrocarbon. Transient, mild CNS depression is common after hydrocarbon ingestion or inhalation. Aspiration is characterized by coughing, which usually is the 1st clinical finding. Chest radiographs may initially be normal, but they often show abnormalities within 6 hr of exposure in patients who have aspirated. Respiratory symptoms can remain mild or progress rapidly to acute respiratory

distress syndrome (ARDS) and respiratory failure. Fever and leukocytosis are common accompanying signs in patients with pneumonitis and do not necessarily imply bacterial superinfection. If hydrocarbon induced pneumonitis develops, respiratory treatment is supportive. Neither corticosteroids nor prophylactic antibiotics have shown any clear benefit.

100.(D) Emesis and lavage are contraindicated given the risk of aspiration. Activated charcoal is not useful because it does not bind the common hydrocarbons and can also induce vomiting. The most important manifestation of kerosene toxicity is aspiration pneumonitis so; whole-bowel irrigation is not indicated. Since kerosene odor is smelt on the cloths, removing all clothing and irrigation of exposed areas is recommended.

101.(D) In the treatment of pediatric poisonings with organophosphates and carbamates insecticides basic decontamination should be performed, including washing all exposed skin with soap and water and immediately removing all exposed clothing. Activated charcoal is unlikely to be of benefit because these are liquids that are rapidly absorbed.

102.(A) Diagnosis of organophosphates and carbamates poisoning is based primarily on history and physical exam findings. Red blood cell cholinesterase and pseudocholinesterase activity levels can be measured in the laboratory. These are only helpful when compared to the patient's known baseline. As such, these assessments are typically limited to farmworkers undergoing ongoing occupational surveillance. Clinical manifestations of organophosphate and carbamate toxicity relate to ACh accumulation at peripheral nicotinic and muscarinic synapses and in the CNS. Symptoms of carbamate toxicity are usually less severe than those seen with organophosphates. A commonly used mnemonic for the symptoms of cholinergic excess at muscarinic receptors is DUMBBELS: diarrhea/defecation, urination, miosis, bronchorrhea/ bronchospasm, bradycardia, emesis, lacrimation, and salivation. Nicotinic signs and symptoms include muscle weakness, fasciculation, tremors, hypoventilation (diaphragm weakness), hypertension, tachycardia, and dysrhythmias. Severe manifestations include coma, seizures, shock, arrhythmias, and respiratory failure.

103.(E) In the treatment of pediatric poisonings with organophosphates and carbamates, the use of succinylcholine for rapid sequence intubation should be avoided because the same cholinesterase enzymes that are poisoned metabolize this neuromuscular blocking agent, leading to prolonged paralysis.

104.(C) Two antidotes are useful in treating cholinesterase inhibitor poisoning: atropine and pralidoxime. Atropine, which antagonizes the muscarinic ACh

receptor, is useful for both organophosphate and carbamate intoxication. Often, large doses of atropine must be administered by intermittent bolus or continuous infusion to control symptoms. Atropine dosing is primarily targeted to drying the respiratory secretions. Pralidoxime breaks the bond between the organophosphate and the enzyme, reactivating AChE. Pralidoxime is only effective if it is used before the bond ages and becomes permanent. Pralidoxime is not necessary for carbamate poisonings because the bond between the insecticide and the enzyme degrades spontaneously.

105.(B) Pyrethrin and pyrithroid exposures can lead to allergic reactions ranging from dermatitis to urticaria to anaphylaxis. Acute exposure can result in headache, nausea, dizziness, tremors, ataxia, choreoathetosis, loss of consciousness, and seizures. The severity of the symptoms depends on the magnitude of the exposure.

106.(A) Compounds with low viscosity and high volatility, such as mineral spirits, naphtha, kerosene, gasoline, and lamp oil, spread rapidly across surfaces and cover large areas of the lungs when aspirated. Only small quantities (<1 mL) of such chemicals need be aspirated to produce significant injury.

Chapter 8

Emergency Medicine and Critical Care Questions

AQEEL MAHDI

1. Which of the following is the **MOST** common reason for a sick-child visit?
   A. Fever
   B. Vomiting
   C. Cough
   D. Diarrhea
   E. Abdominal pain

2. A 4-year-old child presents with stridor, drooling and limitation of neck movement. He had history of upper respiratory infection 4 days ago. Examination shows a febrile child with toxic appearance and limitation of hyperextension of the neck.
Which of the following is the **MOST** likely diagnosis?
   A. Diphtheria
   B. Foreign body aspiration
   C. Bacterial tracheitis
   D. laryngotrachobronchitis
   E. Retropharyngeal abscess

3. What is the normal respiratory rate range in premature babies?
   A. 20-50
   B. 25-55
   C. 30-60
   D. 35-65
   E. 40-70

4. You are in the supermarket where a 2 -year- old girl suddenly develops cough while she was eating candy, she was conscious and mildly dyspneic.
Of the following, the **MOST** appropriate initial action is to
   A. permit to cough spontaneously
   B. open the airway

C. start chest compression
D. start back blows and chest thrusts
E. start abdominal thrusts

5. The recommended time to start intraosseous access (in patient with cardiopulmonary arrest who failed to have intravenous line) is within approximately

A. 1 minute
B. 2 minutes
C. 3 minutes
D. 4 minutes
E. 5 minutes

6. A 13-year-old boy brought to emergency unit after road traffic accident. Examination reveals muffled heart sounds, jugular venous distention, and hypotension (with narrow pulse pressure).
Of the following, the **MOST** likely diagnosis is

A. tension pneumothorax
B. mediastinal emphysema
C. major flail chest
D. massive hemothorax
E. cardiac tamponade

7. Which of the following is the **MOST** common type of shock in children worldwide?

A. Obstructive
B. Distributive
C. Hypovolemic
D. Cardiogenic
E. Anaphylactic

8. A 7-year-old girl brought to emergency unit after road traffic accident. She was dyspneic, cyanosed, distended neck veins, trachea deviated to the left, intercostal retractions, tympanic percussion of the right chest, diminished breath sounds on the right side, and pulse oximetry shows PO_2 84% at room air. of the following the **MOST** likely diagnosis is

A. tension pneumothorax
B. hydropneumothorax

C. major flail chest
D. massive hemothorax
E. cardiac tamponade

9. A 12-year-old boy presents with painful erythematous vesicular lesion in the dorsum of fingers. The patient had history of travel to snowy resort 1day before the appearance of this lesion.
Of the following, the **MOST** likely diagnosis is

A. panniculitis
B. acrocyanosis
C. Raynaud phenomenon
D. chilblains
E. cryoglobulinemia

10. A 12-year-old child presents with severe abdominal pain and vomiting which starts after receiving an elbow blow while playing football with his friends. Examination shows normal vital signs with tender distended abdomen and lap belt mark across the abdomen.
Which of the following abdominal organs is **MOST** likely injured?

A. Liver
B. Pancreas
C. Spleen
D. Bowel
E. Kidney

11. A 6-year-old boy presents with signs of shock after internal bleeding from a car accident. He received IV normal saline boluses reaching 40-60 ml/kg and still in shock.
Which of the following is the **NEXT** appropriate treatment?

A. Ringer lactate solution
B. Albumin
C. Fresh frozen plasma
D. Packed RBC
E. Ionotropic drugs

12. Which of the following represents the mildest spinal cord injury?

A. Spinal shock
B. Transient quadriparesis

C. Loss of sphincter function
D. Sensory level corresponding to the level of injury
E. Injury at the cervical level(C1-C2)

13. Which of the following arteries is **MOST** commonly cannulated for frequent arterial sampling?

A. Radial
B. Ulnar
C. Dorsalis pedis
D. Posterior tibial
E. Femoral

14. Within what time high risk wounds should be closed?

A. 2 hours
B. 4 hours
C. 6 hours
D. 12 hours
E. 24 hours

15. Which of the following intracranial pressure (ICP) is considered significant?

A. >10 mm Hg
B. >20 mmHg
C. >30 mmHg
D. >40 mmHg
E. >50 mmHg

16. A 5-year-old boy brought to causality unconscious after road traffic accident, he opens his eyes in response to voice, utters inappropriate words, he has abnormal flexion to painful stimuli.
According to above data his Glasgow Coma Scale (GCS) score is

A. 7
B. 8
C. 9
D. 10
E. 11

17. Which of the following drugs used in the treatment of shock has potent vasoconstriction with no significant effect on cardiac contractility?

A. Dopamine
B. Dobutamine
C. Epinephrine
D. Norepinephrine
E. Ephedrine

18. A simple guide for pediatric blood pressure is that the lower limit of systolic blood pressure for 1-10 year old should be
A. (Age × 2) +50 mm Hg
B. (Age × 2) +60 mm Hg
C. (Age × 2) +70 mm Hg
D. (Age × 2) +80 mm Hg
E. (Age × 2) +90 mm Hg

19. Which of the following may be the only initial clinical manifestation of shock?
A. Tachycardia
B. Delayed capillary refill
C. Decreased urine output
D. Alteration of mental status
E. Hypotension

20. Which of the following clinical findings is a late sign of shock?
A. Tachycardia
B. Tachypnea
C. Decreased urine output
D. Delayed capillary refill
E. Low blood pressure

21. A 9-month-old infant presents with fever and poor feeding. Examination shows temperature 39°C, heart rate 180 beat/minute, urine output less than 0.5 ml/kg/hour, blood gas analysis shows metabolic acidosis. Blood culture positive for gram negative bacteria.
Of the following, the **MOST** likely diagnosis is
A. systemic inflammatory response syndrome
B. sepsis
C. severe sepsis
D. septic shock

E. multiple-organ dysfunction syndrome

22. Which the following is the **MOST** common cause of syncope in the children?

A. Vasovagal syncope
B. Panic attack
C. Exercise
D. Valsalva
E. Breath-holding spells

23. A 16-year-old boy presents with syncopal attacks after head rotation during shaving his beard.
Which of the following is the **MOST** likely cause?

A. Dysautonomia
B. Exercise
C. Stroke
D. Carotid sinus sensitivity
E. Panic attack

24. Which of the following investigations must be done in all patients presenting with a first episode of syncope?

A. Blood sugar
B. CBC
C. ECG
D. Echocardiography
E. Holter monitoring

25. Which of the following medical conditions catachrestically has rapid and shallow breathing?

A. Pneumonia
B. Laryngotracheitis
C. Renal tubular acidosis
D. Encephalitis
E. Asthma

26. Which of the following diseases is characterized by grunting?

A. Mediastinal mass
B. Laryngomalacia
C. Diaphragmatic hernia

D. Foreign body aspiration
E. Bronchiolitis

27. What is the recommended initial dose of cardioversion in children with VT?
A. 2 joule/kg
B. 4 joule/kg
C. 6 joule/kg
D. 8 joule/kg
E. 10 joule/kg

28. Which of the following terms should be denoted to describe survival of the victim after drowning?
A. Nonfatal drowning
B. Near drowning
C. Secondary drowning
D. Silent drowning
E. Active drowning

29. Accessing venous line is an important step in pediatric resuscitation. Which of the following veins in the upper extremities is the easiest to access?
A. Median antecubital vein
B. Veins on the dorsum of the hand
C. Cephalic vein
D. Median vein of the forearm
E. Basilic vein

30. Which of the following small 2nd degree burn is an indication for admission?
A. Upper chest
B. Lower abdomen
C. Thigh
D. Forearm
E. Hand

31. Which of the following pediatric age groups has the highest death rate from drowning?
A. 1-4 year
B. 5-7 year
C. 8-11 year

D. 12-14 year
E. 15-19 year

32. Which of the following is the **MOST** common place of drowning deaths in infants?

A. Swimming pool
B. Bathtub
C. household bucket
D. Irrigation ditches
E. River

33. A 2-year-old boy brought to emergency department with history of repeated convulsions after drowning in a swimming pool.
Which of the following is considered the anticonvulsant drug of choice?

A. Midazolam
B. Phenobarbital
C. Diazepam
D. Phenytoin
E. Valproic acid

34. Which of the following hand burns should raise suspicion of child abuse?

A. Splash pattern burns
B. Scald burns to the side of the arm
C. Friction burns
D. Glove burns
E. Palm burns

35. Which of the following is the initial drug of choice in drowning victims with cardiopulmonary arrest?

A. Dopamine
B. Dobutamine
C. Epinephrine
D. Norepinephrine
E. Phenylephrine

36. Which of the following is considered as second tier therapy in the management of a child with severe traumatic brain injury (TBI)?

A. Hyperventilation

B. Osmolar agents (mannitol)
C. Controlled mechanical ventilation
D. Head of bed elevation
E. Analgesia

37. A 13-year-old boy presents with light headedness and tunnel vision while assuming upright position, you suspect postural tachycardia syndrome POTS.
Of the following, the **MOST** likely helpful test is
A. Blood pressure measurement
B. Electrocardiogram
C. Head-up tilt test
D. Holter monitoring
E. Echocardiogram

38. A 10-month-old infant diagnosed with septic shock, after receiving IV boluses of normal saline up to 80ml/kg he stills tachypneic, capillary refill time 4 seconds, and his urine output less than 1ml/kg/hour.
Of the following, the **MOST** appropriate next step is
A. Blood transfusion
B. IV frusemide
C. IV hydrocortisone
D. epinephrine
E. norepinephrine

39. A 10-month-old infant develops septic shock; after receiving IV boluses of normal saline up to 80ml/kg and dopamine IV infusion he is still tachypneic with capillary refill time of 4 seconds, and urine output less than 1ml/kg/hour.
Of the following, the **MOST** appropriate next step is
A. IV hydrocortisone
B. Blood transfusion
C. milrinone infusion
D. start activated protein concentrate
E. ECMO

40. Which of the following formulas estimates the proper internal diameter (ID) of endotracheal tube?
A. ID = {age (year) / 2} + 2
B. ID = {age (year) / 3} + 3

C. ID = {age (year) / 4} + 4
D. ID = {age (year) / 5} + 5
E. ID = {age (year) / 6} + 6

41. A 10-month-old infant presents with diarrhea and severe dehydration. Examination shows rapid deep breathing due to metabolic acidosis.
Which of the following formulas indicates a normal respiratory compensatory response?

A. pH 7.15, PaCO2 30 mm Hg, and [HCO3−] 10 mEq/L
B. pH 7.20, PaCO2 15 mm Hg, and [HCO3−] 7.5 mEq/L
C. pH 7.01, PaCO2 10 mm Hg, and [HCO3−] 10 mEq/L
D. pH 6.20, PaCO2 10 mm Hg, and [HCO3−] 5 mEq/L
E. pH 7.27, PaCO2 26 mm Hg, and [HCO3−] 12 mEq/L

42. Which of the following situations causes unreliable pulse oximetry results?

A. Deep sedation
B. Pulmonary edema
C. Mechanical ventilation
D. Poor perfusion
E. Large left-to-right shunts

43. Which of the following symptoms is fundamental in the diagnosis of acute mountain sickness AMS?

A. Nausea
B. Vomiting
C. Headache
D. Dizziness
E. Light headedness

44. Which of the following is the leading cause of burn in children <4 years old?

A. Scald
B. Flame
C. Steam inhalation
D. Chemical
E. Electrical

Chapter 8

Emergency Medicine and Critical Care Answers

AQEEL MAHDI

1.(A) Fever is the most common reason for a sick-child visit. Most cases of fever are the result of self-limited viral infection. However, pediatricians need to be aware of the age-dependent potential for serious bacterial infections, such as urinary tract infection (UTI), sepsis, meningitis, pneumonia, acute abdominal infection, and osteoarticular infection.

2.(E) In toxic-appearing children with stridor, the pediatrician should consider epiglottitis, bacterial tracheitis, or a rapidly expanding retropharyngeal abscess. The incidence of epiglottitis has greatly declined with the advent of the H. influenzae type b(Hib) vaccine, but it remains a possibility in the unimmunized or partially immunized patient. Children with retropharyngeal abscesses may also present with drooling and limitation of neck movement (especially hyperextension) after a recent upper respiratory infection or penetrating mouth injury.

3.(E)

4.(A) A conscious child suspected of having a partial foreign body obstruction should be permitted to cough spontaneously until coughing is no longer effective, respiratory distress and stridor increase, or the child becomes unconscious.

5.(A) If venous access is not available within approximately 1 min in a child with cardiopulmonary arrest, an IO needle should be placed in the anterior proximal tibia (with care taken to avoid traversing the epiphyseal plate).

6.(E) Cardiac tamponade, which is a form of obstructive shock, may be suspected clinically or diagnosed by focused assessment with sonography in trauma (FAST) examination or echocardiography.

Beck's triad: decreased or muffled heart sounds, jugular venous distention, and hypotension (with narrow pulse pressure) are signs of cardiac tamponade. Cardiac tamponade is best managed by thoracotomy or pericardial window, although pericardiocentesis may be necessary as a temporizing maneuver.

7.(C) Hypovolemic shock is the most common type of shock in children worldwide, usually related to fluid losses from severe diarrhea.

8.(A) Tension pneumothorax occurs when air accumulates under pressure in the pleural space. The adjacent lung is compacted, the mediastinum is pushed toward the opposite hemithorax, and the heart, great vessels, and contralateral lung are compressed or kinked. Both ventilation and cardiac output are impaired.

9.(D) Chilblain (pernio) is a form of cold injury in which erythematous, vesicular, or ulcerative lesions occur. The lesions are presumed to be of vascular or vasoconstrictive origin. They are often itchy, may be painful, and result in swelling and scabbing. The lesions are most often found on the ears, the tips of the fingers and toes, and exposed areas of the legs. The lesions last for 1-2 wk but may persist for longer.

10.(D) Specific symptoms and signs give insight into the mechanism of injury and the potential for particular injuries. Pain in the left shoulder may signify splenic trauma. A lap belt mark across the abdomen raises concern for bowel or mesentery injury. An abdominal (and pelvic) CT scan with IV contrast medium enhancement rapidly identifies structural abnormalities and is the preferred study in a stable child. Negative abdominal CT scan has been shown to have a negative predictive value (NPV) of 99.6%.

11.(D) if the patient remains in shock after boluses totaling 40-60 mL/kg of crystalloid, packed red blood cells should be transfused.

12.(B) The mildest injury to the spinal cord is transient quadriparesis evident for seconds or minutes with complete recovery in 24hr. Significant spinal cord injury in the cervical region is characterized by flaccid quadriparesis, loss of sphincter function, and a sensory level corresponding to the level of injury. An injury at the high cervical level (C1-C2) can cause respiratory arrest and death in the absence of ventilatory support.

13.(A) Arterial access is indicated when care providers need frequent blood sampling, particularly to assess adequacy of oxygenation, ventilation, or acid-base balance, and/or continuous blood pressure monitoring. The radial artery, the most commonly cannulated artery, lies on the lateral side of the anterior wrist, just medial to the styloid process of the radius.

14.(C) Although it is accepted that the time from injury to repair should be as brief as possible to minimize the risk of infection, there is no universally accepted guideline as to what length of time is too long for primary wound closure. Also, this length of time varies for different types of lacerations. A prudent recommendation is that higher-risk wounds should be closed within 6 hr at most after the injury but that some low-risk wounds (e.g., clean facial lacerations) may be closed as late as 12-24 hr.

15.(B) Significantly raised ICP (>20mmHg) can occur early after severe traumatic brain injury, but peak ICP generally is seen at 48-72 hr. Need for ICP-directed therapy may persist for longer than a week.

16.(C)

EYE OPENING (TOTAL POSSIBLE POINTS 4):

Spontaneous (4)
To voice (3)
To pain (2)
None (1)

VERBAL RESPONSE (TOTAL POSSIBLE POINTS 5):

Older Children	Infants and Young Children
Oriented (5)	Appropriate words; smiles, fixes, and follows (5)
Confused (4)	Consolable crying (4)
Inappropriate (3)	Persistently irritable (3)
Incomprehensible (2)	Restless, agitated 2
None (1)	None (1)

MOTOR RESPONSE (TOTAL POSSIBLE POINTS 6):

Obeys (6)
Localizes pain (5)
Withdraws (4)
Flexion (3)
Extension (2)
None (1)

17.(D)

Dopamine: ↑ cardiac contractility, at high doses significant peripheral vasoconstriction.
Epinephrine: ↑ heart rate and ↑ cardiac contractility, potent vasoconstrictor.
Dobutamine: ↑ cardiac contractility peripheral vasodilator.
Norepinephrine: Potent vasoconstriction no significant effect on cardiac contractility.
Phenylephrine: Potent vasoconstriction

18.(C) A simple guide for pediatric blood pressure is that the lower limit of systolic blood pressure should be ≥60 mm Hg for neonates; ≥70 mm Hg for 1 mo-1 yr olds; ≥70 mm Hg + (2 × age) for 1-10 yr olds; and ≥90 mm Hg for any child older than 10 yr

19.(A) Shock may initially manifest as only tachycardia, with or without tachypnea. Progression leads to decreased urine output, poor peripheral perfusion, respiratory distress or failure, alteration of mental status, and low BP

20.(E) A significant misconception is that shock occurs only with low BP; hypotension is often a late finding and is not a criterion for the diagnosis of shock because of a complex set of compensatory mechanisms that attempt to preserve BP and peripheral perfusion. Hypotension reflects an advanced state of decompensated shock.

21.(C) Severe Sepsis: Sepsis plus 1 of the following:

Cardiovascular organ dysfunction, defined as: Despite >40 mL/kg of isotonic intravenous fluid in 1 hr:

•Hypotension<5th percentile for age or systolic blood pressure <2 SD below normal for age or

•Need for vasoactive drug to maintain blood pressure

or

Two of the following:

•Unexplained metabolic acidosis: base deficit >5 mEq/L

•Increased arterial lactate:>2 times upper limit of normal

•Oliguria: urine output<0.5 mL/kg/hr

•Prolonged capillary refill:>5 sec

•Core-to-peripheral temperature gap:>3°C

22.(A) Vasovagal syncope is classically associated with a prodrome that includes diaphoresis, warmth, pallor, or feeling lightheaded and is often triggered by a specific event or situation such as pain, medical procedures, or emotional distress This type of syncope is characterized by hypotension and bradycardia. Approximately 30–50% of children will have had a fainting episode before 18 yr of age.

23.(D) Carotid sinus: syncope with head rotation or pressure on the carotid sinus (as in tumors, shaving, tight collars)

24.(C) All patients must have an electrocardiogram obtained, looking primarily for QT interval prolongation, preexcitation, ventricular hypertrophy, T-wave abnormalities, and conduction abnormalities.

25.(A) In diseases of decreased lung compliance, such as pneumonia and pulmonary edema, breathing is characteristically rapid and shallow (decreased tidal volume)

26.(E) Grunting is an expiratory sound caused by sudden closure of the glottis during expiration in an attempt to maintain functional residual capacity FRC and prevent alveolar atelectasis, most commonly heard in diseases with decreased FRC (e.g., pneumonia, pulmonary edema) and peripheral airway obstruction (e.g., bronchiolitis).

27.(A) Shock Energy for Defibrillation: First shock 2 J/kg, second shock 4 J/kg, subsequent shocks 4 J/kg, maximum 10 J/kg or adult dose.

28.(A) The World Congress of Drowning: definition of drowning is "the process of experiencing respiratory impairment from submersion/immersion in liquid." The term drowning does not imply the final outcome—death or survival; the outcome should be denoted as fatal or nonfatal drowning. Use of this terminology should improve consistency in reporting and research; the use of confusing descriptive terms such as "near," "wet," "dry," "secondary," "silent," "passive," and "active" should be abandoned.

29.(A) Veins suitable for cannulation are numerous, but there is considerable anatomic variation from patient to patient. In the upper extremities, the median antecubital vein, located in the antecubital fossa, is often the largest and easiest to access. Many veins on the dorsum of the hand are also suitable for cannulation because they are often large and easily located, and their cannulation is generally well tolerated.

30.(E) Indications for admission:

- ✓ Burns affecting >10% of BSA Burns
- ✓ >10–20% of BSA in adolescent/adult
- ✓ 3rd-degree burns
- ✓ Electrical burns caused by high-tension wires or lightning Chemical burns
- ✓ Inhalation injury, regardless of the amount of BSA burned
- ✓ Inadequate home or social environment
- ✓ Suspected child abuse or neglect
- ✓ Burns to the face, hands, feet, perineum, genitals, or major joints
- ✓ Burns in patients with preexisting medical conditions that may complicate the acute recovery phase
- ✓ Associated injuries (fractures)
- ✓ Pregnancy

31.(A) Drowning rates are consistently highest in 1-4 yr old children, likely because of their curious but unaware nature, coupled with the rapid progression of their physical capabilities. The highest drowning death rates were seen in children age 1-4 yr and 15-19 yr (crude rates of 2.56 and 1.2 per 100,000, respectively).

32.(B) Most (71%) drowning deaths in children younger than 1 yr occur in the bathtub, when an infant is left alone or with an older sibling. The next major risk to children <1 yr is the large (5-gallon) household bucket, implicated in 16% of infant drowning deaths.

33.(D) Fosphenytoin or phenytoin (loading dose of 10-20 mg of phenytoin equivalents/kg, followed by maintenance dosing with 5-8 mg of phenytoin equivalents/ kg/day in 2-3 divided doses; levels should be monitored) may be considered as an anticonvulsant; it may have some neuroprotective effects and may mitigate neurogenic pulmonary edema. Benzodiazepines, barbiturates, and other anticonvulsants may also have some role in seizure therapy, although no conclusive studies have shown improved neurologic outcome.

34.(D) Approximately 18% of burns are the result of child abuse (usually scalds), making it important to assess the pattern and site of injury and their consistency with the patient history. Glove or stocking burns of the hands and feet; single-area deep burns on the trunk, buttocks, or back; and small, full-thickness burns (e.g., cigarette burns) in young children should raise the suspicion of child abuse.

35.(C) Epinephrine is usually the initial drug of choice in victims with bradyasystolic cardiopulmonary arrest (IV dose is 0.01 mg/kg using the 1: 10,000 [0.1 mg/mL] solution given every 3-5 min, as needed). Epinephrine can be given intratracheally (endotracheal tube dose is 0.1-0.2 mg/kg of 1: 1,000 [1 mg/mL] solution) if no IV access is available.

36.(A)

First Tier: intubation and controlled mechanical ventilation, head of bed elevation, sedation and analgesia neuromuscular blockade, osmolar agents (hypertonic saline, mannitol)

Second Tier: barbiturate infusion, decompressive craniectomy, mild hypothermia, hyperventilation, lumbar CSF drainage

37.(C) To diagnose POTS the patient needs to undergo a head-up tilt test for at least 10 min. It is important to have the patient supine for at least 20 min before the tilt test. POTS can also be assessed by a standing test, measuring BP and HR at 1, 3, 5, and 10 min standing, but to have a reliable test similar to the tilt test, the patient needs to be supine for 1 hr before standing.

Other tests may include electrocardiogram, echocardiogram, and Holter monitor when there is concern of a primary cardiac cause of tachycardia, or if there is a need to determine if symptoms correlate with tachycardia. Supine and standing plasma catecholamines help confirm the diagnosis of POTS, as one expects to see either the normal doubling of norepinephrine levels from supine to standing, or a tripling with hyperadrenergic POTS.

38.(D) Rapid fluid resuscitation totaling 60-80 mL/kg or more is associated with improved survival without an increased incidence of pulmonary edema. Fluid resuscitation in increments of 20 mL/kg should be titrated to normalize HR

(according to age-based HRs), urine output (to 1 mL/kg/ hr), capillary refill time (to <2 sec), and mental status. If shock remains refractory following 60-80 mL/kg of volume resuscitation, vasopressor therapy (e.g., norepinephrine, epinephrine) should be instituted while additional fluids are administered. Pediatric guidelines for septic shock unresponsive to fluid resuscitation suggest epinephrine or dopamine whereas adult guideline recommend norepinephrine.

39.(A) Adrenal function is another important consideration in shock, and hydrocortisone replacement may be beneficial. Up to 50% of critically ill patients may have absolute or relative adrenal insufficiency. Patients at risk for adrenal insufficiency include those with congenial adrenal hypoplasia, abnormalities of the hypothalamic-pituitary axis, and recent therapy with corticosteroids (including those with asthma, rheumatic diseases, malignancies, and inflammatory bowel disease). These patients are at high risk for adrenal dysfunction and should receive stress doses of hydrocortisone. Corticosteroids may also be considered in patients with shock that is unresponsive to fluid resuscitation and catecholamines.

40.(C)

41.(E) Normal compensation does not completely correct the pH but rather minimizes a change in pH that would otherwise occur without compensation.

A normal compensation for metabolic acidosis results in a fall in PaCO2 by 1.2 mm Hg for every 1 mEq/L fall in [HCO3−].The most commonly used method to analyze the adequacy of respiratory compensation is Winter's formula:

$PaCO2 = ([HCO3^-] \times 1.5) + 8 \pm 2$

A quick method is to look at the last 2 digits of pH (provided it is not <7.10), which should be within 2 mm Hg of PaCO2.

42.(D) It is unreliable in patients with poor perfusion and poor pulsatile flow to the extremities.

43.(C) Diagnosis AMS is easily identified in older children and adolescents using the Self-Report Lake Louise AMS Scoring System. The criteria require that the individual be in the setting of a recent gain in altitude, be at the new altitude for at least several hours, and report a headache plus at least 1 of the following symptoms: gastrointestinal (GI) upset (anorexia, nausea, or vomiting), general weakness or fatigue, dizziness or lightheadedness, or difficulty sleeping.

44.(A) Scald burns account for 85% of total injuries and are most prevalent in children <4 yr old. Steam inhalation used as a home remedy to treat respiratory infections is another potential cause of burns. Flame burns account for 13%; the remaining are electrical and chemical burns.

Chapter 9

Human Genetics Questions

ZUHAIR ALMUSAWI

1. Which of the following is an Indication for genetic counseling?
   A. Paternal age ≥35 yr
   B. Paternal age ≥40 yr
   C. Paternal age ≥45 yr
   D. Paternal age ≥50 yr
   E. Paternal age ≥55 yr

2. Genetic counseling has a major role in risk assessment for which of the following cancers?
   A. colon
   B. Prostate
   C. Brain
   D. Liver
   E. Skin

3. Which of the following diseases is usually treated with enzyme replacement therapy?
   A. Niemann-Pick disease type B
   B. Neuronal ceroid lipofuscinosis
   C. Pompe disease
   D. Mucopolysaccharidoses MPS IIIA
   E. Metachromatic leukodystrophy

4. Which of the following tests can provide information about the copy number and location of a specific genomic region?
   A. Array-based copy number detection assays
   B. Chromosome analysis (karyotype)
   C. Fluorescent in situ hybridization (FISH)
   D. Whole exome sequencing (WES)
   E. polymerase chain reaction

5. The finding of male-to-male transmission essentially confirms which of the following inheritance?

A. X-linked dominant inheritance
B. X-linked recessive inheritance
C. Autosomal dominant inheritance
D. Autosomal recessive inheritance
E. Multifactorial inheritance

6. Which of the following inheritances is characterized by horizontal transmission?

A. X-linked dominant inheritance
B. X-linked recessive inheritance
C. Autosomal dominant inheritance
D. Autosomal recessive inheritance
E. Multifactorial inheritance

7. Which type of inheritance is expected if a woman with a genetic disorder can have affected offspring of either sex, but an affected father cannot pass on the disease to his offspring?

A. Pseudogenetic Inheritance
B. Digenic inheritance
C. Mitochondrial inheritance
D. Autosomal recessive inheritance
E. Autosomal dominant inheritance

8. When 2 chromosomes fail to separate during meiosis and thus migrate together into one of the new cells, producing 1 cell with 2 copies of the chromosome and another with no copy.
What is the name of the above mechanism of cell division?

A. Nondisjunction
B. Anaphase lag
C. Genetic recombination
D. Crossing over
E. Dislocation

9. Which term is used to describe the presence of 2 or more cell lines in a single individual?

A. Euploidy

B. Mosaicism
C. Polyploidy
D. Triploidy
E. Aneuploidy

10. Which is the **MOST** common and clinically significant type of human chromosome abnormality?
A. Euploidy
B. Mosaicism
C. Polyploidy
D. Triploidy
E. Aneuploidy

11. A couple of 38-year-old husband and a 23-year-old wife with Down syndrome consult you because the husband is planning to have a baby, he asks you about the chance of having a baby with Down syndrome, if his wife becomes pregnant.
Your proper answer, the chance of having Down syndrome is
A. 5%
B. 25%
C. 50%
D. 75%
E. 100%

12. Which of the following is a Hall's criterion to aid in diagnosis of Down syndrome?
A. Upward slanted palpebral fissures
B. Epicanthal folds
C. Speckled irises (Brushfield spots)
D. Three fontanels
E. Mild microcephaly

13. A couple brought their 3-day-old neonate with abnormal features, on examination you find midline cleft lip; flexed fingers with postaxial polydactyly; ocular hypotelorism, bulbous nose; low-set, malformed ears; microcephaly; holoprosencephaly; microphthalmia, and scalp defect.
Of the following, the **MOST** likely cause of these dysmorphic features is
A. Trisomy 13

B. Trisomy 18
C. Trisomy 8
D. Trisomy 9
E. Trisomy 21

14. What is the average time of walking in Down syndrome?
A. 12 months
B. 16 months
C. 20 months
D. 24 months
E. 28 months

15. Which of the following is the **MOST** common congenital heart disease in patient with Down syndrome?
A. Ventricular septal defect
B. Atrial septal defect
C. Patent ductus arteriosus
D. Endocardial Cushing defects
E. Aberrant subclavian artery

16. Which of the following tests is diagnostic and considered as an important advance in prenatal diagnosis of Down syndrome?
A. Free β-human chorionic gonadotropin [β-hCG]
B. Detection of cell-free fetal DNA in maternal plasma
C. Unconjugated estriol
D. α-fetoprotein
E. Fetal nuchal translucency (NT) thickness

17. A 4-year-old boy with Down syndrome presents with progressive pallor, poor weight gain, abdominal distension, loose offensive stool alternating with constipation, and poor appetite for the last 3 months. His blood film shows dimorphic features with normal WBC and platelets count.
Of the following, the **MOST** helpful investigation is
A. serum B12
B. serum folic acid
C. thyroid function test
D. tissue transglutaminase antibodies
E. serum ferritin

18. Limited hip abduction, clinodactyly and overlapping fingers; index over 3rd, 5th over 4th; closed fist, rocker-bottom feet, and hypoplastic nails are features of which of the following trisomies?

A. Trisomy 13
B. Trisomy 18
C. Trisomy 8
D. Trisomy 9
E. Trisomy 21

19. A 3-week old neonate with the following features; hypotonia, short stature, characteristic shrill cry, microcephaly with protruding metopic suture, hypertelorism, bilateral epicanthic folds, high arched palate, and wide flat nasal bridge.

Of the following, the **MOST** likely diagnosis is

A. Wolf-Hirschhorn syndrome
B. Cri du chat syndrome
C. DiGeorge type 2 syndrome
D. Jacobsen syndrome
E. Miller-Dieker syndrome

20. An 8-year-old boy brought by his father complaining from poor school performance. On examination, you find a friendly boy with round face, small upturned nose, long upper lip length, wide mouth, with full cheeks and lips. Lab investigations reveal hypercalcemia, and echo shows supravalvular aortic stenosis.

Of the following, the **MOST** likely diagnosis is

A. DiGeorge syndrome
B. Williams syndrome
C. Kallmann syndrome
D. Angelman syndrome
E. Axenfeld-Rieger syndrome

21. An 18-month-old child presents with following features; microcephaly, ptosis, beaked nose with low-lying philtrum, broad thumbs, large toes, and intellectual disability.

Of the following, the **MOST** likely diagnosis is

A. Rubinstein-Taybi syndrome
B. Williams syndrome

C. Kallmann syndrome
D. Smith-Magenis syndrome
E. Jacobsen syndrome

22. A 3-week-old small for gestational age newborn presents with webbing of the neck, protruding ears, and non-pitting swellings of the hands and feet. Echo shows bicuspid aortic valve with coarctation of the aorta.
Of the following, the **MOST** likely diagnosis is
A. Noonan syndrome
B. Trisomy X syndrome
C. Turner syndrome
D. Jacobsen syndrome
E. Kallmann syndrome

23. Which of the following differentiate Noonan syndrome from Turner syndrome?
A. Short stature
B. Low posterior hairline
C. shield chest
D. Right-sided cardiac lesions
E. Webbed neck

24. What is the **MOST** common sex chromosome aneuploidy in humans?
A. Noonan syndrome
B. Turner syndrome
C. Klinefelter syndrome
D. 47,XYY
E. Fragile X syndrome

25. An 18-month-old boy presents with long palpebral fissures, eversion of lateral portion of lower eyelids, long eyelashes, and blue sclera. On examination, he has global developmental delay, microcephaly, hypotonia with Joint hyperextensibility, prominent fingertip pads, and hypoplastic finger nails.
Of the following, the **MOST** likely diagnosis is
A. Bloom syndrome
B. Kabuki syndrome
C. Axenfeld-Rieger syndrome
D. Kallmann syndrome

E. Smith-Magenis syndrome

26. A 16-year-old adolescent boy has a long face, large cupped ears, high arched palate, a prominent square jaw, hyperextensible finger joints, flat feet, and large testes. He also complains from recurrent otitis media, infrequent seizures, and mild intellectual disability.
Of the following, the **MOST** likely diagnosis is

A. Jacobsen syndrome
B. Fragile X syndrome
C. Noonan syndrome
D. Trisomy X syndrome
E. Pallister-Killian syndrome

27. A preschool child has prominent full cheeks, abnormal ear lobes, sparse hair in the temporal regions, pigmentary skin anomalies, supernumerary nipples, infrequent seizures, and profound intellectual disability. He also had undergone surgery for diaphragmatic hernia in neonatal period.
Of the following, the **MOST** likely diagnosis is

A. Jacobsen syndrome
B. Fragile X syndrome
C. Pallister-Killian syndrome
D. Kallmann syndrome
E. Smith-Magenis syndrome

28. A 1-year-old infant presents with hypochromic microcytic anemia, after full investigations found to have β thalassemia major, Hb electrophoresis were done for parents showing normal father and mother with β thalassemia minor.
What is the type of inheritance in this case?

A. Chromosome breakage syndrome
B. Uniparental disomy
C. Autosomal dominant inheritance
D. Autosomal recessive inheritance
E. Multifactorial inheritance

29. Which of the following is a classic example of imprinting disorder?

A. Prader-Willi syndrome
B. Cartilage hair hypoplasia
C. Bloom syndrome

D. Kallmann syndrome
E. Smith-Magenis syndrome

30. Which of the following is a minor diagnostic criterion for Prader-Willi syndrome?

A. Feeding problems and failure to thrive as an infant
B. Sleep apnea
C. Weight gain at 1-6 yr
D. Characteristic dysmorphic facial features
E. Small genitalia

Chapter 9

Human Genetics
Answers

ZUHAIR ALMUSAWI

1.(B) Indications for genetic counseling
Advanced parental age
• Maternal age ≥35 yr
• Paternal age ≥40 yr

2.(A) Genetic counseling has a major role in risk assessment for cancer, especially breast, ovarian, or colon cancer, for which well-defined risk models and genetic tests are available to assess risk to an individual.

3.(C) Enzyme replacement therapies are available for Gaucher disease and Fabry disease, some mucopolysaccharidoses (MPS I, II, IVA, VI), acid lipase deficiency, and Pompe disease, and are being tested for MPS IIIA, MPS VII, metachromatic leukodystrophy, α-mannosidosis, Niemann-Pick disease type B, and neuronal ceroid lipofuscinosis, late infantile (CLN2).

4.(C) Fluorescent in situ hybridization (FISH) can provide information about the copy number and location of a specific genomic region. Array-based copy number detection assays can be used to screen for chromosomal deletions (large and small) and duplications across the genome, but do not provide information about the orientation or location of genomic regions.
A chromosome analysis (karyotype) can detect relatively large chromosomal deletions and duplications and can also be useful in identifying inversions and chromosomal rearrangements even when they are copy number neutral changes that do not result in a deletion or duplication of genomic material.

5.(C)

6.(D) Autosomal recessive disorders are characterized by horizontal transmission , the observation of multiple affected members of a kindred in the same generation, but no affected family members in other generations.

7.(C) An individual's mitochondrial genome is entirely derived from the mother because sperm contain relatively few mitochondria, and these are degradated after fertilization. It follows that mitochondrial inheritance is essentially maternal inheritance.

8.(A) Two common errors of cell division may occur during meiosis or mitosis, and either can result in an abnormal number of chromosomes. The 1st error is nondisjunction, in which 2 chromosomes fail to separate during meiosis and thus migrate together into one of the new cells, producing 1 cell with 2 copies of the chromosome and another with no copy. The 2nd error is anaphase lag, in which a chromatid or chromosome is lost during mitosis because it fails to move quickly enough during anaphase to become incorporated into 1 of the new daughter cells.

9.(B) Mosaicism is an abnormality defined as the presence of 2 or more cell lines in a single individual. Polyploidy is a common abnormality seen in 1st-trimester pregnancy losses. Triploid cells are those with 3 haploid sets of chromosomes (3n) and are only viable in a mosaic form. Triploid infants can be live born but do not survive long. Triploidy is often the result of fertilization of an egg by 2 sperm (dispermy). Failure of 1 of the meiotic divisions, resulting in a diploid egg or sperm, can also result in triploidy.

10.(E) Abnormal cells that do not contain a multiple of haploid number of chromosomes are termed aneuploid cells.

11.(C) Most males with Down syndrome are sterile, but some females have been able to reproduce, with a 50% chance of having trisomy 21 pregnancies.

12.(A) Hall's criteria to aid in diagnosis of Down syndrome include

- ✓ Hypotonia
- ✓ Poor Moro reflex
- ✓ Flat face
- ✓ Upward slanted palpebral fissures
- ✓ Small dysplastic ears
- ✓ Joint hyperflexibility
- ✓ Short neck, redundant skin
- ✓ Short 5th digit with clinodactyly
- ✓ Single transverse palmar creases
- ✓ Pelvic dysplasia

13. (A)

14. (C)

Milestone	**CHILDREN WITH DOWN SYNDROME**		**UNAFFECTED CHILDREN**	
	Average (mo)	Range (mo)	Average (mo)	Range (mo)
Smiling	2	1.5-3	1	1.5-3
Rolling over	6	2-12	5	2-10
Sitting	9	6-18	7	5-9
Crawling	11	7-21	8	6-11

Creeping	13	8-25	10	7-13
Standing	10	10-32	11	8-16
Walking	20	12-45	13	8-18
Talking, words	14	9-30	10	6-14
Talking, sentences	24	18-46	21	14-32

15.(D)

16.(B) Detection of cell-free fetal DNA in maternal plasma is also diagnostic and replacing conventional 1st- and 2nd-trimester screens. Next-generation DNA sequencing has reduced the cost of this procedure, which has a high degree of accuracy (98% detection rate) and applicability.

17.(D) Congenital and acquired gastrointestinal anomalies (celiac disease) and hypothyroidism are common. Other abnormalities include megakaryoblastic leukemia, immune dysfunction, diabetes mellitus, seizures, alopecia, juvenile idiopathic arthritis, and problems with hearing and vision.

18.(B)

19.(B)

20.(B)

21.(A)

22.(C) Older children and adults with Turner syndrome have short stature and exhibit variable dysmorphic features. Congenital heart defects (40%) and structural renal anomalies (60%) are common. The most common heart defects are bicuspid aortic valves, coarctation of the aorta, aortic stenosis, and mitral valve prolapse. The gonads are generally streaks of fibrous tissue (gonadal dysgenesis). There is primary amenorrhea and lack of secondary sex characteristics. Most patients tend to be of normal intelligence, but intellectual disability is seen in up to 6% of affected children.

23.(D) In contrast to Turner syndrome, Noonan syndrome affects both sexes and has a different pattern of congenital heart disease, typically involving right-sided lesions.

24.(C) Persons with Klinefelter syndrome are phenotypically male; this syndrome is the most common cause of hypogonadism and infertility in males and the most common sex chromosome aneuploidy in humans.

25.(B) Clinical Manifestations of Kabuki Syndrome

Facial

- ✓ Long palpebral tissues and eversions of lateral third of lower eyelids
- ✓ Ptosis
- ✓ Broad, arched eyebrows with sparse hair on lateral third
- ✓ Long eyelashes

- ✓ Blue sclerae
- ✓ Protuberant ears
- ✓ Short nasal columella (depressed nasal tip)

Neurodevelopmental

- ✓ Hypotonia
- ✓ Developmental delay (IQ about 60; >80 in 10%)
- ✓ Low birthweight
- ✓ Postnatal growth deficiency
- ✓ Microcephaly
- ✓ Seizures
- ✓ Autism

Extremity/Skeletal

- ✓ Short, incurved 5th finger
- ✓ Brachydactyly
- ✓ Kyphosis
- ✓ Joint hyperextensibility
- ✓ Persistent fetal fingertip pads
- ✓ Hypoplastic finger nails

Cardiovascular

- ✓ Multiple forms of congenital heart disease

Other

- ✓ Nonimmune hydrops
- ✓ Hypothyroidism
- ✓ Precocious puberty
- ✓ Delayed puberty
- ✓ Lymphatic malformations
- ✓ Feeding difficulties

26.(B) A clinically significant fragile site is on the distal long arm of chromosome Xq27.3 associated with the fragile X syndrome. Fragile X syndrome accounts for 3% of males with intellectual disability.

27.(C) Pallister-Killian syndrome is caused by mosaicism for an isochromosome 12p. The presence of the isochromosome 12p in cells gives 4 functional copies for the short arm of chromosome 12 in the affected cells.

28.(B) Uniparental disomy (UPD) occurs when both chromosomes of a pair or areas from one chromosome in any individual have been inherited from a single parent. Examples of UPD are spinal muscular atrophy, cystic fibrosis, cartilage hair hypoplasia, α- and β-thalassemias, and Bloom syndrome.

29.(A) A classic example of imprinting disorder is seen in Prader-Willi syndrome and Angelman syndrome, 2 very different clinical conditions. These syndromes are usually associated with deletion of the same region in the proximal long arm of chromosome 15. A deletion on the paternally derived chromosome causes Prader-Willi syndrome, while a maternal deletion of the same region as in Prader-Willi syndrome causes Angelman syndrome.

30.(B)

Consensus Diagnostic Criteria for Prader-Willi Syndrome		
MAJOR CRITERIA (1 point each)		**MINOR CRITERIA** (1/2 point each)
1	Neonatal/infantile hypotonia	Decreased fetal movement and infantile lethargy
2	Feeding problems and failure to thrive as an infant	Typical behavior problems
3	Weight gain at 1-6 yr; obesity; hyperphagia	Sleep apnea
4	Characteristic dysmorphic facial features	Short stature for family by 15 yr
5	Small genitalia; pubertal delay and insufficiency	Hypopigmentation for the family
6	Developmental delay/intellectual disability	Small hands and feet for height
7		Narrow hands, straight ulnar border
8		Esotropia, myopia
9		Thick, viscous saliva
10		Speech articulation defects
11		Skin picking

Chapter 10

Metabolic Disorders Questions

QAHTAN ALOBAIDY

1. Which of the following inborn errors of metabolism is associated with neonatal apnea?

A. Hyperphenylalaninemia
B. Tyrosinemia type II
C. Urea cycle disorders
D. Hypermethioninemia
E. Citrullinemia type II

2. Which of the following anticonvulsant drugs may cause hyperammonemia?

A. Valproic acid
B. Paraldehyde
C. Phenobarbital
D. Clobazam
E. Topiramate

3. Which of the following glycogen storage diseases (GSD) is associated with cardiomyopathy?

A. GSD type Ia (glucose-6-phosphatase deficiency)
B. GSD type Ib (impaired glucose-6-phosphate exchanger)
C. GSD type III (glycogen debrancher enzyme deficiency)
D. GSD type VI (liver glycogen phosphorylase deficiency)
E. GSD type IX (phosphorylase kinase deficiencies)

4. Macrocephaly is a pathognomonic clinical finding associated with inborn errors of metabolism.
Which of the following is associated with macrocephaly?

A. Mucopolysaccharidoses
B. Sphingolipidoses
C. Glutaric Acidemia Type 1
D. Wolman Disease

E. Farber Disease

5. A 3-month-old infant presents with macroglossia, hypotonia, increased serum creatinine kinase , and cardiomyopathy.
Of the following, the **MOST** likely diagnosis is

A. glycogen storage disease type II
B. mucopolysaccharidoses
C. oligosaccharidoses
D. sphingolipidoses
E. galactosialidosis

6. A 6-old-male infant presents with exaggerated startle response. His ophthalmological examination revealed macular cherry-red spot.
Of the followings, the **MOST** likely diagnosis is

A. GM1 gangliosidosis
B. Tay-Sachs disease
C. Farber disease
D. galactosialidosis
E. sialidosis

7. Which of the following in born errors of metabolism is characterized by alopecia?

A. Multiple carboxylase deficiency
B. Gaucher disease type 2
C. Steroid sulfatase deficiency
D. Refsum disease
E. Serine deficiency disorders

8. Which of the following inborn errors of metabolism is characterized by sweaty feet urine odor?

A. Maple syrup urine disease
B. Isovaleric academia
C. Phenylketonuria
D. Tyrosinemia type 1
E. Cystinuria

9. Which of the following characterize urea cycle defects with hyperammonemia from other types of inborn error of metabolism with hyperammonemia?

A. Normal serum pH
B. High bicarbonate value
C. Lactic acidosis
D. Ketosis
E. Hypoglycemia

10. A 6-week-old Infant presents with recurrent attacks of hypoglycemia, hepatomegaly, ascites, and jaundice. Blood ammonia, pH, and bicarbonate values are normal.
Of the followings, the **MOST** likely diagnosis is

A. propionic academia
B. classic galactosemia
C. Zellweger spectrum disorders
D. long-chain fatty acid oxidation defects
E. GLUD1 -related hyperinsulinemic hypoglycemia

11. A concerned family had two children with classic phenylketonuria (PKU), consult you about the main vital organ damaged by this disease.
Of the following, the **BEST** answer is the

A. brain
B. kidney
C. skin
D. eye
E. lung

12. What is the **BEST** time to assess phenylketonurea to avoid false negative results?

A. 1st 24 hr of life
B. 24-48 hr of life
C. 48-72 hr of life
D. 72-96 hr of life
E. 96-120 hr of life

13. What is the safe level of plasma phenylalanine during pregnancy?

A. <2 mg/dL

B. <4 mg/dL
C. <6 mg/dL
D. <8 mg/dL
E. <10 mg/dL

14. An infant already on treatment for phenylketonuria (PKU) shows evidence of neurological deterioration despite adequate control of plasma phenylalanine.
Of the following, the **MOST** likely cause is
A. cofactor BH4 deficiency
B. non PKU hyperphenylalaninemia
C. transient neonatal hyperphenylalaninemia
D. severe phenylalanine hydroxylase deficiency
E. hyperphenylalaninemia secondary to liver disease

15. What is the **MOST** common renal manifistation of tyrosinemia type I ?
A. Renal cysts
B. Nephronophthiasis
C. Cortical necrosis
D. Fanconi-like syndrome
E. Renal stones

16. Increased level of α- fetoprotein in the cord blood of affected infants with tyrosinemia indicate
A. liver damage
B. renal damage
C. neural tube defect
D. cardiac defect
E. hydrocephaly

17. Which of the following result in elevated levels of urinary succinylacetone?
A. Galactosemia
B. Hereditary fructose intolerance
C. Neonatal iron storage disease
D. Citrullinemia type II
E. Tyrosinemia type I

18. A-7-month-old infant after weaning from breastfeeding and introduction of a solid feeding including meet and fish; starts to develop severe metabolic acidosis, ketosis, failure to thrive, anemia, mild hepatomegaly, renal tubular acidosis, and an unusual odor. Investigations show elevated level of 4-hydroxyphenylpyruvic acid in their urine.
Of the following, the **MOST** likely diagnosis is

A. tyrosinemia type I
B. tyrosinemia type II
C. tyrosinemia type III
D. hawkinsinuria
E. alkaptonuria

19. A concerned parents counsel you about their infant, who had a black-stained diapers, with ochronosis (dark spots on the sclera and ear cartilage).
Of the following, the **MOST** likely diagnosis is

A. tyrosinemia type I
B. tyrosinemia type II
C. tyrosinemia type III
D. hawkinsinuria
E. alkaptonuria

20.Nitisinone is reasonable drug use as treatment option in

A. alkaptonuria
B. hawkinsinuria
C. propionic acidemia
D. tyrosine hydroxylase deficiency
E. transient tyrosinemia of the newborn

21. A 14-year-old female had generalized albinism; with history of recurrent epistaxis, postsurgical bleeding, and abundant menses. Her bleeding time is prolonged and platelet count is normal?
Of the following, the **MOST** likely diagnosis is

A. Hermansky-Pudlak Syndrome
B. Chédiak-Higashi Syndrome
C. Griscelli syndrome
D. Vici syndrome
E. Waardenburg Syndrome

22. A 10-year-old boy presented to emergency unit with stroke. On examination; he was tall with elongated limbs, arachnodactyly, scoliosis, pectus excavatum, genu valgum, pes cavus, high arched palate, and crowding of the teeth.
Of the following, the **BEST** treatment for this condition is high doses of vitamin

A. B1 (thiamine)
B. B2 (riboflavin)
C. B3 (niacin)
D. B5 (pantothenic acid)
E. B6 (pyridoxine)

23. In classic homocystinuria, thromboembolic episodes involving both large and small vessels, especially those of the brain, are common and may occur at any age.
Which of the following is recommended for preventing vascular events in patients unresponsive to vitamin B6 therapy?

A. Folic acid
B. Betaine
C. Vitamin B12
D. Riboflavin
E. Thiamine

24. A 6-week-old boy present with refusal to feed, vomiting, an exaggerated startle reaction, severe intractable seizures , cortical atrophy with subcortical multicystic lesions, and severe developmental delay. Opthalmological examination reveals bilateral dislocation of ocular lenses.
Of the following, the **MOST** likely diagnosis is

A. sulfite oxidase deficiency
B. Hartnup disorder
C. maple syrup urine disease
D. isovaleric academia
E. biotinidase deficiency

25. Which of the following inborn errors of metabolism, a high protein diet results in a favorable response?

A. Sulfite Oxidase Deficiency
B. Hartnup Disorder
C. Maple Syrup Urine Disease

D. Isovaleric Academia
E. Biotinidase deficiency

26. Which of the following vitamins in high doses have dramatic clinical and biochemical improvement in maple syrup urine disease?
A. B1 (thiamine)
B. B2 (riboflavin)
C. B3 (niacin)
D. B5 (pantothenic acid)
E. B6 (pyridoxine)

27. A 3-month-old infant presents with severe vomiting suggesting pyloric stenosis; but investigations show severe acidosis, hyperammonemia, hypoglycemia, hypocalcemia, and bone marrow suppression. He had the characteristic odor of sweaty feet.
Which of the following amino acid defects is blamed in this presentation?
A. Tryptophan
B. Isoleucine
C. Leucine
D. Valine
E. Methionine

28. Which of the following inborn error of metabolism due to amino acidopathy results in dermatological manifestation in form of pellagra-like rash?
A. Hartnup disorder
B. Biotinidase deficiency
C. Methylglutaconic acidurias
D. Classic maple syrup urine disease
E. Hyperimmunoglobulinemia D syndrome

29. Which of the following inborn error of metabolism due to amino acidopathy results in dermatological manifestation in form of acrodermatitis enteropathica?
A. Hartnup disorder
B. Biotinidase deficiency
C. Methylglutaconic acidurias
D. Classic maple syrup urine disease
E. Hyperimmunoglobulinemia D syndrome

30. Which of the following inborn error of metabolism due to amino acidopathy results in dermatological manifestation in form of morbilliform rash?

A. Hartnup disorder
B. Biotinidase deficiency
C. Methylglutaconic acidurias
D. Classic maple syrup urine disease
E. Hyperimmunoglobulinemia D syndrome

31. Which of the following inborn errors of metabolism due to amino acidopathy results in dermatological manifestation in form of intractable seborrheic dermatitis?

A. Hartnup disorder
B. Biotinidase deficiency
C. Methylglutaconic acidurias
D. Classic maple syrup urine disease
E. Hyperimmunoglobulinemia D syndrome

32. A 10-day-old neonate present as a sepsis-like picture with clinical signs of severe metabolic acidosis, with a high anion gap. Investigations reveal ketonuria, hypoglycemia, anemia, neutropenia, and thrombocytopenia. Brain imaging shows cerebral atrophy, delayed myelination, and abnormalities in the globus pallidus and other parts of the basal ganglia.
Of the following, the **MOST** likely diagnosis is

A. Propionic acidemia
B. Isolated homocystinuria
C. Methylmalonic acidemias
D. Combined malonic and methylmalonic aciduria
E. Combined methylmalonic aciduria and homocystinuria

33. A 10- hour- old neonate presents with poor feeding, failure to suck, lethargy, profound hypotonia, recurrent myoclonic seizures, and hiccups. Blood pH is normal with negative urine assay for organic acids.
Of the following, the **MOST** likely diagnosis is

A. sarcosinemia
B. primary trimethylaminuria
C. nonketotic hyperglycinemia
D. primary hyperoxaluria type I
E. creatine deficiency disorder

34. Which of the following inborn error of metabolism due to amino acidopathy result in body odor that resembles that of a fish?

A. Sarcosinemia
B. Primary trimethylaminuria
C. Nonketotic hyperglycinemia
D. Primary hyperoxaluria type 1
E. Creatine deficiency disorder

35. To assess the pyridoxine responsiveness in patients with primary hyperoxaluria type I, the duration of treatment trial should be at least

A. 1 month
B. 2 months
C. 3 months
D. 4 months
E. 5 months

36. A 3-year-old boy suffers from recurrent otitis media, sinusitis, scaly erythematous maculopapular rash, purpura, and telangiectasia. These skin lesions proceed to recurrent, severe, and painful skin ulcers on hands and legs. Investigation reveals a high level of urinary imidodipeptides.
Of the following, the **MOST** likely diagnosis is

A. prolidase deficiency
B. hyperprolinemia type I
C. hyperprolinemia type II
D. phosphoserine phosphatase
E. phosphoserine aminotransferase deficiency

37. In the 1st few days of life, a full term baby develops a constellation of metabolic acidosis, jaundice, and mild to moderate hemolytic anemia. Urine examination shows 5-oxoprolinuria.
Of the following, the **MOST** likely diagnosis is

A. 5-oxoprolinase deficiency
B. hyperprolinemia type I
C. glutathione synthetase deficiency
D. γ-glutamylcysteine synthetase deficiency
E. γ-glutamyl transpeptidase deficiency

38. Which of the following drugs in patient with renal failure can cause hypoglycemia due to substrate limitation?

A. Trimethoprim-sulfamethoxazole
B. Ampicillin
C. Ciprofloxacin
D. Ceftriaxone
E. Vancomycine

39. Which of the following is the **MOST** common manifestation of hypoglycemia in the newborn?

A. episodes of cyanosis
B. jitteriness
C. seizures
D. lethargy
E. intermittent apneic spells

40. Catabolism of amino acids results in the production of free ammonia, which in high concentration is toxic to the CNS.
In healthy term infants, normal levels can reach as high as

A. 50 µmol/L
B. 100 µmol/L
C. 200 µmol/L
D. 300 µmol/L
E. 400 µmol/L

41. Which value of blood glucose concentration a in sick neonate should be treated vigorously?

A. <25 mg/dL
B. <35 mg/dL
C. <45 mg/dL
D. <55 mg/dL
E. <65 mg/dL

42. Which of the following counter-regulatory hormones act to increase blood glucose level by mobilizing amino acids from muscle?

A. Glucagon
B. Growth hormone
C. Cortisol

D. Epinephrine
E. Thyroxin

43. Which of the following porphyrias causing non-blistering photosensitivity?
A. Congenital erythropoietic porphyria
B. Porphyria cutanea tarda
C. Hereditary coproporphyria
D. Variegate porphyria
E. X-Linked protoporphyria

44. Which of the following anti-epileptic drugs can elevate blood ammonia?
A. Phenobarbital
B. Midazolam
C. Phenytoin
D. Valproic acid
E. Levetiracetam

45. Which of the following is the **MOST** consistent laboratory finding in Hutchinson-Gilford progeria syndrome?
A. Low platelet count
B. High c-reactive protein titer
C. Low serum leptin
D. High liver enzyme
E. High blood urea

46. A 15-month-old boy suffers from progressive spastic diplegia with scissoring of the lower extremities, choreoathetotic movements, loss of already achieved developmental milestones, and failure to thrive
Which of the following inborn error of metabolism mimics the above presentation?
A. Glycine encephalopathy
B. Urea cycle disorders
C. Organic acidemias
D. Fatty acid oxidation defects
E. Mitochondrial respiratory chain defects

47. A 7-year-old boy had night blindness, myopia, loss of peripheral vision, and posterior subcapsular cataract with atrophic lesions in the retina resembling cerebral gyri.
Which of the following is the **MOST** likely diagnosis?

A. Classic citrullinemia
B. Tyrosinemia type I
C. Hyperargininemia
D. Argininosuccinic aciduria
E. Hyperornithinemia

48. A 3- day-old baby presented with generalized seizures unresponsive to conventional anticonvulsant therapies. Mother reports abnormal intrauterine fluttering movements. Laboratory findings show increased concentrations of α-aminoadipic semialdehyde and pipecolic acid in the CSF, plasma, and urine.
Which of the following is the **BEST** line of treatment?

A. Magnesium
B. Vitamin B6
C. Calcium
D. General anesthesia
E. Paraldehyde

49. A 9-month-old boy with macrocephaly and normal developmental milestones, after a minor Infection he suddenly lost head control followed by seizures, generalized rigidity, dystonia, and choreoathetosis. Brain imaging reveals increased extra axial fluid with stretched bridging veins, cortical atrophy, and fibrosis.
Of the following, the **MOST** likely diagnosis is

A. glutaric aciduria type 1
B. Canavan disease
C. leukodystrophy
D. hyperprolinemia type II
E. Alexander disease

50. A 8-month-old boy presents with refusal to feed, nausea, vomiting, and mild diarrhea after ingestion of a high-protein meal. Physical findings reveal moderate hepatosplenomegaly, thin extremities with moderate centripetal adiposity, and growth retardation. Laboratory findings show hyperammonemia,

increased serum ferritin, hypercholesterolemia, and hypertriglyceridemia with increased urinary levels of lysine, arginine, and ornithine.
Of the following, the **MOST** likely diagnosis is

A. Alexander disease
B. cystinurea
C. glutaric aciduria
D. familial protein intolerance
E. argininosuccinic aciduria

51. A 6-month-old boy presents with progressive macrocephaly, severe hypotonia, persistent head lag, and delayed milestones. Head CT scan reveals diffuse white matter degeneration, primarily in the cerebral hemispheres, with less involvement of the cerebellum and brainstem.
Of the following, the **MOST** likely diagnosis is

A. glutaric aciduria type 1
B. Canavan disease
C. leukodystrophy
D. hyperprolinemia type II
E. Alexander disease

52. An adolescent boy after prolonged exercise presented with muscle pain and dark color urine, his serum levels of creatine kinase was highly elevated with normal glucose level, muscle biopsy showed increased deposition of neutral fat.
Which of the following is the **MOST** likely diagnosis?

A. Primary carnitine deficiency
B. Plasma membrane carnitine transport defect
C. Carnitine palmitoyltransferase-II deficiency
D. Carnitine:Acylcarnitine translocase deficiency
A. McArdle disease

53. Which of the following peroxisomal disorders has normal VLCFA levels?

A. Bifunctional enzyme deficiency
B. Zellweger spectrum disorder
C. Acyl-CoA oxidase deficiency
D. Neonatal adrenoleukodystrophy
E. Rhizomelic chondrodysplasia punctate

54. A newborn has short proximal upper and lower limbs; examination reveals cataract with length, weight, and head circumference below 3rd percentile. Radiologic imaging shows metaphyseal cupping and disturbed ossification with vertebral bodies coronal cleft.
Which of the following is the **MOST** likely diagnosis?

A. Achondroplasia
B. Hypochondroplasia
C. Kyphomelic dysplasia
D. Rhizomelic chondrodysplasia punctate
E. Spondylo epiphyseal dysplasia congenital

55. A previously well 7-year-boy presents with hyperactivity, inattention, and worsening school performance, followed by disturbance of vision, ataxia, seizures, and strabismus. Within one year he progressed rapidly to increasing spasticity and paralysis, visual and hearing loss, with loss of ability to speak and swallow.
Of the following, the **MOST** likely diagnosis is

A. multiple sclerosis
B. Alexander disease
C. Adrenoleukodystrophy
D. Refsum disease
E. Charcot-Marie-Tooth disease

56. A 10-year-old boy had xanthomas that cause thickening of the Achilles tendon and extensor tendons of the hands, with cutaneous lesions on the hands, elbows, knees, and buttocks with positive family history of premature heart disease in both parents. Investigations show elevated plasma cholesterol level (800 mg/dL), normal triglyceride level, and slightly decreased HDL level.
Of the following, the **MOST** likely diagnosis is

A. sitosterolemia
B. familial defective ApoB-100
C. polygenic hypercholesterolemia
D. homozygous familial hypercholesterolemia
E. heterozygous familial hypercholesterolemia

57. A 10-year-old boy presented to emergency room as severe acute abdominal pain. Examination shows eruptive xanthomas on the arms, knees, and buttocks,

with hepatosplenomegaly. A milky serum was observed during sample processing.
Of the following, the **MOST** likely diagnosis is

A. Tangier disease
B. hepatic lipase deficiency
C. familial chylomicronemia
D. familial hypertriglyceridemia
E. primary hypoalphalipoproteinemia

58. Secondary causes of transient hypertriglyceridemia should be ruled out before making a diagnosis of familial hypertriglyceridemia.
Which of the following medications is a recognized cause of secondary hypertriglyceridemia?

A. Progesterone
B. Thiazides
C. Anabolic steroids
D. Carbamazepine
E. Cyclosporine

59. Which of the following conditions may cause reduced high-density lipoprotein?

A. Sepsis
B. Stress
C. Hepatitis
D. Malnutrition
E. Renal failure

60. Treatment of hypertriglyceridemia in children rarely requires medications. Dietary restriction of fats, sugars, and carbohydrates, accompanied by increased physical activity is the main steps of management.
Which of the following levels is an indication for medical treatment despite the above mentioned measures?

A. >200 mg/dL
B. >400 mg/dL
C. >600 mg/dL
D. >800 mg/dL
E. >1,000 mg/dL

61. A 9-year-old boy presents with a stinging sensation of the lower limbs, examination shows enlarged orange tonsils and hepatosplenomegaly, his HDL-C levels was extremely low
Of the following, the **MOST** likely diagnosis is

A. Tangier disease
B. hepatic lipase deficiency
C. familial chylomicronemia
D. familial hypertriglyceridemia
E. primary hypoalphalipoproteinemia

62. Which of the following conditions is associated with very low levels of plasma cholesterol and triglycerides?

A. Intestinal lymphangiectasia
B. Friedreich ataxia
C. Abetalipoproteinemia
D. Cerebrotendinous xanthomatosis
E. Familial dysbetalipoproteinemia

63. Fasting status is a pre-requisite in assessment of which of the following?

A. Cholesterol
B. LDL-C
C. HDL-C
D. Triglycerides
E. VLDL

64. Which of the following drugs can be used for the treatment of elevated TG and LDL?

A. Nicotinic acid
B. Ezetimibe
C. Gemfibrozil
D. Statins
E. Gemfibrozil

65. A 6-month-old boy presents with developmental delay, progressive psychomotor retardation and tonic-clonic seizures. He has low-set ears, frontal bossing, depressed nasal bridge, abnormally long philtrum, and hepatosplenomegaly. Ophthalmological examination shows macular cherry-red

spot, skeletal survey reveals anterior beaking of the vertebrae, enlargement of the sella turcica, and thickening of the calvarium.
Of the following, the **MOST** likely diagnosis is

A. mucopolysaccharidoses
B. Tay-Sachs disease
C. Sandhoff disease
D. Gaucher disease
E. GM1 gangliosidosis

66. An Infant was developmentally normal till the age of 5 months, then he developed decreased eye contact, loss of motor skills and an exaggerated startle response to noise. Examination shows macrocephaly, macular pallor and retinal cherry-red spots.
Of the following, the **MOST** likely diagnosis is

A. mucopolysaccharidoses
B. Tay-Sachs disease
C. Niemann-Pick disease
D. Gaucher disease
E. GM1 gangliosidosis

67. Which of the following symptoms of Gaucher disease type 1 is **NOT** reversed by enzyme replacement therapy?

A. Organomegaly
B. Hematologic indices
C. Neurologic progression
D. Bone Pain
E. Pathologic fractures

68. A preadolescent boy had telangiectatic skin lesions distributed in bathing trunk area, hypohidrosis, corneal lenticular opacities, and painful acroparesthesia.
Of the following, the **MOST** likely diagnosis is

A. Farber disease
B. Fabry disease
C. fucosidosis
D. Schindler disease
E. multiple-sulfatase deficiency

69. The finding of lipid inclusions with characteristic birefringent “Maltese crosses” in urinary sediment examination is highly suggestive of

A. Tay-Sachs disease
B. Niemann-Pick disease
C. Gaucher disease
D. Fabry disease
E. GM1 gangliosidosis

70. A 1-year-old child presents with developmental delay and somatic features similar to those of mucopolysaccharidoses. These features include frontal bossing, hepatosplenomegaly, coarse facial features, and macroglossia.
Of the following, the **MOST** likely diagnosis is

A. Farber disease
B. Fabry disease
C. Fucosidosis
D. Schindler disease
E. Krabbe disease

71. A 1-year-old boy presents with myoclonic seizures, irritability, inability to walk, and hyperextension of the knee causing genu recurvatum. Examination reveals absent deep tendon reflexes, muscle wasting and hypotonia, with nystagmus and optic atrophy.
Of the following, the **MOST** likely diagnosis is

A. Krabbe disease
B. metachromatic leukodystrophy
C. multiple-sulfatase deficiency
D. mucolipidoses
E. GM1 gangliosidosis

72. A 9-month-old female presents with failure to thrive, painful joint swelling, and nodule formation misdiagnosed as juvenile rheumatoid arthritis.
Of the following, the **MOST** likely diagnosis is

A. Farber disease
B. Fabry disease
C. Fucosidosis
D. Schindler disease
E. Krabbe disease

73. A 3-week-old male presents with relentless vomiting, abdominal distention, steatorrhea, and hepatosplenomegaly. Investigations show hyperlipidemia and hepatic dysfunction while abdominal ultrasound reveals calcifications of the adrenal glands.
Of the following, the **MOST** likely diagnosis is

A. Wolman disease
B. Fabry disease
C. Fucosidosis
D. Schindler disease
E. Krabbe disease

74. Which of the following laboratory findings is seen in type I glycogen storage disease?

A. Normal blood lactate
B. Low uric acid concentrations
C. Elevated serum creatine kinase levels
D. Prolonged bleeding time
E. Marked elevation in liver transaminase levels

75. A 4-month-old boy presents with feeding difficulties, floppiness, macroglossia, and hepatomegaly. Investigation reveals elevated level of serum creatine kinase while chest x-ray shows massive cardiomegaly.
Of the following, the **MOST** likely diagnosis is

A. Danon disease
B. McArdle disease
C. Pompe disease
D. Tarui disease
E. Schindler disease

76. A 10-year-old boy complains from exercise intolerance with muscle cramps and pain which is relieved after a brief period of rest. He reports burgundy-colored urine after exercise. Lab findings show elevated levels of serum CK at rest, which further increases after exercise.
Of the following, the **MOST** likely diagnosis is

A. Danon disease
B. McArdle disease
C. Pompe disease
D. Tarui disease

E. Schindler disease

77. Which of the following glycogen storage diseases can present as hereditary non-spherocytic hemolytic anemia?

A. Danon disease
B. McArdle disease
C. Pompe disease
D. Tarui disease
E. Andersen disease

78. A 2-year-old boy with history of recurrent stones formation presented with renal colic and hematuria. Abdominal US show multiple stones (6 to 8mm) in both kidneys while KUB is clear. Mother noticed brownish spots on his diaper. Urine examination revealed yellow-brown crystals.
Which of the following is the **MOST** likely diagnosis?

A. Hyperoxaluria
B. Hypercystinuria
C. Hyperclaciuria
D. Hypocitrateuria
E. Dihydroxyadeninuria

79. A concerned parents bring their baby because of his self-injurious behavior. Examination reveals mutilated fingers, mouth, and buccal mucosa. Self-biting is so intense that lead to amputation of right little finger and substantial loss of tissue around the lips. He has dystonic movements and spasticity with moderate range of intellectual disability. Serum levels of uric acid 7mg /dL.
Which of the following is the **MOST** likely diagnosis?

A. Athetoid cerebral palsy
B. Lesch-nyhan disease
C. Dopa-responsive dystonia
D. Myoclonus – dystonia
E. Familial dysautonomia

80. A 10-year-old boy presented to endocrinology department as short stature with normal mentality. Examination reveals short trunk and neck, upper to lower segment (0.7/1), genu valgum, kyphosis, mild corneal clouding, small teeth with abnormally thin enamel, and hepatomegaly.
Which of the following is the **MOST** likely diagnosis?

A. MPS-I: Hurler syndrome
B. MPS-II: Hunter syndrome
C. MPS-III: Sanfilippo syndrome
D. MPS-IV: Morquio syndrome
E. MPS-VI: Maroteaux-Lamy syndrome

81. Which of the following is the **SOLE** manifestation of galactokinase deficiency?
A. Jaundice
B. Cataract
C. Hepatomegaly
D. Hypoglycemia
E. Ataxia

82. Which of the following types of mucopolysaccharidoses usually presents late, because of mild physical features?
A. MPS-I: Hurler syndrome
B. MPS-II: Hunter syndrome
C. MPS-III: Sanfilippo syndrome
D. MPS-IV: Morquio syndrome
E. MPS-VI: Maroteaux-Lamy syndrome

83. Which of the following is an early marker of Hunter syndrome in African and Asian patients?
A. extensive mongolian spots
B. coarse facial features
C. dysostosis multiplex
D. joint stiffness
E. intellectual disability

84. A malnourished infant presents with feeding, swallowing problems, and developmental delay together with radiologic evidence of symmetric lesions affecting the basal ganglia, brainstem, and subthalamic nuclei.
Of the following, the **MOST** likely diagnosis is
A. Leigh Disease
B. Pearson syndrome
C. Barth syndrome
D. mitochondrial DNA Depletion Syndromes

E. nuclear DNA respiratory chain defects

85. A 1-year-old male presents with seizures, global developmental delay, visceromegaly, corneal clouding, and cherry-red maculae. Skeletal survey shows dysostosis multiplex
Of the following, the **MOST** likely diagnosis is

A. sialidosis
B. α-mannosidosis
C. mucopolysacharradiosis
D. aspartylglucosaminuria
E. cerebro-ocular dysplasia–muscular dystrophy

86. A 14-month-old female presents with feeding difficulties and speech delay. Examination shows a characteristic facial features (short nose, long philtrum, and large ears), inverted nipples, and abnormal fat pads distribution, with alternating strabismus, nystagmus, hypotonia, and decreased reflexes.
Of the following, the **MOST** likely diagnosis is

A. phosphomannomutase-2 deficiency
B. mannosephosphoisomerase deficiency
C. glucosyltransferase-1 deficiency
D. cerebro-ocular dysplasia–muscular dystrophy
E. muscle-eye-brain disease

87. Which of the following vitamins are used for the treatment of mitochondrial disease?

A. (B1 , B2 , C)
B. (B1 , B12 , C)
C. (B1 , B12 ,K)
D. (B1 , B2 , K)
E. (B1 , B2 ,A)

88. Which of the following manifestations is a unique finding in all types of mucopolysaccharidoses?

A. Intellectual disability
B. Coarse facial features
C. Visceromegaly
D. Short stature
E. Dysostosis multiplex

Chapter 10

Metabolic Disorders Answers

QAHTAN ALOBAIDY

1.(C) Neonatal apnea is associated with

- ✓ glycine encephalopathy
- ✓ asparagine synthetase deficiency
- ✓ urea cycle disorders
- ✓ organic acidemias
- ✓ disorders of pyruvate metabolism
- ✓ fatty acid oxidation defects
- ✓ mitochondrial respiratory chain defects

2.(A) Medications causes hyperammonemia

- ✓ Valproic Acid
- ✓ Cyclophosphamide
- ✓ 5-Pentanoic Acid
- ✓ Asparaginase

3.(C) Glycogen storage diseases (GSD) associated with cardiomyopathy

- ✓ GSD type II (Pompe disease)
- ✓ GSD type III (glycogen debrancher enzyme deficiency)
- ✓ *PRKAG2* -related disorders (includes lethal congenital glycogen storage disease of heart)

4.(C) Macrocephaly seen in glutaric acidemia type 1 and Canavan disease.

5.(A)

6.(B)

7.(A) Alopecia is seen in multiple carboxylase deficiency (holocarboxylase synthetase deficiency and biotinidase deficiency) while other choices are causes of ichthyosis.

8.(B)

9.(A) Infants with elevated blood ammonia levels from urea cycle defects tend to have normal serum pH and bicarbonate values; without measurement of blood ammonia, they may remain undiagnosed and succumb to their disease.

10.(B) When blood ammonia, pH, and bicarbonate values are normal, other aminoacidopathies (e.g. hyperglycinemia) or galactosemia should be

considered. Galactosemic infants may also manifest cataracts, hepatomegaly, ascites, and jaundice.

11.(A) The brain is the main organ damaged by PKU, but the exact mechanism of injury remains elusive. Both toxic levels of phenylalanine and insufficient tyrosine may play a role.

12.(B) It is recommended that the blood for screening be obtained in the 1st 24-48 hr of life after feeding protein to reduce the possibility of false negative results, especially in the milder forms of the condition.

13.(C) Pregnant women with phenylalanine hydroxylase (PAH)deficiency who are not on a phenylalanine-restricted diet have a very high risk of having offspring with intellectual disability, microcephaly, growth retardation, congenital malformations, and congenital heart disease. These complications are directly correlated with elevated maternal blood phenylalanine levels during pregnancy.

14.(A) Infants with cofactor BH4 deficiency are identified during screening programs for PKU because of evidence of hyperphenylalaninemia. Plasma phenylalanine levels may be as high as those in classic PKU or may be in the milder range. However, the clinical manifestations of the neurotransmitter disorders differ greatly from those of PKU.

15.(D) Renal involvementis manifested as a Fanconi-like syndrome with hyperphosphaturia, hypophosphatemia, normal–anion gap metabolic acidosis, and vitamin D–resistant rickets. Nephromegaly and nephrocalcinosis may be present on ultrasound examination. Glomerular failure may occur in adolescents and older patients.

16.(A) In untreated patients, the blood level of α-fetoprotein is increased, often greatly, and liver-synthesized coagulation factors are decreased in most patients. Increased levels of α- fetoprotein are present in the cord blood of affected infants, indicating intrauterine liver damage.

17.(E) Neonatal screening for hypertyrosinemia using tyrosine alone detects only a fraction of patients with tyrosinemia type I. Succinylacetone, which is assayed by many neonatal screening programs, has higher sensitivity and specificity than tyrosine and is the preferred metabolite for screening.

18.(D) Symptomatic infants and asymptomatic affected children and adults excrete hawkinsin, 4-hydroxyphenylpyruvic acid, and its metabolites in their urine.

19.(E) The diagnosis is confirmed by finding massive excretion of homogentisic acid on urine organic acid testing. Tyrosine levels are normal. The enzyme is expressed only in the liver and kidneys.

20.(A) Nitisinone efficiently reduces homogentisic acid production in alkaptonuria. If presymptomatic individuals are detected, treatment with nitisinone, combined with a phenylalanine- and tyrosine-restricted diet, seems reasonable, although no experience is available regarding long-term efficacy.
21.(A) In Chédiak-Higashi syndrome, the bleeding tendency is typically mild.
22.(E) Classic homocystinuria treatment with high doses of vitamin B6 (100-500 mg/24 hr) causes dramatic improvement in patients who are responsive to this therapy.
23.(B) Betaine (trimethylglycine, 6 g/24 hr for adults or 200-250 mg/kg/day for children) lowers homocysteine levels in body fluids by remethylating homocysteine to methionine, which may result in elevation of plasma methionine levels. This treatment has produced clinical improvement (preventing vascular events) in patients who are unresponsive to vitamin B6 therapy.
24.(A) Diagnosis is confirmed by measurement of sulfite oxidase and molybdenum cofactor in fibroblasts and liver biopsies, respectively or by DNA studies.
25.(B) Treatment with nicotinic acid or nicotinamide (50-300 mg/24 hr) and a high protein diet results in a favorable response in symptomatic patients with Hartnup disorder.
26.(A) Some children with mild or intermediate forms of maple syrup urine disease MSUD who are treated with high doses of thiamine have dramatic clinical and biochemical improvement. Although some respond to treatment with thiamine at 10 mg/24 hr, others may require as much as 100 mg/24 hr for at least 3 wk before a favorable response is observed.
27.(B) In isovaleric acidemia, acute episodes of metabolic decompensation may occur during a catabolic state, such as infection, dehydration, surgery, or high-protein intake. Acute episodes may be mistaken for diabetic ketoacidosis. Some patients may experience acute and recurrent episodes of pancreatitis.
28.(A) The major clinical manifestation in the rare symptomatic patient is cutaneous photosensitivity**.** The skin becomes rough and red after moderate exposure to the sun, and with greater exposure, a pellagra-like rash may develop. The rash may be pruritic, and a chronic eczema may develop. The skin changes have been reported in affected infants as young as 10 days of age.
29.(D) A clinical condition resembling acrodermatitis enteropathica occurs in affected infants whose plasma isoleucine or valine become very low; addition of isoleucine or valine, respectively, to the diet will hasten the recovery of skin rash.

30.(E) These patients have periodic bouts of fever associated with abdominal pain, vomiting, diarrhea, arthralgia, arthritis, hepatosplenomegaly, lymphadenopathy, and morbilliform rash (even petechiae and purpura), which usually start before 1 yr of age.

31.(B) Clinical manifestations are mostly confined to skin and the nervous system. Atopic or seborrheic dermatitis, candidiasis, alopecia, ataxia, seizures (usually myoclonic), hypotonia, developmental delay, optic nerve atrophy, sensorineural hearing loss, and immunodeficiency resulting from impaired T-cell function may occur. A small number of children with intractable seborrheic dermatitis and partial (15–30% activity) biotinidase deficiency, in whom the dermatitis resolved with biotin therapy, have been reported; these children were otherwise asymptomatic.

32.(A) Clinical findings of propionic acidemia are not specific to this disorder only. In the severe form, patients develop symptoms in the 1st few days of life. Poor feeding, vomiting, hypotonia, lethargy, dehydration, a sepsis-like picture, and clinical signs of severe ketoacidosis progress rapidly to coma and death. Neuroimaging shows that these abnormalities, which often occur after an episode of metabolic decompensation, are the result of damage to the basal ganglia, especially to the globus pallidus. This phenomenon has been referred to as metabolic stroke**.**

33.(C) Diagnosis of nonketotic hyperglycinemia (NKH) can be suspected based on the findings of elevated glycine in plasma or CSF and the abnormal CSF/plasma ratio of glycine. The diagnosis is confirmed using molecular analysis of the NKH-related genes.

34.(B) Treatment with oral activated charcoal, short courses of oral metronidazole, neomycin, or lactulose cause temporary reduction in the body odor. Restriction of fish, eggs, liver, and other sources of choline (e.g., nuts, grains) in the diet significantly reduces the odor. Topical use of acidic soaps (pH 5.5) can also help control the odor.

35.(C) Administration of large doses of pyridoxine reduces plasma level and urinary excretion of oxalate.

36.(A) Treatment of prolidase deficiency is supportive. Infectious complications can be fatal and warrant close and proactive antibiotic management.

37.(C) Three forms of this rare condition have been reported. In the mild form, enzyme deficiency causes glutathione deficiency only in erythrocytes. These patients present with hemolytic anemia without chronic metabolic acidosis and demonstrate high residual activity of glutathione synthetase on enzymatic testing. A moderate form has also been observed in which the hemolytic

anemia is associated with variable degrees of metabolic acidosis and 5-oxoprolinuria. Its severe form is distinguished by presence of hemolytic anemia accompanied by severe acidosis, massive 5-oxoprolinuria, and neurologic manifestations.

38.(A)

39.(B) In approximate order of frequency, symptoms include jitteriness or tremors, apathy, episodes of cyanosis, seizures, intermittent apneic spells or tachypnea, weak or high-pitched cry, limpness or lethargy, difficulty feeding (latching on), and eye rolling. Episodes of sweating, sudden pallor, hypothermia, and cardiac arrest and failure may also occur.

40.(B) Normal newborn values are higher than those of the older child or adult. Levels as high as 100 µmol/L can occur in healthy term infants. An ill infant usually manifests a blood ammonia level >150 µmol/L.

41.(D) At any age, hypoglycemia should be considered a cause of an initial episode of convulsions or a sudden deterioration in psych behavioral functioning or level of consciousness.

42.(C)

43.(E)

44.(D) In patients with CPS1, OTC, or ASS deficiency, acute hyperammonemia attacks may be precipitated by valproate administration.

45.(C) The most consistent laboratory findings are low serum leptin below detectable levels (>90%) and insulin resistance (60%). Platelet count is often moderately high. Otherwise, lipid panels, high-sensitivity C-reactive protein, blood chemistries, liver and kidney function tests, endocrine test, and coagulation tests are generally normal.

46.(B)

47.(E) Two genetic disorders feature hyperornithinemia**:** gyrate atrophy of the retina and hyperammonemia-hyperornithinemia homocitrullinemia syndrome.

48.(B) In pyridoxine (vitamin B6)–dependent epilepsy(hypophosphatasia) ;treatment with vitamin B6 (50-100 mg/day) usually results in a dramatic improvement of both seizures and the EEG abnormalities. High doses of pyridoxine can result in peripheral neuropathy and doses >500 mg/day should be avoided. The pyridoxine dependency and thus the therapy are life-long.

49.(A) The onset of the condition is usually heralded by acute encephalopathic findings, such as loss of normal developmental milestones (head control, rolling over, or sitting), seizures, generalized rigidity, opisthotonos, choreoathetosis, and dystonia caused by acute striatal injury. Affected infants are prone to

development of subdural hematoma and retinal hemorrhage following minor falls and head traumas. This can be misdiagnosed as child abuse.

50.(D) This rare autosomal recessive disorder is caused by a defect in the transport of the cationic amino acids lysine, ornithine, and arginine in both intestine and kidneys. Deficiency of the transporter protein (Y+L amino acid transporter 1) in this condition causes multisystem manifestations, which start initially with gastrointestinal (GI) symptoms.

51.(B) Deficiency of aspartoacylase leads to Canavan disease, a severe leukodystrophy characterized by excessive excretion of N -Acetylaspartic acid and spongy degeneration of the white matter of the brain.

52.(C) The first episode usually does not occur until late childhood or early adulthood. Attacks are frequently precipitated by prolonged exercise. There is aching muscle pain and myoglobinuria that may be severe enough to cause renal failure. Serum levels of creatine kinase are elevated to 5,000-100,000 units/L. Hypoglycemia has not been described, but fasting may contribute to attacks of myoglobinuria.

53.(E) It must be emphasized that although plasma VLCFA levels are elevated in many patients with peroxisomal disorders, this is not always the case. The most important exception is rhizomelic chondrodysplasia punctata, in which VLCFA levels are normal, but plasma phytanic acid levels are increased and red blood cell (RBC) plasmalogen levels are reduced.

54.(D) Other findings are vertebral bodies have a coronal cleft filled by cartilage that is a result of an embryonic arrest. Skin changes such as those observed in ichthyosiform erythroderma are present in approximately 25% of patients. The most decisive laboratory test is the demonstration of abnormally low plasmalogen levels in RBCs and an alteration in *PEX7*.

55.(C)

56.(D) The diagnosis may be confirmed genetically or by measuring LDL-R activity in cultured skin fibroblasts.

57.(C) The disease usually presents during childhood with acute pancreatitis.

58.(C) Other options causes secondary hypercholesterolemia.

59.(D) Causes of reduced high-density lipoprotein**:**

- ✓ Smoking
- ✓ Obesity
- ✓ Type 2 diabetes
- ✓ Malnutrition
- ✓ Drugs: β-blockers, anabolic steroids

60.(E) In such patients, the aim is to prevent episodes of pancreatitis.

61.(A) This autosomal co-dominant disease is associated with HDL-C levels <5 mg/dL. It is caused by mutations in ABCA1, a protein that facilitates the binding of cellular cholesterol to apoA-I. This results in free cholesterol accumulation in the reticuloendothelial system, manifested by tonsillar hypertrophy of a distinctive orange color and hepatosplenomegaly. Intermittent peripheral neuropathy may occur from cholesterol accumulation in Schwann cells.

62.(C) This rare autosomal recessive disease is caused by mutations in the gene encoding microsomal triglyceride transfer protein necessary for the transfer of lipids to nascent chylomicrons in the small intestine and VLDL in the liver. This results in absence of chylomicrons, VLDL, LDL, and apoB and very low levels of plasma cholesterol and triglycerides.

63.(D) Data suggest that obtaining a non-fasting lipid profile can be just as useful in detecting severe genetic dyslipidemias as a fasting lipid profile, and thus can be used as first-line screening in children. Fasting lipid profiles may also be used depending on parental, child, and clinician preference, especially if there is concern for hypertriglyceridemia, since triglycerides are affected more by fasting status.

64.(A)

65.(E) The clinical manifestations of the infantile form of GM1 gangliosidosis may be evident in the newborn as hepatosplenomegaly, edema, and skin eruptions (angiokeratoma).

66.(B) The GM2 gangliosidoses include Tay-Sachs disease and Sandhoff disease; each results from deficiency of β-hexosaminidase activity and lysosomal accumulation of GM2 gangliosides, particularly in the CNS.

67.(C)

68.(B) This classic phenotype is caused by the absent activity of the α-galactosidase A and has an estimated prevalence of approximately 1 in 40,000 males. The angiokeratomas usually occur in childhood and may lead to early diagnosis. They increase in size and number with age and range from barely visible to several millimeters in diameter. The lesions are punctate, dark red to blue-black, and flat or slightly raised. They do not blanch with pressure, and the larger ones may show a slight hyperkeratosis.

69.(D) Early in the course of the classic phenotype, casts, RBCs, and lipid inclusions with characteristic birefringent “Maltese crosses” appear in the urinary sediment.

70.(C) Fucosidosis is a rare autosomal recessive disorder caused by the deficient activity of α-fucosidase and the accumulation of fucose-containing

glycosphingolipids, glycoproteins, and oligosaccharides in the lysosomes of the liver, brain, and other organs.

71.(B) This autosomal recessive white matter disease is caused by a deficiency of arylsulfatase A**,** which is required for the hydrolysis of sulfated glycosphingolipids.

72.(A) This rare autosomal recessive disorder results from the deficiency of the lysosomal enzyme acid ceramidase and the accumulation of ceramide in various tissues, especially the joints.

73.(A) This autosomal recessive lysosomal storage diseases result from deficiency of lysosomal acid lipase and accumulation of cholesterol esters and triglycerides in histiocytic foam cells of most visceral organs.

74.(D) The biochemical characteristics of type I GSD are hypoglycemia, lactic acidosis, hyperuricemia, and hyperlipidemia. Despite marked hepatomegaly, the liver transaminase levels are usually normal or only slightly elevated. Easy bruising and epistaxis are common and are associated with a prolonged bleeding time as a result of impaired platelet aggregation and adhesion.

75.(C) Electrocardiographic findings include a high-voltage QRS complex, Wolff-Parkinson-White (WPW) syndrome, and a shortened PR interval. Echocardiography reveals thickening of ventricles and/or the intraventricular septum and/or left ventricular outflow tract obstruction. Muscle biopsy shows the presence of vacuoles that stain positively for glycogen; acid phosphatase is increased, presumably from a compensatory increase of lysosomal enzymes.

76.(B) The standard diagnosis for GSD V includes a muscle biopsy to measure glycogen content as well as enzyme and sequencing of PYGM . An ischemic exercise test offers a rapid diagnostic screening for patients with a metabolic myopathy. Lack of an increase in blood lactate levels and exaggerated blood ammonia elevations indicate muscle glycogenosis and suggest a defect in the conversion of muscle glycogen or glucose to lactate.

77.(D)

78.(E) The renal calculi, composed of 2,8-dihydroxyadenine, are radiolucent, soft, and easily crushed. These stones are not distinguishable from uric acid stones by routine tests but require high-performance liquid chromatography (HPLC), ultraviolet (UV) light, infrared light, mass spectrometry, x-ray crystallography, or capillary electrophoresis for diagnosis, particularly to distinguish from stones in HPRT deficiency

79.(B)

80.(D) Both types of Morquio syndrome are characterized by short-trunk dwarfism, a skeletal dysplasia that is distinct from other mucopolysaccharidoses, and preservation of intelligence.
81.(B) The affected infant is otherwise asymptomatic. Treatment is dietary restriction of galactose.
82.(C) Sanfilippo syndrome is characterized by slowly progressive, severe CNS degeneration with mild somatic disease. Onset of clinical features usually occurs at age 2–6 yr in a child who previously appeared normal.
83.(A) Coarse facial features, short stature, dysostosis multiplex, joint stiffness, and intellectual disability manifest between 2 and 4 yr of age.
84.(A) Leigh disease (subacute necrotizing encephalomyelopathy) is a heterogeneous neurologic disease characterized by demyelination, gliosis, necrosis, relative neuronal sparing, and capillary proliferation in specific brain regions.
85.(A) Sialidosis is an autosomal recessive disorder that results from the primary deficiency of neuraminidase because of mutations in the gene (*NEU1)* that encodes this protein.
86.(A)
87.(A) Mitochondrial cocktails of vitamins and supplements variably include vitamins (B1, B2, C), antioxidants (CoQ10, lipoic acid, vitamin E), and metabolic modifiers (creatine, L -carnitine, L -arginine, folinic acid).
88.(E)

Chapter 11

The Fetus and the Neonatal Infant Questions

AHMED TAWFIQ

1. A concerned mother of 5-day-old precious baby boy, product of IVF with uneventful normal vaginal delivery describes a very brief attacks of rapid rhythmic movement of one of lower limbs occurs during sleep only. Examination was normal.
What is the **MOST** appropriate plan of management?

A. Order initial investigation workup for seizure
B. Admit the baby to neonatal care unit for video monitoring
C. Send for EEG record
D. Start empirical anticonvulsant therapy
E. Reassurance

2. What is the major cause of mortality during infancy?

A. Unsafe sleep practices
B. Congenital malformations
C. Complications of prematurity
D. Accidents
E. Non-accidental injuries and mal-treatment

3. Which of the following organs is affected more often by congenital anomalies?

A. GI tract
B. CNS
C. Heart
D. Kideny
E. Eye

4. Presence of bilateral red eye reflex in a newborn suggest

A. cataract
B. chorioretinitis
C. retinopathy of prematurity

D. persistent hyperplastic primary vitreous
E. absence of intraocular pathology

5. What is the **MOST** prominent sign of neuromuscular disease at birth?
A. Arthrogryposis
B. Microcephaly
C. Hydrocephaly
D. Dislocated hips
E. Undescended testes

6. What is the recommend time for delayed cord clamping in a vigorous full-term infant?
A. 15 seconds
B. 30-60 seconds
C. 1-2 minutes
D. 2-4 minutes
E. Until umbilical pulsations stops

7. You are preparing your tool-set for neonatal resuscitation program (NRP); which tool would be used for oral suction?
A. Mucous extractor
B. Soft cloth
C. Bulb syringe
D. Large bore catheter
E. No tool is required as suction is not needed

8. Which of the following drugs is contraindicated in a nursing mother?
A. Thiouracil
B. Aspirin
C. Amiodarone
D. Domperidone
E. Phenobarbitone

9. Which of the following medical conditions is an absolute contraindication for breast feeding?
A. Active TB (mother on treatment since 21 days)
B. Genital herpes infection
C. HumanT-cell lymphotropic virus type 2

D. Hepatitis C virus infection
E. Varicella zoster infection

10. Which of the following is a leading cause of prematurity?
A. Multiple gestations
B. Cervical shortening
C. Genital infection
D. Presence of fetal fibronectin in cervicovaginal secretions
E. Premature rupture of membranes

11. Polyhyramnios is diagnosed if amniotic fluid index is more than
A. 4 cm
B. 14 cm
C. 24 cm
D. 34 cm
E. 44 cm

12. Which of the following conditions is associated with polyhyramnios?
A. Twin-twin transfusion (donor)
B. Cystic adenomatoid lung malformation
C. Fetal akinesia syndrome
D. Prune-belly syndrome
E. Angiotensin-converting enzyme inhibitors

13. Fetal bradycardia is defined as fetal heart beats less than
A. 50 bpm
B. 70 bpm
C. 90 bpm
D. 110 bpm
E. 130 bpm

14. Which of the following is the **MOST** common form of congenital infections?
A. Cytomegalovirus
B. Rubella
C. Herpes virus
D. Syphilis
E. Toxoplasma

15. A young lady exposed to diagnostic radiation of about 0.1 RAD (took chest x-ray) at 8^{th} wk of gestation; she develops a miscarriage 4 days later, she complains to local health authority.
What is your opinion?

A. Approve the case
B. Dismiss the case
C. Postpone the case meanwhile taking opinion of radiology expert
D. Assessment of mother radiation exposure by biodosimetry
E. Send mother for hematologic screen if negative dismiss the case

16. You are attending a labor of 23-year-old primigravida; the cardiotocography shows slowing of fetal heart rate, beat-to-beat variability declining, and appearance of late deceleration pattern.
Of the following, the **MOST** appropriate action is to

A. start infusion of tocolytic medications
B. start oxytocin infusion
C. counsel parents about possibility of hypoxic-ischemic encephalopathy
D. administer high O_2 concentration to the mother
E. prepare for emergency cesarean section

17. Which of these parameters is a poor predicator in hypoxic-ischemic encephalopathy injury?

A. Low (4-6) 10 min Apgar score
B. Need for CPR in the delivery room
C. Delayed onset of spontaneous breathing > 5 min
D. Seizures onset > 48 hr
E. Absence of EEG findings of burst suppression pattern

18. According to Sarnat classification of hypoxic-ischemic encephalopathy, a term-infant who appears to be hyperactive with mydriatic pupil, exaggerated Moro reflex and myoclonus has

A. ongoing subtle seizure activity
B. HIE stage I
C. HIE stage II
D. HIE stage III
E. unclassified

19. You are assessing a 3-day-old term neonate with hypoxic-ischemic encephalopathy.
Of the following, the **MOST** sensitive test that helps in classification and management is

A. brain MRI
B. brain CT scan
C. brain ultrasound
D. brain and bowel ultrasound
E. brain Doppler study

20. What is the targeted core rectal temperature in therapeutic hypothermia for hypoxic-ischemic encephalopathy?

A. 32 °C
B. 32.5 °C
C. 33 °C
D. 33.5°C
E. 34 °C

21. What is the ideal timing of initiation of therapeutic hypothermia in hypoxic-ischemic encephalopathy?

A. As early as first 30 minutes of life
B. First 6 hours of life
C. First 12 hours of life
D. First 24 hours of life
E. First 48 hours of life

22. What is the duration of therapeutic hypothermia in hypoxic-ischemic encephalopathy?

A. 12 hours
B. 24 hours
C. 48 hours
D. 72 hours
E. 96 hours

23. A useful adjunct to therapeutic hypothermia in hypoxic-ischemic encephalopathy which shows promising results in reducing brain injury and short term motor outcome is

A. erythropoietin

B. high dose phenytoin
C. phenobarbital coma
D. lidocaine infusion
E. amiodarone infusion

24. A full-term baby boy suffered from severe birth asphyxia and hypoxic-ischemic encephalopathy with recurrent seizures not responding to phenobarbitone.
The **MOST** appropriate next choice is to use

A. loading dose phenytoin
B. levetiracetam
C. pyridoxine
D. midazolam
E. lidocaine

25. What is the **MOST** important radiological finding in MRI correlating with poor prognosis in hypoxic-ischemic encephalopathy?

A. Loss of cerebral cortical gray-white differentiation
B. Parasagittal cerebral cortex lesion
C. Basal ganglia/thalamic lesions
D. Occipital lobe lesions
E. Cerebellar Lesions

26. The brain involvement of periventricular area in process of hypoxic-ischemic encephalopathy can lead to future development of

A. cognitive delay
B. visual and auditory processing difficulty
C. dystonia
D. seizure disorder
E. spastic diplegia

27. The brain involvement of parasagittal area in process of hypoxic-ischemic encephalopathy can lead to future development of

A. spastic quadriparesis
B. dystonia
C. seizure disorder
D. ataxia
E. pseudobulbar palsy

28. What is the initial priority in treatment of hypoxic-ischemic encephalopathy?

A. Starting therapeutic hypothermia
B. Correction of acid base status
C. Hemodynamic stabilization
D. Control of seizures
E. Starting parentral nutrition

29. What is the **MOST** common acquired neuronal injury in newborns?

A. Phrenic nerve
B. Brachial plexus
C. Facial nerve
D. Spinal cord transection
E. Horner syndrome

30. You are resuscitating a full term neonate who is depressed at birth, you need to stat bag and mask resuscitation. The fraction of inspired O2 that is needed should be

A. 21%
B. 40%
C. 60%
D. 80%
E. 100%

31. An ex-28-week-preterm baby, at 3 weeks of life is recovering from RDS, extubated since 2 wks, he is developing frequent apneic attacks with bradycardia and desaturation. The baby general condition is good with good capillary refill; he is not pale and appears alert. He is on increasing feeding protocol, TPN had stopped since 4 days, increasing weight, his acid-base balance is normal with euglycemia.
Of the following, the **MOST** appropriate action is to

A. do septic screen and start empirical antibiotics
B. reintubate the baby and put on low ventilatory settings
C. start nasal continuous positive airway pressure (nasal CPAP, 3-5 cm H2O)
D. start heated humidified high-flow nasal cannula (HFNC, 1-4 l/min)
E. start caffeine

32. A well preterm baby recovered from RDS of prematurity, develops gastroesophageal reflux (GER). You are anticipating possible apneic episodes.
Of the following, the **MOST** appropriate option for management is to start

A. omeprazole
B. ranitidine
C. combination of omeprazole and ranitidine
D. domperidone
E. monitoring

33. A near-term baby boy delivered to diabetic mother with no risk factors apart from mother was being poor attendant to antenatal care; the baby birth weight is 2.8 kg. He develops tachypnea and desaturation few hours after birth with no respiratory distress, his heart rate 170 bpm, respiratory rate 72/min, blood pressure 55/35 mmHg. He has hepatomegaly and no cardiac murmur. Chest X-ray shows complete white-out. Blood gas analysis shows mixed respiratory and metabolic acidosis with hypoxia. The baby admitted to NICU, received INSURE therapy with minimal response, he is euglycemic now.
Of the following, the **MOST** appropriate action is to

A. give 2nd dose of survanta
B. intubate and start mechanical ventilation
C. collect septic screen and start empirical antibiotics
D. order an Echocardiography
E. consult metabolic/endocrine team

34. What is the **BEST** type of steroid that should be used antenatally to decrease incidence of RDS of prematurity?

A. Dexamethasone
B. Betamethasone
C. Methylprednisolone
D. Hydrocortisone
E. Clobetasone

35. A 2-day-old full-term baby boy with difficult labor has unilateral scalp swelling which is firm, tense with palpable rims that does not extend to contralateral side, and it has central depression. The baby is hemodynamically stable and neurologically normal; he has jaundice over the face and upper trunk.
Of the following, the **MOST** appropriate action is to

A. closely follow the results of PCV and TSB
B. reassure the family and give appointment follow up within 1 week
C. reassure the family about the benign nature which may disappear after 2weeks-3months
D. order radiological assessment
E. refer to pediatric neurologist

36. A full-term baby girl delivered by vacuum-assisted delivery, attention was coming from accompanied attendant that noticed an increasing scalp swelling overtime, you found a fluctuant mass that straddles cranial sutures involving the scalp and some of subcutaneous tissues of the neck. The baby looks pale.
Of the following, the **MOST** appropriate action is to

A. order serial PCV tests and treat the baby accordingly
B. keep the baby beside the mother and informed staff nurse to monitor vitals and the lesion size
C. admit the baby to NICU
D. reassure the parents about benign nature of lesion
E. anticipate the expected hyperbilirubinemia and anemia by starting single phototherapy and serial PCV

37. You are counseling a mother of large for date baby with difficult labor who was admitted to NICU for complication of meconium aspiration syndrome; now he is improved and the team is willing to discharge the baby; mother is raising a point about the result of head MRI which shows a small subdural hemorrhage in posterior fossa; she asks about the prognosis.
Which one of the following represents the **MOST** appropriate answer?

A. We will arrange an appointment in pediatric neurology clinic for follow-up
B. We will assess neurological status of the baby in next visit and we will order radiological assessment accordingly
C. Serial of MRI will decide the need for evacuation of the lesion
D. Such lesions will calcify leaving asymptomatic cyst which needs no intervention
E. These lesions causes no residual neurological problem

38. All preterm babies should be subjected to cranial ultrasound for detection of intraventricular hemorrhage during the first 3-7 days of life if delivered below

A. 24 weeks of gestation

B. 26 weeks of gestation
C. 28 weeks of gestation
D. 30 weeks of gestation
E. 32 weeks of gestation

39. A 28-week-gestational age preterm baby with a birth weight of 1000 gram received prophylactic surfactant at birth, eventually he needs to be intubated and ventilated on SIMV mode with moderate settings, TPN started on day 1. On day 2 his PCV shows drop from 40 to 28, and he developed hypotension and metabolic acidosis. He received fluid resuscitation and packed RBCs transfusion. Repeated blood gas shows persistence of metabolic acidosis.
Of the following, the **MOST** likely explanation is

A. development of early onset sepsis
B. underlying inborn error of metabolism
C. development of anemia of prematurity
D. development of intracranial hemorrhage
E. TPN related complication

40. A 32-week-preterm baby with 1200 grams birth weight, developed respiratory distress and received rescue INSURE therapy since 4 hours. Now he is on nCPAP with pressure of 6 cm of water and FiO_2 of 60, his saturation ranging between 85-88%; other hemodynamic parameters are accepted.
Of the following, the **MOST** appropriate next step in the management is to

A. keep the nCPAP parameters the same to avoid O_2 toxicity
B. increase nCPAP parameters
C. consider the second dose of surfactant
D. start mechanical ventilation
E. use inhaled nitric oxide

41. Assisted reproductive technology (in vitro fertilization) can increase the risk of

A. Beckwith-Wiedemann syndrome
B. neural tube defect
C. Cornela de-lange syndrome
D. major chromosomal disorders
E. Peirr-robin sequence

42. The **MOST** effective way to increase the O2 saturation in ventilated infant or neonate is by increasing the

A. inspiratory time
B. ventilator rate
C. positive end-expiratory pressure (PEEP)
D. peak inspiratory pressure (PIP)
E. FiO_2

43. The currently recommended oxygen saturation targets in ventilated neonates is

A. 81-85%
B. 86-90%
C. 91–95%
D. constantly above 95%
E. 100%

44. You are preparing to extubate a preterm baby.
Which premedication can enhance the success of extubation?

A. Methylxanthine
B. Dexamethasone
C. Betamethasone
D. IV atropine
E. Endotracheal atropine

45. Which of the following medications proved to have a role in decreasing the risk of development of bronchopulmonary dysplasia in preterm babies?

A. Inhaled corticosteroids therapy
B. Diuretics
C. Vitamin A
D. Low dose systemic dexamethasone
E. Caffeine

46. In delivery room, you received a 28 weeks gestational age preterm baby with a birth weight of 1.28 kg. His APGAR score is good, he needs only routine measures of resuscitation; he develops mild grunting, and his saturation is maintained with minimal nasal O_2.
Of the following, the **MOST** appropriate step in management is to

A. give prophylactic surfactant therapy

B. wait for the signs of severe respiratory distress then give rescue dose of surfactant
C. start nasal CPAP
D. continue on low flow nasal O2 cannula as much as O2 saturation is maintained
E. transfer to neonatal care unit by transport incubator and decision should be made by senior colleague

47. An 18-day-old, ex-preterm baby (30 wks of gestation), his current weight 1.44 kg, had weaned from nasal CPAP, and on increasing feeding protocol. The staff nurse reported an increase need for nasal O_2, increase work of breathing, and decrease urine output < 1ml/kg/hr. By examination, you have noticed bounding pulse and hepatomegaly.
Of the following, the **MOST** appropriate action is to
A. order an echocardiography
B. order renal function and monitor urine output
C. administer diuretics
D. administer ibuprofen
E. refer to pediatric cardiology department

48. You are explaining the effectiveness of supportive measures in spontaneous closure of PDA in extremely preterm infants to a junior colleague.
Which of the followings represent the **MOST** appropriate answer?
A. Most of the infants eventually will needs cyclooxygenase inhibitors
B. About half of them will be cured
C. Most of them will be cured
D. Surgical intervention is high in those who treated by this way
E. It is no longer considered as an option for treatment

49. What is the **MOST** common etiology of tachypnea in newborns?
A. Transient tachypnea of the newborn
B. Hyaline membrane disease
C. Meconium aspiration syndrome
D. Amniotic fluid aspiration
E. Early onset sepsis

50. What is the **MOST** important effective step in prevention of meconium aspiration syndrome?

A. Amnioinfusion
B. Intrapartum nasopharyngeal suction
C. Intubation and suctioning of depressed meconium stained baby
D. Monitoring of fetal distress
E. Liberal rules for cesarean sections

51. A full-term baby boy delivered by cesarean section because of meconium stained amniotic fluid, developed respiratory distress and required mechanical ventilation ultimately. On day 4 discovered to have systolic murmur, his O2 saturation is ranging between 82% and 96%. He had frequent weaning failure from ventilation.
Of the following, the **MOST** likely etiology of cardiac murmur is
A. associated cyanotic congenital heart disease
B. patent ductus arteriosus
C. pulmonary hypertension
D. asphyxiated cardiomyopathy
E. functional mitral regurgitation

52. A full-term baby boy delivered by uneventful simple vaginal delivery, had good APGAR score and discharge home. At the age of 20 hours developed refusal to feed, rapid breathing, and bluish discoloration of the face. He had no dysmorphic features, HR 185 beat/min, RR 70 breath/min, O2 saturation 68% which sometimes increases to 90%, blood gas analysis shows hypoxemia with CO_2 retention. Mother was G4 P3, attending antenatal care regularly but had used ibuprofen frequently for headache without medical consultation. Investigations shows RBS 55mg/dl; Hb 20 gm/dl; WBC 24,000 mm^3 mainly neutrophils; echocardiography shows no congenital heart disease; blood culture and metabolic screen is pending.
Of the following, the **MOST** likely diagnosis is
A. bacterial sepsis
B. concealed congenital heart disease (distal aortic arch anomaly)
C. inborn error of metabolism
D. persistent pulmonary hypertension
E. pulmonary hypoplasia

53. A post-term baby girl ventilated for persistent pulmonary hypertension is suffering from hypotension which is persistent even after adjustment of basic metabolic needs, volume replacement and vasopressor agents.

Of the following, the **MOST** appropriate step in management is to

A. use another bolus of volume replacement therapy
B. use of hydrocortisone
C. use of desmopressin
D. send for serum cortisone
E. order urgent brain ultrasound

54. You are receiving a newborn in delivery room with prenatal diagnosis of diaphragmatic hernia. He delivered with good APGAR score, vitals are normal with normal O2 saturation.

Of the following, the **MOST** appropriate action is to

A. administer O_2 through face mask
B. pass a wide bore nasogastric tube to deflate the stomach
C. intubate and gently ventilate the baby with standards of permissive hypercapnia
D. contact with pediatric surgeon
E. transfer to neonatal care unit

55. What is the best time for operation in ventilated infant with congenital diaphragmatic hernia?

A. As early as possible
B. First 48 hours
C. After 48 hours
D. After neonatal period
E. After total weaning from ventilation

56. An 18-month-old toddler, his weight 12.5kg, with normal development admitted for recurrent lobar infiltration of right lower lobe, chest X-ray shows elevated right dome of diaphragm and paradoxical movement during fluoroscopy.

Of the following, the **MOST** likely cause of recurrent lobar infiltration is

A. eventration of right dome of diaphragm
B. lobar sequestration
C. lung hypoplasia
D. phrenic nerve palsy
E. spinal muscular atrophy

57. A 48-hour-old full-term baby boy, his weight 1.8kg, didn't pass motion yet. He develops an increasing abdominal distension and occasional vomiting, his erect abdominal X-ray shows dilated large bowl, and PR examination is unremarkable. Mother is G3P3, had severe preeclapmsia and received magnesium sulphate, other siblings had no chronic medical problem.
Of the following, the **MOST** likely diagnosis is

A. meconium plug
B. meconium ileus
C. necrotizing enterocolitis
D. hirschsprung disease
E. cystic fibrosis

58. What is the characteristic X-ray finding in abdominal contrast study done for a neonate with meconium ileus?

A. Coil spring sign
B. Dilated distal bowl
C. Microcolon
D. Whirl sign
E. Double bubble sign

59. The usual onset of necrotizing enterocolitis in VLBW preterm babies is at

A. first few hours of life
B. first 48 hours of life
C. first week of life
D. 2^{nd} and 3^{rd} weeks of life
E. 4^{th} week of life

60. What is the **MOST** effective preventive strategy for necrotizing enterocolitis?

A. Human milk
B. Probiotics
C. Synbiotics
D. Pretreatment with H2 receptors antagonist
E. Good antibiotic coverage for bacteria that colonize intensive care unit

61. A well active full-term baby boy, his blood group is B+ve; delivered to 23-year-primigravida mother, her blood group is O+ve with good antenatal care and had no perinatal problems. His complete blood count shows, HB 13 g/dl,

MCV 92 fL, MCHC 36. The blood film, Coomb's test, and serum bilirubin are pending.
From the available data, what is the **MOST** probable diagnosis?

A. ABO incompatibility
B. α-thalassemia trait
C. Maternal iron deficiency
D. Hereditary spherocytosis
E. Normal values for age

62. A primimother, 36 wks gestations who was well throughout pregnancy, reported decreased fetal movements to her obstetrician, for which emergency cesarean section had been done. Her baby was pale and limp and required full resuscitation including volume replacement.
What laboratory test should be done for precise diagnosis?

A. Urgent baby complete blood count and peripheral blood smear
B. Kleihauer-Betke test using maternal blood
C. Reticulocyte count, MCV and MCHC of baby sample
D. Baby and mother Coomb's test
E. Flow cytometry using maternal blood

63. A 3-day-old full-term baby girl presents with history of increasing neonatal jaundice. Examination shows active, pale baby, with splenomegaly. Her investigations show HB 11 gm/dl, MCV 102, MCHC 37, direct antiglobuline test -ve, indirect bilirubin 18 mg/dl, direct bilirubin 0.4mg/dl, blood group B +ve; hematologist reported increased reticulocyte count and bizarre RBCs morphology featuring acanthocytes. The baby developed a stormy course, treated aggressively with phototherapy and frequent blood transfusions throughout neonatal period. Parents are consanguineous and are free from chronic medical problems; mother blood group is O+ve.
Of the following, the **MOST** likely diagnosis is

A. hereditary spherocytosis with incomplete penetrance
B. pyruvate kinase deficiency
C. G6PD deficiency
D. ABO blood groups incompatibility
E. congenital abetalipoproteinmia

64. What is the suggested PCV transfusion threshold for a 5-day-old VLBW baby who has no respiratory support?

A. 40
B. 35
C. 30
D. 25
E. 20

65. Which of the following factors reduce mother sensitization in hemolytic disease of fetus and newborn HDFN resulting from Rh incompatibility?

A. ABO incompatibility
B. Increasing order of pregnancies
C. Type E Rh antigen
D. IgG type maternal antibody response
E. Larger volume of fetomaternal blood transfusion

66. Which of the following is a recognized complication of hemolytic disease of fetus and newborn HDFN?

A. Hypocalcemia
B. Hypoglycemia
C. Hyperkalemia
D. Sepsis
E. Hyperthermia

67. A G5P4 mother who had previous 2 affected babies with hemolytic disease of fetus and newborn (due to Rh incompatility) is asking about how to know whether her coming baby is going to be sffected or not.
Which of the following tests you would use inresponse to her question?

A. Amniocentesis
B. Umbilical blood sampling
C. Fetal DNA from maternal serum
D. Chorionic villous sampling
E. Rising RhD antibody titers

68. A sensitized Rh-negative mother during second trimester is found to have RhD antibody titers of ≥1:16 (15 IU/mL).
What is the **MOST** appropriate next step in management?

A. Give follow up visit after 2 weeks with repeat RhD antibody titer
B. Order umbilical blood sampling
C. Order fetal blood transfusion

D. Administration of Rh-immunoglobulin
E. Order Doppler study of the middle cerebral artery

69. What is the **MOST** appropriate statement that describes the trigger point of pathophysiology of hydrops fetalis?

A. Severe anaemia that causes hypoalbuminemia
B. Extramedullary hematopoiesis that ultimately leads to hepatocellular dysfunction
C. Anaemia that leads to right sided heart failure
D. Renal failure secondary to fetal renal vein thrombosis because of severe hemolysis
E. Generalized tissue hypoxia

70. You are a team leader expecting delivery of a baby with hydrops due to HDFN. The baby is delivered and initial steps of resuscitation were done successfully. He has pallor, upper and lower limbs edema, mild ascites, tachypnea and tachycardia. You started O2 and other supportive therapy.
Which of the following is the **MOST** appropriate step in the management?

A. Whole fresh compatible blood transfusion
B. Small packed RBCs transfusion
C. Small dose of diuretics
D. 20% albumin transfusion
E. Assisted ventilation

71. You are treating a 6- hour-old full term neonate with hemolytic disease of fetus and newborn HDFN due to Rh incompatibility; he received small packed RBCs transfusion, and he is stable right now. His serum bilirubin 8mg/dl (136mmol/l) and hemoglobin 12g/dl.
Of the following, the **MOST** appropriate next step in the management is to

A. proceed for immediate exchange transfusion
B. plot the serial results of total bilirubin on exchange transfusion standard graph and manage accordingly
C. start intensive phototherapy
D. administer IV immunoglobulin
E. administer 20% albumin

72. In which of the following scenarios an immediate exchange transfusion is an indication? (1mg of bilirubin=17mmol/l of bilirubin)

A. A 5 days full-term baby with jaundice, fever, lethargy and hypertonia
B. A 24 hours full-term with Rh negative mother, bilirubin 13 mg/dl, HB 12gm/dl
C. A 48 hours 35 wks high risk baby with bilirubin of 15 mg/dl
D. A 48 hours asphyxiated baby with bilirubin of 15 mg/dl
E. A 72 hours standard risk full-term baby with bilirubin 20 mg/dl

73. Which of the following is the **MOST** common cause for neonatal polycythemia in term infant?
A. Twin-twin transfusion for the recipient
B. Maternal-fetal transfusions
C. Post-maturity
D. Small-for-gestational age
E. Delayed clamping of the umbilical cord

74. A 3-day-old low birth weight neonate brought to emergency room with feeding disturbance and lethargy. Examination shows alert baby with mild tachypnea, cyanosis, and jaundice affecting the face and trunk. His RBS is low, and capillary hematocrit is 68%. No risk factors in the history suggest sepsis.
Of the following, the **MOST** appropriate step in management is to
A. measure central hematocrit
B. start partial exchange transfusion
C. start empirical antibiotics
D. start hydration with normal saline
E. order echocardiography

75. At which age vitamin K–dependent coagulation factors reach adult ranges?
A. End of first week of life
B. End of neonatal period
C. At 4 months of life
D. At 6 months of life
E. At end of first year of life

76. A well and active 3-day-old full term breast fed baby delivered at home, presents to OP clinic with fresh blood in stool, and some nasal bleed.
Of the following, the **MOST** likely diagnosis is
A. early form of hemorrhagic disease of the newborn
B. classic form of hemorrhagic disease of the newborn

C. factor VIII deficiency
D. factor IX deficiency
E. neonatal thrombocytopenia

77. A pregnant mother with familial hypercholesterolemia on cholesterol lowering medication consults you about any possible risk of taking such medications.
What is your **BEST** advice for her?

A. Such medications are safe and causes no possible harm to coming baby
B. We should stop such medications right now
C. A plan is needed for you and your baby to control hypercholesterolemia
D. A plan is needed for you and your baby to control possible bleeding disorder
E. A lipid profile is needed for baby after birth

78. What is the **MOST** common presentation for late onset hemorrhagic disease of the newborn?

A. Cutaneous bleeding
B. Bleeding from injection site
C. Ear-nose-throat and mucosal bleeding
D. Gastrointestinal bleeding
E. Intracranial bleeding

79. Which of the following causes of non-immune hydrops can be treated prenatally?

A. Supraventricular tachycardia
B. Gaucher disease
C. Acardiac twin
D. Fetal akinesia syndromes
E. α-thalassemia

80. What is the **MOST** common remnant of the omphalo-mesenteric duct OMD?

A. Umbilical polyp
B. Umbilical granuloma
C. Umbilical sinus
D. Umbilical fistula
E. Meckel diverticulum

81. What is the **BEST** treatment of umbilical granuloma?

A. Topical use of silver nitrite
B. Topical use of alcohol pads
C. Topical use of povidone-iodine
D. Ligation of the base and excision
E. Frequent soap and water toilet

82. A 5-day-old newborn presents with lethargy, mild umbilical discharge and redness of peri-umbilical tissue.
Of the following, the **MOST** appropriate antimicrobial treatment is

A. Oral amoxcillin
B. Vancomycin pulse amikacin
C. Ampicillin plus gentamicin
D. Vancomycin
E. Cefuroxime

83. A young parents counseled you about the possible risk of antidepressant medication (fluoxetine) during pregnancy.
What is the **MOST** appropriate answer?

A. It's safe to use such medication
B. It's acceptable to be used as so far has no recorded risk registered
C. We can use it with caution, if its benefits outweigh its risk
D. We can use it only in life threatening emergencies
E. We should not use it

84. You are taking first on-call duty in postnatal ward when staff nurse called you for an 8-hour-old baby who has fever, weak suckling, irritability, tremor, and weak cry. Mother is giving history of taking fluoxetine started in third trimester for major depression.
Of the following, the **MOST** appropriate action/advice is to

A. advice the mother about proper positionimg and attachment on breast
B. explain this is not related to side effects of medication
C. order RBS and advice for small frequent breast feedings
D. stop breast feeding and start bottle feeding
E. admit the baby to NICU

85. Which of the following is differentiating primordial dwarfism from pituitary dwarfism?

A. It had in utero growth failure
B. It has hyperbilirubinemia
C. It has hypoglycemia
D. It has micropenis at birth
E. It has associated birth defects

86. A colleague is asking about the maximum time limit to start treatment in congenital hypothyroidism and by which we can still achieve normal development. Your answer would be

A. first 7 days of life
B. first 14 days of life
C. first 21 days of life
D. first 30 days of life
E. first 2 months of life

87. A 7-day-old baby with history of traumatic delivery develops irritability, vomiting, increased tone, and constipation. A firm purple nodule can be appreciated on the trunk. All laboratory work up shows normal results apart from hypercalcemia.
Of the following, the **MOST** likely diagnosis is

A. parathyroid hyperplasia
B. subcutaneous fat necrosis
C. idiopathic hypercalcemia
D. maternal hypoparathyroidism
E. transient hyperparathyroidism

88. What is the confirmative diagnostic test for salt wasting congenital adrenal hyperplasia?

A. Decreased serum glucose
B. Elevated serum potassium
C. Reduced serum cortisone
D. Elevated 17-hydroxyprogesterone
E. Reduced serum aldosterone

89. A female infant with webbing of the neck, lymphedema, hypoplasia of the nipples, cutis laxa, and low hairline at the nape of the neck, low-set ears, high-arched palate, and cubitus valgus should be suspected of having

A. Noonan syndrome

B. Turner syndrome
C. William syndrome
D. Edward syndrome
E. Patau syndrome

90. What is the recommended goal for periconceptual HbA1c in pregestational diabetic mothers to achieve the best outcomes for the mother and the baby?

A. <5%
B. <5.5%
C. <6%
D. <6.5%
E. <7%

91. A full-term macrosomic baby, a product of normal vaginal delivery to a mother with gestational diabetes develops tachypnea, tachycardia, and decreased O_2 saturation. Chest X-ray shows cardiomegaly with pulmonary congestion, and echocardiography shows severe asymmetrical septal hypertrophy with systolic anterior motion (SAM) of anterior mitral leaflet. Baby is on O_2, supportive measures and low dose diuretics, but he is still symptomatic.

Of the following, the **MOST** appropriate action is to start

A. digitalization
B. dopamin
C. propranolol
D. captopril
E. amlodipine

92. A fullterm macrosomic baby boy delivered by difficult and assisted normal vaginal delivery. History reveals, mother had pregestational diabetes with fair control and the baby had passed motion only once since birth. On 36 hours of life, he develops decreased activity and feeding, and red color urine. Examination shows fair hydration, HR 140 b/min, BP 100/75 mmHg, and left sided flank mass. Investigations; RBS 44 mg/dl, TSB 12.9 mg/dl, blood gases normal, HB 20 gm/dl, WBCs 22000/mm^3, and platelets 55000/mm^3.

Of the following, the **MOST** likely diagnosis is

A. small left colon syndrome
B. Hirschsprung disease
C. congenital infantile polycystic kidney disease

D. unilateral dysplastic kidney
E. renal vein thrombosis

93. In mothers who are elected for cesarean section, the **BEST** solution to solve postoperative breast feeding delay problem is to
A. use prenatally expressed breast milk
B. arrange for spinal anesthesia
C. use oral dextrose gel
D. use formula milk with spoon
E. use breast milk bank

94. What is the pre-feeding target plasma glucose concentration in first 48 hrs of life in infants of diabetic mothers?
A. ≥30 mg/dl
B. ≥35 mg/dl
C. ≥40 mg/dL
D. ≥45 mg/dL
E. ≥50 mg/dL

95. What is the plasma glucose level at which IV glucose is indicated in neonatal hypoglycemia in first 4 hours of life?
A. <20mg/dL
B. <25mg/dL
C. <30mg/dL
D. <35mg/dL
E. <40mg/dL

96. Screening for asymptomatic hypoglycemia during first day of life among at-risk infants is indicated for
A. post-term infants
B. infants with congenital infection
C. infants of obese mothers
D. infants with birth asphyxia
E. infant with mothers on beta blockers

97. Which of the following markers have ability to identify neonates with serious bacterial infection?
A. Erythrocyte sedimentation rate

B. Immature-to-total neutrophil count
C. Serum C-reactive protein
D. Procalcitonin
E. Thrombocytopenia

98. What is the empirical antibiotic therapy for early onset neonatal sepsis?
A. Ampicillin
B. Ampicillin + Aminoglycoside
C. Vancomycin + Aminoglycoside
D. Cefotaxime + Aminoglycoside
E. Cefotaxime

99. You are explaining the non- antimicrobial treatment strategies and the role of recombinant GM-CSF and IVIG in treatment of neonatal sepsis to a junior colleague.
Which of the following is the **MOST** appropriate sentence?
A. Recent studies shows encouraging results of their use
B. Recent studies has established their role
C. Studies shows a lot of adverse effects during their use
D. There is insufficient evidence to support their clinical use
E. There is no role in treatment of neonatal sepsis

100. Which of the following is true regarding the dose of empiric antibiotic regime in early onset sepsis?
A. Ampicillin 50 mg/kg/dose BID, gentamicin 5mg/kg/dose BID
B. Ampicillin 50 mg/kg/dose BID, gentamicin 5mg/kg/dose OD
C. Ampicillin 100 mg/kg/dose BID, gentamicin 4mg/kg/dose OD
D. Ampicillin 150 mg/kg/dose BID, gentamicin 4mg/kg/dose OD
E. Ampicillin 50 mg/kg/dose BID, gentamicin 10mg/kg/dose OD

101. You are reviewing the clinical data for a G2 P1 mother who has elected for induction of labor tomorrow. Past obstetrical history reveals GBS infection of her previous baby during early neonatal period.
Of the following, the **MOST** appropriate plan is to
A. order for GBS screening vaginal culture
B. give GBS vaccine to the mother
C. start combination of broad spectrum antibiotics to the mother
D. give intrapartum penicillin chemoprophylaxis

E. collect blood culture for the neonate after birth and treat him accordingly

102. A 36-week-gestation, G1P0 mother presents with labor, she had poor antenatal care visits and her GBS status is unknown. She has neither fever nor rupture of membrane.
Of the following, the **MOST** appropriate action is to

A. order rapid intrapartum nucleic acid amplification test NAAT for GBS
B. start combination of broad spectrum antibiotics to the mother
C. give intrapartum penicillin chemoprophylaxis
D. collect blood culture for the neonate after delivery and treat him accordingly
E. start empiric antibiotic therapy to the neonate

103. Which of the following congenital viral infections causes persistent postnatal infection?

A. Mumps
B. Coxsackievirus B
C. Rubeola
D. Smallpox
E. Hepatitis B virus

104. Which of the following congenital viral infections can cause congenital glaucoma?

A. Cytomegalovirus
B. Rubella virus
C. Treponema pallidum
D. Toxoplasma gondii
E. Human parvovirus B19

105. What is the viral infection if acquired during pregnancy causes mild infection to mother but causes microcephaly, lissencephaly and cerebellar hypoplasia to the baby?

A. Rubella
B. VZV
C. Coxsackievirus B
D. HIV
E. Zika

106. What is the **MOST** common health care acquired infection (nosocomial) in the neonatal intensive care unit?

A. Central line–associated bloodstream infection
B. Ventilator-associated pneumonia
C. Surgical site infection
D. Catheter-associated urinary tract infection
E. Peritoneal dialysis catheter infection

107. What is the predominant microorganism causing the largest fraction of health care acquired infection in the NICU?

A. Coagulase-negative staphylococcus
B. Staphylococcus aureus
C. Enterococcus
D. Escherichia coli
E. Klebsiella pneumonia

108. What is the **MOST** common reported microorganism in ventilation-associated pneumonia?

A. Enterococcus
B. Gram-negative rod
C. Coagulase-negative staphylococcus
D. Staphylococcus aureus
E. Escherichia coli

109. What is the **MOST** effective measure in reducing fungal colonization and invasive infection in NICU for babies < 1000 gm birth weight?

A. Limit the use of broad-spectrum antimicrobials
B. Limitation of postnatal corticosteroid use in VLBW
C. Early enteral feeding with human milk feeding
D. Prophylactic administration of fluconazole
E. Early extubation

Chapter 11

The Fetus and the Neonatal Infant Answers

AHMED TAWFIQ

1.(E) Benign neonatal myoclonic seizure is a self-limiting disorder characterize by neonatal onset myoclonic jerks during non- rapid eye movement sleep and consistent cessation with arousal with absent concomitant EEG findings. Such history in a well-baby requires no investigations.

2.(A) While prematurity is the main cause in early neonatal period (first 7 days of life); the sepsis takes the lead in late neonatal life.

3.(C)

4.(E) Other distracters cause white reflex which is always pathological.

5.(A) Thin ribs and clubfoot are also recognized features.

6.(B) Full-term, vigorous infants may initially be placed on the mother's abdomen after delivery, during which time delayed clamping of the umbilical cord (30-60 sec) is recommended to improve transitional circulation and increase neonatal red blood cell (RBC) volume.

7.(C) Clearing the mouth of secretions with gentle suction with a bulb syringe or soft catheter is indicated if there is an excessive (copious) amount of fluid in the mouth or nares. In resource-poor countries, gentle wiping of the face, nose, and mouth with a soft cloth may be equally effective as a bulb syringe. Spontaneously breathing neonates without distress require no assisted method to clear their airway.

8.(A) Famous drugs that is contraindicated in a nursing mothers are amphetamines, antineoplastic agents, bromocriptine, and chloramphenicol while other distracters can be used with caution.

9.(C) For TB, treatment for 14 days is usually suffice; for all types of hepatitis breast feeding is not contraindicated, while in varicella covering of the lesion is enough.

10.(E)

11.(C) The ultrasound criteria for these diagnoses are based on either the amniotic fluid index (AFI) or a deepest vertical pocket (DVP). The AFI is determined by measuring the vertical dimension of amniotic fluid pockets in 4 quadrants and reporting the sum of these values. An index >24 cm suggests

polyhyramnios, where as an index <5cm suggests oligohydramnios. The DVP method reports the deepest pocket of fluid identified with a value of 2-8cm is considered normal.

12.(B)

13.(D) Fetal bradycardia may occur with fetal hypoxia, placental transfer of local anesthetic agents and β-adrenergic blocking agents, and occasionally, heart block with or without congenital heart disease.

14.(A) Cytomegalovirus (CMV) is the most common congenital infection, affecting 0.2–2.2% of all neonates. Perinatal transmission can occur at any time during the pregnancy; however, the most devastating sequelae occur with first-trimester infection. After a primary infection, 12–18% of neonates will have signs and symptoms at birth, and as many as 25% can develop long-term complications. The most common complication is congenital hearing loss. Severely affected infants have an associated 30% mortality, and 65–80% of survivors develop severe neurologic morbidity**.**

15.(B) Before implantation (0-2 wk postconception), radiation doses of 5-10 rad may result in miscarriage. At 2-8 wk gestation, doses in excess of 20 rad have been associated with congenital anomalies and fetal growth restriction. Severe intellectual disabilities can occur with exposures of ≥25 rad before 25 wk gestation.

16.(D) Start with D then E.

17.(B)

18.(B)

19.(A) MRI is the most sensitive imaging modality for detecting hypoxic brain injury in neonates.

20.(D)

21.(B)

22.(D)

23.(A) High dose erythropoietin.

24.(B) For treatment of refractory seizures in hypoxic-ischemic encephalopathy, levetiracetam is preferable now days as first or second line, third-line agents commonly used include; midazolam, topiramate, and lidocaine. Pyridoxine should also be attempted, particularly in ongoing refractory seizures with highly abnormal EEG background.

25.(C)

26.(E)

27.(A)

28.(C) All are important but in order of sequence of C, B, A, D, E.

29.(B)

30.(A) In the past the recommended inspired gas for neonatal resuscitation had been 100% oxygen. However, resuscitation with room air in term infants is equally effective and may reduce the risk of hyperoxia, which is associated with decreased cerebral blood flow and generation of oxygen free radicals. Room air is the preferred initial gas for neonatal resuscitation in term infants. O2 concentration administered should then be titrated as needed to obtain expected O2 saturations in a term infant after birth, as defined by normal reference range by minute of life.

31.(E) Methylxanthines are good choice for apnea of prematurity for otherwise well preterm baby with good recovery and had no complications, keeping in mind that apnea of prematurity can happen late even after weeks in those who developed RDS while it appears early in the first 7 days in those RDS free disease.

32.(E) Gastroesophageal reflux (GER) is common in neonates, but despite being associated with apnea anecdotally, data do not support a causal relationship between GER and apneic events. In preterm infants, medications that inhibit gastric acid production have potentially harmful side effects (increased incidence of sepsis, necrotizing enterocolitis, death) and may actually increase the incidence of apnea and bradycardia. Therefore the routine use of medications that inhibit gastric acid synthesis or promote gastrointestinal motility to reduce the frequency of apnea in preterm infants should be discouraged.

33.(D) This baby had TAPVR (mostly obstructed type) who is diagnosed by echocardiography. Although infants of diabetic mother can present in term by RDS they can also presented with congenital heart disease. The key here was quite tachypnea without respiratory distress, hypotension, minimal response to surfactant and hepatomegaly. The clinical and radiological picture of TAPVR and RDS are much similar. Congenital pneumonia is a possibility and collecting septic screen and starting empirical antibiotics should have been done initially.

34.(B) Antenatal corticosteroids do not increase the risk of maternal death, chorioamnionitis, or puerperal sepsis. Betamethasone and dexamethasone have both been used antenatally. Betamethasone may reduce neonatal death to a greater extent than dexamethasone.

35.(D) The description is for cephalohematoma which is benign lesion that disappears usually after 2weeks-3months, but a sensation of central depression suggesting but not indicative of an underlying fracture or bony defect. An

underlying skull fracture, usually linear and not depressed. The depressed fracture may be associated in 10–25% of cases.

36.(C) The above scenario is indicative of severe subgaleal (subapponeurotic) hemorrhage, baby should be admitted to NICU, monitored for hypotension, anemia, and hyperbilirubinemia, and consumptive coagulopathy from massive blood loss; secondary causes in such severe bleeding as hereditary coagulopathy (hemophilia) should rolled-out. Less severe cases should be admitted also for close observation of progression of the lesion.

37.(E) Most asymptomatic subdural hemorrhages following labor should resolve by 4 wk of age.

38.(E) Its recommended that premature infants <32 wk of gestation be evaluated with routine real-time cranial ultrasonography (US) through the anterior fontanel to screen for IVH. Infants <1,000 g are at highest risk and should undergo cranial US within the 1st 3-7 days of age, when approximately 75% of lesions will be detectable. All at-risk infants should undergo follow-up US at 36-40 wk postmenstrual age to evaluate adequately for PVL, as cystic changes related to perinatal injury may not be visible for up to 1 mo.

39.(D) Classic presentation of ICH in a preterm baby.

40.(B) Applying nCPAP at 5-10 cm H2O with at FIO2 of ≥40–70% is indicated if there is significant respiratory distress (severe retractions and expiratory grunting) or if SaO2 cannot be kept >90% and usually produces a rapid improvement in oxygenation. Warm, humidified oxygen should be provided at a concentration sufficient to keep PaO2 between 50 and 70 mm Hg (91–95% SpO_2) to maintain normal tissue oxygenation while minimizing the risk of O_2 toxicity. Second dose of surfactant usually considered after 6-12 hours after the first dose.

41.(A) By altered imprinting assisted reproductive technology can lead also to Silver-Russel, Angelman syndromes.

42.(D) During mechanical ventilation, oxygenation is improved by increasing either FiO_2 or the mean airway pressure. The mean airway pressure can be increased by raising the peak inspiratory pressure (PIP), inspiratory time, ventilator rate, or positive end-expiratory pressure (PEEP). Adjustment in pressure is usually most effective (but we should not forget its case dependent). However, excessive PEEP may impede venous return, thereby reducing cardiac output and O2 delivery.

43.(C) Hyperoxia contributes to lung injury in preterm infants. However, a lower target range of oxygenation (85–89%) compared with higher range (91–95%) increases mortality and does not alter rates of BPD, BPD/death, blindness,

or neurodevelopmental impairment. Therefore the currently recommended range of oxygen saturation targets is 91–95%.

44.(A)

45.(E) Avoidance of mechanical ventilation with the early use of nCPAP and early, selective surfactant replacement therapy with rapid extubation decrease the incidence of BPD. Modestly Caffeine therapy for apnea of prematurity has also been associated with a decreased risk of BPD.

46.(C) In the past, intratracheal surfactant replacement for symptomatic premature infants immediately after birth (prophylactic) or during the 1st few hr of life (early rescue) showed reduced air leak and mortality from RDS. However, substantial evidence supports the feasibility and efficacy of prophylactic nCPAP as the primary means of respiratory support for preterm infants with RDS. CPAP started at birth is as effective as prophylactic or early surfactant and is associated with a reduction in BPD. Prophylactic nCPAP is therefore the approach of choice for the delivery room management of a preterm neonate at risk for RDS.

47.(C) All the above mentioned actions are true, but evidently the baby has developed pulmonary edema and deterioration of renal function from undiagnosed PDA and consequent cardiac volume overload. The first action is to administer diuretics and order fluid restriction, meanwhile waiting for an echocardiography and renal function results. Starting cyclooxygenase (COX) inhibitors (indomethacin or ibuprofen) is a later on decision depending on progress and case response to supportive treatment.

48.(C) By the time of discharge in the majority of extremely preterm infants (>90%), the PDA will close spontaneously. Spontaneous ductal closure may be facilitated by general supportive measures, including early (<7 days of age) avoidance of excessive fluid administration and judicious use of diuretics to manage pulmonary edema. Pharmacologic and surgical ductal closure may be indicated in the premature infant with a moderate to large, hemodynamically significant PDA when there is a delay in clinical improvement or deterioration.

49.(A) Transient tachypnea of the newborn is a clinical syndrome of self-limited tachypnea associated with delayed clearance of fetal lung fluid. Although the actual incidence is likely underreported, it is estimated at 3-6 per 1,000 term infant births, making TTN the most common etiology of tachypnea in the newborn.

50.(D) The risk of meconium aspiration may be decreased by rapid identification of fetal distress and initiation of prompt delivery in the presence of late fetal heart rate deceleration or poor beat-to-beat FHR variability. Despite initial

enthusiasm for amnioinfusion, it does not reduce the risk of MAS, cesarean delivery, or other major indicators of maternal or neonatal morbidity. Intrapartum nasopharyngeal suctioning in infants with meconium-stained amniotic fluid does not reduce the risk for MAS. Routine intubation and aspiration of depressed infants (those with hypotonia, bradycardia, or decreased respiratory effort) born through meconium-stained fluid is not effective in reducing the MAS or other major adverse outcomes and is not recommended for neonatal resuscitation.

51.(C) Babies with severe MAS developed severe PHT which causes functional pulmonary and tricuspid regurgitation mimicking congenital heart disease.

52.(D) Persistent pulmonary hypertension of the newborn (PPHN) or persistent fetal circulation occurs in term and postterm infants most often. Predisposing factors include birth asphyxia, MAS, early-onset sepsis, RDS, hypoglycemia, polycythemia, maternal use of nonsteroidal antiinflammatory drugs within utero constriction of the ductus arteriosus, maternal late trimester use of selective serotonin reuptake inhibitors, and pulmonary hypoplasia caused by diaphragmatic hernia, amniotic fluid leak, oligohydramnios, or pleural effusions. PPHN is often idiopathic. Hypoxemia is labile (changing) and out of proportion to the findings on chest radiographs.

53.(B) Inotropic therapy is frequently needed to support blood pressure and perfusion. Whereas dopamine is frequently used as a first-line agent, other agents, such as dobutamine, epinephrine, and milrinone, may be helpful when myocardial contractility is poor. Some of the sickest newborns with PPHN demonstrate hypotension refractory to vasopressor administration. This results from desensitization of the cardiovascular system to catecholamines by overwhelming illness and relative adrenal insufficiency. Hydrocortisone rapidly upregulates cardiovascular adrenergic receptor expression and serves as a hormone substitute in cases of adrenal insufficiency.

54.(E) Infant with diaphragmatic hernia CDH at birth who are stable at birth should take only routine care and should be admitted to neonatal care unit for further workup. Delivery should be at a tertiary center with good experience in the management of CDH. In the delivery room, those with respiratory distress should be rapidly stabilized with endotracheal intubation. Prolonged mask ventilation in the delivery room, which enlarges the stomach and small bowel and thus makes oxygenation more difficult, must be avoided and a naso- or orogastric tube placed immediately for decompression. Gentle ventilation with permissive hypercapnia reduces lung injury, need for ECMO, and mortality.

Factors that contribute to pulmonary hypertension (hypoxia, acidosis, hypothermia) should be avoided.

55.(C) The ideal time to repair the diaphragmatic defect is under debate. Most experts wait at least 48hr after stabilization and resolution of the pulmonary hypertension. Good relative indicators of stability are the requirement for conventional ventilation only, a low peak inspiratory pressure, and FIO2 <50. An echocardiography is indicated to rule out associate congenital heart disease and to assess the pulmonary and systemic pressure.

56.(B) In this scenario the well grown child with normal development and the recurrent infection in the same lobe should rise the possibility of associated lobe sequestration as most of the eventrations is asymptomatic and need no treatment.

57.(A) Meconium plugs are associated with small left colon syndrome in infants of diabetic mothers, CF (40%), Hirschsprung disease (40%), maternal opiate use, magnesium sulfate therapy for preeclampsia, and tocolysis. Meconium plugs syndrome refers to intestinal obstruction, usually in the distal colon, rectum, and anal canal, caused by meconium plugs. Initial treatment may include administration of a glycerin suppository or rectal irrigation with isotonic saline.

58.(C) The characteristic X-ray for meconium ileus shows dilated small bowl due to inspissation of meconium in terminal ileum which eventually will cause microcolon. A in intussusception, B non-specific and occurs in distal bowl obstruction, D in midgut volvulus, and E in duodenal atresia.

59.(D) The onset of NEC is usually in the 2nd or 3rd week of life but can be as late as 3 mo in VLBW infants.

60.(A) The most effective preventive strategy for NEC is the use of human milk . It is well documented that newborns exclusively breastfed have a reduced risk of NEC. While extensive data and meta-analyses would support the use of probiotics to prevent NEC, there is no clear consensus on the safest, most effective formulation, timing of administration, or length of therapy. Other preventive strategies using prebiotics and synbiotics have also been studied, with variable outcomes. Inhibitors of gastric acid secretion (H2 –receptor blockers, proton pump inhibitors) or prolonged empirical antibiotics in the early neonatal period have been associated with increased risk of NEC and should be avoided.

61.(B) MCV <100 fL at birth should prompt consideration of underlying α-thalassemia trait or maternal iron deficiency. 2nd possibility is unlikely if mother had good antenatal care. If the baby is anemic more at birth other severe forms of α-thalassemia syndrome should be considered in the differential diagnosis.

62.(E) Decreased or absent fetal movement is the most common antenatal presentation of large or massive fetomaternal hemorrhage (FMH). It should be associated with a high degree of clinical suspicion. After delivery, the infant usually has pallor, hypotension, and poor perfusion. To diagnose FMH, the classic Kleihauer-Betke test, which identifies fetal erythrocytes containing HbF resistant to acid elution, is technically the gold standard but is labor intensive, highly dependent on the skills of the technician, and often not available as a rapid or point-of-care test. Some advanced laboratories offer a more precise test using flow cytometry to quantify fetal cells in the maternal circulation.

63.(B) This is a typical picture of hemolytic disease of the fetus and newborn (HDFN), and the laboratory results suggesting enzymopathy which is either G6PD deficiency or pyruvate kinase. In G6PD, severe anemia with reticulocytosis is not common, but hyperbilirubinemia can be severe and prolonged. Clinical testing measuring G6PD activity can be performed (<1–2% suggests G6PD deficiency). Pyruvate kinase (PK) deficiency is the 2nd most common RBC enzymopathy and may also be associated with neonatal jaundice and bizarre morphology featuring acanthocytes.

64.(C) Suggested transfusion thresholds

Postnatal age	**Presence of RS**	**Absence of RS**
Week 1	11.5 (35%)	10.0 (30%)
Week 2	10.0 (30%)	8.5 (25%)
Week 3	8.5 (25%)	7.5 (23%)

In addition to these factors, transfusion should be considered for infants with acute blood loss (>20%) or significant hemolysis, as well as before surgery. With no similar evidence-based guidelines for term infants, transfusion should be based on hemodynamic stability, respiratory status, overall clinical condition, and laboratory values.

65.(A) Factors that affect the outcome of antigen-positive fetuses include differential immunogenicity of blood group antigens (RhD antigen being the most immunogenic), a threshold effect of fetomaternal transfusions (a certain amount of the immunizing blood cell antigen is required to induce the maternal immune response), the type of antibody response (IgG antibodies are more efficiently transferred across the placenta to the fetus). Notably, when the mother and fetus are also ABO incompatible, the Rh negative mother is partially protected against sensitization due to rapid removal of the fetal Rh-positive cells by maternal isohemagglutinins (preexisting IgM anti-A or anti-B antibodies that do not cross the placenta). The severity of Rh illness typically worsens with successive pregnancies because of repeated immune stimulation.

66.(B) Hypoglycemia occurs in infants with severe HDFN and may be related to hyperinsulinism and hypertrophy of the pancreatic islet cells in these infants. Other distracters are more related to the complication of treatment of the condition (exchange transfusion).

67.(C) Fetal RBC genotyping provides an accurate prediction for the development of HDFN in sensitized mothers. Fetal Rh status is available by isolating fetal cells or fetal DNA (plasma) from the maternal circulation, which is replacing the more invasive and risky fetal amniocyte testing by amniocentesis and chorionic villus sampling methods. The presence of elevated antibody titers or rising titers indicats an established hemolytic process rather than prediction.

68.(E) If an Rh-negative mother is found to have RhD antibody titers of ≥1 : 16 (15 IU/mL in Europe) at any time during a subsequent pregnancy, the severity of fetal anemia should be monitored by Doppler ultrasonography (US) of the middle cerebral artery (MCA) and then percutaneous umbilical blood sampling (PUBS) if indicated (if signs of hydrops developed). Intravascular (umbilical vein) transfusion of packed erythrocytes (PRBCs) is the preferred treatment of choice for fetal anemia, although intrauterine transfusion into the fetal peritoneal cavity is also effective. Hydrops or fetal anemia (hematocrit <30%) is an indication for umbilical vein transfusion in infants with pulmonary immaturity.

69.(B) Extramedullary hematopoiesis and hepatic congestion compress the intrahepatic vessels and produce venous stasis with portal hypertension, hepatocellular dysfunction, and decreased albumin synthesis. Hydrops is typically present when fetal hemoglobin level is <5 g/dL. Hydrops is also frequently seen with a fetal hemoglobin level <7 g/dL, and in some cases between 7 and 9 g/dL.

70.(B) If clinical signs of severe hemolytic anemia (pallor, hepatosplenomegaly, edema, petechiae, ascites) are evident at birth, immediate resuscitation and supportive therapy, temperature stabilization, and monitoring before proceeding with exchange transfusion may save severely affected infants. Such therapy should include a small transfusion of compatible PRBCs to correct anemia; volume expansion for hypotension, especially in those with hydrops; correction of acidosis with 1-2 mEq/kg of sodium bicarbonate; and assisted ventilation for respiratory failure. Infants with HDFN should be closely monitored with frequent hemoglobin and bilirubin testing to determine their need for phototherapy, simple transfusion, or exchange transfusion.

71.(D) Because of its ability to interfere with immune-mediated clearance of antibody sensitized RBCs, early administration of IVIG may be an effective therapeutic intervention for HDFN. IVIG can prevent immune hemolysis, lower

peak serum bilirubin levels, shorten the duration of phototherapy, and reduce both length of hospitalization and need for exchange transfusion. However, IVIG does not effectively prevent anemia, which results from both immune-mediated RBC destruction and inadequate erythropoiesis. Consequently, simple transfusions are usually needed as an adjunct to IVIG therapy. An IVIG dose of 0.5-1 g/kg is typically used, but optimal dosing has not been established. Treated infants with blood groups A or B should be monitored for worsening hemolysis caused by anti-A or anti-B antibodies present in IVIG.

72.(A) The only 2 indication for immediate exchange transfusion is presence of signs of acute bilirubin encephalopathy (jaundice, fever, lethargy, hypertonia, arching, retrocollis, high pitch cry) and a total bilirubin is more than 5mg/dl of the accepted level for age in hours (the standard graph of hyperbilirubinemia).

73.(E)

74.(A) A capillary Hct >65% should always be confirmed with a venous sample and dehydration should be treated. All polycythemic infants should be closely monitored for intake and output, and blood glucose and bilirubin levels should be closely followed. Asymptomatic infants whose central Hct is 60–70% can be monitored closely and hydrated with adequate enteral intake or administration of intravenous (IV) fluids. Treatment of symptomatic polycythemic newborns is not well defined. A partial exchange transfusion (with normal saline) can be used in infants with severe polycythemia and symptoms of hyperviscosity and should be considered if the Hct is ≥70–75% and symptoms worsen despite aggressive IV hydration.

75.(D) Plasma levels of the vitamin K–dependent coagulation factors (II, VII, IX, X, protein C, protein S) and antithrombin are low at birth and do not reach adult ranges until approximately 6 mo of age.

76.(B) This classic form of hemorrhagic disease of the newborn, which is responsive to (and entirely prevented by) exogenous vitamin K therapy, is characterized by hemorrhage that is most frequently gastrointestinal, nasal, subgaleal, intracranial, or post-circumcision. Prodromal or warning signs (mild bleeding) may occur before serious intracranial hemorrhage. Classically, vitamin K deficiency bleeding occurs early in the newborn period, typically between day 2 and 7 of life, and most often in exclusively breastfeeding infants who did not receive vitamin K prophylaxis at birth. Severe vitamin K deficiency is also more common in premature infants. This baby had home delivery and he didn't received vitamin K prophylaxis.

77.(D) Early-onset vitamin K deficiency bleeding (after birth but in 1st 24 hr) occurs if the mother has been treated chronically with certain drugs (e.g.,

anticoagulant warfarin, anticonvulsant phenytoin or phenobarbital, and cholesterol-lowering medication) that interfere with vitamin K absorption or function. Avoidance of high-risk medications (if possible), antenatal vitamin K to treatment of mother (20 mg) before birth and postnatal administration to infant soon after birth. These infants can have severe bleeding, which is usually corrected promptly by vitamin K administration, although some have a poor or delayed response. If a mother is known to be receiving such drugs late in gestation, an infant PT should be measured using cord blood, and the infant immediately given 1-2 mg of vitamin K IV. If PT is greatly prolonged and fails to improve, or in the presence of significant hemorrhage, 10-15 mL/kg of fresh-frozen plasma should be administered.

78.(E) As early onset type; the late onset usually present with serious intracranial bleeding.

79.(A) In utero treatment has been successful for fetal supraventricular tachycardia(SVT), twin-twin transfusion syndrome, nonimmune fetal anemias, and some surgically treatable fetal conditions.

80.(E) The most common remnant of the OMD is a Meckel diverticulum, where as abnormalities that would become symptomatic in the neonatal period include a sinus or fistula that would drain mucus or intestinal contents through the umbilicus. An umbilical polyp is one of the least common OMD remnants and represents exposed GI mucosa at the umbilical stump. The tissue of the polyp is bright red, firm, and has a mucoid secretion. Therapy for all OMD remnants is surgical excision of the anomaly.

81.(A) Persistence of granulation tissue at the base of the umbilicus is common. The tissue is soft, 3-10 mm in size, vascular and granular, colored dull red or pink, and may have a seropurulent discharge. Granulation tissue is treated by cauterization with silver nitrate, repeated at intervals of several days until the base is dry.

82.(B) The term omphalitis refers to infection of the umbilical cord stump, navel, or the surrounding abdominal wall. The presence of cellulitis is associated with a high incidence of bacteremia, and complicated omphalitis may spread to the peritoneum, the umbilical or portal vessels, or the liver. Treatment of omphalitis includes prompt antibiotic therapy with agents effective against Staphylococcus aureus and Escherichia coli, such as an antistaphylococcal penicillin or vancomycin in combination with an aminoglycoside. If abscess formation has occurred, surgical incision and drainage may be required.

83.(C) All psychotropic agents cross the placenta. Exposure to these medications in utero may lead to a higher risk of congenital malformations, poor neonatal

adaptation syndrome (PNAS), and persistent pulmonary hypertension (PPHN). Some reported defects include anencephaly, atrial septal defect, right ventricular outflow tract obstruction, omphalocele, and gastroschisis. The classification of safety of any medication during pregnancy is graded from A to X. As given the questions A is safe , B when animal studies shows no risk but no available human studies, C as in our scenario the clinical findings of mother should be discussed with psychiatrist and risk VS benefits should be taken in consideration as there is evidence of harm, D for emergencies only and X is prohibited to be use.

84.(C) Poor neonatal adaptation syndrome PNAS related to maternal selective serotonin reuptake inhibitors. Symptoms usually appear within the 1st 8 hr after birth and often persist for the 1st 2-6 days of life. PNAS affects the neurologic, autonomic, respiratory, and gastrointestinal systems. Symptoms include a weak suck reflex, irritability, tremors, hypertonia and hypotonia, hyperthermia, weak or absent cry, sleep disturbances, hypoglycemia, respiratory problems, vomiting and diarrhea, and seizures. Most symptoms are mild, and severe symptoms are rare. No deaths have been reported. Treatment consists of supportive measures, and most cases are mild, of short duration, and self-limiting. Small, frequent, on-demand feedings; swaddling; and skin-to-skin contact are beneficial to support infants through this process. Breastfeeding is protective against developing PNAS and should be encouraged because many antidepressant medications are safe with breastfeeding.

85.(A) Pituitary dwarfism (growth hormone deficiency) is not usually apparent at birth, although male infants with panhypopituitarism may have neonatal hypoglycemia, hyperbilirubinemia, and micropenis. Conversely, primordial dwarfism manifests as in utero growth failure that continues postnatally, with length and weight suggestive of prematurity when born after a normal gestational period; otherwise, physical appearance is normal.

86.(D) Congenital hypothyroidism is one of the most common preventable causes of developmental disability. Congenital screening followed by thyroid hormone replacement treatment started within 30 days after birth can normalize cognitive development in children with congenital hypothyroidism.

87.(B) Subcutaneous fat necrosis can cause hypercalcemia and can occur after a traumatic birth. On examination, firm purple nodules can be appreciated on the trunk or extremities. An infant with hypercalcemia presents with irritability, vomiting, increased tone, poor weight gain, and constipation. Other causes of hypercalcemia in the newborn period are iatrogenic (excess calcium or vitamin

D), maternal hypoparathyroidism, Williams syndrome, parathyroid hyperplasia, and idiopathic.

88.(D) All are true but D is confirmative.

89.(B)

90.(D) The risk of diabetic embryopathy (neural tube defects, cardiac defects, caudal regression syndrome) and spontaneous abortions is highest in those with pregestational diabetes who have poor control (HbA1c >7%) in the first trimester. There is a 4-fold increase in congenital anomalies in infants of mothers with pregestational diabetes**.**

91.(C) A and B are contraindicated as it increases the subaortic obstruction, β-Adrenergic blockers have been shown to relieve the obstruction, but ultimately the condition resolves spontaneously over time.

92.(E) Renal vein thrombosis should be suspected in the infant with a flank mass, hematuria, and thrombocytopenia. IDMs have an increased incidence of hyperbilirubinemia, polycythemia, iron deficiency, and renal vein thrombosis. Small left colon syndrome is a rare anomaly that develops in the second and third trimester because of rapid fluctuations in maternal and therefore fetal glucose, leading to impaired intestinal motility and subsequent intestinal growth.

93.(A) Women can begin to express breast milk before the birth of the baby (≥36 wk gestational age); this will provide an immediate supply of milk to prevent hypoglycemia. Others are still true alternative options.

94.(C) Infants should initiate feedings within 1 hr after birth. A screening glucose test should be performed within 30 min of the first feed. Transient hypoglycemia is common during the 1st 1-3 hr after birth and may be part of normal adaptation to extrauterine life. The target plasma glucose concentration is ≥40 mg/dL before feeds in the 1st 48 hr of life. Clinicians need to assess the overall metabolic and physiologic status, considering these in the management of hypoglycemia. Treatment is indicated if the plasma glucose is <47 mg/dL. Feeding is the initial treatment for asymptomatic hypoglycemia. Oral or gavage feeding with breast milk or formula can be given. An alternative is prophylactic use of dextrose gel, although early feedings may be equally effective.

95.(B) Recurrent hypoglycemia can be treated with repeat feedings or IV glucose as needed. Infants with persistent (and unresponsive to oral therapy) glucose levels <25 mg/dL during the 1st 4 hr after birth and <35 mg/dL at 4-24 hr after birth should be treated with IV glucose, especially if symptomatic. A small bolus of 200 mg/kg of dextrose (2 mL/kg of 10% dextrose) should be administered to infants with plasma glucose below these limits. The small bolus

should be followed by a continuous IV glucose infusion to avoid hypoglycemia. If question arises about an infant's ability to tolerate oral feeding, a continuous peripheral IV infusion at a rate of 4-8 mg/kg/min should be given. Neurologic symptoms of hypoglycemia must be treated with IV glucose. Bolus injections of hypertonic (25%) glucose should be avoided because they may cause further hyperinsulinemia and potentially produce rebound hypoglycemia.

96.(C) Screening asymptomatic hypoglycemia during first day of life among at-risk infants is indicated for late preterm infants, those who are small for gestational age/intrauterine growth restriction, and infants of obese or diabetic mothers. Continue monitoring blood glucose concentrations until 3 consecutive blood glucose concentrations have been ≥45-50 mg/dL. Regarding other distracters, it may be true to do measuring of blood glucose accordingly.

97.(B) Various serum biomarkers have been investigated for their ability to identify infants with serious bacterial infection (SBI). An immature-to-total phagocyte count (I/T ratio) (≥0.2) has the best sensitivity of the neutrophil indices for predicting neonatal sepsis. After the newborn period, serum C-reactive protein (CRP) and procalcitonin have demonstrated reasonable sensitivity and specificity for SBI.

98.(B) Option C for late onset sepsis, option E should be added in case of meningitis in early onset sepsis.

99.(D)

100.(D) This has long been a standard regimen for early-onset sepsis and provides coverage for the most prevalent organisms, predominantly GBS and gram-negative ones. Ampicillin plus cefotaxime (if available) or cefepime may be substituted if the patient presents with infection after discharge from the nursery, or when infection with ampicillin-resistant E. coli is suspected. Ceftriaxone may be substituted if premature infants are ≥41 wk postconception age; it may be used in term infants if they are not receiving intravenous calcium or do not have hyperbilirubinemia.

101.(D) Empiric antibiotic therapy for intrapartum GBS prophylaxis indicated in; previous infant with invasive GBS disease; GBS bacteriuria during any trimester of the current pregnancy; and positive GBS screening culture during current pregnancy. All pregnant women should be screened for GBS at 35-37 wks of gestation.

102.(C) Unknown GBS status at the onset of labor (culture not done, incomplete, or results unknown) and any of the following: delivery at <37 weeks' gestation, amniotic membrane rupture ≥18 hr, intrapartum temperature ≥38.0°c (100.4°f), intrapartum NAAT positive for GBS.

103.(E) CMV, Rubella, VZV, HSV, HBV, HIV and Zika causes persistent postnatal infection; Coxsackie virus causes developmental anomalies; Rubeola and Smallpox causes premaurity.

104.(B)

105.(E) Usually acquired by mosquito bite.

106.(A) Ventilator-associated pneumonia (VAP) is the next most common, followed by surgical site infection and catheter-associated urinary tract infection.

107.(A) It account for about 50% of bloodstream infections in the NICU, Staphylococcus aureus about 10% and others < 5% each.

108.(B) The most commonly reported organisms associated with VAP are gram-negative rods (including *Pseudomonas*), *S. aureus* , and *Enterococcus* . The source of infecting organisms is generally thought to be the infant's oropharynx, although contaminated respiratory equipment and tracheal suction catheters are occasionally implicated.

109.(D) Prophylactic administration of fluconazole during the 1st 6 wk of life reduces fungal colonization and invasive fungal infection in ELBW infants (<1000 g). Neonatal practices that may reduce the risks of invasive candidiasis include limited use of broad-spectrum antimicrobials, use of an aminoglycoside instead of a cephalosporin for empirical therapy when meningitis or antimicrobial resistance is not suspected, and limitation of postnatal corticosteroid use in VLBW infants, early enteral feeding, and establishment of the neonatal gut microbiome with human milk feeding.

Chapter 12

Adolescent Medicine Questions

AHMED TAWFIQ

1. What is the hallmark of puberty in an adolescent girl?
   A. Axillary hair
   B. Breast enlargement
   C. Menarche
   D. Pubic hair
   E. Growth spurt

2. What is the hallmark of puberty in an adolescent boy?
   A. Voice change
   B. Axillary hair
   C. Pubic hair
   D. Testicular enlargement
   E. Growth spurt

3. In a girl who lacks any pubertal signs; what is the age at which prompt evaluation for pubertal delay should be started?
   A. 10 years
   B. 11 years
   C. 12 years
   D. 13 years
   E. 14 years

4. In primary amenorrhea, the standard initial endocrine work up include testing for TSH, prolactin, HCG and
   A. LH
   B. FSH
   C. Thyroxin
   D. free and total testosterone
   E. 17-hydroxyprogesterone

5. A recognizable cause of high FSH in amenorrhea is

A. hypothalamic injury
B. pituitary injury
C. ovarian failure
D. female athlete
E. polycystic ovary

6. In the early postmenarcheal years, the **MOST** common cause of irregular and abnormal uterine bleeding in adolescents is

A. anovulatory cycle
B. eating disorder
C. polycystic ovary disease
D. pelvic inflammatory disease
E. hematologic causes

7. What is the **MOST** common cause of secondary dysmenorrhea in adolescents?

A. Müllerian anomalies
B. Endometriosis
C. Pelvic inflammatory disease
D. Uterine fibroid
E. Ovarian cyst

Chapter 12

Adolescent Medicine Answers

AHMED TAWFIQ

1.(B)

2.(D)

3.(D) Amenorrhea, the absence of menstruation, generally requires evaluation at age 15 yr, or if there has been no menstruation within 3 yr of the onset of puberty (primary amenorrhea), or if there has been no menstruation for the length of 3 previous cycles in a postmenarchal patient (secondary amenorrhea). However, the following caveats exist: lack of any pubertal signs by age 13 yr in a girl should prompt evaluation for pubertal delay.

4.(B) According to FSH level, 2 groups of amenorrhea will be classify, those with low and those with high FSH.

5.(C) Other distracters causing amenorrhea with low FSH.

6.(A) In the early postmenarcheal years, the most common cause of abnormal uterine bleeding in adolescents is anovulation caused by immaturity of the hypothalamic-pituitary ovarian axis. Other distracters are true but less common causes.

7.(B) Secondary dysmenorrhea results from underlying pathology, such as anatomic abnormality, or infection, such as pelvic inflammatory disease. However, the most common cause of secondary dysmenorrhea in adolescents is endometriosis, a condition in which implants of endometrial tissue are found outside the uterus, usually near the fallopian tubes and ovaries. Often, other family members have endometriosis. Although characteristically there is severe pain at menses, adolescents can present with noncyclic pain as well.

Chapter 13

Immunology Questions

USAMA A. AL JUMAILY

1. A 2-year-old child has humoral type immunodeficiency. Which of the following features is **MOST** likely manifested in this child?

F. Recurrent bacterial pneumonias
G. Chronic mucocutaneous candidiasis
H. Absence of lymph nodes and tonsils
I. Recurrent infections with opportunistic organisms
J. Disseminated atypical mycobacterial infection

2. Which of the following types of immunodeficiency is suggestive when a child develops paralysis after vaccination with live-attenuated poliovirus?

A. B-cell defect
B. T-cell defect
C. Complement deficiency
D. Macrophage dysfunction
E. Neutrophils dysfunction

3. A 1-year-old boy has T-cell defect immunodeficiency. His complete blood count **MOST** likely
shows

A. neutropenia
B. neutrophilia
C. lymphopenia
D. eosinophilia
E. monocytosis

4. Which of the following immunoglobulin isotypes has a **MAJOR** role in host defense against parasites?

A. IgG
B. IgM
C. IgA

D. IgE
E. IgD

5. A medical student asks you about postnatal immunoglobulin development. The statement that should be included in this discussion is

A. serum IgM rises sharply at about 6 month after birth
B. serum IgM reaches adult levels by approximately 6yr of age
C. serum IgA remains low throughout infancy
D. maternal serum IgG disappears after 1st yr of life
E. total IgG are reached and maintained by 1 yr of age

6. A 1-year-old boy develops recurrent pneumonias and otitis media. Examination reveals absence of tonsils, and no palpable lymph nodes. Lab tests reveal very low level (<2 standard deviations below the age-appropriate level) of IgG, IgM, IgA, and IgE. Peripheral blood flow cytometry demonstrates absence of circulating B cells.
Of the following, the **MOST** likely diagnosis is

A. common variable immunodeficiency (CVID)
B. transient hypogammaglobulinemia of infancy
C. hyper IgM syndrome
D. Bruton agammaglobulinemia
E. selective IgA deficiency

7. A 2-year-old boy develops pneumonia due to pseudomonas infection. He has been diagnosed with X-linked agammaglobulinemia (XLA) at the age of 1 year.
Of the following, the WBC parameter that is typically seen with such infection is

A. neutropenia
B. neutrophilia
C. lymphopenia
D. eosinopenia
E. eosinophilia

8. Common variable immunodeficiency (CVID) patients may appear similar clinically to those with X-linked agammaglobulinemia (XLA) in the types of infections experienced.
Which of the following infections, is **MORE** likely encountered in XLA than in CVID?

A. Hemophilus influenzae meningitis

B. Streptococcus pneumoniae sinusitis
C. Enterovirus meningoencephalitis
D. Mycoplasma pneumonia
E. Staphylococcus aureus septicemia

9. A 7-year-old girl has recurrent pneumonias and meningitis with encapsulated bacteria. She has normal sized tonsils, enlarged lymph nodes, and splenomegaly. Immunoglobulin level assay reveals very low serum IgG (<2 standard deviations below the age-adjusted norms), with low IgA and IgM levels.
This girl is particularly at increased risk for development of
A. leukemia
B. lymphoma
C. neuroblastoma
D. nephroblastoma
E. medulloblastoma

10. A 4-year-old boy has recurrent attacks of gastroenteritis caused by Giardia lamblia.
Which of the following diseases should be suspected in this boy?
A. common variable immunodeficiency (CVID)
B. hyper IgM syndrome
C. Bruton agammaglobulinemia
D. selective IgA deficiency
E. transient hypogammaglobulinemia of infancy

11. A 7-year-old child with selective IgA deficiency develops car accident requiring blood transfusion. The type of blood transfusion reaction that is **MOST** likely encountered in this child is
A. acute hemolytic transfusion reaction
B. bacterial contamination
C. acute circulatory overload
D. acute anaphylactic reaction
E. delayed hemolytic transfusion reaction

12. Children with X-Linked hyper-IgM syndrome are markedly susceptible to
A. Hemophilus influenzae
B. Streptococcus pneumoniae

C. Mycoplasma pneumoniae
D. Staphylococcus aureus
E. Pneumocystis jiroveci

13. A 5-year-old girl develops fever, leucopenia, and thrombocytopenia three months after allogenic bone marrow transplantation for recurrent ALL. PCR for CMV infection is positive.
Of the following, the 1st line of therapy for this girl is

A. acyclovir
B. ganciclovir
C. foscarnet
D. oseltamivir
E. peramivir

14. Which of the following is the treatment of choice in X-Linked hyper-IgM syndrome?

A. Monthly infusion of intravenous immunoglobulin IVIG
B. Granulocyte colony-stimulating factor
C. Trimethoprim prophylaxis
D. Acyclovir prophylaxis
E. HLA-identical hematopoietic stem cell transplant

15. Which of the following viruses, is often fatal if acquired in children with X-linked lymphoproliferative disorder?

A. Cytomegalovirus (CMV)
B. Epstein-Barr virus (EBV)
C. Coxsackie virus
D. Herpes simplex virus
E. Influenza virus

16. A 9-month-old infant develops disseminated tuberculosis after BCG vaccination. He has previous attacks of diarrhea, pneumonia, and sepsis. Complete blood count reveals low lymphocytes count according to his age. Radiological studies show very small sized thymus and absence of lymph nodes.
Of the following, the **MOST** likely diagnosis is

A. severe combined immunodeficiency (SCID)
B. Bruton agammaglobulinemia
C. common variable immunodeficiency (CVID)

D. autoimmune polyendocrinopathy-candidiasis–ectodermal dysplasia
E. transient hypogammaglobulinemia of infancy

17. An 11-month-old boy develops wide spread eczematous rash. He has a history of 2 episodes of lobar pneumonia caused by streptococcus pneumoniae, and 1 episode of sepsis due to Hemophilus influenzae. There is a history of melena during the neonatal period. Examination shows eczematous rash as well as scattered purpuric rash. Lab tests reveal platelets count of 20000/mm^3, small sized platelets, elevated IgM, low IgA and IgE, and normal IgG level.
Of the following, the treatment of choice for this boy is
A. platelets transfusion
B. appropriate prophylactic antibiotics
C. immunoglobulin replacement
D. aggressive management of eczema
E. hematopoietic stem cell transplantation (HSCT)

18. A 5-year-old boy develops dilated tiny blood vessels involving his eyes, lips, upper neck, and both legs. He has a history of progressive difficulty in walking and unsteady gait since the age of 2 year. He also has progressive delay developmental milestone. Parents are 2^{nd} degree relatives. Examination reveals spider-like blood vessels involving the sclera, mucous membrane of the mouth, and the skin.
Of the following, the **MOST** frequently encountered infection is
A. chronic mucocutaneous candidiasis
B. recurrent sinopulmonary disease
C. Recurrent skin abscesses
D. disseminated atypical mycobacterial infection
E. recurrent meningitis

19. A 7-year-old girl develops recurrent staphylococcal skin abscesses, atopic dermatitis, and pneumatoceles. He has a history of delay in shedding primary teeth and recurrent fractures. Examination shows facial asymmetry, prominent forehead, wide-spaced eyes, and wide fleshy nasal tip. Complete blood count reveals marked eosinophilia.
Of the following, the immunoglobulin class that is remarkably **HIGH** in this girl is
A. IgA
B. IgD
C. IgE

D. IgG
E. IgM

20. An 8-year-old boy develops extensive cutaneous molluscum contagiosum involving the whole trunk. He has a history of frequent viral infections including CMV and EBV. He has also a history of infection with cryptosporidium. Past medical history reveals severe eczema since infancy.
Of the following, the **MOST** likely diagnosis is

A. ataxia-telangiectasias
B. cartilage-Hair hypoplasia
C. Job syndrome
D. Wiskott-Aldrich syndrome
E. DOCK 8 deficiency

21. A 3-year-old girl develops extensive skin infection of the left leg. She has a history recurrent gingivitis and periodontitis with subsequent loss of some primary teeth. She has a history of delayed separation of umbilical cord and omphalitis during the neonatal period. Examination reveals ulcerative skin lesions with limited signs of inflammation and no pus formation.
Of the following, the **MOST** likely lab finding in this girl is

A. neutrophilia
B. neutropenia
C. monocytosis
D. eosinophilia
E. lymphopenia

22. Which of the following leukocyte adhesion deficiency (LAD) syndromes is associated with neutropenia rather than neutrophilia?

A. LAD-1
B. LAD-2
C. LAD-3
D. E-selectin deficiency
E. Rac2 deficiency

23. Which of the following leukocyte adhesion deficiency (LAD) syndromes is associated with bleeding tendency?

A. LAD-1
B. LAD-2

C. LAD-3
D. E-selectin deficiency
E. Rac2 deficiency

24. A 4-year-old boy has recurrent attacks of infections involving skin and respiratory tract. He has light skin and silvery hair. Lab findings reveal neutropenia, and numerous granular inclusions of all WBC.
Of the following, the complication that is **MOST** likely fatal in this disease is
A. overwhelming infections
B. bleeding
C. neuropathy
D. cardiomyopathy (CMP)
E. hemophagocytic lymphohistiocytosis (HLH)

25. The only curative therapy of Chédiak-Higashi syndrome is allogenic hematopoietic stem cell transplantation (HSCT). The complication that is inevitable even after HSCT is
A. recurrent infections
B. bleeding diathesis
C. neuropathy
D. hemophagocytic lymphohistiocytosis
E. growth retardation

26. A 7-year-old boy has persistent fever in spite of 5 days of administration of broad spectrum antibiotics. He has been underwent hematopoietic allogenic bone marrow transplantation for AML 15 days ago. Lab finding reveals neutropenia. CT scan of the chest shows multiple nodules surrounded by a halo of ground-glass attenuation.
Of the following, the **MOST** likely fungal infection is
A. Candida
B. Aspergillus
C. Mucor
D. Fusarium
E. Scedosporium

27. A 5-year-old boy develops fever, productive cough, and shortness of breath consistent with pneumonia. He has been diagnosed with chronic granulomatous disease (CGD) since infancy. You start empirical antibiotics.

Of the following, the **MOST** useful investigation to follow the response to treatment is

A. chest x-ray
B. CT scan of the chest
C. sputum culture
D. Erythrocyte sedimentation rate (ESR)
E. C-reactive protein (CRP)

28. A 5-year-old boy develops jaundice, hepatomegaly, right upper quadrant tenderness, progressive weight gain, and ascites. He has been underwent hematopoietic bone marrow transplantation for neuroblastoma 20 days ago. He has been treated initially for neuroblastoma by chemotherapy and abdominal irradiation.
Of the following, the **MOST** effective treatment for this condition is

A. ursodeoxycholic acid
B. heparin
C. phenobarbital
D. defibrotide
E. vitamin K analogue

29. A 4-year-old boy develops high grade fever, productive cough, and shortness of breath. He has been diagnosed with chronic granulomatous disease (CGD) since infancy. He has a history of recurrent pneumonias since infancy. The mother describes this illness as "unusually tough".
Of the following, the offending organism that should be considered and empirically covered is

A. staphylococcus aureus
B. serratia marcescens
C. borckolderia cepacia
D. candida albicans
E. aspergillus fumigatus

30. Transient neutropenia associated viral infections is frequently encountered in childhood.
Which of the following viruses is accompanied by chronic neutropenia rather than acute one?

A. Influenza A
B. Cytomegalovirus

C. Adenovirus
D. Human herpesvirus 6
E. Measles

31. Which of the following illnesses is associated with minimal risk of infections?
A. Cyclic neutropenia
B. Congenital neutropenia
C. Autoimmune neutropenia
D. Neutropenia following cytotoxic drugs
E. Neutropenia secondary to bone marrow replacement

32. Acute graft-versus-host disease (GVHD) usually develops 2-8 wk after transplantation, manifested as skin, GI, or hepatic features.
Which of the following is the characteristic cutaneous manifestation of acute GVHD?
A. Maculopapular rash
B. Lichen planus
C. Scleroderma
D. Hyperpigmentation
E. Hypopigmentation

33. The term "leukemoid reaction" is applied when WBC counts exceeding 50,000/µL with relatively small proportions of immature myeloid cells.
Which of the following cells constitutes the major component of these immature myeloid cells?
A. Bands
B. Metamyelocytes
C. Myelocytes
D. Promyelocytes
E. Blasts

34. Which of the following conditions of neutrophilia is associated with almost zero leukocyte alkaline phosphatase?
A. Chronic infections
B. Chronic blood loss
C. Vasculitis
D. Postsplenectomy states
E. Chronic myelogenous leukemia

35. A 7-year-old boy with malignancy has been received chemotherapy 2weeks ago. The lab test finding that is typically indicates patient recovering from myeloosuppressive therapy is

A. neutrophilia
B. lymphocytosis
C. monocytosis
D. eosinophilia
E. basophilia

36. What is the lab test finding that is **MOST** likely present in parasitic infections?

A. neutrophilia
B. lymphocytosis
C. monocytosis
D. eosinophilia
E. basophilia

37. Which of the following conditions is **MOST** likely associated with lymphocytosis?

A. Addison disease
B. Polyarteritis nodosa
C. Dermatitis
D. Systemic mastocytosis
E. Asthma

38. A 7-year-old girl develops swelling of the right hand after minor trauma, progressing to involve the forearm. There is no itching, redness, or urticaria. She has a history of previous attack of neck swelling after dental procedure that needed tracheostomy and respiratory support for few days. There is a positive paternal history of recurrent attacks of swelling after trauma or stress conditions.
Of the following, the **MOST** likely complement deficiency describing this condition is

A. C1 inhibitor
B. factor B
C. properdin
D. MASP-1
E. C8b

39. A 12-year-old adolescent female with hereditary angioedema is going to undergo cosmetic nasal surgery.
Which of the following is drug of choice preoperatively?

A. Antihistamines
B. Epinephrine
C. Corticosteroids
D. Ecallantide
E. Eculizumab

40. Allogeneic or autologous hematopoietic stem cells have been used to cure both malignant and nonmalignant disorders.
Which of the following diseases is treated by autologous, rather than allogeneic bone marrow transplantation?

A. Leukemia
B. Relapsed lymphoma
C. Myelodysplastic syndrome
D. B-thalassemia major
E. Severe combined immunodeficiency

Chapter 13

Immunology Answers

USAMA A. AL JUMAILY

1.(A) Disseminated atypical mycobacterial infection is suggestive of macrophage dysfunction. Other distracters are suggestive of T-cell defect.

2.(A)

3.(C) Lymphopenia is seen in the majority of T-cell defects.

4.(D) IgE is also the principal mediator of allergic reactions of the immediate type because of high affinity IgE receptors on basophils and mast cells.

5.(C) Neonates begin to synthesize IgM at an increased rate soon after birth. At about 6 days after birth, the serum concentration of IgM rises sharply. This rise continues until adult levels are achieved by approximately 1 yr of age. Serum IgA remains low throughout infancy. Maternal serum IgG gradually disappears during the 1st 6-8 mo of life until adult concentrations of total IgG are reached and maintained by 7-8 yr. The serum IgG level in infants usually reaches a low point at about 3-4 mo of postnatal life. The rate of development of IgE generally follows that of IgA.

6.(D) Flow cytometry is an important test to demonstrate the absence of circulating B cells , which will distinguish X-linked agammaglobulinemia from most types of CVID, the hyper-IgM syndrome, and transient hypogammaglobulinemia of infancy.

7.(A) Neutropenia, typically seen at diagnosis when infected, can be associated with Pseudomonas or staphylococcal infections.

8.(C) In contrast to XLA, the sex distribution in CVID is almost equal, the age at onset is later, and infections may be less severe**.** Enterovirus meningoencephalitis is rare in patients with CVID.

9.(B) CVID has also been associated with a sprue-like enteropathy with or without nodular lymphoid hyperplasia of the intestine. Other autoimmune diseases include alopecia areata, hemolytic anemia, thrombocytopenia, gastric atrophy, achlorhydria, and pernicious anemia.

Lymphoid interstitial pneumonia, intestinal lung disease, pseudo lymphoma, B-cell lymphomas, amyloidosis, and noncaseating sarcoid-like granulomas of the lungs, spleen, skin, and liver also occur. There is an increased risk of lymphomas.

10.(D) Patients may be asymptomatic or may develop sinopulmonary or gastrointestinal (GI) infections (especially Giardia).
11.(D) Serum antibodies to IgA are reported in as many as 44% of patients with selective IgA deficiency. These antibodies can cause nonhemolytic transfusion reactions. Washed erythrocytes (frozen blood would have this done routinely) or blood products from other IgA-deficient individuals should be administered to patients with IgA deficiency.
12.(E) In addition to opportunistic infections such as P. jiroveci pneumonia, there is an increased incidence of extensive verruca vulgaris lesions, Cryptosporidium enteritis, subsequent liver disease, and an increased risk of malignancy.
13.(B) First-line therapy is usually ganciclovir, with foscarnet as an alternative for resistant strains or ganciclovir intolerance.
14.(E)
15.(B) X-linked lymphoproliferative diseases have susceptibility to EBV and the development of hemophagocytic lymphohistiocytosis (HLH) . There are 3 major clinical phenotypes: (1) fulminant, often fatal, infectious mononucleosis (50% of cases); (2) lymphomas, predominantly involving B-lineage cells (25%); and (3) acquired hypogammaglobulinemia (25%).
16.(A) A combination of opportunistic infections and a persistently low lymphocyte count is an indication to test for SCID.
17.(E) The most likely diagnosis is Wiskott-Aldrich syndrome which is an X-linked recessive disorder characterized by atopic dermatitis, thrombocytopenic purpura with normal-appearing megakaryocytes but small defective platelets, and susceptibility to infection. The curative modality of treatment is HSCT.
18.(B) Ataxia-Telangiectasia is characterized by progressive cerebellar ataxia, oculocutaneous telangiectasias, chronic sinopulmonary disease, a high incidence of malignancy, and variable humoral and cellular immunodeficiency. Ataxia typically becomes evident soon after these children begin to walk and progresses until they are confined to a wheelchair, usually by age 10-12 yr. The telangiectasias begin to develop at 3-6 yr. The most frequent humoral immunologic abnormality is the selective absence of IgA, which occurs in 50–80% of these patients. Recurrent sinopulmonary infections occur in approximately 80% of these patients.
19.(C) Autosomal dominant hyper-IgE syndrome (Job syndrome) is characterized by an exceptionally high serum IgE concentration; an elevated serum IgD concentration; usually normal concentrations of IgG, IgA, and IgM.

20.(E) Deficiency of DOCK 8 (dedicator of cytokinesis 8) is an autosomal recessive condition that most often presents with impressively severe eczema in infancy and toddlerhood. Cutaneous viral infections and susceptibility to CMV, EBV, and cryptosporidia are common.

21.(A) Significant neutrophilic leukocytosis, often >250000/mm^3, is a prominent feature of severe forms of leukocyte adhesion deficiency (LAD). Affected children may have a history of delayed separation of the umbilical cord, usually with associated infection of the cord stump. The presence of significant omphalitis is an important feature that distinguishes these rare patients from the 10% of healthy infants who can have cord separation at age 3 wk or later. Infected areas characteristically have very little neutrophilic infiltration, limited inflammation, and no pus formation.

22.(D)

23.(C) LAD-3 is characterized by a Glanzmann thrombasthenia-like bleeding disorder.

24.(E) The most life-threatening complication of Chédiak-Higashi syndrome is the development of an accelerated phase characterized by pancytopenia, high fever, and lymphohistiocytic infiltration of liver, spleen, and lymph nodes. The onset of hemophagocytic lymphohistiocytosis can occur at any age. This occurs in 85% of patients and usually results in death.

25.(C) HSCT corrects hematopoietic and immunologic function, the NK cell deficiency, and prevent conversion to the accelerated phase, but cannot correct or prevent the neuropathy.

26.(B) Disseminated candidiasis presents frequently as a central venous catheter-associated infection. The most common presentation of invasive aspergillosis is pulmonary aspergillosis. As a nodule enlarges, the dense central core of infarcted tissue may become surrounded by edema or hemorrhage, forming a hazy rim known as the halo sign. When bone marrow function recovers, the infarcted central core may cavitate, creating the crescent sign.

27.(D) Management of infection in CGD is dramatically different than in normal children. CGD patients are always at risk for deep-seated, indolent bacterial infections, so determination of the appropriate treatment can be difficult. The erythrocyte sedimentation rate (ESR) can be quite helpful. If the child does not have a deep-seated infection, the ESR will be normal or will normalize within several days with standard management. Cultures should be obtained, but are usually negative and not helpful, so many support an “antibiotic sensitivity by sedimentation rate response” approach to treatment.

28.(D) Onset of venoocclusive disease is usually within 30 days of transplantation. Risk factors include young age, prior hepatic disease, abdominal radiation, repeated transplantations, neuroblastoma, osteopetrosis, and familial hemophagocytic lymphohistiocytosis. Prophylaxis has traditionally used ursodeoxycholic acid and occasionally heparin. Only defibrotide has demonstrated some efficacy in preventing and treating VOD. Defibrotide is a combination of porcine oligodeoxyribonucleotides that reduces procoagulant activity and enhances fibrinolytic properties of endothelial cells.

29.(C)

30.(B) Viruses causing acute neutropenia include influenzas A and B, adenovirus, respiratory syncytial virus, enteroviruses, human herpesvirus 6, measles, rubella, and varicella. Chronic neutropenia often accompanies infection with Epstein-Barr virus, cytomegalovirus, or HIV.

31.(C) Infections tend to be less frequent in autoimmune neutropenia (AIN) than with the corresponding degree of neutropenia from other causes, probably because remainder of the immune system remains intact and tissue delivery of neutrophils is greater than that in conditions resulting from impaired production. The neutropenia accompanying malignancy or following cancer chemotherapy is frequently associated with compromised cellular immunity and barrier compromise secondary to central venous lines and mucositis, thereby predisposing patients to a much greater risk of infection.

32.(A) Maculopapular rash is the only recognized skin manifestation of acute GVHD. All other distracters are cutaneous manifestations of chronic GVHD.

33.(A) Unlike leukemia, leukemoid reactions show relatively small proportions of immature myeloid cells, consisting largely of band forms, occasional metamyelocytes, and progressively rarer myelocytes, promyelocytes, and blasts. The presence of a left shift, defined as having >5% immature neutrophils in the peripheral blood, is consistent with marrow stress.

34.(E) The leukocyte alkaline phosphatase score of circulating neutrophils can differentiate chronic myelogenous leukemia, in which the level is uniformly almost zero, from reactive or secondary neutrophilia, which features normal to elevated levels.

35.(C) Typically, monocytosis occurs in patients recovering from myeloosuppressive chemotherapy and is a harbinger of the return of the neutrophil count to normal.

36.(D) Hypereosinophilic syndrome and systemic mastocytosis are additional important causes of an elevated eosinophil count.

37.(A) Thyrotoxicosis and Addison disease are endocrine disorders associated with lymphocytosis. Connective tissue disorders, allergic and hyper-inflammatory diseases, pulmonary disorders, and dermatologic conditions, Hypereosinophilic syndrome and systemic mastocytosis are additional important causes of an elevated eosinophil count.

38.(A) Hereditary angioedema is an autosomal dominant disease. It occurs in persons unable to synthesize normal levels of functional C1 inhibitor. Swelling of the affected part progresses rapidly, without urticaria, itching, or redness and often without severe pain. Swelling of the intestinal wall, however, can lead to intense abdominal cramping. Concomitant subcutaneous edema is often absent, and patients have undergone abdominal surgery or psychiatric examination before the true diagnosis was established. Laryngeal edema can be fatal. Attacks last 2-3 days. They may occur at sites of trauma, especially dental, after vigorous exercise, or with menses, fever, or emotional stress. Factor B deficiency is associated with atypical hemolytic uremic syndrome. MASP-1 deficiency is associated with recurrent pneumococcal infections. Properdin and C8b deficiencies are associated with Neisserial infections.

39.(D) An inhibitor of kallikrein (ecallantide) that blocks bradykinin production and an antagonist of the bradykinin receptor (icatibant) are approved in the United States for use in adolescents and adults for long-term prophylaxis, preparation for surgery or dental procedures, or treatment of acute attacks. Eculizumab, a humanized monoclonal antibody to C5, prevents generation of the membrane-attack complex C5b9 and is an effective treatment for paroxysmal nocturnal hemoglobinuria and atypical hemolytic uremic syndrome. Antihistamines, epinephrine, and corticosteroids have no effect.

40.(B) Autologous (from the same individual) transplantation is employed as a rescue strategy after delivering otherwise lethal doses of chemotherapy with or without radiotherapy in children with hematologic malignancies such as relapsed lymphoma or selected solid tumors (e.g., neuroblastoma, brain tumors). Allogeneic (from a donor) transplantation is used to treat children with genetic diseases of blood cells, such as hemoglobinopathies, primary immunodeficiency diseases, various inherited metabolic diseases, and bone marrow failure, hematologic malignancies (leukemia), and myelodysplastic syndromes.

Chapter 14

Allergic Disorders Questions

MARYAM Z. ALMUSAWI

1. Which of following signs is often seen in allergic children rubbing their nose upward with the palm of their hands?

A. Allergic cluck
B. Allergic salute
C. Allergic shiners
D. Dennie-morgan folds
E. Hyper linearity

2. Prominent infraorbital skin folds that extend in an arc from the inner canthu beneath and parallel to the lower lid margin is called

A. allergic shiners
B. chemosis
C. keratosis pilaris
D. Dennie lines
E. allergic salute

3. What is the **MOST** common skin abnormality of allergic children?

A. Xerosis
B. Keratosis pilaris
C. Hyperlinearity
D. Atopic dermatitis
E. Urticaria

4. Which of the following conditions is characterized by peripheral eosinophilia?

A. Sensitized patients after exposure to grass
B. Systemic corticosteroids use
C. Eosinophilic pneumonias
D. Eosinophilic esophagitis
E. Drug reaction

5. Which of the following is associated with decreased risk of asthma?

A. Delay introduction of egg
B. Reduced diversity of the intestinal microbiota during infancy
C. Exposure to dogs in early in childhood
D. Bottle feeding
E. Delivery by cesarean section

6. To avoid false-negative results when ordering epicutaneous skin tests, we should give instruction for the patient to withhold montelukast for

A. 1 day
B. 3 days
C. 5 days
D. 7 days
E. 14 days

7. Which of the following is a **MAJOR** risk factor for development of persistent asthma in childhood?

A. Allergic rhinitis
B. ≥4% peripheral blood eosinophils
C. Food allergen sensitization
D. Eczema
E. Wheezing apart from colds

8. Which of the following is characteristic to consider acute asthma attack as moderate?

A. Peak expiratory flow 50% of predicted
B. Pulse rate 150 beat /min
C. Loud Wheeze throughout inhalation and exhalation
D. Respiratory rate >30breaths/min
E. Confusion

9. A 10-year- boy presents with frequent attacks of shortness of breath, coughing, throat tightness, and audible laryngeal wheezing and stridor with provisional diagnosis of asthma treated with multiple different classes of asthma medications with poor response. His GP doctor referred him to asthma clinic for reevaluation. Spirometry was done which reveals truncated and inconsistent inspiratory and expiratory flow-volume loops while flexible

rhinolaryngoscopy reveals paradoxical vocal cord movements with anatomically normal vocal cords.
Of the following, the **MOST** likely diagnosis is

A. GERD
B. bronchomalacia
C. vocal cord dysfunction
D. hypersensitivity pneumonitis
E. severe persistent asthma

10. Which of the following lung function parameters is **MOST** consistent with asthma?

A. FEV1 /FVC ratio <0.60
B. Worsening in FEV1 ≥12% after exercise challenge
C. Daily peak expiratory flow (PEF) AM -to-PM variation ≥15%
D. Increase in FEV1 >12% after bronchodilator administration
E. High FEV1 (relative to percentage of predicted norms)

11. Which of the following statements regarding FeNO (Fractional Exhaled Nitric Oxide) is **TRUE**?

A. It is an invasive measure of allergic airways inflammation
B. It is a biomarker of allergic eosinophilic airways Inflammation
C. Children as young as 3 years can perform this test
D. Cannot predict future asthma exacerbations
E. It is a significant biomarker of cystic fibrosis

12. A 6-year-old asthmatic patient on medium dose budesonide as MDI since 4 weeks with no improvement in his condition.
Of the following, the **NEXT** step in his management is to

A. add monteleukast
B. reevaluate the inhaler device technique
C. continue same treatment and reevaluate after 2 months
D. increase budesonide dose
E. consider other diagnoses

13. Which of the following is the main component of asthma action plan?

A. Describing regular asthma medication use
B. Standardize assessment of asthma control
C. Understanding of the pathogenesis of asthma

D. Monitoring lung function
E. Determine comorbid conditions

14. Which of the following criteria in early childhood is a risk factor for development of persistent asthma?

A. Sibling asthma
B. High birth weight
C. Female gender
D. Breast feeding
E. Severe pneumonia

15. Which of the following is the most common chronic symptom of asthma?

A. Nasal congestion
B. Non focal chest pain
C. Expiratory wheezing
D. Productive coughing
E. Exercise induced wheeze

16. Which of the following is **TRUE** regarding the use of salmetrol in the treatment of asthma?

A. Used as monotherapy
B. Combined with monteleukast
C. Combined with inhaled corticosteroid
D. Used as reliever therapy
E. Onset of effect 5-10 min after administration

17. A 7- year-old asthmatic boy presents with dry cough, shortness of breath with inability to sleep last night, he used salbutamol inhaler 3 times over one hour with no improvement in symptoms and his PEF <80% of personal.
Of the following, the **MOST** appropriate next step is to

A. continue same treatment
B. use short course oral steroids
C. add montelukast
D. add Inhaled ipratropium
E. use intravenous aminophylline

18. What is the hallmark of atopic dermatitis?

A. Severe dry skin

B. Keratosis pilaris
C. Cradle cap
D. Nonpruritic papules
E. Greasy scaly erythematous lesion

19. Which of the following sites is mainly involved in infantile atopic dermatitis?
A. Extensor limb surface
B. Napkin area
C. Shoulder
D. Hands
E. Scalp

20. A 9-month-boy presents with irritability, erythematous scaly lesion over cheeks, the mother gives a history of egg allergy in her infant.
Which of the following is the first line therapy?
A. Pimecrolimus cream 1%
B. Tar Preparation
C. Diphenhydramine
D. Moisturing cream
E. Topical corticosteroid

21. Which of the following vitamins deficiency is associated with severe atopic dermatitis?
A. E
B. C
C. B1
D. B12
E. D

22. What is the best tolerated sport for a boy with atopic dermatitis?
A. Football
B. Swimming
C. Running
D. Baseball
E. Basketball

23. A 10-year-boy develops small, punctate pruritic wheals surrounded by a prominent erythematous flare after taking a hot shower, once the patient cools down the rash subsides in 30-60 min.
Of the following, the **MOST** likely diagnosis is

A. cholinergic urticaria
B. pressure urticaria
C. contact urticarial syndrome
D. acute papular urticaria
E. dermatographism

24. A 6-year-old boy develops small, yellow-tan to reddish brown macules on scratching his arms, what is the name of this sign?

A. Nikolsky
B. Darier
C. Candle
D. Auspitz
E. Buttonhole

25. Which of the following antibodies are found in patients with chronic urticaria more than general population?

A. Antihepatitis A
B. Antidouble strand
C. Antithyroid
D. Antinuclear
E. Antitransglutaminase

26. What is the main stay of treatment of chronic urticaria?

A. Steroid
B. H1 antihistamine
C. Monoclonal antibody
D. Montelukast
E. Plasmapheresis

27. An 8 year-old-well developed girl presented to outpatient clinic with shortness of breath and increasing progressive swallowing difficulty, there is no history of cough or flue like illness, just a visit to dentist for tooth extraction before one hour.
Of the following, the **MOST** likely cause of her condition is

A. hereditary angioedema
B. spasmodic croup
C. epiglotitis
D. covid 19
E. laryngeal hemangioma

28. A 10-year-boy develops a sudden attack of vomiting, diarrhea, and shortness of breath after attending a birthday party when he ate a cake containing some pistachio butter.
Of the following, the **MOST** appropriate first line measure is
A. ORS solution
B. diphenhydramine
C. gentamycin
D. IM adrenaline
E. ranitidine

29. After managing a patient with severe anaphylaxis in the emergency department, he should be kept for observation before being discharged for at least
A. 2 hour
B. 4 hour
C. 8 hour
D. 12 hour
E. 24 hour

30. Adrenaline is a first line drug used for anaphylactic reaction, its side effect can include
A. bradycardia
B. hypotension
C. headache
D. flushing
E. dry skin

31. Serum sickness is a classic example of
A. type I hypersensitivity reaction
B. type II hypersensitivity reaction
C. type III hypersensitivity reaction
D. type IV hypersensitivity reaction

E. autoimmune disease

32. A 5-year-boy developed fever and sore throat diagnosed as bacterial tonsillitis and received injectable penicillin, after 10 days he gets low grade fever , nausea, diarrhea, malaise, and melena associated with morbilliform rash and arthralgia involving multiple joints. ESR and CRP are high while CBC shows thrombocytopenia.
Of the following, the **MOST** likely diagnosis is

A. Kawasaki disease
B. rheumatic fever
C. sepsis
D. Lyme disease
E. serum sickness

33. Adverse food reaction can be divided into food intolerance and food allergy, which of the following is an example of food intolerance?

A. Food protein‑induced enterocolitis
B. Enteropathy syndromes
C. Galactosemia
D. Oral allergy syndrome
E. Allergic eosinophilic esophagitis

34. A 7- month-old baby boy typically manifest as irritability, intermittent vomiting that occur 1-4 hour after feeding,with bloody diarrhea , anemia, and failure to thrive, the baby is bottle fed with complementary feeding. Investigations show anemia and hypoalbuminemia with normal serum IgE.
Of the following, the **MOST** likely diagnosis is

A. food protein‑induced enterocolitis syndrome
B. proctocolitis
C. enteropathy
D. eosinophilic gasteroenteropathies
E. celiac disease

35. A 2-month-baby girl presented by her anxious mother when she found blood-streak in her daughter stool, she is exclusively breast fed, healthy well looking baby, her investigations are normal apart from mild anemia.
Of the following, the **MOST** likely diagnosis is

A. gastroenteritis

B. food protein–induced allergic proctocolitis
C. enteropathy
D. eosinophilic gasteroenteropathies
E. food protein‑induced enterocolitis syndrome

36. Which of the following vaccines is contraindicated in children with egg allergy?
A. Yellow fever
B. Measles
C. Influenza
D. Rabies
E. MMR

37. An educated family with a history of food allergy seeks your advice to prevent allergy in their expected daughter.
Your proper advice is to
A. avoid allergenic foods during pregnancy
B. use soy-based formulas to prevent allergic diseases
C. delay introduction of potentially highly allergenic foods
D. exclusively breast feed for first 8 months
E. introduce solid (complementary) foods after 4-6 mo

38. Adverse drug reactions can be divided into predictable and unpredictable reactions.
Which of the following is an example of predictable drug reactions?
A. Drug toxicity
B. Allergic (hypersensitivity) reactions
C. Pseudoallergic reactions
D. Idiosyncratic reactions
E. Dose independent reactions

39. When the adverse drug reactions are mediated by drug specific T lymphocytes, this interaction is classified as
A. Immune complex reactions
B. Delayed-type hypersensitivity reactions
C. Cytotoxic antibody reactions
D. Immediate hypersensitivity reactions
E. Serum sickness

40. A 6-year-old boy develops fever, maculopapular rash, facial edema, and generalized lymphadenopathy. Investigations reveal increment in SGPT and SGOT with eosinophilia. He had history of epilepsy treated with valproic acid 2 months ago.

What is the **MOST** likely diagnosis?

A. Kawasaki disease
B. Hodgkins lymphoma
C. DRESS syndrome
D. Brucellosis
E. Typhoid fever

Chapter 14

Allergic Disorders Answers

MARYAM Z. ALMUSAWI

1.(B) Because of nasal pruritus and rhinorrhea, children with allergic rhinitis often perform the allergic salute by rubbing their nose upward with the palm of their hand. This repeated maneuver may give rise to the nasal crease, a horizontal wrinkle over the bridge of the nose.

2.(D) Allergic shiners (blue-gray to purple discolorations beneath their child's lower eyelids, which can be attributed to venous stasis found in up to 60% of allergic patients and almost 40% of patients without allergic disease) Dennie-Morgan folds (Dennie lines) are a feature of atopic dermatitis.

3.(A) Xerosis , or dry skin, is the most common skin abnormality of allergic children.

4.(A) In certain pathologic conditions, such as drug reactions, eosinophilic pneumonias, and eosinophilic esophagitis, significantly increased numbers of eosinophils may be present in the target organ in the absence of peripheral blood eosinophilia.

5.(C) There is also a decreased risk of asthma, AR, and atopic sensitization with early introduction to wheat, rye, oats, barley, fish, and eggs.

6.(A) Montelukast should be withheld for 1 day, most sedating antihistamine preparations for 3-4 days, and nonsedating antihistamines for 5-7 days

7.(D) Early childhood risk factors for persistent asthma have been identified and have been described as major (parent asthma, eczema, inhalant allergen sensitization)and minor (allergic rhinitis, wheezing apart from colds, ≥4% peripheral blood eosinophils, food allergen sensitization) risk factors.

8.(A) Approx. 40–69% or response lasts <2 hr.

9.(C) Vocal cord dysfunction (VCD) can manifest as intermittent daytime wheezing. The vocal cords involuntarily close inappropriately during inspiration and sometimes exhalation, producing shortness of breath, coughing, throat tightness, and often audible laryngeal wheezing and/or stridor. In most cases of VCD, spirometric lung function testing reveals truncated and inconsistent inspiratory and expiratory flow-volume loops, a pattern that differs from the reproducible pattern of airflow limitation in asthma that improves with

bronchodilators. VCD can coexist with asthma. Hypercarbia and severe hypoxia are uncommon in VCD. Flexible rhinolaryngoscopy in the patient with symptomatic VCD can reveal paradoxical vocal cord movements with anatomically normal vocal cords. Prior to the diagnosis, patients with VCD are often treated unsuccessfully with multiple different classes of asthma medications. This condition can be well managed with specialized speech therapy training in the relaxation and control of vocal cord movement.

10.(D)

Spirometry (in clinic):

• Low FEV1 (relative to percentage of predicted norms)

• FEV1 /FVC ratio <0.80

Bronchodilator response (to inhaled β-agonist) assesses reversibility of airflow limitation.

Reversibility is determined by an increase in either FEV1 >12% or predicted FEV1 >10% after inhalation of a short-acting β- agonist (SABA)

Exercise challenge:

• Worsening in FEV1 ≥15%

Daily peak expiratory flow (PEF) or FEV1 monitoring: day-to-day and/or AM -to-PM variation ≥20%*

11.(B)

12.(B)

13.(A) Two main components:

(1) a daily "routine" management plan describing regular asthma medication use and other measures to keep asthma under good control; and

(2) an action plan to manage worsening asthma, describing indicators of impending exacerbations, identifying what medications to take, and specifying when and how to contact the regular physician and/or obtain urgent/emergency medical care.

14.(E)

Early Childhood Risk Factors for Persistent Asthma

Parental asthma*

Allergy: (Atopic dermatitis (eczema)*, Allergic rhinitis, Food allergy, Inhalant allergen sensitization*, Food allergen sensitization)

Severe lower respiratory tract infection: (Pneumonia, Bronchiolitis requiring hospitalization)

Wheezing apart from colds

Male gender

Low birthweight

Environmental tobacco smoke exposure
Reduced lung function at birth
Formula feeding rather than breastfeeding

15.(C) Intermittent dry coughing and expiratory wheezing are the most common chronic symptoms of asthma.

16.(C)

17.(B)

18.(A)

19.(A)

20.(D) Moisturizers are first-line therapy for treatment of atopic dermatitis.

21.(E) Vitamin D deficiency often accompanies severe atopic dermatitis.

22.(B) A sport such as swimming may be better tolerated than others that involve intense perspiration, physical contact, or heavy clothing and equipment. Rinsing off chlorine immediately and lubricating the skin after swimming are important.

23.(A)

24.(B)

25.(C) The incidence of elevated thyroid antibodies in patients with chronic urticaria is approximately 12%, compared with 3–6% in the general population. Although some patients show clinical reduction of the urticaria with thyroid replacement therapy, others do not. The role of thyroid autoantibodies in chronic urticaria is uncertain; their presence may reflect a tendency of the patient to develop autoantibodies, but they may not play a direct role in chronic urticaria.

26.(B)

27.(A) Dental work with the injection of procaine hydrochloride (Novocain) into the gums is a common precipitant for laryngeal edema in patient with hereditary angioedema.

28.(D) Epinephrine is the most important medication, and there should be no delay in its administration in case of anaphylaxis. Epinephrine should be given by the IM route to the lateral thigh (1 : 1000 dilution, 0.01 mg/kg; maximum 0.5 mg).

29.(B) More than 90% of biphasic responses occur within 4 hr, so patients should be observed for at least 4 hr before being discharged from the emergency department.

30.(C)

31.(C)

32.(E) Serum sickness generally begin 7-12 days after injection of the foreign material(drugs or proteins from other species).

33.(C)

34.(A)

35.(B)

36.(A)

37.(E) Prevention of Food Allergy

-Breastfeed exclusively for 4-6 mo.

-Introduce solid (complementary) foods after 4-6 mo of exclusive breastfeeding.

-Introduce low-risk complementary foods 1 at a time.

-Introduce potentially highly allergenic foods (fish, eggs, peanut, milk, wheat) soon after the lower-risk foods (no need to avoid or delay).

-Do not avoid allergenic foods during pregnancy or nursing.

-Soy-based formulas do not prevent allergic disease.

38.(A) Predictable drug reactions , including drug toxicity, drug interactions, and adverse effects, are dose dependent, can be related to known pharmacologic actions of the drug, and occur in patients without any unique susceptibility. Unpredictable drug reactions are dose independent, often are not related to the pharmacologic actions of the drug, and occur in patients who are genetically predisposed. These include idiosyncratic reactions, allergic (hypersensitivity) reactions, and pseudoallergic reactions.

39.(B)

40.(C)

Chapter 15

Rheumatic Diseases of Childhood Questions

ZUHAIR ALMUSAWI

1. If arthralgia is accompanied with a history of dry skin, hair loss, fatigue, growth disturbance, or cold intolerance, testing for which of the following is merited?

A. Infection
B. Thyroid disease
C. Malignancy
D. Orthopedic conditions
E. Pain syndromes

2. An evanescent macular rash associated with fever is part of the diagnostic criteria for which of the following?

A. Henoch-Schönlein purpura
B. Juvenile dermatomyositis
C. Systemic-onset arthritis
D. Systemic lupus erythematosus
E. Mixed connective tissue disease

3. Which of the following antibody tests has a high specificity for SLE; associated with lupus nephritis?

A. Double-stranded DNA (dsDNA)
B. Antinuclear antibody (ANA)
C. Ribonuclease protein (RNP)
D. Antineutrophil cytoplasmic antibodies (ANCAs)
E. Smith (Sm)

4. How much is the sensitivity of rheumatoid factor (RF) as a diagnostic tool in children with Juvenile idiopathic arthritis?

A. <10%
B. <20%
C. <30%

D. <40%
E. <50%

5. What is the **MOST** frequent adverse effect of NSAIDs in children?
A. Gastritis
B. Nausea
C. Mood change
D. Irritability
E. Anemia

6. Which of the following NSAID may cause aseptic meningitis when used to treat patients with lupus?
A. Naproxen
B. Celecoxib
C. Ibuprofen
D. Indomethacin
E. Meloxicam

7. Which of the following is given as an adjunct to minimize adverse effects of methotrexate?
A. Zinc
B. Vitamin B12
C. Vitamin C
D. Folic acid
E. Vitamin D

8. What is the **MOST** significant potential adverse effect of Hydroxychloroquine?
A. Skin discoloration
B. Retinal toxicity
C. Gastric irritation
D. Bone marrow suppression
E. Myositis

9. What is the intravenous corticosteroid of choice to treat severe, acute manifestations of systemic rheumatic diseases?
A. Hydrocortisone
B. Methylprednisolone
C. Dexamethasone

D. Betamethasone
E. Mometasone

10. How long arthritis must be present to make a diagnosis of any Juvenile idiopathic arthritis subtype?
A. ≥2 weeks
B. ≥4 weeks
C. ≥6 weeks
D. ≥8 weeks
E. ≥12 weeks

11. A 4-year-old girl presents with limitation in range of motion, tenderness, and warm swollen right knee joint with left ankle joint for the last 8 weeks. She also complains from easy fatigability and poor sleep quality. ANA is positive.
Of the following, the **MOST** appropriate measure to be done is
A. periodic slit-lamp examination
B. genetic study
C. rheumatoid factor study
D. echo study
E. MRI of involved joints

12. Which of the following Juvenile idiopathic arthritis JIA subtype is associated with a worse prognosis?
A. Persistent oligoarticular JIA
B. Extended oligoarticular JIA
C. Enthesitis related arthritis
D. Polyarticular JIA
E. Systemic JIA

13. An 18-month-old boy presents with arthritis, spiking fever with temperatures ≥39°C for the last 3 weeks, the fever is often present in the evening and is frequently accompanied by a characteristic faint, erythematous, macular rash distributed over the trunk and proximal extremities. On examination, the baby has hepatosplenomegaly, lymphadenopathy, and pericarditis documented by echo.
Of the following, the **MOST** likely diagnosis is
A. oligoarticular JIA
B. rheumatic fever

C. serum sickness
D. systemic JIA
E. Kawasaki disease

14. An 18-month-old child diagnosed as systemic juvenile idiopathic arthritis one month ago and is in remission with treatment, today presents with high-spiking fever, lymphadenopathy, hepatosplenomegaly, purpura, and encephalopathy. Laboratory evaluation shows thrombocytopenia, leukopenia, and hypofibrinogenemia with elevated liver enzymes, lactate dehydrogenase, ferritin, and triglycerides. A bone marrow aspiration shows evidence of hemophagocytosis.
Of the following, the **MOST** likely diagnosis is
A. immune-mediated thrombocytopenia
B. macrophage activation syndrome
C. autoimmune hepatitis
D. familial hypertriglyceridemia
E. visceral leishmaniasis

15. Which of the following laboratory features is useful in distinguishing macrophage activation syndrome (MAS) from a flare of systemic disease?
A. Cytopenias
B. Abnormal liver function tests
C. Coagulopathy
D. Decreased erythrocyte sedimentation rate
E. Hypertriglyceridemia

16. Which of the following viral diseases is a recognized cause of arthritis?
A. Measles
B. Mumps
C. Chickenpox
D. Roseola infantum
E. Hepatitis A

17. If a patient with oligoarticular JIA shows no response to NSAIDs and injections, what is the next step treatment?
A. Etanercept
B. Abatacept
C. Methotrexate

D. Rituximab
E. Anakinra

18. A 5-year-old girl presents with arthritis of her two knees and right ankle, nail pitting and "sausage digit" of the left index finger with lower back pain for the last 3 months, ophthalmic examination shows anterior uveitis. Antinuclear antibody (ANA) is positive.
Of the following, the **MOST** likely diagnosis is
A. juvenile ankylosing spondylitis
B. juvenile psoriatic arthritis
C. inflammatory bowel disease
D. reactive arthritis
E. persistent oligoarticular JIA

19. What is the gold standard imaging for early visualization of sacroiliitis?
A. MRI with gadolinium
B. Short-T1 inversion recovery (STIR) MRI
C. Conventional radiographs
D. CT scan
E. Bone scan

20. Which of the following if used initially and continually may slow the progression of structural damage spondyloarthritis (SpA)?
A. Methotrexate
B. Etanercept
C. Naproxen
D. Rituximab
E. Anakinra

21. A 5-year-old boy presents with acute onset of severe pain in the right groin, referred to the thigh for the last week, the patient had history of an upper respiratory tract infection 2 weeks ago. ESR and white blood cell count are normal, while ultrasound examination confirm widening of the joint space; aspiration of joint fluid was normal and results in dramatic clinical improvement
Of the following, the **MOST** likely diagnosis is
A. toxic synovitis
B. poststreptococcal arthritis
C. septic arthritis

D. reactive arthritis
E. acute rheumatic fever

22. A 5-year-old boy presents with fever, raised tender erythematous plaques and nodules over the face, extremities, and trunk together with arthritis, and localized muscular pain and tenderness. Laboratory investigations reveals ESR 50 mm/hr, positive C-reactive protein test, WBC 20000 leukocytes/mm3 with 75% neutrophils/mm3
Of the following, the **MOST** likely diagnosis is

A. Sweet syndrome
B. pityriasis lichenoides et varioliformis acuta
C. juvenile primary fibromyalgia syndrome
D. granulomatosis with polyangiitis
E. Churg-Strauss syndrome

23. Which of the following is a suggested major criterion for relapsing polychondritis?

A. Eye inflammation
B. Hearing loss
C. Vestibular dysfunction
D. Seronegative inflammatory arthritis
E. Typical inflammatory episodes of nose cartilage

24. ANA is very sensitive for SLE (95–99%), but its specificity is

A. 10%
B. 30%
C. 50%
D. 70%
E. 90%

25. Which of the following is **MORE** common in drug-induced lupus compared with SLE?

A. High percent of positive dsDNA antibodies
B. Hypocomplementemia
C. Significant renal disease
D. Hepatitis
E. Significant neurologic disease

26. Which of the following medications has definite association with drug-induced lupus?

A. Isoniazid
B. Phenytoin
C. Carbamazepine
D. Nitrofurantoin
E. β-blockers

27. Which of the following tests correlate well with SLE disease activity?

A. ANA titers
B. Anti-dsDNA
C. Anti-Smith antibody
D. C-reactive protein (CRP)
E. Antihistone antibodies

28. Which of the following is **MOST** appropriate for the treatment of lupus nephritis?

A. Hydroxychloroquine
B. Cyclophosphamide
C. Corticosteroids
D. Methotrexate
E. Rituximab

29. A 5-week-old infant presents with a characteristic macular rash affecting the periorbital area, trunk, and scalp. The neonate also has thrombocytopenia, leukopenia, and elevated liver enzymes; ECG and echo reveals complete heart block, while maternal serum demonstrates anti-Ro and anti-La antibodies.
Of the following, the **MOST** appropriate management of this infant is

A. IVIG
B. corticosteroids
C. cardiac pacing
D. cardiac transplantation
E. hydroxychloroquine

30. A 7-year-old girl presents with generalized erythema in sun-exposed areas over the chest, neck, knees and elbows with a heliotrope rash of the eyelids that is associated with periorbital edema. The girl also has fever, dysphagia, arthritis,

fatigue, muscle weakness and tenderness, with difficulty in climbing stairs and combing hair.
Of the following, the **MOST** likely diagnosis is

A. juvenile polymyositis
B. juvenile dermatomyositis
C. SLE
D. juvenile psoriatic arthritis
E. enthesitis related arthritis

31. Which of the following is usually elevated on initial presentation of juvenile dermatomyositis?

A. CK
B. ESR
C. Rheumatoid factor
D. ALT
E. CRP

32. A 10-year-old child diagnosed as juvenile dermatomyositis for the last 2 years with poor compliance and under treatment, develops painful ulceration of the skin of right lower calf area near ankle joint with extrusion of crystals.
Of the following, the **MOST** likely cause is

A. cellulitis
B. osteomyelitis
C. lipodystrophy
D. calcinosis
E. trophic ulcer

33. A 6-year-old boy presents with induration of the skin involving the neck, shoulders, face, trunk, and extending to the arms. The child looks well with no constitutional symptoms apart from a febrile streptococcal pharyngitis 10 days ago.
Of the following, the **MOST** likely diagnosis is

A. pseudoscleroderma
B. juvenile systemic sclerosis
C. subcutaneous morphea
D. scleredema
E. En coup de sabre

34. An 8-year-old boy gave history of edema and erythema of left lower limb, now presents with indurated, hypopigmented atrophic lesions extending from middle thigh through knee joint to above ankle resulting in knee contracture with chronic skin breakdown of scar on the lateral knee.
Of the following, the **MOST** likely diagnosis is

A. linear scleroderma
B. generalized morphea
C. subcutaneous morphea
D. En coup de sabre
E. eosinophilic fasciitis

35. What is the **MOST** frequent initial symptom in pediatric systemic sclerosis?

A. Dyspepsia
B. Raynaud phenomenon
C. Dyspnea at rest
D. Dysphagia
E. Proximal muscle weakness

36. A 12-year-old boy presents with recurrent, multiple, painful oral ulcers ranging from 2-10 mm, which last 3-10 days and heal without scarring. He also has blurred vision, redness, periorbital pain, and photophobia, together with papulopustular acneiform lesions, erythema nodosum, folliculitis, purpura, and ulcers.
Which of the following significantly improves oral ulcers and skin features of this disease?

A. Cyclophosphamide
B. Colchicine
C. Corticosteroids
D. Azathioprine
E. Sucralfate

37. A 10-year-old girl presents with recurrent parotid gland enlargement, dry mouth, halitosis, widespread dental caries, dry eyes, recurrent conjunctivitis, and arthralgia. CBC shows leukopenia and high ESR while serological studies reveal high-titer ANA, positive rheumatoid factor, elevated serum amylase, hypergammaglobulinemia, and positive anti–β-fodrin autoantibodies.
Of the following, the **MOST** likely diagnosis is

A. juvenile recurrent parotitis

B. Sjögren syndrome
C. HIV
D. polycystic parotid disease
E. sarcoidosis

38. A 6-year-old boy presents with intermittent unilateral parotid swelling lasting few days associated with fever. Parotid biopsy shows a lack of focal lymphocytic infiltrates.
Of the following, the **MOST** likely diagnosis is

A. juvenile recurrent parotitis
B. Sjögren syndrome
C. eating disorders
D. mumps
E. trauma to the buccal mucosa

39. A 13-year-old Syrian boy presents with recurrent, short-lived (1-3 days), self-limited episodes of fever, unilateral pleuritic chest pain, severe generalized abdominal pain, arthritis of knee, and an erysipeloid erythematous rash that overlies the dorsum of the foot. Other clinical findings include scrotal pain, exertional leg pain. Between flares, the boy is generally symptom free but has persistent elevation of ESR.
Of the following, the **MOST** appropriate treatment for this boy is

A. rilonacept
B. anakinra
C. colchicine
D. corticosteroids
E. azathioprine

40. Which of the following is a diagnostic Indicator of Tumor Necrosis Factor Receptor–Associated Periodic Syndrome (TRAPS)?

A. Recurrent episodes of inflammatory symptoms spanning >6 mo duration
B. Episodes last >10 days on average
C. Responsive to colchicine
D. Affects family members in autosomal recessive pattern
E. More in Caucasians

41. A 5-year-old child presents with recurrent fevers, sterile osteomyelitis, congenital dyserythropoietic anemia (CDA), neutrophilic dermatosis, failure to

thrive, and hepatomegaly. The parents are cousins and genetic study reveals mutations in the LPIN2 gene.
Of the following, the **MOST** likely diagnosis is

A. macrophage activation syndrome
B. Majeed syndrome
C. deficiency of interleukin-36 receptor antagonist
D. deficiency of adenosine deaminase 2
E. Blau syndrome

42. A 4-year-old child presents with recurring episodes of fever, malaise, exudative-appearing tonsillitis, cervical lymphadenopathy, oral aphthae, headache, abdominal pain, and arthralgia. The episodes last 4-6 days and often recur every 3-6 weeks cycles. Laboratory findings reveal mild leukocytosis, elevated acute-phase reactants, and negative throat cultures.
Which of the following drugs shows dramatic response in this child?

A. Azathioprine
B. Cimetidine
C. Anakinra
D. Prednisone
E. Colchicine

43. Which of the following is **MOSTLY** affected by amyloid A (AA) amyloidosis?

A. Testes
B. Adrenals
C. Heart
D. Skin
E. Kidneys

44. What is the **MOST** common chest radiograph finding in sarcoidosis?

A. Parenchymal infiltrates
B. Bilateral hilar adenopathy
C. Miliary nodules
D. Pleural effusion
E. Multiple pneumatoceles

45. A 3-year-old child presents with symmetric polyarticular arthritis involving large and small joints followed by photophobia and red eye. The parents gives a history of skin rash starting in infancy in the form of red-brown to purple

maculopapular lesions <1 cm on the face, upper back, and extremities. CBC reveals anemia, leukopenia, eosinophilia, and high ESR while serological exam shows hypergammaglobulinemia and hypercalcemia.
Of the following, the **MOST** likely diagnosis is

A. juvenile idiopathic arthritis
B. early-onset sarcoidosis
C. Wegener granulomatosis
D. chronic berylliosis
E. chronic granulomatous disease

46. Which of the following is a predictor of poor outcome in Kawasaki disease?

A. Female gender
B. Neutropenia
C. Young age
D. Thrombocytosis
E. Hypernatremia

47. Which of the following clinical features is consistent with Kawasaki disease?

A. Exudative conjunctivitis
B. Erythema and cracking of lips
C. Generalized lymphadenopathy
D. Splenomegaly
E. Vesicular rash

48. Patients with acute Kawasaki disease should be treated with IVIG within how many days of disease onset?

A. 5
B. 7
C. 10
D. 14
E. 21

49. A 5-year-old boy presents with symmetric palpable purpura and petechiae, on lower extremities, extensor aspect of the upper extremities, and buttocks with edema of hands and feet, periorbital area, lips, and scrotum, he also has right knee and left ankle swellings with bouts of severe abdominal pain and melena. CBC is normal with mild elevation of ESR and CRP.
Of the following, the **MOST** appropriate treatment now is

A. ibuprofen
B. corticosteroids
C. intravenous immune globulin
D. azathioprine
E. cyclosporine

50. An 18-month-old well-looking child presents with fever; tender edema of the face, hands, and feet; with large ecchymotic spots on the face and extremities sparing the trunk. CBC and urinalysis results were normal.
Of the following, the **MOST** likely diagnosis is

A. Churgstrauss syndrome
B. hypocomplementemic urticarial vasculitis
C. hypersensitivity vasculitis
D. infantile acute hemorrhagic edema
E. Henoch Schönlein purpura

51. A 15-year-old boy presents with fever, malaise, weight loss, headache, myalgia, arthralgia, dizziness, and abdominal pain; examination reveals decreased peripheral radial pulse with claudication of extremities, blood pressure in right arm 160/105 while in left arm 140/90. Magnetic resonance angiography reveals angiographic abnormalities of the aorta.
Of the following, the **MOST** likely diagnosis is

A. Takayasu arteritis
B. temporal arteritis
C. fibromuscular dysplasia
D. coarctation of aorta
E. polyarteritis nodosa

52. A 10-year-old boy presents with weight loss, severe abdominal pain, hematuria, arthralgia, myalgia with superficial and deep skin ulcers, tender subcutaneous nodules, digital necrosis, and splinter hemorrhages. He gave a history of transient loss of consciousness for few minutes 3 months ago. His blood pressure is 140/100; urinalysis shows proteinuria with red blood cell casts. Abdominal aortogram shows bilateral renal artery aneurysms.
What is the mainstay of therapy for this boy?

A. Oral cyclophosphamide
B. Plasma exchange
C. Methotrexate

D. Oral prednisone
E. IVIG

53. A 14-year-old girl presents with fever, malaise, weight loss, arthralgias, cough, wheezing, dyspnea, and hemoptysis. Examination shows palpable purpura, nasal ulceration, epistaxis, saddle nose, and hearing loss while ophthalmic examination reveals conjunctivitis, scleritis, and uveitis. Her blood pressure 140/100, urinalysis shows proteinuria with hematuria.
Of the following, the **MOST** likely diagnosis is

A. Henoch-Schönlein purpura
B. granulomatosis with polyangiitis
C. Churg-Strauss syndrome
D. microscopic polyangiitis
E. Goodpasture syndrome

54. Which of the following differentiate Churg-Strauss syndrome from granulomatosis with polyangiitis?

A. Small vessel vasculitis
B. Circulating antineutrophil cytoplasmic antibodies
C. Granulomatous inflammation
D. Necrotizing vasculitis
E. Eosinophilia

55. A 7-year-old boy presents with intermittent and bilateral cramping pain, affecting the anterior thigh, shin, and calf mostly in late afternoon or evening. Pain may wake the child from sleep and may last a few minutes to hours, but resolves quickly with massage. A worthy note pain is never present the following morning. Physical findings are normal, and gait is not impaired.
Of the following, the **MOST** likely diagnosis is

A. growing pain
B. restless leg syndrome
C. idiopathic musculoskeletal pain syndrome
D. fibromyalgia
E. complex regional pain syndrome

56. A 15-year-old boy comes to you complaining from uncomfortable painful leg sensations, worsening with periods of rest and at night and is relieved by movement. His mother gives history of same complaint during adolescence.

Which of the following options may benefit this boy?

A. Reassurance
B. Healthy sleep hygiene
C. Massage during the episode
D. Iron supplementation
E. NSAIDs agents

Chapter 15

Rheumatic Diseases of Childhood Answers

ZUHAIR ALMUSAWI

1.(B) Arthralgias are common in childhood and are a frequent reason for referral to pediatric rheumatologists. Arthralgias without physical findings for arthritis suggest infection, malignancy, orthopedic conditions, benign syndromes, or pain syndromes such as fibromyalgia. Although rheumatic diseases may manifest as arthralgias, arthritis is a stronger predictor of the presence of rheumatic disease and a reason for referral to a pediatric rheumatologist.

2.(C)

3.(A)

4.(A) Rheumatoid factor (RF) is present in <10% of children with JIA and thus has poor sensitivity as a diagnostic tool; RF may be elevated by infections such as endocarditis, tuberculosis, syphilis, and viruses (parvovirus B19, hepatitis B and C, mycoplasma), as well as primary biliary cirrhosis and malignancies. In a child with chronic arthritis, RF serves as a prognostic indicator.

5.(B) The most frequent adverse effects of NSAIDs in children are nausea, decreased appetite, and abdominal pain. Gastritis or ulceration occurs less frequently in children. Less common adverse effects (≤5% of children undergoing long-term NSAID therapy), include mood change, concentration difficulty that can simulate attention deficit disorder, sleepiness, irritability, headache, tinnitus, alopecia, anemia, elevated liver enzyme values, proteinuria, and hematuria.

6.(C) Aseptic meningitis has been associated with ibuprofen, primarily in patients with lupus. Naproxen is more likely than other NSAIDs to cause a unique skin reaction called pseudoporphyria.

7.(D)

8.(B) The most significant potential adverse effect is retinal toxicity, which occurs rarely but results in irreversible color blindness or loss of central vision.

9.(B) Methylprednisolone, 10-30 mg/kg/dose up to a maximum of 1 g, given over 1 hr daily for 1-5 days, is the IV preparation of choice.

10.(C)

11.(A) The presence of a positive antinuclear antibody (ANA) test confers increased risk for asymptomatic anterior uveitis, requiring periodic slit-lamp examination. ANA positivity may also be correlated with younger age at disease onset, female sex, asymmetric arthritis, and fewer involved joints over time.

12.(B) Those in whom disease never develops in >4 joints are regarded as having persistent oligoarticular JIA , whereas evolution of disease in >4 joints after 6 mo changes the classification to extended oligoarticular JIA and is associated with a worse prognosis. Isolated involvement of the hip is almost never a presenting sign and suggests enthesitis related arthritis ERA or a nonrheumatic cause.

13.(D) The classic rash is nonpruritic and migratory with lesions lasting <1 hr. Koebner phenomenon, a cutaneous hypersensitivity in which classic lesions are brought on by superficial trauma, is often present.

14.(B) Macrophage activation syndrome (MAS) is a rare but potentially fatal complication of sJIA that can occur at any time (onset, medication change, active or remission) during the disease course. It is also referred to as secondary hemophagocytic syndrome or hemophagocytic lymphohistiocytosis (HLH).

15.(D) The erythrocyte sedimentation rate (ESR) falls because of hypofibrinogenemia and hepatic dysfunction, a feature useful in distinguishing MAS from a flare of systemic disease.

16.(B) Viral illness causing arthritis or extremity pain (parvovirus, rubella, mumps, Epstein-Barr, hepatitis B, chikungunya)

17.(C) A substantial fraction of patients with oligoarthritis show no response to NSAIDs and injections, and therefore require treatment with disease modifying antirheumatic drugs (DMARDs), including methotrexate, and, if no response, TNF inhibitors.

18.(B) Children have psoriatic arthritis if they have arthritis and psoriasis or arthritis and at least 2 of the following: (1) dactylitis, (2) nail pitting or onycholysis, and (3) psoriasis in a first-degree relative. The presence of psoriasis aids in diagnosis but is not required.

19.(B) The gold standard for early visualization of sacroiliitis is evidence of bone marrow edema adjacent to the joint on MRI with fluid sensitive sequences such as short-T1 inversion recovery (STIR). Gadolinium does not add value to the study of the sacroiliac joints if STIR is used. MRI will reveal abnormalities before the plain radiograph.

20.(C) NSAIDs, such as naproxen (15-20 mg/kg/day), are frequently used initially and may slow the progression of structural damage (syndesmophyte formation and growth) if used continually.

21.(A)

22.(A) Diagnostic Criteria for Classic Sweet Syndrome

Major Criteria

Abrupt onset of painful erythematous plaques or nodules

Histopathologic evidence of dense neutrophilic infiltrate without evidence of leukocytoclastic vasculitis

Minor Criteria

Pyrexia >38°C

Association with underlying hematologic or visceral malignancy, inflammatory disease or pregnancy, or preceded by an upper respiratory or gastrointestinal infection or vaccination

Excellent response to systemic corticosteroids or potassium iodide

Abnormal laboratory values at presentation (3 of 4):

Erythrocyte sedimentation rate >20 mm/hr

Positive C-reactive protein test result

>8,000 leukocytes/mm3

>70% neutrophils/mm3

The diagnosis is established by the presence of 2 major criteria plus 2 of the 4 minor criteria.

23.(E) Suggested criteria for relapsing polychondritis

MAJOR

- ✓ Typical inflammatory episodes of ear cartilage
- ✓ Typical inflammatory episodes of nose cartilage
- ✓ Typical inflammatory episodes of laryngotracheal cartilage

MINOR

- ✓ Eye inflammation (conjunctivitis, keratitis, episcleritis, uveitis)
- ✓ Hearing loss
- ✓ Vestibular dysfunction
- ✓ Seronegative inflammatory arthritis

The diagnosis is established by the presence of 2 major or 1 major and 2 minor criteria. Histologic examination of affected cartilage is required when the presentation is atypical.

24.(C) Of note, although a positive antinuclear antibody (ANA) test result is not required for the diagnosis of SLE, ANA-negative lupus is extremely rare. ANA is very sensitive for SLE (95–99%), but it is not very specific (50%). The ANA may be positive many years before a diagnosis of SLE is established. However, most asymptomatic, ANA-positive patients do not have SLE or other autoimmune disease.

25.(D) Hepatitis, which is rare in SLE, is more common in drug-induced lupus. Individuals with drug-induced lupus are less likely to demonstrate antibodies to dsDNA, hypocomplementemia, and significant renal or neurologic disease.

26.(A) Medications associated with drug-induced lupus

Definite Association

Minocycline, procainamide, hydralazine, isoniazid, penicillamine, diltiazem, interferon-α, methyldopa, chlorpromazine, etanercept, infliximab, adalimumab.

Probable Association

Phenytoin, ethosuximide, carbamazepine, sulfasalazine, amiodarone, quinidine, rifampin, nitrofurantoin, β-blockers, lithium, captopril, interferon-γ, hydrochlorothiazide, glyburide, docetaxel, penicillin, tetracycline, statins, gold, valproate, griseofulvin, gemfibrozil, propylthiouracil.

27.(B)

28.(B) Cyclophosphamide, MMF, and azathioprine are appropriate for the treatment of lupus nephritis, whereas MMF and rituximab are often used for significant hematologic manifestations, including severe leukopenia, hemolytic anemia, or thrombocytopenia.

29.(C) Significant conduction system abnormalities after birth are treated with cardiac pacing and occasionally IVIG and corticosteroids, whereas severe cardiomyopathy may require cardiac transplantation. If the conduction defect is not addressed, affected children are at risk for exercise intolerance, arrhythmias, and death. With cardiac pacing, children with conduction system disease in the absence of cardiomyopathy have an excellent prognosis.

30.(B) Clinical features of juvenile dermatomyositis during disease course

FEATURE	%
✓ Muscle weakness	90-100
✓ Dysphagia or dysphonia	13-40
✓ Muscle atrophy	10
✓ Muscle pain and tenderness	30-75
✓ Skin lesions	85-100
✓ Heliotrope rash of eyelids	66-95
✓ Gottron papules	57-95
✓ Erythematous rash of malar/facial area	42-100
✓ Periungual (nail fold) capillary changes	80-90
✓ Photosensitive rash	5-42
✓ Ulcerations	22-30
✓ Calcinosis	12-30
✓ Lipodystrophy	11-14

- ✓ Raynaud phenomenon 2-15
- ✓ Arthritis and arthralgia 22-58
- ✓ Joint contractures 26-27
- ✓ Fever 16-65
- ✓ Gastrointestinal signs and symptoms 8-37
- ✓ Restrictive pulmonary disease 4-32
- ✓ Interstitial lung disease 1-7
- ✓ Cardiac involvement 0-3

31.(D) Elevated serum levels of muscle-derived enzymes (creatine kinase [CK], aldolase, aspartate transaminase, alanine transaminase [ALT], lactate dehydrogenase) reflect muscle inflammation. Not all enzyme levels rise with inflammation in a specific individual; ALT is usually elevated on initial presentation, whereas CK level may be normal. The erythrocyte sedimentation rate (ESR) is often normal, and the rheumatoid factor (RF) test result is typically negative.

32.(D) Lipodystrophy and calcinosis are thought to be associated with long-standing or undertreated disease. Dystrophic deposition of calcium phosphate, hydroxyapatite, or fluoroapatite crystals occurs in subcutaneous plaques or nodules, resulting in painful ulceration of the skin with extrusion of crystals or calcific liquid.

33.(D) Scleredema is a transient, self-limited disease of both children and adults that has sudden onset after a febrile illness (especially streptococcal infections).

34.(A) Linear Scleroderma

Linear lesions can extend through the dermis, subcutaneous tissue, and muscle to underlying bone; more likely unilateral.

35.(B) Raynaud phenomenon (RP) is the most frequent initial symptom in pediatric systemic sclerosis, present in 70% of affected children months to years before other manifestations. RP refers to the classic triphasic sequence of blanching, cyanosis, and erythema of the digits induced by cold exposure and/or emotional stress.

36.(B) In patients without major organ involvement, colchicine significantly improves oral and genital ulcers, skin features, and disease activity of Behçet disease.

37.(B) Proposed Criteria for Pediatric Sjögren Syndrome

Diagnosis requires ≥4 criteria

I. CLINICAL SYMPTOMS

1. Oral: recurrent parotitis or enlargement of parotid gland, dry mouth (xerostomia)

2. Ocular: dry eyes (xerophthalmia) recurrent conjunctivitis without obvious allergic or infectious etiology, keratoconjunctivitis sicca
3. Other mucosal: recurrent vaginitis
4. Systemic: fever, noninflammatory arthralgias, hypokalemic paralysis, and abdominal pain

II. IMMUNOLOGIC ABNORMALITIES

Presence of at least 1 of the following antibodies: anti-SSA, anti-SSB, high-titer antinuclear antibody, rheumatoid factor

III. OTHER ABNORMALITIES OR INVESTIGATIONS

1. Biochemical: elevated serum amylase
2. Hematologic: leukopenia, high erythrocyte sedimentation rate
3. Immunologic: polyclonal hyperimmunoglobulinemia
4. Renal: renal tubular acidosis
5. Histologic proof of lymphocytic infiltration of salivary glands or other organs (i.e., liver)
6. Objective documentation of ocular dryness (rose bengal staining or Schirmer test)
7. Positive findings of parotid gland scintigraphy

IV. Exclusion of all other autoimmune diseases

38.(A) Juvenile recurrent parotitis is characterized by intermittent unilateral parotid swelling typically lasting only a few days. It is frequently associated with fever and may undergo remission with puberty. Unlike in Sjögren syndrome, there is a male predominance, juvenile recurrent parotitis is seen in the younger children (3-6 yr), and there is a lack of focal lymphocytic infiltrates on biopsy.

39.(C) Prophylactic daily oral colchicine decreases the frequency, duration, and intensity of FMF flares. This regimen also prevents the development of systemic AA amyloidosis.

40.(A)

Diagnostic Indicators of Tumor Necrosis Factor Receptor–Associated Periodic Syndrome (TRAPS)

1. Recurrent episodes of inflammatory symptoms spanning >6 mo duration (several symptoms generally occur simultaneously)

a. Fever
b. Abdominal pain
c. Myalgia (migratory)
d. Rash (erythematous macular rash occurs with myalgia)
e. Conjunctivitis or periorbital edema
f. Chest pain

g. Arthralgia or monoarticular synovitis
2. Episodes last >5 days on average (although variable)
3. Responsive to glucocorticosteroids but not colchicine
4. Affects family members in autosomal dominant pattern (although may not always be present)
5. Any ethnicity may be affected

41.(B)

42.(D) Periodic Fever, Aphthous Stomatitis, Pharyngitis, and Adenitis (**PFAPA)** show dramatic response to a single oral dose of prednisone (0.6- 2.0 mg/kg), although this approach does not prevent recurrence and may actually shorten the interval between flares. Cimetidine at 20-40 mg/kg/day is effective at preventing recurrences in approximately one third of cases. Small series have shown that anakinra may be effective during a flare, but because corticosteroids are effective, this may not be a cost-effective approach. Colchicine may extend the time between flares. Complete resolution has been reported after tonsillectomy, although medical management should be the first approach.

43.(E) Although organ involvement may vary, AA amyloidosis most frequently affects the kidneys; 90% of patients have some degree of renal involvement. Unexplained proteinuria may be the presenting feature in some patients. Nephrotic syndrome and renal failure may develop if the underlying inflammatory condition is not controlled or if diagnosis is delayed.

44.(B) In adults and older children, pulmonary involvement is most frequent, with infiltration of the thoracic lymph nodes and lung parenchyma. Isolated bilateral hilar adenopathy on chest radiograph is the most common finding.

45.(B) There appear to be 2 distinct, age dependent patterns of disease among children with sarcoidosis. The clinical features in older children are similar to those in adults (pediatric-onset adult sarcoidosis), with frequent systemic features (fever, weight loss, and malaise), pulmonary involvement, and lymphadenopathy. In contrast, early-onset sarcoidosis manifesting in children <4 yr of age is characterized by the triad of rash, uveitis, and polyarthritis.

46.(C) Several risk stratification models have been constructed to determine which patients with Kawasaki disease (KD) are at highest risk for coronary artery abnormalities CAA. Predictors of poor outcome across several studies include young age, male gender, persistent fever, poor response to IVIG, and laboratory abnormalities, including neutrophilia, thrombocytopenia, transaminitis, hyponatremia, hypoalbuminemia, elevated levels of N-terminal–brain natriuretic protein and elevated C-reactive protein (CRP) levels. Asian and Pacific Islander race and Hispanic ethnicity are also risk factors for CAA.

47.(B) Clinical features that are not consistent with KD include exudative conjunctivitis, exudative pharyngitis, generalized lymphadenopathy, discrete oral lesions (ulceration or exudative pharyngitis), splenomegaly, and bullous, petechial, or vesicular rashes.

48.(C) Patients with acute KD should be treated with 2 g/kg of IVIG as a single infusion, usually administered over 10-12 hr within 10 days of disease onset, and ideally as soon as possible after diagnosis.

49.(B) Glucocorticoids such as oral prednisone (1-2 mg/kg/day), or in severe cases, intravenous (IV) methylprednisolone for 1-2 wk, followed by taper, reduce abdominal and joint pain but do not alter overall prognosis of Henoch Schönlein purpura.

50.(D) The younger age, the nature of the lesions, absence of other organ involvement, and a biopsy may help distinguish infantile acute hemorrhagic edema from Henoch Schönlein purpura.

51.(A)

52.(D) Oral prednisone (1-2 mg/kg/day) or IV pulse methylprednisolone (30 mg/kg/day) are the mainstay of therapy of polyarteritis nodosa. Oral or IV cyclophosphamide are often used as adjunctive therapy, and plasma exchange may be warranted for life-threatening disease.

53.(B)

54.(E) Churg-Strauss syndrome CSS frequently causes inflammation of the upper and lower respiratory tracts, but cartilage destruction is rare. CSS may initially demonstrate chronic or recurrent rhinitis/sinusitis, nasal polyposis, non-fixed pulmonary lesions, and difficult-to-treat asthma. Eosinophilia (>10% of leukocytes) with pulmonary infiltrates may precede a vasculitic phase.

55.(A) Growing pains, more appropriately termed benign nocturnal pains of childhood , growing pains affect 10–20% of children, with peak incidence between age 4 and 12 yr. Pain does not occur during periods of rapid growth or at growth sites. Pain often follows a day with exercise or other physical activities. Physical findings are normal, and gait is not impaired.

56.(D) Restless legs syndrome (RLS , Willis-Ekbom disease), seen more frequently among adolescents and adults, is a sensorimotor disturbance that may be confused with growing pains. Often familial, RLS is a difficult-to-control urge to move the leg that is exacerbated during rest and at night and is relieved by movement. Iron supplementation may benefit pediatric patients with RLS.

Chapter 16

Infectious Diseases Questions

USAMA A. AL JUMAILY

1. A 5-year-old boy with leukemia develops fever and chills; you suspect infection with gram-negative bacilli.
Of the following, the best lab media that support growth of such organisms is

A. sheep blood agar
B. chocolate agar
C. Sabouraud dextrose agar
D. MacConkey agar
E. Inhibitory mold agar

2. A 6-year-old boy develops fever and decrease level of consciousness; he has been diagnosed with posterior fossa low grade glioma since the age of 1 year that was treated successfully by VP shunt insertion and tumor resection. He is fully vaccinated. You suspect shunt infection.
Of the following, the **MOST** common organism isolated from shunt infection is

A. Haemophilus influenzae type b
B. Streptococcus pneumoniae
C. Propionibacterium
D. Mycobacterium tuberculosis
E. Cryptococcus

3. Parasitic infections are usually detected by microscopic examination of clinical specimens like blood, bone marrow, or fecal smears.
Of the following, the parasite that is detected by serologic tests rather than clinical specimens is

A. Plasmodium
B. Trichinella
C. Babesia
D. Leishmania
E. Giardia lamblia

4. A 7-year-old boy has recurrent bacterial respiratory infections; he has been diagnosed with cystic fibrosis since early childhood.
Of the following, the micro-organism that is strongly related with disease severity is

A. Pseudomonas aeruginosa
B. Haemophilus influenzae
C. Staphylococcus aureus
D. Mycoplasma pneumoniae
E. Mycobacterium tuberculosis

5. Which of the following is the **MOST** effective measure to reduce the likelihood of recurrent clostridium difficile-associated diarrhea (CDAD)?

A. Metronidazole
B. Fecal transplantation
C. Vancomycin
D. Avoid using antibiotics
E. Amoxicillin-clavulanate

6. A 5-year-old boy is going to receive immunoglobulin against hepatitis A before travelling with his parents to a country endemic with hepatitis A. The mother is concerned about adverse reactions of immunoglobulin administration.
Of the following, the **MOST** common adverse reaction to immunoglobulin administration is

A. flushing
B. headache
C. chills
D. pain at the injection site
E. nausea

7. A 4-year-old boy is exposed to his relative with varicella infection. He has been diagnosed with primary immune deficiency disorder since infancy. Varicella-zoster immunoglobulin is not available.
Of the following, the **BEST** alternative treatment is

A. intravenous immunoglobulin (IGIV)
B. intramuscular immunoglobulin (IGIM)
C. subcutaneous immunoglobulin (IGSC)
D. palivizumab

E. prophylactic acyclovir

8. Which of the following vaccines consists of whole inactivated microorganisms?

A. Acellular pertussis
B. Hepatitis A
C. Hepatitis B
D. Varicella
E. Rotavirus

9. A 7-year-old boy developed AIDS due to HIV infection. Lab test shows that his $CD4^+$ T-lymphocytes count is 250 cells/mm^3.
Of the following, the live attenuated vaccine that can be given to this child is

A. oral polio
B. small pox
C. yellow fever
D. BCG
E. varicella

10. Children with primary immunodeficiency may not receive vaccines, whether live-attenuated or killed ones. In which of the following diseases, all vaccines can be administered?

A. Chronic granulomatous disease
B. DiGeorge syndrome
C. Properdin deficiency
D. Severe combined immunodeficiency (SCID)
E. Common variable immunodeficiency

11. A 5-year-old girl is going to receive her scheduled BCG vaccine. She has been diagnosed with immune thrombocytopenia 3 weeks ago and has been treated with prednisolone 2mg/kg/day, since that time. What is your recommendation regarding vaccine administration?

A. She can receive her vaccine immediately
B. She should not receive vaccine any more
C. Stop corticosteroids for 2 weeks before resuming her vaccination
D. Stop corticosteroids for at least 1 month before resuming her vaccination

E. She can receive her vaccine immediately once corticosteroids are discontinued

12. A preterm baby is born to a HBsAg-positive mother. His birth weight is 1.9 kg. Your proper management for this baby is to

A. give birth dose of hepatitis B vaccine
B. defer hepatitis B vaccine until chronological age of 1 month
C. give hepatitis B immunoglobulin (HBIG) within 12 hr of birth
D. give hepatitis B vaccine within 12 hr of birth
E. give hepatitis B immunoglobulin (HBIG) and hepatitis B vaccine within 12 hr of birth

13. Which of the following vaccines should be deferred for at least 3 months after immunoglobulin administration?

A. Pertussis
B. Measles
C. Diphtheria
D. Rotavirus
E. Live attenuated influenza vaccine (LAIV)

14. Which of the following is the **MOST** common route of nosocomial infection during hospitalization?

A. Hands
B. Medical equipment
C. Toys
D. Hospital furnishings
E. Phones

15. A 5-year-old boy is admitted to ICU because of pending septic shock. He has been diagnosed with acute myeloblastic leukemia and received intensive chemotherapy with subsequent febrile neutropenia. He is on prolonged course of antibiotics since 3 weeks.
Of the following, the **MOST** common type of infection is

A. rota virus
B. respiratory viruses
C. fungi
D. staphylococci
E. gram-negative bacilli

16. Which of the following pathogens requires droplet isolation rather than contact isolation?

A. Enterohemorrhagic Escherichia coli O157:H7
B. Hepatitis A virus
C. Shigella species
D. Neisseria meningitidis
E. Clostridium difficile

17. A 6-year-old boy is going to receive hepatitis A vaccine before travelling to hepatitis A- endemic country. The parents ask about the time needed for the vaccine to provide adequate protection for safe travel.
Protective immunity after receiving hepatitis A vaccine will develop within

A. 1 week
B. 2 weeks
C. 4 weeks
D. 8 weeks
E. 12 weeks

18. Which of the following is the **MOST** common pathogen that may cause traveler`s diarrhea?

A. Enterotoxigenic Escherichia coli
B. Enteroaggregative Escherichia coli
C. Campylobacter
D. Entamoeba histolytica
E. Shigella

19. A 7-month-old infant develops diarrhea during a travel with his parents to a country in the Southeast Asia.
Of the following, the **MOST** appropriate antibiotic for this infant is

A. amoxicillin
B. trimethoprim-sulfamethoxazole
C. ciprofloxacin
D. azithromycin
E. erythromycin

20. Which of the following drugs is well known to cause fever?

A. Amoxicillin
B. Azithromycin

C. Amphotericin B
D. Cefixime
E. Teicoplanin

21. Which of the following disorders is characterized by periodic fever?
A. Cyclic neutropenia
B. Familial Mediterranean fever
C. TNF receptor–associated periodic syndrome
D. Hyper-IgD syndrome
E. Muckle-Wells syndrome

22. Which of the following conditions is **MOST** likely associated with relative tachycardia rather than bradycardia?
A. Brucellosis
B. Diphtheria
C. Chlamydia pneumonia
D. Legionnaire's disease
E. Leptospirosis

23. Which of the following conditions is classically associated with double quotidian fever (fever that peaks twice in 24 hr)?
A. Familial Mediterranean fever
B. Cyclic neutropenia
C. Behçet disease
D. Inflammatory arthritis
E. Crohn's disease

24. Which of the following CSF parameters is not affected by prior antibiotic administration in bacterial meningitis?
A. PCR
B. Culture
C. Cell
D. Glucose
E. Protein

25. A 3-year-old boy develops fever, drooling of the mouth, difficulty in swallowing, noisy breathing, and neck pain. Examination is unremarkable.
Of the following, the **MOST** appropriate diagnostic study is

A. lumbar puncture
B. MRI of the brain
C. MRI of the spinal cord
D. CT scan of the neck
E. lateral X-ray of the head and neck

26. An 18-month-old girl develops prolonged fever of 3 weeks duration. It has been preceded by a flue like illness. The mother describes that the fever has been subsided after 5 days followed by recurrent spikes every 2-4 days. Other medical history is insignificant. Examination is unremarkable.
Of the following, the **MOST** likely diagnosis is
A. nephrogenic diabetes insipidus
B. familial dysautonomia
C. pseudo-FUO
D. familial Mediterranean fever (FMF)
E. hyper IgD syndrome

27. Which of the following conditions is accompanied by fever in the absence of sweating?
A. Hodgkin lymphoma
B. Tuberculosis
C. Hyperthyroidism
D. Hypoglycemia
E. Familial dysautonomia

28. Which of the following is the **MOST** likely diagnosis in a 1-year-old male child with prolonged fever associated with bulbar conjunctivitis?
A. Measles
B. Lymphogranuloma venereum
C. Kawasaki disease
D. Coxsackievirus infection
E. Tuberculosis

29. Fever, lack of tears, absent corneal reflex, and smooth tongue with absence of fungiform papillae is highly suggestive of which of the following diseases?
A. Familial dysautonomia
B. Ectodermal dysplasia
C. Kawasaki disease

D. Dermatomyositis
E. Scleroderma

30. A 3-year-old boy with chronic granulomatous disease (CGD) develops fever, chills, toxicity, and tachycardia consistent with sepsis.
Of the following, the **MOST** common micro-organism causing sepsis in this child is

A. Staphylococcus aureus
B. Aspergillus
C. Chromobacterium violaceum
D. Streptococcus pneumoniae
E. Candida albicans

31. In which of the following primary immunodeficiency diseases, Pneumocystis jiroveci pneumonia (PCP) infection is more likely to occur?

A. X-linked agammaglobulinemia (XLA)
B. Common variable immunodeficiency
C. Selective IgA deficiency
D. IgG 2 subclass deficiency
E. Hyper IgM syndrome

32. A 12-year-old boy develops fever and severe oral mucositis; he has been diagnosed with acute lymphoblastic leukemia 3 months ago; he has received intensive chemotherapy 1 week before and kept on ciprofloxacin prophylaxis. His absolute neutrophil count (ANC) is 100/mm^3.
Of the following, the **MOST** appropriate antibiotic that should be given to this boy is

A. cefepime
B. vancomycin
C. piperacillin
D. ceftriaxone
E. amphotericin

33. A 12-year-old boy with AML develops fever and oral mucositis 1 week after high dose cytarabine chemotherapy administration. He has been kept on ciprofloxacin prophylaxis after completion of chemotherapy. His ANC is 300/mm^3.
Of the following, the **MOST** commonly encountered pathogen in this boy is

A. Candida albicans
B. Mucor mycosis
C. Aspergillus fumigatus
D. Staphylococcus aureus
E. Streptococcus viridans

34. Which of the following is the **MOST** commonly used intravenous antibiotic as 1st line monotherapy in children with fever and neutropenia?
A. Ceftazidime
B. Cefepime
C. Vancomycin
D. Meropenem
E. Gentamicin

35. A 9-year-old boy develops lymphoma few years after liver transplantation. Which of the following viral infections that may lead to such complication?
A. Cytomegalovirus (CMV)
B. Varicella-zoster virus (VZV)
C. Hepatitis B virus (HBV)
D. Hepatitis C virus (HCV)
E. Epstein-Barr virus (EBV)

36. A 5-year-old boy has greenish discharge at the exit site of central venous line (CVC).
Of the following, the **MOST** likely offending causative micro-organism is
A. Staphylococcus aureus
B. Coagulase-negative staphylococci
C. Streptococcus pneumoniae
D. Candida albicans
E. Mycobacteria

37. A 7-year-old boy has persistent candidemia after removal of central line venous catheter. He is on daily liposomal amphotericin B for the last 2 weeks.
Of the following, the **MOST** appropriate management in this setting is to
A. stop antifungal treatment
B. add itraconazole
C. add fluconazole
D. continue amphotericin for 6 weeks

E. replace amphotericin by fluconazole

38. A 3-year-old boy develops fever. Examination reveals ill child with neck stiffness. Vaccination history is negative upon parental refusal.
Of the following, the **MOST** common causative organism is

A. Haemophilus influenzae type b
B. Escherichia coli
C. Streptococcus pneumoniae
D. Neisseria meningitidis
E. Staphylococcus aureus

39. A 5-year-old boy develops fever. Blood culture reveals Streptococcus pneumoniae pathogens.
Of the following, the **MOST** appropriate antibiotic effective against this pathogen is

A. ampicillin
B. ampicillin-sulbactam
C. amoxicillin-clavulanate
D. ticarcillin-clavulanic acid
E. vancomycin

40. Which of the following cephalosporins is highly effective against pseudomonas aeruginosa?

A. Cefazolin
B. Cefuroxime
C. Ceftriaxone
D. Cefotaxime
E. Ceftazidime

41. A 3-year-old girl develops erythema, swelling and tenderness of the right hand. Swab culture reveals methicillin-resistant staphylococcus aureus (MRSA).
Of the following, the cephalosporin that is **MOST** effective against this pathogen is

A. cefepime
B. ceftazidime
C. ceftaroline
D. cefazolin
E. cefuroxime

42. Which of the following pathogens is resistant to meropenem?

A. Streptococcus pneumoniae
B. Pseudomonas aeruginosa
C. Stenotrophomonas maltophilia
D. Klebsiella
E. Acinetobacter

43. Which of the following aminoglycosides is particularly effective against Francisella tularensis infection?

A. Streptomycin
B. Kanamycin
C. Gentamicin
D. Tobramycin
E. Netilmicin

44. Which of the following is the **MOST** effective antibiotic against extended spectrum beta lactamase (ESBL) organisms?

A. Meropenem
B. Ceftazidime
C. Amikacin
D. Colistin
E. Ertapenem

45. Staphylococcus aureus pathogens can affect the host by direct invasion or through toxin elaboration. Which of the following diseases is caused by direct invasion of the staphylococcus aureus rather than by toxin-mediated process?

A. Food poisoning
B. Pericarditis
C. Scarlet fever
D. Scalded skin syndrome
E. Toxic shock syndrome

46. Skin lesions caused by S. aureus may be indistinguishable from those caused by group A streptococci.
Which of the following manifestations is **MORE** specific for the former?

A. slow expansion
B. less erythema

C. more tenderness
D. more swelling
E. less fluctuation

47. Which of the following organisms may mimics S. aureus infection by causing cavitary pneumonia?

A. Group A streptococcus
B. Mycoplasma pneumoniae
C. Streptococcus pneumoniae
D. Klebsiella pneumoniae
E. Pseudomonas aeruginosa

48. A 2-year-old boy develops gluteal abscess. After draining of the abscess, the culture reveals methicillin susceptible S. aureus (MSSA).
Of the following, the **MOST** appropriate initial antibiotic treatment is

A. ampicillin
B. cefazolin
C. amoxicillin
D. azithromycin
E. ciprofloxacin

49. Toxic shock syndrome (TSS) closely resembles Kawasaki disease clinically. However, some clinical manifestations are more common in TSS.
Of the following, the clinical manifestation that is **MORE** specific for TSS is

A. fever
B. hypotension
C. erythematous rash
D. conjunctival hyperemia
E. skin desquamation

50. Which of the following strains of coagulase-negative staphylococci (CoNS) is commonly causing urinary tract infections (UTI)?

A. Staphylococcus epidermidis
B. Staphylococcus hominis
C. Staphylococcus haemolyticus
D. Staphylococcus saprophyticus
E. Staphylococcus lugdunensis

51. Most of coagulase-negative staphylococci (CoNS) are resistant to methicillin. Which of the following strains is methicillin susceptible?

A. Staphylococcus epidermidis
B. Staphylococcus hominis
C. Staphylococcus haemolyticus
D. Staphylococcus saprophyticus
E. Staphylococcus lugdunensis

52. Treatment of S. epidermidis infections often requires effective antibiotics with removal of indwelling medical devices.
Of the following, the indwelling medical device infection that is treated with antibiotics without its removal is

A. central venous line catheter
B. prosthetic heart valve
C. peritoneal dialysis catheter
D. CSF shunt
E. prosthetic bone

53. A 2-year-old girl develops fever, cough, and shortness of breath secondary to right middle lobe empyema. She is not vaccinated upon parental refusal. You suspect invasive pneumococcal infection.
Of the following, the **MOST** appropriate initial treatment is

A. amoxicillin
B. ampicillin
C. penicillin G
D. ceftriaxone
E. vancomycin

54. A 5-year-old boy develops otitis media due to pneumococcal infection. He has a history of non-type I allergic reaction to penicillin.
Of the following, the **BEST** alternative treatment is

A. cephalexin
B. clindamycin
C. levofloxacin
D. trimethoprim-sulphamethaxazole
E. azithromycin

55. A 4-year-old boy develop bacterial meningitis due to invasive pneumococcal infection.
Of the following, the **MOST** commonly encountered adverse neurological sequel is

A. hearing loss
B. epilepsy
C. intellectual deficits
D. paralysis
E. blindness

56. Which of the following is the **MOST** common infection caused by Streptococcus pyogenes?

A. Pharyngitis
B. Perianal cellulitis
C. Pneumonia
D. Osteomyelitis
E. Suppurative arthritis

57. Erysipelas is an acute infection involving the deeper layers of the skin and the underlying connective tissue.
What is the **MOST** characteristic finding of this infection?

A. Cutaneous swelling
B. Skin erythema
C. Very tender affected skin
D. Sharply defined, slightly elevated border
E. Reddish streaks of lymphangitis projecting out from the margins of the lesion

58. A 5-year-old boy develops nonbullous impetigo involving the face and extremities. The skin lesions have been resolved after antibiotics administration.
Of the following, the **MOST** reliable test that confirms evidence of preceding group A streptococcal infection in this child is

A. anti–streptolysin O
B. anti–DNase B
C. streptozyme test
D. Streptococcal rapid antigen detection
Isothermal loop amplification

59. A 5-year-old boy develops oral pharyngitis secondary to group A streptococcal (GAS) infection. You recommend starting therapy with oral penicillin. To achieve maximal pharyngeal eradication of GAS and prevention of rheumatic fever, the oral penicillin must be administered for a maximal duration of

A. five days
B. seven days
C. ten days
D. fourteen days
E. twenty-one days

60. A 4-year-old girl develops oral pharyngitis due to group A streptococcal (GAS) infection; she has had a history of immediate (anaphylactic-type) hypersensitivity to penicillin.
Of the following, The **MOST** appropriate treatment for this girl is oral

A. cephalosporins
B. clindamycin
C. sulphonamides
D. tetracyclines
E. penicillin

61. A 5-year-old boy develops acute arthritis following an episode of group A streptococcal (GAS) pharyngitis. The characteristic distinct feature that differentiate post streptococcal reactive arthritis (PSRA) from arthritis due to rheumatic fever (RF) is

A. involvement of large joints
B. nonmigratory arthritis
C. long latent period between the antecedent episode of GAS pharyngitis and arthritis
D. dramatic response to aspirin therapy
E. concurrent high grade fever

62. Which of the following Jones criteria is required as the only sole criterion to diagnose acute rheumatic fever (RF)?

A. Acute carditis
B. Polyarthritis
C. Erythema marginatum
D. Subcutaneous nodules

E. Chorea

63. A 5-year-old boy develops acute rheumatic fever manifested by fever and polyarthritis.
Of the following, the **LEAST** commonly involved joint is

A. hip
B. knee
C. ankle
D. elbow
E. wrist

64. Characteristic feature of arthritis due to acute rheumatic fever that distinguishes it from arthritis due to other causes is

A. hotness
B. non tender
C. dramatic response to small dose of salicylates
D. polyarticular
E. deforming

65. Carditis, whether clinical or subclinical, is a major criterion for diagnosis of acute rheumatic fever. The **MOST** commonly involved cardiac valve is

A. mitral
B. aortic
C. pulmonary
D. tricuspid
E. both aortic and pulmonary

66. A 15-year-old male adolescent has been diagnosed with acute rheumatic fever at the age of 5 year. His initial manifestations were clinical carditis and migratory arthritis.
Of the following, the **MOST** common echocardiographic finding at this age is

A. mitral regurgitation
B. mitral stenosis
C. tricuspid regurgitation
D. tricuspid stenosis
E. pulmonary stenosis

67. A 5-year-old boy develops acute rheumatic fever. The parents are concerned about future development of valvular heart disease.
Of the following, the feature that is **highly** correlated with significant rheumatic heart disease is

A. number of initial joints involvement
B. fever more than 38 C°
C. chorea
D. erythema marginatum
E. subcutaneous nodules

68. A 6-year-old girl develops acute rheumatic fever manifested by fever, arthritis and carditis. Chest radiography shows cardiomegaly with pulmonary edema. She is on oral amoxicillin treatment.
Of the following, the **MOST** appropriate initial treatment for this girl is

A. prophylactic benzathine penicillin
B. oral salicylate
C. oral acetaminophen
D. oral prednisolone
E. NSAIDs

69. You are discussing acute rheumatic fever with medical students. Risk of recurrent carditis is raised during the discussion.
Of the following, the statement that should be included in the discussion is

A. the more severe the initial cardiac involvement, the lesser the risk for residual heart disease
B. patients without carditis during the initial episode are less likely to have carditis with recurrent attack
C. there is a stepwise decrease in cardiac involvement as the number of episodes increases
D. patients with initial episode of carditis never have carditis with recurrences
E. the risk for permanent heart damage decreases with each recurrence.

70. A 2-day-old neonate develops recurrent attacks of apnea and respiratory distress; he is lethargic, with bradycardia, and poor feeding. He is a full term baby, a product of normal vaginal delivery. The mother has a history of prolonged rupture of membranes 24 hours before delivery.
Of the following, the **MOST** likely causative pathogen is

A. Staphylococcus aureus
B. group A streptococci
C. Streptococcus pneumoniae
D. group B streptococcus
E. Haemophilus influenzae

71. Invasive group B streptococci (GBS) disease in children beyond early infancy is uncommon. The MOST common infection associated with childhood GBS is
A. bacteremia
B. meningitis
C. pneumonia
D. endocarditis
E. arthritis

72. A 10-day-old neonate develops apnea, bradycardia, and respiratory distress. He is a ex- preterm baby by normal vaginal delivery at a gestational age of 28. He has been admitted to NCU and an intravascular catheter has been inserted.
Of the following, the **MOST** likely causative pathogen is
A. group A Streptococcus
B. group B Streptococcus
C. Staphylococcus aureus
D. Enterococcus
E. Listeria monocytogenes

73. A 3-year-old boy develops sepsis. Blood culture revealed Enterococcus with susceptibility to penicillin. The child is allergic to penicillin.
Of the following, the **MOST** appropriate treatment for this boy is
A. cephalexin
B. azithromycin and gentamicin
C. vancomycin
D. cephalexin and gentamicin
E. vancomycin and gentamicin

74. A 6-year-old child develops fever, toxicity, tachycardia, and borderline blood pressure. He has been diagnosed with acute myeloid leukemia 3 months ago and received intensive chemotherapy. Blood culture revealed vancomycin-resistant Enterococcus faecalis.
Of the following, the **MOST** appropriate initial treatment is

A. linezolid
B. daptomycin
C. quinupristin-dalfopristin
D. tigecycline
E. cephalexin

75. The major virulence of the Corynebacterium diphtheria organism lies in its ability to produce a potent exotoxin. The early local manifestation of this toxin is

A. paralysis of the palate and hypopharynx
B. kidney tubule necrosis
C. thrombocytopenia
D. cardiomyopathy
E. demyelination of nerves

76. Pharyngeal diphtheria may be misdiagnosed with exudative pharyngitis caused by streptococcus pyogenes or Epstein-Barr virus.
Which of the following features is characteristic of pharyngeal diphtheria rather than exudative pharyngitis?

A. Easily detached non adherent membrane
B. Extension of the membrane beyond the faucial area
C. Absence of dysphagia
D. Presence of high grade fever
E. Hoarseness of voice

77. What is the **MOST** common cardiac problem associated with respiratory diphtheria?

A. Myocarditis
B. Dilated cardiomyopathy
C. Hypertrophic cardiomyopathy
D. Cardiac dysrhythmias
E. Heart failure

78. What are the **MOST** frequent ECG changes detected in children with cardiac toxicity due to respiratory diphtheria?

A. first degree heart block
B. second degree heart block
C. third degree heart block

D. sinus tachycardia
E. ventricular tachycardia

79. A 3-year-old boy develops tonsillar diphtheria with extensive membrane formation. The mainstay of management that should be started as early as possible is

A. crystalline penicillin
B. erythromycin
C. equine antitoxin
D. droplet isolation
E. contact isolation

80. A 1-day-old preterm neonate develops lethargy and poor feeding consistent with sepsis associated with diffuse pustular rash. The mother has a history of flu-like illness during the 2nd trimester.
What is **MOST** likely recovered organism by blood culture?

A. Staphylococcus aureus
B. Escherichia coli
C. Group B streptococcus
D. Listeria monocytogenes
E. Chlamydia trachomatis

81. Listeriosis is indistinguishable clinically from neonatal sepsis and meningitis caused by other organisms.
Of the following, the lab finding that suggest Listeria infection is

A. increased peripheral blood monocytes count
B. decreased peripheral blood lymphocytes count
C. increased peripheral blood neutrophils count
D. increased platelets count
E. increased C-reactive protein

82. Listeria monocytogenes CNS infection is usual beyond the neonatal period.
Of the following, the CNS site that is characteristically affected is

A. cerebrum
B. cerebellum
C. brainstem
D. hypothalamus
E. spinal cord

83. Which of the following antibiotic is ineffective against Listeria?

A. Ampicillin
B. Cefotaxime
C. Vancomycin
D. Trimethoprim-sulfamethoxazole
E. Erythromycin

84. A 10-year-old boy develops right submandibular tender mass 3 weeks after dental procedure, associated with pain, fever, and draining fistula with yellow sulfur granules. He has been developed type I diabetes mellitus since the age of 5 year that is poorly controlled.
Of the following, the **MOST** likely causative pathogen is

A. Staphylococcus aureus
B. Streptococcus pneumoniae
C. Actinomyces
D. Nocardia
E. Candida albicans

85. A 6-year-old boy develops fever, cough, and dyspnea, followed by headache, altered mental status, and speech impairment few days later. He has been diagnosed with acute lymphoblastic leukemia 1 year ago and has been on chemotherapy since that time. CT scan of the chest reveals multiple pulmonary cavitary lesions. MRI of the brain shows multiple-ring enhancing lesions suggestive of multiple abscesses.
Of the following, the **MOST** likely causative organism is

A. Staphylococcus aureus
B. Aspergillus fumigatus
C. Mycoplasma pneumonia
D. Actinomyces
E. Nocardia

86. Which of the following is the **MOST** common form of meningococcal infection?

A. Nasopharyngeal asymptomatic carrier
B. Meningitis
C. Septicemia
D. Pneumonia
E. Chronic meningococcemia

87. Which of the following is the **MOST** common complication of meningococcal septicemia?

A. Adrenal hemorrhage
B. Endocarditis
C. Endophthalmitis
D. Focal skin infarction
E. Arthritis

88. Which of the following factors indicates the **POORER** prognosis for invasive meningococcal disease?

A. Hypothermia
B. Hypotension
C. Purpura fulminans
D. Seizures
E. Absence of meningitis

89. You are hospitalizing a 5-year-old boy who develops meningococcal septicemia; the guardian in close contact with him is his mother who is now pregnant at 1st trimester.
Of the following, the drug of choice for prophylaxis of the mother is

A. oral rifampin
B. oral ciprofloxacin
C. intramuscular ceftriaxone
D. intramuscular ampicillin
E. oral azithromycin

90. Which of the following is the **MOST** common type of gonococcal infection in prepubertal girls?

A. Vulvovaginitis
B. Salpingitis
C. Endometritis
D. Perihepatitis
E. Peritonitis

91. Hematogenous disseminated gonococcal infection may begin 7-30 days after primary infection.
Of the following, the **MOST** common initial manifestation is

A. pustular acral skin lesions

B. polyarthralgia with fever
C. tenosynovitis
D. suppurative arthritis
E. osteomyelitis

92. A 5-day-old neonate develops ophthalmia neonatorum due to gonococcal infection acquired from the mother. He is an ex preterm neonate due to premature rupture of membrane, with hyperbilirubinemia.
Of the following, the **MOST** appropriate initial treatment is

A. cefotaxime
B. ceftriaxone
C. cefexime
D. azithromycin
E. tetracycline

93. A 6-year-old boy develops septic arthritis of the left knee joint. He is a case of chronic renal failure and undergoes frequent hemodialysis. Gram stain of the synovial fluid aspirate reveals pairs of gram-negative coccobacilli with tapered ends.
Of the following, the **MOST** likely infected organism is

A. Neisseria gonorrhoea
B. Neisseria meningitides
C. Haemophilus influenzae
D. Kingella kingae
E. Moraxella catarrhalis

94. Although septic arthritis is the most common clinical presentation of Kingella kingae infection, other manifestations may occur with a variable relative frequency.
Which of the following is an exceptional clinical manifestation?

A. Osteomyelitis
B. Bacteremia
C. Endocarditis
D. Spondylodiscitis
E. Pneumonia

95. A 3-year-old girl develops osteomyelitis of left clavicle. Of the following, the pathogen that is frequently infect such site is

A. Staphylococcus aureus
B. Kingella kingae
C. Salmonella typhi
D. Haemophilus influenzae
E. Neisseria gonorrhoea

96. A 5-year-old boy develops right femur osteomyelitis. Lab tests confirm Kingella kingae bacterium. Which of the following antibiotics is ineffective against this pathogen?

A. Vancomycin
B. Ampicillin
C. Ceftriaxone
D. Cefuroxime
E. Oxacillin

97. A 4-year-old girl develops meningitis caused by Hemophilus influenzae type B. Her mother has a concern regarding hearing impairment sequelae.
Of the following, the drug of choice that decreases the incidence of this complication is

A. phenobarbital
B. phenytoin
C. paracetamol
D. dexamethasone
E. rifampicin

98. A 2-year-old boy develops preseptal orbital cellulitis. The characteristic feature that distinguishes this type of infection caused by H. influenzae from that caused by S. aureus and group A streptococcus is

A. fever
B. lid edema
C. lid tenderness
D. warmth of the lid
E. purple discoloration

99. Which of the following infections is mainly caused by nontypeable strains of H. influenzae rather than by H. influenzae type b?

A. Meningitis
B. Pericarditis

C. Suppurative arthritis
D. Acute epiglottitis
E. Otitis media

100. The distinction between preseptal and orbital cellulitis may be difficult clinically. The feature that is more in favor of orbital cellulitis is
A. fever
B. lid edema
C. tenderness
D. warmth of lid
E. limitation of the extraocular movements

101. Which of the following is **MORE** common in neonates with pertussis?
A. High grade fever
B. Prominent cough early in the disease
C. Frequent whooping
D. Apnea
E. Rhinorrhea and sneezing

102. Which of the following is a characteristic feature of pertussis in adolescents?
A. Fever
B. Posttussive vomiting
C. Sore throat and hoarseness
D. Malaise or myalgia
E. Wheezing and rales with tachypnea

103. A 5-year-old boy develops rhinorrhea, sneezing, lacrimation, and conjunctival suffusion. Blood count reveals WBC count of 18,000 cells/µl. You suspect pertussis. Of the following, the blood smear finding that support your diagnosis is
A. absolute neutrophilia
B. high number of band cells
C. absolute lymphocytosis with small normal cells
D. absolute lymphocytosis with large atypical cells
E. absolute eosinophilia

104. Which of the following lab findings predict poor outcome in pertussis?

A. Severe leucopenia
B. Polycythemia
C. Anemia
D. Extreme thrombocytosis
E. Severe thrombocytopenia

105. A 5-year-old boy develops pertussis. He has a history of frequent fainting attacks 2 years ago; the diagnosis of long QT syndrome has been established since then. He lives with her mother in a single house.
Of the following, the **MOST** appropriate antimicrobial regimen is

A. azithromycin for the child only
B. azithromycin for the child and mother
C. trimethoprim-sulphamethaxazole (TMP-SMX) for the child and azithromycin for the mother
D. azithromycin for the child and clarithromycin for the mother
E. TMP-SMX for the child and mother

106. Which of the following is the usual complication that may lead to death in pertussis?

A. Pneumothorax
B. Epistaxis
C. CNS hemorrhage
D. Pulmonary hypertension
E. Bronchiectasis

107. A 5-year-old healthy boy develops nausea, vomiting, and periumbilical crampy abdominal pain 1-day after ingestion poultry products, followed by severe watery diarrhea. Stool culture revealed Salmonella Typhimurium.
Of the following, the **MOST** appropriate management is

A. correction of dehydration and electrolyte disturbances only
B. start oral tetracycline for 7 days
C. start oral cefexime for 10 days
D. start intravenous cefotaxime for 14 days
E. anti emetics and anti diarrheal medications

108. Which of the following subsets of children are at increased risk for salmonella osteomyelitis?

A. Children with HIV infection

B. Children with leukemia
C. Children with sickle cell disease
D. Children with schistosomaisis
E. Children with chronic kidney disease

109. A 5-year-old boy develops high grade fever, myalgia, abdominal pain, and hepatosplenomegaly. He is on oral antibiotics for 1 week requested by a physician. You suspect typhoid fever.
Of the following, the lab test that increases the likelihood of bacteriologic confirmation in this boy is

A. blood culture
B. urine culture
C. stool culture
D. bone marrow culture
E. Widal test

110. Complications pertinent to Salmonella typhi infection may differ in frequency in children comparing to adults.
Which of the following complications is more common in infants?

A. Intestinal hemorrhage
B. Intestinal perforation
C. Acute cerebellar ataxia
D. Deafness
E. Disseminated intravascular coagulopathy

111. Symptoms due to shigellosis and enterohemorrhagic E. coli (EHEC) infection may be similar.
Of the following, the typical symptom with shigellosis that distinguishes it from EHEC is

A. high fever
B. bloody diarrhea
C. abdominal pain
D. vomiting
E. anorexia

112. Which of the following is the **MOST** common complication of shigellosis?

A. Dehydration
B. Seizures

C. Meningitis
D. Sepsis
E. Encephalopathy

113. A 3-year-old boy develops high fever, abdominal cramps, anorexia, emesis, and watery diarrhea of 1 day duration. There is an associated urgency and painful defecation. You suspect bacillary dysentery. Waiting the stool culture result, the lab test that support your diagnosis is

A. absence of stool leukocytes
B. absence of fecal blood
C. marked anemia
D. marked leucopenia
E. leukocytosis with presence of bands cells

114. A 9-year-old girl develops high fever, bloody diarrhea, and colicky abdominal pain. You suspect shigellosis. Of the following, the empirical drug of choice is

A. ampicillin
B. ciprofloxacin
C. TMP-SMX
D. cephalexin
E. cefuroxime

115. A 2-year-old boy develops acute renal shutdown, thrombocytopenia, and microangiopathic hemolytic anemia after a period of acute diarrheal illness.
Of the following, the infectious pathogen that leads to this complication is

A. enterotoxigenic E. coli (ETEC)
B. enteroinvasive E. coli (EIEC)
C. enteropathogenic E. coli (EPEC)
D. Shiga toxin–producing E. coli (STEC)
E. enteroaggregative E. coli (EAEC)

116. A 2-year-old boy develops abdominal pain and watery diarrhea that becomes bloody few days later; there is no accompanying fever. Microbiology test confirms identification of Shiga-toxin E. coli STEC (O157:H7 strains).
Of the following, the **MOST** appropriate step is to

A. give ampicillin
B. start with trimethoprim-sulphamethoxazole TMP-SMX

C. give ciprofloxacin
D. start with oral azithromycin
E. avoid antimicrobial treatment

117. A 2-year-old boy develops sudden onset of profuse watery diarrhea associated with vomiting. There is no crying when passing stool. His mother describes the stool as "rice-water" with a fishy smell, but no blood or mucus.
Of the following, the **MOST** likely causative pathogen is

A. rota virus
B. Vibrio cholerae
C. enterotoxigenic E. coli
D. enteropathogenic E. coli
E. Salmonella enteritidis

118. Campylobacter infections can be both food-borne and water-borne. The main source of this bacterial pathogen is

A. chicken
B. fish
C. oysters
D. red meat
E. swine

119. Which of the following species of campylobacter is **MOST** likely causing endocarditis rather than gastroenteritis?

A. coli
B. fetus
C. rectus
D. C. hypointestinalis
E. upsaliensis

120. A 4-year-old girl develops acute lower limbs flaccid paralysis few days after acute diarrheal illness characterized by fever, abdominal cramps, and watery diarrhea.
Of the following, the **MOST** likely causative organism is

A. Vibrio cholerae
B. Enterotoxigenic E. coli
C. Salmonella enteritidis
D. Campylobacter jejuni

E. Shigella flexneri

121. A 2-year-old boy develops watery diarrhea associated with fever and abdominal pain. He is a case of thalassemia major diagnosed since the age of 6 months, on regular blood transfusion regimen since that time. He has a history of recent blood transfusion. Examination of oral cavity reveals prominent pharyngitis.
Of the following, the **MOST** likely offending micro-organism is

A. Campylobacter jejuni
B. Salmonella typhimurium
C. Yersinia enterocolitica
D. Shigella dysenteriae
E. enterotoxigenic E. coli

122. Adolescents may be infected by Y. enterocolitica; the **MOST** common presentation in such age is

A. enteritis
B. mesenteric lymphadenitis
C. septic arthritis
D. pneumonia
E. meningitis

123. Which of the following drugs is recommended as empirical treatment for children with Yersinia enterocolitis?

A. Trimethoprim-sulfamethoxazole (TMP-SMX)
B. Ampicillin
C. Cephalexin
D. Amoxicillin-clavulanate
E. Cefadroxil

124. A 7-year-old boy develops inguinal lymphadenitis, with remarkable tenderness, fever, chills, and weakness. The overlying skin shows insect bites. As he is living in area endemic with plague, you suspect infection by Y. pestis.
Waiting bacteriologic confirmation by culture of lymph nodes aspirate and blood, the **MOST** appropriate initial management is

A. droplet isolation and streptomycin treatment of the index case, and doxycycline prophylaxis of close contacts
B. droplet isolation and streptomycin treatment of the index case only

C. droplet isolation of the index case only
D. streptomycin treatment of the index case, and doxycycline prophylaxis of close contacts
E. close observation waiting results of bacteriological tests

125. Pseudomonas aeruginosa infections are opportunistic infections that may affect various organs including skin. The characteristic skin lesions of that pathogen is

A. erythema nodosum
B. ecthyma gangrenosum
C. erythema multiforme
D. cellulitis
E. folliculitis

126. P. aeruginosa is an occasional cause of nosocomial bacteremia in newborns. The focus that is frequently preceding bacteremia is

A. ecthyma-like skin lesions
B. perianal abscess
C. nasal discharge
D. conjunctivitis
E. omphalitis

127. A 5-year-old boy develops fever and rigor of 2 days duration. He has been diagnosed with acute leukemia 3 months ago; he is on intensive chemotherapy since that time administered by implanted intravenous catheter. Examination revealed borderline blood pressure, and vasculitic skin lesions. Lab tests revealed: WBC, 500/mm^3; absolute neutrophils count, 75/mm^3.
Of the following, blood culture **MOST** likely reveals

A. S. aureus
B. S. epidermidis
C. coli
D. Klebsiella pneumoniae
E. P. aeruginosa

128. A 6-year-old boy has been diagnosed with cystic fibrosis since early childhood. He develops chronic lung disease and P. aeruginosa infection with frequent pulmonary exacerbations.
Of the following, the drug of choice to decrease pulmonary exacerbations is

A. ceftazidime
B. cefepime
C. piperacillin-tazobactam
D. combined ceftazidime and aminoglycoside
E. azithromycin

129. Brucellosis is a systemic illness that can be very difficult to diagnose in children. Which of the following features occur less frequently in children?
A. Fever
B. Sweating
C. Arthralgia
D. Myalgia
E. Back pain

130. A 6-year-old boy develops fever, arthralgia, myalgia, and back pain of 2 weeks duration. He is living in a village. He has a history of consuming raw dairy products. Examination shows hepatic and splenic enlargement.
Of the following, the **MOST** widely used test to confirm the diagnosis is
A. blood culture
B. bone marrow culture
C. enzyme immunoassay
D. serum agglutination test
E. polymerase chain reaction

131. A 5-year-old girl develops fever, arthralgia, back pain, and hepatosplenomegaly. She has a history of consuming raw milk 3 weeks ago. Serum agglutination test for brucellosis is strongly positive.
Of the following, the **MOST** appropriate treatment is
A. doxycline
B. doxycline and rifampin
C. doxycycline and gentamicin
D. trimethoprim-sulfamethoxazole (TMP-SMX)
E. TMP-SMX and rifampin

132. Duration of treatment for brucellosis is variable according to disease severity and complications. In which of the following complications due to brucella infection, the longest duration of treatment is recommended?
A. Meningitis

B. Endocarditis
C. Arthritis
D. Osteomyelitis
E. Spondylitis

133. Many risk factors may predispose to legionnaire's disease (Legionella pneumonia). The factor that poses the highest risk for infection is
A. asthma
B. bronchitis
C. chronic corticosteroid therapy
D. diabetes
E. renal failure

134. A 3-year-old boy develops malaise, anorexia, and right axillary lymphadenopathy. The lymph nodes are tender with overlying erythema. There are multiple red papules at the base of right middle finger. He has a history of contact with kitten 3 weeks ago.
Of the following, the **MOST** likely infectious pathogen is
A. group B streptococcus
B. Bartonella henselae
C. Toxoplasma gondii
D. Mycobacterium avium
E. Epstein-Barr virus

135. What is the **MOST** common presentation of cat scratch disease caused by Bartonella henselae?
A. Fever
B. Chronic regional lymphadenitis
C. Red papules
D. Headache and malaise
E. Erythema annulare

136. Chronic regional lymphadenitis is the hallmark of cat-scratch disease, affecting the 1st or 2nd set of nodes draining the entry site.
Of the following, the **MOST** commonly affected lymph nodes
A. femoral
B. inguinal
C. epitrochlear

D. axillary
E. submandibular

137. A 3-month-old infant develops poor feeding, weak sucking, feeble cry, and mouth drooling. He is an exclusively breast fed, although he has been given few tea spoons of honey early at the age of 2 months as a racial tradition to increase his body weight. Examination reveals expressionless face, bilateral ptosis, and hypotonia. His vaccination is integral up to his age.
Of the following, the **MOST** likely diagnosis is
A. sepsis
B. myasthenia gravis
C. Guillain-Barré syndrome
D. botulism
E. poliomyelitis

138. Which of the following is a recognized feature of botulism?
A. Asymmetric flaccidity
B. Ascending paralysis
C. Clear sensorium
D. Fever
E. Paresthesia

139. The diagnosis of botulism is unequivocally established by demonstration of toxin in serum or organisms. Which of the following ancillary investigations is helpful in the diagnosis?
A. Electromyography (EMG)
B. Motor nerve conduction study
C. Sensory nerve function
D. Muscle biopsy
E. Nerve biopsy

140. Antibiotic therapy is not part of the treatment of botulism; it is indicated only for the treatment of secondary infections.
Of the following, the class of antibiotics that should be avoided is
A. penicillins
B. cephalosporins
C. macrolides
D. aminoglycosides

E. quinolones

141. Which of the following types of botulism requires aggressive treatment with antibiotics in addition to antitoxin?

A. Infant
B. Foodborne
C. Wound
D. Inhalational
E. Iatrogenic

142. Which of the following is a favorable prognostic factor in tetanus?

A. Long incubation period
B. Presence of fever
C. Generalized disease
D. Onset of trismus <7 days after injury
E. Cephalic tetanus

143. A 10-year-old boy develops crushed injury during biking. The wound is contaminated with dirt. His immunization status is uncertain.
After immediate and thorough cleansing of the wound, the **NEXT** step of management is to

A. administer tetanus toxoid (TD)
B. administer tetanus immunoglobulin (TIG)
C. administer both TD and TIG
D. no TD nor TIG is required
E. administer intravenous immunoglobulin

144. A 7-year-old girl develops diarrhea, fever, and abdominal cramps. The stool contains blood and mucus. She has been diagnosed with non-Hodgkin lymphoma 6 months ago; she is on prophylactic antibiotics after receiving intensive chemotherapy.
Of the following, the valuable investigation to confirm the diagnosis is

A. ultrasonography
B. colonoscopy
C. stool analysis
D. stool culture
E. stool toxin test

145. A 10-year-old boy develops right foot pain after accidental wound injury during walking in the grass. Examination reveals right foot swelling, edema, serosanguineous discharge with a sweet odor, and extensive necrotic tissue around the wound; crepitations are felt by palpation.
Of the following, the **MOST** likely causative pathogen is

A. Staphylococcus aureus
B. Streptococcus pyogenes
C. Klebsiella pneumoniae
D. Clostridium perfringens
E. Pseudomonas aeruginosa

146. Ethambutol is an effective drug against mycobacterial infections. The major adverse effect that renders this drug reserved for use in older children is

A. headache
B. hyperuricemia
C. optic neuritis
D. peripheral neuropathy
E. hepatotoxicity

147. Trimethoprim-sulphamethaxazole is a commonly used drug in some non-tuberculous mycobacterial infections. The **MOST** common adverse effect with prolonged use of this drug is

A. myelosuppression
B. renal abnormalities
C. aseptic meningitis
D. pancreatitis
E. diarrhea

148. A medical student asks you about the distinctive feature of tuberculosis infection.
Of the following, the hallmark of tuberculosis infection (TBI) is

A. fever and night sweating
B. hemoptysis
C. positive tuberculin skin test (TST)
D. radiological evidence of lung cavitations
E. radiological evidence of diffuse lung infiltrates

149. The time between initial infection and clinically apparent TB disease is variable.
Of the following, the organ that has the latest manifestation after TB infection is

A. meninges
B. lymph nodes
C. kidney
D. bones
E. joints

150. A 12-year-old boy develops fever, productive cough, hemoptysis, night sweating, and significant weight loss. He has a history of contact with his grandmother who has have TB disease. Physical examination is insignificant.
Of the following, the **MOST** common radiological findings in this boy is

A. small nodules evenly distributed on the chest
B. unilateral pneumothorax
C. calcified hilar lymphadenopathy alveolar consolidation
D. extensive infiltrates with thick-walled cavities in the upper lobes
E. unilateral pleural effusion

151. Skeletal tuberculosis is a late complication of tuberculosis and is more likely to occur in children than in adults. Which of the following bones is **MOST** likely involved?

A. Skull
B. Vertebrae
C. Humerus
D. Pelvis
E. Femur

152. A 13-year-old girl undergoes tuberculin skin test (TST) as a prerequisite for entering a secondary school. The test shows 9 mm of induration after 12 hours of application. She has been received BCG vaccine at the age of 1 month after birth.
Of the following, your **MOST** appropriate decision is

A. to repeat the test after 1 week
B. the test should be considered positive
C. to consider the test negative
D. the girl is considered to be infected with non TB mycobacterium
E. the test is considered a reaction due to previous BCG vaccine

153. In which of the following groups of children tuberculin skin test (TST) with a duration ≥ 5 mm is considered positive?

A. Those born in high-prevalence regions of tuberculosis
B. Those in close contact with known contagious people with tuberculosis disease
C. Those with Hodgkin disease
D. Those with diabetes mellitus
E. Those with chronic renal failure

154. A medical student asks you about the advantage of using Interferon-γ Release Assay (IGRA) in tuberculosis. Which one of the following statements should be included in your answer?

A. It can differentiate between TB infection and TB disease
B. It has cross-reaction with most other mycobacteria
C. It can be performed well in malnourished infants
D. It can be performed well in children with disseminated TB disease
E. it is preferred in children >2 yr of age who have received BCG vaccine

155. A 4-year-old boy has recent exposure with his grandfather who has active tuberculosis disease. His tuberculin skin test (TST) is negative.
Of the following, your **NEXT** step in the management is to

A. start isoniazid (INH) for 10 weeks then repeat the TST
B. start INH for 9 months
C. observe for early development of tuberculosis infection
D. repeat TST after 2 weeks
E. start rifampin for 6 months

156. A gynecologist asks for your opinion regarding a pregnant woman in 2nd trimester with active TB disease. Your clinical decision is to

A. terminate the pregnancy
B. defer treatment until after delivery
C. start treatment with isoniazid, rifampin, and ethambutol
D. start treatment with aminoglycosides and ethionamide
E. do PCR for tuberculosis of the amniotic fluid

157. A 5-year-old boy develops unilateral enlarging anterior cervical lymph nodes. He has a history of playing in sandboxes. Examination shows firm, non tender, freely movable, and non erythematous lymph node. Radiological

evaluation reveals well capsulated lymph node, about 1.8 cm in diameter, with preservation of hilum. Fine needle aspiration PCR and culture reveals mycobacterium avium. Which of the following is the preferred initial treatment?

A. Wait-and-see approach
B. Complete surgical excision
C. Rifampicin for 3 months
D. Ethambutol for 3 months
E. Combination of clarithromycin and rifabutin for 3 months

158. Which of the following drugs is the only effective treatment for congenital syphilis?

A. Oral penicillin V
B. Oral ampicillin
C. Oral doxycycline
D. Parenteral crystalline penicillin G
E. Parenteral procaine penicillin

159. A 4-year-old girl develops recurrent attacks of high fever and flu-like illness lasting about 4-7 days. At the end of each febrile episode, a diffuse macular rash appears over the trunk and shoulders lasting for 1-2 days. She has poor socio-economic status, living in mud hut. She has been developed pediculosis capitis 2 weeks ago.
Of the following, the **MOST** likely diagnosis is

A. primary syphilis
B. leptospirosis
C. cat-scratch disease
D. relapsing fever
E. toxoplasmosis

160. Which of the following is the drug of choice for a 9-year-old boy with louse-borne relapsing fever?

A. Ampicillin
B. Chloramphenicol
C. Doxycycline
D. Azithromycin
E. Ceftriaxone

161. Which of the following is the 1st characteristic cutaneous manifestation of Lyme disease?

A. Erythema nodosum
B. Erythema migrans
C. Erythema annulare
D. Erythema induratum
E. Erythema toxicum

162. Which of the following cranial nerves is **MOST** commonly affected by Lyme disease?

A. Olfactory
B. Oculomotor
C. Abducent
D. Facial
E. Vagus

163. A school-aged boy developed non-productive cough; he has a preceding headache, malaise, fever, and sore throat few days before. Examination is not significant. Chest radiograph shows bronchopneumonic changes in the right lower lobe.
Of the following, the **MOST** likely causative pathogen is

A. staphylococcus aureus
B. streptococcus pneumoniae
C. mycoplasma pneumoniae
D. parainfluenza virus
E. respiratory syncytial virus

164. A 6-year-old girl develops mycoplasma pneumonia. The first-line therapy for this girl is

A. azithromycin
B. doxycycline
C. levofloxacin
D. trimethoprim
E. linezolid

165. Which of the following electrolytes disturbances is **MOST** likely encountered with amphotericin B deoxycholate therapy?

A. Hyponatremia

B. Hypokalemia
C. Hypochloremia
D. Hypocalcemia
E. Hypophosphatemia

166. A 7-year-old boy with acute lymphoblastic leukemia develops invasive pulmonary aspergillosis after prolonged neutropenia.
Of the following, the **MOST** effective antifungal drug is
A. 5-fluorocytosine (5-FC)
B. fluconazole
C. itraconazole
D. voriconazole
E. amphotericin B deoxycholate

167. What is the **MOST** effective azole antifungal treatment for mucormycosis?
A. Fluconazole
B. Itraconazole
C. Voriconazole
D. Posaconazole
E. Isavuconazole

168. A 7-year-old boy who underwent allogenic bone marrow transplantation develops refractory pulmonary aspergillosis in spite of prolonged therapy with voriconazole.
Of the following, the 2nd line of treatment is
A. fluconazole
B. itraconazole
C. 5-fluorocytosine
D. amphotericin deoxycholate
E. caspofungin

169. Which of the following is the **MOST** common manifestation of invasive candidiasis in neonatal period?
A. Meningoencephalitis
B. Endocarditis
C. Osteomyelitis
D. Arthritis
E. Endophthalmitis

170. Which of the following hematologic parameters is commonly encountered in premature infants with invasive candidiasis?

A. Neutrophilia
B. Neutropenia
C. Thrombocytopenia
D. Thrombocytosis
E. Anemia

171. A 10-day-old infant develops excessive crying, fussiness, and decreased feeding. He is a breast fed baby since delivery. His mother has been received oral antibiotics after episiotomy procedure for 1 week. Examination reveals presence of extensive pearly white, curdish material on the tongue, palate, and buccal mucosa.

Of the following, the **MOST** appropriate treatment is

A. Topical nystatin to the infant
B. Intravenous amphotericin to the infant and mother
C. Topical nystatin to the infant and mother
D. Baking soda to the infant and mother
E. No treatment is required

172. A 3-month-old infant develops diaper dermatitis manifested as a confluent erythematous rash with satellite pustules.

Of the following, the **MOST** appropriate treatment is topical

A. fucidic acid
B. clotrimazole
C. betamethasone
D. zinc oxide
E. vaseline

173. What is the **MOST** commonly recognized manifestation of cryptococcosis?

A. Pneumonia
B. Cutaneous lesions
C. Sepsis
D. Meningitis
E. Arthritis

174. Which of the following is the typical finding seen in MRI of a child with invasive pulmonary aspergillosis?

A. Halo sign
B. Air crescent sign
C. Target sign
D. Lobar emphysema
E. Segmental bronchiectasis

175. Which of the following is the diagnostic test of choice for invasive aspergillosis in children with cancer and hematopoietic stem cell transplantation?

A. Blood culture
B. Histological specimen
C. Eosinophil count
D. Galactomannan test
E. Beta-glucan assay

176. A 5-year-old boy develops fever, headache, chest pain, cough, and myalgias. He is living in a rural area; he has a history of prolonged exposure to chicken coops present nearby his house. Examination reveals hepatosplenomegaly. Chest X-ray shows right side patchy bronchopneumonia and perihilar lymphadenopathy.
Of the following, the **MOST** likely fungal infection is

A. Candida
B. Aspergillus
C. Histoplasma
D. Blastomyces
E. Coccidioides

177. Which of the following radiographic findings is **MORE** likely suggestive of pulmonary mucormycosis rather than other pulmonary invasive fungal infections?

A. Solitary pulmonary nodule
B. Segmental/lobar consolidation
C. Cavitary changes
D. Bronchopneumonic changes
E. Pleural effusions

178. Which of the following is the primary mainstay of treatment for mucormycosis?

A. Liposomal amphotericin B
B. Fluconazole
C. Voriconazole
D. Posaconazole
E. Caspofungin

179. A 9-year-old boy develops abrupt onset of dyspnea, tachypnea, fever, and non productive cough, with decreasing oxygen saturation. Examination is not conclusive. He has been diagnosed with acute lymphoblastic leukemia 9 months ago and he is now on maintenance chemotherapy. Lab tests show decreased WBC count. chest radiograph reveals bilateral diffuse interstitial ground glass infiltrates.
Of the following, the organism that is **MOST** likely detected by bronchoalveolar lavage is

A. Staphylococcus aureus
B. Streptococcus pneumoniae
C. Aspergillus fumigatus
D. Pneumocystis jirovecii
E. Cytomegalovirus

180. Which of the following is the alternative prophylactic treatment for Pneumocystis jirovecii in children who are allergic to TMP-SMX?

A. Benzathine penicillin
B. Dapsone
C. Fluconazole
D. Itraconazole
E. Chloroquine

181. You are meeting a mother of 3-year-old boy who has recently develops measles. She is asking you about the infectivity period to avoid risk of disease transmission to other siblings.
Of the following, the period of disease infectivity is

A. one day before and 3 days after the onset of rash
B. two days before and 4 days after the onset of rash
C. three days before and 6 days after the onset of rash
D. only three days before the onset of rash
E. only four days after the onset of rash

182. Which of the following is a pathognomonic feature of measles?

A. High fever
B. Photophobia
C. Conjunctivitis
D. Koplik spots enanthem
E. Maculopapular exanthem

183. Which of the following features of measles is the longest to last?

A. Fever
B. Cough
C. Conjunctivitis
D. Photophobia
E. Rash

184. A 14-year-old boy develops repetitive attacks of myoclonic jerks not associated with loss of consciousness. He has been experienced poor academic experience, irritability, reduced attention span, and temper outbursts few months ago. He has a history of measles infection at the age of 4 year. EEG shows suppression-burst episodes.
Of the following, his cerebrospinal fluid (CSF) **MOST** likely reveals

A. high WBC count
B. low sugar
C. positive measles culture
D. elevated measles antibody
E. positive measles antigen

185. A 3-year-old girl has been exposed to an index case with measles in the nursery home 4 days ago. She doesn't receive any vaccine yet.
Of the following, the **MOST** appropriate management to prevent or modify infection is

A. vaccine administration
B. immunoglobulin administration
C. vitamin A supplementation
D. ribavirin treatment
E. no treatment is required

186. Which of the following is the **MOST** common finding among infants with congenital rubella syndrome?

A. Deafness

B. Blindness
C. Congenital heart defects
D. Growth retardation
E. Psychomotor delay

187. Which of the following is the **MOST** common ocular abnormality in children with congenital rubella syndrome?

A. Cataract
B. Microphthalmia
C. Iridocyclitis
D. Glaucoma
E. Retinopathy

188. Which of the following is the **MOST** frequently encountered cardiac defect in children with congenital rubella syndrome?

A. Ventricular septal defect
B. Patent ductus arteriosus
C. Pulmonary artery stenosis
D. Pulmonary valve atresia
E. Aortic valve stenosis

189. A pregnant woman in the 1st trimester is exposed to an index case with rubella. Three subsequent blood samples for IgG are collected; the 1st one has been obtained at once, the 2nd one after 3 weeks, and the 3rd one after 6 weeks. Which of the following indicate recent infection of the mother that necessitates termination of pregnancy?

A. If the 1st blood sample is positive
B. If the 1st blood sample is negative with positivity of either the 2nd or 3rd sample
C. If the 1st and 2nd samples are positive and the 3rd one is negative
D. If all 3 samples are positive
E. If all 3 samples are negative

190. A pregnant woman in the 1st trimester is exposed to an index case of rubella. Lab tests confirms recent infection. She refused to terminate pregnancy.
Of the following, the **MOST** appropriate management is

A. close follow up

B. MMR vaccine
C. immunoglobulin administration
D. antiviral therapy
E. obligatory termination of pregnancy

191. Which of the following is the **MOST** common CNS complications of mumps?
A. Meningoencephalitis
B. Transverse myelitis
C. Acute disseminated encephalomyelitis (ADEM)
D. Facial palsy
E. Sensorineural hearing loss

192. A 13-year-old male adolescent develops fever and bilateral parotid swellings consistent with mumps. He has been never received MMR vaccine during childhood.
Of the following, the **MOST** commonly encountered complication is
A. meningoencephalitis
B. orchitis
C. pancreatitis
D. myocarditis
E. thyroiditis

193. Which of the following is a feature of poliomyelitis rather than Guillain-Barré syndrome?
A. Asymmetric paralysis
B. Absence of fever
C. Sensory changes
D. High CSF protein
E. Normal CSF cells

194. A 4-year-old boy develops painful cutaneous lesions involving lips and both hands associated with low grade fever. Examination shows tender vesicles on the tongue, buccal mucosa, lips, and dorsal surface and palms of the hand; some of those vesicles ulcerate leaving shallow lesions with surrounding erythema.
Of the following, the **MOST** likely causative pathogen is
A. Herpes simplex virus

B. Parechovirus
C. Coxsackie A virus
D. Coxsackie B virus
E. Adenovirus

195. Which of the following is the **MOST** common ocular manifestations caused by enteroviruses?
A. Chorioretinitis
B. Uveitis
C. Acute hemorrhagic conjunctivitis
D. Optic neuritis
E. Acute maculopathy.

196. A 6-year-old girl develops low grade fever, headache, and flu-like illness. She then develops bilateral cheeks erythema followed by diffuse macular rash over the trunk and extensors of extremities, with central clearing in some areas giving a reticulate appearance. Soles and palms are spared.
Of the following, the **MOST** likely diagnosis is
A. erythema nodosum
B. erythema multiforme
C. erythema marginatum
D. erythema infectiosum
E. erythema migrans

197. Which of the following is a distinctive feature of Parvo B19 virus infection in children with chronic hemolytic anemias in comparison with erythema infectiosum only?
A. Well-looking appearance
B. Profound anemia
C. Absence of fever
D. Extensive rash
E. Long incubation period

198. Which of the following is the **EARLIEST** clinical feature of herpes labialis in older children?
A. Fever
B. Vesicular lesions
C. Burning sensation
D. Sore throat

E. Local lymphadenopathy

199. A 7-year-old boy develops fever, headache, vomiting, nuchal rigidity, followed by generalized seizures and alteration level of consciousness. CSF analysis reveals WBC, 550/mm^3 (80% lymphocytes, and 20% neutrophils); RBC, 30/mm^3; protein, 50 gm/dL; sugar, 40 gm/dL. MRI of the brain shows right temporal lobe abnormalities including edema and diffuse enhancement.
Of the following, the **MOST** appropriate initial treatment is

A. intravenous ceftriaxone
B. intravenous vancomycin
C. intravenous amphotericin
D. intravenous ganciclovir
E. intravenous acyclovir

200. A full term neonate is born to a mother who develops varicella 3 days before delivery. Examination is normal with no evidence of cutaneous lesions.
Of the following, the **MOST** appropriate management is

A. close observation for the development of rash
B. to give intravenous immunoglobulin (IVIG)
C. to give varicella-zoster immunoglobulin (VZIG)
D. to start acyclovir prophylaxis
E. to give varicella vaccine

201. Which of the following is a characteristic feature of congenital varicella syndrome?

A. Microcephaly
B. Microphthalmia
C. Cataracts
D. Low birth weight
E. Cicatricial skin scarring

202. Which of the following sites of lymphadenopathy is particularly suggestive of infectious mononucleosis?

A. Anterior cervical
B. Posterior cervical
C. Axillary
D. Epitrochlear
E. Inguinal

203. Which of the following malignancies is associated with Epstein-Barr virus infections?

A. Retinoblastoma
B. Medulloblastoma
C. Nasopharyngeal carcinoma
D. Nephroblastoma
E. Neuroblastoma

204. An 8-month-old boy develops high grade fever associated with fussiness and irritability. Three days later, there is abrupt defervescence with the appearance of rose-colored, nonpruritic, 2-3 mm morbilliform rash over the trunk. He is a breast fed infant, and his vaccination history is complete up to his age.

Of the following, the **MOST** likely diagnosis is

A. measles
B. rubella
C. erythema infectiosum
D. erythema toxicum
E. exanthem subitum

205. Which of the following is the **MOST** common complication of the sixth disease?

A. Seizure
B. Acute disseminated demyelination
C. Arthritis
D. Hepatitis
E. Myocarditis

206. Which of the following is the 1st sign of respiratory syncytial virus infection in infants?

A. Fever
B. Rhinorrhea
C. Cough
D. Audible wheezing
E. Peripheral cyanosis

207. Which of the following features differentiate RSV bronchiolitis from Chlamydia trachomatis interstitial pneumonia in infants of less than 4 months of age?

A. Cough
B. Fever
C. Nasal discharge
D. Inspiratory crackles
E. Expiratory wheezing

208. Which of the following is the **MOST** common manifestation of human adenovirus infection in children?

A. Meningoencephalitis
B. Myocarditis
C. Conjunctivitis
D. Hepatitis
E. Hemorrhagic cystitis

209. A 10-month-old healthy male infant develops watery diarrhea of 2 days duration. There has been a preceding mild fever and vomiting that are stopped 1 day ago. He is a breast fed infant with introduction of solid food after the age of 6 month. Examination shows moderate dehydration. Stool analysis reveals absence of blood or white blood cells.
Of the following, the **MOST** important step of management is

A. correction of dehydration
B. stopping breast feeding
C. treatment with TMP-SMX
D. ondansetron antiemetic
E. probiotic therapy

210. A 7-year-old girl is exposed to a bite by a dog. The dog is kept for observation for 10 days.
Of the following, the **MOST** appropriate management is to

A. give immediately killed rabies vaccine
B. give immediately rabies immunoglobulin (RIG)
C. give immediately both vaccine and RIG
D. give immediately both vaccine and RIG if the dog shows 1st sign of rabies
E. euthanize the dog immediately and test the brain tissue for rabies

211. Which of the following microscopical stool findings is specific for Entamoeba histolytica rather than Entamoeba dispar?

A. Presence of leucocytes
B. Presence of fat droplets
C. Presence of phagocytosed erythrocytes
D. Presence of trophozoites
E. Presence of cysts

212. Which of the following immunodeficiencies is particularly associated with chronic Giardia infection?

A. Chronic granulomatous disease
B. X-linked hypogammaglobulinemia
C. Leukocyte adhesion deficiency
D. DiGeorge syndrome
E. Chronic mucocutaneous candidiasis

213. Which of the following has the highest diagnostic sensitivity for detection of Leishmania donovani infection (kala-azar)?

A. Indirect fluorescence assay
B. K39 immunochromatographic strip test
C. Direct agglutination test
D. Bone marrow aspiration
E. Splenic aspiration

214. You are discussing malaria infection in children with a group of medical students.
An important statement that should be included in the discussion is

A. incubation period is the longest with P. falciparum species
B. children with malaria often lack the typical paroxysms seen in adults
C. periodicity of paroxysms is more apparent with P. falciparum
D. complications are the highest with P. knowlesi species infection
E. most common serious complication is bleeding diathesis

215. Which of the following species of malaria causes nephrotic syndrome as a complication?

A. P. vivax
B. P. ovale
C. P. malariae

D. P. falciparum
E. P. knowlesi

216. Which of the following antimalarial drugs is used to prevent relapse in P. vivax and P. ovale infections?

A. Primaquine
B. Chloroquine
C. Quinine sulphate
D. Quinidine gluconate
E. Mefloquine

217. A 9-year-old boy is going to travel with his parents to chloroquine-resistant P. falciparum endemic country. The plan of the parents is to stay there for 1 month. He has medical problems, including seizure disorder and G6PD deficiency that are diagnosed since early childhood.
Of the following, the **MOST** appropriate chemoprophylaxis is

A. atovaquone-proguanil
B. mefloquine
C. primaquine
D. doxycycline
E. sulfadoxine-pyrimethamine

218. Which of the following is the **MOST** common ocular manifestation of toxoplasmosis?

A. Strabismus
B. Microphthalmia
C. Cataracts
D. Nystagmus
E. Chorioretinitis

219. Which of the following sites has a propensity to be involved by calcifications secondary to congenital toxoplasmosis?

A. Frontal cerebral cortex
B. Cerebellum
C. Caudate nucleus
D. Corpus callosum
E. Hypothalamus

220. Which of the following is the drug of choice for intestinal or biliary obstruction caused by Ascaris lumbricoides infestations?

A. Albendazole
B. Mebendazole
C. Lvermectin
D. Piperazine citrate
E. Nitazoxanide

221. Which of the following worms' infestation can cause iron deficiency anemia?

A. Tapeworms
B. Threadworms
C. Roundworms
D. Hookworms
E. Pinworms

222. Which of the following worms' infestation can cause nocturnal perianal pruritis with subsequent sleep disturbances?

A. Tapeworms
B. Threadworms
C. Roundworms
D. Hookworms
E. Pinworms

223. Which of the following is the recommended treatment for schistosomiasis?

A. Piperazine citrate
B. Praziquantel
C. Albendazole
D. Mebendazole
E. Lvermectin

224. Which of the following parasitic infestation can cause megaloblastic anemia?

A. Diphyllobothrium latum
B. Hymenolepis nana
C. Ancylostoma duodenale
D. Taenia saginata
E. Ascaris lumbricoides

225. A 2-month-old infant develops pneumonia described as continuous cough and tachypnea without fever. He has a history of normal vaginal delivery after prolonged rupture of membrane. Examination shows scattered rales. Lab tests reveal absolute eosinophilia (500/µL); chest radiograph shows hyperinflation with minimal infiltrates.

Of the following, the **MOST** likely cause is

A. S. aureus
B. E. coli
C. C. trachomatis
D. respiratory syncytial virus
E. adenovirus

Chapter 16

Infectious Diseases Answers

USAMA A. AL JUMAILY

1.(D) MacConkey agar supports growth of gram-negative rods while suppressing gram-positive organisms. Nutrient-rich media such as sheep blood agar and chocolate agar are used to aid in the recovery of fastidious pathogens. Special media, such as Sabouraud dextrose agar and inhibitory mold agar, are used to recover fungi.

2.(C) Propionibacterium and coagulase-negative staphylococci are the organisms most frequently isolated from shunt infections.

3.(B) Plasmodium and Babesia can be detected in stained blood smears, Leishmania can be detected in stained bone marrow smears. Serologic tests are important in documenting exposure to certain parasites that are not typically found in stool or blood, and thus are difficult to demonstrate in clinical specimens, such as Trichinella.

4.(A) An age-dependent change in lower airway colonization occurs among CF patients, which starts in early childhood with S. aureus and H. influenzae and progressively shifts toward more intrinsically multidrug-resistant organisms, including the notoriously persistent and treatment-refractory bacteria Pseudomonas aeruginosa and Burkholderia cepacia complex which is strongly correlated with disease severity.

5.(B) Fecal transplantation, or administration of feces from healthy donors to CDAD recipients, is cost-effective treatment and superior to antibiotics in reducing the likelihood of recurrent disease.

6.(D)

7.(A) IGIV may be used for varicella after exposure when varicella-zoster immune globulin is not available. IGIM is mainly used for Hepatitis A, measles, and rubella prophylaxis. IGSC is used for treatment of patients with primary immunodeficiencies. Palivizumab monoclonal antibody is used for prophylaxis against respiratory syncytial virus.

8.(B) Vaccines consist of whole inactivated microorganisms (e.g., polio, hepatitis A), parts of the organism (e.g., acellular pertussis, HPV, hepatitis B), polysaccharide capsules (e.g., pneumococcal and meningococcal polysaccharide

vaccines), polysaccharide capsules conjugated to protein carriers (e.g., Hib, pneumococcal, and meningococcal conjugate vaccines), live-attenuated microorganisms (e.g., measles, mumps, rubella, varicella, rotavirus, and live-attenuated influenza vaccines), and toxoids (e.g., tetanus, diphtheria).

9.(E) HIV-infected children may receive varicella vaccine if $CD4^+$ T-lymphocyte percentage is ≥15% for those <6 yr old, or CD4+ T-lymphocyte count ≥200 cells/mm^3 for those ≥6 yr old. MMR may be given to a child with HIV infection provided the child is asymptomatic or symptomatic without evidence of severe immunosuppression.

10.(C) Children with complement deficiency disorders may receive all vaccines, including live-attenuated vaccines. In contrast, children with phagocytic disorders may receive both inactivated and live-attenuated viral vaccines but not live-attenuated bacterial vaccines.

11.(D) Children receiving corticosteroids (≥2 mg/kg/day or ≥20 mg/day of prednisone or equivalent) for ≥14 days should not receive live vaccines until therapy has been discontinued for at least 1 mo. Children on the same dose levels but for <2 wk may receive live virus vaccines as soon as therapy is discontinued.

12.(E) Infants born to HBsAg-negative mother, with a weight ≥2 kg and who are stable should receive a birth dose of hepatitis B vaccine within the 1^{st} 24 hr of life. However, hepatitis B vaccine should be deferred in infants weighing <2 kg at birth until chronological age of 1 mo. All preterm, low-birth weight infants born to HBsAg-positive mothers should receive hepatitis B immunoglobulin (HBIG) and hepatitis B vaccine (at separate anatomic sites) within 12 hr of birth.

13.(B) Immunoglobulin does not interfere with inactivated vaccines. However, immunoglobulin can interfere with the immune response to measles vaccine and by inference to varicella vaccine. In general, immunoglobulin, if needed, should be administered at least 2 wk after the measles vaccine. Depending on the dose of immunoglobulin received, MMR should be deferred for as long as 3-11 mo. Immunoglobulin is not expected to interfere with the immune response to LAIV or rotavirus vaccines.

14.(A) Hand washing is an important method for infection prevention and control.

15.(C) Fungi and multidrug-resistant organisms are common causes of infection in immunocompromised children as well as those requiring intensive care and prolonged hospitalization.

16.(D) Transmission by droplets involves the propulsion of infectious large particles over a short distance (<3 feet), with deposition on another's mucous

membranes or skin. Droplet isolation requires the use of gloves and gowns, as well as masks and eye guards when closer than 3 feet to the patient.

17.(B) A single dose of hepatitis A given to travelers will provide adequate protection. Protective immunity develops within 2 wk after the initial vaccine dose.

18.(A) Enteric protozoa are a much less common cause of traveler's diarrhea than bacterial pathogens; Giardia lamblia is the most likely protozoal cause of persistent diarrhea. Less common travel-associated protozoa include Cryptosporidium spp., Entamoeba histolytica, and Cyclospora.

19.(D) Azithromycin is highly effective against most bacterial pathogens that cause traveler's diarrhea and is the preferred antibiotic among many travel experts. Ciprofloxacin is an alternative for children >1 yr old but should not be prescribed for travelers to the Indian subcontinent or Southeast Asia, where fluoroquinolone resistance is common.

20.(C) Drugs known to cause fever include vancomycin, amphotericin B, and allopurinol.

21.(A) Other disorders are characterized by recurrent episodes of fever that do not follow a strictly periodic pattern.

22.(B) Relative bradycardia can accompany typhoid fever, brucellosis, leptospirosis, or drug fever. It may also be a result of a conduction defect resulting from cardiac involvement with acute rheumatic fever, Lyme disease, viral myocarditis, or infective endocarditis.

23.(D)

24.(A)

25.(D) The presence of drooling and neck or throat pain in an infant or toddler may be suggestive of a retropharyngeal abscess, which is usually confirmed by imaging that may include a lateral radiograph of the soft tissue of the neck or computed tomography (CT) if clinical suspicion is high. CT scan is the definitive diagnostic imaging test. X-ray of the neck often (80% of the time) shows swelling of the retropharyngeal space in affected individuals.

26.(C) Pseudo-FUO is defined as successive episodes of benign, self-limited infections with fever that the parents perceive as 1 prolonged fever episode. It usually starts with a well-defined infection (frequently viral) that resolves but is followed by other febrile viral illnesses that may be less well defined.

27.(E) The continuing absence of sweat in the presence of an elevated or changing body temperature suggests dehydration caused by vomiting, diarrhea, or central or nephrogenic diabetes insipidus. It also should suggest anhidrotic ectodermal dysplasia, familial dysautonomia, or exposure to atropine.

28.(C) Palpebral conjunctivitis in a febrile patient may be a clue to measles, coxsackievirus infection, tuberculosis, infectious mononucleosis, lymphogranuloma venereum, or cat-scratch disease. In contrast, bulbar conjunctivitis in a child with FUO suggests Kawasaki disease or leptospirosis.

29.(A)

30.(C) Sepsis can occur but is more common with certain gram negative organisms such as C. violaceum and F. philomiragia.

31.(E) Unlike a true pure antibody defect, besides being important in T cell–B cell interactions, CD40 ligand is also important in the interaction between T cells and macrophages/monocytes, influencing opportunistic infections such as Pneumocystis jiroveci pneumonia (PCP) and Cryptosporidium intestinal infection in such disease.

32.(B) Indications for using vancomycin in children with febrile neutropenia include severe mucositis, hypotension, quinolones prophylaxis, colonization with methicillin resistant staphylococcus aureus, and obvious catheter related infections.

33.(E) Viridans streptococci are particularly important potential pathogens in patients with the oral mucositis that is often associated with use of cytarabine and in patients who experience selective pressure from treatment with certain antibiotics such as quinolones. Infection caused by this group of organisms can present as acute septic shock syndrome.

34.(B) The Most commonly used antibiotic monotherapy is cefepime or piperacillin-tazobactam. Ceftazidime should not be used as monotherapy if concern exists for gram-positive organisms or resistant gram negative bacteria. Carbapenems such as imipenem/cilastin and meropenem should not be first line, aiming to prevent pressure on carbapenem-resistant Enterobacteriaceae. The addition of a 2^{nd} anti gram-negative bacterial agent (e.g., aminoglycoside) for empirical therapy can be considered in patients who are clinically unstable when multidrug-resistant organisms are suspected.

35.(E) Post transplant lymphoproliferative disorder is more common among children than adults, because primary EBV infection in the immunosuppressed host is more likely to lead to uncontrolled proliferative disorders, including post transplant lymphoma.

36.(E) Green discharge is strongly suggestive of mycobacterial infection, and appropriate stains and culture should be performed.

37.(D) Prolonged therapy (4-6 wk) is indicated for persistent bacteremia or fungemia despite catheter removal, since this may represent unrecognized infective endocarditis or thrombophlebitis.

38.(A) With the advent of conjugate vaccines against Hib, invasive disease has declined dramatically. However, outbreaks still occur, and have been observed in the context of parental refusal of vaccines.

39.(E) MRSA and streptococcus pneumoniae mediate resistance to penicillins through mechanisms other than β-lactamase production, rendering other antibiotics agents of little value for the management of these infections.

40.(E) The third generation cephalosporins (cefotaxime, ceftriaxone, and ceftazidime) are typically used for serious pediatric infections. Ceftazidime is highly active against most strains of P. aeruginosa. A fourth-generation cephalosporin (cefepime) has activity against P. aeruginosa and retains good activity against methicillin-susceptible staphylococcal infections.

41.(C) The first-generation cephalosporins are effective against susceptible strains of S. aureus and group A streptococcus. The second, third, and fourth generation cephalosporins have better activity against gram-negative bacterial infections. Ceftaroline (a fifth generation cephalosporin) has bactericidal activity against resistant gram-positive organisms, including MRSA, and common gram-negative pathogens.

42.(C) Meropenem is not active against Stenotrophomonas maltophilia, rendering their use for cystic fibrosis patients who are infected with this organism problematic. MRSA and E. faecium are not susceptible to carbapenems. Ertapenem is poorly active against P. aeruginosa and Acinetobacter species and should be avoided when these pathogens are encountered.

43.(A) It is also important in the management of Mycobacterium tuberculosis and atypical mycobacterial infections.

44.(D) Colistin is effective for treatment of multidrug resistant gram negative organisms (Enterobacteriaceae including extended-spectrum beta lactamase and carbapenemase producing strains).

45.(B) Direct invasion can result in osteomyelitis, suppurative arthritis, pyomyositis, deep abscesses, pneumonia, empyema, endocarditis, pericarditis, and rarely meningitis.

46.(A) Group A streptococci skin infections are prone to spread more rapidly and can be very aggressive.

47.(D) Other etiology of cavitary pneumonia includes M. tuberculosis.

48.(B) Initial treatment for serious infections thought to be caused by MSSA should include semisynthetic penicillin (e.g., nafcillin) or a first-generation cephalosporin. Penicillin and ampicillin are not appropriate, because >90% of all

staphylococci isolated are resistant to these agents. Quinolone antibiotics have unpredictable activity against MSSA and no activity against MRSA.

49.(B) Other features commonly seen in TSS include diffuse myalgia, vomiting, abdominal pain, diarrhea, azotemia, ARDS, and shock.

50.(D)

51.(D) Unlike most CoNS, S. saprophyticus is usually methicillin susceptible, and UTI can typically be treated with a first generation cephalosporin (cephalexin), amoxicillin–clavulanic acid, or TMP-SMX.

52.(C) Peritonitis caused by S. epidermidis in patients on continuous ambulatory peritoneal dialysis is an infection that may be treated with IV or intraperitoneal antibiotics without removing the dialysis catheter.

53.(D) Empirical therapy with a parenteral third generation cephalosporin should be prescribed for hospitalized infants and children who are not fully immunized, in regions where local epidemiology of invasive pneumococcal strains documents widespread penicillin resistance, or for infants and children with life-threatening infection, including those with empyema. Non–β-lactam agents (vancomycin) have not been shown to be more effective than third-generation cephalosporins in the treatment of pneumococcal pneumonia.

54.(A) For individuals with a non-type I allergic reaction to penicillin, cephalosporins (standard dosing) can be used. For type I allergic reactions (immediate, anaphylactic) to β-lactam antibiotics, clindamycin and levofloxacin are preferred alternatives.

55.(A) Pneumococcal meningitis results in sensorineural hearing loss in 20–30% of patients.

56.(A) Streptococcus pyogenes is a common cause of infections of the upper respiratory tract (pharyngitis) and the skin (impetigo, pyoderma) in children. Less frequently, it causes perianal cellulitis, vaginitis, septicemia, pneumonia, endocarditis, pericarditis, osteomyelitis, suppurative arthritis, myositis, cellulitis, omphalitis, and other infections.

57.(D)

58.(B) The anti–streptolysin O response can be feeble after group A streptococcal skin infection. In contrast, the anti–DNase B responses are generally present after either skin or throat infections. Slide agglutination (Streptozyme) test for the detection of antibodies to several streptococcal antigens is much less well standardized and less reproducible than other antibody tests, and it should not be used as a test for evidence of a preceding GAS infection. Streptococcal rapid antigen detection and isothermal loop

amplification are used for identification of recent GAS infection directly from throat swab.

59.(C)

60.(B) Some penicillin-allergic persons (up to 10%) are also allergic to cephalosporins, and these agents should be avoided in patients with immediate (anaphylactic-type) hypersensitivity to penicillin. Oral clindamycin is an appropriate agent for treating penicillin-allergic patients. An oral macrolide (erythromycin or clarithromycin) or azalide (azithromycin) is also an appropriate agent for patients allergic to penicillins.

61.(B) PSRA usually involves the large joints similar to the arthritis of acute RF, but it may also involve small peripheral joints, as well as the axial skeleton. The latent period between the antecedent episode of GAS pharyngitis and PSRA may be considerably shorter (usually <10 days) than that typically seen with acute RF (usually 14-21 days). PSRA does not respond dramatically to therapy with aspirin or other nonsteroidal anti inflammatory drugs (NSAIDs). In addition, fewer patients with PSRA than with acute RF have temperature >38°C (100.4°F). Very small proportions of patients with PSRA have been reported to develop valvular heart disease and carditis.

62.(E) The diagnosis of acute RF can be made without strict adherence to the Jones Criteria in 3 circumstances: (1) when chorea occurs as the only major manifestation of acute RF, (2) when indolent carditis is the only manifestation in patients who first come to medical attention only months after the apparent onset of acute RF, and (3) in a limited number of patients with recurrence of acute RF in particularly high-risk populations.

63.(A) Arthritis in acute rheumatic fever typically involves larger joints, particularly the knees, ankles, wrists, and elbows. Involvement of the spine, small joints of the hands and feet, or hips is uncommon.

64.(C) Rheumatic joints exquisitely tender, with even the friction of bedclothes being uncomfortable. The joint involvement is characteristically migratory in nature. A dramatic response to even low doses of salicylates is another characteristic feature of the arthritis, and the absence of such a response should suggest an alternative diagnosis.

65.(A)

66.(B) Valvular insufficiency is characteristic of both acute and convalescent stages of acute RF, whereas mitral and/or aortic valvular stenosis usually appears years or even decades after the acute illness.

67.(E)

68.(D) Patients with typical migratory polyarthritis and those with carditis without cardiomegaly or congestive heart failure should be treated with oral salicylates. Patients with carditis and more than minimal cardiomegaly and/or congestive heart failure should receive corticosteroids.

69.(B)

70.(D) Early-onset neonatal group B streptococci (GBS) disease presents within the 1st 6 days of life and is often associated with maternal obstetric complications, including chorioamnionitis, prolonged rupture of membranes, and premature labor.

71.(A)

72.(D) Late-onset enterococcus infection (≥7 days old) is associated with risk factors such as extreme prematurity, presence of an intravascular catheter, or necrotizing enterocolitis (NEC), or it follows an intraabdominal surgical procedure.

73.(E) Cephalosporins should not be relied on in situations where Enterococcus is known or suspected to be involved. Vancomycin can be substituted for the penicillins in allergic patients but should be used with an aminoglycoside because vancomycin alone is not bactericidal.

74.(A) Linezolid is a bacteriostatic antibiotic against most E. faecium and E. faecalis isolates, including VRE isolates. Response rates are generally >90%, including cases of bacteremia and sepsis, and this antibiotic has become the preferred agent in treatment of VRE infections in many institutions. Experience with daptomycin in children is limited, particularly in the setting of Enterococcus infections. Quinupristin-dalfopristin is inactive against E. faecalis. Tigecycline is an expended-spectrum derivative of the tetracycline family. Its efficacy in VRE infections has not yet been demonstrated in clinical trials. Tigecycline may cause discoloration of the teeth, and its use in children <8 yr old should generally be avoided.

75.(A) All other manifestations are systemic due to toxin absorption.

76.(B) The characteristic adherent membrane, extension beyond the faucial area, dysphagia, and relative lack of fever help differentiate diphtheria from exudative pharyngitis.

77.(A) Subtle signs of myocarditis can be detected in most patients.

78.(D) Tachycardia disproportionate to fever is common and may be evidence of cardiac toxicity or autonomic nervous system dysfunction. A prolonged P-R interval and changes in the ST-T wave on an electrocardiographic tracing are relatively frequent findings. Cardiac dysrhythmias can occur. Ventricular tachycardia is also described.

79.(C) Specific antitoxin is the mainstay of therapy and should be administered on the basis of clinical diagnosis. Because it neutralizes only free toxin, antitoxin efficacy diminishes with elapsed time after the onset of mucocutaneous symptoms. The role of antimicrobial therapy is to halt toxin production, treat localized infection, and prevent transmission of the organism to contacts.

80.(D) Early-onset neonatal listeriosis (<5 days of birth), is predominantly septicemic form, characterized by premature delivery or still birth, obstetric complications, and multiorgan involvement including rash but without CNS localization.

81.(A) The presence of elevated peripheral blood monocytes suggests listeriosis. Monocytosis or lymphocytosis may be modest or striking.

82.(C)

83.(B) L. monocytogenes is not susceptible to the cephalosporins, including third-generation cephalosporins. If these agents are used for empirical therapy for neonatal sepsis or meningitis in a newborn, ampicillin must be added for possible L. monocytogenes infection.

84.(C) The hallmark of actinomycosis is contiguous spread that fails to respect tissue or fascial planes, with formation of suppuration, interconnecting abscesses and sinus tracts. The presence of sulfur granules is highly suggestive of the diagnosis. Risk factors for actinomycosis in children include trauma, dental caries, debilitation, and poorly controlled diabetes mellitus.

85.(E) Nocardia organisms are primarily opportunistic pathogens infecting immunocompromised persons. Nocardiosis may be acute, subacute, or chronic suppurative infections. Dissemination after primary lung infection is common. The central nervous system (CNS) is the most concerning and most common secondary site of infection.

86.(A)

87.(D) It typically affects the lower limbs and can lead to substantial scarring and require skin grafting.

88.(E) Poor prognostic factors on presentation include hypothermia or extreme hyperpyrexia, hypotension or shock, purpura fulminans, seizures, leucopenia, thrombocytopenia (including DIC), acidosis, and high circulating levels of endotoxin and TNF-α. The presence of petechiae for <12 hr before admission, absence of meningitis, and low or normal ESR indicate rapid, fulminant progression and poorer prognosis.

89.(C) Ceftriaxone and ciprofloxacin are the most effective agents for prophylaxis. Ciprofloxacin is not recommended in pregnant women. Rifampin is most widely used but fails to eradicate colonization in 15% of cases.

Azithromycin is not recommended routinely. Neither penicillin nor ampicillin treatment eradicates nasopharyngeal carriage and should not be routinely used for prophylaxis.

90.(A) Thin noncornified vaginal epithelium and alkaline pH of the vaginal mucin predispose this age group to infection of the lower genital tract.

91.(B) Meningitis, carditis, and osteomyelitis are rare manifestations.

92.(A) Cefotaxime should be substituted for ceftriaxone in neonates with hyperbilirubinemia (particularly those who are premature) and in those <28 days old if receiving calcium-containing intravenous fluids.

93.(D) Kingella kingae is the most common etiology of septic arthritis, osteomyelitis, and spondylodiscitis in young children. Older children and adults with K. kingae infections often have underlying chronic diseases, immunosuppressing conditions, malignancy, or cardiac valve pathology.

94.(E) Pneumonia is an exceptional clinical presentation.

95.(B) K. kingae osteomyelitis usually involves the long bones of the extremities. The calcaneus, talus, sternum, and clavicle are also frequently affected (and are rarely infected by other bacterial pathogens).

96.(A) K. kingae is always resistant to glycopeptide antibiotics, and the majority of isolates are also resistant to clindamycin**.**

97.(D) Dexamethasone, particularly when given shortly before or concurrent with the initiation of antimicrobial therapy, decreases the incidence of hearing loss.

98.(A) Absence of fever and interruption of the integument (by insect bite or trauma) are more likely to occur with the latter 2 pathogens.

99.(E) Most H. influenzae isolates causing otitis media are nontypeable.

100.(E) Orbital cellulitis may manifest as lid edema but is distinguished by the presence of proptosis, chemosis, impaired vision, limitation of the extraocular movements, decreased mobility of the globe, or pain on movement of the globe.

101.(D) Cough may not be prominent, especially in the early phase, and whoop is infrequent. Apnea and cyanosis can follow a coughing paroxysm, or apnea can occur as the only symptom (without cough).

102.(B) Pertussis should be suspected in any individual who has paroxysms of cough of ≥14 days' duration, with whoop or posttussive vomiting, especially in the absence of fever, malaise or myalgia, exanthem or enanthem, sore throat, hoarseness, tachypnea, wheezes, and rales.

103.(C) Leukocytosis (15,000-100,000 cells/μL) caused by absolute lymphocytosis is characteristic in the catarrhal stage. Lymphocytes are normal

small cells, rather than the large, atypical lymphocytes seen with viral infections. Neutrophilia suggests secondary bacterial infection. Eosinophilia is not a manifestation of pertussis.

104.(D) Severe course and death are correlated with rapid-rise and extreme leukocytosis (median peak white blood cell count in fatal vs nonfatal cases, 94,000 vs 18,000/µL, respectively) and thrombocytosis (median peak platelet count in fatal vs nonfatal cases, 782,000 vs 556,000/µL, respectively), both of which are findings with progressive pulmonary hypertension.

105.(C) Azithromycin is the drug of choice in all age-groups, for treatment or postexposure prophylaxis. However, there is a risk of fatal heart rhythms with use of azithromycin in patients already at risk for cardiovascular events, especially those with prolongation of the QT interval. TMP-SMX is an alternative to azithromycin for infants >2 mo old and children unable to receive azithromycin.

106.(D) Progressive pulmonary hypertension in very young infants and secondary bacterial pneumonia are severe complications of pertussis and are the usual causes of death.

107.(A) Antibiotics are not generally recommended for the treatment of isolated uncomplicated Salmonella gastroenteritis in immunocompetent child, because they may create a chronic carrier state. However, they are recommended in young infants (<3 mo old), and in immune compromised children.

108.(C) Children with HIV infection may develop florid disseminated disease. Children with schistosomaisis are also at a greater risk for development of chronic salmonellosis.

109.(D) The mainstay of the diagnosis of typhoid fever is a positive result of culture. Results of blood cultures are positive in 40–60% of the patients seen early in the course of the disease. Widespread liberal antibiotic use may render bacteriologic confirmation even more difficult. Bone marrow cultures may increase the likelihood of bacteriologic confirmation of typhoid.

110.(E) Diarrhea, toxicity, and complications such as disseminated intravascular coagulation (DIC) are more common in infancy, resulting in higher case fatality rates. However, GI and neurologic complications seen in adults are rare in children.

111.(A)

112.(A) S. dysenteriae serotype 1 infection also frequently causes rectal prolapse, and hemolytic-uremic syndrome.

113.(E) Presumptive data supporting a diagnosis of bacillary dysentery include the finding of fecal leukocytes (usually >50 or 100 PMNs per high-power field, confirming the presence of colitis), fecal blood, and demonstration in peripheral blood of leukocytosis with a dramatic left shift (often with more bands than segmented neutrophils).
114.(B) Ciprofloxacin (20-30 mg/kg/24 hr divided into 2 doses) is the drug of choice recommended by WHO for all patients with bloody diarrhea, regardless of age. Concurrent zinc supplementation is recommended with antibiotic therapy.
115.(D) It is also known as enterohemorrhagic E. coli (EHEC) or verotoxin-producing E. coli (VTEC).
116.(E) The STEC strains represent a particularly difficult therapeutic dilemma; many antibiotics can induce bacterial stress, toxin production, and phage-mediated bacterial lysis with toxin release. Antibiotics should not be given for STEC infection because they can increase the risk of HUS**.**
117.(B)
118.(A) Ingestion of contaminated poultry and raw milk are mainly common sources of this pathogen.
119.(B) C. fetus shows a predilection for vascular endothelium, leading to endocarditis, pericarditis, thrombophlebitis, and mycotic aneurysms. C. hyointestinalis has been associated with proctitis, C. upsaliensis with breast abscesses, and C. rectus with periodontitis.
120.(D) C. jejuni has been identified as the trigger in up to 40% of patients with Guillain-Barré syndrome (GBS). When associated with Campylobacter, GBS is more likely to be the axonal form and has a worse prognosis with slower recovery and more neurologic disability.
121.(C) Y. enterocolitica is transmitted to humans through food, water, animal contact, and contaminated blood products. Conditions associated with iron overload increase risk of infection with this pathogen. Prominent pharyngitis may be seen in 20% of patients at presentation, which may help distinguish it from other causes of gastroenteritis.
122.(B)
123.(A) Y. enterocolitica produces β-lactamases, which are responsible for resistance to penicillins and first-generation cephalosporins.
124.(B) Patients with suspected plague should be placed on droplet isolation until pneumonia is ruled out, sputum cultures are negative, and antibiotic treatment has been administered for 48 hr. The treatment of choice for bubonic plague has been streptomycin for 10 days. Postexposure prophylaxis should be

given to close contacts of patients with pneumonic plague. Contacts of cases of uncomplicated bubonic plague do not require prophylaxis.

125.(B) It begins as pink macules and progress to hemorrhagic nodules and eventually to ulcers with ecchymotic and gangrenous centers with eschar formation, surrounded by an intense red areola.

126.(D)

127.(E) Children with leukemia or other malignancies, particularly those who are receiving immunosuppressive therapy and who are neutropenic, typically with intravascular catheters, are extremely susceptible to septicemia caused by P. aeruginosa**.** Signs of sepsis are often accompanied by a generalized vasculitis, and hemorrhagic necrotic lesions may be found in all organs.

128.(E) Macrolide therapy decreases pulmonary exacerbations in patients with chronic lung disease and P. aeruginosa infection. The mechanism likely relates to altering the virulence properties of P. aeruginosa rather than direct bacterial killing.

129.(B) Fatigue, sweats, chills, anorexia, headache, weight loss, and malaise, are reported in the majority of adult cases but are less frequent in children.

130.(D) Cultures are insensitive and positive only in a minority of cases. Bone marrow cultures may be superior to blood cultures when evaluating patients who have received previous antimicrobial therapy. The enzyme immunoassay should only be used for suspected cases with negative serum agglutination tests or for the evaluation of complicated cases, suspected chronic brucellosis, or reinfection. Polymerase chain reaction assays are not available in most clinical laboratories.

131.(E) Combination therapy is generally recommended because of the risk of relapse with monotherapy. Doxycline is recommended for children more than 8-year-old.

132.(A) Treatment may need to be continued for up to 1 yr in severe cases of central nervous system (CNS) disease.

133.(C) Corticosteroid therapy poses a high risk for infection by interfering with T cell and macrophage function. It is a major risk factor combined with exposure to contaminated potable water.

134.(B) After an incubation period of 7-12 days, 1 or more red papules develop at the site of cutaneous inoculation, often reflecting a linear cat scratch. Lymphadenopathy is generally evident within 1-4 wk. Epstein-Barr virus, cytomegalovirus, and Toxoplasma gondii infections usually cause more generalized lymphadenopathy.

135.(B) Chronic regional lymphadenitis is the hallmark of the disease. Fever occurs in approximately 30% of patients, usually 38-39°C. Other nonspecific symptoms, including malaise, anorexia, fatigue, and headache, affect less than one third of patients.

136.(D) Affected lymph nodes in order of frequency include the axillary, cervical, submandibular, preauricular, epitrochlear, femoral, and inguinal nodes.

137.(D) Ingestion of honey is an identified risk factor of infant botulism. Because relative blood flow and innervations are greatest in the bulbar musculature, botulism manifest neurologically as a symmetric, descending, flaccid paralysis beginning with the cranial nerve musculature and progressing over hours to days.

138.(C) The classic triad of botulism is the acute onset of a symmetric flaccid descending paralysis with clear sensorium, no fever, and no paresthesias.

139.(A) EMG can sometimes distinguish between causes of acute flaccid paralysis. The distinctive EMG finding in botulism is facilitation (potentiation) of the evoked muscle action potential at high-frequency (50 Hz) stimulation. Nerve conduction velocity and sensory nerve function are normal in botulism.

140.(D) This class of antibiotics can potentiate the action of botulinum toxin at the neuromuscular junction.

141.(C) Wound botulism requires aggressive treatment with antibiotics and antitoxin in a manner analogous to that for tetanus and may require wound debridement to remove the source of the toxin.

142.(A) Other favorable prognostic factors are absence of fever, and localized disease. An unfavorable prognosis is also associated with onset of generalized tetanic spasms <3 days after onset of trismus. Cephalic tetanus has an especially poor prognosis because of breathing and feeding difficulties.

143.(C) Non-minor wounds require human TIG except those in a fully immunized patient (i.e., ≥3 doses of adsorbed tetanus toxoid). A tetanus toxoid booster (preferably Tdap) is administered to all persons with any wound if the tetanus immunization status is unknown or incomplete. A booster is administered to injured persons who have completed the primary immunization series if (1) the wound is clean and minor but ≥10 yr have passed since the last booster or (2) the wound is more serious and ≥5 yr have passed since the last booster.

144.(E) Clostridium difficile infection (CDI) is diagnosed by the detection of a C. difficile toxin in the stool by enzyme immunoassay (EIA) or nucleic acid amplification tests (NAATs). Risk factors for CDI include the use of broad-spectrum antibiotics, hospitalization, GI surgery, inflammatory bowel disease

(IBD), chemotherapy, enteral tube feeding, proton inhibitor (PPI) or H2 - receptor antagonist use, and chronic illness.

145.(D) Myonecrosis (Gas Gangrene) is a rapidly progressive anaerobic soft tissue infection, usually affects muscles compromised by surgery, trauma, or vascular insufficiency that become contaminated with C. perfringens spores.

146.(C) Ethambutol should generally be reserved for children old enough to have visual acuity and color discrimination reliably monitored.

147.(A)

148.(C) The hallmark of TBI is a positive TST or interferon-γ release assay (IGRA) result. In this stage the child has no signs or symptoms, a normal physical examination, and the chest radiograph is either normal or reveals only granuloma or calcifications in the lung parenchyma.

149.(C) Disseminated and meningeal tuberculosis are early manifestations, often occurring within 2-6 mo of acquisition. Significant lymph node or endobronchial tuberculosis usually appears within 3-9 mo. Lesions of the bones and joints take several years to develop, whereas renal lesions become evident decades after infection.

150.(D) Reactivation tuberculosis is the most likely diagnosis in this case. All other radiological findings can be seen with primary TB, military TB, and TB pleural effusions.

151.(B) The classic manifestation of tuberculous spondylitis is progression to Pott disease, in which destruction of the vertebral bodies leads to gibbus deformity and kyphosis.

152.(C) The amount of induration in response to the test should be measured by a trained person 48-72 hr after administration. Immediate hypersensitivity reactions to tuberculin or other constituents of the preparation are short-lived (<24 hr) and not considered a positive result. Older children and adults who receive a BCG vaccine are more likely to develop tuberculin reactivity, but most lose the reactivity by 5-10 yr after vaccination.

153.(B) In other groups TST are considered positive when induration is ≥ 10 mm.

154.(E) IGRA has superior specificity (95%) in BCG-immunized, low-risk children. Most experts support the use of IGRAs for the evaluation of TB infection in young children at low risk of infection, especially in those who have received a BCG vaccine.

155.(A) INH should be given to children <5 yr old who have a negative TST or IGRA result but who have a known recent exposure to an adult with potentially contagious TB disease. This practice is often referred to as window prophylaxis. By the time delayed hypersensitivity develops (2-3 months), an untreated child

already may have developed severe tuberculosis. For these children, TST or IGRA testing is repeated 8-10 wk after contact with the source case for tuberculosis has been broken. If the 2nd test result is positive, the child should complete a treatment course for TBI (either 9 mo of INH or one of the shorter treatment options). If the 2nd test result is negative, TBI treatment can be stopped.

156.(C) Because pulmonary tuberculosis is harmful to both the mother and the fetus and represents a great danger to the infant after delivery, tuberculosis in pregnant women always should be treated. The most common regimen for drug-susceptible tuberculosis is isoniazid, rifampin, and ethambutol. The aminoglycosides and ethionamide should be avoided because of their teratogenic effect. the treatment of pregnant women who have asymptomatic TB infection (not disease) is often deferred until after delivery.

157.(B) The preferred treatment of NTM lymphadenitis is complete surgical excision as surgery is more effective than antibiotic treatment. Nodes should be removed while still firm and encapsulated. If surgery of NTM lymphadenitis cannot be performed for some reason, or removal of infected tissue is incomplete, or recurrence or chronic drainage develops, a 3 mo trial of combination chemotherapy is warranted. M. avium complex bacteria are often resistant to rifampin or ethambutol alone. although spontaneous resolution can take several months, a wait-and-see approach can be chosen in selected patient which is not the case in this scenario as the lymph node is enlarging.

158.(D) Parenteral penicillin G is the only documented effective treatment for congenital syphilis, syphilis during pregnancy, and neurosyphilis. Aqueous crystalline penicillin G is preferred over procaine penicillin, because it better achieves and sustains the minimum concentration of 0.018 μg/mL (0.03 units/mL) needed for 7-10 days to achieve the prolonged treponemicidal levels required for the long dividing time of T. pallidum. In penicillin allergic patient, desensitization followed by standard penicillin therapy is the most reliable strategy.

159.(D) Louse-borne relapsing fever is caused by Borrelia recurrentis and is transmitted from person to person by Pediculus humanus, the human body louse. Human infection occurs as a result of crushing lice during scratching. Diagnosis depends on demonstration of spirochetes by darkfield microscopy or in thin or thick blood smears stained with Giemsa or Wright stain and by blood culture. During afebrile remissions, spirochetes are not found in the blood.

160.(C) Oral or parenteral tetracycline or doxycycline is the drug of choice for louse-borne and tick-borne relapsing fever. In children younger than 8 yr of age,

erythromycin (50 mg/kg/day divided every 6 hr PO) for a total of 10 days is recommended.

161.(B) The rash is generally either uniformly erythematous or a target lesion with central clearing. The rash gradually expands (hence the name migrans) to an average diameter of 15 cm and typically remains present for 1-2 weeks.

162.(D) All of the cranial nerves except the olfactory have been reported to be involved with Lyme disease, but the most common are VI and especially VII.

163.(C) The gradual onset in children with atypical pneumonia is in contrast to the sudden onset of lobar pneumonia. Coryza and gastrointestinal complaints are unusual and usually suggest a viral etiology. Examination may be unrevealing. Pneumonia is usually described as interstitial or bronchopneumonic, and involvement is most common in the lower lobes.

164.(A) Mycoplasmas lack a cell wall, so they are resistant to β-lactam agents that act by inhibiting the cell wall synthesis. Other drug classes, such as trimethoprim, rifampin, or linezolid are inactive against M. pneumoniae. Typically, M. pneumoniae is sensitive to macrolides, tetracyclines, and quinolones. Macrolides are preferred in children younger than 8 yr of age. Tetracyclines may be used for children older than 8 yr of age. Fluoroquinolones are effective and bactericidal but have higher minimum inhibitory concentrations (MIC) compared with macrolides and currently are not recommended as a first-line therapy in children.

165.(B) Nephrotoxicity, hypokalemia, and hypomagnesemia are common.

166.(D) Voriconazole is guideline-recommended as the preferred primary therapy against invasive aspergillosis. Antifungal resistance develops quickly to 5-FC monotherapy. Fluconazole is ineffective against Aspergillus and other molds. Itraconazole has the benefit of antifungal activity against Aspergillus species but has erratic oral absorption in high-risk patients and significant drug interactions, and it is no longer recommended for primary therapy of invasive aspergillosis. Amphotericin B was once the preferred treatment for most invasive fungal infections.

167.(D) Posaconazole is active against Zygomycetes such as mucormycosis, and voriconazole is not active against these particular mold infections. Isavuconazole is not as potent against Zygomycetes as posaconazole. Fluconazole plays an important role in the treatment of invasive candidiasis. Itraconazole has a role in treating less-serious infections with endemic mycoses (histoplasmosis, coccidioidomycosis, and blastomycosis).

168.(E) Caspofungin was approved for refractory aspergillosis or intolerance to other therapies and for candidemia and various other sites of invasive Candida infections.

169.(A) Central nervous system involvement is common and is most accurately described as meningoencephalitis. Candida infections involving the central nervous system often result in abscesses**.** Renal involvement commonly complicates neonatal invasive candidiasis. Renal involvement may be limited to candiduria or can manifest with diffuse infiltration of Candida throughout the renal parenchyma or the presence of Candida and debris within the collecting system.

170.(C) Hematologic parameters are sensitive but not specific. Thrombocytopenia occurs in more than 80% of premature infants with invasive candidiasis, but also occurs in 75% of premature infants with Gram-negative bacterial sepsis and nearly 50% of infants with Gram positive bacterial sepsis.

171.(C) Treatment of mild cases might not be necessary. When treatment is warranted, the most commonly prescribed antifungal agent is topical nystatin. In breastfed infants, simultaneous treatment of infant and mother with topical nystatin or oral fluconazole may be indicated.

172.(B) Candida diaper dermatitis is treated with topical antifungal therapy such as nystatin, clotrimazole, or miconazole. If significant inflammation is present, the addition of hydrocortisone 1% may be useful for the 1st 1-2 days, but topical corticosteroids should be used cautiously in infants because the relatively potent topical corticosteroid can lead to adverse effects.

173.(D) CNS disease is the most commonly recognized manifestation of cryptococcosis. Affected patients may develop intracerebral masses (cryptococcomas) and increased ICP. Pneumonia is the 2nd most commonly recognized form of cryptococcosis.

174.(C) Target sign, a nodule with lower central signal compared to the rim-enhancing periphery, is typically seen in MRI. Halo and air crescent signs are usually seen in CT scan.

175.(D) ELISA-based test for galactomannan, one of the components of the Aspergillus cell wall, is the molecular biomarker of choice for the diagnosis of IA in serum, BAL fluid, and CSF. It is the most sensitive test in detecting disease in cancer patients or hematopoietic stem cell transplant recipients. The beta-glucan assay, is a nonspecific fungal assay that detects the major component of the fungal cell wall but it does not discriminate which fungal infection is infecting the patient. Isolation of Aspergillus from blood cultures is uncommon,

likely because fungemia is low-level and intermittent. Obtaining tissue specimens is often impractical in critically ill, often thrombocytopenic, patients.

176.(C) Acute pulmonary histoplasmosis follows exposure to large inocula in closed spaces (e.g., chicken coops or caves) or prolonged exposure (e.g., camping on contaminated soil, chopping decayed wood). The median incubation time is 14 days.

177.(E) The presence of multiple nodules (≥10), pleural effusions, or the reversed halo sign are more suggestive of mucormycosis.

178.(A) Voriconazole is not active against mucormycosis. Posaconazole is active against many of the Mucorales but is presently not recommended for primary therapy; it may be used for salvage therapy or step down from an amphotericin B-based regimen. Echinocandins lack significant activity against the Mucorales.

179.(D) In cases of P. jirovecii pneumonia occurring in children with immunodeficiency due to immunosuppressive medications, the onset of hypoxia and symptoms is often more abrupt. Rales are usually not detected on physical examination. Ground glass appearance in both lungs is the usual radiographic finding.

180.(B) Other alternative prophylactic drugs are atovaquone and pentamidine, but all of these agents are inferior to TMP-SMX.

181.(C)

182.(D) Koplik spots represent the enanthem and are the pathognomonic sign of measles, appearing 1-4 days prior to the onset of the rash. They first appear as discrete red lesions with bluish white spots in the center on the inner aspects of the cheeks at the level of the premolars.

183.(B) Of the major symptoms of measles, the cough lasts the longest, often up to 10 days. With the onset of the rash, symptoms begin to subside. The rash fades over about 7 days in the same progression as it evolved, often leaving a fine desquamation of skin in its wake.

184.(D) Cerebrospinal fluid analysis in SSPE reveals normal cells but elevated IgG and IgM antibody titers in dilutions >1:8.

185.(B) The vaccine is effective in prevention or modification of measles if given within 72 hr of exposure. Immunoglobulin may be given up to 6 days after exposure to prevent or modify infection.

186.(A) Nerve deafness is the single most common finding among infants with CRS.

187.(E) Salt-and-pepper retinopathy is the most common ocular abnormality but have little early effect on vision. Unilateral or bilateral cataracts are the most serious eye finding, occurring in about a third of infants.

188.(B) Patent ductus arteriosus is the most frequently reported cardiac defect, followed by lesions of the pulmonary arteries and valvular disease.

189.(B) If the 1st rubella antibody test result is positive, the mother is likely immune. If all 3 specimens test are negative, infection has not occurred. A negative first specimen and a positive test result in either the 2nd and 3rd specimen indicate that seroconversion has occurred in the mother, suggesting recent infection.

190.(C) The routine use of immunoglobulin for susceptible pregnant women exposed to rubella is not recommended and is considered only if termination of pregnancy is not an option because of maternal preferences, understanding that prophylaxis may reduce the risk for clinically apparent infection but does not guarantee prevention of fetal infection.

191.(A) All other complications are less common. Sensorineural hearing loss is rare.

192.(B) In adolescent and adult males, orchitis is 2nd only to parotitis as a common finding in mumps.

193.(A) In Guillain-Barré syndrome , which is the most difficult to distinguish from poliomyelitis, the paralysis is characteristically symmetric, and sensory changes and pyramidal tract signs are common, contrasting with poliomyelitis. Fever, headache, and meningeal signs are less notable, and the CSF has few cells but elevated protein content.

194.(C) Hand-foot-and-mouth disease, one of the more distinctive rash syndromes, is most frequently caused by coxsackievirus A16.

195.(C) Acute hemorrhagic conjunctivitis, primarily caused by enterovirus D70 and coxsackievirus A24/A24 variant, is explosive and marked by high contagiousness, with spread mainly via eye-hand-fomite-eye transmission.

196.(D) The most common manifestation of Parvo virus B19 is erythema infectiosum (fifth disease). The initial stage is an erythematous facial flushing (slapped-cheek appearance). The rash spreads rapidly or concurrently to the trunk and proximal extremities as a diffuse macular erythema in the second stage. Central clearing of macular lesions occurs promptly, giving the rash a lacy, reticulated appearance.

197.(B) In contrast to children with erythema infectiosum only, patients with aplastic crisis are ill with fever, and have signs and symptoms of profound anemia. Rash is rarely present. The incubation period for transient aplastic crisis is shorter than that for erythema infectiosum because the crisis occurs coincident with the viremia.

198.(C) Older children experience burning, tingling, itching, or pain 3-6 hr (rarely as long as 24-48 hr) before the development of the herpes lesion.
199.(E) HSV is an acute necrotizing infection generally involving the frontal and/or temporal cortex and the limbic system. It is almost always caused by HSV-1 (beyond the neonatal period). Examination of the cerebrospinal fluid (CSF) typically shows a moderate number of mononuclear cells and polymorphonuclear leukocytes, a mildly elevated protein concentration, a normal or slightly decreased glucose concentration, and often a moderate number of erythrocytes. Patients should be promptly treated with intravenous acyclovir.
200.(C) Infants whose mothers demonstrate varicella in the period from 5 days prior to delivery to 2 days afterward are at high risk for severe varicella because the mother has not yet developed a significant antibody response. The infant's rash usually occurs toward the end of the 1st wk. Those infants should receive VZIG as soon as possible after birth. If VZIG is not available, intravenous immunoglobulin (IVIG) may provide some protection. Acyclovir should be used when lesions develop. Vaccine is given to healthy children (not neonates) within 3-5 days after exposure.
201.(E) The characteristic cutaneous lesion has been called a cicatrix, a zigzag scarring, in a dermatomal distribution (zoster-like distribution), often associated with atrophy of the affected limb.
202.(D) Generalized lymphadenopathy is present in 90% of cases; it occurs most commonly in the anterior and posterior cervical nodes and the submandibular lymph nodes and less commonly in the axillary and inguinal lymph nodes. Epitrochlear lymphadenopathy is particularly suggestive of infectious mononucleosis.
203.(C) EBV is associated with lymphoid malignancies, such as Burkitt lymphoma, Hodgkin lymphoma, aggressive NK cell leukemia, T- and NK cell lymphoproliferative disorder, and epithelial cell malignancies such as nasopharyngeal carcinoma and gastric carcinoma.
204.(E) Roseola infantum (exanthem subitum, or sixth disease) is an acute, self-limited disease of infancy and early childhood. It is characterized by the abrupt onset of high fever, which may be accompanied by fussiness. The fever usually resolves acutely after 72 hr (crisis) but may gradually fade over a day (lysis) coincident with the appearance of a faint pink or rose-colored, nonpruritic, 2-3 mm morbilliform rash on the trunk.
205.(A)

206.(B) Typically, the first sign of infection in infants is rhinorrhea. Cough may appear more often after an interval of 1-3 days, at which time there may also be sneezing and a low-grade fever. Soon after, the child begins to wheeze audibly.
207.(E) With C. trachomatis pneumonia, there may be a history of conjunctivitis, and the illness tends to be of subacute onset. Coughing and inspiratory crackles may be prominent; wheezing is not. Fever is usually absent.
208.(C) Conjunctivitis is a familiar illness associated with the human adenoviruses. Other common manifestations are upper and lower respiratory disease, pharyngitis, gastroenteritis. Hemorrhagic cystitis occurs more frequently in young males and typically resolves on its own in 1-2 wk. Myocarditis, hepatitis, or meningoencephalitis occurs less frequently.
209.(A) Rotavirus infection typically begins with mild to moderate fever as well as vomiting, followed by the onset of frequent, watery stools. Vomiting and fever typically abate during the second day of illness, but diarrhea often continues for 5-7 days. The stool is without gross blood or white blood cells. Treating dehydration is the main goal in the treatment of viral enteritis. A secondary goal is maintenance of the nutritional status of the patient. Antibiotics are of no benefit. Therapy with probiotic organisms such as Lactobacillus species has been shown to be helpful only in mild cases and not in dehydrating disease. Antiemetics may help alleviate vomiting but in this case it was stopped early in the course of disease.
210.(D) During the 10-day observation period, at the first sign of rabies in the biting dog, cat, or ferret, treatment of the exposed person with RIG (human) and vaccine should be initiated. The animal should be euthanized immediately and tested.
211.(C) Microscopy cannot differentiate between E. histolytica and E. dispar unless phagocytosed erythrocytes (specific for E. histolytica) are seen. The shape and size of trophozoites and cysts of both are identical and difficult to differentiate.
212.(B) Giardia is a particularly significant pathogen in children with malnutrition and certain immunodeficiencies (IgA deficiency, common variable immunodeficiency, X-linked hypogammaglobulinemia).
213.(E) In patients with visceral leishmaniasis, smears or cultures of material from splenic, bone marrow, or lymph node aspirations are usually diagnostic. In experienced hands, splenic aspiration has a higher diagnostic sensitivity. An immunochromatographic strip test using a recombinant antigen (K39) has a diagnostic sensitivity and specificity for VL of 80–90% and 95%, respectively.

214.(B) Incubation period is the longest with P. malariae species (18-40 days). Periodicity of paroxysms is less apparent with P. falciparum. Plasmodium knowlesi malaria is most often uncomplicated but can lead to severe malaria and death if high-density parasitemia is present. The most common serious complication is severe anemia.

215.(C) Nephrotic syndrome is a rare complication of P. malariae infection that is not observed with any other human malaria species. It is poorly responsive to corticosteroids.

216.(A) Primaquine is used to eradicate any hypnozoites that may remain dormant in the liver, and thus prevent relapses, in P. vivax and P. ovale infections. It is given once daily for 14 days. Testing for glucose-6-phosphate dehydrogenase deficiency must be performed before initiation of primaquine, because it can cause hemolytic anemia in such patients.

217.(D) In areas where chloroquine-resistant P. falciparum exists, atovaquone-proguanil is generally recommended for shorter trips (up to 2 wk) because it must be taken daily. For longer trips, mefloquine is preferred, since it is given only once a week, but it should not be given to children if they have a known hypersensitivity to mefloquine, are receiving cardiotropic drugs, have a history of convulsive or certain psychiatric disorders, or travel to an area where mefloquine resistance exists. Doxycycline is an alternative for children >8 yr old. Primaquine is a daily prophylaxis option for children who cannot tolerate any of the other options but it is contraindicated in children with G6PD deficiency. Sulfadoxine-pyrimethamine is given to infants.

218.(E) T. gondii is estimated to cause 35% of cases of chorioretinitis.

219.(C) Calcifications occur throughout the brain, but there is a propensity for development of calcifications in the caudate nucleus and basal ganglia, choroid plexus, and subependyma.

220.(D) Piperazine citrate causes neuromuscular paralysis of the parasite and rapid expulsion of the worms; it is the treatment of choice for intestinal or biliary obstruction and is administered as syrup through a nasogastric tube.

221.(D) Chronically infected children with moderate and heavy hookworm infections suffer from intestinal blood loss that results in iron deficiency and can lead to anemia as well as protein malnutrition.

222.(E)

223.(B)

224.(A)

225.(C) The absence of fever and wheezing helps to distinguish C. trachomatis pneumonia from respiratory syncytial virus pneumonia.

Chapter 17

The Digestive System Questions

KHALID ALAARJI

1. What is the **MOST** common cause of premature loss of primary teeth?
   A. Malpositioned teeth
   B. Supernumerary teeth
   C. Hypothyroidism
   D. Cleidocranial dysplasia
   E. Premature eruption of the permanent teeth

2. What is the expected time of eruption of first mandibular permanent molar teeth?
   A. 4-5 yr
   B. 6-7 yr
   C. 8-9 yr
   D. 10-11 yr
   E. 12-13 yr

3. Which of following is a recognized cause of severe brownish discoloration of primary teeth?
   A. Hyperbilirubinemia
   B. Tetracycline
   C. Osteogenesis imperfecta
   D. Porphyria
   E. Fluoride

4. A healthy full-term boy found to have cleft palate. Your advice about the preferred time of surgical closure is before age of
   A. 6 months
   B. 12 months
   C. 18 months
   D. 24 months
   E. 30 months

5. Which of the following is the **MOST** cariogenic sugar associated with dental caries?

A. Sucrose
B. Glucose
C. Fructose
D. Lactose
E. Galactose

6. A 4-year-old child presents to out-patient clinic with recurrent painful oral lesions. Examination shows a well-circumscribed ulcerative lesion in buccal mucosa with a white base surrounded by a red halo.
Of the following, the **BEST** palliative therapy is topical application of

A. nystatin
B. steroid
C. acyclovir
D. tetracycline
E. gention violent

7. A 3-year-old child presents with history of fever, malaise, and difficult oral intake. Examination shows cervical lymphadenopathy with clusters of erythematous small vesicles involving oral mucosa and perioral skin.
Of the following, The **MOST** beneficial therapy is

A. topical steroid
B. oral acyclovir
C. oral nyatatin
D. oral co-amoxoclav
E. topical lidocaine

8. What is the **BEST** position of plain radiograph for a newborn with suspected tracheoesophageal fistula?

A. Prone
B. Supine
C. Left lateral
D. Right lateral
E. Semi-sitting

9. A 24-hour-term newborn presents with respiratory distress, cyanosis, and frothy secretion from the mouth, O2 saturation 83% at room air, plain abdomen radiograph show airless abdomen; you are suspecting esophageal atresia (EA).
Of the following, the **MOST** likely type is

A. H type
B. EA with fistula connected to the distal and proximal ends
C. pure EA
D. blind proximal end and fistula connected to the distal end
E. blind distal end and fistula connected to the proximal end

10. A 3-year-old boy presents with low grade fever and large volume bloody diarrhea, he ate a sandwich of hamburger before 4 days.
Of the following, the **MOST** causative pathogen is

A. Salmonella
B. Shigella
C. Campylobacter
D. Antamoeba histolytica
E. Enterohemorrhagic E. coli

11. A 9-year-old boy presents with regurgitation, difficulty in swallowing and weight loss; barium fluoroscopy show bird's beak sign
Of the following, the **MOST** likely diagnosis is

A. repaired oesophegeal fistula
B. GERD
C. acquired pyloric stenosis
D. eosinophilic esophigitis
E. achalasia

12. A 10-year-old boy, a known case of peanut allergy, presents with history of recurrent attacks of difficulty in swallowing, burning sensation in the chest with epigastric pain; investigations reveals high serum IgE. Endoscopy shows food impactions and biopsy send for histological study.
Of the following, the **MOST** likely diagnosis is

A. gastroesophageal reflux disease
B. eosinophilic esophagitis
C. hiatal hernia
D. H-type esophageal atresia
E. achalasia

13. A 3-year-old boy brought to the emergency unit after 12 hour of ingestion of drain cleaner, his mother gave him a lot of milk and water but with difficulty because of pain. Examination shows excessive drooling and oral burn.
Of the following, the **BEST** next step in the management is to

A. observe in hospital
B. start steroid
C. discharge and follow up after 1 week
D. do barium swallow
E. arrange for upper endoscopy

14. A 14-month-old child presents to emergency unit with history of choking, coughing, excessive salivation, and refusal to drink. The AP view of chest radiograph shows a coin facing you and the lateral view shows the edge of coin.
Of the following, the **MOST** likely place of coin impaction is

A. larynx
B. trachea
C. esophagus
D. carina
E. right bronchus

15. A 5-year-old boy presents with sudden onset of severe nausea, vomiting, and abdominal cramps within 3 hours of ingestion of unrefrigerated meats and potato, followed by watery diarrhea.
Of the following, the **MOST** likely causative pathogen is

A. Salmonella
B. Shigella
C. Staphylococcus aureus
D. Campylobacter
E. Shiga toxin-producing E. coli

16. Accidentally esophageal ingestion of button batteries, must be emergently removed within

A. 2 hr
B. 4 hr
C. 6 hr
D. 8 hr
E. 12 hr

17. A 9-month-old infant evaluated for failure to thrive; history reveals frequent vomiting and arching his back during feeding with recurrent hospital visits for wheezing and shortness of breath.
Of the following, the **MOST** likely diagnosis is

A. intuscessption
B. gastroesophageal reflux disease
C. asthma
D. H-type esophageal fistula
E. achalasia

18. Which of the following has the highest risk of pyloric stenosis if given in neonatal period?

A. Cefexime
B. Erythromycin
C. Ampicillin
D. Co-trimoxazole
E. Co-amoxoclav

19. Asymptomatic high risk child for celiac disease (CD) who has positive serologic screening tests and severe mucosal damage consistent with CD is regarded as

A. celiac disease
B. silent celiac disease
C. latent celiac disease
D. potential celiac disease
E. non-celiac gluten sensitivity

20. A 5-year-old child under evaluation for short stature; examination shows pallor, abnormal teething, and wasting of thigh and buttock; CBC reveals (Hb 7gm/dl), serum ferritin (9 mg/dl) and celiac screen reveals anti-TG2 (90 U/mL) but EMA and HLA DQ8/DQ2 not available.
Of the following, the **MOST** appropriate step of management is to

A. search for other diagnosis
B. continue normal diet with follow up serological testing
C. start gluten-free diet
D. do duodenal biopsies
E. regard as non-celiac gluten sensitivity

21. Asymptomatic child of first-degree relative celiac disease patient with positive HLA DQ2 and DQ8 and TG2 Titer (350 U/mL).
Of the following, the **MOST** appropriate step of management is to

A. continue normal diet with further serological testing
B. continue normal diet till became symptomatic
C. send for EMA
D. do esophagogastroduodenoscopy and biopsies
E. start gluten-free diet

22. Which of the following is the **MOST** common extraintestinal manifestation of celiac disease?

A. Iron-deficiency anemia
B. Short stature
C. Enamel hypoplasia
D. Aphthous stomatitis
E. Peripheral neuropathies

23. Which of the following diets should be avoided in child with celiac disease?

A. Soybean
B. Rye
C. Corn
D. Buckwheat
E. Maize

24. What is **MOST** likely causative pathogen of diarrhea after consumption of undercooked eggs?

A. Shiga toxin–producing E. coli
B. Bacillus cereus
C. Staphylococcus aureus
D. Listeria
E. Salmonella

25. The Codex Alimentarius Guidelines define gluten-free food, are those containing gluten less than

A. 20 ppm
B. 30 ppm
C. 40 ppm
D. 50 ppm

E. 60 ppm

26. According to the National Institute for Health and Care Excellence Guidelines, for which of the following conditions celiac testing is recommended?

A. Reduced bone mineral density
B. Unexplained iron deficiency
C. Dental enamel defects
D. Down syndrome
E. Unexplained peripheral neuropathy

27. A 14-year-old child under evaluation for melena; gave history of 2 months of intermittent epigastric dull pain, sometimes awaking him at night.
Of the following, the **MOST** appropriate test for diagnosis is

A. urea breath test
B. stool antigen test for H. pylori
C. upper endoscopy
D. serology for H. pylori
E. abdominal ultrasound

28. What is the **BEST** time to repeat stool test for H. pylori antigen after complete eradication therapy?

A. Immediately after stopping the therapy
B. 1 wk after stopping therapy
C. 2 wk after stopping therapy
D. 4 wk after stopping therapy
E. No need for the test

29. A 13-year-old boy presents with bloody diarrhea, tenesmus, and cramping abdominal pain that interfere with night sleep for the last 2 month, also he had a painful leg ulcer diagnosed as pyoderma gangrenosum.; examination shows pallor, soft abdomen with no palpable mass, and normal anal examination.
Of the following, the **MOST** likely diagnosis is

A. infectious colitis
B. ulcerative colitis
C. Crohn's disease
D. Henoch-Schönlein purpura
E. Behçet disease

30. A 15-year-old boy under evaluation for growth failure and delayed sexual development. Past history reveals recurrent attacks of abdominal pain, diarrhea, mouth ulcer, malaise, and weight loss; examination shows pallor, height and weight below 3rd centile, small testicular size, digital clubbing, and purulent drainage from anus.
Of the following, the **MOST** likely diagnostic tool is

A. sweat test
B. growth hormone assay
C. colonoscopy with biopsy
D. sex hormone study
E. TTG Abs

31. Which of the following is the **MOST** sensitive and specific marker for diagnosis of inflammatory bowel disease?

A. α1-Antitripsin
B. Calprotectin
C. Steatocrit
D. Elastase
E. Chymotrypsin

32. A 15-year-old girl who's known case of ulcerative colitis presents with high grade fever and hematochezia for last 7 days; examination shows pale, thin girl with leg edema. Investigations reveal; Hb (8gm/dl), WBC count (23000/mm^3), platelate count (550000/mm^3), and serum albumin (2.5gm/dl).
Of the following, the first line of treatment is

A. probiotics
B. oral sulfasalazine
C. oral azathioprine
D. IV corticosteroid
E. colectomy

33. Which of the following extraintestinal manifestations tend to occur **MORE** commonly with ulcerative colitis than with Crohn disease?

A. Oral aphthous ulcers
B. Chronic active hepatitis
C. Peripheral arthritis
D. Erythema nodosum
E. Digital clubbing

34. Which of the following is used as a conservative management in pyloric stenosis in patients who are not good surgical candidates?

A. Ondansetron
B. Atropine sulfate
C. Hyoscine bromide
D. Domperidone
E. Omeprazole

35. A 12-month-old infant presents to emergency department with recurrent attacks of bilious vomiting and colicky abdominal pain.
Of the following, the imaging test of choice is

A. CT abdomen
B. abdominal ultrasound
C. abdominal plain film
D. upper gastrointestinal series
E. barium enema

36. The hallmark of duodenal atresia is bilious vomiting and

A. history of polyhydramnios
B. Jaundice
C. no abdominal distention
D. visible peristalsis
E. bile stained amniotic fluid

37. Which of the following should be avoided in the child with acute diarrhea?

A. Yogurt
B. Wheat
C. Rice
D. Fruit juice
E. Potatoes

38. An 18-month-old female presents with recurrent attacks of painless rectal bleeding; she had previous history of intussusception at age of 9 month treated by pneumatic reduction; clotting profile and blood film are normal apart from mild anemia.
Of the following, the **MOST** likely diagnosis is

A. Meckel diverticulum
B. intussusception

C. amoebic dysentery
D. fissure in ano
E. rectal polyp

39. What is the typical abdominal radiograph finding of meconium ileus?
A. Gasless abdomen
B. Double-bubble
C. Multiple air–fluid levels
D. Large, dilated stomach
E. Ground glass appearance

40. Malrotation is incomplete rotation of the intestine during fetal development, which of the following is the **MOST** commonly affected part?
A. Duodenum
B. Jejunum
C. Ileum
D. Colon
E. Cecum

41. Which of the following metabolic abnormalities may cause paralytic ileus?
A. Hyponatremia
B. Hypocalcemia
C. Hypomagnesemia
D. Uremia
E. Alkalosis

42. Which of the following chemotherapy drugs may cause paralytic ileus?
A. Vincristine
B. Methotrexate
C. 6-mercaptopurine
D. Cisplatin
E. Cyclophosphamide

43. An 18-month-old child under evaluatation for abdominal distention, failure to thrive, and constipation starting in infancy with a poor response to medical management. Rectal examination shows empty rectum with explosive passage of stool.
Of the following, the **MOST** appropriate diagnostic tool is

A. abdominal ultrasound
B. plain radiograph of abdomen
C. anorectal manometry
D. unprepared contrast enema
E. CT of the abdomen

44. Constipation is defined as a delay or difficulty in defecation that causes distress to the patient with duration for more than
A. 2 weeks
B. 4 weeks
C. 6 weeks
D. 8 weeks
E. 12 weeks

45. A mother of a 10-month-old infant, who recently underwent hydrostatic reduction of intussusception, asks you if there is a risk of recurrence. You informed them that the recurrence rate is approximately 10% and mostly within
A. 3 days of reduction
B. 7 days of reduction
C. 14 days of reduction
D. 21 days of reduction
E. 28 days of reduction

46. A 16-month-old child presents with lethargy, repeated vomiting, and recurrent inconsolable screaming attacks with leg drawing to the abdomen.
Of the following, the **MOST** appropriate initial diagnostic tool is
A. plain abdominal radiograph
B. upper endoscope
C. CT of abdomen
D. ultrasound of abdomen
E. barium enema

47. A 3-year-old child received treatment with clindamycin for osteomyelitis, presented with severe watery diarrhea containing mucus, fever, and crampy abdominal pain.
Of the following, the **FIRST** line therapy is
A. IV metronidazole
B. oral fidaxomicin

C. probiotic
D. oral vancomycin
E. IV bezlotoxumab

48. Which of the following antiemetic medications is more suitable to be used in children?
A. Prochlorperazine
B. Cyclizine
C. Metoclopramide
D. Domperidone
E. Ondansetron

49. A 4-year-old child presents to emergency unit with seizure, high grade fever, and bloody diarrhea.
What is the **FIRST** line antibiotics therapy?
A. Ciprofloxacin
B. Cefixime
C. Trimethoprim-sulfamethoxazole
D. Metronidazole
E. Ampicillin

50. Which of the following enteropathogens is infectious in small inoculums?
A. Cholera
B. Shigella
C. Nontyphoidal Salmonella
D. Enterotoxogenic E. coli
E. Campylobacter

51. What is the **MOST** common viral cause of acute gastroenteritis in children?
A. Rotavirus
B. Norovirus
C. Sapovirus
D. Adenovirus
E. Astrovirus

52. A 6-year-old boy presents with sudden onset of severe nausea and vomiting within 2 hours of ingestion of improperly refrigerated cooked rice, followed by watery diarrhea.

Of the following, the **MOST** causative pathogen is

A. Salmonella
B. Shigella
C. Campylobacter
D. Bacillus cereus
E. Shiga toxin-producing E. coli

53. Which of the following inherited colorectal cancer syndromes has a **HIGHER** risk of colon cancer?

A. Juvenile polyposis syndrome
B. Peutz-Jeghers syndrome
C. Familial adenomatous polyposis
D. Lynch syndrome
E. Cowden syndrome

54. A 6-year-old boy is under evaluation of recurrent attacks of painless rectal bleeding mainly after defecation with mild abdominal pain. Examination shows dark brown macular lesions around the lips and oral mucosa; endoscopy reveals multiple polyps primarily found in jejunum and ileum.
Of the following, the **MOST** likely diagnosis is

A. Juvenile polyposis syndrome
B. Peutz-Jeghers syndrome
C. Familial adenomatous polyposis
D. Gardner syndrome
E. Cowden syndrome

55. A 10-year-old boy with hepatic failure presents to emergency unit with stuporous but arousable, confused, and incoherent speech. O/E asterixis, hyperreflexia, extensor reflex, and rigidity. EEG shows markedly abnormal triphasic waves.
Of the following, the **MOST** likely stage of hepatic encephalopathy is

A. I
B. II
C. III
D. IVa
E. IVb

56. Which of the following conditions can be associated with the hypoplasia or absence of the gallbladder?

A. Cystic fibrosis
B. Sickle cell anemia
C. Gilbert disease
D. Kawasaki disease
E. Henoch-Schönlein purpura

57. Persistent diarrhea is defined when the diarrhea lasting more than

A. 2 wk
B. 4 wk
C. 6 wk
D. 8 wk
E. 12 wk

58. Which of the following is characteristic of toddler's diarrhea?

A. Painless passage of stool
B. Failure to thrive
C. Perturbed appearance
D. 2 or more wk duration
E. Nighttime defecation is usually present

59. Which of the following pathogens is the **MOST** common cause of chronic infectious diarrhea in developing countries?

A. Shigella
B. Rotavirus
C. Enteroadherent E. coli
D. Campylobacter
E. Salmonella

60. Which of the following is **ONE** of diagnostic criteria for abdominal migraine?

A. Usually less than 1 hr in duration
B. Mainly epigastric
C. May be associated with photophobia
D. Episodes are separated by days
E. Pain does not interfere with normal activities

61. A 7-year-old girl presents with sudden onset of repeated non-bilious vomiting, pallor, and photophobia for the last 48 hr with no significant abnormality in review of other systems; she has had 5 same attacks in the last 6 months and she is completely normal between attacks; her mother complain from migraine.
Which of the following is the **BEST** abortive medication of this condition?

A. Ondansetron
B. Triptans
C. Lorazepam
D. Diphenhydramine
E. Chlorpromazine

62. Which of the following radiographic positions is the **BEST** to determine the type of imperforated anus?

A. Upside-down x-ray
B. Prone cross table lateral plain x-ray
C. Erect x-ray
D. Lateral decubitus x-ray
E. Supine plain x-ray

63. Which of the following are the **MOST** commonly associated malformations with imperforated anus?

A. Tetralogy of Fallot
B. Vesicoureteric reflux
C. Tethered cord
D. Tracheoesophageal fistula
E. Spina bifida

64. What is the liver span of a 12-year-old girl?

A. 4.0-4.5 cm
B. 5.0-5.5 cm
C. 6.0-6.5 cm
D. 7.0-7.5 cm
E. 8.0-8.5 cm

65. Which of the following is **TRUE** regarding pruritus of chronic liver disease?

A. Common in trunk
B. Usually worse at morning

C. Exacerbated with stress
D. Unrelated to the temperature
E. Unrelated to the degree of hyperbilirubinemia

66. What is the **MAIN** area of spider angiomas distribution?
A. Face
B. Abdomen
C. Back
D. Thigh
E. Hand

67. Which of the following is the **MOST** valuable procedure in the evaluation of neonatal hepatobiliary diseases?
A. Abdominal ultrasound
B. Hepatobiliary scintigraphy with technetium-labeled iminodiacetic acid derivatives
C. Endoscopic retrograde cholangiopancreatography
D. Magnetic resonance cholangiopancreatography (MRCP)
E. Liver biopsy

68. A 2-year-old child under evaluation of failure to thrive, recurrent episodes of cholestasis with elevation of serum aminotransferase, alkaline phosphatase, and bile acid levels associated with lymphedema of the lower limbs; between these episodes, the patient is asymptomatic with normal biochemical indices.
Of the following, the **MOST** likely diagnosis is
A. Zellweger syndrome
B. Alagille syndrome
C. Neonatal hemochromatosis
D. Aagenaes syndrome
E. Byler disease

69. A first cousin parents brought their 2-month-old infant because of prolonged jaundice, generalized weakness. Examination reveals high forehead, flattened face, upslanting palpebral fissures, and epicanthal folds; with hypotonia, cataracts, and hepatomegaly; LFT shows conjugated hyperbilirubinemia; abdominal ultrasound reveals renal cortical cysts and radiograph shows stippled calcifications of the patella and greater trochanter.
Of the following, the **MOST** likely diagnosis is

A. Zellweger syndrome
B. Aagenaes syndrome
C. Byler disease
D. neonatal hemochromatosis
E. familial hypercholanemia

70. A 3-week-old infant presents with prolonged neonatal jaundice and recurrent attacks of hypoglycemia; O/E hepatomegaly with no dysmorphic features. Laboratory finding shows blood sugar (42 mg/dL). TSB (18 mg/dL with 13 mg/dL is direct), ALT (140 U/L, AST (122 U/L), serum albumin (2gm/L), serum ferritin (650 mg/dL), PT (28), PTT (58), and INR (3.5). Family history of 2 babies died with same presentation at the neonatal period.
Of the following, the **BEST** method to confirm diagnosis is

A. ultrasonography
B. neonatal screening test
C. buccal mucosal biopsy
D. liver biopsy
E. α1-Antitrypsin phenotype

71. A 6-week-old infant under evaluation of cholestasis. Examination reveals triangular face; broad forehead, deep-set eyes, prominent ears, straight nose with bulbous tip, and pointed chin. Echo study shows peripheral pulmonary artery stenosis. Radiograph of the spine shows "Butterfly" vertebrae. Lab findings; TSB (13 mg/dL with 10 mg/dL is direct) and cholesterol level (760 mg/dL).
Of the following, the **MOST** likely diagnosis is

A. Zellweger syndrome
B. Alagille syndrome
C. Neonatal hemochromatosis
D. Aagenaes syndrome
E. Byler disease

72. In biliary atresia, the success rate for Kasai operation is much higher (90%) if performed before

A. 4 wk of life
B. 6 wk of life
C. 8 wk of life
D. 12 wk of life

E. 16 wk of life

73. A 3-year-old child with chronic cholestasis and poor compliance for supportive measures presents with blurred vision and ataxia. Examination shows decreased vibratory sensation and diminished peripheral reflexes.
Of the following, the **MOST** likely cause for this condition is

A. hepatic encephalopathy
B. Friedreich's ataxia
C. vitamine E deficiency
D. zinc deficiency
E. vitamine A deficiency

74. Which of the following is the **MOST** common presenting manifestation of Wilson disease in children?

A. Jaundice
B. Kayser-Fleischer ring
C. Intention tremor
D. Hemolytic anemia
E. Depression

75. A 6-year-old boy presents to emergency unit with severe epistaxis and abdominal distension for 2 days. There was no history of trauma or bleeding from other site and normal bowel motion. Two previous siblings had died at 6 and 8 years of age with jaundice and abdominal distension of unknown cause. O/E stuporous, severe icterus, ascitis, hepatomegaly 3 cm, and splenomegaly 3 cm. Lab finding reveals Hb 10.8 gm/dl, WBC and platelate counts within normal limits, TSB 18.0 mg/dl mainly direct, SGOT 112 IU/L, SGPT 22 IU/L, ALP 28.4 IU/L, PT 1 min, PTT 2 min, INR 3, and hepatitis viral study was negative.
Of the following, the **MOST** appropriate initial test is

A. serum ceruloplasmin level
B. serum free copper level
C. urinary copper excretion
D. liver MRI
E. liver biopsy

76. Which of the following is the method of choice for diagnosis of Wilson disease?

A. Decreased serum ceruloplasmin level

B. Increased serum free copper level
C. Increased 24hr-urinary copper excretion
D. Demonstration of kayser-fleischer rings
E. Measurement of hepatic parenchymal copper concentration

77. Which of the following is the **MOST** appropriate screening test for asymptomatic first-degree relatives of patients with Wilson disease?
A. Liver function tests
B. Serum ceruloplasmin level
C. Serum copper level
D. Liver biopsy
E. Genetic study

78. Which of the following diets should be **AVOIDED** in patients with Wilson disease?
A. Rice
B. Chocolate
C. Tea
D. Eggs
E. Fresh tomatoes

79. Which of the following agents is **MOST** likely used as adjuvant therapy in patients with Wilson disease?
A. Vitamin E
B. L-carnitine
C. Zinc
D. Medium-chain triglyceride
E. Folic acid

80. Which of the following therapies is **MOST** likely improving the outcome of patients with neonatal hemochromatosis?
A. Intensive phototherapy
B. Exchange transfusion
C. Disferal
D. Vitamine E
E. L-carnitine

81. With recovery of acute viral hepatitis A infection, the liver morphology returns to normal within

A. 3 mo of the acute infection
B. 4 mo of the acute infection
C. 6 mo of the acute infection
D. 9 mo of the acute infection
E. 12 mo of the acute infection

82. A concerned mother asks you about the time of return of her 7-year-old child with acute hepatitis A viral infection to school, the **BEST** time is approximately

A. 3 days after the onset of jaundice
B. 7 days after the onset of jaundice
C. 10 days after the onset of jaundice
D. 14 days after the onset of jaundice
E. 21 days after the onset of jaundice

83. Which of the following is a marker of infectivity?

A. HBsAg
B. Anti-HBc
C. Anti-HBs
D. HBeAg
E. anti-HBe

84. The serological autoantibody profile defines 2 main types of autoimmune hepatitis (AIH); type 1 and 2. Which of the following autoantibodies is positive in AIH type 2?

A. Anti-nuclear antibodies (ANA)
B. Anti–smooth muscle antibody (SMA)
C. Antineutrophil cytoplasmic antibodies
D. Anti–liver kidney microsomal type 1 antibody (anti-LKM-1)
E. Antiactin antibody

85. A 14-year-old girl under evaluation of insidious onset of fatigue, anorexia, malaise, joints pain, rash with secondary amenorrhea and behavioral changes for the last 4 months. Examination shows jaundice, palmar erythema, slightly enlarged tender liver with enlarged spleen. Serum aminotransferase 150 IU/L, TSB 3mg/dL, normal ALP and GGT, elevated serum γ-globulin levels, PT is

prolonged, Serum IgG levels >16 g/L, anti-nuclear antibodies (ANA) and anti–smooth muscle antibody (SMA) are positive with negative hepatitis viral screen. Of the following, the **FIRST** line of treatment is

A. Prednisone
B. Azathioprine
C. D-pencillamin
D. Budesonide
E. Cyclosporine

86. At least 50% of patients with hepatic failure experience serious infection. Which of the following pathogens is the **MOST** likely cause?

A. Staphylococcus aureus
B. E. coli
C. Pseudomonas aeroginosa
D. Streptococcus pneumonia
E. Salmonella

87. Liver failure in the perinatal period can be associated with prenatal liver injury and even cirrhosis. Which of the following conditions is **MOST** common cause of acute liver failure in the neonate?

A. Gestational alloimmune liver disease
B. Galactosemia
C. Tyrosinemia
D. Familial hemophagocytic lymphohistiocytosis
E. Congenital herpes simplex infection

88. Which of the following electrolytes when decreased is a reflection of liver regeneration?

A. Sodium
B. Potassium
C. Magnesium
D. Phosphorus
E. Calcium

Chapter 17

The Digestive System Answers

KHALID ALAARJI

1.(E) While the delayed eruption of the 20 primary teeth can be familial or indicate systemic or nutritional disturbances such as hypopituitarism, hypothyroidism, cleidocranial dysplasia, trisomy 21, and multiple other syndromes. Failure of eruption of single or small groups of teeth can arise from local causes such as malpositioned teeth, supernumerary teeth, cysts, or retained primary teeth.

2.(B)

Permanent **Dentition Eruption**			
Maxillary		**Mandibular**	
Central incisor	7-8 yr	Central incisor	6-7 yr
Lateral incisor	8-9 yr	Lateral incisor	7-8 yr
Canine	11-12 yr	Canine	9-10 yr
First premolar	10-11 yr	First premolar	10-12 yr
Second premolar	10-12 yr	Second premolar	11-12 yr
First molar	6-7 yr	First molar	6-7 yr
Second molar	12-13 yr	Second molar	11-13 yr
Third molar	17-21 yr	Third molar	17-21 yr

3.(E) Anomalies of Color: Discolored teeth can result from incorporation of foreign substances into developing enamel. Neonatal **hyperbilirubinemia** can produce blue to black discoloration of the primary teeth. **Porphyria** produces a red-brown discoloration. **Tetracyclines** are extensively incorporated into bones and teeth and, if administered during the period of formation of enamel, can result in brown-yellow discoloration and hypoplasia of the enamel. Such teeth fluoresce under ultraviolet light. The period at risk extends from approximately 4 mo of gestation to 7 yr of life. Repeated or prolonged therapy with tetracycline carries the highest risk. Excessive **fluoride** during enamel formation affects ameloblastic function, resulting in inconspicuous white, lacy patches on the enamel to severe brownish discoloration and hypoplasia.

4.(B) Surgical closure of a cleft lip is usually performed by 3 mo of age, when the infant has shown satisfactory weight gain and is free of any oral, respiratory, or

systemic infection. In an otherwise healthy child, closure of the palate is usually done before 1 yr of age to enhance normal speech development.

5.(A) Sucrose is the most cariogenic sugar because one of its by-products during bacterial metabolism is glucan, a polymer that enables bacteria to adhere more readily to tooth structures. Dietary behaviors, such as consuming sweetened beverages in a nursing bottle or frequently consuming sticky candies, increase the cariogenic potential of foods because of the long retention of sugar in the mouth.

6.(B) The aphthous ulcer lesions generally last 10-14 days and heal without scarring. Nonprescription palliative therapies, such as benzocaine and topical lidocaine, are effective, as are topical steroids.

7.(B) The symptoms of herpetic gingivostomatitis (herpes simplex virus) usually regress within 2 wk without scarring. Fluids should be encouraged because the child may become dehydrated. Analgesics and anesthetic rinses can make the child more comfortable. Oral acyclovir, if taken within the first 3 days of symptoms in immunocompetent patients, is beneficial in shortening the duration of symptoms. Caution should be exercised to prevent autoinoculation, especially of the eyes.

8.(A) Prone positioning minimizes movement of gastric secretions into a distal fistula, and esophageal suctioning minimizes aspiration from a blind pouch.

9.(C) Plain radiography in the evaluation of respiratory distress might reveal a coiled feeding tube in the esophageal pouch and/or an air-distended stomach, indicating the presence of a coexisting TEF. Conversely, pure EA can manifest as an airless scaphoid abdomen. In isolated TEF (H type), an esophagogram with contrast medium injected under pressure can demonstrate the defect.

10.(E) Enterohemorrhagic Escherichia coli (EHEC) including E. coliO157:H7 and other Shiga toxin–producing E. coli (STEC). Incubation period; 1-9 days (usually 3-4 days). Clinical presentations are watery diarrhea that becomes bloody in 1-4 days in ~40% of infections; in contrast to dysentery, bloody stools are large volume and fever/toxicity are minimal. More common in children <4 yr old. Risk factors are food and water contaminated with feces from ruminants; infected people and animals (fecal-oral); predominantly high-resource countries; undercooked beef especially hamburger, unpasteurized milk and juice, raw fruits and petting zoos, recreational swimming, daycare. Antimotility agents and antibiotics increase risk of hemolytic uremic syndrome.

11.(E) Achalasia manifests with regurgitation and dysphagia for solids and liquids and may be accompanied by undernutrition or chronic cough; retained esophageal food can produce esophagitis. The mean age in children is 8.8 yr,

with a mean duration of symptoms before diagnosis of 23 mo; it is uncommon before school age. Chest radiograph shows an air–fluid level in a dilated esophagus. Barium fluoroscopy reveals a smooth tapering of the lower esophagus leading to the closed LES, resembling a bird's beak. Loss of primary peristalsis in the distal esophagus with retained food and poor emptying are often present. Manometry is the most sensitive diagnostic test and helps differentiate the three types of achalasia.

12.(B) The diagnosis of eosinophilic esophagitis should be considered in the clinical presentation of esophageal dysfunction, associated with esophageal epithelial infiltration of at least 15 eosinophils per high power field. Many patients have other atopic diseases (or a positive family history) and associated food allergies; laboratory abnormalities can include peripheral eosinophilia and elevated immunoglobulin E (IgE) levels.

13.(E) Twenty percent of patients develop esophageal strictures. Absence of oropharyngeal lesions does not exclude the possibility of significant esophagogastric injury, which can lead to perforation or stricture. The absence of symptoms is usually associated with no or minimal lesions; hematemesis, respiratory distress, or presence of at least 3 symptoms predicts severe lesions. An upper endoscopy is recommended as the most efficient means of rapid identification of tissue damage and must be undertaken in all symptomatic children. Dilution by water or milk is recommended as acute treatment, but neutralization, induced emesis, and gastric lavage are contraindicated.

14.(C) Most esophageal foreign bodies lodge at the level of the cricopharyngeus (upper esophageal sphincter), the aortic arch, or just superior to the diaphragm at the gastroesophageal junction (lower esophageal sphincter). Evaluation of the child with a history of foreign body ingestion starts with plain anteroposterior radiographs of the neck, chest, and abdomen, along with lateral views of the neck and chest. The flat surface of a coin in the esophagus is seen on the anteroposterior view and the edge on the lateral view. The reverse is true for coins lodged in the trachea; here, the edge is seen anteroposteriorly and the flat side is seen laterally.

15.(C) Etiology; Staphylococcus aureus (preformed enterotoxin). Incubation period; 1-6 hr. Clinical presentations are sudden onset of severe nausea and vomiting and abdominal cramps; diarrhea and fever may be present. Risk factors; Unrefrigerated or improperly refrigerated meats, potato and egg salads, cream pastries.

16.(A) Sharp objects in the esophagus, multiple magnets or single magnet with a metallic object, or foreign bodies associated with respiratory symptoms

mandate urgent removal within 12 hr of presentation. Button batteries, in particular, must be emergently removed within 2 hr of presentation regardless of the timing of patient's last oral intake because they can induce mucosal injury in as little as 1 hr of contact time and involve all esophageal layers within 4 hr.

17.(B) Most of the common clinical manifestations of esophageal disease can signify the presence of GERD and are generally thought to be mediated by the pathogenesis involving acid GER. Infantile reflux manifests more often with regurgitation (especially postprandially), signs of esophagitis (irritability, arching, choking, gagging, feeding aversion), and resulting failure to thrive; symptoms resolve spontaneously in the majority of infants by 12-24 mo. Older children can have regurgitation during the preschool years; this complaint diminishes somewhat as children age, and complaints of abdominal and chest pain supervene in later childhood and adolescence. Occasional children present with food refusal or neck contortions (arching, turning of head) designated Sandifer syndrome.

18.(B) An association has been found with the use of erythromycin in neonates with highest risk if the medication is given within the first 2 wk of life. There have also been reports of higher incidence of pyloric stenosis among mostly female infants of mothers treated with macrolide antibiotics during pregnancy and breastfeeding.

19.(B)

- ✓ **Symptomatic** ---- Frank malabsorption symptoms and signs (e.g., chronic diarrhea, failure to thrive, weight loss). Extraintestinal symptoms and signs (e.g., anemia, fatigue, hypertransaminasemia, neurologic disorders, short stature, dental enamel defects, arthralgia, aphthous stomatitis)
- ✓ **Silent** ---- No apparent symptoms in spite of histologic evidence of villous atrophy, in most cases identified by serologic screening in at-risk groups
- ✓ **Latent** ---- Subjects who have a normal intestinal histology, but at some other time have shown a gluten dependent enteropathy
- ✓ **Potential** ---- Subjects with positive celiac disease serology but without evidence of altered intestinal histology. Patients may or may not have symptoms and signs of disease and may or may not develop a gluten-dependent enteropathy later.
- ✓ **Non-celiac gluten sensitivity (NCGS)** ---- is a poorly understood condition. Diagnosis is suspected in patients who do not have CD or wheat allergy, and yet show GI and non-GI symptoms upon ingestion of gluten- or wheat-containing food.

20.(D) When anti-TG2 <10 x normal should do esophagogastroduodenoscopy and biopsy; if the result of biopsy is

- ✓ Marsh 0-1---- Unclear case and consider: false positive serology or false negative biopsy or potential CD and extended evaluation of HLA/; serology/biopsies
- ✓ Marsh 2 or 3----diagnosed as a celiac disease and start gluten-free diet

Non-celiac gluten sensitivity (NCGS) ---- is a poorly understood condition. Diagnosis is suspected in patients who do not have CD or wheat allergy, and yet show GI and non-GI symptoms upon ingestion of gluten- or wheat-containing food.

21.(D) Asymptomatic person at genetic risk for CD, HLA positive DQ2 and/or DQ8, TG2 Titer > 3 x normal ------ should do EGD & biopsies from Bulbus & 4 x pars descendens, proper histological work up (Marsh 0 or 1 or Marsh 2 or 3).

22.(A) One of the most common extraintestinal manifestation of CD is iron-deficiency anemia, which is usually unresponsive to iron therapy. Osteoporosis may be present; in contrast to adults, it can be reversed by a gluten-free diet, with restoration of normal peak bone densitometric values. Other extraintestinal manifestations include short stature, delayed puberty, arthritis and arthralgia, epilepsy with bilateral occipital calcifications, peripheral neuropathies, isolated hypertransaminasemia, dental enamel hypoplasia, and aphthous stomatitis.

23.(B)

- ✓ Avoid all foods containing wheat, rye, and barley gluten (pure oats usually safe).
- ✓ Avoid malt unless clearly labeled as derived from corn.
- ✓ Use only rice, corn, maize, buckwheat, millet, amaranth, quinoa, sorghum, potato or potato starch, soybean, tapioca, and teff, bean, and nut flours.
- ✓ Wheat starch and products containing wheat starch should only be used if they contain <20 ppm gluten and are marked “gluten free.”
- ✓ Read all labels and study ingredients of processed foods.
- ✓ Beware of gluten in medications, supplements, food additives, emulsifiers, or stabilizers.
- ✓ Limit milk and milk products initially if there is evidence of lactose intolerance.
- ✓ Avoid all beers, lagers, ales, and stouts (unless labeled gluten free).

24.(E) Salmonella and Shigella (egg salad).

25.(A) The Codex Alimentarius Guidelines define gluten-free food item for food containing <20 ppm (equivalent to 20 mg gluten in 1 kg of product); however, although analytical methods for gluten detection have reached a satisfactory degree of sensitivity, more information is needed on the daily gluten amount that may be tolerated by CD patients. The data available so far seem to suggest that the threshold should be set to <50 mg/day, although individual variability makes it difficult to set a universal threshold

26.(B)

Celiac testing recommended	Celiac testing should be considered
Persistent unexplained abdominal or gastrointestinal symptoms	Metabolic bone disorders (reduced bone mineral density or osteomalacia)
Faltering growth	Unexplained neurological symptoms (particularly peripheral neuropathy or ataxia)
Prolonged fatigue	Unexplained subfertility or recurrent miscarriage
Unexpected weight loss	Persistently increased concentrations of liver enzymes with unknown cause
Severe or persistent mouth ulcers	Dental enamel defects
Unexplained iron, vitamin B12, or folate deficiency	Down syndrome
Type 1 diabetes	Turner syndrome
Autoimmune thyroid disease	William syndrome
Irritable bowel syndrome	Selective IgA deficiency
First degree relatives of people with celiac disease	
Dermatitis herpetiformis	

27.(C) Esophagogastroduodenoscopy is the method of choice to establish the diagnosis of peptic ulcer disease. It can be safely performed in all ages by experienced pediatric gastroenterologists. Endoscopy allows the direct visualization of esophagus, stomach, and duodenum, identifying the specific lesions. Biopsy specimens must be obtained from the esophagus, stomach, and duodenum for histologic assessment as well as to screen for the presence of H. pylori infection. Endoscopy also provides the opportunity for hemostatic therapy including clipping, injection, and the use of thermal coagulation.

28.(C) Patients should stop proton pump inhibitor (PPI) therapy 2 wk prior to testing as negative results on therapy may represent false negatives. Successful H pylori eradication is associated with cure of peptic ulcer disease and very low risk of relapse. Therefore, monitoring the success of therapy is mandatory in these patients 4-6 wk after stopping antibiotics and at least 2 wk after stopping PPI therapy. Eradication can be tested with the 13C-urea breath (13C-UBT) test or stool antigen test.

29.(B) Chronicity is an important part of the diagnosis; it is difficult to know if a patient has a subacute, transient **infectious colitis** or ulcerative colitis when a child has had 1-2 wk of symptoms. Although **Henoch-Schönlein purpura** can manifest as abdominal pain and bloody stools, it is not usually associated with colitis. The most difficult distinction is from **Crohn disease** because the colitis of Crohn disease can initially appear identical to that of ulcerative colitis, particularly in younger children. The gross appearance of the colitis or development of small bowel disease eventually leads to the correct diagnosis; this can occur years after the initial presentation. **Behçet disease** can be distinguished by its typical features

30.(C) Systemic signs and symptoms are more common in Crohn disease than in ulcerative colitis. Fever, malaise, and easy fatigability are common. Growth failure with delayed bone maturation and delayed sexual development can precede other symptoms by 1 or 2 yr and is at least twice as likely to occur with Crohn disease as with ulcerative colitis. Children can present with growth failure as the only manifestation of Crohn disease. Decreased height velocity occurs in about 88% of prepubertal patients diagnosed with Crohn disease, and this often precedes GI symptoms.

31.(B)

- ✓ Fecal **calprotectin** and **lactoferrin** are increasingly being used as more sensitive and specific markers of bowel inflammation as compared to serologic parameters, and these are often elevated.
- ✓ α1-Antitripsin ---- Increased intestinal permeability/protein loss
- ✓ Steatocrit ---- Fat malabsorption
- ✓ Elastase ---- Pancreatic function
- ✓ Chymotrypsin ---- Pancreatic function

32.(D) Fever, severe anemia, hypoalbuminemia, leukocytosis, and more than 5 bloody stools per day for 5 days define fulminant colitis.

Probiotics are effective in adults for maintenance of remission for ulcerative colitis, although they do not induce remission during an active flare. The first drug class to be used with mild or mild-to-moderate colitis is an aminosalicylate. Many children with disease requiring frequent corticosteroid therapy are started on immunomodulators such as **azathioprine** (2.0-2.5 mg/kg/day) or 6-mercaptopurine (1-1.5mg/kg/day). **Steroids** are considered an effective medication for acute flares, but they are not appropriate maintenance medications because of loss of effect and side effects. **Colectomy** is performed for intractable disease, complications of therapy, and fulminant disease that is unresponsive to medical management.

33.(B)

- ✓ Extraintestinal manifestations that tend to occur more commonly with ulcerative colitis than with Crohn disease include pyoderma gangrenosum, sclerosing cholangitis, chronic active hepatitis, and ankylosing spondylitis.
- ✓ Extraintestinal manifestations occur more commonly with Crohn disease than with ulcerative colitis; those that are especially associated with Crohn disease include oral aphthous ulcers, peripheral arthritis, erythema nodosum, digital clubbing, episcleritis, renal stones (uric acid, oxalate), and gallstones.

34.(B) Conservative management with nasoduodenal feedings is advisable in patients who are not good surgical candidates. Oral and intravenous atropine sulfate (pyloric muscle relaxant) has also been described when surgical expertise is not available with 80% success rate described in some studies.

35.(D) The diagnosis of malrotation may be suggested by **ultrasound** but is confirmed by **contrast radiographic studies**. The **abdominal plain film** is usually nonspecific but might demonstrate a gasless abdomen or evidence of duodenal obstruction with a double-bubble sign. **Upper gastrointestinal series** is the imaging test of choice and the gold standard in the evaluation and diagnosis of malrotation and volvulus.

36.(C) The hallmark of duodenal obstruction is bilious vomiting without abdominal distention, which is usually noted on the first day of life. Peristaltic waves may be visualized early in the disease process. A history of polyhydramnios is present in half the pregnancies and is caused by inadequate absorption of amniotic fluid in the distal intestine. This fluid may be bile stained because of intrauterine vomiting. Jaundice is present in one-third of the infants.

37.(D) Soda beverages, fruit juices, tea, and other home fluids are not suitable for rehydration or maintenance therapy because they have inappropriately high glucose concentration and osmolalities and low sodium concentrations. Foods with complex carbohydrates (rice, wheat, potatoes, bread, and cereals), fresh fruits, lean meats, yogurt, and vegetables should be reintroduced while ORS is given to replace ongoing losses from emesis or stools and for maintenance. Fatty foods or foods high in simple sugars (juices, carbonated sodas) should be avoided.

38.(A) Symptoms of a Meckel diverticulum usually arise in the first or second yr of life (average: 2.5 yr), but initial symptoms can occur in the first decade. The majority of symptomatic Meckel diverticula are lined by an ectopic mucosa, including an acid-secreting mucosa that causes intermittent painless rectal

bleeding by ulceration of the adjacent normal ileal mucosa. This ectopic mucosa is most commonly of gastric origin, but it can also be pancreatic, jejunal, or a combination of these tissues. Unlike the upper duodenal mucosa, the acid is not neutralized by pancreatic bicarbonate. The stool is typically described as brick colored or currant jelly colored. Bleeding can cause significant anemia but is usually self-limited because of contraction of the splanchnic vessels, as patients become hypovolemic. Less often, a Meckel diverticulum is associated with partial or complete bowel obstruction. The most common mechanism of obstruction occurs when the diverticulum acts as the lead point of an intussusception.

39.(E) In patients with obstruction caused by jejunoileal atresia or long segment Hirschsprung disease, plain radiographs typically demonstrate multiple air–fluid levels proximal to the obstruction in the upright or lateral decubitus positions. These levels may be absent in patients with meconium ileus because the viscosity of the secretions in the proximal bowel prevents layering. Instead, a typical hazy or ground glass appearance may be appreciated in the right lower quadrant. This haziness is caused by small bubbles of gas that become trapped in inspissated meconium in the terminal ileal region.

40.(E) The most common type of malrotation involves failure of the cecum to move into the right lower quadrant.

41.(D) Ileus is the failure of intestinal peristalsis caused by loss of coordinated gut motility without evidence of mechanical obstruction. In children, it is most often associated with abdominal surgery or infection (gastroenteritis, pneumonia, peritonitis). Ileus also accompanies metabolic abnormalities (e.g., uremia, hypokalemia, hypercalcemia, hypermagnesemia, and acidosis) or administration of certain drugs, such as opiates, vincristine, and antimotility agents such as loperamide when used during gastroenteritis.

42.(A)

43.(D) An unprepared contrast enema is most likely to aid in the diagnosis in children older than 1 mo of age because the proximal ganglionic segment might not be significantly dilated in the first few wk of life. Classic findings are based on the presence of an abrupt narrow transition zone between the normal dilated proximal colon and a smaller-caliber obstructed distal aganglionic segment. In the absence of this finding, it is imperative to compare the diameter of the rectum to that of the sigmoid colon, because a rectal diameter that is the same as or smaller than the sigmoid colon suggests Hirschsprung disease. Radiologic evaluation should be performed without prior preparation (i.e.,

unprepared contrast enema study) to prevent transient dilation of the aganglionic segment.

44.(B)

45.(A) The recurrence rate after hydrostatic reduction of intussusceptions is approximately 10%, and after surgical reduction it is 2–5%; none has recurred after surgical resection. Most recurrences occur within 72 hr of reduction. Corticosteroids may reduce the frequency of recurrent intussusceptions but are rarely used for this purpose.

46.(D) The diagnostic findings of intussusception on ultrasound include a tubular mass in longitudinal views and a doughnut or target appearance in transverse images. Ultrasound has a sensitivity of approximately 98–100% and a specificity of approximately 98% in diagnosing intussusception.

47.(D) Management of pseudomembranous colitis; Oral vancomycin and metronidazole for 7-14 days (first line therapy) displayed equivalent efficacy in a prospective randomized trial.

48.(E) Ondansetron (oral mucosal absorption preparation) reduces the incidence of emesis, thus permitting more effective oral rehydration and is well-established in emergency management of AGE in high resource settings, reducing intravenous fluid requirements and hospitalization. Because persistent vomiting can limit ORS, a single sublingual dose of an oral dissolvable tablet of ondansetron (4 mg for children 4-11 yr old and 8 mg for children older than 11 yr [generally 0.2 mg/kg]) may be given. Antiemetic agents, such as the phenothiazines (such as prochlorperazine), are of little value and are associated with potentially serious side effects (lethargy, dystonia, malignant hyperpyrexia).

49.(A)

First line:

- ✓ Ciprofloxacin 15mg/kg/day PO bid ×3 days; OR
- ✓ Ceftriaxone 50-100mg/kg/day IV or IM, qd ×3 days for severe illness requiring parenteral therapy; OR
- ✓ Azithromycin 12mg/kg once on 1st day, then 6 mg/kg once daily on days 2 through 4 (total course: 4 days)

Second line:

- ✓ Cefixime 8 mg/kg once daily for 3 days; OR
- ✓ Trimethoprim-sulfamethoxazole 4 mg/kg/day of TMP and 20 mg/kg/day SMX twice a day for 5 days (if susceptibility known or likely based on local data).

50.(B) Intrinsic properties of the organism help to define the mode of transmission and incubation period. Enteropathogens that are infectious in **small inocula** (Shigella, STEC, norovirus, rotavirus, G. intestinalis, Cryptosporidiumspp., C. difficile, E. histolytica) are readily transmitted by person to-person contact via the fecal-oral route. Pathogens with **larger infectious** doses, such as cholera, NTS, ETEC, and Campylobacter, generally require food or water vehicles.

51.(A) Rotavirus is the most common cause of AGE among children throughout the world. Several other viruses occur less frequently.

52.(D) Etiology; Bacillus cereus (preformed emetic toxin). Incubation period 1-6 hr. clinical presentations are sudden onset of severe nausea and vomiting; diarrhea may be present 24 hr Soil and water. Risk factors; improperly refrigerated cooked or fried rice, meats.

53.(C) Risk of colon cancer; Juvenile polyposis (10–50%), Peutz-Jeghers syndrome (47–93%.), Familial adenomatous polyposis (100%), Lynch syndrome (not increased), and Cowden syndrome (30%).

54.(B) Peutz-Jeghers syndrome (PJS) is a rare autosomal dominant disorder (incidence: ~1:120,000 total population) characterized by mucocutaneous pigmentation and extensive GI hamartomatous polyposis. Macular pigmented lesions may be dark brown to dark blue and are found primarily around the lips and oral mucosa, although these lesions may also be found on the hands, feet, or perineum. Lesions can fade by puberty or adulthood.

55.(C)

56.(A) The gallbladder is congenitally absent in approximately 0.1% of the population. Hypoplasia or absence of the gallbladder can be associated with extrahepatic biliary atresia or cystic fibrosis. Kawasaki disease and Henoch-Schönlein purpura are associated with hydrops of the gallbladder. Sickle cell anemia and Gilbert disease are associated with cholelithiasis.

57.(A) Chronic diarrhea is defined as stool volume of more than 10 g/kg/day in toddlers/infants and greater than 200 g/day in older children that lasts for 4 wk or more. Persistent diarrhea began acutely but lasts longer than 14 days. In practice, this usually means having loose or watery stools more than 3 times a day.

58.(A) The most benign and common etiology of chronic diarrhea is nonspecific diarrhea that encompasses functional diarrhea (or toddler's diarrhea) in children younger than 4 yr of age and irritable bowel syndromein those 5 yr of age and older. It is the leading cause of chronic diarrhea in an otherwise well child. Toddler's diarrhea is defined by the daily painless recurrent passage of 4

or larger unformed stools, for 4 or more wk, with onset in infancy or preschool years. Nighttime defecation is usually absent. The child appears unperturbed by the diarrhea, there is no evidence of failure to thrive, and the symptoms resolve spontaneously by school age.

59.(C) In developing countries, enteroadherent Escherichia coli and Giardia lamblia have been implicated in chronic diarrhea, whereas, in developed countries, chronic infectious diarrhea usually runs a more benign course and the etiology is often viral, with a major role of rotavirus and norovirus.

60.(C) Diagnostic criteria for abdominal migraine must include all of the following occurring at least twice:

1. Paroxysmal episodes of intense, acute periumbilical, midline, or diffuse abdominal pain lasting 1 hr or more (should be the most severe and distressing symptom)
2. Episodes are separated by weeks to months
3. The pain is incapacitating and interferes with normal activities
4. Stereotypical pattern and symptoms in the individual patient
5. The pain is associated with 2 or more of the following: a. Anorexia b. Nausea c. Vomiting d. Headache e. Photophobia f. Pallor
6. After appropriate evaluation, the symptoms cannot be fully explained by another medical condition.

61.(B) Supportive Care and Abortive Treatment Approaches in Cyclic Vomiting Syndrome

1- Supportive care

- ✓ Fluid and electrolyte management ---- Dextrose containing fluid (D10) and normal saline
- ✓ Nutrition ---- Resume enteral nutrition as soon as possible and if unable to tolerate enteral nutrition and meets criteria, start parenteral nutrition after 3-4 days.
- ✓ Medications
- Antiemetics ---- Ondansetron and alternative: Granisetron
- Sedatives ---- Diphenhydramine, Lorazepam , Chlorpromazine+diphenhydramine IV
- Analgesics ---- Ketorolac
- ✓ Treatment of specific signs and symptoms
- ✓ Epigastric pain ---- H2RA or PPI
- ✓ Diarrhea ---- Antidiarrheals
- ✓ Hypertension ---- captopril
- ✓ Treatment of specific complications

2- Abortive care ---- Antimigraine (triptans), Sumatriptan

3- Recovery and refeeding

62.(B) The invertogram or upside-down x-ray is of little value, but a prone cross table lateral plain x-ray at 24 hr of life (to allow time for bowel distention from swallowed air) with a radiopaque marker on the perineum can demonstrate a low lesion by showing the rectal gas bubble <1 cm from the perineal skin. A plain x-ray of the entire sacrum, including both iliac wings, is important to identify sacral anomalies and the adequacy of the sacrum.

63.(B) There are many anomalies associated with anorectal malformations. The most common are anomalies of the kidneys and urinary tract in conjunction with abnormalities of the sacrum. This complex is often referred to as caudal regression syndrome.

64.(C) The liver span increases linearly with body weight and age in both sexes, ranging from approximately 4.5-5.0 cm at 1 wk of age to approximately 7-8 cm in boys and 6.0-6.5 cm in girls by 12 yr of age.

65.(E) Pruritus; intense generalized itching can occur in patients with chronic liver disease often in association with cholestasis (conjugated hyperbilirubinemia). Symptoms can be generalized or localized (commonly to palms and soles), are usually worse at night, are exacerbated with stress and heat, and are relieved by cool temperatures. Pruritus is unrelated to the degree of hyperbilirubinemia; deeply jaundiced patients can be asymptomatic.

66.(A) Spider angiomas; vascular spiders (telangiectasias), characterized by central pulsating arterioles from which small, wiry venules radiate, may be seen in patients with chronic liver disease. These are usually most prominent in the superior vena cava distribution area (on the face and chest). Their size varies between 1 and 10 mm and they exhibit central clearing with pressure. They presumably reflect altered estrogen metabolism in the presence of hepatic dysfunction.

67.(E) **Abdominal ultrasound** is a helpful diagnostic tool in evaluating neonatal cholestasis because it identifies choledocholithiasis, perforation of the bile duct, or other structural abnormalities of the biliary tree such as a choledochal cyst. **Hepatobiliary scintigraphy with technetium-labeled iminodiacetic acid derivatives** is a sensitive but not specific test for biliary atresia. The role of **ERCP** in the diagnostic process of biliary atresia remains indeterminate. Similarly, the value of **MRCP** in the diagnosis of biliary atresia has not been established. Percutaneous **liver biopsy** is the most valuable procedure in the evaluation of neonatal hepatobiliary diseases and provides the most reliable discriminatory evidence.

68.(D) Aagenaes syndrome is a form of idiopathic familial intrahepatic cholestasis associated with lymphedema of the lower extremities. The relationship between liver disease and lymphedema is not understood and may be attributable to decreased hepatic lymph flow or hepatic lymphatic hypoplasia. Affected patients usually present with episodic cholestasis with elevation of serum aminotransferase, alkaline phosphatase, and bile acid levels. Between episodes, the patients are usually asymptomatic and biochemical indices improve. Compared to other types of hereditary neonatal cholestasis, patients with Aagenaes syndrome have a relatively good prognosis. The locus for Aagenaes syndrome is mapped to a 6.6 cM interval on chromosome 15q.

69.(A) Zellweger (cerebrohepatorenal) syndrome is a rare autosomal recessive genetic disorder marked by progressive degeneration of the liver and kidneys; the disease is usually fatal by 6-12 mo of age. Affected infants have severe, generalized hypotonia and markedly impaired neurologic function with psychomotor retardation.

70.(C) Neonatal iron storage disease (neonatal hemochromatosis, gestational alloimmune liver disease) is a rapidly progressive disease characterized by increased iron deposition in the liver, heart, and endocrine organs without increased iron stores in the reticuloendothelial system. Patients have multiorgan failure and shortened survival. Familial cases are reported, and repeated affected neonates in the same family are common. This is an alloimmune disorder with maternal antibodies directed against the fetal liver. Neonatal hemochromatosis (or fetal loss) seems to be a gestational alloimmune disease, and reoccurrence of severe neonatal hemochromatosis in at-risk pregnancies may be reduced by maternal treatment with weekly high-dose intravenous immunoglobulin (1 g/kg) beginning gestational age 18 wk. The diagnosis is usually confirmed by buccal mucosal biopsy or MRI demonstrating extrahepatic siderosis.

The prognosis is poor; however, liver transplantation can be curative.

71.(B) Alagille syndrome (arteriohepatic dysplasia) is the most common syndrome with intrahepatic bile duct paucity. In addition to others clinical manifestations as ocular abnormalities (posterior embryotoxon, microcornea, optic disk drusen, shallow anterior chamber), cardiovascular abnormalities (usually peripheral pulmonic stenosis, sometimes tetralogy of Fallot, pulmonary atresia, ventricular septal defect, atrial septal defect, aortic coarctation), vertebral defects (butterfly vertebrae, fused vertebrae, spina bifida occulta, rib anomalies), and tubulointerstitial nephropathy. Other findings such as short stature, pancreatic insufficiency, vasculopathy (Moyamoya syndrome, stroke),

and defective spermatogenesis can reflect or produce nutritional deficiency. Patients with Alagille syndrome are likely to have pruritus, xanthomas with markedly elevated serum cholesterol levels, and neurologic complications of vitamin E deficiency if untreated.

72.(C) If flow is not rapidly established in the first month of life, progressive obliteration and cirrhosis ensue. If microscopic channels of patency >150 µm in diameter are found, postoperative establishment of bile flow is likely. The success rate for establishing good bile flow after the Kasai operation is much higher (90%) if performed before 8 wk of life.

73.(C) A degenerative neuromuscular syndrome is found in patients with chronic cholestasis, caused by vitamin E deficiency; affected children experience progressive areflexia, cerebellar ataxia, ophthalmoplegia, and decreased vibratory sensation.

74.(A) Liver disease is the most common disease manifestation in children and can precede neurologic symptoms by as long as 10 yr. Females are 3 times more likely than males to present with acute hepatic failure. When Wilson disease presents after age 20, neurologic symptoms are the most common manifestation. Kayser-Fleischer rings are absent in young patients with hepatic Wilson disease up to 50% of the time but are present in 95% of patients with neurologic symptoms. Coombs-negative hemolytic anemia may be an initial manifestation, possibly related to the release of large amounts of copper from damaged hepatocytes.

75.(A) Most patients with Wilson disease have decreased serum ceruloplasmin levels. The serum free copper level may be elevated in early Wilson disease, and urinary copper excretion (normally <40 µg/day) is increased to >100 µg/day and often up to 1,000 µg or more per day, in equivocal cases, the response of urinary copper output to chelation may be of diagnostic help.

76.(E) Liver biopsy can determine the extent and severity of liver disease and for measuring the hepatic copper content (normally <10 µg/g dry weight) but is only required if clinical signs and noninvasive tests do not allow a final diagnosis or if another liver disorder is suspected. Hepatic copper accumulation is the hallmark of Wilson disease and measurement of hepatic parenchymal copper concentration is the method of choice for diagnosis.

77.(B) This screening should include determination of the serum ceruloplasmin level and 24-hr urinary copper excretion. If these results are abnormal or equivocal, liver biopsy should be carried out to determine morphology and hepatic copper content. Genetic screening by either linkage analysis or direct DNA mutation analysis is possible

78.(B) For patients with Wilson disease, the dietary intake of copper should be restricted to <1 mg/day. High copper content foods such as **liver**, **shellfish**, **nuts**, and **chocolate** should be avoided. If the copper content of the drinking water exceeds 0.1 mg/L, it may be necessary to demineralize the water.

79.(C) Zinc has been used as adjuvant therapy, maintenance therapy, or primary therapy in presymptomatic patients, owing to its unique ability to impair the gastrointestinal absorption of copper. Because penicillamine is an antimetabolite of vitamin B6, additional amounts of this vitamin are necessary.

80.(B) Immune therapy with exchange transfusion and intravenous immunoglobulin has been reported to improve the outcome and reduce the need for liver transplantation in patients with neonatal hemochromatosis.

81.(A) With recovery, the liver morphology returns to normal within 3 mo of the acute infection. If chronic hepatitis develops, the inflammatory infiltrate settles in the periportal areas and often leads to progressive scarring. Both of these hallmarks of chronic hepatitis are seen in cases of HBV and HCV.

82.(B) Patients infected with HAV are contagious for 2 wk before and approximately 7 days after the onset of jaundice and should be excluded from school, childcare, or work during this period. Careful hand-washing is necessary, particularly after changing diapers and before preparing or serving food. In hospital settings, contact and standard precautions are recommended for 1 wk after onset of symptoms.

83.(D)

84.(D)

Characteristic autoantibodies in type 1 AIH

- ✓ Antinuclear antibody
- ✓ Smooth-muscle antibody
- ✓ Antiactin antibody
- ✓ Autoantibodies against soluble liver antigen and liver-pancreas antigen
- ✓ Atypical perinuclear antineutrophil cytoplasmic antibody

Characteristic autoantibodies in type 1 AIH

- ✓ Antibody against liver-kidney microsome type 1
- ✓ Antibody against liver cytosol type 1
- ✓ Antibody against liver-kidney microsomal type 3

85.(A) Prednisone, with or without azathioprine or 6-mercaptopurine, improves the clinical, biochemical, and histologic features in most patients with autoimmune hepatitis and prolongs survival in most patients with severe disease. The goal is to suppress or eliminate hepatic inflammation with minimal side effects.

86.(A) Patients should be monitored closely for infection, including sepsis, pneumonia, peritonitis, and urinary tract infections. At least 50% of patients experience serious infection. Gram-positive organisms (Staphylococcus aureus, Staphylococcus epidermidis) are the most common pathogens, but Gram-negative and fungal infections are also observed.

87.(A) All the options except galactosemia can be associated with prenatal liver injury; gestational alloimmune liver disease (GALD) is the most common cause of acute liver failure in the neonate.

88.(D) Hypophosphatemia, probably a reflection of liver regeneration, and early phosphorus administration are associated with a better prognosis in acute liver failure, whereas hyperphosphatemia predicts a failure of spontaneous recovery.

Chapter 18
The Respiratory System Questions
ZUHAIR ALMUSAWI

1. Which of the following hematological diseases may develop digital clubbing?
   A. Thalassemia
   B. Sickle cell anemia
   C. Iron deficiency anemia
   D. Fanconi anemia
   E. Congenital spherocytosis

2. The following arterial blood gas readings (↓ PCO 2, ↓ PO 2) are seen in
   A. depressed respiratory center
   B. mild intrapulmonary airway obstruction
   C. Ineffective neuromuscular function
   D. central (above the carina) airway obstruction
   E. alveolar–interstitial pathology

3. The utility of MRI of the chest is largely limited to the analysis of
   A. vascular anatomy
   B. early metastases
   C. pleural lesions
   D. cystic parenchymal lesions
   E. bronchiectasis

4. What is the **MOST** common cause of recurrent or persistent cough in children?
   A. Asthma
   B. Chronic rhinitis
   C. Cystic fibrosis
   D. Habit cough
   E. Foreign-body aspiration

5. A 7-year-old boy presents with chronic barking cough all the day with no constitutional symptoms and he sleeps comfortably at night.
Of the following, the **MOST** likely diagnosis is

A. croup
B. tracheomalacia
C. tracheitis
D. foreign body
E. habit cough

6. An 8-year-old boy brought by his father because of throat clearing sound all the day for the last three months, the boy looks healthy with normal ENT examination.
Of the following, the **MOST** likely cause of his problem is

A. gastroesophageal reflux
B. postnasal drip
C. vocal tic
D. sinusitis
E. reactive airway disease

7. A 5-year-old child developed bronchopneumonia 6 weeks ago, treated properly with clinical and radiological improvement. Since that time till now he has persistent cough, for this reinvestigated with chest x-ray, complete blood count, and ESR which were normal. Sputum examination revealed mostly macrophages.
Of the following, the **MOST** likely cause of his persistent cough is

A. asthma
B. cystic fibrosis
C. postinfectious hypersensitivity of cough receptors
D. repeated aspiration
E. plastic bronchitis

8. A 7-year-old child presents with intermittent stridor and wheezing which accompanies physical activity, diagnosed as asthma with no response to asthma therapies.
Of the following, the **MOST** likely cause of his/her problem is

A. allergic (spasmodic) croup
B. laryngeal webs
C. bilateral abductor paralysis of the cords

D. paradoxical vocal cord dysfunction
E. acquired subglottic tracheal stenosis

9. Which of the following is a risk factor associated with sudden infant death syndrome?
A. Age (peak 6-9 mo)
B. Female gender
C. Pacifier (dummy)
D. Postmaturity
E. Recent febrile illness (mild infections)

10. A young mother brought her 6-month-old boy for evaluation, he is completely normal now but she gave a history of sudden attack of cyanosis, irregular breathing, hypotonia, and poor responsiveness which lasted about 30 seconds. This event occurred last night and he returned normal after blowing on his face.
Of the following, the **MOST** likely explanation for this scenario is
A. aborted crib deaths
B. breath holding spell
C. apnea of infancy
D. brief resolved unexplained event
E. prolonged QT syndrome

11. What is the **MOST** common presenting clinical symptom of foreign body in the nose of a child?
A. Mucopurulent nasal discharge
B. Foul nasal odor
C. Epistaxis
D. Nasal obstruction
E. History of insertion of foreign bodies

12. What is he **MOST** common childhood cause of nasal polyposis?
A. Chronic sinusitis
B. Allergic rhinitis
C. Cystic fibrosis
D. Low vitamin D levels
E. Samter triad

13. A change in the color or consistency of the nasal secretions during the course of common cold illness may indicate

A. sinusitis
B. bacterial superinfection
C. accumulation of polymorphonuclear cells
D. vasomotor rhinitis
E. allergic rhinitis

14. Which of the following is no more effective than placebo for the treatment of cold symptoms?

A. Zinc
B. Pseudoephedrine
C. Phenylephrine
D. Vitamin C
E. Saline nose drops

15. Which of the following is a minor symptom of sinusitis?

A. Purulent anterior nasal discharge
B. Nasal congestion or obstruction
C. Facial congestion or fullness
D. Hyposmia or anosmia
E. Dental pain

16. What is the preferred antimicrobial agent used for treatment of group A streptococcal pharyngitis in children?

A. Penicillin
B. Amoxicillin
C. Benzathine penicillin
D. Benzathineprocaine penicillin G combination
E. Azithromycin

17. A 3-year-old child presents with fever, irritability, decreased oral intake, and drooling. Physical examination revealed torticollis, refusal to move the neck with muffled voice and stridor.
Of the following, the **MOST** likely diagnosis is

A. acute epiglottitis
B. foreign body aspiration
C. meningitis

D. retropharyngeal abscess
E. vertebral osteomyelitis.

18. A 2-year-old boy has rhinorrhea, mild cough, and low-grade fever for 2 days, after that he develops barking cough, hoarseness, and inspiratory stridor; crying greatly aggravates his symptoms and the child prefers to sit up in bed. Respiratory rate was 34 breath/min, heart rate 110 beat/min, and oxygen saturation 95%.
Of the following, the **MOST** likely diagnosis is
A. laryngotracheobronchitis
B. acute epiglottitis
C. acute infectious laryngitis
D. spasmodic croup
E. bacterial tracheitis

19. A 3-year-old healthy child suddenly develops a sore throat and fever. Within a matter of hours, the patient appears toxic, swallowing is difficult with drooling, and labored breathing. The neck is hyperextended and the child assumes the tripod position.
Of the following, the **MOST** likely diagnosis is
A. laryngotracheobronchitis
B. acute epiglottitis
C. acute infectious laryngitis
D. spasmodic croup
E. bacterial tracheitis

20. A 3-year-old child awakens with a characteristic barking cough, noisy inspiration, respiratory distress, and anxious frightened appearance.
He was afebrile and was well when went to sleep. He had similar attack 3 months ago which resolve within several hours.
Of the following, the **MOST** likely diagnosis is
A. laryngotracheobronchitis
B. acute epiglottitis
C. acute infectious laryngitis
D. spasmodic croup
E. bacterial tracheitis

21. The effectiveness of oral corticosteroids in viral croup is well established. Of the following, the **LEAST** effective option is

A. single dose of oral dexamethasone
B. Intramuscular dexamethasone
C. nebulized budesonide
D. single dose of oral prednisolone
E. prednisolone 2 mg/kg/day for 3 days

22. A 5-year-old boy looks toxic with high fever, brassy cough, and respiratory distress after a few days of apparent improvement of laryngotracheobronchitis. The patient can lie flat, does not drool, and the usual treatment for croup (racemic epinephrine) is ineffective. His vaccination was up date.
Of the following, the **MOST** commonly isolated pathogen is

A. S. aureus
B. S.pneumoniae
C. Moraxella catarrhalis
D. nontypeable H. influenzae
E. Parainfluenza virus

23. Which of the following conditions should be considered in a child with recurrent or persistent croup-like symptoms?

A. Laryngomalacia
B. Congenital subglottic stenosis
C. Vocal cord paralysis
D. Congenital laryngeal web
E. Subglottic hemangioma

24. A 6-month-old infant with congenital hydrocephalus presents with respiratory distress, high-pitched inspiratory stridor, dysphonic sound, and inspiratory weak cry.
Of the following, the **MOST** likely diagnosis is

A. bilateral vocal cord paralysis
B. congenital laryngeal web
C. subglottic hemangioma
D. laryngocele
E. congenital subglottic stenosis

25. What is the first-line treatment of subglottic hemangiomas?

A. Oral prednisone
B. Oral propranolol
C. Intralesional steroid injection
D. KTP laser endoscopic excision
E. Tracheostomy

26. Acute bronchiolitis in a previously healthy infant presenting with a first episode of wheezing (following a period of upper respiratory symptoms) needs which of the following for the diagnosis?

A. Chest radiography
B. White blood cell and differential counts
C. Polymerase chain reaction
D. Rapid immunofluorescence
E. Good clinical examination

27. Which of the following is of value in the management of children with viral bronchiolitis?

A. Chest physiotherapy
B. Corticosteroids
C. Hypertonic saline
D. Ribavirin
E. Frequent suctioning of nasal and oral secretions

28. What is the **BEST** treatment for cough in an infant with acute bronchitis?

A. Cough suppressants
B. Antihistamines
C. Expectorants
D. Cold medicines
E. Frequent shifts in position

29. Which of the following conditions is proved to cause plastic bronchitis?

A. Pulmonary lymphatic anomalies
B. Toxic inhalation
C. Sickle cell acute chest syndrome
D. Cystic fibrosis
E. Bronchiectasis

30. A 3-year-old healthy child presented with a neck mass noticed while straining or coughing.
Of the following, the **MOST** likely diagnosis is

A. cystic hygroma
B. extrapulmonary sequestration
C. bronchogenic cyst
D. laryngocele
E. Sibson hernia

31. What is the earliest clinical sign of pulmonary edema?

A. Tachypnea
B. Fine crackles and wheezing
C. Gallop rhythm
D. Peripheral edema
E. Jugular venous distention

32. What is the early chest radiographic sign of pulmonary edema?

A. Diffuse streakiness
B. Diffuse patchy densities
C. Cardiomegaly
D. Peribronchial cuffing
E. Pleural effusion

33. What is the **MOST** common underlying problem associated with recurrent pneumonias in hospitalized children?

A. Oropharyngeal incoordination
B. Micrognathia
C. Macroglossia
D. Achalasia
E. Nasal feeding tube

34. What is the **MOST** appropriate test for the diagnosis of recurrent microaspirations?

A. Plain chest radiograph
B. High-resolution CT
C. Videofluoroscopic swallowing study
D. Barium esophagram
E. Esophageal pH monitoring

35. What is the **MOST** helpful test to the clinician in supporting the diagnosis of hypersensitivity pneumonia?

A. High-resolution computerized tomography
B. Elevation of immune globulins
C. Elevation of lactate dehydrogenase
D. Lymphocytosis on bronchoalveolar Lavage
E. Elevation of the erythrocyte sedimentation rate

36. What is the ideal method of treatment and prevention of recurrence of hypersensitivity pneumonia?

A. Removal of the offending antigen
B. 0.5 mg/kg/day of prednisolone
C. High dose pulse intravenous methylprednisolone
D. Cyclosporine
E. Azathioprine

37. A 2-year-old boy presents with cough, dyspnea, migratory peripheral pulmonary infiltrates, and blood eosinophilia. He had a history of pica, chronic abdominal pain, and vomiting.
Of the following, the **MOST** likely diagnosis is

A. acute eosinophilic pneumonia
B. Löffler syndrome
C. chronic eosinophilic pneumonia
D. Churg-Strauss Syndrome
E. allergic bronchopulmonary aspergillosis

38. A 5-year-old boy presents with snoring during sleep, behavioral disturbances, learning difficulties, and excessive daytime sleepiness.
What is the **MOST** likely cause of this problem?

A. Habitual snoring
B. Obstructive sleep apnea
C. Allergic rhinitis
D. Adenotonsillar hypertrophy
E. Bilateral nasal polyps

39. Which strains of the following organisms may be associated with particularly rapid pulmonary deterioration and death in cystic fibrosis?

A. Staphylococcus aureus

B. Haemophilus influenza
C. Pseudomonas aeruginosa
D. Burkholderia cepacia
E. Escherichia coli

40. A 6-month-old infant with history of repeated chest infections, chronic diarrhea, and failure to thrive brought to hospital for checking. The parents are cousin and gave history of passage of meconium after 48 hours. Sweat test was ordered but not available. The resident asked you what other investigation is helpful and you told him to do arterial blood gases.
Which of the following findings aid your diagnosis?

A. Hypernatremic hypochloremic metabolic alkalosis
B. Hyponatremic hypochloremic metabolic alkalosis
C. Hypernatremic hyperchloremic metabolic alkalosis
D. Hyponatremic hypochloremic metabolic acidosis
E. Hypernatremic hyperchloremic metabolic acidosis

41. Which of the following conditions associated with false-negative sweat test results in patients with cystic fibrosis?

A. Eczema (atopic dermatitis)
B. Edema
C. Congenital adrenal hyperplasia
D. Hypothyroidism
E. Nephrogenic diabetes insipidus

42. What is the **BEST** diagnostic test for pancreatic malabsorption in cystic fibrosis?

A. Quantification of fat malabsorption with a 72-hr stool
B. Quantification of elastase-1 activity in a fresh stool sample
C. Oral glucose tolerance testing (OGTT)
D. Spot testing of blood and urine glucose levels
E. Glycosylated hemoglobin levels

43. Which of the following pulmonary function studies has been shown to correlate **MOST** closely with mortality in cystic fibrosis?

A. Residual volume
B. Functional residual capacity
C. Forced expiratory volume in 1 sec

D. Forced vital capacity
E. Total lung capacity

44. Which of the following is a broadly effective oral antibiotic for Pseudomonas infection in cystic fibrosis?
A. Dicloxacillin
B. Linezolid
C. Cephalexin
D. Ciprofloxacin
E. Amoxicillin-clavulanate

45. An important indication for aerosolized antibiotic therapy is to eradicate P. aeruginosa in the airways after initial detection in patients with cystic fibrosis. Which of the following antibiotics is commonly used for this purpose?
A. Ticarcillin
B. Piperacillin
C. Tobramycin
D. Meropenem
E. Ceftazidime

46. Which of the following anti-inflammatory drugs are commonly used in cystic fibrosis?
A. Macrolide antibiotics
B. Systemic corticosteroids
C. Inhaled corticosteroids
D. Ibuprofen
E. Cephalosporin antibiotics

47. An 11-year-old boy with cystic fibrosis presents with wheezing, increased cough, shortness of breath, marked hyperinflation, and a decrease in FEV1 that does not respond to antibiotic therapy. Chest radiograph shows new, focal infiltrates. Total serum immunoglobulin E (IgE) level was 1100 with the presence of eosinophils in rust-colored sputum.
Of the following, the **MOST** appropriate initial treatment is
A. oral corticosteroids.
B. oral antifungals
C. omalizumab
D. montelukast

E. meropenem

48. What vitamins supplementation is recommended in patients with cystic fibrosis?

A. A, C, E, K
B. A, D, E, K
C. A, D, B, K
D. A, D, E, B
E. C, D, E, K

49. What is the **MOST** common type of inheritance of primary ciliary dyskinesia?

A. Autosomal recessive
B. X-linked recessive inheritance
C. Autosomal dominant
D. X-linked dominant inheritance
E. Multifactorial

50. A 48-hour-term newborn developed increased work of breathing, tachypnea, and upper and middle lobe atelectasis with dextrocardia on chest radiograph.
Of the following, the **MOST** likely diagnosis is

A. transient tachypnea of the newborn
B. pneumonia
C. hyaline membrane disease
D. primary ciliary dyskinesia
E. meconium aspiration pneumonia

51. Which of the following is a rare clinical manifestation of primary ciliary dyskinesia (PCD)?

A. Unexplained respiratory distress in term neonate
B. Daily, nonseasonal rhinosinusitis since early infancy
C. Atypical asthma unresponsive to therapy
D. Hydrocephalus
E. Male infertility

52. Which of the following pulmonary hemosiderosis is associated with pulmonary capillaritis?

A. Idiopathic pulmonary hemosiderosis

B. Heiner syndrome
C. Goodpasture syndrome
D. Hemolytic uremic syndromes
E. Pulmonary capillary hemangiomatosis

53. Which of the following laboratory findings is classically associated with pulmonary hemosiderosis?
A. Microcytic hypochromic anemia
B. Increased total iron-binding capacity
C. Decreased ferritin levels
D. Low erythrocyte sedimentation rate
E. Reticulocytopenia

54. What is the first-line treatment of idiopathic pulmonary hemosiderosis(IPH)?
A. Plasmapheresis
B. Methotrexate
C. Systemic corticosteroids
D. Intravenous Immunoglobulin
E. Extracorporeal membrane oxygenation (ECMO)

55. What is the **MOST** common identified risk factor of venous thromboembolic disease (VTE) in children?
A. Obesity
B. Immobility
C. Chronic obstructive pulmonary disease
D. Infection
E. Diabetes mellitus

56. What is the diagnostic test of choice to detect a pulmonary embolism in children?
A. Ventilation–perfusion ($\dot{V} - \dot{Q}$) radionuclide scan
B. Spiral CT with an intravenous contrast agent
C. Pulmonary angiography
D. MRI
E. Doppler ultrasonsgraphy

57. What is the **MAIN** side effect of heparin?
A. Thrombocytopenia

B. Leukopenia
C. Renal impairment
D. Pruritic skin rash
E. Hemolytic anemia

58. What is the **MOST** common lobe involved in atelectasis in children?
A. Left upper lobe
B. Left lower lobe
C. Right upper lobe
D. Right middle lobe
E. Right lower lobe

59. Migrating atelectasis in the newborn infant, a rare and unique presentation, may be secondary to which of the following?
A. Neuromuscular disease
B. Bronchopneumonia
C. Meconium-aspiration pneumonia
D. Bronchopulmonary dysplasia
E. Cystic fibrosis

60. What is the **MOST** common form of pulmonary malignancy in children?
A. Bronchial carcinoid
B. Adenoid cystic carcinoma
C. Mucoepidermoid carcinomas
D. Metastatic lesions
E. Pulmonary blastoma

61. Hydrothorax is usually bilateral, but it can be limited to the right side in which of the following?
A. Cardiac disease
B. Severe nutritional edema
C. Superior vena cava obstruction
D. Ventriculoperitoneal shunt
E. Peritoneal dialysis

62. What is the **MOST** common symptom of pectus excavatum?
A. Fatigue
B. Chest pain

C. Palpitations
D. Wheezing
E. Decreased exercise tolerance

63. What is the **MOST** common cause of death in asphyxiating thoracic dystrophy?
A. Renal failure
B. Hepatic failure
C. Central nervous system depression
D. Pancreatic insufficiency
E. Respiratory failure

64. What are the **MOST** common mucolytic used in ventilated patients?
A. Dornase alfa
B. N-acetylcysteine
C. Hypertonic saline nebulizer
D. Adequate hydration
E. Glycopyrrolate

65. Ventilated patients may have nutritional needs that are equal to, greater, or lesser than those of comparably aged well children. Excessive growth is as harmful as inadequate growth, and excess calories may lead to
A. increased oxygen consumption
B. increased carbon dioxide (CO2) production
C. overhydration
D. obesity
E. metabolic acidosis

66. A 24-hour-fullterm newborn, appropriate weight for gestational age, product of uneventful pregnancy, developed rapid shallow respiration with frequent respiratory pauses which progress in the next few days to apparent cyanosis and respiratory failure. Capillary blood gas analysis during sleep reveals $PaCO_2$ of 95 mm Hg that decline to normal levels after the infants awaken.
Of the following, the **MOST** likely diagnosis is
A. congenital central hypoventilation syndrome
B. diaphragm paralysis
C. autonomic dysregulation
D. Arnold Chiari malformation

E. hypothalamic dysregulation hypoventilation

67. A 9-month-old infant with history of repaired myelomeningocele, big head, and sleep disordered breathing in the form of obstructive sleep apnea; presented with aphonia, hypoventilation, and bradyarrhythmias.
Of the following, the **MOST** likely diagnosis is

A. hypothalamic dysfunction
B. autonomic dysregulation
C. Arnold-Chiari type II malformation
D. central nervous system (CNS) hemorrhage
E. Dandy Walker malformation

Chapter 18

The Respiratory System Answers

ZUHAIR ALMUSAWI

1.(A) Nonpulmonary Diseases Associated With Clubbing

CARDIAC

Cyanotic congenital heart disease
Bacterial endocarditis
Chronic heart failure

HEMATOLOGIC

Thalassemia
Congenital methemoglobinemia (rare)

GASTROINTESTINAL

Crohn disease
Ulcerative colitis
Celiac disease
Chronic dysentery, sprue
Polyposis coli
Severe gastrointestinal hemorrhage
Small bowel lymphoma
Liver cirrhosis (including α1 -antitrypsin deficiency)
Chronic active hepatitis

OTHER

Thyroid deficiency (thyroid acropachy)
Thyrotoxicosis
Chronic pyelonephritis (rare)
Toxic (e.g., arsenic, mercury, beryllium)
Lymphomatoid granulomatosis
Fabry disease
Raynaud disease, scleroderma
Hodgkin disease
Familial

UNILATERAL CLUBBING

Vascular disorders (e.g., subclavian arterial aneurysm, brachial arteriovenous fistula)
Subluxation of shoulder
Median nerve injury
Local trauma

2.(B)

3.(A) The utility of MRI of the chest is largely limited to the analysis of mediastinal, hilar, and vascular anatomy. Parenchymal structures and lesions are not well evaluated by MRI. Chest CT is particularly useful in evaluating very small lesions (e.g., early metastases, mediastinal and pleural lesions, solid or cystic parenchymal lesions, pulmonary embolism, and bronchiectasis).

4.(A) The most common cause of recurrent or persistent cough in children is airway reactivity (asthma). Because cough receptors also reside in the pharynx, paranasal sinuses, stomach, and external auditory canal, the source of a persistent cough may need to be sought beyond the lungs. Specific lower respiratory stimuli include excessive secretions, aspirated foreign material, inhaled dust particles or noxious gases, cold or dry air, and an inflammatory response to infectious agents or allergic processes.

5.(E) Cough all day, never during sleep is habit cough. Causes of barking, brassy cough are croup, habit cough, tracheomalacia, tracheitis, and epiglottitis.

6.(C)

7.(C) Only sputum specimens containing alveolar macrophages should be interpreted as reflecting lower respiratory tract processes. Sputum eosinophilia suggests asthma, asthmatic bronchitis, or hypersensitivity reactions of the lung, but a polymorphonuclear cell response suggests infection; If most of the cells in sputum are macrophages, postinfectious hypersensitivity of cough receptors should be suspected. Sputum macrophages can be stained for hemosiderin content, which is diagnostic of pulmonary hemosiderosis, or for lipid content, which in large amounts suggests, but is not specific for, repeated aspiration. Rarely, children may expectorate partial casts of the airway, which can be characterized in investigating causes of plastic bronchitis.

8.(D) In a child with intermittent stridor (with wheezing) that accompanies physical activity and is not responsive to asthma therapies, paradoxical vocal cord dysfunction may be of consideration. Paradoxical vocal cord dysfunction may be highly supported by history and confirmed by laryngoscopy during an exercise challenge test if symptoms are successfully elicited. Speech therapy and behavior modification may be therapeutic.

9.(E)

MATERNAL AND ANTENATAL RISK FACTORS

Elevated 2nd trimester serum α-fetoprotein
Smoking
Alcohol use
Drug use (cocaine, heroin)
Nutritional deficiency
Inadequate prenatal care
Low socioeconomic status
Younger age
Lower education
Single marital status
Shorter interpregnancy interval
Intrauterine hypoxia
Fetal growth restriction

INFANT RISK FACTORS

Age (peak 1-4 mo)
Male gender
Race and ethnicity (African American, American Indian, Alaska Native, other minorities)
Growth failure
No breastfeeding
No pacifier (dummy)
Prematurity
Prone and side sleep position
Recent febrile illness (mild infections)
Inadequate immunizations
Smoking exposure (prenatal and postnatal)
Soft sleeping surface, soft bedding
Bed sharing with parent(s) or other children
Thermal stress, overheating
Colder season, no central heating

10.(D) Brief Resolved Unexplained Events (BRUE)

A BRUE (pronounced brew) is an event that occurs in an infant younger than 1 yr that typically lasts less than 30 sec and is described by the observer as a sudden, brief, and now-resolved episode that involved at least one of the following:

– cyanosis or pallor
– absent, decreased, or irregular breathing

- marked change in tone, either hyper- or hypotonia
- altered level of responsiveness

The diagnosis of BRUE applies only to infants who were asymptomatic prior to the event and during evaluation, and when no explanation for the event is found through appropriate history and physical examination.

11.(E) Nasal foreign bodies can go unrecognized for long periods of time because they initially produce few symptoms and are difficult to visualize. First symptoms include unilateral obstruction, sneezing, relatively mild discomfort, and, rarely, pain. Presenting clinical symptoms include history of insertion of foreign bodies (86%), mucopurulent nasal discharge (24%), foul nasal odor (9%), epistaxis (6%), nasal obstruction (3%), and mouth breathing (2%).

12.(C) Cystic fibrosis is the most common childhood cause of nasal polyposis, and up to 50% of CF patients experience obstructing nasal polyposis, which is rare in non-CF children. Therefore, CF should be suspected in any child younger than 12 yr old with nasal polyps, even in the absence of typical respiratory and digestive symptoms.

13.(C)

14.(D) Vitamin C, guaifenesin, and inhalation of warm, humidified air are no more effective than placebo for the treatment of cold symptoms.

15.(E)

MAJOR SYMPTOMS

- Purulent anterior nasal discharge
- Purulent or discolored posterior nasal discharge
- Nasal congestion or obstruction
- Facial congestion or fullness
- Facial pain or pressure
- Hyposmia or anosmia
- Fever (for acute sinusitis only)

MINOR SYMPTOMS

- Headache
- Ear pain, pressure, or fullness
- Halitosis
- Dental pain
- Cough
- Fever (for subacute or chronic sinusitis)
- Fatigue

16.(B) Penicillin is inexpensive, has a narrow spectrum of activity, and has few adverse effects. Amoxicillin is often preferred for children because of taste,

availability as chewable tablets and liquid, and the convenience of once-daily dosing. The duration of oral penicillin and amoxicillin therapy is 10 days. A single intramuscular dose of benzathine penicillin or a benzathine procaine penicillin G combination is effective and ensures compliance.

The use of macrolides and related antibiotics should be restricted to patients who cannot safely receive a β-lactam drug for GAS pharyngitis. Tetracyclines, fluoroquinolones, or sulfonamides should not be used to treat GAS pharyngitis.

17.(D) Clinical manifestations of retropharyngeal abscess are nonspecific and include fever, irritability, decreased oral intake, and drooling. Neck stiffness, torticollis, and refusal to move the neck may also be present. The verbal child might complain of sore throat and neck pain. Other signs can include muffled voice, stridor, respiratory distress, or even obstructive sleep apnea. Physical examination can reveal bulging of the posterior pharyngeal wall, although this is present in <50% of infants with retropharyngeal abscess. Cervical lymphadenopathy may also be present.

Lateral pharyngeal abscess commonly presents as fever, dysphagia, and a prominent bulge of the lateral pharyngeal wall, sometimes with medial displacement of the tonsil.

18.(A)

19.(B) A brief period of air hunger with restlessness may be followed by rapidly increasing cyanosis and coma. Stridor is a late finding and suggests near-complete airway obstruction. Complete obstruction of the airway and death can ensue unless adequate treatment is provided. The barking cough typical of croup is rare. Usually no other family members are ill with acute respiratory symptoms.

20.(D) Spasmodic croup occurs most often in children 1-3 yr of age and is clinically similar to acute laryngotracheobronchitis, except that the history of a viral prodrome and fever in the patient and family are often absent. The cause is viral in some cases, but allergic and other factors may also contribute.

21.(D) Corticosteroids decrease the edema in the laryngeal mucosa through their antiinflammatory action. Oral steroids are beneficial, even in mild croup, as measured by reduced hospitalization, shorter duration of hospitalization, and reduced need for subsequent interventions such as epinephrine administration. Most studies that demonstrated the efficacy of oral dexamethasone used a single dose of 0.6 mg/kg, a dose as low as 0.15 mg/kg may be just as effective. Intramuscular dexamethasone and nebulized budesonide have an equivalent clinical effect; oral dosing of dexamethasone is as effective as intramuscular administration. A single dose of oral prednisolone is less effective; 1 randomized

controlled trial found no difference in the effectiveness of prednisolone 2 mg/kg/day for 3 days versus 1 dose of dexamethasone 0.6 mg/kg.

22.(A) Bacterial tracheitis is an acute bacterial infection of the upper airway that is potentially life-threatening. S. aureus is the most commonly isolated pathogen, with isolated reports of methicillin-resistant S. aureus. S. pneumoniae, S. pyogenes, Moraxella catarrhalis, nontypeable H. influenzae; anaerobic organisms have also been implicated. The mean age is between 5 and 7 yr. There is a slight male predominance.

23.(B) Subglottic stenosis is a narrowing of the subglottic larynx and manifests in the infant with respiratory distress and biphasic or primarily inspiratory stridor. It may be congenital or acquired.

24.(A) Vocal cord paralysis is the third most common congenital laryngeal anomaly that produces stridor in infants and children. Congenital central nervous system lesions such as Chiari malformation, myelomeningocele, and hydrocephalus or birth trauma may be associated with bilateral paralysis.

25.(B) Propranolol was introduced in 2008 and rapidly became the first-line treatment of infantile and subglottic hemangiomas, including in a recent randomized clinical trial comparing it with systemic steroids. The mechanism is thought to be through VEGF or adrenergic vasoconstriction pathways and can involute the lesion in a few days. Typically, treatment is with 1-3 mg/kg/day of propranolol for 4-12 mo, based on clinical monitoring as noted in a 2011 consensus guideline.

26.(E) The diagnosis of acute bronchiolitis is clinical, particularly in a previously healthy infant presenting with a first episode of wheezing following a period of upper respiratory symptoms. Chest radiography is not routinely indicated in children with suspected bronchiolitis. Areas of atelectasis associated with bronchiolitis are often observed on chest radiographs and may be difficult to distinguish from bacterial pneumonia; as a result, obtaining chest radiography in a patient whose clinical course and exam are consistent with bronchiolitis may encourage unnecessary antibiotic use. Laboratory testing is also not routinely indicated; the white blood cell and differential counts are usually normal and are not predictive of bacterial superinfection. Viral testing (polymerase chain reaction or rapid immunofluorescence) is not routinely recommended in the diagnosis of bronchiolitis but may be helpful if such testing prevents more invasive evaluations. Concurrent serious bacterial infection (sepsis, pneumonia, meningitis) is unlikely, although confirmation of viral bronchiolitis may obviate the need for a sepsis evaluation in the young febrile infant.

27.(E) The treatment of children with viral bronchiolitis is supportive management. Hypoxemic children should receive supplemental oxygen. There is a developing consensus surrounding target oxygen saturations; national guidelines in the United States propose a threshold of 90%.

Some children may also require support with supplemental hydration. Fluid can be administered intravenously or enterally via nasogastric tube, with some preference given to the latter due to an association between better outcomes and continued provision of enteral nutrition.

Frequent suctioning of nasal and oral secretions often provides relief of distress and improves work of breathing and ability to feed. Chest physiotherapy has been extensively evaluated and provides no benefit to children with bronchiolitis. Pharmacologic agents have largely proven ineffective in the management of bronchiolitis.

There is debate over the use of hypertonic saline in children with bronchiolitis. Racemic epinephrine has not been found to improve length of stay or clinical outcomes among inpatients with bronchiolitis. Ribavirin is also not currently recommended, because of minimal impact on disease outcomes, and because it is costly, difficulty to administer, and associated with important toxicities.

28.(E) Frequent shifts in position can facilitate pulmonary drainage in infants. Older children are sometimes more comfortable with humidity, but this does not shorten the disease course. Cough suppressants can relieve symptoms but can also increase the risk of suppuration and inspissated secretions and therefore should be used judiciously. Antihistamines dry secretions and are not helpful; expectorants are likewise not indicated. Nonprescription cough and cold medicines should not be used in children younger than 4 yr of age, and their use is cautioned in children age 4-11 yr.

29.(A) Conditions Associated With Plastic Bronchitis

***PROVEN CONDITIONS**

Congenital heart disease with Fontan physiology

Pulmonary lymphatic anomalies

Influenza A pulmonary infection

***POSSIBLE CONDITIONS**

Toxic inhalation

Sickle cell acute chest syndrome

Hypersecretory and near-fatal asthma (eosinophilic casts)

***UNLIKELY AND UNPROVEN CONDITIONS**

Cystic fibrosis

Chronic obstructive pulmonary disease

Bronchiectasis
Bacterial pneumonia

30.(E) The presenting sign of a cervical hernia (Sibson hernia) is usually a neck mass noticed while straining or coughing. Some lesions are asymptomatic and detected only when a chest film is taken for another reason. Findings on physical examination are normal except during Valsalva maneuver, when a soft bulge may be noticed in the neck.

31.(A) The earliest clinical signs of pulmonary edema include increased work of breathing, tachypnea, and dyspnea. As fluid accumulates in the alveolar space, auscultation reveals fine crackles and wheezing, especially in dependent lung fields. In cardiogenic pulmonary edema, a gallop may be present, as well as peripheral edema and jugular venous distention.

32.(D) Chest radiographs can provide useful ancillary data, although findings of initial radiographs may be normal. Early radiographic signs that represent accumulation of interstitial edema include peribronchial and perivascular cuffing. Diffuse streakiness reflects interlobular edema and distended pulmonary lymphatics. Diffuse, patchy densities, the so-called butterfly pattern, represent bilateral interstitial or alveolar infiltrates and are a late sign. Cardiomegaly is often seen with cardiogenic causes of pulmonary edema. Heart size is usually normal in noncardiogenic pulmonary edema.

33. (A) In 2 reports, between 26% and 48% of such children were found to have dysphagia with aspiration as the underlying problem.

34.(C) A modified barium swallow study with videofluoroscopy (videofluoroscopic swallowing study) is generally considered the gold standard for evaluating the swallowing mechanism.

35.(D) Bronchoalveolar Lavage is one of the most sensitive tests and very helpful to the clinician in supporting the diagnosis of hypersensitivity pneumonia HP. Lymphocytosis frequently exceeding 50% of the recovered cells is seen on the BAL and should alert the clinician to the possibility of HP.

36.(A) The control of environmental exposure to the offending antigen is a key to curing hypersensitivity pneumonia HP and remains the ideal method of treatment and prevention of recurrence. The clinical and pathologic manifestations of acute and subacute HP are reversible with removal of the offending antigen.

37.(B) The transient pulmonary infiltrates with eosinophilia syndrome that is most often seen in children (formerly known as Löffler syndrome) is characterized by migrating pulmonary infiltrates with peripheral blood

eosinophilia caused by the helminthic infections. A. lumbricoides or roundworm is the most common parasite causing this disease.

38.(B)

39.(D) Infection with certain strains of B. cepacia and other multidrug resistant organisms may be associated with particularly rapid pulmonary deterioration and death.

40.(B) These children may present with hypochloremic alkalosis. Hyponatremia is a risk particularly in warm climates.

41. (B)

WITH FALSE-POSITIVE RESULTS

- ✓ Eczema (atopic dermatitis)
- ✓ Ectodermal dysplasia
- ✓ Malnutrition/failure to thrive/deprivation
- ✓ Anorexia nervosa
- ✓ Congenital adrenal hyperplasia
- ✓ Adrenal insufficiency
- ✓ Glucose-6-phosphatase deficiency
- ✓ Mauriac syndrome
- ✓ Fucosidosis
- ✓ Familial hypoparathyroidism
- ✓ Hypothyroidism
- ✓ Nephrogenic diabetes insipidus
- ✓ Pseudohypoaldosteronism
- ✓ Klinefelter syndrome
- ✓ Familial cholestasis syndrome
- ✓ Autonomic dysfunction
- ✓ Prostaglandin E infusions
- ✓ Munchausen syndrome by proxy

WITH FALSE-NEGATIVE RESULTS

- ✓ Dilution
- ✓ Malnutrition
- ✓ Edema
- ✓ Insufficient sweat quantity
- ✓ Hyponatremia
- ✓ Cystic fibrosis transmembrane conductance regulator mutations with preserved sweat duct function

42.(B) The diagnosis of pancreatic malabsorption can be made by the quantification of elastase-1 activity in a fresh stool sample by an enzyme-linked

immunosorbent assay specific for human elastase. The quantification of fat malabsorption with a 72-hr stool collection is rarely necessary in the clinical setting.

43.(C) Standard pulmonary function studies are usually obtained starting at about 4 yr of age and are routinely done by age 6. Forced expiratory volume in 1 sec (FEV1) is the measurement that has been shown to correlate most closely with mortality and shows a gradual decline averaging 2–3% per year throughout childhood.

44.(D) The quinolones are the only broadly effective oral antibiotics for Pseudomonas infection, but resistance against these agents may emerge. Macrolides may reduce the virulence properties of P. aeruginosa, such as biofilm production, and contribute antiinflammatory effects. Long-term therapy with azithromycin 3 times a week improves lung function in patients with chronic P. aeruginosa infection.

45.(C) Aerosolized tobramycin inhalation solution or powder, or aztreonam inhalation solution used as a suppressive therapy (on 1 mo, off 1 mo), may reduce symptoms, improve pulmonary function, and decrease the occurrence of pulmonary exacerbations.

46.(A) Macrolide antibiotics have an antiinflammatory effect, and 3 days/wk azithromycin has been shown to reduce the likelihood of development of pulmonary exacerbations, especially in patients with chronic Pseudomonas airway infection, so this is a commonly used therapy.

47.(A) Allergic bronchopulmonary aspergillosis occurs in 5–10% of patients with CF. The presence of rust-colored sputum, the recovery of Aspergillus organisms from the sputum, a positive skin test for A. fumigatus , the demonstration of specific IgE and IgG antibodies against A. fumigatus, or the presence of eosinophils in a fresh sputum sample supports the diagnosis.

Treatment is directed at controlling the inflammatory reaction with oral corticosteroids. Oral antifungals are usually reserved for patients who relapse after initial steroid treatment. For refractory cases, omalizumab, humanized monoclonal anti-IgE, has been effective.

48.(B) Because pancreatic insufficiency results in malabsorption of fat-soluble vitamins (A, D, E, K), vitamin supplementation is recommended.

49.(A) PCD typically has autosomal recessive patterns of inheritance, although X-linked inheritance has been reported.

50.(D) Primary ciliary dyskinesia PCD has several characteristic clinical features. Neonatal respiratory distress (NRD) is a common feature, and most affected

term newborns develop increased work of breathing, tachypnea, and upper and middle lobe atelectasis on chest radiographs.

51.(D) A few case reports have associated neonatal hydrocephalus with PCD. The ependyma of the brain ventricles are lined by ciliated epithelium and are important for cerebrospinal fluid flow through the ventricles and aqueduct of Sylvius. The finding of enlarged brain ventricles on sonograms, when linked with situs inversus totalis, has been proposed as a prenatal diagnostic marker for PCD.

52.(C)

53.(A) Pulmonary hemorrhage is classically associated with a microcytic, hypochromic anemia. Reductions of serum iron levels, decreased or normal total iron-binding capacity, and normal to increased ferritin levels may be found with chronic disease. An elevated erythrocyte sedimentation rate is a nonspecific finding. The reticulocyte count is frequently elevated.

54.(C) In IPH, systemic corticosteroids are frequently utilized as first-line treatment and are expected to be of particular benefit in the setting of immune-mediated disease. Steroids modulate neutrophil influx and the inflammation associated with hemorrhage; consequently, they may decrease progression toward fibrotic disease.

55.(D) The most common identified risk factors in children include infection, congenital heart disease, and the presence of an indwelling central venous catheter.

56.(B) Although a ventilation–perfusion ($\dot{V} - \dot{Q}$) radionuclide scan is a noninvasive and potentially sensitive method of pulmonary embolus detection , the interpretation of $\dot{V} - \dot{Q}$ scans can be problematic. Helical or spiral CT with an intravenous contrast agent is valuable and the diagnostic test of choice to detect a PE.

57.(A) Side effects include heparin-induced thrombocytopenia as well as bleeding and osteoporosis.

58.(C) In contrast to atelectasis in adult patients, in whom the lower lobes and, in particular, the left lower lobe are most often involved, 90% of cases in children involve the upper lobes and 63% involve the right upper lobe.

59.(A)

60.(D) Metastatic lesions are the most common forms of pulmonary malignancy in children; primary processes include Wilms tumor, osteogenic sarcoma, and hepatoblastoma.

61.(A) Hydrothorax is usually bilateral, but in cardiac or hepatic disease it can be limited to the right side or greater on the right than on the left side.

62.(E) The deformity is present at or shortly after birth in one-third of cases but is usually not associated with any symptoms at that time. In time, fatigue, chest pain, palpitations, recurrent respiratory infections, wheezing, stridor, and cough may be present. Decreased exercise tolerance is one of the most common symptoms.

63.(E) Most patients with this disorder die shortly after birth from respiratory failure.

64.(C) Hypertonic saline is the most common mucolytic, but a number of other agents have been tried, such as dornase alfa and N-acetylcysteine.

65.(B) Excessive growth is as harmful as inadequate growth, and excess calories may lead to increased carbon dioxide (CO_2) production.

66.(A)

67.(C) Arnold-Chiari type II malformation is associated with myelomeningocele, hydrocephalus, and herniation of the cerebellar tonsils, caudal brainstem, and the 4th ventricle through the foramen magnum. Sleep disordered breathing, including OSA and hypoventilation, has been reported. Direct pressure on the respiratory centers or brainstem nuclei or increased intracranial pressure because of the hydrocephalus may be responsible. Vocal cord paralysis, apnea, hypoventilation, and bradyarrhythmias have also been reported. Patients with Arnold-Chiari type II malformation have blunted responses to hypercapnia, and, to a lesser degree, hypoxia.

Chapter 19

The Cardiovascular System Questions

AHMED TAWFIQ

1. During fetal life, the umbilical venous partial pressure of O_2 is about
   A. 20-25 mmhg
   B. 30-35 mmhg
   C. 50-55 mmhg
   D. 60-65 mmhg
   E. 70-75 mmhg

2. What is the percentage of right ventricular output that enters to the lung during fetal life?
   A. 5%
   B. 10%
   C. 15%
   D. 20%
   E. 35%

3. A 10-day-old full-term boy has been diagnosed with large VSD; you had informed parents that close follow-up is essential. Heart failure would be expected at the time of
   A. starting decrease pulmonary vascular resistance
   B. nadir of normal physiological anemia of infancy
   C. pulmonary hypertension
   D. any coincident upper respiratory tract infection
   E. at onsent of lower respiratory tract infections

4. A 7-year-old boy with inoperable complex cyanotic heart disease, complaining of headache and occasional chest pain for the last one month. His current CBC shows HB of 21.5 gm/dl and PCV of 66, his serum ferritin 85mg/dl. Of the following, the **NEXT** step in the management would be
   A. follow up visit after 2 weeks for another workup
   B. advice about appropriate hydration and stop diuretics medications

C. partial exchange transfusion
D. iron therapy
E. reassurance

5. You are preparing a child with inoperable cyanotic heart disease for partial exchange transfusion. You would replace the aspirated blood with
A. 5% water dextrose
B. 1/2 glucose saline
C. normal saline
D. fresh whole blood with lower PCV
E. fresh frozen plasma

6. Which of the following is an indication for screening fetal echocardiography?
A. All pregnant women
B. Gestational diabetes on oral hypoglycemic agent
C. Suspecting fetal chromosomal abnormality
D. History of exposure to teratogens during late pregnancy
E. 2^{nd} degree relative with congenital heart disease

7. A 7-year-old child with familial hypertrophic cardiomyopathy is being evaluated with cardiac exercise testing.
Which of the following results is considered as abnormal?
A. ST segment elevation > 2 mm for at least 0.2 second
B. Q wave > 4 mm
C. Appearance of U wave
D. ST segment depression of > 2 mm for at least 0.06 second
E. An increase in blood pressure before maximal exercise is reached

8. A 10-day-old boy with a recent diagnosis of D-TGA with patent foramen ovale and intact interventricular septum develops increasing cyanosis. In order to decrease cyanosis and improve the general condition until total surgical correction; the appropriate action at this moment is to
A. start prostaglandin E_1
B. do stent procedure to reopen ductus arteriosus
C. do atrial septostomy
D. start high flow nasal O_2
E. arrange for home nasal CPAP

9. A 7-year-old boy with fully corrected tetralogy of Fallot (TOF), on regular visit to cardiology clinic, he had increasing complaint of orthopnea and shortness of breath. Echocardiography assessment shows significant pulmonary insufficiency (pulmonary regurgitation).
Of the following, the **MOST** appropriate management is to

A. offer an open heart surgery with valve replacement
B. give antifailure medications as long term therapy
C. offer balloon dilatation of distal branch artery stenosis
D. advice reassurance and follow-up
E. use melody valve

10. What is the **MOST** common congenital heart disease?

A. Tetralogy of Fallot
B. Transposition of great arteries
C. Atrial septal defect
D. Ventricular septal defect
E. Pulmonary stenosis

11. Which of the following congenital heart diseases can cause fetal hemodynamic disturbance and hydrops?

A. Ebstein anomaly
B. Pulmonary atresia
C. D-transposition of great arteries
D. Severe type of hypoplastic left heart syndrome
E. Patent ductus arteriosus

12. A young parents are feeling guilty and want to know the cause of their baby heart problem (ventricular septal defect).
Of the following, the **MOST** reasonable answer is

A. Most cases of congenital heart disease are multifactorial
B. It is related to racial factors and chromosomal disorders
C. It runs in certain families with unknown inheritance pattern
D. It follow classical mendelian inheritance
E. The cause of most heart defect is still unknown

13. A 3-day-old baby boy admitted with poor feeding and recurrent seizures, he had receding chin, cleft palate, heart murmur, and absent thymic shadow on chest x-ray.

Of the following, the **MOST** helpful diagnostic test is

A. total and ionized calcium
B. blood gas analysis
C. chromosomal karyotyping
D. fluorescence in situ hybridization analysis
E. echocardiography

14. Which of the following congenital heart defects may be associated with CATCH 22 syndromes?

A. Perimemranous VSD
B. Tetralogy of Fallot
C. Double inlet single ventricle
D. D- transposition of great arteries
E. Pulmonary atresia

15. You are assessing a full-term boy in the postnatal ward with features of DiGeorge syndrome, father (who is a pharmacist) is highly concerned as he had previously similar affected baby who died because of recurrent infections. Currently the baby is stable.
Of the following, the **MOST** appropriate way for genetic counseling should be done by

A. sending the baby for FISH analysis
B. sending the baby for chromosomal karyotyping
C. sending the baby and parents for FISH analysis
D. reassurance of the family as the course of illness may be mild
E. sending for metabolic screen

16. You received a call from staff nurse about the result of routine screening pulse oximetry saturation for asymptomatic 24-hour-old full-term baby in postnatal ward. The staff nurse reported that the result of oximetry as follow ; Right hand 92%, left foot 90% in 2 occasions. You have evaluated the baby and found no murmur, pulses and pressure between upper limbs and lower limbs have marginal difference.
Of the following, the **MOST** appropriate next step is to

A. discharge the baby home with reassurance
B. discharge the baby home with follow-up appointment after 3 days
C. send for erect chest X-ray
D. send for an ECG

E. send for an echocardiography

17. Which of the following congenital heart lesions is ductal dependent?
A. Tetralogy of Fallot
B. Pulmonary atresia
C. D-transposition of great arteries with VSD
D. L-transposition of great arteries
E. ASD with pulmonary stenosis

18. Which of the following congenital heart diseases can cause pressure overload?
A. Dilated cardiomyopathy
B. VSD
C. Ebstein anomaly
D. Atrioventricular septal defects
E. Coarctation of the aorta

19. Which of the following statements is true regarding patent foramen ovale?
A. It is a recognized type of atrial septal defects
B. It usually causes significant hemodynamic left-right shunt
C. It can persist in up to 50% of adults
D. Device closure is needed in case of thromboembolic stroke
E. Left-right shunt is common beyond neonatal period

20. A 4-year-old girl with asymptomatic small ASD secundum, echocardiography shows no dilatation of cardiac chambers. Family concerned about the timing of closure of the defect.
What is the **MOST** appropriate answer?
A. The best time for closure is now
B. We need to watch the girl closely anticipating the complications
C. We need not to worry as it is a small defect and closure is not indicated
D. The closure is needed before school entry even it is small to prevent Eisenmenger syndrome
E. We can wait till adolescent period when her weight will be good enough to bear operation

21. A 3-year-old girl diagnosed as a case of large ASD secundum, not fit for transcatheter closure and need surgery. Echocardiography shows right atrial

and right ventricular dilatation. Family shows no much interest in repairing the defect as they see their child has no much symptoms; they want to postponed it till adulthood.
What is your **BEST** advice for the family?

A. It is better to be done as early as possible
B. There is good chance for spontaneous closure; so I would agree to wait till adulthood
C. We can wait as long as she is well and having normal growth parameters
D. We can do appointment echocardiography every 3 months for close follow-up
E. The best period for closure is just before adolescence

22. Which of the following structures remains normal in size in patients with large ASD?

A. Pulmonary artery
B. Right atrium
C. Left atrium
D. Right ventricle
E. Left ventricle

23. Which of the following is the characteristic finding in a child with moderate–large size ASD secundum?

A. Right ventricular systolic lift at left sternal border
B. Grade 3/6 systolic ejection murmur at pulmonary area
C. Wide and fixed splitting of S2
D. Wide splitting of S2 during inspiration
E. pulmonic ejection click

24. Scimitar syndrome represents the anomalous connection between one of the pulmonary veins and

A. superior vena cava
B. inferior vena cava
C. right atrium
D. left atrium
E. pulmonary artery

25. Which type of ASDs is **MOSTLY** associate with partial anomalous pulmonary venous return?

A. Sinus venosus
B. Secundum
C. Common canal (atrioventricular)
D. Primum
E. Patent foramen ovale

26. What is the **MOST** common cardiac defect associated with ostium primum?
A. Gooseneck deformity of mitral valve
B. Cleft tricuspid valve
C. Cleft mitral valve
D. Supracristal type of VSD
E. Sinus venosus type of ASD

27. What is the ideal time of surgical closure of an atrioventricular septal defect in Down syndrome?
A. Neonatal period
B. 3-6 months of life
C. Late infancy
D. Preschool age
E. During second decad

28. In babies affected by atrioventricular septal defects; the associated Gerbode shunt can occur between
A. left atrium to right atrium
B. right atrium to left atrium
C. left atrium to right ventricle
D. left ventricle to right atrium
E. left ventricle to right ventricle

29. The VSD type that is **MOST** commonly associated with aortic regurgitation is
A. supracristal
B. membranous
C. muscular
D. swiss cheese
E. inlet

30. In which type of ventricular septal defects, an aneurysm can form limiting left-to-right shunt?

A. supracristal
B. membranous
C. muscular
D. swiss cheese
E. inlet

31. You are explaining the role of PDA in a case of pulmonary atresia to your junior colleague.
The **MOST** appropriate statement is that
A. absence of PDA is such lesion reduces the risk of surgery
B. it represents the only source of pulmonary blood flow
C. pharmacological trial for it is closure is indicated
D. the smaller the size of PDA; the better course of disease
E. heart failure is usually secondary to persistence of such lesion

32. Which of the following conditions simulate clinically patent ductus arteriosus?
A. Coronary artery fistula
B. Large ASD
C. Severe pulmonary stenosis
D. Pulmonary stenosis with ASD
E. Congenital mitral stenosis

33. A 3-year-old boy with mild developmental delay referred to cardiology clinic with heart murmur. He has hypertelorism, down-slanting eyes, webbed neck, short stature, and chest deformity.
What is the **MOST** likely cardiac lesion associated with this condition?
A. Supravalvular aortic stenosis
B. ASD
C. VSD
D. Coaractation of aorta
E. Pulmonary stenosis

34. A 12-day-old baby boy who had uneventful perinatal history with smooth delivery, presents with increasing feeding difficulties, cyanosis, cardiac murmur, and hepatomegaly. Chest X-ray shows cardiac prominence of the right ventricle and right atrium with decrease intrapulmonary vascularity. ECG shows peaked R waves in right chest leads, with tall spiked P waves.

Of the following, the **MOST** appropriate treatment for this condition is

A. to start high flow nasal O_2
B. to start nasal CPAP
C. to start PG E1 infusion
D. to start antifailure medications
E. to arrange for balloon valvuloplasty

35. What is the **MOST** common cardiac anomaly that is associated with coaractation of aorta?

A. Bicuspid aortic valve
B. Parachute mitral valve
C. Subaortic stenosis
D. Anomalous right subcalvian artery
E. VSD

36. A 4-year-old boy underwent elective surgical correction for coaractation of aorta, as he recovers from anesthesia; he develops severe abdominal pain, vomiting and hematochezia, his blood pressure 130/95 mmhg, HB 14 gm/dl, and WBC 24,000/mm^3.

What is the **MOST** likely cause?

A. Malignant hypertensive crises
B. Acute peritonitis
C. Mesenteric arteritis
D. Fibrinous adhesions
E. Thrombo-embolic complication

37. A 5-year-old boy, a known case of coaractation of aorta, developed sudden very severe headache, nausea, vomiting, blurred vision, photophobia, with gradual deterioration in the level of consciousness which ended with coma. Examination shows stiff neck, B.P 126/86.

Of the following, the **MOST** likely diagnosis is

A. malignant hypertensive crises
B. ruptured cranial aneurysm
C. thrombotic cerebrovascular accident
D. acute meningoencephalitis
E. brain abscess

38. An 18-month-old boy had history of recurrent wheezing, frequent hospital admissions, and failure to gain weight. Examination reveals pallor, mild tachypnea loud and split S2. Left atrial and right ventricular enlargements with pulmonary congestion are seen in chest X-ray, and the ECG shows bifid P wave.
Of the following, the **MOST** likely diagnosis is

A. large ASD secundum type
B. moderate primum ASD
C. coaractation of aorta
D. interrupted aortic arch
E. mitral stenosis

39. A 12-week-old baby boy, a known case of congenital heart disease had history of wheezing and recurrent lung collapses. Examination shows underweight baby, O_2 saturation 72%, tachypnea and tachycardia, and diastolic murmur at left upper sternal border.
Of the following, the **MOST** likely diagnosis is

A. critical pulmonary stenosis
B. Ebstein anomaly
C. tricuspid atresia
D. absence of pulmonary valve
E. atrioventricular septal defect

40. An 8-year-old girl presents with occasional left sided chest pain and palpitation, examination shows a healthy girl with apical late systolic murmur preceded by a click, more prominent in sitting position, her chest X-ray is normal and ECG shows biphasic T waves in leads aVF and V6, with occasional unifocal premature ventricular contractions.
Of the following, the **MOST** likely diagnosis is

A. mitral valve stenosis
B. ventricular ectopics
C. mitral valve prolapse
D. tricuspid valve regurgitation
E. small ventricular septal defect

41. What is the finding that is considered as a risk factor for graft dysfunction in cardiac transplantation?

A. Tricuspid regurgitation
B. Mitral regurgitation

C. Aortic regurgitation
D. Pulmonary regurgitation
E. Thrombo-embolic phenomena

42. An 18-month-old boy who underwent open heart surgery for repair of tetralogy of Fallot; developed increased needs for postoperative narcotics that is associated with sweating and irritability. Lab studies shows increased AST, ECG shows ST elevation in left chest leads, and echocardiography assessment is pending.
Of the following, the **MOST** likely cause is
A. transient heart block
B. postcardiomyotomy syndrome
C. interruption of an aberrant coronary artery
D. significant residual VSD with left-to-right shunt
E. right ventricular failure

43. You received a call from a colleague in remote area describing a 5-day-old neonate who is having cardiac murmur, cyanosis, cardiomegaly, and an ECG shows left axis deviation with bifid P waves. No availability of echocardiography in that remote hospital.
Of the following, the **MOST** likely diagnosis is
A. tricuspid atresia
B. pulmonary atresia
C. atrioventricular septal defects
D. mitral atresia
E. transposition of great arteries

44. You are counseling parents of a 21-day-old boy who underwent pulmonary artery banding for tricuspid atresia and large VSD; he is stable now, showing no signs of heart failure, and start to gain weight.
They are asking about the ideal time of next operation for their baby.
Of the following, the **BEST** response will be
A. during first 2-6 months of life to avoid LV dysfunction
B. as late as 10-12 months to gain adequate weight to stand operation
C. during second year of life
D. 3-4 years is the ideal time to do total correction
E. no time limit is important as we are keeping close follow-up for the baby

45. Congenital heart diseases may have characteristic shape on radiographic examination.
Which of the following is usually seen in Ebstein anomaly?

A. Egg on a string
B. Boot shape
C. Snow man appearance
D. Box shape
E. Gooseneck shape

46. You are explaining the reason behind early surgical correction in D-TGA to a junior colleague. Which of the following is the **BEST** statement describing that reason?

A. Intervention should be started before pulmonary hypertension become evident
B. For early correction of associated coronary artery anomalies
C. Left ventricular mass may become less with time
D. Early intervention results in better future growth parameters
E. Better neurological prognosis is observed with early intervention

47. What is the procedure of choice for correction of D-transposition of great arteries?

A. Arterial switch
B. Atrial switch
C. Ventricular switch
D. Sano
E. Norwood

48. The clinical course of patients with double-outlet right ventricle without pulmonary stenosis is usually similar to

A. VSD with a large left-to-right shunt
B. pulmonary atresia
C. hypoplastic left heart syndrome
D. tricuspid atresia
E. D-transposition of great arteries

49. What is the **MOST** common type of total anomalous pulmonary venous return that is associated with obstruction?

A. Supracardiac type

B. Coronary sinus type
C. Mixed type
D. Infracardiac type
E. Right atrial type

50. A full-term baby boy, product of uneventful normal vaginal delivery, soon after birth developed increasing cyanosis and respiratory difficulty, chest X-ray shows a very dramatic perihilar pattern of pulmonary edema and a small heart, blood gas analysis shows hypoxemia and hypercapnia. Mother attended antenatal period regularly with no history of maternal illnesses.
Of the following, the **MOST** likely diagnosis is
A. persistent pulmonary hypertension
B. respiratory distress syndrome
C. total anomalous pulmonary venous return with obstruction
D. congenital group B streptococcal pneumonia
E. hypoplastic left heart syndrome

51. A full-term baby boy, product of uneventful normal vaginal delivery, soon after birth develops increasing cyanosis and respiratory difficulty, chest X-ray shows a very dramatic perihilar pattern of pulmonary edema and a small heart, blood gas analysis shows hypoxemia and hypercapnia. Mother attended antenatal period regularly with no history of maternal illnesses.
Of the following, the **MOST** appropriate action is to
A. start prostaglandin E1 infusion
B. escalate FiO2 accordingly
C. start antifailure treatment
D. refer for urgent surgical intervention
E. start mechanical ventilation

52. A 2-month-old infant, has history of mild tachypnea and excessive sweating with feeding since birth increasing with time, he presents with moderate-severe respiratory distress, and cyanosis. Examination shows gallop rhythm with systolic murmur along the left sternal border, chest X-ray shows cardiomegaly with perihilar edema, blood gas analysis shows hypoxemia and hypercapnia, color Doppler shows pulmonary veins are draining away from the heart.
Of the following, the **MOST** likely diagnosis is
A. truncus arteriosus
B. large VSD

C. total anomalous venous return
D. single ventricle
E. hypoplastic left heart syndrome

53. A 3-day-old neonate, a product of normal vaginal delivery, presents with sudden change in color and poor feeding. Examination shows webbed neck, poor peripheral pulses, cold extremities, palpable right parasternal lift, and hepatomegaly. ECG shows right ventricular dominance with peaked P wave. Chest X-ray shows plethoric lung with cardiomegaly. Mother was poor attendant to the primary health care.
Of the following, the **MOST** likely diagnosis is
A. tricuspid atresia
B. total anomalous pulmonary venous return
C. L-transposition of great arteries
D. D-transposition of great arteries
E. hypoplastic left heart syndrome

54. Which of the following congenital heart diseases shows encouraging antenatal interventional results?
A. Tricuspid atresia
B. Hypoplastic left heart syndrome
C. Total anomalous pulmonary venous return
D. Severe coaractation of aorta
E. D-transposition of great arteries

55. A 7-month-old boy has history of intermittent vomiting, recurrent wheezing started soon after birth that is exacerbated by crying, feeding, or flexion of the neck. The mother informs you that extension of the neck tends to relieve the noisy respiration. Past medical history reveals mild atopy and history of hospital admission twice because of pneumonia.
Of the following, the **MOST** likely diagnosis is
A. GERD
B. Sandifier syndrome
C. severe atopic asthma
D. vascular ring
E. lobe sequestration

56. A 5-week-old girl brought to you complaining from recurrent bouts of crying, pallor and sweating, the mother describes her baby as "nothing make him comfortable", during examination you find a borderline growth parameters, gallop rhythm, pansystolic murmur mainly at apex, cardiomegaly on chest X-ray, and ST segment elevation and deep Q wave in V5/V6 in ECG, echocardiography shows dilated LV with reduced ejection fraction.
Of the following, the **MOST** likely diagnosis is

A. coaractation of aorta
B. mitochondrial cardiomyopathy
C. dilated cardiomyopathy
D. large VSD
E. anomalous origin of left coronary artery from pulmonary artery

57. A 6-year-old girl referred to cardiology clinic from asthma clinic for further evaluation because of "treatment failure". She had history of recurrent symptoms of dyspnea, fatigability, chest pain, and syncope for the last year. During examination you find left parasternal heave with loud S_2, increase jugular venous pressure, cardiomegaly on chest X-ray, and tricuspid insufficiency with increase right ventricular pressure on echocardiography. Cardiac catheterization reveals elevated both pulmonary wedge pressure and left ventricular end-diastolic pressure.
Of the following the **MOST** likely cause is

A. primary pulmonary hypertension
B. pulmonary venous stenosis
C. mitral stenosis
D. restrictive cardiomyopathy
E. cor triatriatum

58. Which of the following can develop early pulmonary vascular disease (Eisenmenger)?

A. Atrioventricular septal defect with Down syndrome
B. Moderate VSD with Edward syndrome
C. ASD with Holtoram syndrome
D. ASD with Noonan syndrome
E. Very large VSD with Moderate pulmonary stenosis

59. A 6-year-old intelligent child with small ASD brought for follow-up in cardiology clinic; mother claiming he is not eating well and he is not interested

in play, these symptoms developed after his grandfather death due to coronary arterial disease. Mother is worried about prognosis of her child.
Of the following, the **BEST** advice/plan is to

A. restrict child physical exercise and encourage intake
B. refer the child to cardiac center for further evaluation
C. emphasize the nature of the child's congenital heart disease
D. call for whole family counseling session
E. make strict rules for the family to protect the child

60. The mother of a 2-month-old infant, who is a known case of cyanotic heart disease, asks whether vaccination is recommended or not.
What is your **BEST** advice?

A. Routine immunization may lead to infective endocarditis
B. We will postpone the immunization till after surgical correction
C. Herd immunity will be the best protection for your child
D. Routine immunization plus influenza vaccine are needed
E. Selected schedule for your baby should be implemented

61. A young parents of a 4-month-old baby boy with tetralogy of Fallot, has scheduled operation for total correction at the age of 8 months, they have plan to visit mountainous area, they are asking about any expected precautions to be taken.
What is your **BEST** advice?

A. It is safe journey as all airplanes are pressurized
B. It is better to put the baby on antibiotic prophylaxis during the journey time
C. It is the time to travel now as there is still 4 months to surgery
D. Travel but take care of environmental precautions
E. Postpone the travel till after corrective surgery

62. A 13-year-old girl presents with chest pain after strenuous exercise; with completely normal physical examination, assessed by family doctor who order an ECG, chest X-ray and echocardiography; all shows normal results apart from accidental finding of tiny PDA.
Of the following, the **BEST** advice is to

A. go forward for Holter monitoring assessment
B. refer to pediatric cardiologist for closure of PDA
C. start decongestive cardiac treatment

D. describe simple analgesia and reassurance about cardiac lesion
E. ensure regular follow-up for cardiac lesion on 6-monthly bases

63. A 7-year-old boy referred to cardiac clinic because of chest pain. His ECG is normal and echocardiography shows bicuspid aortic valve with normal other cardiac structures. Father is worried and asking about the prognosis.
What is your **BEST** answer?

A. This is normal finding and "nothing to worry about"
B. Annual checkup is recommended
C. Life expectancy is usually affected
D. Bacterial endocarditis is the most frequent complication
E. He may have some degree of stenosis during adulthood

64. A 4-year-old girl with history of PDA closure by catheter intervention 2 years ago; referred from dentist asking for antibiotic prophylaxis of bacterial endocarditis before proceeding to a filling of premolar tooth.
What is the **MOST** appropriate answer?

A. This is a low risk prophylaxis need only oral amoxicillin
B. This is a high risk prophylaxis and need parental ampicillin and gentamicin
C. Dental prophylaxis is no longer recommended for all congenital heart diseases
D. Prophylaxis is needed right now, but it's not recommended after 5 years from catheter intervention
E. No prophylaxis is needed any more for her

65. Which of the following is the **MOST** common form of cardiomyopathy?

A. Restrictive
B. Hypertrophic obstructive
C. Hypertrophic non-obstructive
D. Dilated
E. Non-compaction

66. What is the **MOST** common etiology of dilated cardiomyopathy?

A. Idiopathic
B. Genetic
C. Metabolic
D. Viral

E. Ischemic

67. A 2-month-old girl presents with 2 weeks history of feeding and respiratory difficulties with excessive sweating. Examination shows tachypnea, tachycardia, poor peripheral pulses, gallop rhythm, and hepatomegaly; cardiomegaly on chest X-ray and dilated LV with poor ejection fraction on echocardiography. Routine investigational workup shows hypoglycemia, and increase liver transaminases.
Of the following, the **MOST** likely etiology is

A. genetic
B. metabolic
C. viral
D. mitochondrial
E. ischemic

68. A 14-month-old boy presents to emergency room with mild respiratory distress. Examination shows RR 65/min, HR 170/min, BP 50/30, poor peripheral pulses, gallop rhythm, and hepatomegaly. Cardiomegaly on chest X-ray and dilated LV with poor ejection fraction on echocardiography. Supportive measures started soon in emergency room.
Of the following, the **MOST** appropriate step is to start

A. valsartan
B. captopril
C. carvedilol
D. dopamin
E. digoxin

69. Which of the following is the drug of choice for hypertrophic cardiomyopathy?

A. Diuretics
B. β-Adrenergic blocking agents
C. Digoxin
D. Angiotensin-converting enzyme (ACE) inhibitors
E. Angiotensin receptor blockers

70. A 7-year-old child referred by school doctor complaining from recurrent episodes of chest pain, palpitation, and occasional syncope. The child is doing well in school and quite intelligent. During examination you find an overactive

precordial impulse, systolic ejection murmur in the aortic region not associated with an ejection click, and an apical blowing murmur. Referring doctor mentions in the referring letter that there is family history of sudden death of his sib during football play.
Of the following, the **MOST** likely diagnosis is

A. severe aortic stenosis
B. severe pulmonic stenosis
C. coaractation of aorta
D. dilated cardiomyopathy
E. hypertrophic obstructive cardiomyopathy

71. A 6-year-old girl had history of recurrent episodes of cough, mild shortness of breath, chest pain, and occasional syncope in the last 12 months, all episodes were treated symptomatically. Examination shows gallop rhythm, hepatomegaly, and mild pitting pedal edema. Mild cardiomegaly on chest X-ray, with dilated atria on echocardiography.
Of the following, the **MOST** likely diagnosis is

A. dilated cardiomyopathy
B. hypertrophic obstructive cardiomyopathy
C. severe aortic stenosis
D. pulmonary hypertension
E. restrictive cardiomyopathy

72. What are the **MOST** common pediatric cardiac tumors?

A. Rhabdomyomas
B. Fibromas
C. Myxomas
D. Sarcomas
E. Metastatic tumors

73. Which of the following is an ominous complication in patients with acute myocarditis?

A. Shock
B. Arrhythmia
C. Thrombo-embolism
D. Congenstive heart failure
E. Dilated cardiomyopathy

74. You are starting digitalization for a baby with heart failure. Basic metabolic, electrolyte, and an ECG strip were done before the first digitalization dose. Now, after 8 hours you are willing to give the 2nd digitalizing dose.
Of the following, the **MOST** appropriate next step is to

A. repeat echocardiography
B. measure digoxin level
C. repeat ECG strip
D. check renal function tests
E. repeat serum potassium

75. Which of the following electrolyte disturbances can increase digoxin toxicity?

A. Hypokalemia
B. Hypocalcemia
C. Hyponatremia
D. Hyperkalemia
E. Hypernatremia

76. You are managing a 2-year-old boy who develops heart failure after an open cardiac surgery for tetralogy of Fallot. He has only minor response to dopamine.
Of the following, the **MOST** appropriate next step is to add

A. epinephrine
B. isoproterenol
C. milrinone
D. digoxin
E. furosemide

77. Which of the following inotropic medications used for cardiogenic shock can increase urine output when given at low doses?

A. Epinephrine
B. Isoproterenol
C. Milrinone
D. Dopamin
E. Dobutamine

78. A 6-year-old boy presents to emergency room with SVT which is controlled successfully by 2nd dose of adenosine. Post episodic ECG reveals short P-R

interval and slow upstroke of the QRS. His echocardiography came normal. Antecedent history reveals syncope.
Of the following, the **MOST** appropriate next action is to

A. start digoxin therapy
B. start amiodarone prophylaxis
C. start propranolol
D. order Holter monitor
E. refer for electrophysiological study

79. A 6-month-old girl presents to emergency room with history of irritability, excessive crying, and off-color appearance. Her pulses is intact and capillary refill is <2 seconds, cardiac monitor shows narrow complex tachycardia with heart rate of 220/min. you are unsuccessful to convert the rhythm with a trial of an ice pack over the face.
Of the following, the **MOST** appropriate next step is

A. carotid massage
B. adenosine
C. verapamil
D. propranolol
E. synchronized DC cardioversion

80. A 3-year-old boy who presents to emergency room with SVT successfully converted to sinus rhythm after 3 doses of adenosine. His post episodic ECG and echocardiography are normal.
Of the following, the **MOST** appropriate next step is to

A. send for Holter monitoring
B. send for electrophysiological study
C. start digoxin
D. start amiodarone
E. start propranolol

81. A 5-year-old boy who is a known case of WPW syndrome on amiodarone treatment for the last 3 months came for follow-up.
Of the following, the **MOST** likely indicated investigation is

A. thyroid function test
B. renal function test
C. Hb A1c
D. echocardiography

E. ECG

82. You are on duty in PICU when you notice rhythm conversion in a 9-month-old infant with acute myocarditis. You noticed an emergence of regular wide QRS complex tachycardia. Vital signs are; HR 180 beat/min, B.P 80/60 mmHg with good peripheral pulses.
Of the following, the **MOST** appropriate management is to

A. start CPR
B. give DC defibrillation
C. give synchronized DC cardioversion
D. start amiodarone
E. start dysopyramide

83. Which of the following antibiotics can cause prolong QT interval?

A. Cefuroxime
B. Trimethoprim/sulfamethoxazole
C. Amoxicillin-clavulanic acid
D. Ceftriaxone
E. Tobramycin

84. All the following medications can cause prolonged QT interval **EXCEPT**

A. fluconazole
B. imipramine
C. domperidone
D. ondansetron
E. metocloporamide

85. A 5-year-old boy brought to emergency room after he experienced syncopal episode after exercise. The patient had been diagnosed as seizure disorder and he is on valproate for previous 2 similar episodes. His older sib who was described as smart swimmer died as a result of drowning. His ECG shows a heart rate–corrected QT interval of 0.5 sec.
Of the following, the **MOST** likely diagnosis is

A. LQTS type 1 (**LQT1**)
B. LQTS type 2 (**LQT2**)
C. LQTS type 3 (**LQT3**)
D. 2nd degree heart block
E. complete heart block

86. A 4-year-old boy with tetralogy of Fallot and B-T shunt had dental procedure 1 week ago. Now he has spikes of high grade fever, rigors, and splenomegaly. Echocardiography shows moderate size vegetation attached to pulmonary valve.
Of the following, the **MOST** likely causative organism is

A. Staphylococcus aureus
B. Streptococcus viridans
C. group D enterococci
D. Streptococcus pneumonia
E. Coagulase-negative Staphylococci

87. First episode of acute rheumatic carditis often results in

A. complete healing of the valvular lesions
B. chronic rheumatic heart disease
C. subacute prolonged carditis
D. involvement of almost all valves of the heart
E. bacterial endocarditis

Chapter 19

The Cardiovascular System Answers

AHMED TAWFIQ

1.(B) The placenta is not as efficient an oxygen-exchange organ as the lungs, so that umbilical venous partial pressure of oxygen (PO_2), the highest O_2 level provided to the fetus, is only 30-35 mm Hg vs to about 75-100 mmHg in post-natal period. Approximately 50% of the umbilical venous blood enters the hepatic circulation, whereas the rest bypasses the liver and joins the inferior vena cava (IVC) via the ductus venosus, where it partially mixes with poorly oxygenated IVC blood derived from the lower part of the fetal body.

2.(A) Approximately 5% of right ventricular (RV) outflow enters the lungs. The major portion of this blood bypasses the lungs and flows right-to-left through the ductus arteriosus into the descending aorta to perfuse the lower part of the fetal body, including providing flow to the placenta via the 2 umbilical arteries.

3.(B) In acyanotic infants with large left-to-right shunts, the onset of heart failure often coincides with the nadir of the normal physiologic anemia of infancy. Decreasing pulmonary vascular resistance is a dynamic and continuous process starting after birth and continued till few weeks later.

4.(C) In cyanotic patients with inoperable conditions, partial exchange transfusion may be required to treat symptomatic (most often headache or chest pain) individuals whose hematocrit has risen to the 65–70% level. Polycythemic patients with concomitant iron deficiency are at even greater risk for cerebrovascular accidents, and needs treatment with iron but in this scenario serum ferritin is normal.

5.(E) This procedure is not without risk, especially in patients with an extreme elevation in peripheral vascular resistance. Because these patients do not tolerate wide fluctuations in circulating blood volume, blood should be replaced with fresh frozen plasma or albumin.

6.(C) Screening fetal echocardiogram is recommended for women with a previous child or first-degree relative with CHD, for those who are at higher risk of having a child with cardiac disease (e.g., insulin-dependent diabetic patients, women exposed to teratogenic drugs during early pregnancy), and in any fetus in whom a chromosomal abnormality is suspected or confirmed. Mothers with

gestational diabetes need neonatal echocardiography for their babies for checking severity of asymmetrical septal hypertrophy. For fetuses with ductal dependent lesions, delivery can be planned at a tertiary care center, avoiding the requirement for postnatal transport of an unstable infant.

7.(D) The exercise ECG is considered abnormal if the ST segment depression is >2 mm and extends for at least 0.06 sec after the J point (onset of the ST segment) in conjunction with a horizontal, upward, or downward-sloping ST segment. A decrease in blood pressure before maximal exercise is reached is a risk indicator in patients with hypertrophic cardiomyopathy.

8.(C) Balloon atrial septostomy (Rashkind procedure) is the procedure of choice. Option A is good in first few days as the ductus arteriosus is opened. Option B is good for lesions with RVOT obstruction (pulmonary atresia). Options D and E are for respiratory rather than cardiac problem.

9.(E) One of the greatest advances in interventional catheterization over the past decade has been transcatheter valve implantation . Typically, a porcine valve is sewn into an expandable stent, which is then collapsed around a balloon catheter. At this time, the most common application in children is replacement of the pulmonary valve (Melody Valve) in patients who have had prior surgery for tetralogy of Fallot (usually because of residual pulmonary insufficiency) and antifailure medications can be used as short-term therapy meanwhile arrange the procedure.

10.(D) Ventricular septal defect accounts to about 30-35% of congenital heart diseases; atrial septal defect 6-8%; tetralogy of Fallot and pulmonary stenosis 5-7% and lastly transposition of great arteries 3-5%.

11.(A) Most congenital defects are well tolerated in the fetus because of the parallel nature of the fetal circulation. Even the most severe cardiac defects, such as hypoplastic left heart syndrome, can usually be well compensated for by the fetal circulation. One notable exception is the case of severe regurgitant lesions, most frequently of the tricuspid valve. In these lesions, such as Ebstein anomaly of the tricuspid valve or severe right ventricular outflow obstruction, the parallel fetal circulation cannot compensate for the volume load imposed on the right side of the heart.

12.(E) The massage should reach to the parents is that, the cause in most of congenital heart defects is still unknown and parents are not responsible for the problem. Many cases of CHD are multifactorial and results from a combination of genetic predisposition and an as-yet-to-be-determined environmental stimulus. A small percentage of congenital heart lesions are related to known chromosomal abnormalities. Other genetic factors may have a role in CHD; for

example, certain types of VSDs (supracristal) are more common in Asian children.

13.(D) The investigations of choice for management is RBS, total and ionized calcium and echocardiography; but the investigation that leads you to diagnosis in DiGeorge syndrome is fluorescence in situ hybridization (FISH) analysis. Comparative genome hybridization has in many cases replaced routine karyotyping in the clinical workup of newborns with CHD.

14.(B) Question is asking indirectly about conotruncal cardiac defects. Cardiac lesions associated with 22q11.2 deletions are most often seen in association with either the DiGeorge syndrome or the Shprintzen (velocardiofacial) syndrome. The acronym CATCH 22 has been used to summarize the major components of these syndromes: cardiac defects, abnormal facies, thymic aplasia, cleft palate, and hypocalcemia. The specific cardiac anomalies are conotruncal defects (tetralogy of Fallot, truncus arteriosus, double-outlet right ventricle, subarterial VSD) and branchial arch defects (coarctation of the aorta, interrupted aortic arch, right aortic arch). Congenital airway anomalies such as tracheomalacia and bronchomalacia are sometimes present.

15.(C) Although the risk of recurrence is extremely low in the absence of a parental 22q11.2 deletion, it is 50% if 1 parent carries the deletion. In this scenario there is high possibility of one parent is carrier.

16.(E) Screening for duct-dependent cyanotic congenital heart disease is performed between 24 and 48 hr of life and before discharge in asymptomatic newborns. A pulse oximetry saturation of 90–94% in the right hand or either foot requires urgent echocardiography. A pulse oximetry saturation <95% in either location or a saturation difference >3% between the right hand and either foot is considered a positive test and should be repeated in an hour; if positive again, it should be repeated in another hour. If it remains positive, echocardiography is indicated. In addition, a careful reexamination of the pulses and blood pressure in the upper and lower extremities as well as cardiac auscultation are indicated in children with an initial positive screen.

17.(B) Multiple studies demonstrate the benefit of routine pulse oximetry screening for all newborns to detect unsuspected critical cyanotic CHD; lesions include hypoplastic left heart syndrome, pulmonary atresia, tetralogy of Fallot, total anomalous pulmonary venous return, transposition of the great arteries, tricuspid atresia, truncus arteriosus, neonatal coarctation of the aorta, and aortic arch hypoplasia/atresia.

18.(E) The most common lesions resulting in increased volume load are those that cause left-to-right shunting : atrial septal defect (ASD), ventricular septal

defect (VSD), atrioventricular septal defects and patent ductus arteriosus. It also includes regurgitant lesions and the dilated cardiomyopathies. The pathophysiologic common denominator of lesions resulting in increased pressure load is an obstruction to normal blood flow .The most frequent are obstructions to ventricular outflow: valvular pulmonic stenosis, valvular aortic stenosis, and coarctation of the aorta. Less common are obstructions to ventricular inflow: tricuspid or mitral stenosis, cor triatriatum, and obstruction of the pulmonary veins.

19.(D) An isolated valve-incompetent patent foramen ovale (PFO) is a common echocardiographic finding during infancy. It is usually of no hemodynamic significance and is not considered an ASD; a PFO may play an important role if other structural heart defects are present. PFO may be present in 15–30% of adults. An isolated PFO does not require surgical treatment, although it may be a risk for paradoxical (right to left) systemic embolization. Device closure of these defects is one treatment option considered in adults with a history of thromboembolic stroke.

20.(C) In patients with small secundum ASDs and minimal left-to-right shunts without RV enlargement, the consensus is that closure is not required. Infants with small to moderate-sized ASDs can be watched closely, since these defects will often grow smaller in the 1st yr of life. Transcatheter device or surgical closure is advised for all symptomatic patients, as well as for asymptomatic patients with Qp:Qs ratio of at least 2 : 1 and those with RV enlargement. The timing for elective closure is usually after the 1st yr of life and before entry into school.

21.(A) For large defects, repair is preferred during early childhood because surgical mortality and morbidity are significantly greater in adulthood; the long-term risk of arrhythmia caused by chronic atrial dilation is also greater after ASD repair in adults.

22.(E) The increased blood flow through the right side of the heart results in enlargement of the right atrium and ventricle and dilation of the pulmonary artery. The left atrium may also be enlarged as the increased pulmonary blood flow returns to the left atrium, but the left ventricle and aorta are normal in size.

23.(C) All except D are true, but C is characteristic. In most patients with an ASD, the characteristic finding is that the second heart sound (S2) is widely split and fixed in its splitting during all phases of respiration.

24.(B) Occasionally, in PAPVR, an anomalous vein draining into the IVC is visible on chest radiography as a crescentic shadow of vascular density along the right border of the cardiac silhouette (scimitar syndrome).
25.(A) When an associated ASD is present, it is generally of the sinus venosus type but can be secundum.
26.(C) In most cases a cleft in the anterior leaflet of the mitral valve is also noted. The tricuspid valve is usually functionally normal, although some anatomic abnormality of the septal leaflet is present. The ventricular septum is intact.
27.(B) The risk for development of pulmonary vascular disease is greater in patients with Down syndrome, and therefore surgical correction is usually considered early in these patients, within the 1st 3-6 mo of life, while in non-syndromic babies the timing of operation is between 6-12 months.
28.(D) In complete AV septal defects, left-to-right shunting occurs at both the atrial and the ventricular level. Additional shunting may occur directly from the left ventricle to the right atrium (known as a Gerbode shunt) because of absence of the AV septum.
29.(A) A supracristal (subarterial) VSD can be complicated by prolapse of the aortic valve into the defect and aortic insufficiency, which may eventually develop in 50–90% of these patients. Although supracristal VSD accounts for approximately 5% of all patients with VSD, the incidence is higher in Asian children and in males. Although most common with supracristal VSDs, aortic insufficiency is occasionally associated with VSDs located in the membranous septum
30.(B) In defects of the membranous septum , a thin membrane (called a ventricular septal aneurysm but consisting of abnormal tricuspid valve tissue) can partially cover the defect and limit the volume of the left-to-right shunt.
31.(B) PDA is seen in 10% of patients with other congenital heart lesions and often plays a critical role in providing a source of pulmonary blood flow when the right ventricular outflow tract is stenotic or atretic or in providing systemic blood flow in the presence of aortic coarctation or interruption. It has beneficial effects and works as palliation.
32.(A) A congenital fistula may exist between a coronary artery and an atrium, ventricle (especially the right), or pulmonary artery. Regardless of the recipient chamber, the clinical signs are similar to those of PDA, although the machinery-like murmur may be more diffuse.

33.(E) Pulmonary stenosis as a result of valve dysplasia is the most common cardiac abnormality in Noonan syndrome and is associated, in approximately 50% of cases. Surgery is the treatment of choice because of valve dysplasia.

34.(E) In a neonate with critical pulmonic stenosis, urgent treatment by either balloon valvuloplasty or surgical valvotomy is warranted. The condition which can present in similar way is persistent pulmonary hypertension but smooth delivery make this possibility is unlikely.

35.(A) The anomaly occurs twice as often in males as in females. Coarctation of the aorta may be a feature of Turner syndrome and is associated with a bicuspid aortic valve in >70% of patients. Mitral valve abnormalities (a supravalvular mitral ring or parachute mitral valve) and subaortic stenosis are potential associated lesions. When this group of left sided obstructive lesions occurs together, they are referred to as the Shone complex.

36.(C) Postcoarctectomy Syndrome; Postoperative mesenteric arteritis may be associated with acute hypertension and abdominal pain in the immediate postoperative period. The pain varies in severity and may occur in conjunction with anorexia, nausea, vomiting, leukocytosis, intestinal hemorrhage, bowel necrosis, and small bowel obstruction. Relief is usually obtained with antihypertensive drugs (e.g.,nitroprusside, esmolol, captopril) and intestinal decompression; surgical exploration is rarely required for bowel obstruction or infarction.

37.(B) Severe neurologic damage or even death may rarely occur from associated cerebrovascular disease. Subarachnoid or intracerebral hemorrhage may result from rupture of congenital aneurysms in the circle of Willis, rupture of other vessels with defective elastic and medial tissue, or rupture of normal vessels; these accidents are secondary to hypertension.

38.(E) If congenital mitral stenosis is moderate to severe, symptoms usually appear within the 1st 2 yr of life. These infants have failure to thrive and various degrees of dyspnea and pallor. In some patients, wheezing may be a dominant symptom, and a misdiagnosis of bronchiolitis or reactive airway disease may have been made. Heart enlargement because of dilation and hypertrophy of the right ventricle and left atrium is common. Most patients have rumbling apical diastolic murmurs, but the auscultatory findings may be relatively obscure. S2 is loud and split. An opening snap of the mitral valve may be present. The ECG reveals RVH and may show bifid or spiked P waves indicative of left atrial enlargement. Radiographs usually show left atrial and RV enlargement and pulmonary congestion in a perihilar or venous pattern.

39.(D) Congenital absence of the pulmonary valve is usually associated with a ventricular septal defect (VSD), often in the context of tetralogy of Fallot. In many of these neonates, the pulmonary arteries become widely dilated and compress the bronchi, with subsequent recurrent episodes of wheezing, pulmonary collapse, and pneumonitis. The presence and degree of cyanosis are variable. Florid pulmonary valvular incompetence may not be well tolerated, and death may occur from a combination of bronchial compression, hypoxemia, and heart failure

40.(C) Mitral valve prolapse results from an abnormal mitral valve mechanism that causes billowing of 1 or both mitral leaflets, especially the posterior cusp, into the left atrium toward the end of systole. The abnormality is predominantly congenital but may not be recognized until adolescence or adulthood. Mitral valve prolapse is usually sporadic, is more common in girls, and may be inherited as an autosomal dominant trait with variable expression. This lesion is not progressive in childhood, and specific therapy is not indicated. Antibiotic prophylaxis is no longer recommended during surgery and dental procedures.

41.(A) Tricuspid regurgitation is seen in up to 30% of children after heart transplantation, which can be a risk factor for graft dysfunction.

42.(C) Uncommon immediate postoperative problems include RV failure, transient heart block, residual VSD with left-to-right shunting, and myocardial infarction from interruption of an aberrant coronary artery. Scenario shows classical features of myocardial infarction results from interruption of an aberrant coronary artery, the anomaly that is present in about 5-10% of TOF cases.

43.(A) The key for diagnosis in this scenario is association of cyanosis with superior axis or left axis and the age of presentation which fits tricuspid atresia; AV canal have the same ECG finding but neither the age nor the associated cyanosis is fitting.

44.(A) The next stage of palliation for patients with tricuspid atresia involves the creation of an anastomosis between the superior vena cava and the pulmonary arteries (bidirectional Glenn shunt). This procedure is performed usually between 2 and 6 mo of age.

45.(D) The cardiovascular imaging signs of congenital anomalies that are most often seen in radiologic practice include, egg on a string (side) in transposition of the great arteries, snowman in total anomalous pulmonary venous return, Scimitar in partial anomalous pulmonary venous return, Gooseneck in endocardial cushion defect, figure of three and reverse figure of three in aortic

coarctation, boot-shaped heart in tetralogy of Fallot, box-shaped heart in Ebstein anomaly.

46.(C) The arterial switch (Jatene) procedure is the surgical treatment of choice for neonates with d-TGA and an intact ventricular septum and is usually performed within the first 2 weeks of life. The reason for this time frame is that as PVR declines after birth, pressure in the left ventricle (connected to the pulmonary vascular bed) also declines. This pressure drop results in a decrease in left ventricular (LV) mass over the first few weeks of life. If the arterial switch operation is attempted after LV pressure (and mass) has declined too far, the left ventricle will be unable to generate adequate pressure to pump blood to the high pressure systemic circulation.

47.(A) Sano and Norwood procedures are used for hypoplastic left heart syndrome.

48.(A) In double-outlet right ventricle without pulmonary stenosis, both the aorta and the pulmonary artery arise from the right ventricle. The only outlet from the left ventricle is through a VSD. In the absence of obstruction to pulmonary blood flow, clinical manifestations are similar to those of an uncomplicated VSD with a large left-to-right shunt. Systemic desaturation may result from mixing of oxygenated and deoxygenated blood in the right ventricle.

49.(D) Infracardiac type associated with obstruction in 95-100% of cases.

50.(C) In neonates with marked pulmonary venous obstruction, the chest radiograph demonstrates a very dramatic perihilar pattern of pulmonary edema and a small heart. In option A the presentation is usually similar but the cardiac shadow is normal or increased with decreased to normal perihilar shadow. In option B the patient is usually preterm baby. In C it is typical for TAPVR with obstruction. In D neither antenatal history nor CXR finding are suggestive. In E it is ductal dependent so the presentation will be later (after few days).

51.(D) Obstructed TAPVR is a pediatric cardiac surgical emergency because prostaglandin therapy is usually not effective. Option B, C, and E all are important supportive measures but delay in decision taken in such situations may lead to death.

52.(C) This is the classical presentation of TAPVR without obstruction. In most cases without obstruction, the heart is enlarged, the pulmonary artery and right ventricle are prominent, and pulmonary vascularity is increased. Demonstration of any vein with Doppler flow away from the heart is pathognomonic of TAPVR. Option A, B, and D have similar course of events with some differences in the clinical findings.

53.(E) This is classical presentation of LHHS when shock and signs of heart failure dominate the clinical picture, ECG usually shows right ventricular dominance with absent left ventricular forces. This lesion may be isolated or associated in 5–15% of patients with known genetic syndromes, such as Turner syndrome, trisomy 13, 18, or 21, Jacobsen syndrome (11q deletion), Holt-Oram syndrome, and Rubinstein-Taybi syndrome. In these circumstances, noncardiac manifestations of the syndrome may be evident and influence the clinical outcomes. Occasionally it is familial and inherited as an autosomal recessive trait.

54.(B) Serial fetal echocardiographic studies demonstrate that in some fetuses, HLHS may be a progressive in utero lesion, beginning with simple valvar aortic stenosis in midgestation. The decreased flow through the stenotic aortic valve reduces flow through the left ventricle during development, resulting in gradual ventricular chamber hypoplasia. The potential for preventing this hypoplasia has been demonstrated by performing in utero aortic balloon valvuloplasty in midgestation fetuses. Early results are encouraging, although even if the aortic valve is successfully opened, adequate ventricular growth occurs in only about 30% of patients. At present, this procedure is regarded as experimental.

55.(D) If the vascular ring produces compression of the trachea and esophagus, symptoms are frequently present during infancy. Chronic wheezing is exacerbated by crying, feeding, and flexion of the neck. Extension of the neck tends to relieve the noisy respiration. Vomiting may also be a component. Affected infants may have a brassy cough, pneumonia, or rarely, sudden death from aspiration. Echocardiography in combination with either MRI or CT used for diagnosis.

56.(E) In anomalous origin of the left coronary artery from the pulmonary artery (**ALCAPA**), the blood supply to the left ventricular (LV) myocardium is severely compromised. Soon after birth, as pulmonary artery pressure falls, perfusion pressure to the left coronary artery (LCA) becomes inadequate; myocardial ischemia, infarction, and fibrosis result. In some cases, interarterial collateral anastomoses develop between the right coronary artery (RCA) and LCA. Blood flow in the LCA is then reversed, and it empties into the pulmonary artery, a condition known as the “myocardial steal” syndrome. The left ventricle becomes dilated, and its performance is decreased. Mitral insufficiency is a frequent complication secondary to a dilated valve ring or infarction of a papillary muscle.

57.(D) This description is fit for all the mentioned distracters with only minor changes. The echocardiography and cardiac catheterization can differentiate

between different types of left-sided obstructive lesions (pulmonary venous stenosis, mitral stenosis, restrictive cardiomyopathy) that result in pulmonary venous hypertension. The presence of pulmonary arterial hypertension (PAH) with a normal pulmonary capillary wedge pressure is diagnostic of PAH. If the wedge pressure is elevated and left ventricular end-diastolic pressure (LVEDP) is normal, obstruction at the level of the pulmonary veins, left atrium, or mitral valve should be suspected. If LVEDP is also elevated, the diagnosis of restrictive cardiomyopathy should be entertained. The risks associated with cardiac catheterization are increased in severely ill patients with primary PH.

58.(A) Pulmonary vascular disease occurs more rapidly in patients with trisomy 21 who have left-to-right shunts.

59.(C) Emphasizing the nature of CHD will reduce anxiety of the parents and also reduces the reflected anxiety on the child himself. Calling for whole family counseling session is not indicated and may cause horror. Most patients who have mild congenital heart disease (CHD) do not require treatment. The parents and child should be made aware that a normal life is expected, and that no restriction of the child's activities is necessary. Overprotective parents may use the presence of a mild congenital heart lesion or even a functional heart murmur as a means to exert excessive control over their child's activities. Although fears may not be expressed overtly, the child may become anxious regarding early death or debilitation, especially when an adult member of the family acquires unrelated symptomatic heart disease. The family may have an unexpressed fear of sudden death, and the rarity of this manifestation should be emphasized in discussions directed at improving their understanding of the child's congenital heart defect. The difference between CHD and degenerative coronary disease in adults should be emphasized. General health maintenance, including a well-balanced “heart-healthy” diet, aerobic exercise, and avoidance of smoking, should be encouraged.

60.(D) Routine immunizations should be given, with the inclusion of influenza vaccine during the appropriate season. Prophylaxis against the respiratory syncytial virus (RSV) is recommended during RSV season in young infants with unrepaired CHD and significant hemodynamic abnormalities. Careful consideration of the timing of administration of live-virus vaccination is required in patients who are potential candidates for heart or heart-lung transplantation, and these patients cannot receive live-virus vaccines after they have received their transplant.

61.(E) In cyanotic patients high altitudes and sudden changes in the thermal environment should also be avoided, as both can result in either respiratory

infection which could be severe or precipitation of TET spells due to high altitude. In general cyanotic patients should also avoid situations where dehydration may occur, which leads to increased viscosity and increases the risk of stroke. Diuretics may need to be decreased or temporarily discontinued during episodes of acute gastroenteritis. Treatment of iron deficiency anemia is important in cyanotic patients, who may have a low mean corpuscular hemoglobin concentration despite polycythemia.

62.(D) The pain follow strenuous exercise in a healthy adolescent is usually muscular in origin. A silent PDA is a tiny defect that cannot be heard by auscultation and is only detected by other means such as echocardiography. Life expectancy is always normal in this population and the risk for endocarditis is extremely low.

63.(E) Bicuspid aortic valve is the most common congenital heart disease that manifest during adult life. By the age of 45 yr, approximately 50% of bicuspid aortic valves will have some degree of stenosis. Usually have normal life expectancy.

64.(E) Only patients with cardiac conditions associated with the highest risk for adverse outcomes should continue antibiotic prophylaxis before surgery: previous endocarditis; unrepaired cyanotic CHD, including palliative shunts and conduits; completely repaired congenital heart defects with prosthetic material or device, surgically placed or by catheter intervention, during the 1st 6 mo after the procedure; and repaired CHD with residual defects at or adjacent to the site of a prosthetic patch or prosthetic device (which inhibits endothelialization). Except for the conditions just listed, antibiotic prophylaxis is no longer recommended for other forms of CHD.

65.(D) Dilated cardiomyopathy, the most common form of cardiomyopathy in children, is the cause of significant morbidity and mortality as well as the most common indication for cardiac transplantation.

66.(A) Although the most common etiology of DCM remains idiopathic, its likely that undiagnosed familial/genetic conditions and myocarditis predominate. Unlike adult patients with DCM, ischemic etiologies are rare in children. The annual incidence of DCM in children younger than 18 yr is 0.57 cases per 100,000 per year. Incidence is higher in males, blacks, and infants <1 yr old.

67.(B) In dilated cardiomyopathies, the presence of hypoglycemia, acidosis, hypotonia, or signs of liver dysfunction suggests an inborn error of metabolism. Neurologic or skeletal muscle deficits are associated with mitochondrial disorders or muscular dystrophies.

68.(D) In patients presenting with extreme degrees of heart failure or circulatory collapse, intensive care measures are often required, including intravenous inotropes and diuretics, mechanic ventilatory support, and on occasion, mechanical circulatory support.

69.(B) Restriction from competitive sports and strenuous physical activity is highly recommended, and additional recreational exercise activities should be tailored to each individual based on their overall clinical status. β-Adrenergic blocking agents (propranolol, atenolol, metoprolol) or calcium channel blockers (verapamil) may be useful in diminishing LVOT obstruction, modifying LV hypertrophy, and improving ventricular filling. They also confer an antiarrhythmic benefit and may reduce symptoms. Digoxin is contraindicated.

70.(E) Presence of family history of sudden death and absence of ejection click makes diagnosis of severe aortic stenosis is unlikely. Many patients with hypertrophic cardiomyopathies are asymptomatic, and 50% of cases present with a heart murmur or during screening when another family member has been diagnosed with HCM. Symptoms of HCM may include palpitations, chest pain, easy fatigability, dyspnea, dizziness, and syncope. Sudden death is a well-recognized but uncommon manifestation that occurs during physical exertion. Characteristic physical examination findings include an overactive precordial impulse with a lift or heave, a systolic ejection murmur in the aortic region not associated with an ejection click, and an apical blowing murmur of mitral insufficiency. In critical PS cyanosis is predominate.

71.(E) Dramatic atrial dilation can result from the abnormal ventricular myocardial compliance and high ventricular diastolic pressure. Abnormal ventricular filling, sometimes referred to as diastolic heart failure, is manifest in the systemic venous circulation with edema, hepatomegaly, or ascites.

72.(A) The vast majority of tumors originating from the heart are benign. Rhabdomyomas are the most common pediatric cardiac tumors and are associated with tuberous sclerosis in 70–95% of cases.

73.(B) All types of arrhythmia can presents in viral myocarditis, the most dangerous and can lead to death is ventricular tachycardia.

74.(C) The ECG must be closely monitored, and rhythm strips obtained before each of the 3 digitalizing doses. Digoxin should be discontinued if a new rhythm disturbance is noted. Prolongation of the P-R interval is not necessarily an indication to withhold digitalis, but a delay in administering the next dose or a reduction in the dosage should be considered, depending on the patient's clinical status. Minor ST segment or T-wave changes are frequently noted with digitalis administration and should not affect the digitalization regimen.

75.(A) Baseline serum electrolyte levels should be measured before and after digitalization. Hypokalemia and hypercalcemia exacerbate digitalis toxicity. Because hypokalemia is relatively common in patients receiving diuretics, potassium levels should be monitored closely in those receiving a potassium-wasting diuretic in combination with digitalis. In patients with active myocarditis, some cardiologists recommend avoiding digitalis altogether and if used, maintenance digitalis should be started at half the normal dose without digitalization because of the increased risk of arrhythmia in these patients.

76.(C) Milrinone is useful in treating patients with low cardiac output who are refractory to standard therapy. It has been shown to be highly effective in managing the low-output state present in children after open heart surgery.

Milrinone has both positive inotropic effects on the heart and peripheral vasodilator effects and has generally been used as an adjunct to dopamine or dobutamine therapy in the intensive care unit.

77.(D)

78.(E) Patients with WPW syndrome have a small but real risk of sudden death. Risk stratification, including 24 hr Holter monitoring and exercise study, may help differentiate patients at higher risk for sudden death from WPW. Syncope is an ominous symptom in WPW, and any patient with syncope and WPW syndrome should have an electrophysiology study (EPS) and likely catheter ablation. Although most often presenting in patients with a normal heart, WPW syndrome may also be associated with Ebstein anomaly of the tricuspid valve, hypertrophic cardiomyopathy or L-TGA.

79.(B) Do not loss a lot of time in vagal stimulation, adenosine is the drug of choice. Verapamil can cause cardiac arrest and severe hypotension in children less than one year and contraindicated in patients with WPW. Synchronized cardioversion reserved for patients who shows signs of severe heart failure.

80.(E) Once the patient has been converted to sinus rhythm, a longer-acting agent is selected for maintenance therapy. In patients without an antegrade accessory pathway (non-WPW), the β-adrenergic blockers are the mainstay of drug therapy. Digoxin is also popular and may be effective in infants, but less so in older children. In children with WPW, digoxin or calcium channel blockers may increase the rate of antegrade conduction of impulses through the bypass tract, with the possibility of ventricular fibrillation, and are therefore contraindicated.

81.(A) Hypothyroidism or hyperthyroidism, elevated triglycerides, hepatic toxicity, and pulmonary fibrosis all are possible side effects of amiodarone.

82.(D) For patients with ventricular tachycardia VT who are hemodynamically stable, intravenous amiodarone, lidocaine, or procainamide is the initial drug of choice. If treatment is to be successful, it is critical to search for and correct any underlying abnormalities, such as electrolyte imbalance, hypoxia, or drug toxicity. Hemodynamically unstable patients with VT should be immediately treated with DC cardioversion. Option A and D is reserved for patients suffering from VF.

83.(B) Antibiotics; erythromycin, clarithromycin, azithromycin, telithromycin, trimethoprim/sulfamethoxazole, and fluoroquinolones.

84.(E)

85.(A) LQTS type 1 (LQT1) events are usually induced by stress or exertion, whereas events in LQT3 often occur at rest, especially during sleep. LQT2 events have an intermediate pattern, often occurring in the postpartum period or with auditory triggers. LQT3 has the highest probability for sudden death, followed by LQT2 and then LQT1.

86.(B) No relationship exists between the infecting organism and the type of congenital defect, duration of illness, or age of the child. Staphylococcal endocarditis is more common in patients with no underlying heart disease. Viridans group streptococcal infection is more common after dental procedures; group D enterococci are seen more often after lower bowel or genitourinary manipulation.

87.(A) A single episode of acute rheumatic carditis often results in complete healing of the valvular lesions, while repeated episodes, especially involving previously affected valves, result in chronic rheumatic heart disease (RHD), which is the rationale for secondary prophylaxis.

Chapter 20

Diseases of the Blood Questions

HASANEIN H. GHALI

1. Erythrocytes in the fetus are larger than in adults, and at 22-23 wk gestation the mean corpuscular volume can be as high as

A. 105 femtoliters (fL)
B. 115 femtoliters (fL)
C. 125 femtoliters (fL)
D. 135 femtoliters (fL)
E. 145 femtoliters (fL)

2. How much is the normal ratio of HbA to HbA_2 throughout life?

A. 15 : 1
B. 20 : 1
C. 25 : 1
D. 30 : 1
E. 35 : 1

3. Which of the following conditions is associated with increased Levels of HbA_2>3.4%?

A. Megaloblastic anemia
B. Iron-deficiency
C. α-thalassemia
D. Hemolytic anemias
E. Diamond Blackfan anemia

4. Microcytes are seen in blood films of which of the following conditions?

A. Aplastic anemia
B. Liver disease
C. Anemia of chronic disease
D. Down syndrome
E. Hypothyroidism

5. Basophil stippling is seen in blood films of which of the following conditions?

A. Hereditary spherocytosis
B. Aplastic anemia
C. Liver disease
D. Thalassemia
E. Xerocytosis

6. A 4-month-old infant presents with progressive pallor,microcephaly, snub nose, high-arched palate, hypertelorism, low-set ears, and triphalangeal thumb. Peripheral blood smear shows macrocytic RBCs with low reticulocyte count. Of the following, the **MOST** likely diagnosis is

A. Diamond-Blackfan anemia
B. Fanconi anemia
C. Shwachman-Diamond syndrome
D. Pearson marrow-pancreas syndrome
E. Aase syndrome

7. Which of the following is the mainstay therapy of Diamond-Blackfan anemia?

A. Chronic red cell transfusions
B. Corticosteroids
C. Hematopoietic stem cell transplantation
D. Hydroxyurea
E. spenectomy

8. Which of the following differentiate transient erythroblastopenia of childhood from iron-deficiency anemia?

A. MCV
B. Reticulocytopenia
C. Neutrophil count
D. Platelet number
E. Age of onset

9. Which of the following is the **BEST** diagnostic test used to distinguish anemia of chronic disease (ACD) from iron-deficiency anemia (IDA)?

A. Serum iron
B. Serum transferrin
C. Serum ferritin
D. Soluble transferring receptor

E. MCV

10. A 7-year-old child presents with pallor, jaundice, frontal bossing hepatosplenomegaly, , supernumerary toes, and gall stones revealed by US. Laboratory findings revealed hemoglobin 8 g/dL, MCV 110 fL, normoblasts, anisopoikilocytosis and basophilic stippling of RBCs. The bone marrow aspirate shows erythroid hyperplasia, megaloblastosis, with incompletely divided cells with thin chromatin bridges between nuclei of pairs of erythrocytes. Electron microscopy revealed erythroblasts with a characteristic "Swiss cheese" heterochromatin pattern.
Of the following, the **MOST** likely diagnosis is

A. congenital dyserythropoietic anemia
B. Diamond-Blackfan anemia
C. Fanconi anemia
D. hereditary spherocytosis
E. hereditary elliptocytosis

11. A 10-week-old baby is brought by his mother because of pallor, he has been delivered normally at 38 weeks with weight of 3.6 Kg, now he looks well but his Hb is 10 mg/dl.
Of the following, the **MOST** appropriate management is

A. iron therapy
B. folic acid therapy
C. vitamin E therapy
D. reassurance
E. erythropoietin therapy

12. Which of the following milk is folate deficient?

A. Human breast milk
B. Infant formula
C. Pasteurized cow's milk
D. Goat's milk
E. Camel milk

13. Which of the following anticonvulsant drugs can impair folic acid absorption?

A. Phenobarbital
B. Oxcarbazepine

C. Ethosuximide
D. Lamotrigine
E. Levetiracetam

14. A 7-month-old boy presents with progressive pallor, irritability, chronic diarrhea, and poor weight gain. He exclusively breast fed till the age of two months, then shifted to boiled goat milk because of mother illness. Hb 7 gm/dl, MCV 105 fl, reticulocyte count 1%, WBCs and platelets normal.
Of the following, the **MOST** important investigation is

A. serum B12
B. serum folic acid
C. serum ferritin
D. tissue trasglutaminase IgA Ab
E. serum zinc

15. A 7-month-old boy presents with progressive pallor, irritability, chronic diarrhea, and poor weight gain. He was exclusively breast fed till the age of two months, then shifted to boiled goat milk because of mother illness. Hb 7 gm/dl, MCV 105 fl, reticulocyte count 1%, WBCs and platelets were normal.
Of the following, the **MOST** appropriate treatment for this boy is

A. folic acid therapy 1.0 mg/day for 1wk
B. folic acid therapy 5.0 mg/day for 2 wk
C. folic acid therapy 1.0 mg/day for 4 wk
D. folic acid therapy 1.0 mg/day for 8 wk
E. multivitamin dropper (containing 0.2 mg of folate)

16. A 1-year-old boy presents with progressive pallor, weakness, lethargy, feeding difficulties, failure to thrive, and irritability. On examination he has a tinge of jaundice, glossitis, hypotonia, and developmental delay. His peripheral blood shows macro-ovalocytosis of the RBCs with neutropenia and thrombocytopenia. His mother has peptic ulcer disease and on proton pump inhibitors.
Of the following, the **MOST** likely cause is

A. B_{12} deficiency
B. folate deficiency
C. hereditary ovalocytosis
D. iron deficiency
E. Imerslund-Grasbeck syndrome

17. A 4-year-old child presents with pallor, hypotonia, developmental delay, and movement disorders. Laboratory findings revealed megaloblastic anemia and proteinuria. The parents are 1st degree relatives and the child has two normal siblings.
Of the following, the **MOST** likely diagnosis is

A. B12 deficiency
B. folate deficiency
C. Imerslund-Grasbeck syndrome
D. Rogers syndrome
E. Oroticaciduria

18. A 3-year-old child presents with megaloblastic anemia, sensorineural deafness, and diabetes mellitus. He has visual problems, congenital heart disease, and his height is below 3rd percentile. The bone marrow revealed megaloblastic changes with ringed sideroblasts.
Of the following, the **MOST** likely diagnosis is

A. B_{12} deficiency
B. Transcobalamin deficiency
C. Imerslund-Grasbeck syndrome
D. Rogers syndrome
E. Oroticaciduria

19. How much is the necessary daily dietary intake of iron to maintain positive iron balance in childhood?

A. 10 mg
B. 20 mg
C. 30 mg
D. 40 mg
E. 50 mg

20. Irritability, anorexia, lethargy, and systolic flow murmurs are often heard, when the hemoglobin level falls below

A. 8 g/dL
B. 7 g/dL
C. 6 g/dL
D. 5 g/dL
E. 4 g/dL

21. Which of the following provides very useful and early indicators of iron deficiency?

A. Serum ferritin
B. Serum iron
C. Serum transferrin
D. Free erythrocyte protoporphyrins
E. Reticulocyte hemoglobin concentration

22. Iron deficiency is best prevented to avoid both its systemic manifestations and anemia. Breastfeeding should be encouraged, with the addition of supplemental iron at the age of

A. 2 months
B. 4 months
C. 6 months
D. 8 months
E. 10 months

23. Iron therapy in developing countries may increase the virulence of certain

A. viruses
B. gram negative bacteria
C. gram positive bacteria
D. fungi
E. atypical bacteria

24. Blood film of a 2-year-old child shows poikilocytosis, microcytosis, fragmented erythrocytes, and elliptocytes.
Of the following, the **MOST** likely diagnosis is

A. sickle cell disease
B. liver disease
C. hereditary spherocytosis
D. hereditary elliptocytosis
E. hereditary pyropoikilocytosis

25. Which of the following RBC indices is **MOST** helpful in the diagnosis of hereditary spherocytosis?

A. RBC count
B. MCV
C. MCH

D. MCHC
E. RDW

26. Which of the following closely mimics hereditary spherocytosis in the neonatal period?

A. ABO incompatibility
B. Autoimmune hemolytic anemia
C. Clostridial sepsis
D. Wilson disease
E. Congenital dyserythropoietic anemia type II

27. When splenectomy is indicated for hereditary spherocytosis, it should be performed after age of

A. 3years
B. 4 years
C. 5 years
D. 6 years
E. 7 years

28. What is the **MOST** important lab finding that differentiate hereditary pyropoikilocytosis from hereditary elliptocytosis?

A. Extreme microcytosis
B. Extreme anisocytosis
C. Extreme macrocytosis
D. Extreme elliptocytosis
E. Extreme ovalocytosis

29. A 2-year-old child presents with mild compensated macrocytic hemolytic anemia with splenomegaly 2 cm below costal margin with intermittent jaundice. CBC shows elevated MCHC, MCV, decreased erythrocyte osmotic fragility, and decreased K+ concentration, blood film reveals small numbers of stomatocytes, target cells, and contracted RBCs.
Of the following, the **MOST** likely diagnosis is

A. Hereditary hydrocytosis
B. Hereditary xerocytosis
C. Hereditary cryohydrocytosis
D. Tangier disease
E. Sitosterolemia

30. A 10-year-old girl presents with large orange tonsils, hepatosplenomegaly, lymphadenopathy, cloudy corneas, and peripheral neuropathy. Her peripheral blood smear reveals moderate stomatocytic hemolytic anemia and thrombocytopenia.
Of the following, the **MOST** likely diagnosis is

A. Phytosterolemia
B. Rh Deficiency Syndrome
C. Cryohydrocytosis
D. Tangier disease
E. Sitosterolemia

31. Pancytopenia with bone marrow infiltration is seen in

A. B_{12} deficiency
B. CMV infection
C. systematic lupus erythematosus
D. osteopetrosis
E. aplastic anemia

32. What is the **MOST** common inherited pancytopenia syndrome?

A. Dyskeratosis congenita
B. Fanconi anemia
C. Shwachman-Diamond syndrome
D. Diamond-Blackfan anemia
E. Congenital amegakaryocytic thrombocytopenia

33. Which of the following pancytopenia is transmitted through autosomal dominant manner?

A. Congenital amegakaryocytic thrombocytopenia
B. Amegakaryocytic thrombocytopenia with radioulnar synostosis
C. Nijmegen breakage syndrome
D. Seckel syndrome
E. Dubowiz syndrome

34. Which of the following pancytopenia is transmitted through mitochondrial inheritance?

A. Diamond-Blackfan anemia
B. Amegakaryocytic thrombocytopenia with radioulnar synostosis
C. Pearson marrow-pancreas syndrome

D. Schimke syndrome
E. Cartilage-hair hypoplasia

35. What is the **MOST** common congenital anomaly in Fanconi anemia?
A. Cardiac
B. Gastrointestinal
C. Eye
D. Renal
E. Skeletal

36. What is the latest hematological abnormality evolved in Fanconi anemia?
A. Anemia
B. Thrombocytopenia
C. Neutropenia
D. Macrocytosis
E. Increased hemoglobin F

37. What is the **MOST** common available test for diagnosis of Fanconi anemia?
A. Complete blood count
B. Bone marrow aspiration and biopsy
C. Chromosomal breakage test
D. Mutation screening
E. Next-generation sequencing

38. Which of the following options provides curative outcome for hematological abnormalities in patients with Fanconi anemia?
A. Androgen therapy
B. Hematopoietic stem cell transplantation
C. Granulocyte colony stimulating factor
D. Erythropoietin
E. Blood / blood products transfusions

39. Which of the following malignancies is considered as a complication of Fanconi anemia?
A. Acute lymphoblastic leukemia
B. Acute myeloid leukemia
C. Osteosarcoma
D. Rhabdomyosarcoma

E. Retinoblastoma

40. Initial response of androgen therapy in Fanconi anemia is heralded by
A. rise in hemoglobin
B. increase in neutrophil count
C. increase in lymphocyte count
D. reticulocytosis
E. increase in platelet counts

41. What is the **MOST** commonly used androgenic drug in Fanconi anemia?
A. Testosterone
B. Oxymetholone
C. Methenolone acetate
D. Nandrolone decanoate
E. Boldenone undecenoate

42. Which of the following is **TRUE** regarding Pros and cons of androgen therapy in Fanconi?
A. They can produce a response in about 30% of patients
B. Treatment is curative in some patients
C. Dose modification is always required according to hematological response
D. Once response is achieved, it usually continues for many years
E. Liver tumors are reported as possible consequences of androgen use

43. Granulocyte colony-stimulating factor (G-CSF) in Fanconi anemia can usually induce an increase in the neutrophil count; however, a notable limitation to starting therapy is
A. severe bone pain
B. local bleeding at site of subcutaneous injection
C. risk of expansion of bone marrow cells with clonal cytogenetic abnormality
D. the rise of counts involves only white blood cells
E. possible loss of response after few doses due to disease progression

44. A 4-year-old male develops recurrent epistaxis and easy bruising.. The patient was the first child of unrelated healthy parents born after 41 weeks of pregnancy. Physical examination of the patient reveal short stature, clinodactyly with brachy mesophalangia on bilateral 5th fingers, multiple café-au-lait spots on both thighs and right buttock. He has no other abnormalities. His initial complete blood cell count results are as follows: white blood cell, 5.0×10^9/L; hemoglobin 9.5 g/dL; platelets 80×10^9/L.
Of the following, the **MOST** likely diagnosis is

A. megaloblastic anemia
B. Pearson syndrome
C. congenital Amegakaryocytic Thrombocytopenia
D. Fanconi anemia
E. Diamond-Blackfan anemia

45. What is the **MOST** common hematological abnormality in Shwachman-Diamond syndrome?

A. Anemia
B. Neutropenia
C. Lymphopenia
D. Thrombocytopenia
E. Pancytopenia

46. What is the **MOST** common non-hematological abnormality in Shwachman-Diamond syndrome?

A. Pancreatic insufficiency
B. Elevated transaminase
C. Skeletal abnormality
D. Short stature
E. Rib cage abnormality

47. A 24-month-old girl was admitted to the hospital because of steatorrhea and short stature. At the age of three months, she presented with convulsion and neutropenia. Since then she has had frequent purulent otitis media and pneumonias. She has angular stomatitis, and many dental caries. The results of the laboratory studies were as follows: hemoglobin 11.3 g/dL, leukocytes 3,000/μL (neutrophils 17.3%, lymphocytes 59.5%, monocytes 22.9%, basophils 0.3%, and eosinophils 0%), platelets 152,000/μL.
Of the following, the **MOST** likely diagnosis is

A. cystic fibrosis
B. primary lactase deficiency
C. Shwachman-diamond syndrome
D. alpha-one antitrypsin deficiency
E. Pearson syndrome

48. Which of the following feature are more specific of Fanconi anemia than Shwachman-diamond syndrome?
A. Metaphyseal dysplasia
B. Delayed appearance of secondary ossification centers
C. Triphalangeal thumb
D. Short or flared ribs
E. Thoracic dystrophy

49. What is the **MOST** common reported chromosomal abnormality in Shwachman-diamond syndrome?
A. Isochromosome 7q
B. Monosomy 7
C. Isochromosome 7q combined with monosomy 7
D. Deletions of 7q
E. Deletions of 20q

50. What is the old diagnostic triad for dyskeratosis congenita?
A. Nail dysplasia, reticular pigmentation of the chest, and oral leukoplakia
B. Bone marrow failure, nail dysplasia, and oral leukoplakia
C. Reticular pigmentation of the chest, bone marrow failure, and nail dysplasia
D. Avascular necrosis of hip, nail dysplasia, and bone marrow failure
E. Optic atrophy, nail dysplasia, and oral leukoplakia

51. What is the **MOST** common clinical manifestation of dyskeratosis congenita?
A. Leukoplakia
B. Epiphora
C. Blepharitis
D. Skin pigmentation
E. Phimosis

52. Revesz syndrome has many of the features of dyskeratosis congenital DC and presents in early childhood in addition to

A. blepharitis
B. conjunctivitis
C. exudative retinopathy
D. cataract
E. optic atrophy

53. Which of the following inherited bone marrow failure syndromes has the lowest survival and highest mortality?

A. Fanconi anemia
B. Shwachman-Diamond syndrome
C. Dyskeratosis congenita
D. Congenital amegakaryocytic thrombocytopenia
E. Diamond-Blackfan Anemia

54. Congenital amegakaryocytic thrombocytopenia patients usually have normal phenotype. Yet, about 10% of cases involve physical anomalies.
Of the following, the **MOST** common anomalies are

A. eye anomalies
B. kidney malformations
C. skeletal malformations
D. cardiac anomalies
E. dysmorphic features

55. An 8-year-old boy presents with fever, headache, chills, and painful throat for five days. Leukoplakia is noted on his tongue and some of his finger and toe nails are markedly dystrophic. His skin seem spotted with pigmentation on the anterior chest and neck. Vitally, his blood pressure 106/74 mm Hg, pulse rate 106/min, respiratory rate 28b/min, and body temperature 38.9 C. His WBC count was 0.300/mm^3 with 37% neutrophils, platelets 21,000 and Hb 9.2 g/dl.
Of the following, the **MOST** likely diagnosis is

A. congenital amegakaryocytic thrombocytopenia
B. dyskeratosis congenita
C. reticular dysgenesis
D. Fanconi anemia
E. cartilage-hair syndrome

56. Which of the following viruses is implicated as a possible cause of acquired prolonged aplastic anemia?

A. Rubella virus
B. Measles virus
C. Cytomegalovirus
D. Herpes virus
E. Parvovirus B19

57. Which of the following antibacterial agents is implicated as a possible cause of acquired aplastic anemia?

A. Ampicillin
B. Trimethoprim-sulfamethoxazole
C. Cefotaxime
D. Gentamycin
E. Azithromycin

58. Which of the following findings is a criterion of severe aplastic anemia?

A. Absolute neutrophil count 600/mm^3
B. Platelet count 18,000/mm^3
C. Reticulocyte count 1.5 %
D. Bone marrow biopsy cellularity 33%
E. Hemoglobin level 6.0 gm/dl

59. Cases of Aplastic anemia should be assessed for paroxysmal nocturnal hemoglobinuria. The **MOST** sensitive test is

A. direct antiglobulin test
B. hemoglobin F level
C. erythrocyte CD55 and CD59
D. haptoglobin
E. urine for hemosiderin

60. What is the **MOST** common chromosomal abnormality found within the marrow of patients with myelodysplastic syndrome?

A. Monosomy 7
B. Monosomy 8
C. Trisomy 11
D. Trisomy 13
E. Trisomy 21

61. Which of the following lab findings suggest polycythemia due to heart or lung disease?

A. Normal red cell mass with low plasma volume
B. Elevated red cell mass with high carboxyhemoglobin
C. Elevated red cell mass with low arterial oxygen
D. Normal red cell mass with abnormal hemoglobin study
E. Elevated red cell mass with low erythropoietin

62. Which of the following is a major WHO criterion for diagnosis of Polycythemia Vera?

A. Red cell mass 20% above mean normal predicted value
B. Bone marrow trilineage myeloproliferation
C. Subnormal serum erythropoietin level
D. Endogenous erythroid colony growth
E. Presence of JAK2 or similar mutation

63. What is the usual physical finding in polycythemia?

A. Hepatosplenomegaly
B. Skin ecchymosis
C. Pruritus marks
D. Hypertension
E. lymphadenopathy

64. What is the **MOST** convenient initial treatment of polycythemia?

A. Phlebotomy
B. Aspirin
C. Hydroxyurea
D. Interferon-gamma
E. Iron

65. Among infants less than 4 months of age, which of the following is an indication of blood transfusion?

A. Hemoglobin less than 14.0 gm/dl in patient with severe pulmonary disease
B. Hemoglobin less than 10.0 g/dl in patient with moderate pulmonary disease
C. Hemoglobin less than 14.0 g/dl in patients with severe cardiac disease
D. Hemoglobin less than 11.0 g/dl preoperatively and during major surgery

E. Hemoglobin less than 10.0 g/dl postoperatively

66. What is the key reason that the nadir hemoglobin values of premature infants are lower than those of term infants?

A. Bleeding is more common among premature infants
B. Bone marrow is relatively lazy in premature infants
C. Rate of hemolysis in premature infants is greater than term
D. Diminished plasma erythropoietin in response to anemia
E. Fetal hemoglobin replacement is slow among premature infants

67. According to the guidelines of pediatric platelet transfusion, patients with marrow failure and hemorrhagic risk factors (infection, organ failure, and clotting abnormalities) should have their platelets count maintained above

A. 80,000/mm^3
B. 60,000/mm^3
C. 40,000/mm^3
D. 20,000/mm^3
E. 10,000/mm^3

68. A 1-month-old infant with sepsis on aggressive antibiotic therapy, while he is clinically unstable requires a platelet count to be maintained above

A. 100,000/mm^3
B. 70,000/mm^3
C. 50,000/mm^3
D. 30,000/mm^3
E. 10,000/mm^3

69. In patients with inherited platelet disorders, platelet transfusions are justified only for overt bleeding and high risk of bleeding. This is because

A. the risk of bleeding is less than acquired platelet disorders
B. no response to platelet transfusions
C. life threatening bleeding is seldom in these disorders
D. possible association with transfusion reaction
E. repeated transfusions lead to refractoriness

70. Granulocyte transfusion for a 3-day-old newborn is indicated with a threshold of neutrophil count below?

A. 1,000/mm^3

B. 1,500/mm^3
C. 2,000/mm^3
D. 2,500/mm^3
E. 3,000/mm^3

71. Which of the following factors reduces the level of von Willebrand factor?
A. Stress
B. Infections
C. Hypothyroidism
D. Exercise
E. Pregnancy

72. Which of the following types of von Willebrand disease is misdiagnosed as hemophilia due to low factor VIII level?
A. Type 1C
B. Type 2A
C. Type 2B
D. Type 2M
E. Type 2N

73. Which type of von Willebrand disease presents with thrombocytopenia?
A. Type 1C
B. Type 2A
C. Type 2B
D. Type 2M
E. Type 2N

74.Which of the following subtypes of von Willebrand disease is likely to benefit from Desmopressin?
A. Type 1
B. Type 1C
C. Type 2A
D. Type 2B
E. Type 3

75. Which of the following conditions causes thrombocytopenia through impaired platelets production?
A. Kasabach-Merritt syndrome

B. Hypersplenism
C. Hypothermia
D. Infection-induced thrombocytopenia
E. Osteopetrosis

76. A 3-year-old boy presents with sudden onset of generalized petechiae and purpura for the last two days, he is otherwise healthy and playful. Parents deny any previous similar episodes and family illnesses. His general examination showed active boy with normal vitals and no abnormality in general exam apart from these skin lesions. His heart exam is normal, chest note is normal and abdominal exam fails to identify organ enlargement.
Of the following, the **MOST** likely diagnosis is

A. aplastic anemia
B. immune thrombocytopenia
C. acute leukemia
D. Glanzmann thrombasthenia
E. von Willebrand disease

77. Which of the following lab tests results is worrisome in immune thrombocytopenia and should raise the suspicion of another diagnosis?

A. A platelet count of 1.000 /mm^3
B. A WBCs count of 2.500 /mm^3
C. A hemoglobin count of 8.0 gm/dl
D. A bone marrow examination showing increase number of megakaryocytes
E. A negative Coombs test

78. Autoimmune screen, in the context of immune thrombocytopenia, is required in which of the following clinical conditions?

A. Five-year-old girl with long history of skin and mucosal bleeding lasting more than one year
B. Acute history of severe profound epistaxis, heralding possible need for blood transfusion
C. Acute presentation of intracranial hemorrhage in a background of two weeks skin bleeding that wasn't without a prior problem
D. Periorbital bleeding suggesting basal skull fracture after road traffic accident in a newly diagnosed immune thrombocytopenia with one month of the accident

E. An adolescent girl presented with severe menorrhagia and pallor, with a background of few weeks skin ecchymosis with reluctance for medical counseling for social reasons

79. A 6-year-old girl presents with recurrent skin ecchymosis and petechial rash over the abdomen and lower limbs for the last few weeks. Her past medical history is negligible apart from frequent neonatal exchange transfusion for severe Rh incompatibility. Her regional exam shows a clear chest note, normal heart sounds, and soft palpable spleen of 5 cm BCM with no hepatomegaly and no palpable masses. His CBC film shows isolated thrombocytopenia, normal other indices and normal film.
Of the following, the **MOST** likely diagnosis is

A. acute lymphoblastic leukemia
B. portal vein thrombosis
C. immune thrombocytopenia
D. chronic liver disease
E. aplastic anemia

80. Autoimmune thrombocytopenia may be the initial manifestation of which of the following malignancies?

A. Neuroblastoma
B. Lymphoma
C. Wilms' tumor
D. Rhabdomyosarcoma
E. Ewing's sarcoma

81. A 3-year-old boy presents with skin bleeding (ecchymosis and petechial rash) over the lower and upper limbs for two-days, no other site of bleeding and no constitutional symptoms. His mother denies any similar illness in his history and in the family. Examination of this child was normal. CBC film shows isolated thrombocytopenia (33.000/mm^3).
Of the following, the **MOST** appropriate initial treatment is

A. observation
B. steroid
C. IVIG
D. rituximab
E. thrombopoietin receptor agonist

82. What is the **BEST** treatment option for a newly diagnosed immune thrombocytopenia in a 6-year-old naughty child, diagnosed as ADHD in his early childhood?

A. Observation
B. Splenectomy
C. IVIG
D. Rituximab
E. Thrombopoietin receptor agonist

83. What is the **BEST** curative option for a chronic immune thrombocytopenia in a 10-year-old male child, with recurrent epistaxis and gum bleeding that impairs his daily active life style?

A. Observation
B. Splenectomy
C. IVIG
D. Rituximab
E. Thrombopoietin receptor agonist

84. Which of the following is a **TRUE** fact regarding therapy of idiopathic immune thrombocytopenia?

A. The majority of patient have moderate to severe symptoms of bleeding
B. Early therapy does not prevent intracranial hemorrhage
C. Initiating early therapy affect the outcome of the disease
D. Antiplatelets antibodies bind to autologous rather than transfused platelets
E. Aggressive way of management is preferable so as to control bleeding

85. Splenectomy in Immune thrombocytopenia (ITP) is considered in which of the following conditions?

A. A two-year old boy with newly diagnosed ITP whose bleeding symptoms failed to be ameliorated with steroids and IVIG
B. A ten-year old girl with chronic ITP with recurrent symptoms of skin bleeding that is diffuse, not responding to steroids, rituximab and other medical therapy.
C. A six-year old boy with acute ITP developed intracranial hemorrhage from a road traffic accident who is hemodynamically stable and his bleeding is controlled with IVIG and steroids

D. A thirteen-year old girl with chronic ITP and severe vaginal bleeding that necessitated blood transfusion and failed to respond to steroids, IVIG, rituximab and contraceptive therapy.
E. A five-year old boy with acute ITP on initial therapy with steroid and presented with severe epistaxis and consequent anemia declared the need for blood transfusion

86. Passing into the chronic phase of the disease, a patient with immune thrombocytopenia (ITP) should be carefully evaluated to exclude
A. aplastic anemia
B. acute leukemia
C. HIV infection
D. non-hodgkin lymphoma
E. medical child abuse

87. Thrombotic thrombocytopenic purpura is a rare thrombotic microangiopathy characterized by the pentad of
A. Fever, microangiopathic hemolytic anemia, thrombocytopenia, abnormal renal function, and central nervous system changes
B. Pneumonia, microangiopathic hemolytic anemia, thrombocytosis, abnormal renal function, and central nervous system changes
C. Fever, autoimmune hemolytic anemia, thrombocytopenia, abnormal liver function, and central nervous system changes
D. Fever, autoimmune hemolytic anemia, thrombocytosis, abnormal renal function, and central nervous system changes
E. Pneumonia, autoimmune hemolytic anemia, thrombocytosis, abnormal liver function, and central nervous system changes

88. A 14-year-old boy presents with 3-weeks history of fever, infrequent vomiting and irritability, followed by weakness in both lower limbs then progressed to pallor and diffuse petechial rash of the skin, trunk, and lower limbs. He was treated initially as nonspecific infection with injectable antibiotics, but seems fruitless. Regrettably, the last few days witnessed episodes of abnormal movement and disturbed vision. His general examination showed a semiconscious blind adolescent boy with aggressive behavior, having pallor, diffuse skin bleeding, without any edema or lymph nodes enlargement. His meningeal signs were negative and regional exam ENT, chest and abdomen were normal. His neurological exam showed only a lower limb weakness of

grade 3 with diminished reflexes. His CBC showed abnormal RBCs, with schistocytes, spherocytes, helmet cells, and an elevated reticulocyte count in association with thrombocytopenia. Blood urea nitrogen and creatinine are slightly elevated.
Of the followings, the **MOST** likely diagnosis is

A. hemolytic uremic syndrome
B. thrombotic thrombocytopenic purpura
C. acute leukemia
D. aplastic anemia
E. meningococcemia

89. What is the **MOST** appropriate effective initial management of acquired thrombotic thrombocytopenic purpura?

A. Steroids
B. Splenectomy
C. Rituximab
D. IVIG
E. Plasmapheresis

90. A 1-month-old female presented with diffuse skin ecchymosis and petechial rash since early days of life, she was noticed to have occasional blood spits from the mouth. No significant antenatal, neonatal, and family history. Initial CBC shows thrombocytopenia with normal other indices. Other workup tests were normal. BMA shows normal hematopoietic elements with frank absence of megakaryocytes.
Of the following, the **MOST** likely diagnosis is

A. Fanconi anemia
B. congenital dyserythropoietic anemia
C. congenital cytomegalovirus infection
D. congenital leukemia
E. congenital amegakaryocytic thrombocytopenia

91. Which of the following conditions has been linked with Thrombocytopenia-absent radius (TAR)?

A. Cow's milk intolerance
B. Diabetes Mellitus
C. Congestive heart failure
D. Chronic renal failure

E. Bronchiectasis

92. A mother of a 3-month-old girl consulted the pediatric clinic for a concern of accidental discovery of thrombocytopenia during checkup for pallor, which later labeled to be physiological anemia. She denied any clinical symptoms of bleeding in her daughter, further workup failed to reveal any abnormality in the girl, physical examination was completely normal. On further inquiry of the mother who has sensorineural deafness since early life, she reported that she had mild to moderate thrombocytopenia during early childhood that persisted to adulthood with no significant bleeding. Medical archives of both of them showed platelets count of 25,000 – 50,000 /mm^3, sometimes accompanied by neutrophil inclusion bodies, with normal other indices.
Of the following, the **MOST** likely diagnosis is

A. thrombocytopenia-absent radius
B. MYH9-related thrombocytopenia
C. congenital aplastic anemia
D. Wiskott-Aldrich syndrome
E. congenital leukemia

93. Which of the following is considered as diagnostic of splenic hypofunction?

A. Target cells
B. Decreased osmotic fragility
C. Heinz bodies
D. Poikilocytosis
E. Pitted erythrocytes

94. Banti syndrome (congestive splenomegaly) is seen in

A. acute myeloid leukemia
B. amyloidosis
C. juvenile idiopathic arthritis
D. alpha1-antitrypsin deficiency
E. Niemann-Pick disease

95. Which of the following abnormalities constitute Ivemark syndrome?

A. Congenital heart defects, renal anomalies, lung hypoplasia, asplenia and levocardia
B. Congenital heart defects, dextrocardia, bilateral trilobed lungs, asplenia and heterotopic abdominal organs

C. Cyanotic heart defect, skeletal anomalies, retinal detachment, brain cyst and gastroschises
D. Congenital renal anomalies, liver cysts, bowel atresia, omphalocele and congenital abnormalities of the lower limbs
E. Congenital renal anomalies, multiple brain cysts, imperforated anus, prune belly syndrome and lung cysts

96. What are the characteristic RBC changes in peripheral blood smear of patients with splenic hypofunction?

A. Howell-Jolly bodies
B. Basophilic stippling
C. Pappenheimer bodies
D. Cabot rings
E. Hemoglobin crystals

97. What is the **MOST** common pathogen implicated in the etiology of lymphangitis?

A. Group A streptococci
B. Escherichia coli
C. Pseudomonas aeruginosa
D. Clostridium difficile
E. Proteus species

98. Regardless of the presence or absence of systemic symptoms or other abnormal physical findings, which of the following properties should always raise the question of malignancy in a lymph node?

A. Axillary in site
B. Firm and fixed
C. Matted
D. Red overlying skin
E. Tender

99. Which of the following features signifies adenopathy caused by bacteria rather than other infectious agents?

A. Atypical anatomic areas
B. A draining sinus
C. Lack of prior pyogenic infection
D. A short course

E. Unusual clues in the history

100. Which of the following is an unusual site of tuberculous lymphadenopathy?
A. Hilar
B. Supraclavicular
C. Abdominal
D. Cervical
E. Inguinal

101. Which of the following viruses has been implicated as an association with Castleman disease?
A. Measles virus
B. Human herpes virus 8
C. Epstein-Barr virus
D. Parvovirus B19
E. Cytomegalovirus

102. TAFRO syndrome stands for
A. Tetany, Anasarca, Facial nerve paralysis, Rhinitis, and Otitis media
B. Typhlitis, Allergy, Fluid retention, Rheumatological problems, and Organomegaly
C. Thrombocytopenia, Alopecia, Facial nerve paralysis, Rhinitis, and Otitis media
D. Thrombocytopenia, Anasarca, Fever, Reticulum fibrosis, and Organomegaly
E. Tetany, Allergy, Fluid retention, Fever, Reticulum fibrosis, and Otalgia

103. POEMS syndrome stands for
A. Polyneuropathy, organomegaly, endocrinopathy, M-proteins, and skin lesions
B. Polyuria, organomegaly, endocarditis, Myasthenia, and skin lesions
C. Parotid swelling, otitis externa, endocrinopathy, M-proteins and Serositis
D. Polyneuropathy, orchitis, endocrinopathy, Myasthenia, and Serositis
E. Polyuria, organomegaly, edema, M-proteins and skin lesions

Chapter 20

Diseases of the Blood
Answers

HASANEIN H. GHALI

1.(D) Erythrocytes in the fetus are larger than in adults, and at 22-23 wk gestationthe mean corpuscular volume can be as high as 135 femtoliters (fL). Similarly, the mean corpuscular hemoglobin is very high at 22-23wk and falls relatively linearly with advancing gestation. In contrast, the mean corpuscular hemoglobin concentration is constantthroughout gestation at 34 ±1 g/dL.

2.(D) The minor adult hemoglobincomponent, HbA_2 , contains delta (δ) chains and has the structure α2 δ2 . Atbirth, <1.0% of HbA_2 is detected, but by 12 mo of age the normal level is 2.0–3.4%. Throughout life, the normal ratio of HbA to HbA_2 is about 30 : 1.

3.(A) The normal adult level of HbA_2 (2.0–3.4%) is seldom altered. Levels of HbA_2>3.4% are found in most persons with the β-thalassemia trait and in those with megaloblastic anemias secondary to vitamin B12 and folic acid deficiency. Decreased HbA_2 levels are found in those with iron-deficiency anemia and α-thalassemia.

4.(C) Microcytes are seen in Iron deficiency, thalassemias, Lead toxicity, and anemia of chronic disease. Macrocytes are seen in newborns, vitamin B12 or folate deficiency, Diamond-Blackfan anemia, Fanconi anemia, aplastic anemia, liver disease, Down syndrome, and hypothyroidism.

5.(D) Basophil stippling are seen in thalassemia, lead intoxication, and myelodysplasia.

6.(A) Diamond-Blackfan anemia (DBA) is a rare, congenital bone marrow failure syndrome that usually becomes symptomatic in early infancy. More than 90% of cases are recognized in the 1st yr of life. The disorder is characterized by anemia, usually normochromic and macrocytic; reticulocytopenia; and insufficient or absent red blood cell (RBC) precursors in an otherwise normallycellular bone marrow. Up to 50% of affected individuals have additional, extrahematopoietic anomalies.

7.(B) Corticosteroids are a mainstay of therapy, and approximately 80% of patients initially respond. Because corticosteroids impair linear growth as well asphysical and neurocognitive development, many hematologists maintain

infantson chronic transfusion therapy and delay the start of steroids until after age 1 yr.

8.(A) The temporary suppression of erythropoiesis results in reticulocytopenia and moderate to severe normocytic anemia. Some degree of neutropenia occurs in up to 20% of cases. Platelet numbers are normal or elevated.

9.(D) Soluble transferring receptor (sTfR) is a diagnostic test used to distinguish ACD from iron-deficiency anemia (IDA); sTfR levels are high in IDA and normal in ACD.

10.(A) Congenital dyserythropoietic anemia type I CDA I may be diagnosed at any age, most cases are recognized during childhood or adolescence. CDA I is rarely diagnosed in utero. In addition to anemia related symptoms, other findings often include splenomegaly, jaundice, and hepatomegaly. Hemoglobin concentrations generally range between 7 and 11 g/dL. The anemia is usually macrocytic (mean corpuscular volume: 100-120 fL), but normocytic indices may be seen during childhood. Anisopoikilocytosis is appreciated on the peripheral blood smear. In some cases, normoblasts and basophilic stippling of RBCs may be seen. The bone marrow aspirate shows binucleated and, more rarely, multinucleated polychromatophilic erythroblasts are also appreciated.

Electron microscopy is the gold standard for diagnosis, revealing erythroblasts with a characteristic “Swiss cheese” heterochromatin pattern.

11.(D) In the full-term infant, physiologic anemia requires no therapy beyond ensuring that the infant’s diet contains essential nutrients for normal hematopoiesis.

12.(D) Human breast milk, infant formulas, and pasteurized cow’s milk provide adequate amounts of folic acid. Goat’s milk is folate deficient, and supplementation must be given when it is the child’s main food.

13.(A) Certain anticonvulsant drugs (e.g., phenytoin, primidone, phenobarbital) can impair folic acid absorption, and many patients treatedwith these drugs have low serum levels. Frank megaloblastic anemia is rare and readily responds to folic acid therapy, even when administrationof the offending drug is continued.

14.(B) Normal serum folic acid levels are 5-20 ng/mL; with deficiency, levels are <3 ng/mL.

15.(C) Folic acid therapy (0.5-1.0 mg/day) should be continued for 3-4 wk until a definite hematologic response has occurred. Maintenance therapy with a multivitamin (containing 0.2 mgof folate) is adequate.

16.(A) Vitamin B_{12} deficiency in infants is most often nutritional, resulting from low Cbl levels in the breast milk of B_{12}-deficient mothers. Associated

megaloblastic anemia often appears during the 1st yr of life. Maternalm deficiency may be caused by pernicious anemia or gastrointestinal disorders such as Helicobacter pylori infection, celiac disease, Crohn disease, or pancreatic insufficiency. Previous gastric bypass surgery, treatment with proton pump inhibitors, or inadequate intake from a strict unsupplemented vegetarian diet has also been implicated.

17.(C) Imerslund-Grasbeck syndrome is a rare, recessively inherited pediatric disorder resulting in selective vitamin B_{12} malabsorption in the ileum and consequent vitamin B_{12} deficiency. It usually becomes clinically apparent within the 1st 6 yr of life. In addition to megaloblastic anemia, the patient may also have neurologic defects (e.g., hypotonia, developmentaldelay, brain atrophy, movement disorders, dementia) and/or proteinuria.

18.(D) Thiamine-responsive megaloblastic anemia (Rogers syndrome) is a very rare autosomal recessive disorder characterized by megaloblastic anemia, sensorineural deafness, and diabetes mellitus. Congenital heart defects, arrhythmias, visual problems, short stature, trilineage myelodysplasia, and strokes are also described. Thiamine-responsive megaloblastic anemia usually presents in infancy but may occasionally developin childhood and adolescence and occurs in several ethnically distinctpopulations. The bone marrow is characterized not only by megaloblastic changes but also by ringed sideroblasts. Continuous thiamine supplementation usually reverses the anemia and diabetes, but not existinghearing defects.

19.(A) It is therefore necessary to absorb approximately 1 mg daily to maintain positive iron balance in childhood. Because <10% of dietary iron is usually absorbed, a dietary intake of 8-10 mg of iron daily is necessary to maintain iron levels.

20.(D) When the hemoglobin level falls to <5 g/dL, irritability, anorexia, and lethargy develop, and systolic flow murmurs are often heard. If the hemoglobin continues to fall, tachycardia and high output cardiac failure can occur.

21.(E) Detection of increased soluble transferrin receptor and decreased reticulocyte hemoglobin concentration provides very useful and early indicators of iron deficiency, but availability of these tests is more limited.

22.(B)

23.(B) Iron therapy may increase the virulence of malaria and certain gram negative bacteria, particularly in developing countries. Iron over dose is associated with Yersinia infection.

24.(E) Sickle cells: sickle cell disease

Target cells: hemoglobinopathies (HbC, HbS, thalassemia), liver disease

Schistocytes/burr cells/helmet cells/RBC fragments: microangiopathic hemolytic anemia (DIC, HUS, TTP)
Spherocytes: hereditary spherocytosis, autoimmune hemolyticanemia
Cigar-shaped cells: hereditary elliptocytosis
“Bite” cells: G6PD deficiency
Poikilocytosis, microcytosis, fragmented erythrocytes, elliptocytes:hereditary pyropoikilocytosis

25.(D) The mean corpuscular volume (MCV) of HS erythrocytes is low normal or even slightly decreased, and the mean corpuscular hemoglobin concentration (MCHC) is usually increased (>35 g/dL). An MCHC >35.4 g/dL combined with a red cell distribution width (RDW) <14% has been suggested as a screening test for HS.

26.(A) Isoimmune hemolytic disease of the newborn, particularly when a result of ABO incompatibility, closely mimics the appearance of HS. The detectionof antibody on an infant’s RBCs using a direct antiglobulin (Coombs)test should establish the diagnosis of immune hemolysis.

27.(D) Most experts recommend splenectomy for patients with severe HS and believe it should be strongly considered for patients with moderate HS and frequent hypoplastic or aplastic crises, poor growth, or cardiomegaly.
It is generally not recommended for patients with mild HS. When splenectomy is indicated, it should be performed after age 6 yr.

28.(A) HPP is characterized by extreme microcytosis (mean corpuscular volume, 50-65 fL/cell), extraordinary variation in cell size and shape, and microspherocytosis with occasional elliptocytosis.

29.(B) Hereditary xerocytosis (HX), the most common type of the hereditary stomatocytosis syndromes, is a dominant disorder of erythrocyte dehydration.
Affected patients exhibit a mild compensated macrocytic hemolyticanemia with variable degrees of splenomegaly and intermittent jaundice. The MCHC and MCV are elevated, erythrocyte osmotic fragility isdecreased, and K+ concentration and total monovalent cation contentare decreased. There are small numbers of stomatocytes, target cells,and contracted RBCs with hemoglobin puddled to the side on peripheralblood smear.

30.(D) Tangier disease, familial deficiency of high-density lipoproteins(HDLs) is a rare recessive disorder that results from mutationsin the cholesterol and phospholipid transport protein ABCA1, leading to perturbations of cellular cholesterol transport, and resulting in the accumulation of cholesterol esters in many tissues. Hematologic manifestations include a mild to moderate stomatocytic hemolytic anemia and thrombocytopenia. Affected patients can

also have large orange tonsils, hepatosplenomegaly, lymphadenopathy, cloudy corneas, peripheralneuropathy, and premature atherosclerosis.

31.(D) Pancytopenia with bone marrow infiltration can be seen in metastatic solid tumors, myelofibrosis, hemophagocytic lymphohistiocytosis, and osteopetrosis.

32.(B) Inherited pancytopenias account for approximately 30% of cases of pediatric bone marrow failure. Fanconi anemia is considered the most common of the inherited pancytopenias.

33.(B) Amegakaryocytic thrombocytopenia with radioulnar synostosis characterized by thrombocytopenia with progression to pancytopenia. It could be associated with acute myeloid leukemia. The gene is HOXA11.

34.(C) Pearson marrow-pancreas syndrome is characterized by neutropenia with progression to pancytopenia. The gene is mtDNA deletion.

35.(E) The most common congenital anomalies in FA are skeletal and include absence of radii and/or thumbs that are hypoplastic, supernumerary, bifid, or absent. Anomalies of the feet, congenital hip dislocation, and leg abnormalities can also be seen.

36.(A) Thrombocytopenia, red blood cell (RBC) macrocytosis, and increased hemoglobin F, as a result of bone marrow stress, often appear first. Subsequently, patients develop neutropenia and then anemia.

37.(C) Complete blood count shows pancytopenia; bone marrow aspiration and biopsy confirms the hypoplasia of the marrow; chromosomal breakage test is the most commonly available and detects the increased fragility with and without addition of the cross-linking agents but still 10-15% have somatic mosaicism and may not show the high degree of chromosomal fragility; Next-generation sequencing has largely replaced the need for 2-step genetic testing (complementation group testing followed by targeted gene testing) and is most often used.

38.(B)

39.(B) AML, MDS, and rare head, neck and skin tumors.

40.(D) Reticulocytosis and a rise in hemoglobin within 1-2 mo. White blood cell (WBC) counts may increase next, followed by platelet counts.

41.(B) Oral oxymetholone and danazol are the 2 most commonly used androgenic drugs.

42.(E) A response is expected in 70% of patients, treatment is a bridge rather than curative while awaiting for a suitable donor for HSCT or while weighing the risks and benefits of transplant, If a low dose is initially employed, the androgen dose can be increased every 3-4 wk as long as no major side effects

are seen and until the desired response is achieved. If a high dose is initially employed, androgen dosage can be slowly reduced to the minimum dose that maintains the required blood counts. Side effects of androgens include masculinization, increased linear growth, increased mood swings or aggressiveness, elevated hepatic enzymes, cholestasis, peliosishepatis, and liver tumors. Screening for these should be performed regularly. Patients typically stop responding to androgens after several months or years, as their bone marrow failure progresses or as they develop MDS or AML.

43.(C) A heightened risk of expansion of bone marrow cells with clonal cytogenetic abnormalities such as monosomy 7. All other items are true but not a limitation for initiating therapy.

44.(D)

45.(B) Neutropenia 90%, anemia 46%, thrombocytopenia 42%, and pancytopenia 21%.

46.(A) Exocrine pancreatic insufficiency 98%, elevated transaminase 61%, skeletal abnormality 70%, rib cage abnormality 35% and short stature 66%.

47.(C)

48.(C) Although skeletal abnormalities are variable in Shwachman-diamond syndrome, classic findings are metaphyseal dysplasia, osteopenia, delayed appearance of secondary ossification centers, short or flared ribs, and thoracic dystrophy.

49.(A) Isochromosome 7q is particularly common, suggesting that it is a fairly specific clonal marker of SDS and might be related to the presence of mutant SBDS on 7q11. Other clonal chromosome abnormalities include monosomy 7, i(7q) combined with monosomy 7, deletions or translocations involving part of 7q, and deletions of 20q [Del(20q)].

50.(A) A diagnostic triad of mucocutaneous features was proposed when the disorder was first described and included dysplastic nails, lacy reticular pigmentation of the upper chest and/or neck, and oral leukoplakia. However, the triad is not present in all individuals.

51.(D) Skin pigmentation is the commonest (89%) followed by nail dystrophy (88%).

52.(C) Revesz syndrome has many of the features of DC and presents in early childhood in addition to Bilateral exudative retinopathy is required to establish a diagnosis. Patients may also have intracranial calcifications, IUGR, developmental delay, and bone marrow failure.

53.(C) Owing to additional risk of liver and pulmonary fibrosis.

54.(D) The most common anomalies are neurologic and cardiac. Findings related to cerebellar and cerebral atrophy are frequent, and developmental delay is a prominent feature. Congenital heart disease includes atrial and ventricular septal defects, patent ductus arteriosus, tetralogy of Fallot, and coarctation of the aorta.

55.(B)

56.(C) Cytomegalovirus, Epstein-Barr, Hepatitis B, Hepatitis C, Hepatitis non-A, non-B, non-C (seronegative hepatitis) and HIV. Parvovirus B19 is classically associated with isolated red blood cell (RBC) aplasia, but in patients with sickle cell disease or immunodeficiency, it can result in transient pancytopenia.

57.(B) Dose dependent (Chloramphenicol, dapsone, fluorocytosine), and dose independent (Chloramphenicol, dapsone, sulfonamides, tetracycline, methicillin, amphotericin, quinacrine, chloroquine, pyrimethamine).

58.(B) Severe aplastic anemia is defined as a condition in which ≥2 cell components have become seriously compromised (absolute neutrophil count <500/mm3, platelet count <20,000/mm3, reticulocyte count <1% after correction for hematocrit) in a patient whose bone marrow biopsy material has <30% cellularity.

59.(C)

60.(A)

61.(C)

62.(E) Presence of JAK2 or similar mutation. (A) is a major criterion if >25%.

63.(A)

64.(A) Phlebotomy is the initial treatment of choice to alleviate symptoms of hyperviscosity and decrease the risk of thrombosis.

65.(B) Maintain hemoglobin >12.0 g/dl and severe pulmonary disease; maintain hemoglobin >12.0 g/dl during extracorporeal membrane oxygenation; maintain hemoglobin >10.0 g/dl and moderate pulmonary disease; maintain hemoglobin >12.0 g/dl and severe cardiac disease; maintain hemoglobin >10.0 g/dL preoperatively and during major surgery; maintain hemoglobin >7.0 g/dL postoperatively; and maintain hemoglobin >7.0 g/dL and symptomatic anemia.

66.(D) Another factor is the rapid disappearance of EPO from infant plasma (i.e., accelerated metabolism).

67.(D)

68.(C)

69.(E) In patients with inherited disorders, PLT transfusions are justified only if the risk of significant bleeding is quite high or if bleeding is overt, because

inherited PLT dysfunction often is lifelong and repeated transfusions may lead to alloimmunization and refractoriness.

70.(E) For infants below 4 months, granulocyte transfusion is indicated with blood neutrophil count $<3.0 \times 10^9$ /L in 1st wk of life or $<1.0 \times 10^9$ /L thereafter and fulminant bacterial infection.

71.(C) Certain diseases, such as hypothyroidism, and medications, such as valproic acid, can lower VWF levels in affected patients. Repeat testing may be required to rule out or confirm a diagnosis of VWD. Other choices increase the level.

72.(E) Characterized by a defect in the ability of VWF to bind FVIII, also low factor VIII is seen in Type 3.

73.(C) 2B and platelet-type VWD.

74.(A) Desmopressin is beneficial in Type 1 and some type 2VWD with caution.

75.(E)

76.(B)

77.(B) Leukopenia suggests another diagnosis.

78.(A) Chronic Immune thrombocytopenia is an indication for autoimmune assay.

79.(B) Isolated enlargement of the spleen suggests the potential for hypersplenism caused by liver disease or portal vein thrombosis.

80.(B) Autoimmune thrombocytopenia may be an initial manifestation of SLE, HIV infection, common variable immunodeficiency, and rarely, lymphoma or autoimmune lymphoproliferative syndrome.

81.(A) Observation in this condition is best and inexpensive.

82.(C) Rapid elevation of platelets is needed in cases of social hyperactivity.

83.(B) It gives 80% cure.

84.(B) The majority of patients have mild symptoms despite severe thrombocytopenia, early therapy does not affect the outcome of disease, transfused platelets are liable for destruction so as the autologous platelets, and observation and watchful waiting is the best current strategy in cases of mild bleeding.

85.(D) The role of splenectomy in ITP should be reserved for 2 circumstances: (1) the older child (≥4 yr) with severe ITP that has lasted >1 yr (chronic ITP) and whose symptoms are not easily controlled with therapy and (2) when life-threatening hemorrhage (ICH) complicates acute ITP, if the platelet count cannot be corrected rapidly with transfusion of platelets and administration of IVIG and corticosteroids.

86.(C) Autoimmune disease (e.g., SLE), chronic infectious disorders (e.g., HIV), and nonimmune causes of chronic thrombocytopenia, such as type 2B and platelet- type von Willebrand disease, X-linked thrombocytopenia, autoimmune lymphoproliferative syndrome, common variable immunodeficiency syndrome, autosomal macrothrombocytopenia, and Wiskott-Aldrich syndrome

87.(A)

88.(B)

89.(E) Effective in 80–95% of patient.

90.(E) Findings in the BMA study are diagnostic.

91.(A) Intolerance to cow's milk formula (present in 50%) may complicate management by triggering gastrointestinal bleeding, increased thrombocytopenia, eosinophilia, and a leukemoid reaction.

92.(B) MYH9-related thrombocytopeniacomprises a number of diverse hereditary thrombocytopenia syndromes (e.g., Sebastian, Epstein, May-Hegglin, Fechtner) characterized by autosomal dominant macrothrombocytopenia, neutrophil inclusion bodies, and a variety of physical anomalies, including sensorineural deafness, renal disease, and eye disease.

93.(E)

94.(D) Splenomegaly may result from obstruction in the hepatic, portal, or splenic veins leading to hypersplenism. Wilson disease, galactosemia, biliary atresia, and α1-antitrypsin deficiency may result in hepatic inflammation, fibrosis, and vascular obstruction.

95.(B)

96.(A) Howell-Jolly bodies is seen in sickle cell disease, basophilic stippling in lead and heavy metal poisoning, Pappenheimer bodies in sideroblastic anemia, Cabot rings in megaloblastic anemia, and hemoglobin crystals in hemoglobin C disease.

97.(A) Group A streptococci and Staphylococcus aureus.

98.(B) A firm, fixed node should always raise the question of malignancy, regardless of the presence or absence of systemic symptoms or other abnormal physical findings.

99.(D) A prolonged course is more suggestive of infectious agents rather than bacteria.

100.(E)

101.(B) HHV-8 may stimulate excessive production of interleukin-6 (IL-6).

102.(D)

103.(A)

Chapter 21

Cancer and Benign Tumors Questions

HASANEIN H. GHALI

1. Which one of the following malignancies is associated with the use of alkylating agents?
   A. Brain tumors
   B. Hodgkin disease
   C. Osteosarcoma
   D. Neuroblastoma
   E. Hepatoblastoma

2. Which of the following viruses is associated with the development of Kaposi sarcoma?
   A. Epstein-Barr virus
   B. Human herpes virus
   C. Hepatitis B virus
   D. Hepatitis C virus
   E. Human papilloma virus

3. The development of cancer that is linked to genomic imprinting is best represented by:
   A. Bloom syndrome
   B. Trisomy 21
   C. Beckwith-Wiedemann syndrome
   D. Ataxia telangiectasia
   E. Wiskott-Aldrich syndrome

4. Which of the following agents has a side effect of pseudotumor cerebri?
   A. Methotrexate
   B. Vinblastine
   C. Cytarabine
   D. Tretinoin
   E. Etoposide

5. Which of the following agents has a significant possibility of allergic reaction?

A. Etoposide
B. Methotrexate
C. Cisplatin
D. L-Asparaginase
E. Vinblastine

6. Which of the following cancers is more common in school-age children?

A. Hodgkin lymphoma
B. Neuroblastoma
C. Retinoblastoma
D. Wilms tumor
E. Hepatoblastoma

7. Which of the following lymphadenopathies, region-wise, are highly suggestive of malignancy?

A. Anterior upper cervical
B. Post auricular
C. Supraclavicular
D. Axillary
E. Inguinal

8. A “new-onset asthma” symptoms during adolescence may be a presentation of

A. acute lymphoblastic leukemia
B. lymphoma
C. neuroblastoma
D. askin tumor
E. brain tumor

9. PET scan as a part of workup in staging of pediatric cancers is indicated in which of the following malignancies?

A. CNS tumor
B. Neuroblastoma
C. Retinoblastoma
D. Hodgkin lymphoma
E. Hepatoblastoma

10. CSF analysis as a part of work up in staging of pediatric cancers is indicated in which of the following malignancies?

A. Hodgkin lymphoma
B. Wilms tumor
C. Bladder rhabdomyosarcoma
D. Non-Hodgkin lymphoma
E. Osteosarcoma

11. Which of the following chromosomal translocations is associated with acute promyelocytic leukemia?

A. t(9;22)
B. t(1;19)
C. t(15;17)
D. t(2;13)
E. t(11;22)

12. A 6-year-old boy under current treatment of acute lymphoblastic leukemia has exposed to a brother with active chicken pox.
Of the following, the **MOST** appropriate management is

A. oral acyclovir
B. intravenous acyclovir
C. varicella-zoster immunoglobulin
D. close observation for any skin lesions
E. close observation for any temperature rise

13. Among all the aspects of palliative care, the most serious cause of suffering of a cancer child is

A. hair loss
B. psychological trauma
C. pain
D. fear of death
E. fear of loss of function

14. Which of the following agents has been associated with neurocognitive deficits as late effects of cancer treatment?

A. Cyclophosphamide
B. Etoposide
C. Vincristine

D. Daunomycin
E. Methotrexate

15. Which of the following agents has been associated with hearing loss as late effects of cancer treatment?
A. cyclophosphamide
B. carboplatin
C. etoposide
D. vincristine
E. daunomycin

16. Which of the following matches is **TRUE** regarding late effects of cancer therapy and uses of chemotherapies?
A. Renal insufficiency and daunomycin
B. Cardiomyopathy and busulfan
C. Pulmonary fibrosis and melphalan
D. Gonadal dysfunction and procarbazine
E. Peripheral neuropathy and carboplatin

17. Which of the following matches is **TRUE** regarding oncological emergencies and cancers?
A. Hyperkalemia and medulloblastoma
B. Hyponatremia and rhabdomyosarcoma
C. Disseminated intravascular coagulation and lymphoma
D. Spinal cord compression and neuroblastoma
E. Superior vena cava syndrome and hepatoblastoma

18. Which of the following agents has the **LEAST** myelosuppressive effect?
A- Dactinomycin
B- Daunomycin
C- Methotrexate
D- Vincristine
E- Carmustine

19. What is the MOST common primary modality of treatment of childhood cancer?
A. Chemotherapy
B. Surgery

C. Radiotherapy
D. Biologic agent therapy
E. Laser therapy

20. Which of the following tissue cells is **MORE** susceptible to effects of chemotherapy?
A. Neurons
B. Muscle cells
C. Connective tissue
D. Epidermis
E. Bone

21. What is the most common adverse effect of radiation therapy in children?
A. Dermatitis
B. Mucositis
C. Somnolence
D. Alopecia
E. Nausea and diarrhea

22. Which of the following agents is used as prophylaxis in patients with cancer to prevent infection with Pneumocystis jiroveci?
A. Amoxiclavulanic acid
B. Trimethoprim-sulfamethoxazole
C. Fluconazole
D. Ciprofloxacin
E. Acyclovir

23. Patients receiving immunosuppressive therapy should receive irradiated blood products to prevent
A- graft-versus-host disease
B- transfusion-associated reactions
C- infections
D- febrile neutropenia
E- thrombocytopenia

24. Which of the following presenting features is common in acute lymphoblastic leukemia as compared to acute myeloid leukemia?
A. Subcutaneous nodules

B. Disseminated intravascular coagulation
C. Granulocytic sarcoma
D. Mediastinal mass
E. Gingival hypertrophy

25. Which of the following features of acute myeloid leukemia is specific for infants?
A. Blueberry muffin lesions
B. Disseminated intravascular coagulation
C. Chloroma
D. Splenomegaly
E. Gingival hypertrophy

26. Which of the following features is **MORE** common in acute promyelocytic leukemia rather than other types of acute myeloid leukemia?
A. Subcutaneous nodules
B. Disseminated intravascular coagulation
C. Granulocytic sarcoma
D. Splenomegaly
E. Gingival hypertrophy

27. Which of the following features of acute myeloid leukemia is associated with t(8;21)?
A. Subcutaneous nodules
B. Disseminated intravascular coagulation
C. Chloroma
D. Gingival hypertrophy
E. Huge splenomegaly

28. Which of the following chromosomal abnormalities is usually seen in acute promyelocytic leukemia?
A. t(8;21)
B. inv(16)
C. t(15;17)
D. 11q23 abnormalities
E. del(7q), −7

29. Which of the following agents has improved the survival of acute promyelocytic leukemia dramatically and deferred the need for bone marrow transplantation?

A. All-trans-retinoic acid
B. Arsenic trioxide
C. Cytarabine
D. Anthracycline
E. Etoposide

30. A 3-day-old boy with Down syndrome is noted by a family pediatrician as having enlarged liver and spleen during the first checkup visit. The mother states that he is active and good feeder, with no obvious features of sickness. General physical and regional examination was normal apart from hepatosplenomegaly. An initial complete blood count showed Hb 12.0 gm/dl, WBC 55,000/cmm with many blast cells, and platelets 70,000/cmm. BMA showed acute leukemia.
Which of the following facts discussed during counseling is inadmissible?

A. Ten percent of neonates with Down syndrome develop a transient leukemia.
B. These features usually resolve within the first three months of life
C. At time being, less intensive chemotherapy should be instituted to induce remission
D. After resolution, 20-30% will develop acute leukemia in the next 3 years
E. Later on, such cases will develop acute megakaryocytic leukemia

31. Ninety-nine percent of chronic myeloid leukemia cases are characterized by which of the following specific translocation?

A. t(1;19)
B. t(4;11)
C. t(9;22)
D. t(12;21)
E. t(10;14)

32. A 6-year-old boy is referred to the pediatric clinic from a general physician for fever, fatigue and weight loss for the last few weeks. The patient experience pain in the abdomen below the left side of the chest. During examination, he is pale, febrile 38.3 C, no lymphadenopathy, but frank abdominal distension. Abdominal examination shows a huge spleen reaching umbilicus, slightly tender

on palpation. Cardiovascular and neurological examination are normal. Complete blood count shows Hb 9.5 gm/dl, WBC 53,000/cmm, platelets 560,000/cmm. The peripheral blood demonstrated normocytic, normochromic anemia, with leukocytosis, 5% blasts, 17% myelocytes, 5% metamyelocytes, 10% lymphocytes, 2% monocytes, 5% eosinophils and 3% basophils and thrombocytosis. The bone marrow is characteristically hypercellular, with expansion of the myeloid cell line and its progenitor cells. Megakaryocytes are increased.

Of the following, the **MOST** likely diagnosis is

A. Acute lymphoblastic leukemia
B. Toxoplasmosis
C. Acute promyelocytic leukemia
D. Chronic myeloid leukemia
E. Kala-azar

33. Which of the following genetic syndromes has a predilection for juvenile myelomonocytic leukemia?

A. Edward syndrome
B. Noonan syndrome
C. Turner syndrome
D. Klinefelter syndrome
E. Angelman syndrome

34. Which of the following factors is stratified as high risk in treatment of acute lymphoblastic leukemia?

A. Age at diagnosis of 9 years
B. WBC at diagnosis of 33,000/cmm
C. Pre-B-cell detected by flowcytometry
D. Philadelphia chromosome detected by cytogenetic study
E. Rapid response to therapy

35. Which of the following agents is added to the higher risk patient during induction therapy of acute lymphoblastic leukemia?

A. Prednisolone
B. Daunomycin
C. Asparaginase
D. Vincristine
E. Intrathecal methotrexate

36. Which of the following infectious agents requires prophylactic treatment during chemotherapy for acute lymphoblastic leukemia?

A. Pneumocystis jiroveci
B. Streptococcus viridans
C. Leishmania donovani
D. Escherichia coli
E. Pseudomonas auroginosa

37. Which of the following clinical presentations is more frequently encountered in T-cell rather than B-cell acute lymphoblastic leukemia?

A- Hepatosplenomegaly
B- Respiratory distress
C- Bone pain
D- Pallor
E- Fever

38. Which of the following is a B-symptom in Hodgkin lymphoma?

A. Pruritus
B. Anorexia
C. Painful limbs
D. Lethargy
E. Drenching night sweating

39. Which of the following is the typical presentation of Hodgkin lymphoma?

A. Left cervical progressive swelling for 3 months in a six-year old boy; no other constitutional symptoms; active daily living is perfect.
B. A nine-month old boy with shortness of breath due to rapidly progressing mediastinal mass.
C. A twelve-year old girl with fever for 4 months and abdominal distension; examination showed generalized lymphadenopathy and hepatosplenomegaly.
D. Chronic abdominal pain in a ten-year old boy with appearance of jaundice in the last few weeks, sonogram showed big lymph node at the porta hepatis
E. An axillary swelling of few months in a nine-years old girl, with fever and weight loss; history of contact with tuberculosis patient.

40. A 10-year-old boy is recently diagnosed with Hodgkin lymphoma. He has right sided cervical and axillary lymph nodes involvement. No history of fever in the last weeks, with remarkable weight loss about 15% in the last 4 months. Work up and staging have failed to identify more involved lymph nodes groups.
What is the correct stage of the disease for this patient?

A. IB
B. IIA
C. IIB
D. IIIA
E. IIIB

41. A newly diagnosed Hodgkin disease in a thirteen-year old girl with bilateral cervical lymph nodes involvement. The family reported one episode of high fever (40.0C) in the last days, but without weight loss or sweating. Work up showed liver multiple hypoechoic lesion by sonography with high metabolic activity by PET scan suggestive of liver involvement.
What is the correct stage of the disease for this patient?

A. IIB
B. IIIB
C. IIIA
D. IVA
E. IVB

42. What is the cutoff time to use standard treatment rather than myeloablative autologous stem cell transplantation in children with relapsed Hodgkin lymphoma?

A. 6 months
B. 12 months
C. 18 months
D. 24 months
E. 30 months

43. In the perspectives of International Pediatric Non-Hodgkin Lymphoma Staging System, which of the following matches is **CORRECT**?

A. Mediastinal tumor is stratified as stage II.
B. A primary completely resected gastrointestinal tumor is stratified as stage I
C. Any paraspinal tumor is stratified as stage II

D. Two or more nodal sites on the same site of the diaphragm is stratified as stage III
E. A single bone lesion with regional lymph node involvement is stratified as stage II

44. A 6-year-old boy presents with irritant coughing that progressed to shortness of breath for the last week with low grade fever and night sweating. The mother noted that his face had become swollen and slightly dusky in color. On examination, he is febrile (38.5C), no pallor, and no skin bleeding, in moderate respiratory distress, with puffy face and engorged neck vessels; there are dilated superficial veins on the upper chest. Lymph node examination revealed enlarged left supraclavicular lymph nodes. Abdominal examination shows normal soft abdomen with no palpable masses or organomegaly. Initial radiological evaluation shows big mediastinal mass; Complete blood count and bone marrow aspiration are normal, liver and renal function are normal, lactate dehydrogenase is high.
Of the following, the **MOST** likely diagnosis is
A. acute lymphoblastic leukemia
B. neuroblastoma
C. germ cell tumor
D. sarcoma
E. lymphoblastic lymphoma

45. Which of the following types of lymphoma is treated with the same therapeutic approach of childhood acute lymphoblastic leukemia?
A. Burkitt lymphoma
B. Germinal center B-cell like lymphoma
C. Anaplastic large cell lymphoma
D. Lymphoblastic lymphoma
E. Diffuse large B-cell lymphoma

46. Which of the following scenarios of newly diagnosed pediatric non-Hodgkin lymphoma is indicated for radiotherapy as part of treatment?
A. A six-year old girl with huge jaw mass causing difficult breathing and swallowing, diagnosed as Burkitt lymphoma
B. A three-year old boy with big abdominal mass causing pressure effect on the ureters and consequent hydronephrosis diagnosed as diffuse large B-cell lymphoma

C. A two-year boy with intestinal mass with bilateral facial palsies, diagnosed as Burkitt lymphoma
D. A ten-year old boy with acute superior mediastinal syndrome, diagnosed as lymphoblastic lymphoma
E. A six-year-old boy with disseminated anaplastic large cell lymphoma including liver, spleen, and lungs.

47. Which of the following is a recognized complication seen in newly diagnosed patient with bulky Burkitt lymphoma after starting treatment?
A. Disseminated intravascular coagulopathy
B. Septicemia
C. Tumor lysis syndrome
D. CNS encephalopathy
E. Hemorrhagic pancreatitis

48. A 6-year-old boy is recently diagnosed with Burkitt lymphoma. Treatment is started with intravenous fluid hydration and chemotherapy. Three day later, the boy develops tremulousness and unusual spasm of both hands. Review of his charts shows reduced urine output for the last 12 hours with slightly elevated blood pressure. The body weight is increased by 500 gm comparing to that at time of admission. These findings are consistent with:
A. acute renal failure due to chemotherapy
B. tumor lysis syndrome
C. postrenal urinary tract obstruction by the tumor
D. renal failure from lymphomatous infiltration
E. acute renal injury from dehydration

49. What is the most common malignancy below age of 19 years?
A. Leukemia
B. CNS tumor
C. Renal Tumors
D. Lymphoma
E. Sarcomas

50. Lhermitte-Duclos disease (Dysplastic gangliocytoma of the cerebellum) is seen in which of the following familial syndromes?
A. Tuberous sclerosis
B. Von Hippel-Lindau syndrome

C. Li-Fraumeni syndrome
D. Cowden syndrome
E. Turcot syndrome

51. Which of the following familial syndromes' defects is located on Chromosome 17?
A. Neurofibromatosis type 1
B. Neurofibromatosis type 2
C. Tuberous sclerosis
D. Cowden syndrome
E. Von Hippel-Lindau syndrome

52. What is the most common primary brain tumor in children (0-14) year old?
A. Pilocytic astrocytoma
B. Glioblastoma
C. Ependymal tumors
D. Meningioma
E. Pituitary tumors

53. What are the most common congenital and neonatal brain tumors?
A. Teratomas (mature and immature)
B. Embryonal tumors
C. Astrocytic tumors
D. Neuronal tumors
E. Mixed neuronal-glial tumors

54. In young children, the diagnosis of brain tumor may be delayed because
A. the tumor increases in size very slowly due to low proliferation rate
B. the symptoms are similar to common illnesses in this age group such as vomiting
C. there is a good space for these tumors to grow in before producing symptoms
D. the symptoms are usually neglected by the family of the child
E. the diagnosis for brain tumors is difficult in this age group

55. Which of the following features is more frequently seen in supratentorial tumor as compared to infratentorial tumor?
A. Headache

B. Vomiting
C. Focal motor weakness
D. Nausea
E. Torticollis

56. What is the standard neuroimaging modality for diagnosis of primary brain tumors?

A. CT scan
B. MRI
C. PET scan
D. MRA
E. Sonography

57. In addition to the routine evaluation of brain tumors, special consideration should be taken in tumor of Pituitary, suprasellar and optic chiasmal region to look for

A. metabolic assessment
B. respiratory function evaluation
C. endocrine function
D. hematological evaluation
E. cardiac function

58. What is the preferential site for CNS germ cell tumors?

A. Suprasellar
B. Pontine
C. Cerebral cortex
D. Cerebellum
E. Medulla

59. Which of the following statements regarding ependymomas in children is **TRUE**?

A. The median age of presentation is 10 years
B. The majority occur in the supratentorial location
C. Radiologically, it appears like a well-circumscribed lesion with variable enhancement.
D. Tumor in the posterior fossa is associated with better outcome
E. Surgery alone is usually curative

60. What is the **MOST** common CNS embryonal tumor?
    A. Medulloblastoma
    B. Supratentorial PNET
    C. Ependymoblastoma
    D. Medulloepithelioblastoma
    E. Atypical teratoid/rhabdoid tumor

61. Which of the following malignancies is associated with Chloroma?
    A. Acute lymphoblastic leukemia
    B. Acute myeloid leukemia
    C. Non-Hodgkin lymphoma
    D. Neuroblastoma
    E. Medulloblastoma

62. What is the most common extracranial solid tumor in children?
    A. Wilms tumor
    B. Neuroblastoma
    C. Non-Hodgkin lymphoma
    D. Hodgkin Lymphoma
    E. Rhabdomyosarcoma

63. What is the most commonly diagnosed malignancy in infants?
    A. Wilms tumor
    B. Neuroblastoma
    C. Non-Hodgkin lymphoma
    D. Hodgkin Lymphoma
    E. Rhabdomyosarcoma

64. A 4-year-old boy is brought by his mother complaining of bone pain and limping with fever for the last three weeks. The mother notice that he lost weight during this period and his skin color became lighter with significant loss of his usual activity. Also, she noticed that he has been developed a bluish discoloration around both eyes. On examination, he is ill, febrile, pale, with bilateral periorbital ecchymoses, no Lymph node enlargement and no other skin discoloration. Abdominal examination is notable for upper right sided deep-seated abdominal mass that is below the liver. Liver is 2 cm below costal margin and spleen is not palpable.
Of the following, the **MOST** likely diagnosis is

A. Wilms tumor
B. non-Hodgkin lymphoma
C. neuroblastoma
D. leukemia
E. systematic lupus erythematosus

65. The syndrome of Neuroblastoma with massive liver involvement is called
A. Pepper syndrome
B. Horner syndrome
C. Hutchinson syndrome
D. Kerner-Morrison syndrome
E. Neurocristopathy syndrome

66. Limping and irritability in young child with neuroblastoma associated with bone and bone marrow metastasis referred to as
A. Pepper syndrome
B. Horner syndrome
C. Hutchinson syndrome
D. Kerner-Morrison syndrome
E. Neurocristopathy syndrome

67. An 18-month-old boy who is referred to the pediatric hospital for evaluation of chronic diarrhea and failure to thrive for the past few months with no response to any medical therapy or therapeutic formulas. All diarrhea-related lab tests are negative. The accompanied sonography shows a small right sided adrenal gland mass, which is confirmed by CT scan.
Of the following, the **MOST** likely cause of diarrhea is
A. food allergy
B. vasoactive intestinal peptides
C. associated celiac disease
D. small intestinal bacterial overgrowth
E. pancreatic insufficiency

68. Spontaneous regression of neuroblastoma is reported in which of the following clinical scenarios?
A. A three-year old boy with large suprarenal tumor invading the adjacent vessels with isolated bone marrow metastasis.

B. A five-year old girl with small suprarenal mass and diffuse bone marrow involvement.
C. A sixteen-month boy with small suprarenal tumor and widespread subcutaneous tumor nodules, massive liver involvement without bone involvement.
D. A four-month old girl with small mediastinal tumor and Horner syndrome
E. A six-year old girl with paraspinal tumor causing acute paralysis from spinal cord compression

69. What is the **MOST** common metastatic site of Wilms tumor?
A. Lung
B. Bone marrow
C. Brain
D. Spleen
E. Bone

70. A 9-month-old boy develops abdominal distension noticed by the mother for the last few weeks. Past history reveals a birth weight of 5.5 kg and neonatal surgical correction of omphalocele. On examination; he has large tongue, abdominal distention, and asymmetrical appearance of the lower limbs giving an impression of larger left lower limb than the right side. Ultrasonography shows large liver and pancreas.
Of the following, the **MOST** likely diagnosis is
A. WAGR syndrome
B. Denys-Drash syndrome
C. Beckwith-Wiedemann syndrome
D. Perlman syndrome
E. Trisomy 18

71. Features of (Male pseudohermaphroditism with female external genitalia, focal segmental glomerulosclerosis and gonadoblastoma) are linked to Wilms tumor. These features are suggestive of
A. Perlman syndrome
B. Denys-Drash syndrome
C. Frasier syndrome
D. Schimke syndrome
E. Galloway-Mowat syndrome

72. A 10-year-old girl is referred to the hematology/oncology unit after initial diagnosis of Wilms' tumor based on imaging studies. On examination, she has short stature, microcephaly, and multiple café-au-lait spots distributed on the trunk.
Of the following, the **MOST** likely diagnosis is

A. Diamond-Blackfan Anemia
B. Dyskeratosis Congenita
C. Shwachman-Diamond syndrome
D. Cartilage hair hypoplasia
E. Fanconi anemia

73. A 3-year-old boy is referred after initial diagnosis of Wilms tumor based on the imaging studies. He has a history of early onset renal failure and renal mesangial sclerosis and pseudohermaphrodism.
Of the following, the **MOST** likely diagnosis is

A. WAGR syndrome
B. Denys-Drash syndrome
C. Beckwith-Wiedemann Syndrome
D. Frasier Syndrome
E. Perlman Syndrome

74. The clinical characteristics of Polyhydramnios, macrosomia, distinctive facial features, renal dysplasia, nephroblastomatosis, and multiple congenital anomalies is indicative of

A. Simpson-Golabi-Behmel syndrome
B. Costello syndrome
C. Perlman syndrome
D. Sotos syndrome
E. Beckwith-Wiedemann syndrome

75. What is the **MOST** common initial clinical presentation of Wilms tumor?

A. Abdominal pain
B. Abdominal mass
C. Hematuria
D. Hypertension
E. Fever

76. Which one of the following conditions is stratified as stage V disease of Wilms tumor?

A. Bilateral renal involvement
B. Lung metastasis
C. A tumor thrombus extended into the inferior cava
D. Liver metastasis
E. Lymph node metastasis outside the abdominopelvic region

77. What is the **MOST** common solid renal tumor identified in the neonatal period?

A. Mesoblastic nephroma
B. Wilms tumor
C. Clear cell sarcoma
D. Rhabdoid tumor of the kidney
E. Renal cell carcinoma

78. What is the **MOST** common anatomic site affected by Rhabdomyosarcoma?

A. Head and Neck
B. Orbit
C. Hepatobiliary
D. Extremities
E. Retroperitoneal

79. Sarcoma botryoides is a grape like mass of Rhabdomyosarcoma locat in the

A. Vagina
B. Rectum
C. Neck
D. Orbit
E. Cheek

80. Which of the following is a benchmark to differentiate between Ewing and osteogenic sarcomas

A. Age of onsent is the second decades for Ewing, but the third decade for osteogenic
B. Females gender predominates in Ewing while male do so in osteogenic sarcoma
C. Site of predilection is diaphysis of long bones in Ewing while metaphysis of the long bones in osteogenic

D. Fever is rare in Ewing while it is common in osteogenic
E. Lung and bones are common metastatic sites in Ewing while liver and bone marrow in osteogenic sarcoma

81. Rothmund-Thomson syndrome is a rare condition associated with which of the following group of manifestations?

A. Short stature, skin telangiectasia, small hands, and hypoplasticity of the thumbs
B. Short stature, skin melanoma, large hands and feet, and absence of the kidneys
C. Tall stature, skin telangiectasia, large feet, and hypoplasticity of the thumbs
D. Short stature, skin telangiectasia, small hands and feet, and extra-thumbs
E. Tall stature, skin rash, large hands and feet, and hypoplasticity of the thumbs

82. A 13-year-old boy presents with 3-month history of left lower thigh pain and swelling that grew up slowly. He has no history of trauma or injury to the site and no fever or other constitutional features. Activity is normal except for the last few weeks where he started to feel aches and movement restriction. Initial CBC and other biochemical tests were normal but elevated alkaline phosphatase. X-ray of the left thigh shows a lytic lesion in the lower left femur with the classical sunburst pattern.
Of the following, the **MOST** likely diagnosis is

A. Ewing Sarcoma
B. Osteosarcoma
C. Non-Hodgkin lymphoma
D. Simple bone cyst
E. Histiocytosis

83. What is the **MOST** important prognostic factor in osteosarcoma?

A. Age
B. Gender
C. High LDH level
D. Histologic response to chemotherapy
E. Primary site of tumor

84. Why patients with bone tumors may have delay in diagnosis?

A. The pathological differentiation of bone tumors is rather difficult
B. Patients are usually adolescent and reluctant to further diagnostic work up
C. Clinical features are usually nonspecific of the disease itself
D. Symptoms are likely to be attributed to a sport injury
E. Apart from biopsy, no specific diagnostic test is available

85. A 12-year-old boy presented with 2-week history of fever, mild lower back pain, and troublesome standing and walking with fever at day and night. This progressed to complete inability to move his lower limbs that ensued suddenly within the last 24 hours. There was no antecedent history of trauma. On examination, there was bilateral spastic paraplegia with only a flicker of movement at the right ankle joint. Sensory examination revealed mild hypoesthesia to all sensations below the level of D10 dermatome. Tendon reflexes of his lower extremities were exaggerated and Babinski response was extensor bilaterally. Magnetic resonance imaging revealed a lesion in the D9 vertebra compressing the spinal cord.

Of the following, the **MOST** likely diagnosis is

A. Ewing sarcoma
B. rhabdomyosarcoma
C. osteogenic sarcoma
D. leukemia
E. Burkitt Lymphoma

86. What is the characteristic radiographic sign of Ewing sarcoma?

A. Moth-eaten appearance
B. Codman triangles
C. Onion skin appearance of periosteum
D. Sunburst appearance of bone lesions
E. Popcorn calcifications

87. Which of the following malignancies is radioresistant?

A. Ewing Sarcoma
B. Lymphoma
C. Neuroblastoma
D. Osteosarcoma
E. Wilms' tumor

88. Which of the following benign bone lesions can be diagnosed by radiographs only?

A. Langerhans cell histiocytosis
B. Fibrous dysplasia
C. Osteomyelitis
D. Osteochondroma
E. Osteoblastoma

89. A 6-year-old boy presents with right lower thigh swelling that has been noticed for the last few months. Initial radiograph shows lesion in the right femur.
Which of the following symptoms is suggestive of malignant nature of the lesion?

A. Night sweatig
B. Night pain
C. Poor appetite
D. Local erythema
E. Impaired daily activities

90. Which of the following benign bone lesions can be treated by observation only?

A. Osteoblastoma
B. Chondroblastoma
C. Aneurysmal bone cyst
D. Chondromyxoid fibroma
E. Osteochondroma

91. You are the physician responsible for counselling with a family of a 10-year-old boy who is recently diagnosed as having Osteochondroma by radiographic suggestion.
Which one of the following statements regarding his condition is **TRUE**?

A. Surgery is not performed unless the lesion is symptomatic
B. The diagnosis requires core needle biopsy
C. Malignant degeneration is a possible squeal in childhood
D. It is not common during childhood
E. Usually arise from vertebrae and pelvic bone

92. Which of the following conditions has features closely resemble bone infection?

A. Osteochondroma
B. Osteoid osteoma
C. Eosinophilic granuloma
D. Angiosarcoma
E. Aneurysmal bone cyst

93. Maffucci syndrome refers to a condition of angiomas of the soft tissue with multiple

A. Osteochondroma
B. Enchondroma
C. Chondroblastoma
D. Chondromyxoid fibroma
E. Osteoid osteoma

94. What is the classical presenting feature of retinoblastoma?

A. Pain
B. Decreased vision
C. Strabismus
D. Leukocoria
E. Pupil irregularity

95. The diagnosis of retinoblastoma is essentially established by

A. ophthalmological examination
B. MRI brain
C. CT scan brain
D. local Sonography
E. biopsy

96. A common differential diagnosis of retinoblastoma is

A. glaucoma
B. endophthalmitis for toxoplasma gondi
C. idiopathic retinopathy
D. raised intraocular pressure
E. hyperplastic primary vitreous

97. Which of the following conditions is associated with an increased risk for mediastinal germ cell tumors?

A. Down syndrome
B. Klinefelter syndrome
C. Inguinal hernia
D. Testicular microlithiasis
E. Cryptorchidism

98. What is the most common site for teratoma?
A. Ovaries
B. Testes
C. Sacrococcygeal
D. Mediastinal
E. Brain

99. What is the **MOST** common site for Embryonal carcinoma?
A. Ovaries
B. Testes
C. Sacrococcygeal
D. Mediastinal
E. Brain

100. Which of the following gonadal tumors can cause virilization or precocious puberty?
A. Yolk sac tumor
B. Dysgerminoma
C. Sertoli-Leydig cell tumor
D. Seminoma
E. Gonadoblastoma

101. Radiotherapy is important in which of the following Germ Cell Tumors?
A. Testicular tumors
B. Ovarian tumors
C. Mediastinal tumors
D. Central nervous system tumors
E. Sacrococcygeal tumors

102. Which of the following markers provides important confirmation of the diagnosis of Endodermal sinus tumor as well as to monitor the patient for tumor response and recurrence?

A. Human Chorionic Gonadotropin
B. Lactate dehydrogenase
C. Alpha fetoprotein
D. Ferritin
E. C-reactive protein

103. Which of the following subtypes of hepatoblastoma is associated with normal alpha-fetoprotein level and worse outcome?
A. Fetal type
B. Embryonal type
C. Mixed histology type
D. Small cell undifferentiated type
E. Mesenchymal element type

104. Which of the following syndromes is a risk factor for development of hepatocellular carcinoma?
A. Down syndrome
B. Fragile X- syndrome
C. Fanconi syndrome
D. Patau syndrome
E. Alagille syndrome

105. What is the **MOST** common benign tumor of infancy?
A. Neurofibroma
B. Fibrosarcoma
C. Hemangioma
D. Ganglioneuroma
E. Lymphangioma

106. What is the **MOST** common complication of cutaneous hemangioma?
A. Ulceration
B. Secondary infections
C. Bleeding
D. Pain
E. Anemia

107. A 15-year-old boy who recently has been referred as a case of thyroid mass suggestive of tumor by sonography for evaluation and preoperative assessment.

Upon further history gathering and physical examination, you noticed his father was affected with thyroid cancer and underwent surgical resection and medical treatment. On examination, he has marfanoid features and multiple oral mucosal neuroma of the tongue with central midline thyroid swelling of 6*6 cm that moves with swallowing and left sided prominent cervical lymphadenopathy. The mother reported that he has long history of constipation and being unable to cry tears since early childhood. Two years ago, he underwent thorough investigation for multiple endocrine neoplasia.
Of the following, the **MOST** likely diagnosis is

A. differentiated thyroid carcinoma
B. papillary thyroid carcinoma
C. medullary thyroid carcinoma
D. follicular thyroid carcinoma
E. benign thyroid lesion

108. Which of the following organs is highly affected by Langerhans Cell Histiocytosis?

A. Skin
B. Bones
C. Lymph nodes
D. Liver and spleen
E. Lung

109. Which of the following organ involvements in Langerhans Cell Histiocytosis carries a high risk of mortality?

A. Lungs
B. Pancreas
C. Bone marrow
D. Kidneys
E. Brain

110. Which of the following complications of Langerhans cell histiocytosis is irreversible?

A. Pulmonary fibrosis
B. Pancytopenia
C. Chronic Otitis Media
D. Diabetes Insipidus
E. Primary Hypothyroidism

111. A 3-year-old boy presented to the pediatric clinic with history of fever, malaise, weight loss, and irritability for few weeks, with no obvious diagnosis after visiting their family physician and doing initial primary investigations. The history showed no other significant data, but only chronic recurrent “on and off” ear discharge. He also has long history of chronic recurrent atopic dermatitis that was not responding to medical treatment prescribed by dermatologist. On examination, he was febrile, pale, non-reactive, with low growth parameters, and bilateral cervical lymphadenopathies with left sided axillary palpable lymph node. The scalp, armpits and diaper regions showed features of seborrheic dermatitis.
Of the following, the **MOST** likely diagnosis is

A. Rosai-Dorfman disease
B. non-Hodgkin lymphoma
C. Langerhans Cell Histiocytosis
D. immune deficiency syndrome
E. multicentric Castleman disease

112. What is the most common consistent sign of Hemophagocytic Lymphohistiocytosis?

A. Hepatosplenomegaly
B. Lymphadenopathy
C. Respiratory distress
D. Jaundice
E. Neurological signs

Chapter 21

Cancer and Benign Tumors Answers

HASANEIN H. GHALI

1.(C) Osteosarcoma and acute myeloid leukemia.
2.(B) Epstein-Barr virus is associated with lymphoproliferative disorders, Hepatitis B virus and Hepatitis C virus are associated with hepatocellular carcinoma, and Human papilloma virus with cervical cancer.
3.(C)
4.(D)
5.(D)
6.(A) School-age children might present with lymphoma or with brain tumors
7.(C)
8.(B) Lymphoma, especially during adolescence, often manifests as an anterior mediastinal mass. Symptoms such as chronic cough, unexplained shortness of breath, or "new-onset asthma" are typical with this presentation and are often overlooked.
9.(D)
10.(D)
11.(C) t(9;22) is for CML, t(1;19) is for ALL, t(2;13) is for rhabdomyosarcoma, and t(11;22) is for Ewing sarcoma.
12.(C)
13.(C) Pain is a serious cause of suffering among patients with cancer. It may be the result of organ obstruction or compression or bone metastasis, or it may be neuropathic.
14.(E) Methotrexate and cranial radiation.
15.(B) Cisplatin and Carboplatin
16.(D) Renal insufficiency is associated with the use of Ifosfamide, cisplatin and carboplatin. Cardiomyopathy is linked to the use of daunomycin and Adriamycin. Pulmonary fibrosis is linked to the use of bleomycin, busulfan and lomustine. Peripheral neuropathy is linked to vincristine and vinblastine.
17.(D) Hyperkalemia due to tumor lysis syndrome (leukemia), hyponatremia due to SIDH (leukemia and CNS tumors), disseminated intravascular coagulation

due to sepsis and tumor factors (promyelocytic leukemia), superior vena cava syndrome due to superior mediastinal mass (lymphoma).

18.(D)

19.(A)

20.(D) Most susceptible are tissues or organs with high rates of cell turnover: bone marrow, oral and intestinal mucosa, epidermis, liver, and spermatogonia. Least susceptible to chemotherapy and radiation therapy are cells that do not replicate or that replicate slowly, such as neurons, muscle cells, connective tissue, and bone.

21.(A) Dermatitis is the most common general adverse effect, because skin is always in the treatment field.

22.(B)

23.(A) They should receive leukoreduced blood products to prevent transfusion-associated reactions and infections.

24.(D) T-cell leukemia

25.(A) Blueberry muffin lesions or subcutaneous nodules

26.(B)

27.(C) They are discrete masses, known as chloromas or granulocytic sarcomas. These masses can occur in the absence of apparent bone marrow involvement and typically are associated with a t(8;21) translocation. Chloromas also may be seen in the orbit and epidural space.

28.(C) PML-RARA, of favorable prognosis, is a gene rearrangement involving the retinoic acid receptors.

29.(A) Acute promyelocytic leukemia, characterized by a gene rearrangement involving the retinoic acid receptor [t(15;17); PML-RARA], is very responsive to all-trans -retinoic acid (ATRA, tretinoin) combined with anthracyclines and cytarabine. The success of this therapy makes marrow transplantation in first remission unnecessary for patients with this disease.

30.(C) No treatment is indicated.

31.(C) Philadelphia chromosome t(9;22) (q34;q11), resulting in a BCR-ABL fusion protein. This translocation, although characteristic of CML, is also found in a small percentage of patients with ALL.

32.(D)

33.(B) It's association with Noonan syndrome has been shown to have spontaneous resolution.

34.(D) Children who are younger than 1 yr or older than 10 yr or who have an initial leukocyte count of >50,000/µL are considered to be high risk. Additional characteristics that adversely affect outcome include T-cell immunophenotype

or a slow response to initial therapy. Chromosomal abnormalities, including hypodiploidy, the Philadelphia chromosome t(9;22), and MLL gene rearrangements, portend a poorer outcome. More favorable characteristics include a rapid response to therapy, hyperdiploidy, trisomy of specific chromosomes (4, 10, and 17), and rearrangements of the ETV6-RUNX1

35.(B) Daunomycin (Daunorubicin)

36.(A)

37.(B) Respiratory distress usually is related to anemia but can occur in patients as the result of a large anterior mediastinal mass (e.g., in the thymus or nodes). This problem is most frequently seen in adolescent boys with T-cell ALL. T-ALL also usually has a higher leukocyte count.

38.(E) Systemic symptoms**,** classified as B symptoms, that are considered important in staging are unexplained fever >38°C (100.4°F), weight loss >10% total body weight over 6 mo, and drenching night sweats.

39.(A) Typically, patient presented with cervical or supra clavicular swelling.

40.(C) According to Lugano classification for Hodgkin lymphoma.

41.(D) According to Lugano classification for Hodgkin lymphoma.

42.(B) Patients whose disease relapses >12 mo after chemotherapy alone or combined-modality therapy have the best prognosis, and their relapses usually respond to additional standard therapy.

43.(E) Mediastinal tumor is stratified as stage III; a primary completely resected gastrointestinal tumor is stratified as stage II; any paraspinal tumor is stratified as stage III; two or more nodal sites on the same site of the diaphragm is stratified as stage II.

44.(E) Lymphoblastic lymphoma typically manifests as a symptomatic mediastinal mass and also has a predilection for spreading to the bone marrow, CNS, and testes in males.

45.(D) Localized or advanced disease requires 12-24 mo of therapy, including chemotherapy, intrathecal therapy, and cranial radiation in CNS-positive lymphoma

46.(D) Radiation therapy is used only in special circumstances, such as CNS involvement in lymphoblastic lymphoma or the presence of acute superior mediastinal syndrome or paraplegias

47.(C) Newly diagnosed patients, especially those with BL or LBL, are at high risk for TLS. These patients require vigorous hydration, frequent electrolyte monitoring, and either an xanthine oxidase inhibitor (e.g., allopurinol, 10 mg/kg/day orally in 3 divided doses daily) or a recombinant urate oxidase (e.g., rasburicase, 0.2 mg/kg/day intravenously once daily for up to 5 days)

48.(B)
49.(B) The overall mortality among this group approaches 30%. Patients with CNS tumors have the highest morbidity—primarily neurologic—of all children with malignancies.
50.(D) On chromosome 10q23.
51.(A) 17q11, the gene NF1.
52.(A) Pilocytic astrocytoma and medulloblastoma/primitive neuroectodermal tumors are the most common tumors in children 0-14 years.
53.(A) During the 1st yr of life, supratentorial tumors predominate and most often include choroid plexus complex tumors and teratomas.
54.(B)
55.(C) Supratentorial tumors are more frequently associated with focal motor weakness, focal sensory changes, language disorders, focal seizures, and reflex asymmetry. The classical triad of headache, vomiting and nausea is more in favor of infratentorial tumors.
56.(B) MRI with and without gadolinium is the standard neuroimaging.
57.(C)
58.(A) The suprasellar and pineal regions are preferential sites for germ cell tumors.
59.(C) The mean age is 6 years. The majority occurs in the posterior fossa. The most important prognostic factor is the extent of tumor resection. The younger the patient, the poorer the prognosis. Tumor in the posterior fossa is associated with poorer outcome. Surgery is rarely curative.
60.(A)
61.(B)
62.(B)
63.(B)
64.(C)
65.(A) Massive involvement of the liver with metastatic disease, with or without respiratory distress.
66.(C)
67.(B) Intractable secretory diarrhea caused by tumor secretion of vaso-intestinal peptides. Tumors are generally biologically favorable.
68.(C) Infants <18 mo old also can present in unique fashion, termed stage MS (previously 4S; see later), with widespread subcutaneous tumor nodules, massive liver involvement, limited bone marrow disease, and a small primary tumor without bone involvement or other metastases. The stage MS disease can spontaneously regress.

69.(A) Lung, regional lymph nodes and liver.
70.(C)
71.(C)
72.(E) Associated with BRCA2 and PALB2.
73.(B) WT1 missense mutation.
74.(C)
75.(B) Hypertension is present in about 25% of patients at presentation and has been attributed to increased renin activity. Abdominal pain (40%), gross painless hematuria (18%), and constitutional symptoms such as fever, anorexia, and weight loss are other findings at diagnosis.
76.(A) Bilateral renal involvement. B, D, and E are stage IV while C is stage III.
77.(A) Mesoblastic nephroma is the most common solid renal tumor identified in the neonatal period and the most frequent benign renal tumor in childhood.
78.(A) These tumors may occur at virtually any anatomic site but are usually found in the head and neck (25%), orbit (9%), genitourinary tract (24%), and extremities (19%); retroperitoneal and other sites account for the remainder of primary sites. The incidence at each anatomic site is related to both patient age and tumor type.
79.(A) Vaginal rhabdomyosarcoma may manifest as a grapelike mass of tumor tissue bulging through the vaginal orifice, known as sarcoma botryoides, and can cause urinary tract or large bowel symptoms. Vaginal bleeding or obstruction of the urethra or rectum may occur. Similar findings can be noted with uterine primaries.
80.(C) Both occurred in the second decade, both have a slight male predilection, fever is common in Ewing and rare in osteogenic sarcoma, lung and bones are common metastatic site for both.
81.(A) Rothmund-Thomson syndrome is a rare condition associated with short stature, skin telangiectasia, small hands and feet, hypoplasticity or absence of the thumbs, and a high risk of osteosarcoma.
82.(B) The lesion may be mixed lytic and blastic in appearance, but new bone formation is usually visible. The classic radiographic appearance of osteosarcoma is the sunburst pattern.
83.(D) One of the most important prognostic factors in osteosarcoma is the histologic response to chemotherapy; a poor histologic response is ≥10% viable tumor.
84.(D)
85.(A) The differential diagnosis for a paraspinal/perivertebral mass in a child is wide and includes benign and malignant etiology. Malignant lesions such as

chondrosarcoma, Ewing sarcoma, peripheral neuroectodermal tumor (PNET) and neuroblastoma metastasis, amongst others. Benign lesions would not usually show bone destruction.

86.(C) A, B and D are for osteosarcoma, while A and D are for chondrosarcoma.

87.(D) The other choices are radiosensitive.

88.(D) Osteochondroma, nonossifying fibroma, unicameral bone cyst, and enchondroma can be diagnosed by radiographs only.

89.(B) Night pain that awakens a child suggests malignancy, but relief of such pain with aspirin is common with osteoid osteomas

90.(E) Other choices are locally aggressive and need to be treated.

91.(A) It is the most common benign lesion in childhood, usually arise from the long bones, with very rare malignant degeneration. It is diagnosed by radiograph alone unless it is suspicious of malignant condition.

92.(C) And Ewing Sarcoma

93.(B) It has a high rate of malignant transformation

94.(D) Retinoblastoma classically presents with leukocoria, a white pupillary reflex, which often is first noticed when a red reflex is not present at a routine newborn or well-child examination or in a flash photograph of the child.

95.(A) The diagnosis is established by the characteristic ophthalmologic findings of a chalky, white-gray retinal mass with a soft, friable consistency. Imaging studies are not diagnostic, and biopsies are contraindicated.

96.(E) The differential diagnosis of retinoblastoma includes other causes of leukocoria, including persistent hyperplastic primary vitreous, Coats disease, vitreous hemorrhage, cataract, endophthalmitis from *Toxocara canis*, choroidal coloboma, retinopathy of prematurity, and familial exudative vitreoretinopathy.

97.(B) All other distractors are associated with increased risk of testicular cancer.

98.(C)

99.(B)

100.(C) Sertoli-Leydig cell tumors and granulosa cell tumors produce hormones that can cause virilization, feminization, or precocious puberty, depending on pubertal stage and the balance between Sertoli cells (estrogen production) and Leydig cells (androgen production).

101.(D) Except for tumors of the central nervous system, radiation therapy is limited to those tumors that are not amenable to complete excision and are refractory to chemotherapy.

102.(C) The serum α-fetoprotein level is elevated with endodermal sinus tumors and may be minimally elevated with teratomas.

103.(D) The pure fetal histology subtype predicts a more favorable outcome, and the small cell undifferentiated subtype is associated with normal AFP levels and predicts a worse outcome.

104.(E) Hepatocellular carcinoma occurs mostly in adolescents and often is associated with hepatitis B or C infection. It also occurs in the chronic form of hereditary tyrosinemia, galactosemia, glycogen storage disease, α1 -antitrypsin deficiency, progressive familial intrahepatic cholestasis, and biliary cirrhosis. Alagille syndrome and aflatoxin B contamination of food are associated risk factors.

105.(C) Hemangiomas, the most common benign tumors of infancy, occur in approximately 5–10% of term infants.

106.(A) Ulceration may lead to secondary infection.

107.(C) Medullary thyroid carcinoma is an uncommon disease in childhood that almost always occurs in the context of an autosomal dominant, hereditary endocrine tumor syndrome that arises secondary to activating mutations in the RET protooncogene: multiple endocrine neoplasia type 2a (MEN2A) or type 2b (MEN2B).

108.(B) The skeleton is involved in 80%, Skin in 50%, Lymph nodes in 33%, Hepatosplenomegaly in 20%, and lungs in 10%.

109.(C) The risk organs are liver, spleen, and the hematopoietic (bone marrow) system.

110.(A) Late (fibrotic) complications, whether hepatic or pulmonary, are irreversible and require organ transplantation to be definitively treated.

111.(C)

112.(A) Physical examination often reveals hepatosplenomegaly (70–100%), lymphadenopathy (20–50%), respiratory distress (40–90%), jaundice, and symptoms of CNS involvement (50%) that are not unlike those of aseptic meningitis or acute demyelinating encephalomyelitis.

Chapter 22

Nephrology Questions

QAHTAN ALOBAIDY

1. When the blood passes through the glomerular capillaries, plasma is filtered through the glomerular capillary walls.
Which of the following plasma molecules filter freely through the glomeruli?

A. Peptides
B. Albumin
C. Globulins
D. Interleukin-6
E. Myoglobin

2. The “bedside” Schwartz formula is the most widely used pediatric formula for GFR estimation, and is based on the an empirical constant, patient height, and which of the following?

A. Inulin
B. Cystatin c
C. Blood urea
D. Serum creatinine
E. Fructose polymer

3. Which of the following urinalysis results is considered as significant hematuria in school age children?

A. > 10 RBCs/HPF
B. > 20 RBCs/HPF
C. > 30 RBCs/HPF
D. > 40 RBCs/HPF
E. > 50 RBCs/HPF

4. Which of the following causes heme-positive red urine?

A. Homogentisic acid
B. Myoglobin
C. Methemoglobin

D. Porphyrin
E. Urates

5. What is the characteristic finding in upper urinary tract hematuria in urine examination?
A. Leukocytes casts
B. Isomorphic RBCs
C. Bright red urine
D. Blood clots in urine
E. Proteinuria>100 mg/dL

6. Which of the following distinguish IgA nephropathy from post infectious glomerulonephritis?
A. Gross hematuria
B. Proteinuria
C. Normal C_3 level
D. RBC casts
E. Serum IgA level

7. A 13-year-old boy presents with progressive proteinuria, his brother on regular hemodialysis sessions, wearing glasses for his eyes problem and is also suffering from hearing impairment.
Which of the following ophthalmological findings is pathognomonic for their disease?
A. Macular flecks
B. Recurrent corneal erosions
C. Anterior lenticonus
D. Optic nerve atrophy
E. Retinitis pigmentosa

8. A 7-year-old male presents with sudden onset of gross hematuria and edema. Blood pressure is 130/90 mmHg. Investigations results; blood urea 160mg/dl, low C3 level, and urinalysis shows RBC casts. He had history of treatment for acute tonsillitis with penicillin for 10 days.
Which of the following is the **MOST** likely diagnosis?
A. IgA nephropathy
B. Alport syndrome
C. Essential mixed cryoglobulinemia

D. Post infectious glomerulonephritis
E. Membranoproliferative glomerulonephritis

9. Which of the following neurologic sequelae is seen in patients with post streptococcal glomerulonephritis and severe prolonged hypertension?
A. Stroke
B. Seizures
C. Brain edema
D. Intracranial bleeding
E. Basal ganglions calcification

10. Which of the following can cause low serum complement (C3, CH 50)?
A. Polyaretritis nodosa
B. Goodpasture syndrom
C. Hypersensitivity vasiculitis
D. Henoch-Schönlein Purpura
E. Subacute bacterial endocarditis

11. A 4-year-old boy had three days history of hematuria and mild puffy face. Blood pressure was 140/90 mmHg. He underwent surgery for hydrocephalus one month ago. Urinalysis: RBC +++, protein +, granular cast +, hyaline cast +, blood urea 80 mg/dl, serum creatinine 0.9 mg/dl and low complement level.
Which of the following is the **MOST** likely organism causing this complication?
A. Streptococcus viridans
B. Staphylococcus epidermidis
C. Staphylococcus aureus
D. Klebsiella pneumoniae
E. Pseudomonas aeruginosa

12. A 15-year-old female presented with fever, fatigue, and generalized edema. On examination you find malar discoid rash, investigations shows Hb 7 g/dl, WBC $10*10^3$uL, platelet $50*10^3$uL, positive antinuclear antibody test, serum albumin 2.2mg/dl, total serum protein 4mg/dl, and urinalysis revealed +++ proteinuria.
Which of the following is the **MOST** likely class of this disease?
A. I
B. II
C. III

D. IV
E. V

13. A 10-year-old girl, a known case of beta thalassemia and hepatitis C infection, presents with puffiness. Blood pressure was 140/90 mmHg, urinalysis RBC+++, blood urea 95 mg/dl, serum creatinine 0.9 mg/dl.
Of the following, the **MOST** likely diagnosis is

A. membranous nephropathy
B. membranoproliferative glomerulonephritis
C. lupus nephritis
D. post streptococcal glomerulonephritis
E. Immunoglobulin A nephropathy

14. Which of the following are the **MOST** common post-renal transplant bacterial infections?

A. Urinary tract infections
B. Osteomyelitis
C. Peritonitis
D. Meningitis
E. Sinusitis

15. Which of the following is an option for treatment of acute cellular rejection post renal transplant?

A. Plasmapheresis
B. Intravenous immunoglobulin
C. Thymoglobulin
D. Rituximab
E. Proteasome inhibitors

16. The natural history of most forms of crescentic glomerulonephritis (CGN) is rapid and relentless progression to end-stage renal failure.
Which of the following **RARELY** progresses to CGN?

A. Henoch-Schönlein Purpura
B. Microscopic polyangiitis
C. Immunoglobulin A nephropathy
D. Goodpasture disease
E. Systemic lupus erythematosus

17. Which of the following microorganisms is blamed in causing hemolytic-uremic syndrome?

A. Streptococcus pneumonia
B. Mycoplasma hominis
C. Urea plasma urealyticum
D. Staphylococcus epidermidis
E. Klebsiella pneumoniae

18. A 2-year-old toddler is admitted to hospital with diarrhea and lethargy. There is a known local outbreak of E coli 0157:H7 and his initial blood investigations showed evidence of acute renal failure and anemia.
Which of the following investigation results would be expected?

A. Prolong prothrombin time
B. Fragmented red blood cells
C. Thrombocytosis
D. Right-shift of the white blood cells
E. Raised serum haptoglobins

19. What is the characteristic lab finding of pneumococci-induced hemolytic-uremic syndrome?

A. Prolong prothrombin time
B. Macroscopic hematuria
C. Thrombocytosis
D. Coombs positive hemolytic anemia
E. High-grade proteinuria

20. A 14-year-old girl with cystinuria disease on D-penicillamine treatment for the last 6 months, presented with generalized edema, decreased urine output, and blood pressure of 140/95 mmHg. Urinalysis; pus cell ++, RBC +, albumin+++, blood urea 60 mg/dl, total serum protein 4.5mg/dl, serum albumin 2.2mg/dl, serum cholesterol 300mg/dl, and normal serum C3 and C4.
Of the following, the **MOST** likely diagnosis is

A. focal segmental glomerulonephritis
B. membarenoprolifrative glomerulonephritis
C. membranous glomerulopathy
D. minimal change nephrotic syndrome
E. rapidly progressive glomerulonephritis

21. A full term baby born to a 33-year-old diabetic mother presents with gross hematuria and a palpable mass in the right flank. His blood pressure 100/60 mmHg, blood urea 80 mg/dl, serum creatinine 1.3 mg/dl with a maternal serum creatinine prior to delivery of 0.9 mg/dl, and his complete blood count: HGB 12 g/dl, WBC $17*10^3$ uL, platelet count $53*10^3$uL with normal PTT, PT, and fibrinogen.
Of the following, the **MOST** likely diagnosis is

A. Wilms tumor
B. multicystic dysplastic kideny
C. renal vein thrombosis
D. polycystic kidney disease
E. neuroblastoma

22. You are reviewing a 15-year-old boy who had recently been diagnosed with autosomal dominant polycystic kidney disease (ADPKD) in the renal clinic. You proceed to examine his cardiovascular system.
What other features you expect to find on examination?

A. Dilated cardiomyopathy
B. Mitral stenosis
C. Aortic stenosis
D. Renal bruit secondary to renal artery stenosis
E. Mitral valve prolapse

23. An 8 -year-old boy presents with severe anemia and polyuria. Examination revealed growth failure, retinal degeneration with blood urea of 66 mg/dl and serum creatinine of 3 mg/dl.
Of the following, the **MOST** likely diagnosis is

A. infantile polycystic disease
B. autosomal recessive polycystic kidney disease
C. nephronophthisis
D. Bardet-biedl syndrome
E. Von hippel-lindau syndrome

24. What is the **MOST** common virus causing hemorrhagic cystitis?

A. Para influenza
B. Adenovirus
C. Parvo virus
D. Enterovirus

E. Rhinovirus

25. Which of the following immunosuppressant drugs had a high rate of aphthous ulcers, dyslipidemia, and poor wound healing?

A. Tacrolimus
B. Corticosteroids
C. Cyclosporine
D. Mycophenolate mofetil
E. Sirolimus

26. Which of the following is the **BEST** method to predict proteinuria in patient with diabetes mellitus?

A. Microalbuminuria test
B. Dipstick testing
C. Boiling test
D. Spot urine for protein/creatinine ratio
E. 24 hr urine for protein and creatinine excretion

27. Which of the following can cause false-negative proteinuria?

A. Alkaline PH
B. Dilute urine
C. Oliguria
D. Hematuria
E. Myoglobulinuria

28. Which of the following conditions can be associated with microalbuminuria?

A. Obesity
B. Hyperlipidemia
C. Renal vein thrombosis
D. Cirrhosis
E. Leukemia

29. An 8-year-old boy with steroid dependent nephrotic syndrome presented to the emergency unit with fever and abdominal pain. Examination reveals moderate ascites and abdominal tenderness; aspiration of peritoneal fluid shows peritoneal leukocyte counts 350 cells/µL.
Of the following, the **MOST** likely microorganism isolated from the ascitic fluid is

A. Streptococcus viridans

B. Staphylococcus epidermidis
C. Escherichia coli
D. Streptococcus pneumonia
E. Enterococcus

30. Which of the following types of nephrotic syndrome had the worst prognosis?
A. Focal segmental glomerulosclerosis
B. Membranoproliferative glomerulonephritis
C. C3 glomerulopathy
D. Membranous nephropathy
E. Minimal change disease

31. Which of the following predispose to the development of venous thromboembolism in patients with nephrotic syndrome?
A. Decreased platelet number
B. Loss of antithrombin III
C. Hemodilution
D. Loss of fibrinogen
E. Reduced metabolism of vitamin K

32. A 10-year-old child with steroid dependent nephrotic syndrome and frequent relapses, his height below third centile. The parents are concerned about his short stature.
Which of the following is the **MOST** appropriate strategy to improve linear growth in their child?
A. Reduce the total dose of steroid
B. Use single dose regimen
C. Start growth hormone therapy
D. Follow steroid-sparing strategies
E. Combine calcium with steroid therapy

33. Which of the following is a preventable side effect of cyclophosphamide?
A. Neutropenia
B. Alopecia
C. Hemorrhagic cystitis
D. Premature gonadal failure
E. Increased risk of future malignancy

34. During cyclophosphamide therapy in children with frequently relapsing and steroid-dependent nephrotic syndrome; the white blood cell count must be monitored weekly and the drug should be withheld if the count falls below

A. 2,000/mm^3
B. 3,000/mm^3
C. 4,000/mm^3
D. 5,000/mm^3
E. 6,000/mm^3

35. Which of the following is the **MOST** appropriate initial treatment to reduce proteinuria in minimal change disease?

A. Protein restriction in diet
B. Lipid lowering agent
C. Angiotensin converting enzyme inhibitor
D. Loop diuretics
E. Prednisolone

36. Which of the following drug is recommended as initial therapy for children with steroid-resistant nephrotic syndrome?

A. Tacrolimus
B. Mycophenolate mofetil
C. Levamisole
D. Chlorambucil
E. Azathioprine

37. A 17-year-old girl with nephrotic syndrome is noted to have symmetrical marked loss of adipose tissue, first manifests in the face and then gradually progresses to the upper extremities, thorax and upper abdomen. Investigations reveal low complement level.
Which type of nephrotic syndrome is associated with these manifestations?

A. Membranoproliferative glomerulonephritis type II
B. Focal segmental glomerulonephritis
C. Minimal change disease
D. Renal vein thrombosis
E. Membranous glomerulonephritis

38. Which of the following urine findings can differentiate classic distal renal tubular acidosis (type I) from proximal renal tubular acidosis (type II)?

A. Phosphaturia
B. Aminoaciduria
C. Hypercalciuria
D. Glycosuria
E. Uricosuria

39. A 4-year-old mentally handicapped boy presents by his parents to outpatient clinic as severe rickets and poor vision. Examination shows severe bow legs, growth failure, and hypotonia. Investigations reveal ABG: PH 7.2, Na^{2}132 mg/dl, k^{+}2.5 mg/dl, HCO_3 6 mEq/l, and Cl^{-}115 mg/dl, Blood urea 65mg/dl, urinalysis show glucose ++, phosphate +, and urine PH 5.
Of the following, the **MOST** likely diagnosis is

A. cystinosis
B. Lowe syndrome
C. chronic renal failure
D. vitamin D resistant rickets
E. Dent's disease

40. Which of the following is the **BEST** way to confirm the diagnosis of cystinosis?

A. Detection of cystine crystals in the cornea
B. Measurement of leukocyte cysteine content
C. Normal anion gap (hyperchloremic) metabolic acidosis
D. Measurementof amino acids in urine
E. Cysteine urine concertation

41. Which of the following findings can differentiate renal tubular acidosis (type IV) from other types?

A. Hyperchloremia
B. Hyperkalemia
C. Nephrocalcinosis
D. Normal anion gap metabolic acidosis
E. Normal glomerular filtration rate

42. Which of the following types of renal tubular acidosis (RTA) typically cause urine pH >6?

A. Distal
B. Proximal

C. Combined proximal and distal
D. Hyperkalemic type
E. Cystinosis

43. Which of the following types of renal tubular acidosis (RTA) is associated with hypercalciuria?

A. Distal (type I) RTA
B. Proximal (type II) RTA
C. Combined proximal and distal (type III) RTA
D. Hyperkalemic (type IV) RTA
E. Nephropathic cystinosis

44. Which of the following causes false low bicarbonate level in interpretation of arterial blood gas analysis?

A. Traumatic blood draws
B. Large volumes of blood sampling
C. Addison disease
D. Chronic diarrhea
E. Diabetic ketoacidosis

45. A 6-month-old boy presents with polyuria. Examination shows irritability, tachycardia, tachypnea, and moderate dehydration. Investigations reveal blood urea 66 mg/dl, serum creatinine 0.5mg/dl, ABG: PH 7.36, HCO3 26mEq/l, Na^{+2} 155mg/dL, CL^{-} 105mg/dL, K^{+}3.8mg/dL, lactate 2.9mmol/L, serum osmolality 293 mOsm/kg, and urine osmolality 280 mOsm/kg.
Of the following, the **MOST** likely diagnosis is

A. Bartter syndrome
B. acute kidney injury
C. diabetes insipidus
D. distal renal tubular acidosis
E. medullary sponge kidney

46. A first cousin parents brought their 13-month-old toddler with hyperthermia and massive polyuria. Examination reveals failure to thrive, tachycardia, tachypnea, and normal blood pressure. Investigations reveal blood urea 70mg/dL, serum creatinine 0.5mg/dL, ABG pH 7.55, HCO3 32 mEq/l, Na^{+2} 127mg/dL, CL^{-} 80mg/dl, K^{+}2.8mg/dL, and lactate 2mmol/L, serum osmolality

290 mOsm/kg, with a urine osmolality of 280 mOsm/kg; abdominal ultrasound shows renal pyramids calcifications
Of the following, the **MOST** likely diagnosis is

A. Bartter syndrome
B. Gitelman Syndrome
C. diabetes insipidus
D. EASTsyndrome
E. medullary sponge kidney

47. Which of the following tubulopathy is characterized by low-molecular-weight proteinuria, hypercalciuria, glycosuria, aminoaciduria, and phosphaturia?

A. Gordon syndrome
B. Cystinuria
C. Liddle syndrome
D. Dent disease
E. Bartter syndrome type V

48. A mother with positive history of polyhydramnios in her pregnancy consults you about her premature baby boy with massive polyuria, on careful examination; you find he is suffering from sensory neural deafness. Investigations reveal arterial pH 7.55, HCO3 32 mEq/l, Na^{+2} 127 mg/dl, Cl^{-} 80mg/dl, K^{+} 2.8 mg/dl.
Of the following, the **MOST** likely diagnosis is

A. Liddle syndrome
B. Gordon syndrome
C. medullary sponge kidney
D. Bartter syndrome type IV
E. chronic renal failure

49. Which of the following syndromes is characterized by hyperaldosteronism, hypertension, hypokalemia, and alkalosis?

A. Liddle
B. Bartter
C. Gordons
D. Gitelman
E. milk-alkali

50. You have a call from the senior house officer about a newborn admitted before 3 days in the neonatal care unit, asking you about his serum creatinine which has been increased 2.5 times of the baseline and his urine output <0.5 mL/kg/hr. for last 12 hr.
Which of the following stage of renal injury he had?

A. Acute kidney injury stage 1
B. Acute kidney injury stage 2
C. Acute kidney injury stage 3
D. Chronic renal failure stage 1
E. Chronic renal failure stage 2

51. Which of the following tests is **MOST** useful in determining prerenal azotemia from acute tubular necrosis?

A. Serum urea level
B. Hemoglobin concentration
C. Plasma osmolality
D. Urinary urea
E. Urinary sodium

52. A 14-year-old boy presents to the emergency department after a crush injury to his right leg. Radiography reveals no underlying fractures. One hour after the injury, he develops worsening pain, swelling, and paresthesia of the injured leg. Investigations reveal, high lactate, serum creatinine kinase, and myoglobin level; arterial pH, 7.29; potassium, 5.8 mmol/L; and low glomerular filtration rate.
Of the following, the **MOST** appropriate initial management is to

A. estimate urine output
B. start high rate intravenous fluid
C. give intravenous mannitol 20%
D. give loading dose furosemide
E. arrange for urgent dialysis

53. A 4-year-old boy presents to emergency unit as dark color urine following ingestions of fava bean. Examination shows pallor, jaundice and hypotension. Investigations reveal blood urea 80 mg/dl, serum creatinine 1.3 mg/ dl, and urinalysis show broad, brownish granular casts, protein nil, high both urinary sodium and fractional excretion of sodium, and high urinary osmolality.
Which of the following is the **MOST** likely cause of his acute kidney injury?

A. Hypovolemia
B. Acute tubular necrosis
C. Acute interstitial nephritis
D. Glomerulonephritis
E. Tubular obstruction

54. A 5-year-old boy presents with fever, maculopapular rash, and mild arthralgia 3 days following ingestion of ibuprofen. Investigations reveal blood urea 80 mg/dl, serum creatinine 1.3 mg/ dl, and urinalysis show white blood cells, eosinophils, and cellular casts.
Which of the following is the **MOST** likely cause of his acute kidney injury?

A. Hypovolemia
B. Acute tubular necrosis
C. Acute interstitial nephritis
D. Glomerulonephritis
E. Tubular obstruction

55. Which of the following is the **MOST** likely cause of raised phosphate level in patients with chronic kidney disease?

A. Decreased renal excretion
B. Increase gut absorption
C. Hypervitaminosis D
D. Secondry hyperparathyroidism
E. Decreased 25-alpha hydroxylation of vitamin D

56. Which of the following is the leading cause of death in childhood chronic renal failure?

A. Central nervous disease
B. Cardiovascular disease
C. Respiratory disease
D. Hematological disease
E. Gastrointestinal disease

57. Which of the following is not restricted regarding the dietary management of chronic renal disease?

A. Protein
B. unsaturated Fat
C. Salt

D. Saturated Fat
E. Fluid

58. What is the sufficient level of vitamin D 25-hydroxycalciferol in patients with chronic renal failure?
A. ≥20 ng/ mL
B. ≥ 30 ng/mL
C. ≥ 40 ng/mL
D. ≥ 50 ng/mL
E. ≥ 60 ng/mL

59. Which of the following is the **BEST** time for ambulatory blood pressure monitoring to diagnose hypertension in patient with chronic renal disease?
A. Twice/day
B. Over 24 hr
C. Twice/week
D. Weekly
E. Every two week

60. A 4-year-old male with stage 4 chronic renal disease, serum calcium 8 mg/dl, serum phosphorus 7 mg/dl. You advised the patient to follow a low-phosphorus diet and to enhance gastrointestinal phosphate excretion by using phosphate binders.
Which of the following is the **BEST** time to give such treatment?
A. Before meals
B. After meals
C. With meals
D. Before sleep
E. Early morning

61. Which of the following is the **BEST** level of serum bicarbonate to be maintained in chronic renal disease?
A. ≥ 14 mEq/L
B. ≥ 16 mEq/L
C. ≥ 18 mEq/L
D. ≥ 20 mEq/L
E. ≥ 22 mEq/L

62. You are evaluating a child on hemodialysis with refractory anemia in spite using iron supplementation and erythropoietin. His parathyroid hormone level is elevated in the last year.
Which of the following is the **MOST** common cause of his refractory anemia?

A. Occult blood loss
B. Chronic infection
C. Folate deficiency
D. Bone marrow fibrosis
E. Vitamin B12 deficiency

63. Which of the following are the antihypertensive medications of choice in all children with pediatric CKD?

A. ACE inhibitors
B. β-adrenergic blockers
C. Vasodilators
D. Loop diuretics
E. Calcium channel blockers

64. Which of the following is a modifiable risk factor for CKD progression?

A. Older age
B. CKD severity
C. Onset of puberty
D. Glomerular etiology of renal disease
E. Persistent nephrotic range proteinuria

65. Which of the following substances is responsible for osmotic gradient created in the dialysis fluid used in peritoneal dialysis?

A. Sodium
B. Potassium
C. Protein
D. Dextrose
E. Calcium

66. Parents are concerned about immunization of their son who recently underwent renal transplant.
Which of the following vaccines should be avoided to this child as well as his family members?

A. Varicella-Zoster

B. Inhaled influenza
C. Hepatitis A
D. MMR
E. Measles

67. Which of the following immunosuppressant drugs can cause new-onset diabetes after transplant (NODAT)?

A. Tacrolimus
B. Corticosteroids
C. Cyclosporine
D. Mycophenolate mofetil
E. Azathioprine

Chapter 22

Nephrology Answers

QAHTAN ALOBAIDY

1.(A) Small plasma molecules filter freely (e.g., electrolytes, glucose, phosphate, urea, creatinine, peptides, low-molecular weight proteins), whereas larger molecules are retained in the circulation (such as albumin and globulins).

2.(D) Because true measurement of the GFR is expensive and time consuming, the GFR is commonly estimated (eGFR) by the clearance of endogenous creatinine.

The "bedside" Schwartz formula is the most widely used pediatric formula and is based on the serum creatinine (Scr), patient height, and an empirical constant:

eGFR = 0.413× height (cm)/S_{cr} (mg/dL).

3.(E) Hematuria, defined as the persistent presence of more than 5 red blood cells (RBCs)/high power field (HPF) in uncentrifuged urine, occurs in 4–6% of urine samples from school-age children. The presence of 10-50 RBCs/µL may suggest underlying pathology, but significant hematuria is generally considered as > 50 RBCs/HPF.

4.(B) Most important causes of heme positive red urine are hemoglobin and myoglobin while the metabolites that cause heme negative red urine are homogentisic acid, melanin, methemoglobin, porphyrin, tyrosinosis and urates.

5.(E) Hematuria from within the glomerulus is often associated with brown, cola- or tea-colored, or burgundy urine, proteinuria > 100 mg/dL via dipstick, urinary microscopic findings of RBC casts, and deformed urinary RBCs (particularly acanthocytes). Hematuria originating within the tubular system may be associated with the presence of leukocytes or renal tubular casts. Lower urinary tract sources of hematuria may be associated with gross hematuria that is bright red or pink, terminal hematuria (gross hematuria occurring at the end of the urine stream), blood clots, normal urinary RBC morphology, and minimal proteinuria on dipstick (<100 mg/dL).

6.(C) IgA nephropathy is the most common chronic glomerular disease in children. Normal serum levels of C3 in IgA nephropathy help to distinguish this disorder from postinfectious glomerulonephritis. Serum IgA levels have no diagnostic value because they are elevated in only 15% of pediatric patients.

7.(C) A combination of a thorough family history, a screening urinalysis of first-degree relatives, an audiogram, and an ophthalmologic examination are critical in making the diagnosis of Alport syndrome(AS). The presence of anterior lenticonus is pathognomonic.

8.(D) Acute post-streptococcal glomerulonephritis (APSGN) is a classic example of the acute nephritic syndrome characterized by the sudden onset of gross hematuria, edema, hypertension, and renal dysfunction. It is one of the most common glomerular causes of gross hematuria in children and is a major cause of morbidity in group A β-hemolytic streptococcal infection.

9.(D) Hypertension is seen in 60% of patients and is associated with hypertensive encephalopathy in 10% of cases. Although the neurologic sequelae are often reversible with appropriate management, severe prolonged hypertension can lead to intracranial bleeding.

10.(E) Systemic diseases causes of low Serum complement (C3, CH 50):

- ✓ Lupus nephritis (focal 75%)
- ✓ Subacute bacterial endocarditis (90%)
- ✓ Shunt nephritis (90%)
- ✓ Essential mixed cryoglobulinemia (85%)
- ✓ Visceral abscess

While renal diseases causes of low Serum complement (C3, CH 50):

- ✓ Acute post infectious glomerulonephritis (>90%)
- ✓ Membranoproliferative glomerulonephritis (50-80%)

11.(B) Glomerulonephritis with hematuria is a recognized complication of various chronic infections. Classic examples that are uncommonly encountered today include bacterial endocarditis caused by viridans group streptococci and other organisms, and ventriculoatrial shunts infected with Staphylococcus epidermidis.

12.(E) Patients with class V lupus nephritis commonly present with nephrotic syndrome.

13.(B) Membranoproliferative glomerulonephritis (MPGN), also known as mesangiocapillary glomerulonephritis, most commonly occurs in older children or young adults. MPGN can be classified into primary (idiopathic) and secondary forms of glomerular disease. Secondary forms of MPGN are most commonly associated with subacute and chronic infection, including hepatitis B and C, syphilis, subacute bacterial endocarditis, and infected shunts, especially ventriculoatrial shunts (shunt nephritis)

14.(A) Pneumonia and urinary tract infection are the most common post-transplant bacterial infections. Urinary tract infections can progress rapidly to

urosepsis and may be confused with episodes of acute rejection. Trimethoprim-sulfamethoxazole is used for urinary tract infection prophylaxis as well as Pneumocystis jirovecii pneumonia prophylaxis for 3-6 mo after transplant.

15.(C) Steroid-resistant or high-grade rejection can treated with thymoglobulin (1.5 mg/kg/day) for 7-14 days, high-dose tacrolimus (trough levels > 20 ng/dL for 1-2 wk), or local allograft irradiation.

16.(C) Immunoglobulin (Ig) A nephropathy, a common glomerulonephritis, only rarely is rapidly progressive.

17.(A) HUS secondary to systemic infections include the following

- ✓ Neuraminidase (Streptococcus pneumoniae)
- ✓ Human immunodeficiency virus
- ✓ Influenza
- ✓ Human herpes virus 6
- ✓ Parvovirus B19
- ✓ Malaria

18.(B) Red blood cell (RBC) destruction may occur in hemolytic anemias because of mechanical injury as the cells traverse a damaged vascular bed. Damage may be microvascular when RBCs are sheared by fibrin in the capillaries during intravascular coagulation or when renovascular disease accompanies the hemolytic-uremic syndrome (HUS).

19.(D) Thrombocytopenia is an invariable finding in the acute phase, with platelet counts usually 20,000-100,000/mm3. Partial thromboplastin and prothrombin times are usually normal. The Coombs test is negative, with the exception of pneumococci-induced HUS, where the Coombs test is usually positive. Leukocytosis is often present and significant. Urinalysis typically shows microscopic hematuria and low-grade proteinuria.

20.(C) Nephrotic syndrome has also developed during therapy with numerous drugs and chemicals. The histologic picture can resemble membranous glomerulopathy (penicillamine, captopril, gold, nonsteroidal antiinflammatory drugs, mercury compounds), minimal change nephrotic syndrom (probenecid, ethosuximide, methimazole, lithium), or proliferative glomerulonephritis (procainamide, chlorpropamide, phenytoin, trimethadione, paramethadione).

21.(C) The development of renal vein thrombosis is classically heralded by the sudden onset of gross hematuria and unilateral or bilateral flank masses. However, patients can also present with any combination of microscopic hematuria, flank pain, hypertension, or a microangiopathic hemolytic anemia with thrombocytopenia or oliguria.

22.(E) Mitral valve prolapse is seen in approximately 12% of children; aortic and coronary artery aneurysms and aortic valve insufficiency are noted in affected adults.

23.(C) Associated external findings include retinal degeneration (Senior-Loken syndrome), cerebellar ataxia (Joubert syndrome), and hepatic fibrosis (Boichis disease).

24.(B) Hemorrhagic cystitis is defined as the presence of sustained hematuria and lower urinary tract symptoms as dysuria, frequency, and urgency. Hemorrhagic cystitis can occur in response to viruses (adenovirus types 11 and 21 and influenza A). The polyoma BK virus present latently in immunocompetent hosts is associated with the development of drug induced cystitis in immunosuppressed patients.

25.(E) Mammalian Target of Rapamycin (mTOR) Inhibitors (sirolimus more commonly than everolimus) are used primarily as adjunctive immune-suppression in combination with MMF in order to avoid tacrolimus toxicity or with tacrolimus and MMF to spare steroids.

26.(A) Microalbuminuria is defined as the presence of albumin in the urine above the normal level but below the detectable range of conventional urine dipstick methods. Microalbuminuria in children has been found to be associated with obesity and to predict, with reasonable specificity, the development of diabetic nephropathy in type 1 diabetes mellitus.

27.(B) Total protein concentration in urine can be estimated with chemically impregnated plastic strips that contain a pH-sensitive colorimetric indicator that changes color when negatively charged proteins, such as albumin, bind to it. False-negative test results can occur in patients with a low urine pH (<4.5), dilute urine or a large volume of urine output, or in disease states in which the predominant urinary protein is not albumin.

28.(A)

29.(D) Spontaneous bacterial peritonitis presents with fever, abdominal pain, and peritoneal signs. Although Pneumococcus is the most frequent cause of peritonitis, Gram-negative bacteria also are associated with a significant number of cases. Children with nephrotic syndrome and fever or other signs of infection must be evaluated aggressively, with appropriate cultures drawn, and should be treated promptly and empirically with antibiotics. Peritoneal leukocyte counts > 250 cells/µL are highly suggestive of spontaneous bacterial peritonitis.

30.(A) Lesions consistent with FSGS may be seen secondary to HIV infection, vesicoureteral reflux, and intravenous use of heroin and other drugs of abuse.

Only 20% of patients with FSGS respond to prednisone. The disease is often progressive, ultimately involving all glomeruli with end-stage renal disease in most patients.

31.(B) Nephrotic syndrome is a hypercoagulable state resulting from multiple factors: vascular stasis from hemoconcentration and intravascular volume depletion, increased platelet number and aggregability, and changes in coagulation factor levels. There is an increase in hepatic production of fibrinogen along with urinary losses of antithrombotic factors such as antithrombin III and protein S.

32.(D) Growth may be affected in children who require long-term corticosteroid therapy. Steroid-sparing strategies may improve linear growth in children who require prolonged courses of steroids.

33.(C) The potential side effects of the drug (neutropenia, disseminated varicella, hemorrhagic cystitis, alopecia, sterility, increased risk of future malignancy) should be carefully reviewed with the family before initiating treatment. Attention to adequate hydration can attenuate the risk of hemorrhagic cystitis.

34.(D)

35.(E)

36.(A) Calcineurin inhibitors (cyclosporine or tacrolimus) are recommended as initial therapy for children with steroid-resistant nephrotic syndrome.

37.(A) In many patients with type II MPGN, C3 nephritic factor (anti–C3 convertase antibody) is present. This factor activates the alternative complement pathway. In unusual cases, patients with type II MPGN demonstrate an associated systemic disease called partial lipodystrophy, where there is diffuse loss of adipose tissue and decreased complement in the presence of C3 nephritic factor.

38.(C) Renal tubular acidosis (RTA) is a disease state characterized by a normal anion gap (hyperchloremic) metabolic acidosis in the setting of normal or near-normal glomerular filtration rate. Hypercalciuria is usually present in distal RTA and can lead to nephrocalcinosis or nephrolithiasis. Chronic metabolic acidosis also impairs urinary citrate excretion. Hypocitraturia further increases the risk of calcium deposition in the tubules.

39.(B) Lowe syndrome (oculocerebrorenal syndrome of Lowe) is a rare X-linked disorder characterized by congenital cataracts, mental retardation, and Fanconi syndrome.

40.(B) The diagnosis of cystinosis is suggested by the detection of cystine crystals in the cornea and confirmed by measurement of increased leukocyte cysteine content. Prenatal testing is available for at-risk families.

41.(B) True hyperkalemic acidosis is consistent with type IV RTA, whereas the finding of normal or low potassium suggests type I or II.

42.(A) Once the presence of a non–anion gap metabolic acidosis is confirmed, the urine pH can help distinguish distal from proximal causes. A urine pH < 5.5 in the presence of acidosis suggests proximal RTA, whereas patients with distal RTA typically have a urine pH > 6.

43.(A) Patients with distal RTA should be monitored for the development of hypercalciuria. Those with symptomatic hypercalciuria (recurrent episodes of gross hematuria), nephrocalcinosis, or nephrolithiasis can require thiazide diuretics to decrease urine calcium excretion.

44.(A) Traumatic blood draws (such as heel stick specimens), small volumes of blood in adult-size specimen collection tubes, or a prolonged specimen transport time at room temperature can lead to falsely low bicarbonate levels, often in association with an elevated serum potassium value.

45.(C) The diagnosis is suggested in a male infant with polyuria, hypernatremia, and dilute urine. Simultaneous serum and urine osmolality measurements should be obtained. If the serum osmolality value is 290 mOsm/kg or higher with a simultaneous urine osmolality value of < 290 mOsm/kg, a formal water deprivation test is not necessary. Because the differential diagnosis includes causes of central diabetes insipidus, the inability to respond to ADH (and thus the presence of NDI) should then be confirmed by the administration of vasopressin (10-20 µg intranasally) followed by serial urine and serum osmolality measurements hourly for 4 hr.

46.(A) Consanguinity suggests the presence of an autosomal recessive disorder. Older children can have a history of recurrent episodes of polyuria with dehydration, failure to thrive, nonspecific fatigue, dizziness, and chronic constipation. Older children may also present with muscle cramps and weakness secondary to chronic hypokalemia. The blood pressure is usually normal. Serum chemistry reveals the classic biochemical abnormalities of a hypokalemic hypochloremic metabolic alkalosis. The renal function is typically normal.

47.(D) Although some patients develop nephrocalcinosis, nephrolithiasis, progressive renal failure, and hypophosphatemic rickets, patients with Dent disease typically do not have proximal renal tubular acidosis or extrarenal manifestations.

48.(D)

49.(A) Liddle syndrome is an autosomal dominant disorder that results from an activating mutation of the distal nephron sodium channel that is normally upregulated by aldosterone. Patients have the characteristics of hyperaldosteronism—hypertension, hypokalemia, and alkalosis—but low serum renin and aldosterone levels. These patients respond to the potassium-sparing diuretics (triamterene and amiloride) that inhibit this sodium channel.

50.(B) This According to KDIGO staging of acute kidney injury(AKI). AKI can be defined as an abrupt (1 to 7 days) and sustained (more than 24 hours) decrease in kidney function.

51.(E) The sensitivity and specificity of urine sodium of < 20 mEq/L in differentiating prerenal azotemia from acute tubular necrosis are 90% and 82%, respectively.

52.(B) A case of acute lower limb compartment syndrome; his paraesthesia, elevated blood lactate, elevated creatine kinase, acidosis, and myoglobinaemia support this diagnosis. Reduced estimated glomerular filtration rate and elevated serum creatinine and urea level suggest that acute kidney injury due to rhabdomyolysis.

53.(B)

54.(C) The presence of white blood cells and white blood cell casts with low-grade hematuria and proteinuria suggests tubule-interstitial disease. Urinary eosinophils may be present in some children with drug-induced tubule-interstitial nephritis.

55.(A)

56.(B) Childhood-onset ESRD still carries significant morbidity and a 30-fold increased mortality rate as compared with healthy peers, with cardiovascular and infectious diseases as the leading causes of death.

57.(A) Dietary protein restriction is not suggested for children with CKD because of the concern about adverse effects on growth and development; in fact, the recommended protein intake is often 100% (or more for those receiving dialysis) of the dietary reference intake for ideal weight for children.

58.(B)

59.(B) Ambulatory blood pressure monitoring (ABPM) over 24 hr, the gold standard of blood pressure evaluation, is recommended in patients with renal disease to diagnose and treat hypertension, especially masked hypertension.

60.(C) CKD patients of all ages should typically follow a low-phosphorus diet with the goal of maintaining age-appropriate serum phosphorus values. Infants should be provided with a low-phosphorus formula (Similac PM 60/40).

Phosphate binders (given with meals) are used to enhance GI phosphate excretion, and at present are recommended to be started at the onset of hyperphosphatemia.

61.(E) Metabolic acidosis develops because of a decreased net acid excretion by the failing kidneys. Either Bicitra (1 mEq sodium citrate/mL) or sodium bicarbonate tablets (650 mg = 8 mEq of base) may be used to maintain the serum bicarbonate level ≥ 22 mEq/L.

62.(D) Patients who appear to be resistant to ESA should be evaluated for iron deficiency, occult blood loss, a chronic infection or inflammatory state, vitamin B12 or folate deficiency, or bone marrow fibrosis related to secondary hyperparathyroidism.

63.(A) ACE inhibitors (e.g., enalapril or lisinopril) and angiotensin II receptor blockers (ARB; e.g., losartan) are the antihypertensive medications of choice in all children with pediatric CKD, irrespective of the level of proteinuric renal disease, because of their potential ability to slow CKD and their superiority in controlling blood pressure as noted in various observational and research studies.

64.(E) Non modifiable risk factors associated with more rapid CKD progression include older age, glomerular etiology of renal disease, CKD severity, and onset of puberty. In terms of potential modifiable risk factors (in addition to an elevated blood pressure), persistent nephrotic range proteinuria, anemia, and dyslipidemia, as well as no ACE/ARB use, were important predictors of CKD progression.

65.(D) Peritoneal dialysis utilizes the patient's peritoneal membrane to transport fluid and solutes. Excess body water is removed by an osmotic gradient created by the relatively high dextrose concentration in the dialysis fluid; wastes are removed by diffusion from the peritoneal capillaries into the dialysis fluid.

66.(B) It is recommended that all immunizations be current prior to transplantation. All live vaccines (measles-mumps-rubella and varicella) should be given prior to transplantation, and antibody titers should be checked for a response because these vaccines should not be given to immunosuppressed patients. Measles-mumps-rubella may be given as early as 6 mo of age. Inhaled (live-attenuated virus) influenza vaccine should not be given to transplant patients, family members, or healthcare providers.

67.(A) Tacrolimus also appears to cause less dyslipidemia, though other side effects such as new-onset diabetes after transplant (NODAT), tremor, seizure,

alopecia, and sleep disturbance seem to be more common in patients treated with tacrolimus than cyclosporine.

CHAPTER 23

Urologic Disorders Questions

QAHTAN ALOBAIDY

1. The parents of a 10-year-old boy with a single kidney ask you if he can participate in contact sports.
Which of the following arguments is against such participation?

A. Infection
B. Renal injury
C. Hypertension
D. Vascular injury
E. Reflux nephropathy

2. What is the mode of inheritance in multicystic dysplastic kidney (MCDK)?

A. Autosomal recessive
B. Autosomal dominant
C. X-linked recessive
D. X-linked dominant
E. Not inherited

3. Which of the following renal cystic disorders needs surgical removal?

A. Multicystic kidney
B. Benign multilocular cyst
C. Simple cyst
D. Medullary sponge kidney
E. Calyceal diverticulum

4. Which of the following systems must be checked thoroughly in a newborn with single umbilical artery?

A. Renal
B. Central nervous
C. Respiratory
D. Gastrointestinal
E. Endocrine

5. In the delivery room, the doctor consults you about a baby born with external ear anomalies, imperforate anus, and scoliosis. Your advice is to exclude renal anomalies.
Which of the following investigations should be done in this newborn?

A. Renal function test
B. Renal sonogram
C. General urine exam
D. Abdominal CT scan
E. Intravenous pyelography

6. What is the **MOST** common microorganism causing urinary tract infections?

A. Enterococcus
B. Pseudomonas
C. Group B streptococcus
D. Escherichia coli
E. Staphylococcus saprophyticus

7. Which of the following is a second line treatment of oxalate renal stones?

A. Ferrous sulphate
B. Thiazide diuretics
C. Lithium
D. Pyridoxine
E. Allopurinol

8. Which of the following may be the only manifestation of pyelonephritis?

A. Flank pain
B. Fever
C. Malaise
D. Rigor
E. Vomiting

9. A 7-year-old boy presents with renal mass, he had complicated pyelonephritis secondary to renal calculi. Renal histopathology shows foamy histiocytes, you consult a urological surgeon who recommends total nephrectomy.
Of the following, the **MOST** likely diagnosis is

A. neuroblastoma
B. Wilms tumor
C. acute lobar nephronia

D. acute lobar nephritis
E. xanthogranulomatous pyelonephritis

10. Which of the following stones is treated with D-penicillamine?
A. Calcium oxalate
B. Indinavir
C. Cystine
D. Struvite
E. Uric acid

11. The nitrites test is of little value in the diagnosis of urinary tract infections caused by which of the following microorganism?
A. E coli
B. Proteus
C. Klebsiella
D. Enterococcus
E. Pseudomonas

12.Which of the following is a recognized cause of sterile pyuria?
A. Renal transplant
B. Ulcerative colitis
C. Kawasaki disease
D. Refrigerated urine
E. Fungal infection

13. Which of the following antibiotics does not achieve significant renal tissue level in the treatment of pyelonephritis?
A. Cefepime
B. Ciprofloxacin
C. Cephalexin
D. Nitrofurantoin
E. Gentamicin

14.after initiation of antibiotic therapy for treatment of UTI, urine cultures are typically negative within
A. 6 hr
B. 12 hr
C. 18 hr

D. 24 hr
E. 30 hr

15. What is the mode of inheritance of primary vesicoureteral reflux?
A. Autosomal dominant
B. Autosomal recessive
C. X-linked dominant
D. X-linked recessive
E. Multifactorial

16. What is the primary treatment option in a patient with symptomatic renal stone size 2.2 cm?
A. Shock wave lithotripsy
B. Ureteroscopic removal
C. Percutaneous nephrolithotomy
D. Laparoscopic removal
E. Open surgery

17. Which of the following procedures has the lowest radiation risk for the diagnosis of vesicoureteral reflux?
A. Radionuclide cystogram
B. Contrast voiding cystourethrography
C. Indirect cystography
D. Renal MRI
E. Renal scintigraphy

18. During routine renal sonogram in the second trimester; antenatal hydronephrosis was detected. You explain to the family that the anterior-posterior diameter of the renal pelvis determines the degree of severity.
Which of the following diameter is regarded as severe hydronephrosis?
A. >4 mm
B. >6 mm
C. >8 mm
D. >10 mm
E. >12 mm

19. Which of the following is the **MOST** likely cause of urinary ascites in newborn?

A. Megaureters
B. Posterior urethral valve
C. Multicystic dysplastic kidney
D. Ureteropelvic junction obstruction
E. Ureterovesical junction obstruction

20. Which of the following is associated with patent draining urachus?
A. Urethral obstruction
B. Multicystic dysplastic kidney
C. Ureterovesical junction obstruction
D. Ureteropelvic junction obstruction
E. Megaureter

21. A mother was concerned about fetal ultrasound which reveals dilation of the renal pelvis. Your advice is to repeat sonographic study after birth to confirm the prenatal findings.
Which of the following is the ideal time to perform postnatal ultrasound?
A. After first day
B. After third day
C. After first week
D. After 2 weeks
E. After 1 month

22. A neonate had deficient abdominal muscles, undescended testes, and urinary tract abnormalities. His parents worried about the prognosis of their baby.
The prognosis ultimately depends on severity of
A. pulmonary hypoplasia
B. cardiac abnormalities
C. bowel malrotation
D. renal dysplasia
E. urological abnormalities

23. What is the **MOST** common cause of severe obstructive uropathy in children?
A. Urethral atresia
B. Urethral hypoplasia
C. Posterior urethral valves

D. Ureteropelvic junction obstruction
E. Primary obstructed nonrefluxing megaureter

24. What is the percentage of occurrence of vesicouretral reflux in posterior urethral valve disease?
A. 10%
B. 30%
C. 50%
D. 70%
E. 90%

25. Which of the following syndromes is associated with bladder diverticula?
A. Marfan syndrome
B. Fragile X syndrome
C. Prader-willi syndrome
D. Ehlers-Danlos syndrome
E. Rett syndrome

26. A 6-year-old boy presents with suprapubic pain, fever, irritative voiding symptoms, and an erythematous infraumbilical mass.
Which of the following is the **MOST** likely diagnosis?
A. Patent urachus
B. Urachal cyst
C. Vesicourachal diverticulum
D. Umbilical–urachal sinus
E. Cloacal exstrophy

27. Which of the following surgical complications develops in augmented cystoplasty in response to mucus accumulation?
A. Urinary tract infection
B. Metabolic acidosis
C. Spontaneous perforation
D. Bladder calculi
E. Malignant neoplasm

28. Which of the following is the **MOST** appropriate initial test to exclude sacral agenesis in a patient with imperforate anus?
A. Spinal ultrasound

B. Sacral X-ray
C. MRI
D. C-T scan
E. Myelogram

29. What is the **MOST** common cause of daytime incontinence?
A. overactive bladder
B. voiding postponement
C. detrusor–sphincter discoordination
D. cystitis
E. overflow incontinence

30. A 6-year-old boy exhibit a staccato stream, day and night wetting, recurrent UTIs, constipation, and encopresis.
Of the following, the **MOST** likely diagnosis is
A. diurnal urge syndrome
B. nonneurogenic neurogenic bladder
C. posterior urethral valves
D. epispadias
E. ectopic ureter

31. Medical student ask you about the calculation of the bladder capacity in milliliters in a 4-year-old boy.
Which of the following equation can be used for this purpose?
A. age + 1 (in years) X 30
B. age + 2 (in years) X 30
C. age + 3 (in years) X 30
D. age + 4 (in years) X 30
E. age + 5 (in years) X 30

32. The ideal age at which active treatment for nocturnal enuresis can be started is
A. 4 years
B. 5 years
C. 6 years
D. 7 years
E. 8 years

33. Desmopressin acetate is a synthetic analog of antidiuretic hormone that reduces urine production overnight. This medication is FDA-approved in children and is preferable to be used as

A. tablet
B. IM injection
C. IV injection
D. nasal spray
E. skin patch

34. Which of the following types of renal stones are radiolucent on plain abdominal radiograph?

A. Calcium oxalate
B. Calcium phosphate
C. Cystine
D. Struvite
E. Uric acid

35. What is the **MOST** common cause of priapism in children?

A. Sickle cell disease
B. Sildenafil ingestion
C. Leukemia
D. Perineal trauma
E. Idiopathic

36. Which of the following syndromes causes micropenis due to hypergonadotropic hypogonadism?

A. Robinow syndrome
B. Kallmann syndrome
C. Prader-Willi syndrome
D. Bardet–Biedl syndrome
E. Lawrence-Moon-Bardet-Biedl syndrome

37. What is the maximum age the testes can be descended spontaneously in a baby with cryptorchidism?

A. 2 months
B. 4 months
C. 6 months
D. 9 months

E. 12 months

38. A 4-year-old boy with cryptorchidism presents with inguinal pain and swelling.
Which of the following is the **MOST** likely diagnosis?

A. Seminoma
B. Femoral hernia
C. Testicular torsion
D. Germ cell malignancy
E. Varicocele

39. A 10-year-old girl presents with loin pain and hematuria. Urine dipstick demonstrates blood ++++, nitrates +ve, leukocyte +++, and protein ++, urine culture shows Proteus infection. KUB demonstrates a stag-horn calculus in the left renal pelvis.
What is the **MOST** likely composition of this renal stone?

A. Calcium oxalate
B. Xanthine
C. Cystine
D. Struvite
E. Urate

40. Which of the following is the only complication cystinuria?

A. Renal tubular acidosis
B. Chronic renal failure
C. Renal calculi
D. Nephrocalcinosis
E. Recurrent UTI

41. Which of the following is the **MOST** common organism in a 9-year-old boy with scrotal pain, erythema, and swelling?

A. Escherichia coli
B. Chlamydia
C. Gonococcus
D. Enterovirus
E. Adenovirus

42. Which of the following can differentiate testicular torsion from an incarcerated hernia?

A. Tender testis
B. Swollen scrotum
C. Absent cremasteric reflex
D. Absent swelling in the inguinal area
E. Associated nausea and vomiting

43. Which of the following anatomical sites of renal stones is asymptomatic?

A. Ureteropelvic junction
B. Mid-ureter
C. Ureterovesical junction
D. Urinary bladder
E. Urethra

44. What is the **MOST** appropriate imaging modality in suspected renal colic?

A. KUB
B. U/S
C. spiral CT scan
D. MRI
E. IV pyelography

CHAPTER 23

Urologic Disorders Answers

QAHTAN ALOBAIDY

1.(B) The arguments against such participation are that the contralateral normal kidney is hypertrophic and not as well protected by the ribs, and a serious renal injury could have serious lifelong consequences.

2.(E) MCDK is a congenital condition in which the kidney is replaced by cysts and does not function; it can result from ureteral atresia. Kidney size is highly variable. The incidence is approximately 1 in 2,000. MCDK usually is unilateral and generally is not inherited.

3.(B) A multilocular cyst (multilocular cystic nephroma) is a lesion in the kidney that falls in a spectrum of diseases, along with multilocular cyst with partially differentiated Wilms tumor, multilocular cyst with nodules of Wilms tumor, or cystic Wilms tumor. The multilocular cyst is considered benign and is unrelated to the multicystic dysplastic kidney. More than 95% occur in children < 4 yr, and most are discovered during evaluation for an abdominal or flank mass. The lesion should be removed.

4.(A) Upper urinary tract anomalies are more common in children with certain physical findings. The incidence of renal anomalies is increased if there is a single umbilical artery and an abnormality of another organ system (congenital heart disease).

5.(B) External ear anomalies (particularly if the child has multiple congenital anomalies), imperforate anus, and scoliosis are associated with renal anomalies. Infants with these physical findings should undergo a renal sonogram.

6.(D) Urinary tract infections are caused primarily by colonic bacteria. Escherichia coli cause 54–67% of all UTIs, followed by Klebsiella spp. and Proteus spp., Enterococcus, and Pseudomonas. Other bacteria known to cause UTIs include Staphylococcus saprophyticus, group B streptococcus, and, less commonly, Staphylococcus aureus, Candida spp., and Salmonella spp.

7.(D)

8.(B) Fever may be the only manifestation; particular consideration should occur for a temperature > 39C without another source lasting more than 24 hr for males and more than 48 hr for females.

9.(E) Xanthogranulomatous pyelonephritis is a rare type of renal infection characterized by granulomatous inflammation with giant cells and foamy histiocytes. It can manifest clinically as a renal mass or an acute or chronic infection. Renal calculi, obstruction, and infection with Proteus spp. or E. coli contribute to the development of this lesion, which usually requires total or partial nephrectomy.

10.(C) D-penicillamine is a chelating agent that binds to cysteine or homocysteine, increasing the solubility of the product. Although poorly tolerated by many patients, it has been reported to be effective in dissolving cystine stones and in preventing recurrences when hydration and urinary alkalinization fail.

11.(D) Nitrites and leukocyte esterase are often positive in infected urine. Bacteria generally require 4 hr for metabolism of nitrates to nitrites. Nitrites may not be detected in cases of UTI if the organism does not convert nitrates to nitrites (most notably enterococcus) or if the child has urinary frequency, where there may not be enough time for the conversion to nitrites. In febrile infants less than 60 days old, the presence of pyuria, nitrites, or leukocyte esterase has a high sensitivity and specificity for a UTI.

12.(C) Sterile pyuria (positive leukocytes, negative culture) may occur in partially treated bacterial UTIs, viral infections, urolithiasis, renal tuberculosis, renal abscess, UTI in the presence of urinary obstruction, urethritis as a consequence of a sexually transmitted infection, inflammation near the ureter or bladder (appendicitis, Crohn disease), Kawasaki disease, schistosomiasis, neoplasm, renal transplant rejection, or interstitial nephritis (eosinophils).

13.(D) Nitrofurantoin should not be used routinely in children with a febrile UTI, because it does not achieve significant renal tissue levels.

14.(D) The rate of renal scarring increases between days 2 and 3 of fever; this makes the prompt evaluation and appropriate treatment of a recurrent UTI important. Urine cultures are typically negative within 24 hr of initiation of antibiotic therapy, and therefore a urine culture during treatment is almost invariably negative.

15.(A) Primary VUR appears to be an autosomal dominant inherited trait with variable penetrance.

16.(C)

17.(A) Radiation exposure during a radionuclide cystogram is significantly less than that from a contrast VCUG.

18.(D)

19.(B)

20.(A)

21.(B) Because neonatal oliguria can cause temporary decompression of a dilated renal pelvis, it is ideal to perform the first postnatal US after the 3rd day of life. Delaying the US may be impractical.

22.(A) The prognosis ultimately depends on the degree of pulmonary hypoplasia and renal dysplasia. One third of children with prune-belly syndrome are stillborn or die in the first few mo of life because of pulmonary hypoplasia.

23.(C) The most common cause of severe obstructive uropathy in children is PUVs, affecting 1 in 8,000 males. While the most severe form of obstructive uropathy in males is urethral atresia, a rare condition.

24.(C)

25.(D) They occur commonly in children with connective tissue disorders, including Williams syndrome, Ehlers-Danlos syndrome, and Menkes syndrome.

26.(B) A patent urachus results in continuous urinary drainage from the umbilicus. Vesicourachal diverticulum, which is a diverticulum of the bladder dome, and umbilical–urachal sinus, which is a blind external sinus that opens at the umbilicus. These lesions should be excised.

27.(D) Bladder calculi have developed in as many as 70% of children followed for 10 yr after enterocystoplasty. The calculi develop in response to mucus that accumulates in the bladder and act as a nidus for stone formation. This complication can be prevented by daily irrigation of the bladder with sterile saline.

28.(A) Approximately 30–45% of children with a high imperforate anus have a neuropathic bladder, often because of sacral agenesis. Newborns with imperforate anus should undergo a spinal ultrasound during their initial evaluation.

29.(A) The most common causes of daytime incontinence are overactive bladder (urge incontinence) and bladder–bowel dysfunction.

30.(B) Hinman syndrome is a very serious but uncommon disorder involving failure of the external sphincter to relax during voiding in children without neurologic abnormalities. Children with this syndrome also called nonneurogenic neurogenic bladder, typically exhibit a staccato stream, day and night wetting, recurrent UTIs, constipation, and encopresis.

31.(B) In children up to the age of 14 yr, the mean bladder capacity in milliliters is equal to the age + 2 (in years) times 30 (e.g., the bladder capacity of a 6 yr old should be 240 mL).

32.(C) Reassure the child and parents that the condition is self-limited and to avoid punitive measures that can affect the child's psychological development

adversely. Active treatment should be avoided in children younger than 6 yr of age, because enuresis is extremely common in younger children.

33.(A) In the past a nasal spray was used, but some children experienced hyponatremia and convulsions with this formulation, and the nasal spray is no longer recommended for nocturnal enuresis. Hyponatremia has not been reported in children using the oral tablets.

34.(E) These stones are radiolucent on x-ray. The diagnosis should be suspected in a patient with persistently acid urine and urate crystalluria.

35.(A) The most common cause of priapism in children is sickle cell disease, which is characterized by a predominance of sickle cell hemoglobin. As many as 27.5% of children with sickle cell disease develop priapism. The priapism is generally related to a low-flow state, secondary to sickling of red blood cells within the sinusoids of the corpora cavernosa during normal erection, resulting in venous stasis.

36.(A) Primary testicular failure can result from gonadal dysgenesis or rudimentary testes syndrome and also occurs in Robinow syndrome (characterized by hypoplastic genitalia, shortening of the forearms, frontal bossing, hypertelorism, wide palpebral fissures, short broad nose, long philtrum, small chin, brachydactyly, and a normal karyotype).

37.(B) Spontaneous descent occurs secondary to a temporary testosterone surge (termed a minipuberty) during the first 2 mo, which also results in significant penile growth. If the testis has not descended by 4 mo, it will remain undescended.

38.(C) Inguinal pain and/or swelling in a male with an undescended testis should raise the suspicion of an inguinal hernia or torsion of the undescended testis.

39.(D) Urinary tract infections caused by urea-splitting organisms (most often Proteus spp., and occasionally Klebsiella spp., Escherichia coli, Pseudomonas spp., and others) result in urinary alkalinization and excessive production of ammonia, which can lead to the precipitation of magnesium ammonium phosphate (struvite) and calcium phosphate. In the kidney, the calculi often have a staghorn configuration, filling the calyces. The calculi act as foreign bodies, causing obstruction, perpetuating infection, and causing gradual kidney damage.

40.(C) Cystinuria is a rare autosomal recessive disorder of the epithelial cells of the renal tubule that prevents absorption of the four dibasic amino acids (cystine, ornithine, arginine, lysine) and results in excessive urinary excretion of these products. The only known complication of this familial disease is the

formation of calculi, because of the low solubility of cystine. The patients usually have acidic urine, which leads to a higher rate of precipitation.

41.(A) In younger males, the responsible organismis often *Escherichia coli*. After puberty, bacterial epididymitis becomes progressively more common and can cause acute painful scrotal swelling in young sexually active males. Urinalysis usually reveals pyuria. Epididymitis can be infectious (usually gonococcus or *Chlamydia*), but often the organism remains undetermined. Additional etiologies include familial Mediterranean fever, enterovirus, and adenoviruses.

42.(D) Testicular torsion produces acute pain and swelling of the scrotum. On examination, the scrotum is swollen, and the testis is exquisitely tender and often difficult to examine. The cremasteric reflex nearly always is absent. The position (lie) of the testis is abnormal, and the testis position often is high in the scrotum. In addition, often there is associated nausea and vomiting. The condition can be differentiated from an incarcerated hernia because swelling in the inguinal area typically is absent with torsion.

43.(D) Children with urolithiasis usually have gross or microscopic hematuria, severe flank pain and can have irritative symptoms of dysuria, urgency, and frequency. If the calculus is in the distal ureter, the child can have irritative symptoms of dysuria, urgency, and frequency. If the stone passes into the bladder, the child usually becomes asymptomatic. If the stone is in the urethra, dysuria and difficulty voiding can result, particularly in males.

44.(C) In a child with suspected renal colic, there are multiple imaging options. The most accurate study is an unenhanced spiral CT scan of the abdomen and pelvis. This study takes only a few minutes to perform, has 96% sensitivity and specificity in delineating the number and location of calculi, and demonstrates whether the involved kidney is hydronephrotic.

CHAPTER 24

Gynecologic Problems of Childhood Questions

QAHTAN ALOBAIDY

1. Neonatal vaginal bleeding requires further evaluation if it persists beyond
   A. 5 days
   B. 10 days
   C. 20 days
   D. 30 days
   E. 45 days

2. Which of the following types of hymens associated with urinary obstruction?
   A. Crescentic
   B. Redundant
   C. Microperforate
   D. Septated
   E. Imperforate

3. Which of the following clitoral width in a newborn need further evaluation?
   A. >2 mm
   B. >3 mm
   C. >4 mm
   D. >5 mm
   E. >6 mm

4. What is the **MOST** common cause of ambiguous genitals in the neonatal period?
   A. Congenital adrenal hyperplasia
   B. Complete androgen insensitivity
   C. Male pseudohermaphrodite
   D. True hermaphrodite
   E. Idiopathic

5. A 15-year-old girl presented with external genital pruritus and skin lesion. Examination reveals skin-colored papules, some with a shaggy, cauliflower-like appearance.
Of the following, the **MOST** likely diagnosis is

A. molluscum contagiosum
B. condyloma acuminata
C. herpes simplex
D. lichen sclerosus
E. contact dermatitis

6. Which of the following antibiotics is recommended as first line therapy in vulvovaginal Infections caused by Haemophilus influenza?

A. Cotrimoxazole
B. Clarithromycin
C. Azithromycin
D. Amoxicillin
E. Erythromycin ethyl succinate

7. Which of the following antibiotics is recommended as first line therapy in vulvovaginal Infections caused by Yersinia?

A. Cotrimoxazole
B. Clarithromycin
C. Penicillin V
D. Amoxicillin
E. Erythromycin ethyl succinate

8. A 3-month-old girl presents with napkin rash treated by increasing the frequency of diaper changes, allowing her to be diaper free, frequent bathing, and application of zinc oxide but without significant benefit.
Which of the following can decrease the inflammation of his diaper dermatitis when applied topically?

A. Antifungal
B. Steroid
C. Coconut oil
D. Gention violet
E. Antibiotic

9. Which of the following is **FIRST-LINE** therapy in patients with labial adhesions and difficulty voiding?

A. Topical estrogen
B. Surgical correction
C. Topical antibiotic
D. Systemic antibiotic
E. Systemic anticholinergic

10. Which of the following syndromes is the **MOST** common cause of vaginal agenesis?

A. Fraser
B. McKusick-Kaufman
C. Johanson-Blizzard
D. Renal-genital-middle ear anomalies
E. Mayer-Rokitansky-Küster-Hauser

11. Which of the following dermatologic conditions affect the vulvar area in children and can lead to destruction and scarring of normal genital architecture?

A. Vulvar psoriasis
B. Lichen sclerosis
C. Idiopathic vulvar aphthoses
D. Contact dermatitis
E. Lichen planus

12. History is often more predictive than physical exam in the diagnosis of which of the following conditions?

A. Vaginal foreign body
B. Pinworms infestation
C. Labial agglutination
D. Molluscum contagiosum
E. Condyloma acuminata

13. A 10-year-old girl presents with blood-tinged and foul-smelling vaginal discharge fails to respond to hygiene measures.
Of the following, the **MOST** likely diagnosis is

A. vaginal foreign body
B. acute genital ulcer

C. herpes simplex infection
D. molluscum contagiosum
E. condyloma acuminata

14. Lack of development of the breast is considered delayed and warrants evaluation at which of the following ages?
A. 10 year
B. 11 year
C. 12 year
D. 13 year
E. 14 year

15. Which of the following is characterized by amastia and aplasia of the pectoralis muscles?
A. Ectodermal dysplasia
B. Congenital adrenal hyperplasia
C. Gonadal dysgenesis
D. Hypogonadotropic hypogonadism
E. Poland syndrome

16. Which of the following is the **MOST** common cause of breast pain in adolescent's girls?
A. Exercise
B. Pregnancy
C. Prolactinoma
D. Fibroadenoma
E. Polycystic ovary syndrome

17. Which of the following drugs can cause unilateral thelarche?
A. Diazepam
B. Cimetidine
C. Haloperidol
D. Cyproheptadine
E. Oral contraceptives

18. Which of the following is the **MOST** common solid mass seen in the breast of an adolescent girl?
A. Lipoma

B. Intraductal papilloma
C. fibroadenoma
D. Breast carcinoma
E. Intramammary lymph node

19. Which of the following is the **MOST** common ovarian neoplasm in children and adolescents?

A. Dysgerminoma
B. Cystadenomas
C. Gonadoblastoma
D. Gynandroblastoma
E. Mature cystic teratoma

20. A 10-year-old girl presents with acute lower abdominal pain, nausea, and vomiting. Examination revels abdominal tenderness. Pelvic ultrasound shows enlargement of left adnexa and demonstrate the presence of Doppler flow, free pelvic fluid, and "whirlpool sign". Of the following ,the **MOST** likely diagnosis is

A. tubo-ovarian abscess
B. endometriomas
C. adnexal torsion
D. ovarian cyst
E. uterine rhabdomyosarcoma

21. Which of the following heritable syndromes associated with müllerian anomalies is X-linked type of inheritance?

A. Camptobrachydactyly
B. Hand-foot-genital
C. Fraser
D. Uterine hernia
E. Renal-genital-middle ear anomalies

22. Which of the following is the **MOST** sensitive and specific imaging technique used for evaluating müllerian anomalies?

A. MRI
B. Ultrasound
C. Hysterosalpingogram
D. Sonohysterography
E. CT scan

CHAPTER 24

Gynecologic Problems of Childhood Answers

QAHTAN ALOBAIDY

1.(D) Estrogenic effects commonly notable in neonates include prominence of the labia majora and a white vaginal discharge. The labia minora and hymen may protrude slightly from the vestibule. A small amount of neonatal vaginal bleeding from endometrial sloughing following maternal hormone withdrawal might occur. Bleeding that is excessive or persistent beyond 1 mo of life requires further evaluation.

2.(E) Cervico-vaginal mucus secretions can accumulate behind the blocked outflow tract of an imperforate hymen and manifest as a mucocolpos. In this instance and if urinary obstruction occurs, correction of the imperforate hymen in the neonatal period is indicated.

3.(E) The clitoris may appear large in proportion to the other genital structures, especially in premature infants. If the clitoris appears enlarged, the clitoral width should be measured; values > 6 mm in a newborn indicate a need for further evaluation. If clitoromegaly and ambiguous genitals are present, the obstetrician and pediatrician should immediately obtain expert consultation for evaluation of the infant and to counsel the parents.

4.(A) Congenital adrenal hyperplasia is the most common cause of ambiguous genitals (accounting for > 90% of cases), and the salt-wasting forms can lead to rapid dehydration with subsequent fluid and electrolyte imbalance. Delay in the diagnosis and treatment of congenital adrenal hyperplasia may be life-threatening.

5.(B)

6.(D)

7.(A)

8.(A) First-line treatment includes hygiene measures such as increasing the frequency of diaper changes, allowing the infant to be diaper free, frequent bathing, and application of water-repellant barriers such as zinc oxide. If diaper dermatitis persists after these conservative measures, or if the classic satellite lesions of Candida are present, treatment with a topical antifungal can decrease the inflammation.

9.(A) First-line therapy in patients with difficulty voiding, persistent infections, or pain includes topical estrogen (Premarin or Estrace cream 0.01%) or a topical steroid (Betamethasone 0.05% ointment) applied twice daily to the midline raphe under gentle traction. Surgical correction is rarely necessary, but recurrence is common until the age of puberty.

10.(E) The most common cause of vaginal agenesis is MayerRokitansky-Küster-Hauser (MRKH) syndrome, with an incidence reported at 1 in 4,000-5,000 female births. After gonadal dysgenesis, müllerian agenesis is the second most common cause of primary amenorrhea. The etiology is believed to be multigenetic and multifactorial. This condition is present at birth but often not diagnosed until mid-adolescence.

11.(B) Lichen sclerosus is commonly seen in the anogenital region and has a characteristic appearance of white skin changes associated with areas of erosion, ulceration, and petechiae. This disease can cause severe discomfort and most commonly presents with vulvar or perianal pruritus, dysuria, and constipation.

12.(B) If pinworms are suspected, transparent adhesive tape or an anal swab should be applied to the anal region in the morning before defecation or bathing and then placed on a slide. Eggs seen on microscopic examination confirm the diagnosis, and sometimes the pinworms can be seen at the anal verge. Clinical history is often more indicative of disease than physical exam, and a negative tape test does not rule out this pathogen as a cause.

13.(A) Vaginal foreign bodies are a common finding in children presenting with blood-tinged and foul-smelling discharge. Quick identification and removal of the foreign body avoids potential complications, including recurrent urinary tract infections, dermatologic abnormalities, vaginal perforation, or fistula formation. The most common object found in the prepubertal vagina is retained toilet paper.

14.(D)

15.(E) Amastia is usually unilateral and can be congenital or associated with systemic disorders (e.g., ectodermal dysplasia), endocrine disorders (e.g., congenital adrenal hyperplasia, gonadal dysgenesis, hypogonadotropic hypogonadism), or novel gene mutations. It can be associated with anomalies of the underlying mesoderm, such as abnormal pectoralis muscles seen in Poland syndrome (aplasia of the pectoralis muscles, rib deformities, webbed fingers, and radial nerve aplasia).

16.(A) The most common causes of breast pain in adolescents are exercise and benign breast changes. Physiologic swelling and tenderness occur on a cyclic

basis, most commonly during the premenstrual phase, and are secondary to hormonal stimulation and resulting proliferative changes.

17.(B) Unilateral thelarche has also been reported as a side effect of cimetidine and is reversible when the medication is stopped.

18.(C) The most common solid mass seen in adolescent girls is the fibroadenoma. Fibroadenomas are most often located in the upper outer quadrant of the breast. The average size is 2-3 cm, and 10–25% of patients have multiple lesions. The physical examination is usually diagnostic because these lesions are well circumscribed, rubbery, mobile, and not tender. In equivocal cases, an ultrasound may be helpful in making the diagnosis.

19.(E) The most common ovarian neoplasm in children and adolescents is the mature cystic teratoma (dermoid cyst). Most are benign and contain mature tissue of ectodermal (skin, hair, sebaceous glands, neuroectodermal tissue), mesodermal (muscle, bone, cartilage, fat, teeth), and/or endodermal (thyroid, salivary, respiratory, gastrointestinal) origin.

20.(C) Adnexal torsion of the ovary and/or fallopian tube is the fifth most common gynecologic emergency and occurs more often in children and adolescents than in adults. It can occur in individuals with normal adnexa but more often occurs in adnexa enlarged by cystic (follicular, tubal) changes or ovarian (teratoma, cystadenoma) neoplasms.

21.(D)

22.(A) MRI is considered standard of care and best suited for complex anomalies because of its noninvasive, high-quality capabilities. MRI is the most sensitive and specific imaging technique used for evaluating müllerian anomalies because it can image nearly all reproductive structures, blood flow, external contours, junctional zone resolution on T2-weighted images, and associated renal and other anomalies. MRI also has a high correlation with surgical findings because of its multiplanar capabilities and high spatial resolution.

Chapter 25

The Endocrine System Questions

HAYDER ALMUSAWI

1. What is the most sensitive test for primary thyroid dysfunction?
   A. Free T3
   B. T4
   C. TSH
   D. TRH
   E. Thyroglobulin

2. Thyroxine-Binding Globulin (TBG) level usually decrease in
   A. pregnancy
   B. administration of glucocorticoids
   C. newborn period
   D. hepatitis
   E. administration of estrogens

3. The result of screening program of a child is low level of total T4 and normal levels of free T4 and TSH.
What is the most likely diagnosis?
   A. Primary hypothyroidism
   B. Secondary hypothyroidism
   C. Tertiary hypothyroidism
   D. TBG deficiency
   E. Thyroditis

4. What is the dose of L-T4 for a 7-year old child with acquired hypothyroidism?
   A. 1-2 µg/kg
   B. 3-5 µg/kg
   C. 6-8 µg/kg
   D. 9-11 µg/kg
   E. 12-15 µg/kg

5. Children treated with amiodarone should have serial monitoring of
   A. renal function
   B. thyroid function
   C. liver function
   D. platelet count
   E. blood sugar

6. Which of the following is a characteristic finding in familial dysalbuminemic hyperthyroxinemia?
   A. Increased serum concentrations of T4
   B. Decreased serum concentrations of TSH
   C. Increased serum concentrations of free T4
   D. Increased serum concentrations of free T3
   E. Increased serum concentrations of TSH

7. What is the **most** common cause of permanent congenital hypothyroidism?
   A. Defects in thyroid hormone synthesis
   B. Defective Iodide transport
   C. Thyroid dysgenesis
   D. Defects of Iodine organification
   E. Defects of thyroglobulin synthesis

8. A 10-month-old boy found to have bilateral fallopian tubes and a rudimentary uterus during repair of herniorrhaphy. Biopsy of the gonads reveals normal testicular tissue. On examination, his phallus is normal in size and appearance, and his testes size is 2 mL for each.
What is the **MOST** likely sex chromosome pattern for this child?
   A. XY
   B. XX/XY
   C. XXY
   D. XO/XY
   E. XYY

9. Which of the following play a role in inhibiting GH release?
   A. Sleep
   B. Exercise
   C. Physical stress
   D. Trauma

E. Hyperglycemia

10. Which of the following inhibit release of CRH and ACTH?
A. Atrial natriuretic peptide
B. Arginine vasopressin
C. Oxytocin
D. Angiotensin II
E. Cholecystokinin

11. A 9-year-old boy has been hospitalized for 1 month following road traffic accident. He is in a full body cast and has not been able to ambulate. In the last 2 days, he becomes irritable, anorectic, with frequent urination.
Of the following, the **MOST** likely cause is
A. hyperglycemia
B. hyponatremia
C. hypercalcemia
D. hypokalemia
E. cytitis

12. Which of the following is an indication for rhGH treatment to promote linear growth?
A. Chronic renal failure after transplantation
B. Idiopathic short stature
C. Constitutional growth delay
D. Edward syndrome
E. Thalassemia

13. Which of the following is a reasonable Criterion for stopping GH treatment?
A. A decision by the patient that he or she is tall enough
B. A growth rate <3 inch/yr
C. A bone age >16 yr in girls
D. A bone age >18 yr in boys
E. Recurrent abdominal pain

14. Which of the following can be caused by recombinant IGF-1?
A. Hypertension
B. Hypocalcaemia
C. Hypoglycemia

D. Anemia
E. Acidosis

15. Which of the following results can establish the diagnosis of diabetis inspidis (DI)?

A. Serum potassium <3.5 mmol/ L and serum sodium <145 mmol/L
B. Serum potassium >4.5 mmol/L and serum sodium >135 mmol/L
C. Serum osmolality is <270 mOsm/kg and the urine osmolality is >600 mOsm/kg
D. Serum osmolality is >300 mOsm/kg and the urine osmolality is <300 mOsm/kg
E. Serum osmolality is >270 mOsm/kg and the urine osmolality is >330 mOsm/kg

16. The initial approach to a patient with hyponatremia begins with determination of

A. volume status
B. urine sodium
C. urine specific gravity
D. serum potassium
E. BUN

17. Which of the following causes of hyponatremia is associated with hypovolemia and high urine sodium?

A. Systemic dehydration
B. SIADH
C. Renal primary salt loss
D. Runner's hyponatremia
E. Factitious hyponatremia

18. Syndrome of inappropriate antidiuretic hormone secretion (SIADH) is characterized by

A. hypernatremia
B. concentrated urine (>100 mosm/kg)
C. decreased plasma volume
D. low urine sodium
E. high serum uric acid

19. What is the maximum amount of water a person with normal renal function can consume daily?

A. 4 L/m2
B. 6 L/m2
C. 8 L/m2
D. 10 L/m2
E. 12 L/m2

20. What is the **MOST** commonly diagnosed adenoma during childhood?

A. Prolactinoma
B. Corticotropinoma
C. Somatotropinoma
D. Thyrotropinoma
E. Gonadotropinoma

21. The nurse in NICU called you because of persistent hypoglycemia in a 3-day-old male neonate, on examination the neonate had eye proptosis with periorbital fullness, mid-glabellar capillary malformation , earlobe creases, macroglossia, hepatosplenomegaly, and omphalocele.
What is the most likely cause of hypoglycemia?

A. Poor feeding
B. Systemic infection
C. Adrenal insufficiency
D. Pancreatic β-cell hyperplasia
E. Fatty acid oxidation disorder

22. An 11-year-old boy presents with history of headache, mood disturbance, behavioral swings, difficulty with sleep, decrease in attention span, and a decline in school performance for the last 3 months , examination shows diffuse smooth goiter.
What is the **MOST** appropriate next step in his management?

A. TSH and T4 level
B. CT brain
C. ESR
D. CBC and blood film
E. Neck ultrasound

23. Cancer risk is high in patients with Beckwith-Wiedemann syndrome until 8 years of age, so they need regular surveillance with abdominal ultrasound and

A. α-fetoprotein assay
B. HCG assay
C. 17 hydroxyprogesteron assay
D. α-1-antitrypsin assay
E. serum uric acid

24. Which of the following is a diagnostic criteria for diabetes mellitus?

A. Fasting plasma glucose 100-125 mg/dL
B. 2 hr plasma glucose during OGTT ≥ 140 mg/dL
C. Hemoglobin A1C ≥ 6.5%
D. Symptoms of diabetes mellitus plus random plasma glucose ≥ 160 mg/dL
E. Low C peptide

25. The clearest evidence of a role for viral infection in human T1DM is seen in

A. congenital rubella syndrome
B. enteroviral infection
C. mumps infection
D. RSV infection
E. HSV infection

26. A 12-years-old girl presents to the ER with history of repeated vomiting and abdominal pain , in the preceded 4 days she suffered from frequent urination and excessive thirst.
What is the next step in her management?

A. Intravenous fluid
B. Analgesia
C. Ultrasound abdomen
D. Blood glucose
E. Surgical consultation

27. An 8-year-old boy presents with abdominal pain and rapid breathing, his blood sugar is 483 mg/dl.
What is the **MOST** appropriate next step in his management?

A. Subcutaneous insulin 1 unit/kg
B. 45% glucose saline 20 ml/kg/hr
C. Intravenous insulin 0.2 unit/kg/hr

D. 0.9% normal saline 10 ml/kg/hr
E. Intravenous analgesia

28. In treatment of patient with diabetic ketoacidosis and after receiving 0.9% normal saline, blood glucose 445 mg/dl, PH 7.3 venous, and CO2 18 meq/l. What is the **MOST** appropriate type of fluid to be used next?

A. 0.9% normal saline
B. 0.45% saline
C. 0.18% glucose saline
D. 5% glucose water
E. Ringer lactate

29. What is the type of intravenous fluid to be used when the blood glucose dropped to 220mg/dl while treating a patient with diabetic ketoacidosis?

A. 0.9% normal saline
B. 45% glucose saline
C. 18% glucose saline
D. 5% glucose water
E. Ringer lactate

30. Propylthiouracil is not recommended in children due to its potential to cause

A. renal failure
B. respiratory failure
C. agraniolocytosis
D. DIC
E. liver failure

31. In patients who might have transient congenital hypothyroidism, a trial of stopping L -T4 for 4 weeks may be undertaken after the age of

A. 6 months
B. 9 months
C. 18 months
D. 24 months
E. 36 months

32. What percent of children with type 1 diabetes mellitus may develop thyroid autoantibodies?

A. 5%
B. 10%
C. 20%
D. 40%
E. 80%

33. A 13-year-old boy presents with history of headache, mood disturbance, difficulties with sleep, decrease in attention span, and a decline in school performance for the last 3 months , examination shows small thyroid size, investigations reveals low TSH , high free T4 , and very low serum thyroglobulin with 123 I radioiodine uptake suppression.
Of the following, the **MOST** likely diagnosis is
A. Graves' disease
B. thyrotoxicosis factitia
C. Hashimoto's thyroiditis
D. myxedema
E. thyroid carcinoma

34. Which of the following indicate a high likelihood of GH deficiency?
A. Exotropia
B. Proptosis
C. Cleft lip
D. Micrognathia
E. Saddle nose

35. A 1-year- old child presents with severe short stature with lengths >4 SD below the mean, his birth length was 1 SD below the mean, his GH level is high while insulin-like growth factor (IGF) 1 level is low.
Of the following, the **MOST** likely diagnosis is
A. Laron syndrome
B. Prader Willi syndrome
C. Silver Russell syndrome
D. Noonan syndrome
E. SHOX gene variant

36. Which of the following is a physical sign of growth hormone deficiency in a 3-year-old boy?
A. Proportionate short stature

B. Normal height velocity
C. Weight for length decreased
D. Low-pitched voice
E. Large penis

37. Provocative tests that rapidly increase the level of GH in normal children include administration of
A. thyroxine
B. arginine
C. metoclopramide
D. somatostatin
E. histadine

38. Which of the following is an indication for rhGH treatment to promote linear growth?
A. Down syndrome
B. Digeorge syndrome
C. Noonan syndrome
D. Patue syndrome
E. Edward syndrome

39. Which of the following is a criterion for stopping GH treatment?
A. A growth rate <2 inch/yr
B. A bone age >12 yr in girls
C. A bone age >16 yr in boys
D. Sever bone pain
E. Uncontrolled hypertension

40. All patients diagnosed with GH deficiency should undergo periodic evaluation of
A. renal function
B. liver function
C. PT, PTT
D. adrenal function
E. lipid profile

41. Which of the following drugs may induce nephrogenic diabetes insipidus?
A. Cisplatin

B. Vincristine
C. Cyclosporine
D. Cyclophosphamide
E. Doxorubicin

42. Extracellular fluid tonicity is regulated by
A. sodium intake and excretion
B. potassium intake and excretion
C. water intake and excretion
D. calcium metabolism
E. environmental temperature

43. Which of the following epiphysis is present at birth in a full-term newborn?
A. Proximal femoral
B. Proximal tibial
C. Proximal ulnar
D. Proximal fibular
E. Proximal radial

44. Acquired nephrogenic diabetes insipidus can result from
A. hypocalcemia
B. hyperkalemia
C. rifampin
D. isoniazid
E. streptomycin

45. Hyponatremia with decrease intravascular volume and low urine sodium is usually seen in
A. systemic dehydration
B. SIADH
C. primary polydipsia
D. cerebral salt wasting
E. factitious hyponatremia

46. Syndrome of inappropriate antidiuretic hormone secretion is characterized by
A. hypernatremia
B. an inappropriately diluted urine

C. decrease plasma volume
D. low urine sodium
E. low serum uric acid

47. Which of the following overgrowth syndromes need tumor surveillance?
A. Marfan syndrome
B. Loeys-Dietz syndrome
C. Perlman syndrome
D. Homocystinuria
E. Lujan syndrome

48. What is the **FIRST** sign of puberty in female?
A. Breast bud
B. Pubic hair
C. Axillary hair
D. Menstrual activity
E. Peak height velocity

49. Peak height velocity occurs in females at breast stage
A. I-II
B. II–III
C. III-IV
D. IV-V
E. V

50. The mean age of menarche is about
A. 11.75 yr
B. 12.25 yr
C. 12.75 yr
D. 13.25 yr
E. 13.75 yr

51. Levothyroxine (L -T4) is given orally for the treatment of congenital hypothyroidism , the recommended initial dose is
A. 2-5 µg/kg/day
B. 5-10 µg/kg/day
C. 10-15 µg/kg/day
D. 15-25 µg/kg/day

E. 100-150 µg/kg/day

52. Levothyroxine (L -T4) tablets should be crushed and mixed with a small volume (1-2 mL) of liquid and should not be mixed with

A. breast milk
B. soy protein formula
C. cow milk
D. lactose free formula
E. hydrolyzed formula

53. DiGeorge/velocardiofacial syndrome has been reported in infants born to mothers treated early in pregnancy with

A. ACE inhibitors
B. retinoic acid
C. cocaine
D. thidomide
E. warfarin

54. A teenager male diagnosed recently with diabetes insipidus, he had history of T1DM and optic atrophy since early childhood.
Which of the following manifestations is seen in this patient?

A. Deafness
B. Eczema
C. Seizure
D. Anosmia
E. Vetiligo

55. A 10-year-old boy presented with distal radius fracture, he had history of previous fracture since 2 years. As part of his evaluation, height and weight are both at the 75th percentile and physical examination results are unremarkable ,serum calcium concentration 7.2 mg/dL, serum phosphorus concentration 2.6 mg/dL, serum magnesium concentration 1.9 mg/dL, and serum alkaline phosphatase 845 .
Of the following, the **MOST** appropriate next step is to measure

A. serum creatinine
B. serum 1,25-dihydroxyvitamin D
C. serum 25-hydroxyvitamin D
D. serum parathyroid hormone

E. urine calcium to creatinine ratio

56. A 12-year-old boy diagnosed with T1DM since 2 months, he established glucose control in the last 2 weeks.
What investigation is recommended now?

A. Fasting lipid profile
B. Protein c peptide
C. Hb A1c
D. Serum creatinin
E. Serum albumin

57. A 6-year-girl presents with history of polyuria, polydipsia, polyphagia and decrease body weight, her investigation reveal blood glucose 321 mg/dl, Hb A1c 8 and ketones in urine –ve.
What is the **NEXT** step in her management?

A. Reassurance and reevaluation after 2 weeks
B. Start metformin
C. Check for other autoimmune disease
D. Start intravenous normal saline
E. Start subcutaneous insulin

58. What is the starting subcutaneous insulin dose in newly diagnosed 4- year-old boy with T1DM?

A. 0.1-0.25 unit\kg\day
B. 0.25-0.5 unit\kg\day
C. 0.5-0.7 unit\kg\day
D. 0.7-1 unit\kg\day
E. 1-1.2 unit\kg\day

59. In diabetic ketoacidosis bicarbonate therapy may increase the risk of

A. hypokalemia
B. hyperkalemia
C. hyponatremia
D. hypernatremia
E. hypocalcemia

60. What is the recommended percent of energy from protein in diabetic patient diet?

A. 4-10%
B. 12-20%
C. 22-28%
D. 30-36%
E. 38-44%

61. What is the target pre-meal blood glucose range in an 8- year-old patient with IDDM?

A. 100-200 mg\dl
B. 90-150 mg\dl
C. 80-130 mg\dl
D. 70-120 mg\dl
E. 65-120 mg\dl

62. Nutrition plays an essential role in the management of patients with T1DM. The caloric mixture should comprise approximately

A. 35% carbohydrate, 30% fat, and 35% protein
B. 45% carbohydrate, 30% fat, and 25% protein
C. 55% carbohydrate, 30% fat, and 15% protein
D. 55% carbohydrate, 20% fat, and 25% protein
E. 45% carbohydrate, 25% fat, and 35% protein

63. Diets with high fiber content are useful in improving control of blood glucose. Daily recommended fiber intake can be determined using the equation: Gram of fiber daily = age in years +

A. 3
B. 5
C. 7
D. 9
E. 11

64. What is the most helpful test to assess glycemic control in thalassemic patient with newly diagnosed T1DM?

A. HbA1c
B. Fructosamine
C. Prealbumin
D. Galactosamine
E. Hb F

65. What is the major complication of exercise in patients with diabetes?

A. Hypoglycemia
B. Hypercalcemia
C. Hypernatremia
D. Hypokalemia
E. Hypomagnesaemia

66. Acute orchitis is common in pubertal or adult males with mumps and may lead to subfertility in

A. 3%
B. 8%
C. 13%
D. 18%
E. 23%

67. Transient gynecomastia occurs in 60–90% of male newborns, most cases resolve within

A. first wk
B. 1-2 wk
C. 4-8 wk
D. 9-12 wk
E. 4-8 mo

68. A 9- month- old girl brought by her mother due to stunted growth, on examination extremities are short, and head size is normal, the anterior fontanel is large, protruded tongue, dry scaly pale yellow skin, with clear sclera. Which of the following is the **MOST** appropriate initial test?

A. GH assay
B. Serum calcium
C. TSH
D. LFT
E. CBC

Chapter 25

The Endocrine System Answers

HAYDER ALMUSAWI

1.(C) Serum TSH levels are the most sensitive test for primary thyroid dysfunction. Serum TSH levels are elevated in primary hypothyroidism and suppressed in hyperthyroidism. In central (secondary) hypothyroidism, serum TSH is either low or inappropriately in the normal range despite a low serum T4 or free T4 level.

2.(B) Estimation of TBG binding is occasionally necessary because it is increased or decreased in a variety of clinical situations, with effects on the level of total T4 and T3. TBG binds approximately 70% of T4 and 50% of T3. TBG levels increase in pregnancy, in the newborn period, with hepatitis, and with administration of estrogens (oral contraceptives), selective estrogen receptor modulators, heroin or methadone mitotane, and 5-fluorouracil. TBG level decrease with androgens, anabolic steroids, glucocorticoids, nicotinic acid, and L asparaginase. These effects are the results of modulation of hepatic synthesis of TBG.

3.(D) Congenital TBG deficiency is an X-linked dominant trait. It is most often discovered through screening programs for neonatal hypothyroidism. Affected patients have low levels of total T4 (usually<4µg/dL) and elevated triiodothyronine (T3) resin uptake, but levels of free T4 and thyrotropin (TSH) are normal.

4.(B) The dose on a weight basis gradually decreases with age. For children age 1-3 yr, the average daily L - T4 dose is 4-6 µg/kg; for age 3-10 yr, 3-5 µg/kg; and for age 10-16 yr, 2-4 µg/kg.

5.(B) Amiodarone, a drug used for cardiac arrhythmias and consisting of 37% iodine by weight, causes hypothyroidism in approximately 20% of treated children. Children treated with amiodarone should have serial monitoring of thyroid function.

6.(A) Familial dysalbuminemic hyperthyroxinemia is an autosomal dominant disorder that may be mistaken for hyperthyroidism. Patients have an abnormal albumin variant with a markedly increased affinity for T4 that leads to increased serum concentrations of T4. Levels of T3 are normal or slightly elevated.

However, levels of free T4, free T3, and TSH are normal, and affected patients are euthyroid.

7.(C) Thyroid dysgenesis is the most common cause of permanent congenital hypothyroidism, accounting for 80–85% of cases. In approximately 33% of cases of dysgenesis no thyroid tissue is present (agenesis). In the other 66% of infants rudiments of thyroid tissue are present either in the normal position (hypoplasia) or in an ectopic location.

8.(A) The boy who has normal descended testes and a normal penis and is discovered to have fallopian tubes and a rudimentary uterus, has the persistent müllerian duct syndrome. This is an autosomal recessive disorder and is associated with an XY chromosomal pattern.

9.(E) Sleep, exercise, physical stress, trauma, acute illness, puberty , fasting, and hypoglycemia stimulate the release of GH, whereas hyperglycemia, hypothyroidism, and glucocorticoids inhibit GH release.

10.(A) Arginine vasopressin, oxytocin, angiotensin II, and cholecystokinin stimulate release of CRH and ACTH, whereas atrial natriuretic peptide and opioids inhibit release of CRH and ACTH.

11.(C) Prolonged immobilization can lead to hypercalcemia.

12.(B) The U.S. Food and Drug Administration (FDA) has approved 8 pediatric indications for rhGH treatment to promote linear growth. They are GH deficiency, Turner syndrome, chronic renal failure before transplantation, idiopathic short stature, small-for-gestational-age short stature, Prader-Willi syndrome, SHOX gene abnormality, and Noonan syndrome.

13.(A) Criteria for stopping GH treatment include a decision by the patient that he or she is tall enough, a growth rate <1 inch/yr , and a bone age >14 yr in girls and >16 yr in boys.

14.(C) The risk of hypoglycemia is reduced by giving the injections concurrently with a meal or snack.

15.(D) The diagnosis of DI is established if the serum osmolality is >300 mOsm/kg, and the urine osmolality is <300 mOsm/kg. DI is unlikely if the serum osmolality is <270 mOsm/kg or the urine osmolality is >600 mOsm/kg. If the patient's serum osmolality is <300 mOsm/kg (but >270 mOsm/kg) and pathologic polyuria and polydipsia are present, a water deprivation test is indicated to establish the diagnosis of DI and to differentiate central from nephrogenic causes.

16.(A) The initial approach to a patient with hyponatremia begins with determination of the volume status.

17.(C) Systemic dehydration and runner hyponatremia associated with hypovolemia and low urine sodium, SIADH with hypervolemia and high urine sodium while factitious hyponatremia associated with normovolemia and normal urine sodium.

18.(B) Syndrome of inappropriate antidiuretic hormone secretion (SIADH) is characterized by hyponatremia, an inappropriately concentrated urine (>100 mOsm/kg), normal or slightly elevated plasma volume, normal-to-high urine sodium, and low serum uric acid.

19.(D) The maximum amount of water a person with normal renal function can consume daily is 10 L/m2.

20.(A) The most commonly diagnosed adenoma during childhood is prolactinoma, followed by corticotropinoma, and then somatotropinoma.

21.(D) Beckwith-Wiedemann syndrome clinical features include eye proptosis with periorbital fullness, mid-glabellar capillary malformation (nevus flammeus), earlobe creases and pits, and macrosomia, including macroglossia, hepatosplenomegaly , nephromegaly , and omphalocele. They also have hypoglycemia secondary to hyperinsulinemia as a result of pancreatic β-cell hyperplasia.

22.(A) In Graves disease, serum TSH is suppressed and free T4 and T3 are elevated.

23.(A) Children with Beckwith-Wiedemann syndrome are predisposed to embryonal tumors, including Wilms tumor , hepatoblastoma, neuroblastoma, and adrenocortical carcinoma. Cancer risk is high until 8 yr of age, and regular surveillance with abdominal ultrasound and measurement of α-fetoprotein is recommended every 3 mo until age 8 yr .

24.(C)

25.(A) Prenatal infection with rubella is associated with β-cell autoimmunity in up to 70%, with development of T1DM in up to 40% of infected children. The time lag between infection and development of diabetes may be as high as 20 yr. Interestingly, there appears to be no increase in risk of diabetes when rubella infection develops after birth or when live-virus rubella immunization is used.

26.(D)

27.(D)

28.(B)

29.(D) Diabetic Ketoacidosis Treatment Protocol 1st hr 10-20 mL/kg IV bolus 0.9% NaCl or LR then 2nd hr until DKA resolution 0.45% NaCl and 5% glucose if blood sugar <250 mg/dL.

30.(E) Methimazole is the first line ATD for children with Graves disease. Propylthiouracil is a similar ATD that is effective in hyperthyroidism, but its use is not recommended in children due to its potential to cause liver failure.

31.(E) About 35% of infants with congenital hypothyroidism and a normally located thyroid gland have transient disease and do not require lifelong therapy. In patients who might have transient disease, a trial of stopping L -T4 for 4 wk may be undertaken after 3 yr of age for 3-4 wk to assess whether the TSH rises significantly, indicating the presence of permanent hypothyroidism.

32.(C) About 20% of children with type 1 diabetes mellitus develop thyroid autoantibodies, and about 5% become hypothyroid.

33.(B) When thyrotoxicosis is caused by exogenous thyroid hormone (thyrotoxicosis factitia), levels of free T4 and TSH are the same as those seen in Graves disease but, in contrast to Graves disease, thyroid size is small, serum thyroglobulin is very low, and 123 I radioiodine uptake is suppressed.

34.(C) Midline facial anomalies (cleft lip, palate) or the finding of a solitary maxillary central incisor indicate a high likelihood of GH or other anterior or posterior pituitary hormone deficiency.

35.(A) Laron syndrome involves mutations of the GH receptor. Children with this condition clinically resemble those with severe IGHD. Birth length tends to be about 1 SD below the mean, and severe short stature with lengths >4 SD below the mean is present by 1 yr of age. Resting and stimulated GH levels tend to be high and insulin-like growth factor (IGF) 1 levels are low.

36.(A) Linear growth failure (may be the only clinical feature present), proportionate short stature, low height velocity, weight for length appropriate or increased, micropenis in males, small midface, high-pitched voice, and delayed dental eruption.

37.(B) A variety of provocative tests have been devised that rapidly increase the level of GH in normal children. These include administration of insulin, arginine, clonidine, levodopa, or glucagon.

38.(C) The U.S. Food and Drug Administration (FDA) has approved 8 pediatric indications for rhGH treatment to promote linear growth. These are GH deficiency, Turner syndrome, chronic renal failure before transplantation, idiopathic short stature, small-for-gestational-age short stature, Prader-Willi syndrome, SHOX gene abnormality, and Noonan syndrome.

39.(C) Criteria for stopping GH treatment include a decision by the patient that he or she is tall enough, a growth rate <1 inch/yr, and a bone age >14 yr in girls and >16 yr in boys.

40.(D) Periodic evaluation of thyroid and adrenal function is indicated for all patients diagnosed with GH deficiency.
41.(A) Demeclocycline, lithium, cisplatin and methoxyflurane.
42.(C) Extracellular fluid tonicity is regulated almost exclusively by water intake and excretion, whereas extracellular volume is regulated by sodium intake and excretion. The control of plasma tonicity and intravascular volume involves a complex integration of endocrine, neural, behavioral, and paracrine systems.
43.(B) The distal femoral and proximal tibial epiphyses, normally present at birth.
44.(C) Acquired NDI can result from hypercalcemia or hypokalemia and is associated with lithium, demeclocycline, foscarnet, clozapine, amphotericin, methicillin, and rifampin.
45.(A) Systemic dehydration, decreased effective plasma volume, primary salt loss (nonrenal) and runner's hyponatremia all of them associated with hyponatremia, decrease intravascular volume and low urine sodium.
46.(E) Syndrome of inappropriate antidiuretic hormone secretion (SIADH) is characterized by hyponatremia, an inappropriately concentrated urine (>100 mOsm/kg), normal or slightly elevated plasma volume, normal-to-high urine sodium, and low serum uric acid.
47.(C) Perlman syndrome associated with macrosomia, unusual facies, nephroblastosis, severe hypotonia and very high risk of Wilms tumor.
48.(A)
49.(B)
50.(C) In females, the breast bud (thelarche) is usually the first sign of puberty (10-11 yr), followed by the appearance of pubic hair (pubarche) 6-12 mo later. The interval to the onset of menstrual activity (menarche) is usually 2-2.5 yr but may be as long as 6 yr. Peak height velocity occurs early (at breast stage II–III, typically between 11 and 12 yr of age) in females and always precedes menarche. The mean age of menarche is about 12.75 yr.
51.(C)
52.(B) L-T4 tablets should be crushed and mixed with a small volume (1-2 mL) of liquid. L-T4 tablets should not be mixed with soy protein formulas, concentrated iron, or calcium, because these can inhibit L-T4 absorption. Although it is recommended to administer L -T4 on an empty stomach and avoid food for 30-60 min, this is not practical in an infant.
53.(B) DiGeorge/velocardiofacial syndrome occurs in 1 in 4,000 newborns. In 90% of patients, the condition is caused by a deletion of chromosome 22q11.2. This syndrome has also been reported in a small number of patients with a

deletion of chromosome 10p13, in infants of diabetic mothers, and in infants born to mothers treated with retinoic acid for acne early in pregnancy.

54.(A) Wolfram syndrome (DIDMOD: diabetes insipidus, diabetes mellitus, optic atrophy, deafness) is an autosomal recessive disease due predominantly to mutations in the WFS1 gene and is a progressive neurodegenerative disease. Case definition requires the presence of T1DM and optic atrophy. This syndrome may be present in ~5% of patients with T1DM.

55.(C) The most common cause of hypocalcemia and hypophosphatemia is prolonged vitamin D deficiency. This deficiency can best be determined by measuring 25-hydroxyvitamin D (25[OH]D) levels because it is the primary storage form of vitamin D. Although 1,25-dihydroxyvitamin D (1,25[OH]2D) is the active metabolite of vitamin D, measuring 1,25(OH)2D levels is less useful in the diagnosis of vitamin D deficiency because hypocalcemia drives parathyroid hormone (PTH) secretion and increases the conversion of 25(OH)D to 1,25(OH)2D.

56.(A) There is an increased risk of cardiovascular disease associated with diabetes, so it is recommended to obtain a fasting lipid profile in children ≥10 yr of age once glucose control has been established.

57.(E)

58.(B)

59.(A) Bicarbonate therapy may increase the risk of hypokalemia and cerebral edema so should be considered only in situations with severe acidosis unresponsive to standard DKA management.

60.(B) High-protein intakes may contribute to diabetic nephropathy. Low intakes may reverse preclinical nephropathy. Therefore, 12–20% of energy is recommended; lower end of range is preferred.

61.(C) <5 yr: 100-200 mg\dl , 5-11 yr: 80-150 mg\dl, 12-15 yr: 80-130 mg\dl, 16-18 yr: 70-120 mg\dl.

62.(C) The caloric mixture should comprise approximately 55% carbohydrate, 30% fat, and 15% protein, but must be individualized to meet specific patient needs.

63.(B)

64.(B) HbA1c values may be elevated in thalassemia (or other conditions with elevated hemoglobin F) and lower in sickle cell disease (or other conditions with high red blood cell turnover). Fructosamine can be used instead of HbA1c in these patients.

65.(A) A major complication of exercise in patients with diabetes is the presence of a hypoglycemic reaction during or within hours after exercise. In patients

who are in poor metabolic control, vigorous exercise may precipitate ketoacidosis because of the exercise-induced increase in the counterregulatory hormones.

66.(C) Acute orchitis is common in pubertal or adult males with mumps and may lead to subfertility in 13% of cases, though infertility is rare. Testosterone secretion usually remains normal. The incidence of mumps orchitis in postpubertal males has increased in some areas as a result of a decrease in measles, mumps, and rubella vaccination uptake.

67.(C) Most cases resolve within 4-8 wk of birth, but a few can last as long as 12 mo.

68.(C)

Chapter 26

The Nervous System Questions

ZUHAIR ALMUSAWI

1. Which of the following is regarded as a discomforting test for the child?
   A. Measurement of weight
   B. Measurement of length
   C. Measurement of head circumference
   D. Measurement of body temperature
   E. Examination of deep tendon reflexes

2. At What age smell can be tested reliably?
   A. 32nd wk of gestation
   B. Full-term newborn
   C. Three months
   D. Six months
   E. One year

3. What is the gestational age at which a premature infant can blinks in response to a bright light?
   A. 26 wk of corrected gestational age
   B. 28 wk of corrected gestational age
   C. 30 wk of corrected gestational age
   D. 32 wk of corrected gestational age
   E. 34 wk of corrected gestational age

4. Which of the following is a sequelae of complete paralysis of the oculomotor nerve?
   A. Ptosis
   B. Constriction of the pupil
   C. Displacement of the eye inward
   D. Displacement of the eye upward
   E. Impairment of adduction

5. Which of the following terms is used to describe involuntary, chaotic, conjugate oscillations of the eyes?

A. Pendular nystagmus
B. Jerk nystagmus
C. Horizontal nystagmus
D. Ocular bobbing
E. Opsoclonus

6. Which of the following is usually caused by unilateral injury of the vagus nerve?

A. Hoarse voice
B. Respiratory distress
C. Nasal regurgitation
D. Pooling of secretions
E. Immobile, low-lying soft palate

7. Which of the following cause an elevated polymorphonuclear WBC count in the CSF?

A. Early phase of aseptic meningitis
B. Tuberculous meningitis
C. Fungal meningitis
D. Spinal cord tumor
E. Immunologic disorders

8. The normal CSF protein is 10-40 mg/dL in a child and as high as 120 mg/dL in a neonate. The CSF protein falls to the normal childhood range by

A. 1 mo of age
B. 2 mo of age
C. 3 mo of age
D. 4 mo of age
E. 5 mo of age

9. What is the imaging method of choice for detecting intracranial hemorrhage in infancy?

A. Skull x-ray
B. Cranial ultrasonography
C. Cranial CT
D. Cranial CT angiography

E. Brain MRI

10. What is the preferred imaging procedure to detect infarcts lasting more than 24 hr?

A. Trans cranial Doppler
B. Cranial ultrasonography
C. Cranial CT
D. Cranial CT angiography
E. Brain MRI

11. What is the preferred imaging procedure to detect brain tumor?

A. Skull x-rays
B. Cranial ultrasonography
C. Cranial CT
D. Cranial CT angiography
E. Cranial MRI

12. What is the preferred brain imaging procedure to detect inborn errors of metabolism?

A. Proton MR spectroscopy (MRS)
B. Positron emission tomography (PET)
C. Cranial CT
D. Cranial CT angiography
E. MRI with gadolinium

13. Generalized epileptiform discharges typically occur in children with

A. cerebral dysgenesis
B. Brain cysts
C. slow growing brain tumors
D. glial scar tissue
E. structurally normal brains

14. What is the **BEST** investigation for cases of occult spinal dysraphism?

A. MRI
B. ultrasonography
C. spine x-ray
D. CT scan
E. alpha feto-protein

15. A 25-year-old mother gives birth to a full-term boy with myelomeningocele, she is reluctant to be pregnant again, and she asked you about chance of recurrence.
What is your proper answer?

A. It is negligible
B. It is 3–4%
C. It is 6–8%
D. It is 10%
E. It is 25%

16. Maternal periconceptional use of folic acid supplementation reduces the incidence of neural tube defects (NTDs) in pregnancies at risk by at least 50%. To be effective, folic acid supplementation should be initiated before conception and continued until at least the

A. 4th wk of gestation
B. 8th wk of gestation
C. 12th wk of gestation
D. 16th wk of gestation
E. 20th wk of gestation

17. A 33-year-old woman with history of previously affected child with myelomeningocele is planning for a new pregnancy, she read about prevention and she asks you how?
Your appropriate answer is, supplementation should be started 1 mo before the time of the planned conception with

A. 0.4 mg of folic acid daily
B. 0.8 mg of folic acid daily
C. 1 mg of folic acid daily
D. 4 mg of folic acid daily
E. 5 mg of folic acid daily

18. What is the term applied for the condition in which there is absence of cerebral convolutions and a poorly formed sylvian fissure?

A. Lissencephaly
B. Schizencephaly
C. Septooptic dysplasia
D. Porencephaly
E. Polymicrogyria

19. A 4-month-old female infant presents with repeated seizures resistant to anticonvulsants, electroencephalogram shows hemihypsarrhythmia, X-ray spine shows partially developed (hemivertebra). Fundoscopy reveals abnormalities of the retina, including lacunae and coloboma of the optic disc. MRI brain shows agenesis of the corpus callosum.
Of the following, the **MOST** likely diagnosis is

A. Colpocephaly
B. Aicardi syndrome
C. Holoprosencephaly
D. syntelencephaly
E. Sotos syndrome

20. What is the first thing to be done if the cause of microcephaly is unknown?

A. Mother's serum phenylalanine
B. Chromosome microarray
C. MRI
D. Fasting plasma and urine amino acid analysis
E. Serum ammonia determination

21. What is the approximate total volume of CSF in infants?

A. 25 mL
B. 50 mL
C. 75 mL
D. 100 mL
E. 150 mL

22. What is the **MOST** common cause of communicating hydrocephalus?

A. Tumors in the spinal cord
B. Subarachnoid hemorrhage
C. Leukemic infiltrates
D. Pneumococcal meningitis
E. Tuberculous meningitis

23. A 6-month-old Infant presents with a rapid increase in head size and a prominent occiput.
Of the following, the **MOST** likely diagnosis is

A. Dandy-Walker malformation
B. type I Chiari malformation

C. type II Chiari malformation
D. posterior fossa brain tumor
E. aqueductal stenosis

24. An 18-month-old obese child presented with high forehead, frontal bossing, down slanting palpebral fissures, hypertelorism, long narrow face, prominent mandible, and malar flushing. He is macrocephalic, hypotonic with poor coordination, and moderate cognitive impairment.
Of the following, the **MOST** likely diagnosis is
A. Simpson-Golabi-Behmel syndrome
B. Fragile X syndrome
C. Weaver syndrome
D. Gorlin syndrome
E. Sotos syndrome

25. What is the **MOST** common cause of anatomic megalencephaly?
A. Sotos syndrome
B. fragile X syndrome
C. benign familial megalencephaly
D. Weaver syndrome
E. Gorlin syndrome

26. What is the **MOST** common form of craniosynostosis?
A. Frontal plagiocephaly
B. Occipital plagiocephaly
C. Scaphocephaly
D. Trigonocephaly
E. Turricephaly

27. What is the cause of trigonocephaly?
A. Premature fusion of the lambdoid suture
B. Premature fusion of the coronal suture
C. Premature fusion of the sphenofrontal suture
D. Premature fusion of the metopic suture
E. Premature fusion of the frontoethmoidal suture

28. A 6-year-old child presents with brachycephaly, underdeveloped orbits, ocular proptosis, hypoplasia of the maxilla, and orbital hypertelorism.

Of the following, the **MOST** likely diagnosis is

A. Apert syndrome
B. Carpenter syndrome
C. Chotzen syndrome
D. Pfeiffer syndrome
E. Crouzon syndrome

29. Which of the following is a factor increasing the risk for deformational plagiocephaly?

A. Female
B. Postmaturity
C. Sleep position is prone at birth and at 6 wk
D. Breast feeding
E. Tummy time < 3 times/day

30. What is the **BEST** treatment for a 6-month-old infant with severe deformational plagiocephaly?

A. Watch-and-wait management
B. Repositioning and physiotherapy
C. Increasing tummy time
D. Helmet therapy
E. Surgery

31. Which of the following is a major risk factor for recurrence of febrile seizures?

A. Family history of febrile seizures
B. Family history of epilepsy
C. Complex febrile seizure
D. Male gender
E. Age < 1 yr

32. Which of the following risk factors has the highest incidence for occurrence of subsequent epilepsy after a febrile seizure?

A. Recurrent febrile seizures
B. Fever < 1 hr before febrile seizure
C. Family history of epilepsy
D. Complex febrile seizures (focal)
E. Neurodevelopmental abnormalities

33. A 6-month-old infant develops high fever followed by prolonged focal seizure for more than 30 minutes after vaccination, subsequently the seizures start to occur with lower grade of fever and then without fever. During the second year of his life, atypical absences occur frequently with developmental delay.
Of the following, the **MOST** likely diagnosis is

A. Generalized epilepsy with febrile seizures plus (GEFS+),
B. Dravet syndrome
C. Temporal lobe epilepsy secondary to mesial temporal sclerosis
D. Focal complex febrile seizures
E. Febrile infection–related (or refractory) epilepsy (FIRES)

34. What is the **MOST** frequent cause of febrile status epilepticus?

A. Norovirus
B. Enteroviruses
C. Shigella
D. COVID 19
E. HHV-6B

35. A 10-month-old boy presented with febrile seizure which lasted about 10 minutes with no clinical signs of meningeal irritation, his immunization status was unknown.
A medical student asks you about lumbar puncture LP for this baby; your answer should be

A. No need for LP at time being
B. LP should be done because his immunization status was unknown
C. LP should be done after brain ultrasound
D. LP should be done if he developed another seizure within 24 hour
E. LP should be done because he is a male gender

36. An 18-month-old boy is presenting with the first simple febrile seizure and is otherwise neurologically healthy; his father read in the net that EEG has a significant role in the management of his son.
Your proper answer should be

A. EEG should not be performed as part of the evaluation
B. EEG would predict the future recurrence of febrile seizures
C. EEG would predict the future recurrence of epilepsy
D. EEG should be done after 2 wk of a febrile seizure

E. EEG should be done after second attack of febrile seizure

37. Which of the following blood studies is routinely recommended in the workup of a child with a first simple febrile seizure?

A. Serum electrolytes
B. Calcium
C. Magnesium
D. Complete blood count
E. Blood glucose

38. Which of the following nutritional deficiencies is associated with an increased risk of febrile seizures?

A. Iron
B. Zinc
C. B12
D. B6
E. Folate

39. A 5-year-old child wakes up at night due to a focal seizure with throat tingling and clonic contractions of one side of the face, with drooling and inability to speak but with preserved consciousness and comprehension. EEG shows typical wide-based spikes that are markedly increased in frequency during drowsiness and sleep with normal MRI.

Of the following, the **MOST** likely diagnosis is

A. atypical benign childhood epilepsy with centrotemporal spikes
B. benign childhood epilepsy with centrotemporal spikes
C. benign epilepsy with occipital spikes
D. nocturnal autosomal dominant frontal lobe epilepsy
E. familial benign epilepsy syndromes

40. Which of the following is **TRUE** regarding typical absence seizures?

A. They usually start at 3-4 yr of age
B. They can occur up to hundreds of times per day
C. They usually have an aura
D. They usually have florid automatisms
E. They usually have a postictal period

41. A 7-month-old infant presents with epileptic spasms (that usually occur in clusters), developmental regression, and a high voltage, slow, chaotic background with multifocal spikes EEG.
Of the following, the **MOST** likely diagnosis is

A. Ohtahara syndrome
B. early myoclonic infantile encephalopathy
C. Dravet syndrome
D. West syndrome
E. Lennox-Gastaut syndrome

42. A 5-year-old boy presents with developmental delay, tonic seizures that occur in sleep with EEG findings of 1- to 2-Hz spike and slow waves, polyspike bursts in sleep, and a slow background in wakefulness. The seizures are intractable despite multiple therapies.
Of the following, the **MOST** likely diagnosis is

A. Ohtahara syndrome
B. Doose syndrome
C. Dravet syndrome
D. West syndrome
E. Lennox-Gastaut syndrome

43. A 3-week-old newborn presents with recurrent generalized tonic seizures that progress to status epilepticus which is not controlled by multiple antiepileptic drugs. The mother reports increased fetal movements in utero. Plasma and CSF showed elevated α-aminoadipic semialdehyde and pipecolic acid levels.
Of the following, the **MOST** likely diagnosis is

A. pyridoxine dependent epilepsy
B. folinic acid–responsive seizures
C. pyridoxal phosphate–responsive neonatal epileptic encephalopathy
D. cerebral folate deficiency
E. tetrahydrobiopterin deficiencies

44. Which of the following drugs is the first choice for focal seizures?

A. Oxcarbazepine
B. Ethosuximide
C. Valproate
D. Lamotrigine

E. Topiramate

45. Which of the following drugs is **MOST** often used as initial treatment of absence seizures?

A. Valproate
B. Lamotrigine
C. Ethosuximide
D. Acetazolamide
E. Clonazepam

46. Which of the following antiepileptic drugs may cause apnea?

A. Benzodiazepines
B. Carbamazepine
C. Lamotrigine
D. Levetiracetam
E. Phenobarbital

47. Which of the following antiepileptic drugs may cause irreversible visual field deficit?

A. Benzodiazepines
B. Carbamazepine
C. Vigabatrin
D. Levetiracetam
E. Phenobarbital

48. Which of the following antiepileptic drugs may cause aplastic anemia?

A. Benzodiazepines
B. Carbamazepine
C. Vigabatrin
D. Levetiracetam
E. Phenobarbital

49. Hepatic and pancreatic toxicity are serious side effects in which of the following antiepileptic drugs?

A. Benzodiazepines
B. Valproic acid
C. Vigabatrin
D. Levetiracetam

E. Gabapentin

50. Which of the following antiepileptic drugs is used in an epileptic patient with migraine?

A. Benzodiazepines
B. Valproic acid
C. Vigabatrin
D. Levetiracetam
E. Gabapentin

51. Which of the following antiepileptic drugs is used in an epileptic obese patient?

A. Topiramate
B. Valproic acid
C. Vigabatrin
D. Levetiracetam
E. Gabapentin

52. Which of the following antiepileptic drugs is better to be used in a patient with both absence and generalized tonic-clonic seizures?

A. Topiramate
B. Valproic acid
C. Phenytoin
D. Ethosuximide
E. Gabapentin

53. Which of the following is the safest antiepileptic drug to use during pregnancy?

A. Levetiracetam
B. Valproic acid
C. Phenytoin
D. Ethosuximide
E. Gabapentin

54. Which of the following antiepileptic drugs requires frequent (even weekly) monitoring of liver function and blood counts throughout the therapy?

A. Levetiracetam
B. Valproic acid

C. Felbamate
D. Ethosuximide
E. Gabapentin

55. Which of the following antiepileptic drugs requires giving vitamin D supplementation with it?
A. Levetiracetam
B. Gabapentin
C. Felbamate
D. Ethosuximide
E. Phenytoin

56. Which of the following is an absolute contraindication to ketogenic diet?
A. Primary carnitine deficiency
B. Myoclonic-astatic epilepsy
C. Tuberous sclerosis complex
D. Rett syndrome
E. Dravet syndrome

57. Discontinuation of antiepileptic drugs AEDs is usually indicated when children are free of seizures for at least
A. 1 year
B. 2 year
C. 3 year
D. 4 year
E. 5 year

58. Antiepileptic drug AED therapy should be discontinued gradually; often over a period of
A. 1-2 months
B. 2-4 months
C. 3-6 months
D. 4-8 months
E. 5-10 months

59. Which of the following neonatal seizures is frequently not associated with electrographic discharges?
A. Spasms

B. Focal clonic
C. Focal tonic
D. Generalized myoclonic
E. Generalized tonic

60. What is the **MOST** common cause of neonatal seizures?
A. Vascular events
B. Hypoxic-ischemic encephalopathy
C. Intracranial infections
D. Brain malformations
E. Metabolic disturbances

61. A full-term newborn of a malnourished mother presents with hypocalcemic seizure not responding to IV calcium.
Of the following, the **MOST** likely cause is
A. hyponatremia
B. hypernatremia
C. hypomagnesemia
D. hypoglycemia
E. pyridoxine dependency

62. What is the first-choice long-acting drug in neonatal seizures?
A. Lorazepam
B. Phenobarbital
C. Phenytoin
D. Fosphenytoin
E. Levetiracetam

63. A 3-week-old newborn is well and ready to be discharged from NICU; he developed seizure at first day of life due to difficult labor and treated with phenobarbital loading then maintenance on 5mg/kg/day twice daily. An EEG done before discharge and does not show evidence of epileptiform activity. Parents asked you about the fate of anticonvulsant and when can be stopped.
Of the following, the **MOST** appropriate answer is that phenobarbital should be
A. stopped immediately
B. tapered at time of discharge
C. tapered after 3 months
D. tapered after 6months

E. tapered after 12 months

64. What is the **MOST** common type of status epilepticus in children?

A. Convulsive status epilepticus
B. Febrile status epilepticus
C. Nonconvulsive status epilepticus
D. Epilepsia partialis continua
E. Refractory status epilepticus

65. What is the first-line agent recommended by the American Epilepsy Society Guidelines to control status epilepticus?

A. Phenobarbital
B. Lorazepam
C. Phenytoin
D. Valproate
E. Levetiracetam

66. Which of the following investigations is recommended to a 1-year-old boy presents with recurrent severe breath-holding spells?

A. EEG
B. ECG
C. Serum calcium
D. Serum Zinc
E. Echocardiography

67. How you treat a 1-year-old boy presenting with recurrent severe cyanotic breath-holding spells?

A. Education and reassurance of the parents
B. Atropine sulfate
C. Anti-seizure drug therapy
D. Cardiac pacemaker
E. Basic cardiopulmonary resuscitation

68. A 9-year-old boy presents with pallor and sweating followed by a gradual collapse with loss of consciousness triggered by standing for a long time without movement when taking a hot shower.
What is the **MOST** likely cause of this event?

A. Complex partial seizure

B. Vasovagal syncope
C. Temporal lobe epilepsy
D. Exercise-induced anaphylaxis
E. Long QT syndrome

69. A 5-year-old boy presents with postural imbalance and nystagmus, he reported that objects seem to be moving toward him. The child appears frightened during the episode with diaphoresis, nausea, and vomiting. MRI and EEG are normal.
Of the following, the **MOST** likely diagnosis is
A. cyclic vomiting syndrome
B. long QT syndrome
C. benign paroxysmal vertigo of childhood
D. The Alice in Wonderland syndrome
E. hyperekplexia

70. A 6-month-old infant presents with paroxysmal episodes of generalized stiffening and opisthotonic posturing accompanied by apnea, staring, and minimal jerking of the extremities. These episodes often occur 30 min after a feed.
What is the **MOST** likely cause of these events?
A. Benign paroxysmal torticollis of infancy
B. Exaggerated startle reflex
C. Stiff baby syndrome
D. Sandifer syndrome
E. Paroxysmal dyskinesia

71. A 4-month-old boy infant presents with repeated head drops, hundreds times per day, without concurrent EEG epileptic activity. His parents asked about prognosis, your answer is
A. It persists for life
B. It subsides when child walks without support
C. It subsides by 1st year of life
D. It may lead to developmental delay
E. It may convert to epileptic myoclonus

72. A 2- year-old girl brought by her mother complaining from rhythmic rocking movements in lying and sometimes sitting position often associated with perspiration, irregular breathing, and grunting, but no loss of consciousness. Which of the following is the **MOST** likely cause of this event?

A. Tic
B. Mannerism
C. Anxiety attack
D. Infantile gratification
E. Hyperventilation spell

73. What is the **MOST** common presenting complaint of intramedullary spinal cord tumors?

A. Gait disturbance
B. Back pain
C. Sensory deficits
D. Scoliosis
E. Bladder disturbance

74. Status migrainosus is defined as migraine that persists beyond

A. 24 hr
B. 48 hr
C. 72 hr
D. 96 hr
E. one week

75. What is the **MOST** frequent symptom of idiopathic intracranial hypertension?

A. Vomiting
B. Transient visual obscuration
C. Chronic progressive frontal headache
D. Diplopia
E. Pulsatile tinnitus

76. What is the preferred initial drug to treat migraine headache?

A. Acetaminophen
B. Naproxen sodium
C. Aspirin
D. Triptan

E. Ibuprofen

77. Which of the following differentiate tension-type headache from migraine?
A. Focal in location
B. Worsened by physical activity
C. Nonthrobbing quality
D. Frequently associated with nausea
E. Moderate to severe intensity

78. Which of the following neurocutaneous syndromes is X-linked dominant?
A. Neurofibromatosis type 2 (NF2)
B. Tuberous sclerosis complex (TSC)
C. Sturge- Weber syndrome (SWS)
D. von Hippel–Lindau disease (VHL)
E. Incontinentia pigmenti

79. What is the **MOST** frequent lesion associated with neurofibromatosis type 2?
A. Bilateral vestibular schwannomas
B. optic nerve schwannomas
C. Intracranial meningiomas
D. Spinal tumors
E. Peripheral neuropathy

80. Definite tuberous sclerosis complex TSC is diagnosed when at least two major or one major plus two minor features are present.
Which of the following is a minor feature?
A. Subependymal nodules
B. Facial angiofibromas (≥3)
C. Ungual fibromas (≥2)
D. Retinal achromic patch
E. Shagreen patch

81. What is the **MOST** common neurologic manifestation of tuberous sclerosis complex?
A. Epilepsy
B. Nystagmus
C. Sleep disorder

D. Attention deficit hyperactivity disorder
E. Depression

82. What is the first-line therapy for infantile spasms in Tuberous Sclerosis Complex TSC?
A. Adrenocorticotropic hormone (ACTH)
B. Vigabatrin
C. Epilepsy surgery
D. Valproate
E. Everolimus

83. A young mother brought her 6-month-old boy with infantile spasm secondary to tuberous sclerosis complex TSC with good response to vigabatrin, his cardiac echo revealed cardiac rhabdomyoma. The mother counsels you about the fate of this cardiac lesion.
Your best answer should be
A. It usually ends with congestive heart failure
B. It tends to slowly resolve spontaneously
C. It usually ends with arrhythmias
D. If started in fetal life, it is fatal in infancy
E. It usually needs surgical intervention

84. What is the imaging modality of choice for demonstrating the extension of pial capillary malformation in Sturge-Weber syndrome (SWS)?
A. Brain MRI
B. Brain MRI with contrast
C. Head CT
D. Head CT with contrast
E. Brain US

85. What is the **MOST** common cause of death in von Hippel–Lindau disease?
A. Pheochromocytoma
B. Renal carcinoma
C. Cerebellar hemangioblastoma
D. Hemangioblastoma of the spinal cord
E. Retinal angiomas

86. A 9-month-old infant presents with developmental delay, hypotonia, abnormal eye movements, abnormal respirations, and a distinctive facial feature. Axial MRI brain demonstrates “molar tooth sign”.
Of the following, the **MOST** likely diagnosis is

A. agenesis of vermis of the cerebellum
B. Chiari malformation
C. Dandy-Walker syndrome
D. Leigh disease
E. Joubert syndrome

87. A 3-year-old child presents with vomiting, truncal ataxia, horizontal nystagmus, and dysarthria. Examination of the cerebrospinal fluid is normal. The child had varicella 3 weeks ago.
What is the **MOST** likely diagnosis?

A. Infectious cerebellitis
B. Acute labyrinthitis
C. Neuroblastoma
D. Acute cerebellar ataxia
E. Hartnup disease

88. A 6-year-old child presents with steatorrhea, failure to thrive, ataxia, peripheral neuritis, and retinitis pigmentosa. Blood smear shows acanthocytosis, Serum chemistries reveal decreased levels of cholesterol and triglycerides and absent serum β-lipoproteins.
Of the following, the **MOST** likely diagnosis is

A. Bassen-Kornzweig disease
B. arginosuccinic aciduria
C. Hartnup disease
D. Refsum disease
E. Friedreich ataxia

89. A 12-year-old boy presents with slowly progressive ataxia which affect the lower extremities more than the upper extremities. Examination demonstrates a positive Romberg test, absent ankle deep tendon reflexes; and extensor plantar response. The boy has explosive dysarthric speech, nystagmus, with preserved intelligence. He also has high-arched feet and hammer toes.
Of the following, the **MOST** likely diagnosis is

A. Roussy-Levy disease

B. Ramsay Hunt syndrome
C. Friedreich ataxia
D. ataxia-telangiectasia
E. Refsum disease

90. A 9- year-old girl awakened yesterday morning with clumsiness, dropped the tea cup from her hand on breakfast then she started to develop generalized abnormal body movement involving the face with crying and unintelligible speech. When the patient asked to hold the upper limbs overhead, arms and palms turn outward. Examination of heart is normal.
Of the following, the **MOST** likely diagnosis is
A. benign hereditary chorea
B. Sydenham chorea
C. chorea secondary to systemic lupus erythematosus
D. chorea secondary to antiphospholipid antibody syndrome
E. paraneoplastic chorea

91. Which of the following nutritional disorders may cause secondary intracranial hypertension without an obstructive lesion on MRI?
A. Nutritional rickets
B. Hypovitaminosis A
C. Zinc deficiency
D. Copper deficiency
E. Vitamin K deficiency

92. A 5-week-old infant presents with repetitive focal myoclonic jerks occurring during sleep, more prominent in the upper extremities continued for few seconds, these jerks began during the 1st week of life, waking the baby causes the movements to abruptly cease. Neurologic examination is normal.
Of the following, the **MOST** likely cause of these movements is
A. Opsoclonus myoclonus (ataxia) syndrome
B. Benign neonatal sleep myoclonus
C. Benign myoclonus of early infancy
D. Hypoglycemia
E. Myoclonic epilepsy with ragged red fibers (MERRF)

93. What is the **MOST** common cause of spastic diplegia?
A. Prematurity

B. Ischemia
C. hyperbilirubinemia
D. Thrombophilic disorders
E. Periventricular hemorrhagic infarction

94. What is the **MOST** common clinical manifestation of burn encephalopathy?
A. Altered states of consciousness
B. Hallucinations
C. Coma
D. Seizures
E. Irritability

95. A 14-year-old girl complained from rapidly progressive anxiety, agitation, delusional thoughts, bizarre behavior, language disintegration, and insomnia. After few weeks, additional symptoms occur, including a decreased level of consciousness, seizures, choreoathetoid movements, and autonomic instability. Brain MRI studies shows nonspecific cortical and subcortical T2–fluid attenuated inversion recovery (FLAIR) signal abnormalities, the cerebrospinal fluid shows moderate lymphocytic pleocytosis, the electroencephalogram (EEG) shows diffuse slow activity in the delta and theta ranges while the abdominal ultrasound revealed teratoma of the ovary.
Of the following, the **MOST** likely diagnosis is
A. anti-N -Methyl-D-Aspartate Receptor Encephalitis
B. neuroleptic malignant syndrome
C. limbic encephalitis
D. Kleine-Levin syndrome
E. encephalitis lethargica

96. A 7-year-old child presents with progressive refractory focal seizures, cognitive deterioration, and focal neurologic deficits. MRI shows atrophy of right brain hemisphere.
Of the following, the **MOST** likely diagnosis is
A. Bickerstaff Encephalitis
B. acute disseminated encephalomyelitis (ADEM)
C. Rasmussen encephalitis
D. Ophelia syndrome
E. autoimmune limbic encephalitis

97. What is the **MOST** effective treatment for control of seizures in Rasmussen encephalitis?

A. High-dose steroids
B. Plasma exchange
C. IVIG
D. Tacrolimus
E. Functional hemispherectomy

98. A 15-month-old child presents with progressive loss of motor and language milestones beginning at 6 months of age with seizures, macrocephaly, and doll-like face. Abdominal examination reveals splenomegaly while fundoscopy shows cherry-red spots, and the auditory brainstem responses shows prolonged latencies.

Of the following, the **MOST** likely diagnosis is

A. Sandhoff disease
B. Juvenile GM2 gangliosidosis
C. Tay-Sachs disease TSD
D. Krabbe disease
E. Metachromatic leukodystrophy

99. A 10-month-old boy infant presents with excessive irritability and crying, unexplained episodes of hyperpyrexia, vomiting, and difficulty feeding, diagnosed by GP doctor as colic with frequent milk formula change with no improvement, now he develops generalized seizures , alterations in body tone with rigidity and opisthotonos and visual inattentiveness as a result of optic atrophy.

Of the following, the **MOST** likely diagnosis is

A. Sandhoff disease
B. juvenile GM2 gangliosidosis
C. Tay-Sachs disease
D. Krabbe disease
E. Metachromatic leukodystrophy

100. A 2-year-old boy has insidious onset of gait disturbance, frequent falls, with feeding and swallowing impairment. The extremities are hypotonic, and the deep-tendon reflexes are absent. The parents noticed deterioration in intellectual function and dysarthric slurred speech. Examination of the retina shows optic atrophy.

Neurophysiologic evaluation shows slowing of peripheral nerve conduction velocities. CT and MRI images of the brain indicate diffuse symmetric attenuation of the cerebellar and cerebral white matter.
Of the following, the **MOST** likely diagnosis is

A. Sandhoff disease
B. Juvenile GM2 gangliosidosis
C. Tay-Sachs disease
D. Krabbe disease
E. Late infantile metachromatic leukodystrophy

101. A 3-year-old microcephalic female with regression of language and motor milestones, presents with ataxic gait and fine tremor of hand movements with sighing respirations, and intermittent periods of apnea. The patient develops generalized tonic-clonic convulsions with autistic behavior.
Of the following, the **MOST** likely diagnosis is

A. occipital horn syndrome
B. Rett Syndrome
C. Menkes disease
D. Tay-Sachs disease
E. Krabbe disease

102. What is the hallmark of acute disseminated encephalomyelitis ADEM?

A. Headache
B. Meningeal signs
C. Seizures
D. Cranial neuropathies
E. Encephalopathy

103. Which of the following drugs may cause secondary intracranial hypertension without an obstructive lesion on MRI?

A. Nitrofurantoin
B. Ceftriaxone
C. Ampicillin
D. Cefixime
E. Vancomycin

104. A 9-year-old boy presents with periocular pain and headache after a non-specific viral illness, few days later, he develops bilateral visual loss.

What is the **MOST** likely cause of this visual loss?

A. Sarcoidosis
B. Behçet disease
C. Lyme disease
D. Leber hereditary optic neuropathy
E. Optic neuritis

105. Which of the following is characterized by visual loss; sensorineural hearing loss; encephalopathy; headache; memory loss; behavioral disturbances and brain MRI shows involvement of corpus callosum with snowball lesions?

A. Acute disseminated encephalomyelitis
B. Neuromyelitis optica spectrum disorders
C. Neurosarcoidosis
D. Susac syndrome
E. Hypoxic-ischemic vasculopathies

106. Which of the following imaging studies can demonstrate arterial ischemic stroke (AIS) few minutes following the onset?

A. Brain CT
B. Cerebral MRI
C. Diffusion-weighted MRI
D. MR angiography
E. Ultrasound brain

107. Which of the following may cause hand dominance within the first year of life?

A. Perinatal stroke
B. Megaloblastic anemia
C. Sickle cell anemia
D. Moyamoya disease
E. Focal cerebral arteriopathy

108. What is the **MOST** common cause of childhood subarachnoid hemorrhagic stroke?

A. Hereditary hemorrhagic telangiectasia
B. Intracranial aneurysm
C. Arteriovenous malformations
D. Idiopathic thrombocytopenic purpura

E. Hemolytic-uremic syndrome

109. Which of the following CNS infections may have the highest CSF protein level?

A. Tuberculous meningitis
B. Viral meningitis
C. Acute bacterial meningitis
D. Partially treated bacterial meningitis
E. Fungal meningitis

110. Which of the following viral diseases may cause low glucose in CSF?

A. Herpes simplex virus
B. Arboviruses
C. Mumps
D. Adenoviruses
E. Rabies

111. Which of the following bacterial meningitis increased in children with cochlear implants?

A. Staphylococcal bacterial meningitis
B. Anaerobic bacterial meningitis
C. Gram-negative enteric bacterial meningitis
D. Pneumococcal bacterial meningitis
E. Pseudomonas aeruginosa bacterial meningitis

112. What is the **MOST** frequently identified pathogen of bacterial meningitis in children beyond the neonatal period?

A. Streptococcus pneumoniae
B. Neisseria meningitides
C. Haemophilus influenzae Type b
D. Staphylococcus aureus
E. Pseudomonas aeruginosa

113. What are the predominant organisms that cause brain abscesses in children?

A. Staphylococcus aureus
B. Haemophilus spp.
C. Escherichia coli

D. Klebsiella pneumoniae,
E. Streptococcus anginosus

114. Which of the following tests can differentiate between bacterial and viral meningitis?
A. C-reactive protein
B. Erythrocyte sedimentation rate
C. Procalcitonin
D. Blood culture
E. WBC count

115. What is the recommended treatment duration for N. meningitidis meningitis?
A. 7 days
B. 10 days
C. 14 days
D. 21 days
E. 28 days

116. Steroids reduced hearing loss in children with meningitis due to
A. Streptococcus pneumoniae
B. Neisseria meningitides
C. Haemophilus influenzae Type b
D. Staphylococcus aureus
E. Pseudomonas aeruginosa

117. What is the **MOST** common neurologic sequelae from bacterial meningitis?
A. Hearing loss
B. Cognitive impairment
C. Recurrent seizures
D. Delay in acquisition of language
E. Visual impairment

118. What is the **MOST** common CNS manifestation of Varicella-zoster infection?
A. Acute encephalitis
B. Cerebellar ataxia
C. Deafness

D. Flaccid paralysis
E. Aseptic meningitis

119. Deafness is a recognized complication of which of the following viral infections?

A. Polio
B. Measles
C. Mumps
D. Rubella
E. Herpes simplex virus

120. Which of the following viral encephalitis may have normal CSF glucose, protein, and cell counts?

A. Herpes simplex virus
B. Parecho viruses
C. Epstein-Barr virus
D. Cytomegalo virus
E. Mumps

Chapter 26

The Nervous System Answers

ZUHAIR ALMUSAWI

1.(C) The physician should approach the child slowly, reserving any invasive, painful, or discomforting tests for the end of the examination (e.g., measurement of head circumference, gag reflex).

2.(A) Although not a routine component of the examination, smell can be tested reliably as early as the 32nd wk of gestation by presenting a stimulus and observing for an alerting response or withdrawal, or both. Care should be taken to use appropriate stimuli, such as coffee or peppermint, as opposed to strongly aromatic substances (e.g., ammonia inhalants) that stimulate the trigeminal nerve. Each nostril should be tested individually by pinching shut the opposite side.

3.(B) Evaluation of vision in the premature infant presents unique challenges. At 28 wk of corrected gestational age, a premature infant blinks in response to a bright light, and at 32 wk, the infant maintains eye closure until the light source is removed. The pupil reacts to light by 29-32 wk of corrected gestational age; however, the pupillary response is often difficult to evaluate, because premature infants resist eye opening and have poorly pigmented irises. A normal 37-wk infant turns the head and eyes toward a soft light, and a term infant is able to fix on and follow a target, such as the examiner's face.

4.(A) The oculomotor nerve innervates the superior, inferior, and medial recti, as well as the inferior oblique and levator palpebrae superioris muscles. Complete paralysis of the oculomotor nerve causes ptosis, dilation of the pupil, displacement of the eye outward and downward, and impairment of adduction and elevation.

5.(E) Ocular bobbing is characterized by a downward jerk followed by a slow drift back to primary position and is associated with pontine lesions. Opsoclonus describes involuntary, chaotic, conjugate oscillations of the eyes, which are often seen in the setting of neuroblastoma or viral infection.

6.(A) Unilateral injury of the vagus nerve results in weakness of the ipsilateral soft palate and a hoarse voice; bilateral lesions can produce respiratory distress as a result of vocal cord paralysis, as well as nasal regurgitation of fluids, pooling

of secretions, and an immobile, low-lying soft palate. Isolated lesions to the vagus nerve may be a complication of thoracotomies or may be seen in neonates with type II Chiari malformations.

7.(A) An elevated polymorphonuclear count suggests bacterial meningitis or the early phase of aseptic meningitis. CSF lymphocytosis can be seen in aseptic, tuberculous, or fungal meningitis; demyelinating diseases; brain or spinal cord tumor; immunologic disorders, including collagen vascular diseases; and chemical irritation (following myelogram, intrathecal methotrexate).

8.(C) The CSF protein falls to the normal childhood range by 3 mo of age. The CSF protein may be elevated in many processes, including infectious, immunologic, vascular, and degenerative diseases; blockage of CSF flow; as well as tumors of the brain (primary CNS tumors, systemic tumors metastatic to the CNS, infiltrative acute lymphoblastic leukemia) and spinal cord. With a traumatic tap, the CSF protein is increased by approximately 1 mg/dL for every 1,000 red blood cells/mm3.

9.(B) Cranial ultrasonography is the imaging method of choice for detecting intracranial hemorrhage, periventricular leukomalacia, and hydrocephalus in infants with patent anterior fontanels.

10.(E) CT or MRI can detect infarcts more than 24 hr old, although MRI is generally preferred to avoid exposure to ionizing radiation.

11.(E) MRI with and without gadolinium is generally preferred because it provides a more detailed view of the anatomy without exposure to ionizing radiation.

12.(A) Proton MR spectroscopy (MRS) is a molecular imaging technique in which the unique neurochemical profile of a preselected brain region is displayed in the form of a spectrum. Many metabolites can be detected, the most common of which are N -acetylaspartate, creatine and phosphocreatine, choline, myoinositol, and lactate. Changes in the spectral pattern of a given area can yield clues to the underlying pathology, making MRS useful in the diagnosis of inborn errors of metabolism, as well as the preoperative and post therapeutic assessment of intracranial tumors. MRS can also detect areas of cortical dysplasia in patients with epilepsy, because these patients have low N–acetylaspartate:creatine ratios.

13.(E) EEG abnormalities can be divided into two general categories: epileptiform discharges and slowing. Epileptiform discharges are paroxysmal spikes or sharp waves, often followed by slow waves, which interrupt the background activity. They may be focal, multifocal, or generalized. Focal discharges are often associated with cerebral dysgenesis or irritative lesions,

such as cysts, slow growing tumors, or glial scar tissue; generalized discharges typically occur in children with structurally normal brains. Generalized discharges can occur as an epilepsy trait in children who have never had a seizure and, by themselves, are not an indication for treatment.

14.(A) Occult spinal dysraphism is often associated with more significant developmental abnormalities of the spinal cord, including syringomyelia, diastematomyelia, lipoma, fatty filum, dermal sinus, and/or a tethered cord. A spine x-ray in these cases might show bone defects or may be normal. All cases of occult spinal dysraphism are best investigated with MRI. Initial screening in the neonate may include ultrasonography, but MRI is more accurate at any age.

15.(B) The risk of recurrence after one affected child is 3–4% and increases to 10% with two prior affected children.

16.(C) To be effective, folic acid supplementation should be initiated before conception and continued until at least the 12th wk of gestation, when neurulation is complete. The mechanisms by which folic acid prevents NTDs remain poorly understood.

17.(D) The United States Public Health Service recommends that all women of childbearing age who can become pregnant take 0.4 mg of folic acid daily. If, however, a pregnancy is planned in high-risk women (previously affected child), supplementation should be started with 4 mg of folic acid daily, beginning 1 mo before the time of the planned conception.

18.(A)

19.(B) Aicardi syndrome represents a complex disorder that affects many systems and is typically associated with agenesis of the corpus callosum, distinctive chorioretinal lacunae, and infantile spasms. Patients are almost all female, suggesting a genetic abnormality of the X chromosome (it may be lethal in males during fetal life). Seizures become evident during the first few months and are typically resistant to anticonvulsants. An electroencephalogram shows independent activity recorded from both hemispheres as a result of the absent corpus callosum and often shows hemihypsarrhythmia. All patients have severe intellectual disability and can have abnormal vertebrae that may be fused or only partially developed (hemivertebra). Abnormalities of the retina, including circumscribed pits or lacunae and coloboma of the optic disc, are the most characteristic findings of Aicardi syndrome.

20.(A) If the cause of the microcephaly is unknown, the mother's serum phenylalanine level should be determined. High phenylalanine serum levels in an asymptomatic mother can produce marked brain damage in an otherwise normal nonphenylketonuric infant.

21.(B) The total volume of CSF approximates 50 mL in an infant and 150 mL in an adult.

22.(B) Nonobstructive or communicating hydrocephalus most commonly follows a subarachnoid hemorrhage, which is usually a result of intraventricular hemorrhage in a premature infant. Blood in the subarachnoid spaces can cause obliteration of the cisterns or arachnoid villi and obstruction of CSF flow.

23.(A) The Dandy-Walker malformation consists of a cystic expansion of the fourth ventricle in the posterior fossa and midline cerebellar hypoplasia, which results from a developmental failure of the roof of the fourth ventricle during embryogenesis. Infants present with a rapid increase in head size and a prominent occiput.

24.(E) Sotos syndrome (cerebral gigantism) is the most common megalencephalic syndrome, with 50% of patients having prenatal macrocephaly and 100% of patients having macrocephaly by age 1 yr. Early postnatal overgrowth normalizes by adulthood.

25.(C) Benign familial megalencephaly is easily diagnosed by a careful family history and measurement of the parents' head circumferences (occipitofrontal circumferences).

26.(C) Premature closure of the sagittal suture produces a long and narrow skull, or scaphocephaly, the most common form of craniosynostosis. Scaphocephaly is associated with a prominent occiput, a broad forehead, and a small or absent anterior fontanel. The condition is sporadic, is more common in males, and often causes difficulties during labor because of cephalopelvic disproportion. Scaphocephaly does not produce increased ICP or hydrocephalus, and results of neurologic examination of affected patients are normal.

27.(D) Trigonocephaly is a rare form of craniosynostosis caused by premature fusion of the metopic suture. These children have a keel-shaped forehead and hypotelorism and are at risk for associated developmental abnormalities of the forebrain. Milder forms of metopic ridging are more common.

28.(E) Crouzon syndrome is characterized by premature craniosynostosis and is inherited as an autosomal dominant trait. The shape of the head depends on the timing and order of suture fusion but most often is a compressed back-to-front diameter or brachycephaly resulting from bilateral closure of the coronal sutures. The orbits are underdeveloped, and ocular proptosis is prominent. Hypoplasia of the maxilla and orbital hypertelorism are typical facial features.

29.(E) Factors That Increase the Risk for Deformational Plagiocephaly

- ✓ Male

- ✓ First-born child
- ✓ Prematurity
- ✓ Limited passive neck rotation at birth (e.g., congenital torticollis)
- ✓ Developmental delay
- ✓ Sleep position is supine at birth and at 6 wk
- ✓ Bottle feeding only
- ✓ Tummy time < 3 times/day
- ✓ Lower activity level, slower milestone achievement
- ✓ Sleeping with head to same side, positional preference

30.(D) Infants with severe deformational plagiocephaly DP should be considered for helmet therapy at any age. Studies suggest helmet therapy should be started for significant DP between 4 and 8 mo and continued for 7-8 mo. Parents should be counseled on the commitment involved in this treatment because helmets need to be worn up to 23 hr per day Noncompliance has been documented in 80% of study patient populations in as little as 4 mo.

31.(E)

MAJOR

- ✓ Age < 1 yr
- ✓ Duration of fever < 24 hr
- ✓ Fever 38-39°C (100.4-102.2°F)

MINOR

- ✓ Family history of febrile seizures
- ✓ Family history of epilepsy
- ✓ Complex febrile seizure
- ✓ Daycare
- ✓ Male gender
- ✓ Lower serum sodium at time of presentation

32.(E)

- ✓ Simple febrile seizure 1%
- ✓ Recurrent febrile seizures 4%
- ✓ Complex febrile seizures (>15 min in duration or recurrent within 24hr) 6%
- ✓ Fever < 1 hr before febrile seizure 11%
- ✓ Family history of epilepsy 18%
- ✓ Complex febrile seizures (focal) 29%
- ✓ Neurodevelopmental abnormalities 33%

33.(B) The majority of patients who had prolonged febrile seizures and encephalopathy after vaccination and who had been presumed to have suffered

from vaccine encephalopathy (seizures and psychomotor regression occurring after vaccination and presumed to be caused by it) turn out to have Dravet syndrome mutations, indicating that their disease is caused by the mutation and not secondary to the vaccine. This has raised doubts about the very existence of the entity termed vaccine encephalopathy.

34.(E) Febrile seizures often occur in the context of otitis media; roseola and human herpesvirus (HHV) 6 infections; and infections with norovirus, enteroviruses, Shigella, or similar agents, making the evaluation more demanding. In patients with febrile status epilepticus, HHV-6B (more frequently) and HHV-7 infections account for 30% of the cases.

35.(B) Meningitis should be considered in the differential diagnosis, and lumbar puncture should be performed for all infants younger than 6 mo of age who present with fever and seizure, if the child is ill-appearing, or at any age if there are clinical signs or symptoms of concern. A lumbar puncture is an option in a child 6-12 mo of age who is deficient in Haemophilus influenzae type b and Streptococcus pneumoniae immunizations or for whom the immunization status is unknown. A lumbar puncture is an option in children who have been pretreated with antibiotics.

36.(A) If the child is presenting with the first simple febrile seizure and is otherwise neurologically healthy, an EEG need not be performed as part of the evaluation. An EEG would not predict the future recurrence of febrile seizures or epilepsy even if the result is abnormal. Spikes during drowsiness are often seen in children with febrile seizures, particularly those older than age 4 yr, and these do not predict later epilepsy. EEGs performed within 2 wk of a febrile seizure often have nonspecific slowing, usually posteriorly. Thus, in many cases, if an EEG is indicated, it is delayed until or repeated after more than 2 wk have passed. An EEG should, therefore, generally be restricted to special cases in which epilepsy is highly suspected, and generally, it should be used to delineate the type of epilepsy rather than to predict its occurrence.

37.(E) Blood studies (serum electrolytes, calcium, phosphorus, magnesium, and a complete blood count) are not routinely recommended in the workup of a child with a first simple febrile seizure. Blood glucose should be measured initially and with prolonged postictal obtundation or with poor oral intake (prolonged fasting).

38.(A) Iron deficiency is associated with an increased risk of febrile seizures, and thus screening for that problem and treating it appears appropriate.

39.(B) Benign childhood epilepsy with centrotemporal spikes (BECTS), which typically starts during childhood (ages 3-10 yr) and is outgrown by adolescence.

The child typically wakes up at night due to a focal seizure with preserved awareness causing buccal and throat tingling and tonic or clonic contractions of one side of the face, with drooling and inability to speak but with preserved consciousness and comprehension. Focal seizures with impaired awareness and secondary generalized seizures can also occur. EEG shows typical wide-based centrotemporal spikes that are markedly increased in frequency during drowsiness and sleep. MRI is normal. Patients respond very well to antiepileptic drugs (AEDs) such as oxcarbazepine and carbamazepine. In some patients who only have rare and mild seizures, treatment might not be needed.

40.(B) Typical absence seizures usually start at 5-8 yr of age and are often, due to their brevity, overlooked by parents for many months even though they can occur up to hundreds of times per day. Unlike focal seizures with impaired awareness they do not have an aura, usually last for only a few seconds, and are accompanied by eyelid flutter or upward rolling of the eyes but typically not by the usually more florid automatisms seen in focal seizures with impaired awareness (absence seizures can have simple automatisms such as lip smacking or picking at clothing, and the head can very minimally fall forward). Absence seizures do not have a postictal period and are characterized by immediate resumption of what the patient was doing before the seizure. Hyperventilation for 3-5 min can precipitate the seizures and the accompanying 3-Hz spike-and–slow-wave discharges.

41.(D) Patients with cryptogenic West syndrome have normal development before onset, whereas patients with symptomatic West syndrome have preceding developmental delay owing to perinatal encephalopathies, malformations, underlying metabolic disorders, or other etiologies. In males, West syndrome can also be caused by ARX gene mutations (often associated with ambiguous genitalia and cortical migration abnormalities). West syndrome, especially in cases of unknown etiology (cryptogenic cases, i.e., cases that are not symptomatic of a metabolic or structural brain disorder), is a medical emergency because a delay in diagnosis of 3 wk or longer can affect the long-term prognosis. The spasms are often overlooked by parents and by physicians, being mistaken for startles caused by colic or other benign paroxysmal syndromes

42.(E) Lennox-Gastaut syndrome typically starts between the ages of 2 and 10 yr and consists of a triad of developmental delay, multiple seizure types that as a rule include atypical absences, and myoclonic, astatic, and tonic seizures, as well as specific EEG abnormalities. The tonic seizures occur either in wakefulness (causing falls and injuries) or also, typically, in sleep. The third

component is the EEG findings: 1- to 2-Hz spike and slow waves, polyspike bursts in sleep, and a slow background in wakefulness. Patients commonly have tonic, myoclonic, atonic, and other seizure types causing falls and are difficult to control. Most are left with long-term cognitive impairment and intractable seizures despite multiple therapies. Some, but not all, patients start with Ohtahara syndrome, develop West syndrome, and then progress to Lennox-Gastaut syndrome.

43.(A) Pyridoxine dependent epilepsy typically presents with a neonatal or infantile (and rarely childhood) onset of encephalopathy with, at times, reports of increased fetal movements (seizures) in utero. There are recurrent focal motor seizures, generalized tonic seizures, and myoclonus. Seizures progress to status epilepticus if no pyridoxine is used. Diagnosis is confirmed by the presence of elevated plasma, urine, and CSF α-aminoadipic semialdehyde and elevated plasma and CSF pipecolic acid levels. The presence of either homozygous or compound heterozygous mutations in ALDH7A1 alleles (which encode the protein antiquitin) confirms the diagnosis. The use of pyridoxine 100 mg daily orally (higher doses, up to 500-600 mg/day, have been used) or intravenously helps stop the seizures. Mutations of the PROSC gene can also cause pyridoxine dependent epilepsy.

44.(A) In general, the drugs of first choice for focal seizures and epilepsies are oxcarbazepine and levetiracetam.

45.(C) Absence seizures are most often initially treated with ethosuximide, which is as effective as, but less toxic than, valproate; both are more effective than lamotrigine (which has fewer side effects than valproate).

46.(A) Benzodiazepines side effects:

- ✓ **Nuisance:** dose-related neurotoxicity (drowsiness, sedation, ataxia), hyperactivity, drooling, increased secretions
- ✓ **Serious:** apnea

47.(C) Vigabatrin side effects:

- ✓ **Nuisance:** hyperactivity
- ✓ **Serious:** irreversible visual field deficits, retinopathy that requires frequent ophthalmologic evaluations and follow-up

48.(B) Carbamazepine side effects:

- ✓ **Nuisance:** tics, transient leukopenia; hyponatremia, weight gain, nausea; dizziness
- ✓ **Serious:** Stevens-Johnson syndrome, agranulocytosis, aplastic anemia, liver toxicity

49.(B) Valproic acid side effects:

- ✓ **Nuisance:** weight gain; hyperammonemia tremor, alopecia, menstrual
- ✓ irregularities
- ✓ **Serious:** hepatic and pancreatic toxicity

50.(B) The presence of migraine in a patient with epilepsy can lead to the choice of a medication that is effective against both conditions, such as valproate, topiramate, or zonisamide.

51.(A) In an obese patient, a medication such as valproate might be avoided, and a medication that decreases the appetite, such as topiramate or zonisamide, might be used instead.

52.(B) In a patient with both absence and generalized tonic-clonic seizures, a drug that has a broad spectrum of anti-seizure effects, such as lamotrigine or valproate, could be used rather than medications that have a narrow spectrum of efficacy, such as phenytoin and ethosuximide.

53.(A) Based on available evidence, levetiracetam and lamotrigine are FDA pregnancy category C drugs and probably the safest AEDs to use during pregnancy.

54.(C) One exception requiring frequent (even weekly) monitoring of liver function and blood counts throughout the therapy is felbamate, owing to the high incidence of liver and hematologic toxicity (1 in 500 children under 2 yr of age with complex neurologic disorders who are taking the drug).

55.(E) Other potential side effects are rickets from phenytoin, phenobarbital, primidone, and carbamazepine (enzyme inducers that reduce the 25-hyrdroxyvitamin D level by inducing its metabolism).

56.(A) The diet is absolutely contraindicated in carnitine deficiency (primary); carnitine palmitoyltransferase I or II deficiency; carnitine translocase deficiency; β-oxidation defects; medium-chain acyl dehydrogenase deficiency; long-chain acyl dehydrogenase deficiency; short-chain acyl dehydrogenase deficiency; long-chain 3-hydroxyacyl-coenzyme A deficiency; medium-chain 3- hydroxyacyl–coenzyme A deficiency; pyruvate carboxylase deficiency; and porphyrias. Thus, an appropriate metabolic workup, depending on the clinical picture, usually needs to be performed before starting the diet (e.g., acyl carnitine profile, total and free carnitine levels).

57.(B) Discontinuation of AEDs is usually indicated when children are free of seizures for at least 2 yr. In more severe syndromes, such as temporal lobe epilepsy secondary to mesial temporal sclerosis, Lennox-Gastaut syndrome, or severe myoclonic epilepsy, a prolonged period of seizure freedom with treatment is often warranted before AEDs are withdrawn, if withdrawal is

attempted at all. In self-limited (benign) epilepsy syndromes, the duration of therapy can often be as short as 6 mo.

58.(C) AED therapy should be discontinued gradually; often over a period of 3-6 months. Abrupt discontinuation can result in withdrawal seizures or in status epilepticus.

59.(E) There are five main neonatal seizure types: subtle, clonic, tonic, spasms, and myoclonic. Spasms, focal clonic, focal tonic, and generalized myoclonic seizures are, as a rule, associated with electrographic discharges (epileptic seizures), whereas motor automatisms and subtle, generalized tonic, and multifocal myoclonic episodes are frequently not associated with discharges and thus are thought to often represent release phenomena secondary to brain injury rather than true epileptic seizures.

60.(B) Hypoxic-ischemic encephalopathy is the most common cause of neonatal seizures, accounting for 50–60% of patients. Seizures secondary to this encephalopathy occur within 12 hr of birth.

61.(C) Hypomagnesemia (<1.5 mg/dL) is often associated with hypocalcemia and occurs particularly in infants of malnourished mothers. In this situation, the seizures are resistant to calcium therapy but respond to intramuscular magnesium, 0.2 mL/kg of a 50% solution of MgSO4.

62.(B) Phenobarbital is considered by many as the first-choice long-acting drug in neonatal seizures. Whether to use a benzodiazepine first depends on the clinical situation. The usual loading dose is 20 mg/kg. If this dosage is not effective, then additional doses of 5-10 mg/kg can be given until a cumulative dose of 40 mg/kg is reached.

63.(B) In general, if the EEG before the time of discharge does not show evidence of epileptiform activity, medications are usually tapered at that time. If the EEG remains paroxysmal, the decision is usually delayed until several months after discharge.

64.(B)

65.(B) The American Epilepsy Society SE Guidelines recommend using either intravenous lorazepam, intravenous diazepam, or intramuscular midazolam as a first-line agent. Less evidence supports the use of phenytoin/fosphenytoin, phenobarbital, valproate, or levetiracetam as alternative first-line agents. Additionally, in some infants, a trial of pyridoxine may be warranted.

66.(B) Education and reassurance of the parents is usually all that is needed because these episodes are, as a rule, self-limited and are outgrown within a few years. However, screening for anemia and for electrical cardiac disturbances with an electrocardiogram is recommended because the spells are

worsened by iron-deficiency anemia and can rarely be the presenting sign of long-QT syndromes.

67.(A) Education and reassurance of the parents is usually all that is needed because these episodes are, as a rule, self-limited and are outgrown within a few years.

68.(B) Vasovagal (neurocardiogenic) syncope is usually triggered by dehydration, heat, standing for a long time without movement, hot showers, the sight of blood, pain, swallowing, vomiting, sudden exposure to cold as with cold water immersion, and a sudden episode of stress. The history is usually the clue to distinguishing syncope from epileptic seizures: there is initially pallor and sweating followed by blurring of vision, dizziness, and nausea and then a gradual collapse with loss of consciousness.

69.(C) Benign paroxysmal vertigo of childhood is a common migraine equivalent that consists of brief seconds-to-minutes episodes of vertigo that is often accompanied by postural imbalance and nystagmus. It is important to note that vertigo does not always refer to a spinning motion; it can also refer to a backward or forward motion (vertigo titubans) where children sometimes report that objects seem to be moving toward them. The child appears frightened during the episode. Diaphoresis, nausea, vomiting, and, rarely, tinnitus may be present. Episodes usually remit by 6 yr of age. MRIs and EEGs are normal, but caloric testing, if done, can show abnormal vestibular function.
Diphenhydramine, 5 mg/kg/day (maximum of 300 mg/day) may be used for a cluster of attacks. Preventive therapy with cyproheptadine may rarely be needed for frequent attacks.

70.(D) Gastroesophageal reflux in infants may cause paroxysmal episodes of generalized stiffening and opisthotonic posturing that may be accompanied by apnea, staring, and minimal jerking of the extremities. Episodes often occur 30 min after a feed. In older children, this syndrome manifests with episodic dystonic or dyskinetic movements consisting of laterocollis, retrocollis, or torticollis, the exact pathophysiology of which remains elusive.

71.(C) Infantile head atonic attacks consist of repeated head drops, hundreds to thousands per day, usually appearing at 3-6 mo of life and spontaneously subsiding by the 1st yr of life, without concurrent EEG epileptic activity.

72.(D) Pleasurable behaviors similar to masturbation may occur from infancy onward, and may consist of rhythmic rocking movements in the sitting or lying position or rhythmic hip flexion and adduction. Infantile gratification (masturbation), which is more common in girls, usually occurs at 2-3 yr of age and is often associated with perspiration, irregular breathing, and grunting, but

no loss of consciousness. Occasionally this is associated with child abuse or with other psychopathology.

73.(B) Back pain related to the level of the tumor is a common presenting complaint. It is likely that this pain will awaken the child from sleep and improve as the day progresses.

74.(C) Some prolonged headaches may still be migraine, but a migraine that persists beyond 72 hr is classified as a variant termed status migrainosus.

75.(C) The most frequent symptom is chronic (weeks-to-months), progressive, frontal headache that may worsen with postural changes or a Valsalva maneuver.

76.(E) Ibuprofen has been well documented to be effective at a dose of 7.5-10.0 mg/kg and is often preferred; however, acetaminophen (15 mg/kg) can be effective in those with a contraindication to NSAIDs.

77.(C) Migraines are typically moderate to severe, are focal in location, are worsened by physical activity or limit physical activity, and have a throbbing quality, Tension-type headaches TTHs are mild to moderate in severity, are diffuse in location, are not affected by activity (although the patient may not feel like being active), and are nonthrobbing (often described as a constant pressure). TTH is much less frequently associated with nausea, photophobia, or phonophobia and is never associated with more than one of these at a time or with vomiting.

78.(E)

79.(A)

Bilateral vestibular schwannomas 90–95%
Other cranial nerve schwannomas 24–51%
Intracranial meningiomas 45–58%
Spinal tumors 63–90%
Extramedullary 55–90%
Intramedullary 18–53%
Peripheral neuropathy Up to 66%

80.(D)

Minor Features of Tuberous Sclerosis Complex

Dental enamel pits (>3)
Intraoral fibromas (≥2)
Retinal achromic patch
Confetti skin lesions
Nonrenal hamartomas
Multiple renal cysts

81.(A) The most common neurologic manifestations of tuberous sclerosis complex TSC consist of epilepsy, cognitive impairment, and autism spectrum disorder. TSC may present during infancy with infantile spasms and a hypsarrhythmic electroencephalogram pattern.

82.(B) Vigabatrin is the first-line therapy for infantile spasms. Adrenocortico-tropic hormone (ACTH) can be used if treatment with vigabatrin fails.

83.(B) Approximately 50% of children with TSC have cardiac rhabdomyomas, which may be detected in the fetus by an echocardiogram, usually by 20-30 wk gestation. The rhabdomyomas may be numerous and located throughout the ventricular myocardium, and although they can cause congestive heart failure and arrhythmias in a minority of patients, they tend to slowly resolve spontaneously.

84.(B)

85.(B)

86.(E) Joubert syndrome and related disorders are autosomal recessive disorders marked by developmental delay, hypotonia, abnormal eye movements, abnormal respirations, and a distinctive malformation of the cerebellum and brainstem that manifests as the “molar tooth sign” on axial MRI.

87.(D) Acute cerebellar ataxia occurs primarily in children 1-3 yr of age and is a diagnosis of exclusion. The condition often follows a viral illness, such as varicella virus, coxsackievirus, or echovirus infection, by 2-3 wk. It is thought to represent an autoimmune response to the viral agent affecting the cerebellum.

88.(A) Abetalipoproteinemia (Bassen-Kornzweig disease) is an autosomal recessive disorder caused by a mutation in the microsomal triglyceride transfer protein (MTP). This disorder begins in childhood with steatorrhea and failure to thrive. A blood smears shows acanthocytosis, which consists of spiculated red blood cells. Serum chemistries reveal decreased levels of cholesterol and triglycerides and absent serum β-lipoproteins. Neurologic signs become evident by late childhood and consist of ataxia, retinitis pigmentosa, peripheral neuritis, abnormalities of position and vibration sense, muscle weakness, and intellectual disability.

89.(C)

90.(B) Sydenham chorea (St. Vitus dance) is the most common acquired chorea of childhood. It occurs in 10–20% of patients with acute rheumatic fever, typically weeks to months after a group A β-hemolytic streptococcal infection. Peak incidence is at age 8-9 yr, with a female predominance of 2:1.

91.(B) The following nutritional disorders can cause secondary intracranial hypertension without an obstructive lesion on MRI;

- ✓ Hypovitaminosis A
- ✓ Vitamin A intoxication
- ✓ Hyperalimentation in malnourished patient
- ✓ Vitamin D–dependent rickets

92.(B) Benign neonatal sleep myoclonus begins during the 1st week of life, diminishes in the 2nd mo, and is usually gone before 6 months of age. The movements are most likely to occur during quiet (non-REM) sleep, but have been described in all sleep stages.

93.(A) Spastic diplegia is bilateral spasticity of the legs that is greater than in the arms. Spastic diplegia is strongly associated with damage to the immature white matter during the vulnerable period of immature oligodendroglia between 20-34 wk of gestation.

94.(D) Seizures are the most common clinical manifestation of burn encephalopathy, but altered states of consciousness, hallucinations, and coma may also occur.

95.(A) Anti-N -Methyl-D-Aspartate Receptor Encephalitis is considered the second most common cause of autoimmune encephalitis after acute disseminated encephalomyelitis in children and adolescents.

96.(C) Rasmussen encephalitis is an inflammatory encephalopathy characterized by progressive refractory focal seizures, cognitive deterioration, and focal neurologic deficits that occur with gradual atrophy of one brain hemisphere. The disorder frequently presents in children 6-8 yr old, although adolescents and adults can be affected.

97.(E) The most effective treatment for control of the seizures is functional hemispherectomy, which consists of surgical disconnection of the affected hemisphere.

98.(A) The diagnosis of Sandhoff disease is established by finding deficient levels of hexosaminidases A and B in serum and leukocytes. Children usually die by 3 yr of age. Sandhoff disease is caused by mutations in the HEXB gene located on chromosome 5q13.

99.(D)

100.(E)

101.(B)

102.(E) Encephalopathy is the hallmark of ADEM, ranging from changes in behavior and persistent irritability to coma.

103.(A) The following drugs can cause secondary intracranial hypertension without an obstructive lesion on MRI; Tetracyclines, Sulfonamides, Nalidixic acid, Fluoroquinolones, Corticosteroid therapy and withdrawal, Nitrofurantoin, Cytarabine, Cyclosporine, Phenytoin, Mesalamine, Isotretinoin, Amiodarone, Oral contraceptive pills/implants.

104.(E) The typical presentation of optic neuritis is unilateral or bilateral visual loss over hours to days, abnormal color vision, visual field loss, and sometimes a relative afferent pupillary defect. The visual loss can be quite severe, with the majority of children at 20/200 VA or worse. Periocular pain or pain with eye movement and, at times, a headache are common features.

105.(D) Susac syndrome characterized by all mentioned features with the following MRI findings (Focal and small lesions in supratentorial and infratentorial regions (both white matter and gray matter); involvement of corpus callosum (snowball lesions); leptomeningeal enhancement).

106.(C) Diffusion-weighted MRI demonstrates arterial ischemic stroke (AIS) from minutes to 7 days following the onset; MR angiography can confirm vascular occlusion and suggest possible arteriopathy. Diffusion-weighted MRI can also demonstrate wallerian degeneration in the descending corticospinal tract, which correlates with chronic hemiparesis.

107.(A) Hand dominance within the first year of life is abnormal and may be the result of perinatal stroke.

108.(C) Arteriovenous malformations are the most common cause of childhood subarachnoid and intraparenchymal hemorrhagic stroke and may occur anywhere.

109.(A) CSF protein (mg/dL) in tuberculous meningitis is 100-3,000; may be higher in presence of block.

110.(C) CSF glucose generally normal; may be decreased to < 40 in some viral diseases, particularly mumps (15–20% of cases).

111.(D) The risk of pneumococcal bacterial meningitis was historically increased by more than 30-fold in children with cochlear implants, though advances in implant design have reduced this risk.

112.(A) Although the incidence of pneumococcal meningitis has been reduced, S. pneumoniae remains the most frequently identified pathogen of bacterial meningitis in the United States and in other countries that have adopted similar vaccination strategies.

113.(E) The predominant organisms that cause brain abscesses are streptococci, which account for one third of all cases in children, with members of the

Streptococcus anginosus group (S. anginosus, Streptococcus constellatus, and Streptococcus intermedius) being the most common streptococci.

114.(D) Blood cultures should be performed in all patients with suspected meningitis. Blood cultures reveal the responsible bacteria in up to 80–90% of cases of meningitis. Elevations of the C-reactive protein, erythrocyte sedimentation rate, and procalcitonin can be seen in both bacterial and viral meningitis and should not be used to routinely determine which patients should receive antimicrobials.

115.(A) Currently, the recommended treatment duration for uncomplicated S. pneumoniae meningitis is 10-14 days with a 3rd-generation cephalosporin or intravenous penicillin (300,000-400,000 units/kg/day, divided every 4-6 hr) used for penicillin-sensitive isolates, or vancomycin if the isolate is resistant to penicillins and cephalosporins. For N. meningitidis meningitis, the recommended treatment duration is 5-7 days with intravenous penicillin (300,000 units/kg/day) for strains with a minimum inhibitory concentration (MIC) of penicillin < 0.1 μg/mL, or ceftriaxone for strains with an MIC of 0.1-1 μg/mL. Uncomplicated H. influenzae type b meningitis should be treated for 7-10 days with ampicillin for β-lactamase–negative strains, or a 3rd-generation cephalosporin for β-lactamase–positive isolates. Gram-negative bacillary meningitis should be treated for 3 wk or at least 2 wk after CSF sterilization, which may occur after 2- 10 days of treatment.

116. (C) In a Cochrane review of using steroids in meningitis treatment, steroids reduced hearing loss in children with meningitis due to H. influenzae type b but not due to other pathogens. The use of steroids in children did not reduce mortality rates?

117.(A) Sensorineural hearing loss is the most common sequela of bacterial meningitis and, usually, is already present at the time of initial presentation. It is a result of cochlear or auditory nerve inflammation and occurs in as many as 30% of patients with pneumococcal meningitis, 10% with meningococcal meningitis, and 5–20% of those with H. influenzae type b meningitis. All patients with bacterial meningitis should undergo careful audiologic assessment before or soon after discharge from the hospital. Frequent reassessment on an outpatient basis is indicated for patients who develop a hearing deficit.

118.(B) The most common manifestation of CNS involvement is cerebellar ataxia and the most severe is acute encephalitis.

119.(C) Mumps meningoencephalitis is typically mild, but deafness from damage of the 8^{th} cranial nerve can occur.

120.(B)

Chapter 27

Neuromuscular Disorders Questions

ZUHAIR ALMUSAWI

1. A 9-month-old floppy infant with bulbar and oculomotor muscles involvement.
Which of the following is the **MOST** likely cause?

A. Chromosomal disorder
B. Spinal muscular atrophy
C. Peripheral neuropathy
D. Myasthenia syndrome
E. Congenital myotonic dystrophy

2. A 6-month-old floppy infant presents with generalized weakness which spares the diaphragm, facial muscles, and sphincters.
Which of the following is the **MOST** likely cause?

A. Chromosomal disorder
B. Spinal muscular atrophy
C. Peripheral neuropathy
D. Myasthenia syndrome
E. Congenital myotonic dystrophy

3. Ptosis is a prominent feature in which of the following congenital myopathies?

A. Central core disease
B. Nemaline myopathy
C. Myotubular myopathy
D. Congenital fiber type disproportion
E. Muscular dystrophy type C1C

4. A 2-year-old child presents with generalized hypotonia, weakness, very thin muscle mass, and arthrogryposis. The head is dolichocephalic with high arched and opened mouth. The mother reported decreased fetal movements.
The serum CK level is normal but the muscle biopsy was diagnostic.

Of the following, the **MOST** likely diagnosis is

A. congenital muscle fiber–type disproportion
B. central core disease
C. Nemaline rod myopathy
D. myotubular myopathy
E. centronuclear myopathy

5. A 19-month-old child presents with hypotonia, joint laxity, motor developmental delay, and hip girdle weakness. He can sit without support, but unable to stand on his feet, he has recurrent shoulder dislocations, and congenital hip dysplasia. His father developed malignant hyperthermia with anesthesia for repair of inguinal hernia.

Of the following, the **MOST** likely diagnosis is

A. congenital muscle fiber–type disproportion
B. central core disease
C. Nemaline rod myopathy
D. myotubular myopathy
E. centronuclear myopathy

6. Central core myopathy is consistently associated with malignant hyperthermia which is treated with

A. dantrolene
B. salbutamol
C. albuterol
D. N-acetylcysteine
E. correction of a specific gene defect

7. Which of the following muscles is **COMMONLY** involved in amyoplasia?

A. Palmaris longus
B. Sternocleidomastoid muscle
C. Pectoralis major muscle
D. Flexor carpi radialis
E. Deltoid muscle

8. In Duchenne muscular dystrophy, Gowers' sign is nearly always evident by age of

A. 1 year
B. 3 year

C. 5 year
D. 7 year
E. 9 year

9. After the calves, what is the next **MOST** common site of muscular hypertrophy in Duchenne muscular dystrophy?
A. Tongue
B. Forearm
C. Neck
D. Shoulder
E. Hip

10. Initiation of steroids in Duchenne muscular dystrophy is indicated when a child shows
A. plateau in development
B. severe regression in motor development
C. cardiomyopathy
D. scoliosis
E. increased CK even if asymptomatic

11. A 3-year-old child presented with narrow head, high arched palate, inverted-V–shaped upper lip, thin cheeks, and scalloped concave temporalis muscles with progressive wasting of distal muscles and flattened thenar and hypothenar eminences. The child has difficulty with climbing stairs and the Gowers' sign is positive.
Of the following, the **MOST** likely diagnosis is
A. Duchenne muscular dystrophy
B. Classic myotonic dystrophy (type 1)
C. Emery-Dreifuss muscular dystrophy
D. Becker muscular dystrophy
E. Nemaline myopathy

12. Which of the following is characterized by grip myotonia?
A. Duchenne muscular dystrophy
B. Classic myotonic dystrophy (type 1)
C. Emery-Dreifuss muscular dystrophy
D. Becker muscular dystrophy
E. Nemaline myopathy

13. Which of the following endocrine abnormalities is common in myotonic dystrophy?

A. Hyperthyroidism
B. Adrenocortical insufficiency
C. Testicular atrophy
D. Ovarian atrophy
E. Hypothyroidism

14. Which of the following is characterized by weakness and generalized muscular hypertrophy so that affected children resemble bodybuilders (Herculean appearance)?

A. Generalized myotonia of Becker
B. Paramyotonia congenita
C. Myotonia congenita (Thomsen disease)
D. Calcium channel periodic paralysis
E. Sodium channel periodic paralysis

15. Which of the following is the first line treatment for the nondystrophic myotonias?

A. Carbamazepine
B. Phenytoin
C. Gabapentin
D. Acetazolamide
E. Mexiletine

16. Which of the following can cause rhabdomyolysis and myoglobinuria?

A. Statins
B. Colchicine
C. Labetalol
D. Cyclosporine
E. Vincristine

17. Which of the following can cause hyperkalemic periodic paralysis?

A. Addison disease
B. Thyrotoxicosis
C. Bartter syndrome
D. Laxative abuse
E. Nontropical sprue

18. A 10-year-old child consulted you because of episodic event of inability to move after awakening but gradually recovers muscle strength during the next few minutes or hours. All four extremities are involved and he is normal between attacks.
Of the following, the **MOST** likely diagnosis is

A. Andersen-Tawil syndrome
B. periodic paralysis
C. thyrotoxic periodic paralysis
D. myotonia congenita
E. paramyotonia congenital

19. A 6-month-old boy presented with poor feeding, hypotonia, respiratory distress, and hepatomegaly. Chest x-ray revealed cardiomegaly, the serum CK level was greatly elevated, and the ECG showed short P-R interval with wide QRS complex.
Of the following, the **MOST** likely diagnosis is

A. Andersen disease
B. McArdle disease
C. Tarui disease
D. Pompe disease
E. Cori-Forbes disease

20. A 10-year-old boy develops muscular cramps, weakness, and myoglobinuria after exercise but strength is normal between attacks. The serum CK level is elevated only during exercise.
Of the following, the **MOST** likely diagnosis is

A. Andersen disease
B. McArdle disease
C. Tarui disease
D. Pompe disease
E. Cori-Forbes disease

21. What is the best screening test for diagnosis of mitochondrial cytopathies?

A. Serum lactate
B. Cerebrospinal fluid lactate
C. Serum 3-methyl-glutaconic acid
D. Hepatic enzymes (transaminases)
E. Coenzyme-Q10

22. A 9-year-old boy present with asymmetric ptosis, some degree of extraocular muscle weakness, diplopia, and preserved pupillary response to light. He has poor head control with progressive weakness involving limb–girdle muscles and distal muscles of the hands. Tendon stretch reflexes are diminished.
Of the following, the **MOST** likely diagnosis is

A. facioscapulohumeral muscular dystrophy
B. spinal muscular atrophy
C. motor neuron disease
D. mitochondrial myopathy
E. myasthenia gravis

23. A 3-year-old myasthenic boy presented with abdominal cramps, diarrhea, profuse sweating, salivation, bradycardia, increased weakness, and miosis for the last two hours.
Of the following, the **MOST** likely cause for this presentation is

A. botulism
B. chronic fatigue syndrome
C. myasthenic crisis
D. organophosphorus poisoning
E. cholinergic crisis

24. Which of the following diagnostic tests is highly specific for myasthenia gravis?

A. Electromyography
B. Muscle biopsy
C. Nerve biopsy
D. Anti-AChR antibodies
E. Electroneurography

25. Which of the following drugs is used as a clinical test for myasthenia gravis?

A. Atropine sulfate
B. Edrophonium chloride
C. Pyridostigmine
D. Neostigmine
E. Propranolol

26. Which of the following antibiotics should be avoided in patients with myasthenia gravis?

A. Cephalosporines
B. Penicillins
C. Macrolids
D. Aminoglycosides
E. Vancomycin

27. A 7-month-old boy infant presented with constipation, poor feeding, and a weak cry. On evaluation, the baby appears hypotonic, with facial weakness, dysphagia, and a poor gag. The baby was exclusive breast feeding and his mother started giving him rice soup with honey, his vaccination is up to date.
Of the following, the **MOST** likely diagnosis is

A. viral encephalitis
B. poliomyelitis
C. infantile botulism
D. spinal muscular atrophy
E. myasthenia gravis

28. What is the common cause of Bell palsy?

A. Herpes simplex virus type 1
B. Epstein-Barr virus
C. Cytomegalovirus
D. Mumps
E. Human herpesvirus 6

29. A 3-month-old boy presented with severe hypotonia, symmetric generalized muscle weakness affecting the lower limbs more than the upper limbs, absence of deep tendon reflexes with evident tongue fasciculations. He looks alert with no involvement of the facial and extraocular muscles.
Of the following, the **MOST** likely diagnosis is

A. Kugelberg-Welander disease
B. poliomyelitis
C. infantile botulism
D. spinal muscular atrophy type I
E. myasthenia gravis

30. A 3-day-old neonate presented with severe muscle weakness, respiratory distress, feeding problems, and generalized arthrogryposis. There is a perception of decreased intrauterine movements by the mother. The baby died after 6 weeks from presentation.
Of the following, the **MOST** likely type of SMA is

A. I
B. II
C. III
D. IV
E. 0

31. A 2-year-old boy presents with gross motor developmental delay, proximal muscle weakness more prominent in the lower extremities. He can sit without support but is unable to walk independently.
Of the following, the MOST likely type of SMA is

A. I
B. II
C. III
D. IV
E. 0

32. What is the most simple and practical tool for diagnosis of patients with spinal muscular atrophy SMA?

A. Electrocardiography
B. Electromyography
C. Nerve conduction study
D. Muscle biopsy
E. Serum creatine kinase

33. What is the **MOST** definitive first-step diagnostic test in a patient with a clinical suspicion of spinal muscular atrophy SMA?

A. Muscle biopsy
B. Genetic marker from blood sample
C. Electromyography
D. Nerve conduction study
E. Serum creatine kinase

34. Which of the following is diagnostic for Charcot-Marie-Tooth disease?

A. Nerve conduction velocities
B. Electromyography
C. Muscle biopsy
D. Serum creatine kinase
E. Sural nerve biopsy

35. A 7- year-old girl presents with intermittent motor and sensory neuropathy, ataxia, progressive neurosensory hearing loss, retinitis pigmentosa with loss of night vision, ichthyosis, liver dysfunction, and skeletal malformations. Motor and sensory nerve conduction velocities are delayed.
Of the following, the **MOST** likely diagnosis is

A. Refsum disease
B. Roussy-Lévy syndrome
C. Déjèrine-Sottas disease
D. Peroneal muscular atrophy
E. Charcot-Marie-Tooth disease

36. A 13-year-old boy presents with recurrent episodes of burning pain and paresthesias of the feet and lower legs so severe that he is unable to walk; these episodes are often precipitated by physical activity. Raised red-black lesions are seen in the buttocks, and periumbilical area.
Of the following, the **MOST** likely diagnosis is

A. Refsum disease
B. Roussy-Lévy syndrome
C. Déjèrine-Sottas disease
D. Fabry disease
E. Charcot-Marie-Tooth disease

37. A 4-year-old child present with nystagmus, progressive ataxia, weakness, dysphagia, dysarthria, optic neuropathy, and kinky hair.
Of the following, the **MOST** likely diagnosis is

A. Hypermyelinating (Tomaculous) Neuropathy
B. Giant axonal neuropathy
C. Refsum disease
D. Roussy-Lévy syndrome
E. Menkes disease

38. Which of the following may cause peripheral neuropathy?

A. Nalidixic acid
B. Sulphamethaxazole
C. Cefixime
D. Amoxicillin
E. Nitrofurantoin

39. A 9-year-old boy presented with fever, cough , shortness of breath diagnosed as pneumonia and treated properly with good response, 10 days later he developed numbness and paresthesia, followed by symmetric weakness in the lower extremities and progressively involving the trunk and the upper limbs with loss of ability to walk.

Of the following, the **MOST** likely diagnosis is

A. acute transverse myelitis
B. poliomyelitis
C. acute disseminated encephalomyelitis
D. Guillain-Barré syndrome
E. acute flaccid myelitis

40. An 8-year-old girl presented with acute external ophthalmoplegia, ataxia, areflexia with involvement of 6th cranial nerve, and very mild lower extremity weakness.

Of the following, the **MOST** likely diagnosis is

A. acute motor axonal neuropathy
B. Bickerstaff brainstem encephalitis
C. Miller-Fisher syndrome
D. polyneuritis cranialis
E. acute transverse myelitis

41. What is the **MOST** sensitive test to detect early signs of peripheral nerve inflammation in Guillain-Barré syndrome?

A. Serum antiganglioside antibodies
B. Nerve conduction study
C. Serologic testing for Campylobacter and Helicobacter infections
D. Serum creatine kinase level
E. CSF study

42. What is the first line treatment for Guillain-Barré syndrome with severe or rapidly progressive muscle weakness?

A. Plasmapheresis
B. Immunosuppressive drugs
C. Intravenous immunoglobulin
D. Steroids
E. Gabapentin

Chapter 27

Neuromuscular Disorders Answers

ZUHAIR ALMUSAWI

1.(D) In Neuromuscular junction diseases (Myasthenia syndromes, Infantile botulism), Bulbar and oculomotor muscles exhibit greater degree of involvement.

2.(B)

3.(C)

4.(C) In the infantile form, generalized hypotonia and weakness, which can include bulbar-innervated and respiratory muscles, and a very thin muscle mass are characteristic. The head is dolichocephalic, and the palate high arched or even cleft. Muscles of the jaw may be too weak to hold it closed. Decreased fetal movements are reported by the mother, and neonates suffer from hypoxia and dysphagia; arthrogryposis may be present.

5.(B) The phenotypical spectrum of core myopathies ranges from mild to severe. Hypotonia, joint laxity, motor developmental delay, hip girdle or axial muscle weakness, orthopedic complications such as recurrent shoulder or patellar dislocations, congenital hip dislocation or dysplasia, or foot deformities may be presenting features.

6.(A) Central core myopathy CCD is consistently associated with malignant hyperthermia (MH), which can precede the diagnosis of CCD. All patients and asymptomatic carriers should be counseled in terms of a potentially fatal adverse reaction to volatile anesthetics and muscle relaxants. Preoperative anesthetic consultation in patients known to be subject to general anesthesia should be considered. Treatment of MH requires dantrolene and additional supportive care measures.

7.(A) Congenital absence of individual muscles is common and is often asymmetric. A common aplasia is the palmaris longus muscle of the ventral forearm, which is absent in 30% of normal subjects and is fully compensated for by other flexors of the wrist.

8.(C) An early Gowers' sign may be seen by age 3 yr, but nearly always is evident by age 5 or 6 yr.

9.(A) After the calves, the next most common site of muscular hypertrophy is the tongue, followed by muscles of the forearm.

10.(A) Initiation of steroids is indicated when a child shows a plateau in development and/or a regression in motor development as compared with peers. This typically occurs by 4-6 yr old. Recommended doses are prednisone 0.75 mg/kg/day or deflazacort 0.9 mg/kg/day.

11.(B) Classic myotonic dystrophy (type 1) (DM1, or Steinert disease) infants can appear almost normal at birth, or facial wasting and hypotonia can already be early expressions of the disease.

12.(B) During physical examination, myotonia may be demonstrated by asking the patient to make tight fists and then to quickly open the hands (grip myotonia).

13.(E) Endocrine abnormalities involve many glands and appear at any time during the course of the disease so that the endocrine status must be reevaluated annually. Hypothyroidism is common; hyperthyroidism occurs rarely. Adrenocortical insufficiency can lead to an addisonian crisis even in infancy. Diabetes mellitus is common in patients with myotonic dystrophy; some children have a disorder of insulin release rather than defective insulin production. The onset of puberty may be precocious or, more often, delayed. Testicular atrophy and testosterone deficiency are common in adults and are responsible for a high incidence of male infertility. Ovarian atrophy is rare. Frontal baldness is also characteristic in male patients and often begins in adolescence.

14.(C) Myotonia congenita (Thomsen disease), a type of channelopathy, is the most common of the nondystrophic myotonia syndromes and is characterized by weakness and generalized muscular hypertrophy so that affected children resemble bodybuilders (Herculean appearance).

15.(E) Treatments for the nondystrophic myotonias include mexiletine as the first line (both for sodium channel and chloride channel myotonias). Mexiletine has been shown to improve stiffness as well as decrease handgrip myotonia.

16.(A) The following can cause rhabdomyolysis and myoglobinuria

Cholesterol-lowering drugs (especially statins)
Alcohol
Heroin
Amphetamine
Toluene
Cocaine
ε-Aminocaproic acid

Pentazocine
Phencyclidine

17.(A) Causes of hyperkalemic periodic paralysis
Addison disease
Hypoaldosteronism
Excessive potassium supplementation
Potassium-sparing diuretics
Chronic renal failure

18.(B) Muscles that remain active in sleep, such as the diaphragm, extraocular muscles (rapid eye movements), and cardiac muscle, are not affected. Patients are normal between attacks, but in adult life the attacks become more frequent, and the disorder causes progressive myopathy with permanent weakness even between attacks. The usual frequency of attacks in childhood is once a week.

19.(D) The infantile form is a severe generalized myopathy and cardiomyopathy. Patients have cardiomegaly and hepatomegaly and are diffusely hypotonic and weak. The serum CK level is greatly elevated. A muscle biopsy specimen reveals a vacuolar myopathy with abnormal lysosomal enzymatic activities such as acid and alkaline phosphatases.

20.(B) Glycogenosis V (McArdle disease) is caused by muscle glycogen phosphorylase deficiency inherited as an autosomal recessive trait at locus 11q13, encoded by the PMGM gene. Exercise intolerance is the cardinal clinical feature. Physical exertion results in cramps, weakness, and myoglobinuria, but is normal between attacks. The serum CK level is elevated only during exercise.

21.(C) Serum 3-methyl-glutaconic acid often is increased in mitochondrial cytopathies in general, demonstrated in more than 50 different genetic mutations, and hence is a good screening measurement; it rarely is increased in other metabolic diseases.

22.(E) In juvenile autoimmune MG, unilateral or bilateral but usually asymmetric ptosis and some degree of extraocular muscle weakness are the earliest and most constant signs. Extraocular weakness is not confined to muscles innervated by just one or two of the three corresponding brainstem nuclei; it is progressive. Older children might complain of diplopia, and young children might hold open their eyes with their fingers or thumbs if the ptosis is severe enough to obstruct vision.

23.(E) Cholinergic crisis secondary to overdosing with anticholinesterase medications. The muscarinic effects include abdominal cramps, diarrhea, profuse sweating, salivation, bradycardia, increased weakness, and miosis.

Cholinergic crisis requires only supportive care and withholding of further doses of cholinergic drugs, and it passes within a few hours; the dose of medication to be restarted should be reconsidered, unless the patient had taken an overdose that was not prescribed.

24.(A) Myasthenia Gravis is one of the few neuromuscular diseases in which electromyography (EMG) is more specifically diagnostic than a muscle or nerve biopsy. A decremental response is seen to repetitive nerve stimulation; the muscle potentials diminish rapidly in amplitude until the muscle becomes refractory to further stimulation.

25.(B) A clinical test for MG is administration of a short-acting cholinesterase inhibitor, usually edrophonium chloride. Ptosis and ophthalmoplegia improve within a few seconds, and the fatigability of other muscles decreases.

26.(D) Certain antibiotics can potentiate myasthenia and should be avoided; these include the aminoglycosides, beta blocking agents, procainamide, chloroquine, and fluoroquinolones.

27.(C) In infantile botulism, which classically presents between the ages of 4 and 7 mo, honey as well as spores from dirt (e.g., near construction sites) are common sources of contamination.

28.(A) Numerous viruses have been linked with Bell palsy. Active or reactivations of herpes simplex or varicella-zoster virus are probably the most common causes of Bell palsy.

29.(D)

30.(E) Infants who are symptomatic prenatally or at birth are classified as having a rare phenotype, SMA type 0 (<1%); they can present with severe muscle weakness, respiratory distress, feeding problems, and cranial nerve involvement. Congenital contractures, ranging from simple clubfoot to generalized arthrogryposis, occur in approximately 10% of severely involved neonates. There is a perception of decreased intrauterine movements by the mother, and these infants usually die within the first months of life.

31.(B) In type II SMA, affected infants are usually able to suck and swallow, and respiration is adequate in early infancy. Developmental delay in gross motor milestones or stagnation of motor development between the ages of 6 and 18 mo is rather typical for this form.

32.(A) Electrocardiography (EKG) may serve as a simple and practical tool in patients with SMA to demonstrate a baseline tremor as an artefact representing muscle fibrillations more prominent on lead II. Although seen in mainly lower motor neuron diseases, including poliomyelitis, recognition of this EKG pattern

may prevent further electrophysiologic tests (electromyography [EMG] and nerve conduction studies [NCSs]) in SMA patients.

33.(B) The simplest, most definitive first-step diagnostic test in a patient with a clinical suspicion of SMA and normal and/or mildly elevated serum CK levels, is a molecular genetic marker in the blood for the homozygous deletion in SMN1.

34.(E) Sural nerve biopsy is diagnostic. Large- and medium-size myelinated fibers are reduced in number, collagen is increased, and characteristic onion bulb formations of proliferated Schwann cell cytoplasm surround axons. This pathologic finding is called interstitial hypertrophic neuropathy.

35.(A)

36.(D) The presentation is in late childhood or adolescence, with recurrent episodes of burning pain and paresthesias of the feet and lower legs so severe that patients are unable to walk. These episodes are often precipitated by fever or by physical activity. Objective sensory and motor deficits are not demonstrated on neurologic examination, and reflexes are preserved. Autonomic nerve involvement is almost universal and may cause cardiac rhythm abnormalities, cutaneous mottling, and gastrointestinal peristaltic abnormalities, but autonomic expression is variable between patients.

Characteristic skin lesions are seen in the perineal region, scrotum, buttocks, and periumbilical zone as flat or raised red-black telangiectasias known as angiokeratoma corporis diffusum. Hypohidrosis may be present. Recurrent strokes result from vascular wall involvement.

37.(B) Giant axonal neuropathy is a rare autosomal recessive disease with onset in early childhood. It is a progressive mixed peripheral neuropathy and degeneration of central white matter, similar to the leukodystrophies. Ataxia and nystagmus are accompanied by signs of progressive peripheral neuropathy. A large majority of affected children have frizzy or kinky hair, which microscopically shows variation in the diameter of the shaft and twisting, similar to that in Menkes disease; hence, microscopic examination of a few scalp hairs provides a simple screening tool in suspected cases.

38.(E) Amiodarone, Chloramphenicol, Chloroquine, Cisplatin, Colchicine, Dapsone, Ethambutol, Ethanol, Fluoroquinolones, Gold, Hydralazine, Isoniazid, Metronidazole, Nitrofurantoin, Nitrous oxide, Nucleosides (antiretroviral agents dideoxycytidine [ddC], didanosine [ddI], d4T, others), Penicillamine, Pentamidine, Phenytoin, Pyridoxine (excessive), Statins, Stilbamidine, Suramin, Tacrolimus, Taxanes (paclitaxel, docetaxel), Thalidomide, Tryptophan (eosinophilia–myalgia syndrome), Vincristine.

39.(D) Initial symptoms include numbness and paresthesia, followed by weakness. Radicular back pain and myalgia are common in the initial stages; affected children can be very irritable. Weakness usually begins in the lower extremities and progressively involves the trunk, the upper limbs, and finally the bulbar muscles, but weakness is sometimes proximally prominent. Extraocular muscle involvement is rare, but many patients develop facial weakness. In most patients, weakness is essentially symmetric. Weakness progresses over days or weeks, the clinical nadir occurring in less than 4 wk. Approximately 60% of children lose the ability to walk at some point in their illness; a small proportion progress to flaccid tetraplegia.

40.(C) Miller-Fisher syndrome (MFS) is an uncommon GBS variant associated with acute external (and occasionally internal) ophthalmoplegia, ataxia, and areflexia. The 6th cranial nerve is most often involved in MFS. Although areflexia is seen in MFS, patients have no or only very mild lower extremity weakness, compared with GBS. Distal paresthesias are common in MFS. Urinary incontinence or retention is a complication in approximately 20% of cases but is usually transient. MFS overlaps clinically with Bickerstaff brainstem encephalitis.

41.(B) Nerve conduction studies and electromyography are sensitive to early signs of peripheral nerve inflammation in GBS. Motor and sensory nerve conduction velocities are reduced to a variable extent, reflecting the patchy nature of nerve involvement in this disorder, which is also reflected in the presence of focal conduction block and dispersed responses. Electromyography may show acute denervation of muscle.

42.(C) Patients with milder weakness and slow progression may be treated expectantly, with observation for stabilization and spontaneous remission. Severe or rapidly progressive muscle weakness is treated with intravenous immunoglobulin (IVIG); common protocols include IVIG 0.4 g/kg/day for 5 consecutive days or 1g/kg/day for 2 days. Plasmapheresis and/or immunosuppressive drugs are alternatives if IVIG is ineffective. Steroids are not effective for weakness but may help with pain.

Chapter 28

Disorders of the Eye Questions

ZUHAIR ALMUSAWI

1. At what age, the cornea in newborns attains the adult size?
   A. 2 years
   B. 4 years
   C. 8 years
   D. 12 years
   E. 16 years

2. At what age, persistent deviation of an eye in an infant requires evaluation?
   A. Birth
   B. 3 months
   C. 6 months
   D. 9 months
   E. 12 months

3. What is the approximate visual acuity in newborns?
   A. 20/20
   B. 20/50
   C. 20/100
   D. 20/200
   E. 20/400

4. At what age, most children attain 20/20 vision?
   A. 2 years
   B. 4 years
   C. 6 years
   D. 8 years
   E. 10 years

5. Which of the following abnormalities of refraction is more common in infants with a history of retinopathy of prematurity?

A. Hyperopia
B. Myopia
C. Astigmatism
D. Anisometropia
E. Amblyopia

6. What is the best way to treat children with amblyopia?
A. Part-time patching
B. Use of atropine drops
C. Full-time occlusion
D. Surgery
E. Watchful observation

7. Which of the following is an important cause of progressive loss of vision in children?
A. Craniopharyngioma
B. Hypertensive encephalopathy
C. Optic neuritis
D. Leukemia
E. Increased intracranial pressure

8. What may be the first clue to amaurosis in an infant?
A. Timidity
B. Clumsiness
C. Strabismus
D. Excessive cry
E. Behavioral change

9. Which of the following is a cause of prominent anisocoria in darkness than light?
A. Third nerve palsy
B. Tonic pupil
C. Pharmacologic mydriasis
D. Disorder of the iris
E. Horner syndrome

10. What is the most common ophthalmic finding in shaken baby syndrome?
A. Ecchymosis of the lids

B. Hemorrhage about the eye
C. Retinal detachment
D. fracture of the orbit
E. Retinal hemorrhage

11. Which term is used to denote to 2 irides of different color?
A. Heterochromia iridium
B. Heterochromia iridis
C. Dyscoria
D. Corectopia
E. Anisocoria

12. A young mother brought her 3-week-old well looking baby complaining from intermittent misalignment of the eyes, she seeks your advice regarding the prognosis of this problem.
O the following, the **MOST** appropriate advice is that it usually
A. resolves without treatment before 6 wk of age
B. resolves without treatment before 12 wk of age
C. resolves without treatment before 24 wk of age
D. resolves without treatment before 1 year of age
E. persists for life

13. Which of the following can cause severe visual impairment in childhood?
A. Rifampicin
B. Isoniazid
C. Streptomycin
D. Ethambutol
E. Pyrazinamide

14. The mother of an 18-month-old boy noticed that he has excessive problems going to sleep in a dark room, while he sleeps easily and comfortably when the lights are on. Clinical examination shows a well built, cheerful, cooperative normal baby.
Of the following, the **MOST** likely cause of this behavior is
A. separation anxiety
B. primary retinal degeneration
C. night blindness
D. autism

E. vitamin A deficiency

15. A 2-year-old boy presents with outward drifting of right eye, which usually occurs when he is fixating at distance. The deviation is more frequent with fatigue or illness. The visual acuity is good in both eyes.
Of the following, the **MOST** likely cause is

A. infantile esotropia
B. accommodative esotropia
C. intermittent exotropia
D. pseudostrabismus
E. heterophoria

16. A young mother brought her 3-day-old neonate with mild crossed eyes with limited ability to move the right eye laterally. The baby is a product of difficult vaginal delivery, admitted to NICU for 24 hours and discharged home in good condition.
Of the following, the **MOST** likely cause is

A. congenital 6th nerve palsy
B. transient 6th nerve paresis
C. sequel of hypoxic ischemic encephalopathy
D. sequel of intracranial hemorrhage due to vitamin K deficiency
E. damage to lateral rectus ocular muscle

17. A mother brought her 4-year-old boy complaining from impaired hearing and expressionless face. Examination reveals incomplete, bilateral asymmetric facial paresis; with ptosis and esotropia.
Of the following, the **MOST** likely diagnosis is

A. Duane syndrome
B. Möbius syndrome
C. Brown syndrome
D. Parinaud syndrome
E. sylvian aqueduct syndrome

18. A 14-month-old boy presents with fine, rapid, pendular nystagmus, head nodding, and torticollis.
Of the following, the **MOST** likely diagnosis is

A. opsoclonus
B. gaze-paretic nystagmus

C. downbeat nystagmus
D. spasmus nutans
E. Ocular bobbing

19. Which of the following may be the first sign of neuroblastoma?
A. Opsoclonus
B. Ocular dysmetria
C. Ocular flutter
D. Ocular bobbing
E. Ocular myoclonus

20. A 4-month-old infant with left upper lid ptosis, the mother noticed that the affected lid raises up while the baby sucks from the bottle.
Of the following, the **MOST** likely cause of this ptosis is
A. myasthenia gravis
B. muscular dystrophy
C. blepharophimosis syndrome
D. Marcus Gunn jaw-winking phenomenon
E. congenital 3rd nerve palsy

21. Which of the following is the **MOST** appropriate initial treatment of rapidly expanding lid capillary hemangioma?
A. Topical timolol
B. Intralesional corticosteroid
C. Systemical corticosteroid
D. Systemic propranolol
E. Surgical excision

22. What is the usual presentation of children with hyphema?
A. Loss of vision
B. Subconjuctival hemorrhage
C. Strabismus
D. Leukocoria
E. Local pain

23. What is the **MOST** common cause of tearing in infants?
A. Nasolacrimal obstruction
B. Glaucoma

C. Intraocular inflammation
D. Corneal abrasion
E. Foreign body

24. A 10-day-old newborn admitted to NICU with diagnosis of late onset sepsis, 1 day later the patient develops edema, erythema of the lids, purulent discharge, pannus formation, and endophthalmitis with rapid deterioration to septic shock.
Of the following, the **MOST** likely pathogen is

A. Staphylococcus aureus
B. Chlamydia trachomatis
C. N. gonorrhoeae
D. Pseudomonas aeruginosa
E. E.coli

25. What is the **MOST** frequent cause of bacterial purulent conjunctivitis in children?

A. Pneumococci
B. Staphylococci
C. Gonococci
D. Meningococci
E. Nontypable haemophilus influenza

26. A 10-year-old boy presents with low grade fever, sore throat, sensation of a foreign body beneath the lids, itching, with edema and photophobia. Examination reveals large oval follicles within the conjunctiva, preauricular adenopathy, and a pseudomembrane on the conjunctival surface.
Of the following, the **MOST** likely pathogen is

A. enterovirus
B. coxsackie virus
C. adenovirus
D. diphtheria
E. chlamydia

27. A 12-year-old boy comes to outpatient clinic in summer complaining from intense itching, clear watery discharge, and conjunctival edema.
Of the following, the **MOST** appropriate initial treatment is topical

A. mast cell stabilizers

B. prostaglandin inhibitors
C. corticosteroids
D. antihistamine drops
E. antibiotics

28. Which of the following should be avoided in dendritic keratitis?
A. Mechanical debridement
B. Topical ganciclovir
C. Systemic acyclovir
D. Cycloplegic agent
E. Topical corticosteroids

29. Which of the following may cause cataract?
A. Spherocytosis
B. Thalassemia
C. Sickle cell disease
D. G6PD deficiency
E. Iron deficiency anemia

30. An 8-year-old febrile toxic looking boy presents with proptosis, painful limitation of movement of the right eye, and decreased visual acuity, together with edema of the conjunctiva and swelling of the eyelids.
Of the following, the **MOST** likely diagnosis is
A. idiopathic orbital inflammation
B. granulomatous vasculitis
C. orbital cellulitis
D. idiopathic orbital inflammation
E. metastatic tumor

31. Which of the following causes of anterior uveitis may remain asymptomatic?
A. Kawasaki disease
B. Ulcerative colitis
C. Brucellosis
D. Herpes simplex
E. Juvenile idiopathic arthritis

32. Which of the following represent a major risk factor in retinopathy of prematurity?

A. Oxygenation
B. Retinal immaturity
C. Apnea
D. Hypercarbia
E. Anemia

33. What is the initial sign in the majority of patients with retinoblastoma?
A. Strabismus
B. Pseudohypopyon
C. Hyphema
D. Leukocoria
E. Vitreous hemorrhage

34. What is the first clinical manifestation of retinitis pigmentosa?
A. Impairment of night vision
B. Progressive loss of peripheral vision
C. Loss of central vision
D. Strabismus
E. Leukocoria

35. The diagnosis of which of the following disorders, in a patient with presumed retinitis pigmentosa, is important?
A. Scheie syndrome
B. Sanfilippo syndrome
C. Kearns-Sayre syndrome
D. Laurence-Moon syndrome
E. Refsum disease

36. A cherry-red spot typically occurs in which of the following conditions?
A. Metachromatic leukodystrophy
B. Niemann-Pick disease
C. Tay-Sachs disease
D. Galactosialidosis
E. Mucolipidoses

37. What is the **MOST** frequent presenting sign of Coats disease?
A. Blurring of vision
B. Rubeosis of the iris

C. Glaucoma
D. Cataract
E. Vitreous hemorrhage

38. Which of the following hypertensive retinal changes is irreversible?
A. Generalized constriction of the arterioles
B. Flame-shaped hemorrhages
C. Cotton-wool spots
D. Papilledema
E. Copper-wire appearance

39. Which of the following is a principal sign of orbital tumors?
A. Ptosis
B. Optic nerve head congestion
C. Optic atrophy
D. Proptosis
E. Loss of vision

40. A 2-year-old child presents with severe headache, fever, severe malaise, and decreased vision. Examination reveals neck rigidity, positive kernig sign, bilateral ptosis, proptosis, swelling, and ophthalmoplegia. CSF result is normal.
Of the following, the **MOST** likely cause of the eye lesion is
A. orbital cellulitis
B. subperiosteal abscess
C. orbital abscess
D. septic cavernous sinus thrombophlebitis
E. inflammatory edema

41. A 4-year-old child presents with pain, redness, swelling, increase in tearing, and discharge over the lateral one third of the right upper eyelid associated with concurrent preauricular lymphadenopathy on the same side.
Of the following, the **MOST** likely diagnosis is
A. dacryocystitis
B. dacryoadenitis
C. preseptal cellulitis
D. hordeolum
E. chalazion

Chapter 28

Disorders of the Eye
Answers

ZUHAIR ALMUSAWI

1.(A) The cornea is relatively large in newborns (averaging 10 mm) and attains adult size (nearly 12 mm) by the age of 2 yr or earlier.

2.(C) Many normal infants may have imperfect coordination of the eye movements and alignment during the early days and weeks, but proper coordination should be achieved by 3-6 mo, usually sooner. Persistent deviation of an eye in an infant at 6 mo of age requires evaluation.

3.(E) The visual acuity in newborns is estimated to be approximately 20/400. This poor vision is a result of the immature, multilayered foveal anatomy.

4.(C) visual **acuity** of 20/40 is generally accepted as normal for 3 yr old children. At 4 yr of age, 20/30 is acceptable. By 5 or 6 yr of age, most children attain 20/20 vision.

5.(B) Myopia is infrequent in infants and preschool-age children. It is more common in infants with a history of retinopathy of prematurity.

6.(C) Although full-time occlusion has historically been considered the best way to treat children with amblyopia, a series of prospective studies has shown that some children can achieve similar results with part-time patching or the use of atropine drops.

7.(A) Gliomas of the optic nerve and chiasm and craniopharyngiomas are primary diagnostic considerations in children who show progressive loss of vision.

8.(C) The first clue to amaurosis in an infant may be nystagmus or strabismus, with the vision deficit itself passing undetected for some time. Timidity, clumsiness, or behavioral change may be the initial clues in the very young. Deterioration in school progress and indifference to school activities are common signs in an older child.

9.(E) Patient who has more prominent anisocoria in light than darkness, indicating that the pupillary constrictor of the larger pupil is abnormal (i.e., it fails to constrict in light)as in third nerve palsy, tonic pupil, pharmacologic mydriasis, or a disorder of the iris while a patient who has more prominent anisocoria in darkness than light, indicating that the pupillary dilator of the

smaller pupil is abnormal (i.e., it fails to dilate in darkness) could be Horner syndrome or simple anisocoria.

10.(E) Retinal hemorrhage is the most common ophthalmic finding and occurs at all levels of the retina. The pattern of hemorrhage helps distinguish this disorder from other causes of retinal hemorrhage or from accidental injuries.

11.(A) In heterochromia, the 2 irides are of different color (heterochromia iridium) or a portion of an iris differs in color from the remainder (heterochromia iridis).

12.(B) Between 2 and 4 mo of age, many infants have infantile esotropia (neonatal misalignments), which in most resolve spontaneously. Those that resolve without treatment do so before 10-12 wk of age and have intermittent or variable deviations. Those most likely to benefit from active treatment have persistent esotropia (10 wk-6 mo of age) and constant esotropia (40 PD), in combination with a refractive error ≤ +3.00 D, and the absence of prematurity, developmental delay, meningitis, nystagmus, eye anomalies, and incomitant or paralytic strabismus.

13.(D)

14.(C) Nyctalopia, or night blindness, is vision that is defective in reduced illumination. It generally implies impairment in function of the rods, particularly in dark adaptation time and perceptual threshold. Stationary congenital night blindness may occur as an autosomal dominant, autosomal recessive, or X-linked recessive condition. It may be associated with myopia and nystagmus. Children may have excessive problems going to sleep in a dark room, which may be mistaken for a behavioral problem. Progressive night blindness usually indicates primary or secondary retinal, choroidal, or vitreoretinal degeneration; it occurs also in vitamin A deficiency or as a result of retinotoxic drugs such as quinine.

15.(C) Exodeviations are the second most common type of misalignment. The divergent deviation may be intermittent or constant. Intermittent exotropia is the most common exodeviation in childhood. It is characterized by outward drifting of 1 eye, which usually occurs when a child is fixating at distance. The deviation is generally more frequent with fatigue or illness. Exposure to bright light may cause reflex closure of the exotropic eye. Because the eyes initially can be kept straight most of the time, visual acuity tends to be good in both eyes and binocular vision is initially normal. The age at onset of intermittent exotropia varies but is often between age 6 mo and 4 yr.

16.(B) In neonates, a transient 6th nerve paresis can occur; it usually clears spontaneously by 6 wk. It is believed that increased intracranial pressure associated with labor and delivery is the contributing factor.

17.(B) The distinctive features of Möbius syndrome are congenital facial paresis and abduction weakness. The facial palsy is commonly bilateral, frequently asymmetric, and often incomplete, tending to spare the lower face and platysma. Ectropion, epiphora, and exposure keratopathy may develop. The abduction defect may be unilateral or bilateral. Esotropia is common.

18.(D) Spasmus nutans is a special type of acquired nystagmus in childhood. In its complete form, it is characterized by the triad of pendular nystagmus, head nodding, and torticollis. The nystagmus is characteristically very fine, very rapid, horizontal, and pendular; it is often asymmetric, sometimes unilateral. Signs usually develop within the first year or two of life. Components of the triad may develop at various times. In many cases, the condition is benign and self-limited, usually lasting a few months, sometimes years.

19.(A) Opsoclonus is most often associated with infectious or autoimmune encephalitis. It may be the first sign of neuroblastoma or other tumors producing a paraneoplastic syndrome.

20.(D) Marcus Gunn jaw-winking ptosis (maxillopalpebral synkinesis) accounts for 5% of ptosis in children. In this syndrome, an abnormal synkinesis exists between the 5th and 3rd cranial nerves; this causes the eyelid to elevate with movement of the jaw. The wink is produced by chewing or sucking and may be more noticeable than the ptosis itself.

21.(D) In the case of a rapidly expanding lesion, which may cause amblyopia by obstructing the visual axis or inducing astigmatism, treatment should be considered. Systemic propranolol has been shown to be an effective treatment without the risks associated with corticosteroid use. Other treatment options include topical timolol, corticosteroids (systemically or by direct injection), and surgical excision.

22.(A) Hyphema is the presence of blood in the anterior chamber of the eye. It may occur with either a blunt or perforating injury and represents a situation that may threaten vision. Hyphema appears as a bright or dark red fluid level between the cornea and iris, or as a diffuse murkiness of the aqueous humor. Children with hyphema present with acute loss of vision, with or without pain.

23.(A)

24.(D) Conjunctivitis caused by Pseudomonas aeruginosa is uncommon, acquired in the nursery, and a potentially serious process. It is characterized by

the appearance on days 5-18 of edema, erythema of the lids, purulent discharge, pannus formation, endophthalmitis, sepsis, shock, and death.

25.(E) The most frequent causes are nontypable Haemophilus influenzae (60–80%; associated with ipsilateral otitis media), pneumococci (20%), and staphylococci (5–10%).

26.(C) Epidemic keratoconjunctivitis is caused by adenovirus serotypes 8, 19, or 37, and is transmitted by direct contact.

27.(D) Allergic conjunctivitis is usually accompanied by intense itching, clear watery discharge, and conjunctival edema (chemosis). It is commonly seasonal (spring-summer). Cold compresses and topical antihistamine drops give symptomatic relief. Topical mast cell stabilizers or prostaglandin inhibitors may also help. In selected cases, topical corticosteroids are used under an ophthalmologist's supervision but should not be used routinely or for a long time.

28.(E) Topical use of corticosteroids causes exacerbation of superficial herpetic disease of the eye and may lead to corneal perforation; eye drops combining steroids and antibiotics are therefore to be avoided in treatment of red eye, unless there are clear-cut indications for their use and close supervision during therapy.

29.(A) Miscellaneous disorders causing cataract

- ✓ Atopic dermatitis
- ✓ Drugs (corticosteroids)
- ✓ Radiation
- ✓ Trauma
- ✓ Juvenile idiopathic arthritis
- ✓ Retinopathy of prematurity
- ✓ Spherocytosis

30.(C) Inflammation of the tissues of the orbit, characterized by the triad of proptosis, painful limitation of movement of the eye, and potentially decreased visual acuity, is termed orbital cellulitis. Edema of the conjunctiva (chemosis) and inflammation and swelling of the eyelids may be seen.

31.(E) Iridocyclitis that occurs in children with juvenile idiopathic arthritis deserves special mention. Unlike most forms of anterior uveitis, it rarely creates pain, photophobia, or conjunctival hyperemia. Loss of vision may not be noticed until severe and irreversible damage has occurred.

32.(B) The risk factors associated with ROP are not fully known, but prematurity and the associated retinal immaturity at birth represent the major factors. Oxygenation, respiratory distress, apnea, bradycardia, heart disease, infection,

hypercarbia, acidosis, anemia, and the need for transfusion are thought by some to be contributory factors. Generally the lower the gestational age, the lower the birth weight, and the sicker the infant, the greater the risk for ROP.

33.(D) The clinical manifestations of retinoblastoma vary depending on the stage at which the tumor is detected. The initial sign in the majority of patients is a white pupillary reflex (leukocoria). Leukocoria results because of the reflection of light off the white tumor. The second most frequent initial sign of retinoblastoma is strabismus.

34.(A) Impairment of night vision or dark adaptation is often the first clinical manifestation. Progressive loss of peripheral vision, often in the form of an expanding ring scotoma or concentric contraction of the field, is usual. There may be loss of central vision.

35.(E) The retinal manifestations of abetalipoproteinemia (Bassen-Kornzweig syndrome; and Refsum disease are also similar to those found in retinitis pigmentosa. The diagnosis of the latter 2 disorders in a patient with presumed retinitis pigmentosa is important because treatment is possible.

36.(C) A cherry-red spot typically occurs in certain sphingolipidoses, principally in Tay-Sachs disease (GM2 type 1), in the Sandhoff variant (GM2 type 2), and in generalized gangliosidosis (GM1 type 1). Similar but less distinctive macular changes occur in some cases of metachromatic leukodystrophy (sulfatide lipidosis), in some forms of neuronopathic Niemann- Pick disease, in galactosialidosis, and in certain mucolipidoses.

37.(A) Coats disease is exudative retinopathy of unknown nonhereditary cause is characterized by telangiectasia of retinal vessels with leakage of plasma to form intraretinal and subretinal exudates and by retinal hemorrhages and detachment. The condition is usually unilateral. It predominantly affects boys, usually appearing in the 1st decade. The condition is nonfamilial and, for the most part, occurs in otherwise healthy children. The most frequent presenting signs are blurring of vision, leukocoria, and strabismus. Rubeosis of the iris, glaucoma, and cataract may develop. Treatment with photocoagulation or cryotherapy may be helpful.

38.(E) In long-standing hypertension, irreversible changes may occur. Thickening of the vessel wall may produce a silver- or copper-wire appearance. Hypertensive retinal changes in a child should alert the physician to renal disease, pheochromocytoma, collagen disease, and cardiovascular disorders, particularly coarctation of the aorta.

39.(D) The effects of orbital tumors vary with their locations and growth patterns. The principal signs are proptosis, resistance to retroplacement of the

eye, and impairment of eye movement. A palpable mass may be found. Other significant signs are ptosis, optic nerve head congestion, optic atrophy, and loss of vision. Bruit and visible pulsation of the globe are important clues to vascular lesions.

40.(D)

41.(B) Dacryoadenitis is inflammation of the lacrimal gland; it most commonly occurs in the pediatric population and in some young adults and is related to a variety of infectious pathogens. Pain, redness, swelling, increase in tearing, and discharge over the lacrimal gland are noted and usually visible at the lateral one third of the upper eyelid; concurrent preauricular lymphadenopathy may be noted. It may occur with mumps (in which case it is usually acute and bilateral, subsiding in a few days or weeks), with influenza, infectious mononucleosis, and herpes zoster. Staphylococcus aureus may produce a suppurative dacryoadenitis, and other bacterial causes include streptococci and Neisseria gonorrhoeae.

Chapter 29

The Ear
Questions

MUSTAFA ALANBAKI

1. What is the **MOST** common cause of otalgia in children?
   A. Foreign body in the auditory canal
   B. Trauma
   C. Otitis externa
   D. Acute otitis media
   E. Mastoiditis

2. What is the **MOST** common cause of conductive hearing loss in children?
   A. Tympanic membrane perforation
   B. Middle ear effusion
   C. Impacted cerumen
   D. Foreign body in the auditory canal
   E. Cholesteatoma

3. What is the **MOST** common infectious cause of congenital sensorineural hearing loss?
   A. Congenital rubella
   B. Congenital toxoplasmosis
   C. Congenital syphilis
   D. Congenital cytomegalovirus
   E. Congenital herpes simplex

4. Which of the following is the average threshold level (db) of severe hearing loss?
   A. 16-25
   B. 26-30
   C. 31-50
   D. 51-70
   E. More than 70

5. What is the correct answer regarding criteria for referral of a child for audiologic assessment?

A. No use of single words in the age of 12 months
B. Single-word vocabulary of ≤10 words in the age of 18 months
C. No evidence of 2-word combinations; unintelligible in the age of 30 months
D. <600 words; clarity ≤80% in the age of 36 months
E. No use of telegraphic sentences in the age of 48 months

6. At what age a child with a known risk factor for hearing loss should be evaluated?

A. First week of life
B. First month of life
C. First 3 months of life
D. First 6 months of life
E. First year of life

7. What is the most common cause of vertigo in children?

A. Acute serous labyrinthitis
B. Acute suppurative labyrinthitis
C. Vestibular neuritis
D. Vestibular migraine
E. Benign paroxysmal vertigo

8. What is the most common causative agent of otitis externa?

A. Staphylococcus Aureus
B. Pseudomonas aeruginosa
C. Proteus mirabilis
D. Klebsiella pneumoniae
E. Candida albicans

9. Which of the following differentiate between otitis externa and otitis media in children?

A. Otitis externa is more common
B. Fever is more prominent in otitis externa
C. Periauricular lymphadenitis is more common in otitis externa
D. Otoscopic examination of the tympanic membrane should be normal in otitis externa

E. Hearing loss is more common in otitis externa

10. A 2-year-old toddler is brought by his parents with 5 days history of fever, severe earache of the right side, associated with redness of right eye with greenish secretions from it. Exam: Temperature 38.4, PR 100, RR 28, BP 100/65. He is active, alert to his surroundings and otherwise in no distress. Right tympanic membrane is erythematous and bulging, with poor mobility on pneumatic otoscopy.
What is the most common causative agent?

A. Streptococcus pneumoniae
B. Nontypeable Haemophilus influenzae
C. Moraxella catarrhalis
D. Chlamydia trachomatis
E. Staphylococcus aureus

11. What is the most specific tympanic membrane finding in acute otitis media?

A. Erythema
B. Bulging
C. Opacification
D. Perforation
E. Immobility

12. A 7-month-old infant presented to you 3 days ago with second attack of acute otitis media and you gave him azithromycin suspension orally as he had urticarial rash following amoxicillin therapy in the first attack, but no improvement of his symptoms, so you want to change his antibiotic.
Of the following, the **MOST** appropriate choice is

A. trimethoprim-sulfamethoxazole
B. cefdinir
C. levofloxacin
D. ceftriaxone
E. clindamycin

13. What is the antibiotic of choice for acute otitis media (AOM), if β-lactamase-producing pathogen is suspected?

A. Ceftriaxone
B. Cefdinir
C. Amoxicillin-clavulanate

D. Levofloxacin
E. Clarithromycin

14. What is the most common causative pathogen in uncomplicated acute mastoiditis in children?

A. Staphylococcus aureus
B. Streptococcus pyogenes
C. Pseudomonas aeruginosa
D. Haemophilus influenzae
E. Streptococcus pneumoniae

15. A 12-year-old boy presented to the emergency room with a 5-day-history of left sided otalgia, with 2 days of left temporal and parietal headache, vomiting, and vertigo. He received co-amoxiclav with a diagnosis of "ear infection. On examination, temp 39.8°C. He has neck stiffness but no focal neurological signs but left side papilledema. The left tympanic membrane is red and bulging. There are retroaurcular redness, tenderness and swelling.
Of the following, the **MOST** appropriate next step is

A. change the antibiotic to IV ceftriaxone and follow up after 72 hours
B. change the antibiotic to IV ceftriaxone and vancomycin and perform ultrasound to the left mastoid area.
C. change the antibiotic to IV ceftriaxone and vancomycin and perform CT scan of the left temporal bone
D. change the antibiotic to IV ceftriaxone and vancomycin and perform contrast-enhanced CT scan of the brain.
E. change the antibiotic to IV ceftriaxone and vancomycin and perform MRI and MRV of the brain.

16. An 18- month-old toddler had recently a cochlear implant due to severe sensorineural hearing loss, you suggest pneumococcal vaccination.
What is the recommended schedule?

A. 3 doses of 13-valent pneumococcal conjugate vaccine (PCV13), 2 months apart.
B. 3 doses of 13-valent pneumococcal conjugate vaccine (PCV13), 2 months apart, with 1 extra dose of PCV13 at ≥24 months of age.
C. 3 doses of 13-valent pneumococcal conjugate vaccine (PCV13), 2 months apart, with 1 dose of 23-valent pneumococcal polysaccharide vaccine (PPV23) at ≥24 months of age.

D. 2 doses of 13-valent pneumococcal conjugate vaccine (PCV13), 2 months apart, with 1 extra dose of PCV13 at ≥24 months of age.

E. 2 doses of 13-valent pneumococcal conjugate vaccine (PCV13), 2 months apart, with 1 dose of 23-valent pneumococcal polysaccharide vaccine (PPV23) at ≥24 months of age.

Chapter 29

The Ear
Answers

MUSTAFA ALANBAKI

1.(D) Otalgia usually is associated with inflammation of the middle ear (about 50% of cases) or external ear, but it can represent pain referred from involvement of the teeth, temporomandibular joint, or pharynx.
2.(B) Most conductive hearing loss (CHL) is acquired, with middle-ear fluid the most common cause.
3.(D) The most common infectious cause of congenital sensorineural hearing loss (SNHL) is cytomegalovirus (CMV).
4.(D) Hearing loss is categorized according to average threshold level (db) at 500-2,000 hz (ansi) into the following levels:
Normal range 0-15 db
Slight hearing Loss 16-25 db
Mild hearing Loss 26-30 db
Moderate hearing Loss 31-50 db
Severe hearing Loss 51-70 db
Profound hearing Loss > 70 db
5.(C) The Criteria for Referral for Audiologic Assessment is as following:
At 12 months: No differentiated babbling or vocal imitation
At 18 months: No use of single words
At 24 months: Single-word vocabulary of ≤10 words
At 30 months: <100 words; no evidence of 2-word combinations; unintelligible
At 36 months: <200 words; no use of telegraphic sentences; clarity <50%
At 48 months: <600 words; no use of simple sentences; clarity ≤80%
6.(D) Any child with a known risk factor for hearing loss should be evaluated in the first 6 months of life.
7.(E) Benign paroxysmal vertigo, the most common cause of vertigo in pediatric patients, is characterized by short periods of vertigo or dizziness lasting seconds to a few minutes and associated with imbalance and nystagmus; tinnitus or hearing loss is unusual.
8.(B) External otitis (swimmer's ear, although it can occur without swimming) is caused most commonly by P. aeruginosa (up to 60%), but S. aureus,

Enterobacter aerogenes, Proteus mirabilis, Klebsiella pneumoniae, streptococci, coagulase-negative staphylococci, diphtheroids, and fungi such as Candida and Aspergillus also may be isolated.

9.(C) Otitis media is more common than otitis externa and is a leading reason for physician visits and for use of antibiotics and figures importantly in the differential diagnosis of fever. The predominant symptom of otitis externa is acute rapid onset (typically within 48 hr) of ear pain (otalgia), often severe, accentuated by manipulation of the pinna or by pressure on the tragus and by jaw motion. The severity of the pain and tenderness (tragus or pinna, or both) may be disproportionate, If the tympanic membrane (TM) can be visualized (in otitis externa), it may appear either normal or opaque, TM mobility may be normal or, if the TM is thickened, mobility may be reduced in response to positive and negative pressure. Other physical findings may include palpable and tender lymph nodes in the periauricular region, and erythema and swelling of the pinna and periauricular skin. Conductive hearing loss (CHL) may result from edema of the skin and tympanic membrane (TM), serous or purulent secretions, or the canal skin thickening associated with chronic external otitis, but on the other hand middle ear disease and on the top of the list otitis media with effusion is the most common cause of conductive hearing loss.

10.(B) Conjunctivitis-associated otitis media, simultaneous appearance of purulent and erythematous conjunctivitis with an ipsilateral OM is a well-recognized presentation, caused by nontypeable H. Influenzae in most children. The disease often is present in multiple family members and affects young children and infants.

11.(B) Bulging of the TM is the most specific finding of AOM (97%) but has lower sensitivity (51%).

12.(C) For children with a non–type I reaction in which cross reactivity with cephalosporins is of less concern, first-line therapy with cefdinir would be an appropriate choice. In children with a type I reaction or known sensitivity to cephalosporin antibiotics, there are far fewer choices. Resistance to trimethoprim-sulfamethoxazole by many strains of both nontypeable H. influenzae and S. pneumoniae and a reported high clinical failure rate in children with AOM treated initially with this antimicrobial argue against its use. Similarly, increasing rates of macrolide resistance argue against the efficacy of azithromycin. Although not approved by the FDA for use in children, many clinicians have used quinolones in this patient population. Resistance to macrolides also results in resistance to clindamycin, which otherwise is generally effective against resistant strains of S. pneumoniae.

13.(C) In cases of antibiotic resistance which is attributable to production of β-lactamase, can be overcome by combining amoxicillin with a β-lactamase inhibitor (clavulanate).

14.(E) The most common causative pathogen in uncomplicated acute mastoiditis in children is Streptococcus pneumoniae (10-51%), Streptococcus pyogenes (0–12%), Staphylococcus aureus (2–10%), Pseudomonas aeruginosa (10%), and Haemophilus influenzae (2–3%).

15.(D) The provisional diagnosis of this case is acute otitis media complicated by acute mastoiditis and intracranial (mostly epidural) abscess. ultrasound can be helpful to differentiate postauricular erythema from a postauricular abscess and avoids the risk of ionizing radiation exposure. However, ultrasound is not as sensitive as CT scanning and will underdiagnose postauricular abscess formation and will provide no information as to whether there is an intracranial complication present such as a brain abscess. CT scan of the temporal bone can confirm the diagnosis of acute mastoiditis, if there is any concern about the possibility of an intracranial complication (as in this case), a contrasted CT scan is the most sensitive test readily available and should be ordered upon presentation. Magnetic resonance imaging is generally reserved for patients in whom there is a suspected intracranial complication like sagittal vein thrombosis. Regarding antibiotic therapy If intracranial infection is suspected, broader spectrum antimicrobial coverage (e.g., vancomycin plus a 3rd-generation cephalosporin) should be initiated.

16.(E)

Recommended pneumococcal vaccination schedule for persons with cochlear implants			
Age at first pcv13 dose (months)	PCV13 primary series	PCV13 additional dose	PPV23 dose
2-6	3 doses, 2 months apart	1 dose at 12-15 months of age	Indicated at ≥24 months of age
7-11	2 doses, 2 months apart	1 dose at 12-15 months of age	Indicated at ≥24 months of age
12-23	2 doses, 2 months apart	Not indicated	Indicated at ≥24 months of age
24-59	2 doses, 2 months apart	Not indicated	Indicated
≥60	Not indicated	Not indicated	Indicated

Chapter 30

Skin Questions

ZUHAIR ALMUSAWI

1. What is the usual scalp hair growth per month?
   A. 0.5 cm
   B. 1 cm
   C. 1.5 cm
   D. 2 cm
   E. 2.5 cm

2. How long it takes for a complete fingernail to regrow?
   A. 2 months
   B. 4months
   C. 6 months
   D. 9 months
   E. 12 months

3. A 2-year-old boy diagnosed as acute myelogenous leukemia 2 months ago, and now on treatment, presents with fever and oral ulcers; with tender, erythematous, and edematous pustular nodules on the arms, legs, and trunk. CBC reveals anemia, and absolute neutrophilia.
Of the following, the **MOST** likely diagnosis is
   A. pyoderma gangrenosum
   B. necrolytic migratory erythema
   C. langerhans cell histiocytosis
   D. Behçet disease
   E. Sweet syndrome

4. When medication eruptions usually begin after exposure?
   A. Immediately
   B. 1-3 days
   C. 4-6 days
   D. 7-10 days

E. 11-14 days

5. A 3-year-old boy with β-thalassemia major underwent bone marrow transplant 2- weeks ago, presents with fever, mucositis, diarrhea, erythematous maculopapular eruption that starts at the head and neck including the ears, and subsequently spread to the trunk, extremities, palms, and soles.
Of the following, the **MOST** likely diagnosis is

A. Sweet syndrome
B. acute graft versus host disease
C. drug eruption
D. necrolytic migratory erythema
E. Langerhans cell histiocytosis

6. A 4-year-old boy diagnosed 5 weeks ago as focal epilepsy treated with carbamazepine with good response. Today presents with fever, malaise, diffuse exanthem of pruritic, morbilliform papules on the head, upper trunk, and arms, with facial edema and cervical lymphadenopathy. CBC shows eosinophilia and atypical lymphocytosis with elevated liver transaminase.
Of the following, the **MOST** likely diagnosis is

A. medication reaction
B. Sweet syndrome
C. DRESS syndrome
D. serum sickness–like reaction
E. Stevens-Johnson syndrome

7. Which of the following is a common offending medication causing fixed-drug eruption?

A. Penicillin
B. Sulfonamide
C. Macrolide
D. Cephalosporin
E. Aminoglycoside

8. A 7-year-old boy visited the primary health center due to sore throat with low grade fever, azithromycin was prescribed for him, 2 days after the drug therapy is started, he develops generalized erythema with numerous, small, discrete, nonfollicular, pruritic, sterile pustules beginning on the face and intertriginous regions.

Of the following, the **MOST** likely diagnosis is

A. generalized pustular psoriasis
B. bullous impetigo
C. IgA pemphigus
D. subcorneal pustular dermatosis
E. acute generalized exanthematous pustulosis

9. Which of the following topical medications vehicle is preferable for dry scaly skin?

A. Lotion
B. Cream
C. Ointment
D. Gel
E. Solution

10. Which of the following topical antibiotics is **MOST** effective in treatment of moderate impetigo?

A. Neomycin
B. Nitrofurazone
C. Fusidic acid
D. Polysporin
E. bacitracin

11. Which of the following is a medium-potency topical corticosteroid?

A. Mometasone furoate
B. Hydrocortisone
C. Desonide
D. Fluocinonide
E. Betamethasone dipropionate

12. A 3-day-fullterm, well looking boy brought by his mother because she noticed numerous firm, yellow-white, 1-2 mm papules and pustules over his body surface. Intralesional aspirate demonstrates eosinophils with sterile cultures.

Of the following, the **MOST** likely diagnosis is

A. pyoderma
B. candidiasis
C. transient neonatal pustular melanosis

D. miliaria
E. erythema toxicum

13. An 11-month-old boy brought by his mother complaining from episodes of high fever, more during summer time with poor response to conventional management. Examination shows dry, hypopigmented skin, frontal bossing, a flattened nasal bridge, thick everted lips, and prominent low-set ears, with sparse scalp hair, eyebrows and lashes.
Of the following, the **MOST** likely diagnosis is

A. hypohidrotic ectodermal dysplasia
B. hidrotic ectodermal dysplasia
C. Fabry disease
D. Sjögren syndrome
E. hereditary sensory autonomic neuropathy

14. A 9-month-old infant brought by his mother complaining from a macular, sharply circumscribed, pink lesion localized to the right forehead and upper eyelid, with repeated seizures and left hemiparesis. The mother seeks your advice about the skin lesion.
Your advice is to

A. start pulsed-dye laser therapy
B. start topical corticosteroid ointment
C. start calcineurin-inhibiting antiinflammatory agents
D. reassure the mother that it will fade with time
E. advise the mother to use masking cosmetics

15. Which of the following sites of infantile hemangioma seem to persist most often?

A. Lip
B. Check
C. Scalp
D. Back
E. Anterior chest

16. A 6-month-old infant presents with enlarging infantile hemangioma of the lower eyelid obscuring the medial canthus.
Of the following, the **MOST** appropriate treatment for this infant is

A. expectant observation

B. topical timolol solution
C. oral propranolol
D. oral corticosteroids
E. intralesional corticosteroid injection

17. Which of the following drugs decreases the blood level of propranolol?
A. Cimetidine
B. Ciprofloxacin
C. Isoniazid
D. Theophylline
E. Phenytoin

18. A 9-month-old boy presents with pallor, ecchymoses, and petechiae with rapid increase in the size of a large vascular lesion on axilla. The platelet count is depressed, but the bone marrow contains increased numbers of normal megakaryocytes.
Of the following, the **FIRST**-line therapy is
A. surgical excision
B. systemic steroid
C. antiplatelet
D. antifibrinolytic
E. sirolimus

19. A 5-year-old child presents with a small red, glistening, sessile papule with a weeping and crusted surface on left check.
Of the following, the **MOST** likely diagnosis is
A. spider angioma
B. angiokeratoma of Mibelli
C. pyogenic granuloma
D. spindle cell hemangioma
E. congenital hemangioma

20. A 10-year-old boy presents with 1-3 mm red to blue-black papules with a hyperkeratotic surface over the genitalia and in the umbilical and inguinal regions. The boy gives history of recurrent episodes of fever, paresthesia, agonizing pain, cyanosis and flushing of the hands and feet with hypohidrosis.
Of the following, the **MOST** likely diagnosis is
A. Fabry disease

B. Osler-Weber-Rendu disease
C. Maffucci syndrome
D. angiokeratoma of Mibelli
E. Kaposiform hemangioendothelioma

21. A 3-year-old boy presents with pigmented macules of few millimeters in size on the lips, buccal mucosa, nose, hands, feet, and around the mouth, with diffuse hyperpigmentation of the nails. The mother gave history of episodic abdominal pain, diarrhea, and melena.
Of the following, the **MOST** likely diagnosis is
A. Gardner syndrome
B. Laugier-Hunziker syndrome
C. Peutz-Jeghers syndrome
D. Cronkhite-Canada syndrome
E. multiple lentigines syndrome

22. Which of the following has strong association with café-au-lait macules?
A. Ataxia telangiectasia
B. Bloom syndrome
C. Fanconi anemia
D. Russell-Silver syndrome
E. McCune-Albright syndrome

23. Postnatal growth retardation, microcephaly, dysmorphic facies, congenital cardiac defects, malabsorption, anal stenosis, genitourinary anomalies, congenital hip dysplasia, hirsutism, mental retardation, and café-au-lait spots are features of which of the following syndromes?
A. Johanson-Blizzard syndrome
B. Rubinstein-Taybi syndrome
C. Kabuki syndrome
D. Russell-Silver syndrome
E. Bloom syndrome

24. What is the **MOST** common form of albinism seen worldwide?
A. Oculocutaneous albinism type 1 (OCA1)
B. Oculocutaneous albinism type 2 (OCA2)
C. Brown Oculocutaneous albinism
D. Oculocutaneous albinism type 3 OCA3 (rufous albinism)

E. Oculocutaneous albinism type 4 OCA4

25. Hypopigmentation of the skin, eyes, and hair; prolonged bleeding times and easy bruising; recurrent infections; abnormal natural killer cell function; and peripheral neuropathy are features of which of the following syndromes?

A. Hermansky-Pudlak syndrome
B. Chédiak-Higashi syndrome
C. Bloom syndrome
D. Cross-McKusick-Breen syndrome
E. Johanson-Blizzard syndrome

26. A 5-year-old boy presents with sharply demarcated amelanotic patches on the forehead and anterior scalp with a white forelock, there are Islands of darker pigmentation within the amelanotic areas.
Of the following, the **MOST** likely cause of this disorder is

A. piebaldism
B. vitiligo
C. nevus depigmentosus
D. Waardenburg syndrome
E. oculocutaneous albinism type 2

27. What is the hallmark of Waardenburg syndrome type 1?

A. White forelock
B. Depigmented skin
C. Deafness
D. Heterochromia irides
E. Limb abnormalities

28. An 8-year-old boy presents with bizarre patterned, hypopigmented macules arranged over his abdomen in sharply demarcated whorls and streaks. The palms, soles, and mucous membranes are spared with no history of inflammatory or vesicular lesions preceding the development of the pigmentary changes. The patient is hypotonic with nystagmus and intellectual disability.
Of the following, the **MOST** likely cause of this disorder is

A. oculocutaneous albinism type 3 OCA3 (rufous albinism)
B. hypomelanosis of Ito
C. systematized nevus depigmentosus
D. incontinentia pigmenti

E. Vogt-Koyanagi-Harada syndrome

29. A 10-year-old girl presents with a remarkably symmetric pattern of white patches with hyperpigmented margins on periocular area and dorsum of both feet. The patient gave history of frequent carpo-pedal spastic attacks relieved by IV calcium.
Of the following, the **MOST** appropriate treatment is

A. topical steroid
B. topical pimecrolimus
C. narrow-band ultraviolet light
D. cover-up cosmetics
E. minimizing sun exposure of affected areas

30. Which of the following is the **MOST** common implicated factor in the etiology of erythema multiforme?

A. Sulfonamides
B. Herpes simplex virus
C. Mycoplasma pneumoniae
D. Amoxicillin
E. Acetaminophen

31. A 12-year-old boy develops abrupt, fixed papules with a central dusky zone on the palms and extensor upper extremities, followed by oral lesions on vermilion border of the lips and buccal mucosa. The boy gives history of herpetic mouth lesion 10 days ago.
Of the following, the **MOST** likely diagnosis is

A. Stevens-Johnson syndrome
B. pemphigus vulgaris
C. erosive lichen planus
D. erythema multiforme
E. Behçet syndrome

32. Which of the following differentiate toxic epidermal necrolysis from Stevens-Johnson syndrome?

A. Skin tenderness
B. Corneal ulceration
C. Pneumonitis
D. Mouth ulcers

E. Genital ulcers

33. Which of the following should be considered in early management of Stevens-Johnson syndrome?

A. Systemic antibiotics
B. Corticosteroids
C. IV immunoglobulin
D. Azathioprine
E. Dapsone

34. A 2-year-old child presents with easy blistering on palms and soles manifested when the child begins to walk, also the patient has oral mucosal erosions.
Of the following, the **MOST** likely diagnosis is

A. epidermolysis bullosa simplex generalized (formerly Koebner)
B. epidermolysis bullosa simplex localized (formerly Weber-Cockayne)
C. epidermolysis bullosa simplex Dowling-Meara
D. epidermolysis bullosa simplex with mottled pigmentation
E. epidermolysis bullosa simplex superficialis

35. A 5-year-old boy with past history of painful oral ulcers for the last 6 weeks presents with large, flaccid bullae on nonerythematous skin of the face and trunk. The lesions rupture and enlarge peripherally, producing painful, raw, denuded areas that have little tendency to heal. Examination shows malodorous granulomatous lesions at sites of ruptured bullae with positive Nikolsky sign.
Of the following, the **MOST** likely diagnosis is

A. erythema multiforme
B. bullous pemphigoid
C. Stevens-Johnson syndrome
D. pemphigus vulgaris
E. toxic epidermal necrolysis

36. What is the best initial treatment for pemphigus vulgaris?

A. Systemic methylprednisolone
B. Azathioprine
C. Cyclophosphamide
D. Methotrexate
E. IVIG

37. A 7-year-old boy presents with symmetric, grouped, small, intensely pruritic papules and vesicles on both knees and elbows; with hemorrhagic lesions on palms and soles. The boy looks short, thin, with chronic abdominal pain and diarrhea.
Of the following, the **MOST** likely diagnosis is

A. scabies
B. papular urticaria
C. dermatitis herpetiformis
D. contact dermatitis
E. papular eczema

38. What is the **MOST** common type of eczema in children?

A. Seborrheic dermatitis
B. Irritant contact dermatitis
C. Nummular eczema
D. Atopic dermatitis
E. Acute palmoplantar eczema

39. What is the first-line therapy for diaper dermatitis?

A. Topical sucralfate
B. Low-potency topical corticosteroids
C. Zinc oxide ointment
D. Topical anticandidal agent
E. Topical preparations containing triamcinolone-nystatin

40. What is the recommended age for earlobes piercing?

A. Immediately after birth
B. One month
C. First birth day
D. After 10 year of age
E. No age limit

41. Which of the following is the **MOST** common cause of allergic contact dermatitis after topical application?

A. Neomycin
B. Antihistamines
C. Anesthetics
D. Corticosteroids

E. Dye in temporary tattoos

42. A 5-year-old boy presents with hypopigmented, ill-defined, round, mildly erythematous, and finely scaly patches on the face with minimal Itching, the eruption is exacerbated by dryness.
Of the following, the **MOST** likely diagnosis is

A. vitiligo
B. tinea versicolor
C. tinea corporis
D. pityriasis alba
E. contact dermatitis

43. A young mother brought her 3-month-old well looking smiley baby with diffuse scaling and crusting of the scalp followed by a greasy, scaly, erythematous papular non-pruritic dermatitis involving the face, neck, and retroauricular areas. The mother is asking about the prognosis of this lesion.
What is your **MOST** appropriate answer?

A. It resolves by 1 year
B. It resolves by late childhood
C. It will persist through adulthood
D. It reflects systemic dysfunction of the immune system
E. It may progress to psoriasis

44. Which of the following drugs may cause photoallergic drug eruptions?

A. Nalidixic acid
B. Furosemide
C. Sulfonamides
D. Naproxen
E. Piroxicam

45. A 2-year-old child presents with erythema, scaling, crusting, freckles, and keratosis on sun-exposed areas of the body following minimal UV exposure, the patient also has photophobia, lacrimation, blepharitis, and sensorineural deafness.
Of the following, the **MOST** likely diagnosis is

A. Cockayne syndrome
B. trichothiodystrophy
C. xeroderma pigmentosum

D. actinic prurigo
E. polymorphous light eruption

46. A 5-year-old boy had history of plaques of erythema in a butterfly distribution on the face, neck, dorsal portions of the hands, and buttocks started from age of 3 months. These lesions are replaced gradually by reticulated, atrophic, hyperpigmented and hypopigmented telangiectatic patches. The boy looks short with small hands and feet; sparse eyebrows, eyelashes, and alopecia. Eye examination reveals subcapsular bilateral cataracts.
Of the following, the **MOST** likely diagnosis is

A. Bloom syndrome
B. Rothmund-Thomson syndrome
C. cockayne syndrome
D. xeroderma pigmentosum
E. actinic prurigo

47. A 10-year-old boy presents with an eczematous eruption on the face and extremities in a glove-and-stocking photo distribution followed by hyperpigmentation and hyperkeratosis intensified by further exposure to sunlight. He is mentally intact with episodic cerebellar ataxia. The mother mentioned that these cutaneous signs initially developed during the early months of life. The urine contains increased amounts of monoamine monocarboxylic amino acids.
Of the following, the **MOST** likely diagnosis is

A. pellagra
B. Hartnup disease
C. ataxia-telangiectasia
D. xeroderma pigmentosum
E. hydroa vacciniforme

48. A young child, after few weeks of streptococcal pharyngitis, develops an acute eruption of many erythematous papules which coalesce to form round papules smaller than 1.5 cm on the trunk and face with sharply demarcated, irregular borders and a thick silvery scale, when removed result in pinpoint bleeding.
Of the following, the **MOST** likely diagnosis is

A. viral exanthems

B. secondary syphilis
C. pityriasis rosea
D. pityriasis lichenoides chronica
E. guttate psoriasis

49. What is the first-line topical agent for psoriatic skin lesions?
A. Tazarotene
B. Tar preparations
C. Salicylic acid
D. Calcineurin inhibitors
E. calcitriol

50. A 15-year-old dark-skinned boy develops a widespread, symmetric eruption involving mainly the trunk and proximal limbs that are round, pink to brown <1 cm in diameter, slightly raised, and covered by a fine scale. He also gave history of a 7 cm annular patch with raised border and fine, adherent scales on the chest, 7 days prior to this eruption.
Of the following, the **MOST** likely diagnosis is
A. drug eruptions
B. viral exanthem
C. guttate psoriasis
D. nummular dermatitis
E. pityriasis rosea

51. A 13-year-old boy presents with small, firm, skin-colored, warty papules changed to yellow, pruritic, malodorous, greasy crusts coalesced forming large, symmetrical, gray-brown, plaques on the scalp, face, chest, limb flexures, and groin with hyperkeratosis of the palms and soles and nail dystrophy. These lesions worsen in summertime.
Of the following, the **MOST** likely diagnosis is
A. seborrheic dermatitis
B. acanthosis nigricans
C. Hailey-Hailey disease
D. Darier disease
E. flat warts

52. A 4-year-old child, 1 week after a non-specific viral illness, develops numerous, flat-topped, red variable size papules on the face, buttocks, and

limbs, sparing the trunk and scalp. There is also a line of papules on the right shin at site of minor local trauma.
Of the following, the **MOST** likely diagnosis is

A. Gianotti-Crosti syndrome
B. erythema infectiosum
C. lichen planus
D. erythema multiforme
E. Henoch-Schönlein purpura

53. A 10-year-obese female with diabetis mellitus presents with symmetric, hyperpigmented, velvety, hyperkeratotic plaques with exaggerated skin lines in the posterior neck and axillae.
Of the following, the **MOST** likely diagnosis is

A. reticulated papillomatosis
B. Addison disease
C. acanthosis nigricans
D. pellagra
E. erythrasma

54. An 11-month-old female baby brought by her mother in January complaining from slight roughening of the skin surface and scaling most prominent on the extensor aspects of the legs sparing abdomen, neck, and face. There is no accompanying disorder of hair, teeth, mucosal surfaces, or other organ systems.
Of the following, the **MOST** likely diagnosis is

A. X-Linked Ichthyosis
B. ichthyosis vulgaris
C. Harlequin ichthyosis
D. lamellar ichthyosis
E. epidermolytic ichthyosis

55. A 2-year-old child with history of generalized pruritic ichthyosis accentuated on the flexures and the lower abdomen with palmoplantar hyperkeratosis.
The patient has spastic diplegia with delayed speech and primary retinal degeneration. Urinalysis reveals elevation of leukotriene B4 (LTB4).
Of the following, the **MOST** likely diagnosis is

A. Netherton syndrome
B. Sjögren-Larsson syndrome

C. Refsum syndrome
D. chondrodysplasia punctate
E. CHILD syndrome

56. A 15-year-old boy had history of generalized ichthyosis from early childhood, presents with progressive paralysis, ataxia, retinitis pigmentosa, anosmia, and deafness. Lipid analysis of the blood shows elevated phytanic acid values.
Of the following, the **MOST** likely diagnosis is
A. Netherton syndrome
B. Sjögren-Larsson syndrome
C. Refsum syndrome
D. chondrodysplasia punctate
E. CHILD syndrome

57. A 6-month-old infant presents with mild hyperkeratosis, cataract, hypertelorism, optic nerve atrophy, saddle nose, frontal bossing, disproportionate shortening of the proximal extremities, psychomotor retardation, failure to thrive, and spasticity; X-ray shows stippled humeral head epiphysis.
Of the following, the **MOST** likely diagnosis is
A. chondrodysplasia punctate
B. keratitis with ichthyosis and deafness (KID syndrome)
C. Chanarin-Dorfman syndrome
D. Papillon-Lefèvre Syndrome
E. Richner-Hanhart syndrome

58. A 15-year-old diabetic female presents with irregularly shaped, sharply demarcated, yellow, sclerotic plaques with central telangiectasia and a violaceous border located on the shins. These skin lesions started as erythematous papules 3 months ago.
Of the following, the **MOST** likely diagnosis is
A. necrobiosis lipoidica
B. xanthomas
C. morphea
D. granuloma annulare
E. pretibial myxedema

59. A 7-year-old boy presents with brawny edema of the face and neck that spreads rapidly to involve the thorax and arms. The face has a waxy, mask-like appearance. The involved areas feel indurated, woody, non-pitting, and are not sharply demarcated from normal skin. The overlying skin is normal in color and is not atrophic. The patient gives history of proceeding febrile streptococcal pharyngitis 7 days ago.
Of the following, the **MOST** likely diagnosis is

A. morphea
B. myxedema
C. scleredema of Buschke
D. dermatomyositis
E. subcutaneous fat necrosis

60. What is the classic sign of lipoid proteinosis?

A. Hoarseness
B. Beaded papules on the eyelids
C. Patchy alopecia
D. Enlarged firm tongue
E. Diffusely thickened palms

61. What is the **MOST** common symptom seen in the classic infantile type of urticaria pigmentosa?

A. Intense pruritus
B. Headache
C. Colic
D. Diarrhea
E. Flushing

62. Which of the following may exacerbate mast cell mediator release in patients with mastocytosis?

A. Macrolids
B. Cephalosporins
C. Vancomycin
D. Penicillins
E. Sulphonamides

63. Skin hyperelasticity, joint hypermobility, easy bruising, velvet skin, Joints hypermobility, scoliosis, and mitral valve prolapse are features of

A. Loeys-Dietz syndrome
B. MASS syndrome
C. cutis laxa
D. pseudoxanthoma elasticum
E. classic Ehlers-Danlos Syndrome

64. Which of the following Infectious agents is a common cause of erythema nodosum?

A. Group A streptococcus
B. Yersinia
C. Mycoplasma
D. leprosy
E. Brucellosis

65. A 2-week-old full-term infant, product of difficult vaginal labor with previous admission to NICU for 3 days, presents with asymptomatic indurated, erythematous sharply demarcated plaque on the right cheek.
Of the following, the **MOST** likely diagnosis is

A. sclerema neonatorum
B. panniculitis
C. cellulitis
D. hematoma
E. subcutaneous fat necrosis

66. A 7-week-old infant with perinatal history of difficult vaginal breech delivery, repeated seizures 12 hours after birth controlled by phenobarbitone, developed indurated, erythematous sharply demarcated plaque on the right shoulder, presents with lethargy, poor feeding, vomiting, failure to thrive, irritability, and seizure, electrocardiography reveals shortening of the QT interval.
Of the following, the **MOST** appropriate investigation is

A. lumbar puncture
B. blood glucose
C. serum calcium
D. blood culture
E. ultrasound brain

67. Munchausen syndrome by proxy should be considered in young children in which of the following conditions?

A. Factitial panniculitis
B. Pernio
C. Cold urticarial
D. Cold panniculitis
E. Buccal cellulitis

68. A 10-year-old diabetic girl on insulin presents with thin emaciated extremities and gluteal region with normal appearance of face, neck, and trunk together with golden yellow papules on both elbows. Blood tests reveal hyperlipidemia.
Of the following, the **MOST** likely diagnosis is

A. Type I familial partial lipodystrophy
B. Type 2 familial partial lipodystrophy
C. Type 3 familial partial lipodystrophy
D. Type 4 familial partial lipodystrophy
E. Type 5 familial partial lipodystrophy

69. Which of the following intoxications produces necrosis of sweat glands, resulting in anhidrosis with or without erythema and bullae?

A. Lead
B. Atropine
C. Scopolamine
D. Diazepam
E. Salicylate

70. Which of the following drugs may cause hyperhidrosis?

A. Ciprofloxacin
B. Theophylline
C. Azithromycin
D. Diazepam
E. Valproic acid

71. Which of the following have dramatic response in miliaria?

A. Cooling
B. Antipyretics
C. Topical steroids
D. Topical clindamycin
E. Topical aluminum

72. An adolescent obese heavy smoker female presents with multiple pruritic painful erythematous nodules progressed to deep abscesses then contracted scars confined to axillae, anogenital area, and breasts. These lesions are found for the last 2 years punctuated by relapses and partial remissions with episodic inflammatory arthritis.
Of the following, the **MOST** likely diagnosis is

A. furuncles
B. scrofuloderma
C. actinomycosis
D. hidradenitis suppurativa
E. lymphogranuloma venereum

73. Which of the following drugs causes hypertrichosis?

A. Phenytoin
B. Valproate
C. Phenobarbitone
D. Diazepam
E. Ethosuximide

74. Which of the following conditions may be complicated by trichobezoars?

A. Traction alopecia
B. Hair pulling
C. Trichotillomania
D. Tinea capitis
E. Alopecia areata

75. A 4-month-old infant brought by his young mother with complaint of fussiness and loss of hair from occipital area for the last 2 weeks, the baby is bottle fed, diagnosed to have atopic dermatitis of the face, 1 month ago and on low potent topical steroid.
Of the following, the **MOST** likely cause of this hair loss is

A. anagen effluvium
B. telogen effluvium
C. alopecia ophiasis
D. alopecia areata
E. anhidrotic ectodermal dysplasia

76. Which of the following is a possible etiology of telogen effluvium?
    A. Chemotherapy
    B. Radiation therapy
    C. Lead
    D. Colchicine
    E. discontinuation of high-dose corticosteroids

77. Which of the following cause diffuse milky white nails?
    A. Anemia
    B. Hypocalcemia
    C. Malnutrition
    D. Pellagra
    E. Zinc deficiency

78. Which of the following conditions is characterized by large nails?
    A. Rubinstein-Taybi syndrome
    B. Ectodermal dysplasias
    C. Ellis–van Creveld
    D. Incontinentia pigmenti
    E. Otopalatodigital

79. Which of the following conditions is characterized by pseudoclubbing?
    A. Gardner's syndrome
    B. Maffucci's syndrome
    C. Apert's syndrome
    D. Diamond's syndrome
    E. Pachydermoperiostosis

80. Which of the following conditions is associated with Lindsay nails (half-and-half nails)?
    A. Renal disease
    B. Cirrhosis
    C. congestive heart failure
    D. Hypoalbuminemia
    E. Lichen planus

81. Which of the following diseases is associated with yellow nail syndrome?
    A. Subacute bacterial endocarditis

B. Langerhans cell histiocytosis
C. Evere rheumatoid arthritis
D. Cirrhosis
E. Bronchiectasis

82. Which of the following drugs is a cause of onycholysis?
A. Cyclophosphamide
B. Adriamycin
C. Vincristine
D. Methotrexate
E. 6-mercaptopurine

83. A 10-year-old boy presents with longitudinal ridging, pitting, fragility, thinning, distal notching, and opalescent discoloration of all the nails. The patient has no associated skin or systemic diseases.
Of the following, the **MOST** likely cause of these nail changes is
A. Trachyonychia
B. Onychomycosis
C. Acute paronychia
D. Onycholysis
E. Koilonychia

84. A 5-year-old girl presents with multiple sharply demarcated, irregular, smooth red patches surrounded by an elevated white border on the dorsum of the tongue. The pattern of lesions changes over days and is asymptomatic but may cause a burning sensation to cold foods.
Of the following, the **MOST** likely cause of these tongue changes is
A. black hairy tongue
B. geographic tongue
C. fissured tongue
D. Canker sores
E. oral hair leukoplakia

85. A 5-year-old boy presents with a tiny pustule on the skin of right check after an insect bite, which rapidly progress into a honey-colored crusted plaque 1.5 cm in diameter with little pain, no constitutional symptoms but submandibular adenopathy, CBC reveals WBC 16000/cmm with 70% neutrophils.
Of the following, the **MOST** likely diagnosis is

A. herpes simplex
B. varicella-zoster
C. tinea corporis
D. kerion
E. nonbullous impetigo

86. Which empirical antibiotics are used in neonates with cellulitis?
A. I V vancomycin and gentamicin
B. I V gentamicin and cefotaxime
C. I V ampicillin and gentamycin
D. I V nafcillin and vancomycin
E. I V clindamycin

87. A 2-year-old boy develops cellulitis of left palm after an accidental trauma, he has no fever, lymphadenopathy, or other constitutional signs.
What is the initial antibiotics treatment?
A. clindamycin
B. trimethoprim-sulfamethoxazole
C. cephalexin
D. amoxicillin
E. azithromycin

88. An 18-month-old boy develops malaise, fever, irritability, and exquisite tenderness of the skin followed by diffuse scarlatiniform erythema and purulent conjuctivitis. The skin rapidly acquire a wrinkled appearance with development of diffuse flaccid blisters and erosions with circumoral erythema and radial crusting and fissuring around the eyes, mouth, and nose, followed by large sheets of epidermis peel away.
Of the following, the **MOST** likely diagnosis is
A. bullous impetigo
B. epidermolysis bullosa
C. pemphigus
D. staphylococcal scalded skin syndrome
E. toxic epidermal necrolysis

89. A poorly controlled 10-year-old diabetic boy develops a red macule on the right buttock that vesiculates and then ulcerates with black, depressed, crusted

center and a surrounding rim of raised violaceous edges. CBC reveals severe neutropenia and high HbA_1C.
Of the following, the **MOST** likely cause of this lesion is

A. S. aureus
B. Enterobacter spp.
C. P. aeruginosa
D. Burkholderia cepacia
E. Serratia marcescens

90. What is the **MOST** common cause of blistering distal dactylitis?

A. Group A streptococcus
B. S. aureus
C. H. influenza
D. Salmonella
E. S. epidermidis

91. A 7-year-old boy presents with superficial, erythematous, well marginated, nonindurated, tender, pruritic, and confluent rash from the anus outward. The boy has rectal pain described as burning inside the anus during defecation, with blood-streaked stools. His 5-year-old brother has the same complaint.
Of the following, the **MOST** likely diagnosis is

A. psoriasis
B. seborrheic dermatitis
C. pinworm infestation
D. perianal infectious dermatitis
E. sexual abuse

92. Which of the following furuncles sites may cause intense pain?

A. Face
B. Axillae
C. Buttocks
D. Groin
E. External auditory canal

93. A 10-year-old obese diabetic boy presents with mildly pruritic, sharply demarcated, irregularly bordered, and slightly scaly brownish red patch in right axilla. The diagnosis is readily made on Wood lamp examination.
Of the following, the **MOST** likely diagnosis is

A. erysipeloid
B. erythrasma
C. tinea versicolor
D. seborrheic dermatitis
E. erysipelas

94. A young child develops cervical lymphadenitis in the submandibular region which enlarges over several weeks, ulcerate, and drain. The local reaction is nontender and circumscribed with no constitutional symptoms or evidence of lung or other organ involvement.
Of the following, the **MOST** likely cause is
A. Mycobacterium scrofulaceum
B. syphilitic gumma
C. deep fungal infection
D. actinomycosis
E. hidradenitis suppurativa

95. A malnourished white adolescent male develops reddish brown macules covered with a fine scale enlarged forming non pruritic confluent patches on the neck and upper chest.
Of the following, the **MOST** likely diagnosis is
A. seborrheic dermatitis
B. pityriasis alba
C. tinea versicolor
D. pityriasis rosea
E. secondary syphilis

96. A 6-year-old black boy develops a small papule at the base of a scalp hair follicle then spreads peripherally, forming an erythematous and scaly circular plaque within which the hairs become brittle and broken, with a confluent pruritic patch of alopecia.
Of the following, the **MOST** likely diagnosis is
A. seborrheic dermatitis
B. psoriasis
C. alopecia areata
D. tinea capitis
E. trichotillomania

97. A young boy presents with a dry, mildly erythematous, elevated, scaly plaque spreading centrifugally and clears centrally forming an annular lesion on right antecubital area for the last 2 weeks. The lesion does not fluoresce with a Wood lamp.
Of the following, the **MOST** likely diagnosis is

A. granuloma annulare
B. nummular eczema
C. pityriasis rosea
D. tinea corporis
E. erythema chronicum migrans

98. What is the hallmark of candidal diaper dermatitis?

A. Intensely erythematous confluent plaque
B. Satellite pustules
C. Involvement of inguinal folds
D. Involvement entire scrotum and penis
E. Involvement the vaginal mucosa and labia

99. Which of the following is key feature uniformly observed in kwashiorkor?

A. Moon facies
B. Diffuse fine reddish brown scaling
C. Linear fissures and erosions
D. Thin soft nails
E. Sparse, thin, and depigmented hairs

100. A 3-year-old boy presents with 4 discrete, pearly, skin-colored, dome-shaped, papules, with central umbilication 2 to 4 mm in diameter on the right check.
Of the following, the **MOST** likely diagnosis is

A. ectopic sebaceous glands
B. keratoacanthoma
C. juvenile xanthogranulomas
D. molluscum contagiosum
E. warty dyskeratoma

101. What is the classic lesion of scabies?

A. Excoriated red papules
B. Threadlike burrows

C. Bullae
D. Wheals
E. Vesicles

102. A 7-year-old boy presents with intense generalized pruritus, worse at night; inflammatory pruritic excoriated papules localized to flexor aspects of wrists, axillae, and buttocks.
Of the following, the **MOST** likely diagnosis is
A. dermatitis herpetiformis
B. eczema
C. pediculosis corporis
D. pityriasis rosea
E. scabies vulgaris

103. What is the treatment of choice for scabies?
A. Permethrin 5% cream
B. Sulfur ointment 5–10%
C. Crotamiton 10% lotion
D. Lindane 1% lotion
E. Benzoyl benzoate 10–25% lotion

104. What is the treatment of choice for pediculosis capitis?
A. Malathion
B. Spinosad
C. Benzyl alcohol lotion
D. Ivermectin
E. Permethrin

105. Which of the following is recommended as a topical monotherapy for mild acne?
A. Benzoyl peroxide
B. Erythromycin
C. Clindamycin
D. Retinoid
E. Adapalene

106. Which of the following antibiotics is **MOST** effective in the treatment of acne?

A. Tetracycline
B. Doxycycline
C. Erythromycin
D. Azithromycin
E. Amoxicillin

107. Which of the following vitamins can induce acneiform lesions in susceptible individuals?
A. B12
B. E
C. C
D. D
E. A

108. A 2-year-old otherwise normal white boy brought to you because of a firm, dome-shaped, orange papule, 0.7cm in diameter on upper trunk with few café-au-lait macules and normal blood lipid values.
Of the following, the **MOST** likely diagnosis is
A. papulonodular urticarial pigmentosa
B. dermatofibromas
C. xanthomas of hyperlipoproteinemia
D. juvenile xanthogranuloma
E. dermatofibroma

109. A 19-month-old female presents with asymmetric truncal lipomatous mass since birth with macrodactyly, linear epidermal nevus, and scoliosis.
Of the following, the **MOST** likely diagnosis is
A. Proteus syndrome
B. Klippel-Trenaunay syndrome
C. Bannayan-Riley-Ruvalcaba syndrome
D. CLOVES syndrome
E. Gorlin syndrome

110. A 7-month-old boy weaned recently from breast milk to cow's milk due to maternal malignancy, presents with chronic diarrhea, stomatitis, failure to thrive, and cutaneous eruption in the form of vesiculobullous, eczematous, dry, scaly, lesions symmetrically distributed in the perioral, acral, and perineal areas, with reddish sparse hair. He also has conjunctivitis and blepharitis.

Of the following, the **MOST** likely nutrient deficiency is

A. biotin
B. vitamin B2
C. vitamin B6
D. essential fatty acids
E. zinc

Chapter 30

Skin

Answers

ZUHAIR ALMUSAWI

1.(B)

2.(C) Nail growth is relatively slow; complete fingernail regrowth takes 6 mo, while complete toenail regrowth requires 12-18 mo.

3.(E) Malignancy-associated Sweet syndrome is most commonly associated with hematologic malignancies, especially acute myelogenous leukemia . It manifests abruptly before, during, or after the malignancy course and is characterized by tender, erythematous, edematous plaques or nodules that may be pustular or targetoid, often accompanied by fever, anemia, and leukocytosis. Oral ulcers are more common in malignancy-associated Sweet syndrome than in other forms of the disease, and extracutaneous manifestations involving various organ systems may also occur.

4.(D) The majority of medication reactions are mild morbilliform or exanthematous eruptions of little clinical consequence. Identifying the suspect medication may be difficult owing to the many medications used in immunosuppressed patients. Features that may help identify suspect medications include rash onset relative to exposure, character of distribution and spread, associated symptoms, and laboratory data. Medication eruptions begin on the trunk 7-10 days after exposure; they spread peripherally and are associated with pruritus and, less commonly, with fever, arthralgia, and lymphadenopathy. Eosinophilia may support a diagnosis of drug eruption but may be absent in the setting of bone marrow suppression. Penicillins, sulfa drugs, cephalosporins, nonsteroidal antiinflammatory drugs, anticonvulsants, and aminoglycosides are common offenders. Medication eruptions may resolve despite continued use of the offending agent, or they may progress to more severe involvement.

5.(B)

6.(C) Drug rash with eosinophilia and systemic symptoms (DRESS syndrome) It is classically seen 2-6 wk after initial exposure to an anticonvulsant (carbamazepine, phenobarbital, phenytoin, lamotrigine) or other drugs

(allopurinol, minocycline, sulfonamides [dapsone, sulfasalazine], other antibiotics) and often manifests as the triad of fever, rash, and hepatitis.

7.(B) Offending medications include sulfonamides, tetracyclines, nonsteroidal antiinflammatory drugs (NSAIDs), and acetaminophen.

8.(E) Acute generalized exanthematous pustulosis is often drug-related (most commonly aminopenicillins, macrolides, sulfonamides), occurring within hours to days after drug exposure. It is characterized by many nonfollicular sterile pustules with underlying edema and erythema, typically beginning on the face and intertriginous regions. Neutrophilia and fever are common, whereas eosinophilia is less common than in DRESS syndrome. The rash may burn or itch; mucous membrane involvement is rare and often mild. Internal organ involvement is not common and often is asymptomatic. A pustular smear is always indicated to rule out infection in the setting of leukocytosis, fever, and a pustular rash. Therapy consists of stopping the causative drug and offering symptomatic relief with moist dressings, emollients, and mid-potency topical corticosteroids (applied twice daily for 1 wk).

9.(C) Acute weeping lesions respond best to wet compresses, followed by lotions or creams. For dry, thickened, scaly skin, or for treatment of a contact allergic reaction possibly the consequence of a component of a topical medication, an ointment base is preferable, as it helps to occlude and moisten the affected area. Gels and solutions are most useful for the scalp and other hairy areas because of their faster absorption.

10.(C) The sensitizing potential of certain topical antibiotics, such as neomycin and nitrofurazone, should be kept in mind and avoided when possible. Mupirocin, fusidic acid, and retapamulin are the most effective topical agents currently available and are as effective as oral erythromycin in treatment of mild to moderate impetigo. Polysporin and bacitracin are not as effective.

11.(A) Corticosteroids can be divided into 7 different categories on the basis of strength, but for practical purposes, 4 categories can be used: low, moderate, high, and super. Low-potency preparations include hydrocortisone, desonide, and hydrocortisone butyrate. Medium-potency compounds include amcinonide, betamethasone, flurandrenolide, fluocinolone, mometasone furoate, and triamcinolone. High-potency topical steroids include fluocinonide and halcinonide. Betamethasone dipropionate and clobetasol propionate are superpotent preparations and should be prescribed with care.

12.(E) A benign, self-limited, evanescent eruption, erythema toxicum occurs in approximately 50% of full-term infants; preterm infants are affected less commonly. The lesions are firm, yellow-white, 1-2 mm papules or pustules with

a surrounding erythematous flare. At times, splotchy erythema is the only manifestation. Lesions may be sparse or numerous and either clustered in several sites or widely dispersed over much of the body surface. The palms and soles are usually spared. Peak incidence occurs on the 2nd day of life, but new lesions may erupt during the first few days as the rash waxes and wanes. Onset may occasionally be delayed for a few days to weeks in premature infants.

13.(A) In hypohidrotic ectodermal dysplasia, affected patients are unable to sweat and may experience episodes of high fever in warm environments, which may be mistakenly considered to be fevers of unknown origin. This error is particularly common in infancy, when the facial changes are not easily appreciated. Diagnosis at this time may be made using the starch-iodine test or palmar or scalp biopsy. Scalp biopsy is the most sensitive and is 100% specific.

14.(A) The most effective treatment for capillary malformations is with the pulsed-dye laser. This therapy is targeted to hemoglobin within the lesion and avoids thermal injury to the surrounding normal tissue. After such treatment, the texture and pigmentation of the skin are generally normal without scarring. Therapy can begin in infancy, when the surface area of involvement is smaller. There may be advantages to treating within the 1st year of life. Although this approach is quite effective, redarkening of the stain may occur over time, making ongoing treatments useful. Masking cosmetics may also be used.

15.(A) The course of a particular lesion is unpredictable, but approximately 60% of these lesions reach maximal involution by 5 yr of age, and 90–95% by 9 yr. Spontaneous involution cannot be correlated with size or site of involvement, but lip lesions seem to persist most often.

16.(C) In a disfiguring, life- or vision-threatening, or ulcerated IH that is not responding to other treatment, oral propranolol is the first-line treatment.

17.(E) Increased propranolol levels occur with inhibitors of CYP2D6 (cimetidine, amiodarone, fluoxetine, quinidine, ritonavir) and CPY1A2 (cimetidine, ciprofloxacin, isoniazid, ritonavir, theophylline); decreased blood levels occur with inducers of hepatic drug metabolism (rifampin, phenytoin, phenobarbital).

18.(B) Kasabach-Merritt Phenomenon is a life-threatening combination of a rapidly enlarging Kaposiform hemangioendothelioma (KHE), thrombocytopenia, microangiopathic hemolytic anemia, and an acute or chronic consumption coagulopathy. The clinical manifestations are usually evident during early infancy. The vascular lesion is usually cutaneous and is only rarely located in viscera. The associated thrombocytopenia may lead to precipitous hemorrhage accompanied by ecchymoses, petechiae, and a rapid increase in the size of the vascular lesion. Severe anemia from hemorrhage or microangiopathic hemolysis

may ensue. Treatment includes surgical excision of small lesions, although this is often difficult because of coagulopathy. Additional pharmacologic treatments include systemic steroids with or without vincristine as first-line therapy in most cases.

19.(C) A pyogenic granulomas (PG) are relatively common in children, particularly on the face, arms, and hands. Such a lesion located on a finger or hand may appear as a subcutaneous nodule. PGs may arise at sites of injury, but a history of trauma often cannot be elicited.

20.(A) Angiokeratoma Corporis Diffusum (Fabry Disease) is an inborn error of glycolipid metabolism (α-galactosidase), angiokeratoma corporis diffusum is an X-linked recessive disorder that is fully penetrant in males and is of variable penetrance in carrier females. Angiokeratomas appear before puberty and occur in profusion over the genitalia, hips, buttocks, and thighs and in the umbilical and inguinal regions. They consist of 0.1-3.0 mm red to blue-black papules that may have a hyperkeratotic surface. Telangiectasias are seen in the mucosa and conjunctiva. Additional clinical manifestations include recurrent episodes of fever and agonizing pain, cyanosis and flushing of the acral limb areas, paresthesias of the hands and feet, corneal opacities detectable on slit-lamp examination, and hypohidrosis. Renal involvement and cardiac involvement are the usual causes of death.

21.(C) The Peutz-Jeghers syndrome is characterized by melanotic macules on the lips and mucous membranes and by gastrointestinal (GI) polyposis. It is inherited as an autosomal dominant trait (STK11 gene). Onset is noted in infancy and early childhood when pigmented macules appear on the lips and buccal mucosa. The macules are usually a few millimeters in size but may be as large as 1-2 cm. Macules also occasionally appear on the palate, gums, tongue, and vaginal mucosa. Cutaneous lesions may develop on the nose, hands, and feet; around the mouth, eyes, and umbilicus; and as longitudinal bands or diffuse hyperpigmentation of the nails. Polyposis usually involves the jejunum and ileum but may also occur in the stomach, duodenum, colon, and rectum. Episodic abdominal pain, diarrhea, melena, and intussusception are frequent complications.

22.(E) Strong association

- ✓ Neurofibromatosis type 2
- ✓ Multiple familial café-au-lait
- ✓ Legius (NF-1–like) syndrome
- ✓ McCune-Albright syndrome
- ✓ Mismatch repair cancer syndrome

- ✓ Ring chromosome syndromes
- ✓ LEOPARD/multiple lentigines syndrome
- ✓ Cowden syndrome (multiple hamartoma syndrome)
- ✓ Bannayan-Riley- Ruvalcaba syndrome

23.(C)

24.(B) OCA2 ranges from nearly normal to closely resembling type 1 albinism. This is the most common form of albinism seen worldwide. Little or no melanin is present at birth, but pigment, particularly red-yellow pigment, may accumulate during childhood to produce straw-colored or light brown skin in white individuals. Pigmented nevi may develop. Progressive improvement in visual acuity and nystagmus occurs with aging.

25.(B)

26.(A) A congenital autosomal dominant disorder, piebaldism is characterized by sharply demarcated amelanotic patches that occur most frequently on the forehead, anterior scalp (producing a white forelock), ventral trunk, elbows, and knees. Islands of normal or darker-than-normal pigmentation may be present within the amelanotic areas.

27.(A) Waardenburg syndrome also manifests at birth as localized areas of depigmented skin and hair. There are four types of Waardenburg syndrome. The hallmark of Waardenburg type 1 is the white forelock, which is seen in 20–60% of patients. Only 15% of patients have areas of depigmented skin. Deafness occurs in 9–37%, heterochromia irides in 20%, and unibrow (synophrys) in 17–69% of those affected. Dystopia canthorum (i.e., telecanthus) is seen in all patients with Waardenburg type 1.

28.(B) Hypomelanosis of Ito is a rare congenital skin disorder affecting children of both sexes that can have associated defects in several organ systems. Hypomelanosis of Ito is currently a descriptive rather than definitive diagnosis. The skin lesions of hypomelanosis of Ito are generally present at birth but may be acquired in the first 2 yr of life. The hypopigmentation remains unchanged throughout childhood but fades during adulthood.

29.(A) Localized areas of vitiligo may respond to potent topical steroid, topical tacrolimus, or topical pimecrolimus. In patients with more extensive involvement, narrow-band ultraviolet light B (UVB) [UVB311] is the treatment of choice.

30.(B) Among the numerous factors implicated in the etiology of erythema multiforme (EM), infection with herpes simplex virus (HSV) is the most common.

31.(D) EM is characterized by an abrupt, symmetric cutaneous eruption, most commonly on the extensor upper extremities; lesions are relatively sparse on

the face, trunk, and legs. Lesions can be seen on the palms and soles. The eruption often appears initially as red macules or urticarial plaques that expand centrifugally to form lesions up to 2 cm in diameter with a dusky to necrotic center.

32.(A) Pain from mucosal ulceration is often severe, but skin tenderness is minimal to absent in SJS, in contrast to pain in TEN.

33.(C) IV immunoglobulin (IVIG; 1.5-2.0 g/kg/day × 3 days) should be considered in early disease. Total dose >2 g/kg has shown improved but not statistically significant outcomes in children compared with adults.

34.(B) EBS–localized (formerly Weber-Cockayne) predominantly affects the hands and feet and often manifests when a child begins to walk; onset may be delayed until puberty or early adulthood, when heavy shoes are worn or the feet are subjected to increased trauma. Bullae are usually restricted to the hands and feet; rarely, they occur elsewhere, such as the dorsal aspect of the arms and the shins. The disorder ranges from mildly incapacitating to crippling at times of severe exacerbations.

35.(D)

36.(A) The disease is best treated initially with systemic methylprednisolone 1-2 mg/kg/day. Azathioprine, cyclophosphamide, mycophenolate mofetil, and methotrexate therapy all have been useful in maintenance regimens. IVIG given in cycles may be beneficial to patients whose disease does not respond to steroids. Rituximab with IVIG replacement has been effective in the management of severe pemphigus.

37.(C)

38.(D)

39.(C) First-line therapy for diaper dermatitis is application of a protective barrier agent (ointment or paste) containing petroleum or zinc oxide at every diaper change.

40.(D) Nickel dermatitis develops from contact with jewelry, metal closures on clothing, or even cell phones. Some children are exquisitely sensitive to nickel, with even the trace amounts found in gold jewelry provoking eruptions. The most frequently involved sites from jewelry are the earlobes from nickel containing earrings. Early ear piercing increases risk of sensitization, and it is recommended to delay piercing until after 10 yr of age.

41.(A) Neomycin sulfate is present in many nonprescription topical antibiotic preparations, and thus children are frequently exposed at an early age.

42.(D) Pityriasis alba lesions wax and wane but eventually disappear, and normal pigmentation often takes months to return.

43.(A) Seborrheic dermatitis may begin in the 1st month of life and typically self-resolves by 1yr.

44.(C) Photoallergic drug eruptions:

•Systemic drugs include tetracyclines, psoralens, chlorothiazides, sulfonamides, barbiturates, griseofulvin, thiazides, quinidine, phenothiazines

Phototoxic drug eruptions:

•Systemic agents include nalidixic acid, furosemide, nonsteroidal antiinflammatory agents (naproxen, piroxicam), and high doses of agents causing photoallergic eruptions

45.(C) Xeroderma pigmentosum is a rare autosomal recessive disorder that results from a defect in nucleotide excision repair. Skin changes are first noted during infancy or early childhood in sun-exposed areas though lesions may occur at other sites, including the scalp. The skin lesions consist of erythema, scaling, bullae, crusting, ephelides (freckles), telangiectasia, keratoses, basal and squamous cell carcinomas, and malignant melanomas. Interestingly, although most patients experience exaggerated acute sunburn reactions following minimal UV exposure, up to half of affected patients do not and instead develop progressive freckling. This difference in presentation depends on genetic subtype. Ocular manifestations include photophobia, lacrimation, blepharitis, symblepharon, keratitis, corneal opacities, tumors of the lids, and possible eventual blindness. Neurologic abnormalities such as cognitive deterioration and sensorineural deafness develop in approximately 20% of patients.

46.(B) Rothmund-Thomson syndrome is also known poikiloderma congenital because of the striking skin changes. It is inherited as an autosomal recessive trait.

47.(B) Hartnup disease is a rare inborn error of metabolism with autosomal recessive inheritance. Neutral amino acids, including tryptophan, are not transported across the brush border epithelium of the intestine and kidneys due to mutation of the SLC6A19 gene encoding the transporter. Administration of nicotinamide and protection from sunlight results in improvement of both cutaneous and neurologic manifestations.

48.(E) Guttate psoriasis has also been observed after perianal streptococcal infection, viral infections, sunburn, and withdrawal of systemic corticosteroid therapy or tumor necrosis factor (TNF)-α inhibitors.

49.(E) The first-line topical agents for lesions on the body are emollients, vitamin D analogs (calcipotriene or calcitriol, although calcitriol is less irritating for children), and mid- to high-potency corticosteroids.

50.(E) This benign, common eruption occurs most frequently in children and young adults. Although a prodrome of fever, malaise, arthralgia, and pharyngitis may precede the eruption, children rarely complain of such symptoms. A herald patch classically precedes the generalized eruption and may occur anywhere on the body. Herald patches are generally larger than other lesions and vary from 1 to 10 cm in diameter; they are annular in configuration and have a raised border with fine, adherent scales. Approximately 5-10 days after the appearance of the herald patch, a widespread, symmetric eruption involving mainly the trunk and proximal limbs becomes evident. Lesions may appear in crops for several days.

51.(D) Darier disease usually occurs in late childhood and persists throughout life. Several exacerbating triggers have been identified: sweating, UV light exposure, heat, friction, surgery, and infections; thus, Darier disease has a chronic relapsing course that usually worsens in summertime.

52.(A) Gianotti-Crosti Syndrome (Papular Acrodermatitis),this distinctive eruption is benign and predominantly occurs in children younger than 5 yr old about 1 wk after a viral illness. Cases are usually sporadic, but epidemics have been recorded. Lines of papules (Koebner phenomenon) may be noted on the extremities following minor local trauma. The eruption resolves spontaneously but may take up to 2 mo.

53.(C) The clinical severity and histopathologic features of acanthosis nigricans correlate positively with the degree of hyperinsulinism and with the degree of obesity.

54.(B) Ichthyosis vulgaris is the most common of the disorders of keratinization, with an incidence of 1/250 live births. Onset generally occurs in the 1st yr of life.

55.(B) The clinical picture of Sjögren-Larsson syndrome consists of ichthyosis, cognitive impairment, and spasticity.

56.(C) Refsum syndrome is a multisystem disorder that becomes symptomatic in the 2nd or 3rd decade of life. The ichthyosis may be generalized, is relatively mild, and resembles ichthyosis vulgaris. The ichthyosis may also be localized to the palms and soles. Chronic polyneuritis with progressive paralysis and ataxia, retinitis pigmentosa, anosmia, deafness, bony abnormalities, and electrocardiographic changes are the most characteristic features.

57.(A) These heterogeneous disorders are marked by ichthyosis and bone changes. Nearly all patients with the X-linked dominant form and approximately 25% of those with the recessive type have cutaneous lesions, ranging from severe, generalized erythema and scaling to mild hyperkeratosis. Rhizomelic CPD is associated with cataracts, hypertelorism, optic nerve atrophy, disproportionate shortening of the proximal extremities, psychomotor

retardation, failure to thrive, and spasticity; most affected patients die in infancy.

58.(A) This disorder manifests as erythematous papules that evolve into irregularly shaped, sharply demarcated, yellow, sclerotic plaques with central telangiectasia and a violaceous border. Scaling, crusting, and ulceration are frequent. Lesions develop most commonly on the shins.

59.(C)

60.(B) Lipoid proteinosis may be noted initially in early infancy as hoarseness. Skin lesions appear during childhood and consist of yellowish papules and nodules that may coalesce to form plaques. The classic sign is beaded papules on the eyelids.

61.(E) Systemic signs of histamine release, such as anaphylaxis-like episodes, hypotension, syncope, headache, episodic flushing, tachycardia, wheezing, colic, and diarrhea, are uncommon and occur most frequently in the more severe types of mastocytosis. Flushing is by far the most common symptom seen.

62.(C) Pharmacologic Agents and Physical Stimuli That May Exacerbate Mast Cell Mediator Release in Patients With Mastocytosis

Immunologic Stimuli

- ✓ Venoms (immunoglobulin E–mediated bee venom)
- ✓ Complement-derived anaphylatoxins
- ✓ Biologic peptides (substance P, somatostatin)
- ✓ Polymers (dextran)

Nonimmunologic Stimuli

- ✓ Physical stimuli (heat, cold, rubbing, trauma, sunlight)
- ✓ Acetylsalicylic acid and related nonsteroidal analgesics*
- ✓ Thiamine
- ✓ Ketorolac tromethamine
- ✓ Alcohol
- ✓ Vancomycin
- ✓ Dextromethorphan
- ✓ Narcotics (codeine, morphine)*
- ✓ Radiographic dyes (iodine containing)

Emotional Issues

- ✓ Anxiety
- ✓ Sleep deprivation
- ✓ Stress

63.(E) EDS represents a portion of the hereditary connective tissue disorders, many of which have unique features that enable clinical differentiation. The

primary differential diagnosis would include Loeys-Dietz syndrome, which has features of both vEDS and Marfan syndrome. EDS has also been confused with MASS syndrome (mitral valve prolapse, aortic root dilation, skeletal changes, skin changes), cutis laxa, and pseudoxanthoma elasticum. In general the skin of patients with cutis laxa hangs in redundant folds, whereas the skin of those with EDS is hyperextensible and snaps back into place when stretched. Other disorders that impact the integrity of the connective tissues—such as exposure to corticosteroids and osteogenesis imperfecta or mild myopathic disorders (Bethlem myopathy, Ullrich congenital muscular dystrophy)—can be indistinguishable in the early stages of disease.

64.(A) Etiology of Erythema Nodosum

Viruses

Epstein-Barr, hepatitis B, mumps

Fungi

Coccidioidomycosis, histoplasmosis, blastomycosis, sporotrichosis

Bacteria and Other Infectious Agents

Group A streptococcus,* tuberculosis,* Yersinia, cat-scratch disease, leprosy, leptospirosis, tularemia, mycoplasma, Whipple disease, lymphogranuloma venereum, psittacosis, brucellosis

Other

Sarcoidosis, inflammatory bowel disease,* estrogen-containing oral contraceptives,* systemic lupus erythematosus, Behçet syndrome, severe acne, Hodgkin disease, lymphoma, sulfonamides, bromides, echinacea, Sweet syndrome, pregnancy, idiopathic*

65.(E) The cause of subcutaneous fat necrosis (SCFN) is unknown. The disease in infants may be a result of ischemic injury from various perinatal complications, such as maternal preeclampsia, birth trauma, asphyxia, and prolonged hypothermia. Whole-body cooling for neonatal encephalopathy is increasingly associated with SCFN.

66.(C) A rare but potentially serious complication of subcutaneous fat necrosis (SCFN) is hypercalcemia .

67.(A) Treatment of factitial panniculitis must address the primary reason for the performance of this self-destructive act. Munchausen syndrome by proxy should be considered in young children.

68.(A)

69.(D) At the level of the sweat gland, anticholinergics (drugs such as atropine and scopolamine) may paralyze the sweat glands. Acute intoxication with

barbiturates or diazepam has produced necrosis of sweat glands, resulting in anhidrosis with or without erythema and bullae.

70.**(A)** Causes of Hyperhidrosis

Cortical

- ✓ Emotional
- ✓ Familial dysautonomia
- ✓ Congenital ichthyosiform erythroderma
- ✓ Epidermolysis bullosa
- ✓ Nail-patella syndrome
- ✓ Jadassohn-Lewandowsky syndrome
- ✓ Pachyonychia congenita
- ✓ Palmoplantar keratoderma
- ✓ Stroke

Hypothalamic

Drugs:

- ✓ Alcohol
- ✓ Antipyretics
- ✓ Cocaine
- ✓ Emetics
- ✓ Insulin
- ✓ Opiates (including withdrawal)
- ✓ Ciprofloxacin
- ✓ Exercise

Infection:

- ✓ Defervescence
- ✓ Chronic illness

Metabolic:

- ✓ Carcinoid syndrome
- ✓ Debility
- ✓ Diabetes mellitus
- ✓ Hyperpituitarism
- ✓ Hyperthyroidism
- ✓ Hypoglycemia
- ✓ Obesity
- ✓ Pheochromocytoma
- ✓ Porphyria
- ✓ Pregnancy
- ✓ Rickets

- ✓ Infantile scurvy

Cardiovascular:

- ✓ Heart failure
- ✓ Shock
- ✓ Vasomotor
- ✓ Cold injury
- ✓ Raynaud phenomenon
- ✓ Rheumatoid arthritis

Neurologic:

- ✓ Abscess
- ✓ Familial dysautonomia
- ✓ Postencephalitic
- ✓ Tumor

Miscellaneous:

- ✓ Chédiak-Higashi syndrome
- ✓ Compensatory
- ✓ Lymphoma
- ✓ Phenylketonuria
- ✓ Vitiligo
- ✓ Frey syndrome

Medullary

- ✓ Physiologic gustatory sweating
- ✓ Encephalitis
- ✓ Granulosis rubra nasi
- ✓ Syringomyelia
- ✓ Thoracic sympathetic trunk injury

Spinal

- ✓ Cord transection
- ✓ Syringomyelia

Changes in Blood Flow

- ✓ Maffucci syndrome
- ✓ Arteriovenous fistula
- ✓ Klippel-Trénaunay syndrome
- ✓ Glomus tumor
- ✓ Blue rubber-bleb nevus syndrome

71.(A) All forms of miliaria respond dramatically to cooling of the patient by regulation of environmental temperatures and by removal of excessive clothing;

administration of antipyretics is also beneficial to patients with fever. Topical agents are usually ineffective and may exacerbate the eruption.

72.(D)

73.(A) Drugs causes hypertrichosis: Diazoxide, phenytoin, corticosteroids, cortisporin, cyclosporine, androgens, anabolic agents, hexachlorobenzene, minoxidil, psoralens, penicillamine, and streptomycin.

74.(C) Trichophagy, resulting in trichobezoars, may complicate trichotillomania.

75.(B) Telogen effluvium accounts for the loss of hair by infants in the 1st few months of life; friction from bed sheets, particularly in infants with pruritic, atopic skin, may exacerbate the problem. There is no inflammatory reaction; the hair follicles remain intact, and telogen bulbs can be demonstrated microscopically on shed hairs. Because >50% of the scalp hair is rarely involved, alopecia is usually not severe. Parents should be reassured that normal hair growth will return within approximately 3-6 mo.

76.(E) Telogen effluvium manifests as sudden loss of large amounts of hair, often with brushing, combing, and washing of hair. Diffuse loss of scalp hair occurs from premature conversion of growing, or anagen, hairs, which normally constitute 80–90% of hairs, to resting, or telogen, hairs. Hair loss is noted 6 wk to 3 mo after the precipitating cause, which may include childbirth; a febrile episode; surgery; acute blood loss, including blood donation; sudden severe weight loss; discontinuation of high-dose corticosteroids or oral contraceptives; hypo- or hyperthyroidism; and psychiatric stress.

77.(D) White Nail or Nail Bed Changes

DISEASE	**CLINICAL APPEARANCE**
Anemia	Diffuse white
Arsenic	Mees lines:transverse white lines
Cirrhosis	Terry nails:most of nail, zone of pink at distal end
Darier disease	Longitudinal white streaks
Half-and-half nail	Proximal white, distal pink azotemia
High fevers (some diseases)	Transverse white lines
Hypoalbuminemia	Muehrcke lines: stationary paired transverse bands
Hypocalcemia	Variable white
Malnutrition	Diffuse white
Pellagra	Diffuse milky white
Punctate leukonychia	Common white spots
Tinea and yeast	Variable patterns
Thallium toxicity(rat poison)	Variable white
Trauma	Repeated manicure: transverse striations

Zinc deficiency Diffuse white

78.(A) Large nails seen in

- ✓ Pachyonychia congenita
- ✓ Rubinstein-Taybi syndrome
- ✓ hemihypertrophy

79.(C) Pseudoclubbing is seen in

- ✓ Apert's syndrome
- ✓ Pfeiffer's syndrome
- ✓ Rubinstein-Taybi syndrome

80.(A) When the proximal portion of the nail is white and the distal 20–50% of the nail is red, pink, or brown, the condition is called half-and-half nails or Lindsay nails; this is seen most commonly in patients with renal disease but may occur as a normal variant.

81.(E) Yellow nail syndrome manifests as thickened, excessively curved, slow growing yellow nails without lunulae. All nails are affected in most cases. Associated systemic diseases include bronchiectasis, recurrent bronchitis, chylothorax, and focal edema of the limbs and face. Deficient lymphatic drainage, caused by hypoplastic lymphatic vessels, is believed to lead to the manifestations of this syndrome.

Splinter hemorrhages most often result from minor trauma but may also be associated with subacute bacterial endocarditis, vasculitis, Langerhans cell histiocytosis, severe rheumatoid arthritis, peptic ulcer disease, hypertension, chronic glomerulonephritis, cirrhosis, scurvy, trichinosis, malignant neoplasms, and psoriasis.

82.(C) Onycholysis indicates separation of the nail plate from the distal nail bed. Common causes are trauma, long-term exposure to moisture, hyperhidrosis, cosmetics, psoriasis, fungal infection (distal onycholysis), atopic or contact dermatitis, porphyria, drugs (bleomycin, vincristine, retinoid agents, indomethacin, chlorpromazine [Thorazine]), and drug-induced phototoxicity from tetracyclines or chloramphenicol.

83.(A) Trachyonychia is characterized by longitudinal ridging, pitting, fragility, thinning, distal notching, and opalescent discoloration of all the nails. Patients can have no associated skin or systemic diseases and no other ectodermal defects.

84.(B)

85.(E) The differential diagnosis of nonbullous impetigo includes viruses (herpes simplex, varicella-zoster), fungi (tinea corporis, kerion), arthropod bites, and

parasitic infestations (scabies, pediculosis capitis), all of which may become impetiginized.

86.(A) Neonates with cellulits should receive an intravenous antibiotic with a β-lactamase–stable antistaphylococcal antibiotic such as nafcillin, cefazolin, or vancomycin, and an aminoglycoside such as gentamicin or a 3rd-generation cephalosporin such as cefotaxime.

87.(C) In infants and children older than 2 mo with mild to moderate infections, particularly if fever, lymphadenopathy, and other constitutional signs are absent, treatment of cellulitis may be initiated orally on an outpatient basis with a penicillinase-resistant penicillin such as dicloxacillin or a 1st-generation cephalosporin such as cephalexin or, if MRSA is suspected, with clindamycin.

88.(D)

89.(C) Ecthyma gangrenosum is a necrotic ulcer covered with a gray-black eschar. It is usually a sign of P. aeruginosa infection, most often occurring in immunosuppressed patients. Neutropenia is a risk factor for ecthyma gangrenosum.

90.(A) Blistering distal dactylitis is a superficial blistering infection of the volar fat pad on the distal portion of the finger or thumb that typically affects infants and young children. More than 1 finger may be involved, as may the volar surfaces of the proximal phalanges, palms, and toes. Blisters are filled with a watery purulent fluid; polymorphonuclear leukocytes and Gram-positive cocci are identified on Gram stain.

91.(D) Perianal infectious dermatitis presents most commonly in boys (70% of cases) between the ages of 6 mo and 10 yr as perianal dermatitis (90% of cases) and pruritus (80% of cases). In boys, the penis may be involved. Fecal retention is a frequent behavioral response to the infection. Although local induration or edema may occur, constitutional symptoms, such as fever, headache, and malaise, are absent, suggesting that subcutaneous involvement, as in cellulitis, is absent. Familial spread of perianal infectious dermatitis is common, particularly when family members bathe together or use the same water.

92.(E) Sites of predilection are the hair-bearing areas on the face, neck, axillae, buttocks, and groin. Pain may be intense if the lesion is situated in an area where the skin is relatively fixed, such as in the external auditory canal or over the nasal cartilages.

93.(B) Erythrasma is a benign chronic superficial infection caused by Corynebacterium minutissimum. Predisposing factors include heat, humidity, obesity, skin maceration, diabetes mellitus, and poor hygiene. Approximately

20% of affected patients have involvement of the toe webs. Other frequently affected sites are moist, intertriginous areas such as the groin and axillae.

94.(A) Mycobacterium scrofulaceum causes cervical lymphadenitis (scrofuloderma) in young children, typically in the submandibular region. Nodes enlarge over several weeks, ulcerate, and drain. The local reaction is nontender and circumscribed, constitutional symptoms are absent, and there generally is no evidence of lung or other organ involvement. Other atypical mycobacteria may cause a similar presentation, including Mycobacterium avium complex, Mycobacterium kansasii, and Mycobacterium fortuitum. Treatment is accomplished by excision and administration of antituberculous drugs.

95.(C) The lesions of tinea versicolor vary widely in color. In white individuals, they are typically reddish brown, whereas in black individuals they may be either hypopigmented or hyperpigmented. The characteristic macules are covered with a fine scale. They often begin in a perifollicular location, enlarge, and merge to form confluent patches, most commonly on the neck, upper chest, back, and upper arms.

96.(D) Tinea capitis can be confused with seborrheic dermatitis, psoriasis, alopecia areata, trichotillomania, and certain dystrophic hair disorders. When inflammation is pronounced, as in kerion, primary or secondary bacterial infection must also be considered.

97.(D) Tinea corporis, defined as infection of the glabrous skin, excluding the palms, soles, and groin, can be caused by most of the dermatophyte species, although T. rubrum and Trichophyton mentagrophytes are the most prevalent etiologic organisms. In children, infections with M. canis are also common. Tinea corporis can be acquired by direct contact with infected persons or by contact with infected scales or hairs deposited on environmental surfaces. M. canis infections are usually acquired from infected pets.

98.(B) Satellite pustules, those that stud the contiguous skin, are a hallmark of localized candida infections.

99.(A) The cutaneous manifestations may closely resemble those of acrodermatitis enteropathica; however, edema of the extremities and face (“moon facies”) and a protuberant abdomen (“pot belly”) are key features uniformly observed in kwashiorkor.

100.(D) Molluscum contagiosum are discrete, pearly, skin-colored, smooth, dome-shaped, papules vary in size from 1 to 5 mm. They typically have a central umbilication from which a plug of cheesy material can be expressed. The papules may occur anywhere on the body, but the face, eyelids, neck, axillae, and thighs are sites of predilection. They may be found in clusters on the

genitals or in the groin of adolescents and may be associated with other venereal diseases in sexually active individuals. Lesions commonly involve the genital area in children but are not acquired by sexual transmission in most cases.

101.(B) In an immunocompetent host, scabies is frequently heralded by intense pruritus, particularly at night. The first sign of the infestation often consists of 1-2 mm red papules, some of which are excoriated, crusted, or scaling. Threadlike burrows are the classic lesion of scabies but may not be seen in infants. In infants, bullae and pustules are relatively common. The eruption may also include wheals, papules, vesicles, and a superimposed eczematous dermatitis.

102.(E) Clinical manifestations of classic scabies (scabies vulgaris) include: Intense generalized pruritus, worse at night; inflammatory pruritic papules localized to finger webs, flexor aspects of wrists, elbows, axillae, buttocks, genitalia, female breasts; lesions and pruritus spare the face, head, and neck; secondary lesions include eczematization, excoriation, and impetigo.

103.(A) The treatment of choice for scabies is permethrin 5% cream (Elimite) applied to the entire body from the neck down, with particular attention to intensely involved areas. Scabies is frequently found above the neck in infants (younger than 2 yr old), necessitating treatment of the scalp. The medication is left on the skin for 8-12 hr and should be reapplied in 1 wk for another 8-12 hr period.

104.(A) Malathion 0.5% in isopropanol is the treatment of choice and should be applied to dry hair until hair and scalp are wet, and left on for 12 hr. A second application, 7-9 days after the initial treatment may be necessary. This product is flammable, so care should be taken to avoid open flames.

105.(A) Benzoyl peroxide or combinations with erythromycin or clindamycin are effective acne treatments and are recommended as monotherapy for mild acne, or in conjunction with a topical retinoid, or systemic antibiotic therapy for moderate to severe acne.

106.(B) Doxycycline and minocycline are more effective than tetracycline. Although oral erythromycin or azithromycin can be effective in treating acne, its use should be limited to those who cannot use the tetracyclines (i.e., pregnant women or children <8 yr of age). Erythromycin use should be restricted because of its increased risk of bacterial resistance.

107.(A) Drugs that can induce acneiform lesions in susceptible individuals include isoniazid, phenytoin, phenobarbital, trimethadione, lithium carbonate, androgens (anabolic steroids), and vitamin B12.

108.(D) Xanthogranulomatous infiltrates occur occasionally in ocular tissues. This process may result in glaucoma, hyphema, uveitis, heterochromia iridis, iritis, or sudden proptosis. When seen in patients <2 yr of age, multiple lesions and periocular location may heighten concerns for intraocular involvement. There appears to be an association among juvenile xanthogranuloma, neurofibromatosis, and childhood leukemia, most frequently juvenile chronic myelogenous leukemia. There is no need to remove the benign lesions of juvenile xanthogranuloma because most of them regress spontaneously in the first few years. Residual dyspigmentation and atrophy may result.

109.(D) CLOVES syndrome (congenital lipomatous overgrowth, vascular malformations, epidermal nevi, and scoliosis/skeletal and spinal anomalies) is usually a sporadic disorder caused by a mutation in the PIK3CA gene with an asymmetric truncal lipomatous mass present at birth. Additional features include macrodactyly, vascular malformations (low flow), linear epidermal nevus, and renal anomalies.

110.(E) Acrodermatitis enteropathica is a rare autosomal recessive disorder caused by an inability to absorb sufficient zinc from the diet. The genetic defect is in the intestinal zinc-specific transporter gene SLC39A4. Initial signs and symptoms usually occur in the 1st few months of life, often after weaning from breast milk to cow's milk. The cutaneous eruption consists of vesiculobullous, eczematous, dry, scaly, or psoriasiform skin lesions symmetrically distributed in the perioral, acral, and perineal areas and on the cheeks, knees, and elbows. The hair often has a peculiar, reddish tint, and alopecia of some degree is characteristic. Ocular manifestations include photophobia, conjunctivitis, blepharitis, and corneal dystrophy detectable by slit-lamp examination. Associated manifestations include chronic diarrhea, stomatitis, glossitis, paronychia, nail dystrophy, growth retardation, irritability, delayed wound healing, intercurrent bacterial infections, and superinfection with Candida albicans.

Chapter 31

Bone and Joint Disorders Questions

ZUHAIR M. ALMUSAWI

1. What is approximate ratio of head height to total height at birth?
   A. 1:3
   B. 1:4
   C. 1:5
   D. 1:6
   E. 1:7

2. By what age, birth height usually doubles?
   A. 3 yr
   B. 4 yr
   C. 5 yr
   D. 6 yr
   E. 7 yr

3. Which of the following is a cause of painless limp in a 2-year-old toddler?
   A. Septic arthritis
   B. Transient synovitis
   C. Intervertebral diskitis
   D. Malignancy
   E. Cerebral palsy

4. Which of the following imaging modalities is **BEST** for diagnosis of early septic arthritis?
   A. Bone scan
   B. Computed Tomography
   C. Plain X- ray
   D. Ultrasonography
   E. Magnetic Resonance Imaging

5. Which of the following congenital foot deformities has the highest association with developmental dysplasia of the hip?

A. Metatarsus adductus
B. Calcaneovalgus foot
C. Congenital talipes equinovarus
D. Congenital vertical talus
E. Flatfoot

6. A 2-year-old toddler has a normal longitudinal foot arch when examined in a non–weight-bearing position or standing on the toes, but the arch disappears when standing flat.
Of the following, the **MOST** likely diagnosis is

A. Hypermobile pes planus
B. Tarsal coalition
C. Cavus foot
D. Persian slipper foot
E. Rocker-bottom foot

7. What is the **MOST** likely cause of unilateral cavus foot?

A. Tethered cord
B. Myelodysplasia
C. Occult intraspinal anomaly
D. Charcot-Marie-Tooth
E. Refsum disease

8. What is the **MOST** common cause of heel pain in children?

A. Iselin disease
B. Köhler disease
C. Freiberg infraction
D. Sever disease
E. Pes cavus

9. What is the proper time for removal of well-formed extra digit of foot?

A. At birth
B. Between 3 and 6 mo of age
C. Between 6and 9 mo of age
D. Between 9 and 12 mo of age
E. Between 12 and 18 mo of age

10. A 4-year-old boy brought by his mother because of flexion at the proximal interphalangeal joint of the 2nd toe with hyperextended metatarsal-phalangeal joint and a painful callus over the dorsum of the toe.
Of the following, the **MOST** likely diagnosis is

A. mallet toe
B. hammer toe
C. claw toe
D. macrodactyly
E. subungual exostosis

11. Which of the following is the **MOST** common cause of foot pain in an 8-year-old child?

A. Poorly fitting shoes
B. Foreign body
C. Osteomyelitis
D. Plantar fasciitis
E. Dactylitis

12. A young parents brought their 2-year-old 1st boy concerned with his "bowed" appearance of legs and in-toeing gait. On examination he had also congenital metatarsus varus.
Of the following, the **MOST** likely cause of his gait is

A. femoral retroversion
B. external femoral torsion
C. slipped capital femoral epiphysis
D. medial (internal) tibial torsion
E. active rickets

13. An 18-month-old boy brought by his concerned young mother because of his bow legs, on examination in recumbent position, legs do not appear bowed, but in erect position during weight bearing and with ankles in apposition, the legs are bowed.
Of the following, the **MOST** likely cause of boy's bow legs is

A. Blount disease
B. nutritional rickets
C. hypophosphatasia
D. physiologic bowleg
E. metaphyseal dysplasia

14. Which of the following is a secondary form of osteoporosis in children?
    A. Gaucher disease
    B. Osteogenesis imperfecta
    C. Ehlers-Danlos syndrome
    D. Marfan syndrome
    E. Homocystinuria

15. The appearance of symmetric bilateral genu valgum as part of the normal physiologic process of leg development is most pronounced around the age of
    A. 2 years
    B. 4 years
    C. 6 years
    D. 8 years
    E. 10 years

16. Which of the following conditions causes lower extremity shortening?
    A. Rheumatoid arthritis
    B. Hemarthrosis (hemophilia)
    C. Osteomyelitis
    D. Neurofibromatosis
    E. Tuberculosis

17. A 12-year-old boy presents with a fairly large asymptomatic mass behind the knee which is firm but compressible located medially and distal to the popliteal crease. The mass is usually most prominent when the knee is extended.
Of the following, the only necessary diagnostic test to confirm the diagnosis is knee
    A. CT
    B. radiographs
    C. ultrasonography
    D. MRI
    E. aspiration

18. What is the **BEST** management for the boy in the above scenario at the time being?
    A. Reassurance
    B. Rest and leg elevation
    C. Aspiration

D. Corticosteroid injection
E. Surgical excision

19. A 13-year-old boy presents with anterior knee pain, over the tibial tubercle aggravated by sports activities but may often persist with regular daily activities. Physical examination reveals point tenderness over the tibial tubercle and the distal portion of the patellar tendon. Radiographs reveal fragmentation of the tibial tubercle and soft tissue swelling. There is no acute traumatic inciting event.
Of the following, the **MOST** likely diagnosis is
A. osteochondritis dissecans
B. Osgood-Schlatter disease
C. Sinding-Larsen-Johansson syndrome
D. patellofemoral pain syndrome
E. acute patellofemoral dislocation

20. An adolescent female complains from pain beneath the patella aggravated by walking up and down stairs, and when sitting in a flexed knee position for an extended period of time. Pain is often relieved through knee extension. The onset of symptoms was gradual with no history of trauma. Physical exam reveals tenderness with palpation about the medial aspect of the patella.
Of the following, the **MOST** likely cause of pain is
A. osteochondritis dissecans
B. Osgood-Schlatter disease
C. Sinding-Larsen-Johansson syndrome
D. patellofemoral pain syndrome
E. acute patellofemoral dislocation

21. Which of the following is a risk factor regarding developmental dysplasia of the hip (DDH)?
A. Male gender
B. Cephalic presentation
C. Polyhydramnios
D. Small birth weight
E. First pregnancy

22. What is the **MOST** reliable sign of a dislocated hip in a 4-month-old infant?
A. Limitation of abduction

B. Apparent shortening of the thigh
C. Proximal location of the greater trochanter
D. Asymmetry of the gluteal or thigh folds
E. Hip click

23. What is the presenting complaint of DDH in a walking child?
A. Limp
B. Waddling gait
C. Excessive lordosis
D. Leg-length discrepancy
E. Toe-walks on the affected side

24. A 4- week-old newborn during routine examination found to be Ortolani positive (dislocated but reducible left hip). Ultrasonographic image of the left hip demonstrates an α angle of 42 degree.
Of the following, the **MOST** appropriate treatment at this age is
A. Pavlik harness
B. triple diapers
C. abduction diapers
D. Frejka pillow
E. von Rosen splint

25. A 6-year-old girl presents with pain in the right groin and anterior thigh but she is able to bear weight on the affected limb. She is afebrile with history of nonspecific upper respiratory tract infection 10 days ago. Erythrocyte sedimentation rate, serum C-reactive protein, and white blood cell counts are normal, and Ultrasonography of the hip demonstrates a small joint effusion.
Of the following, the **MOST** likely diagnosis is
A. transient synovitis
B. septic arthritis
C. Legg-Calvé-Perthes disease
D. osteochondromatosis
E. Schwartz-Jampel syndrome

26. Which of the following radiographic signs have been reported to describe a "head at risk" for severe deformity in Legg-Calvé-Perthes disease?
A. Decreased size of the ossification center
B. Lateralization of the femoral head

C. Subchondral fracture
D. Lateral extrusion of the epiphysis
E. Physeal irregularity

27. What is the earliest abnormality noted on physical exam in patients with scoliosis?
A. Posterior rib hump on the convex side of the spinal curve
B. Asymmetry of the posterior chest wall on forward bending
C. Apparent leg-length discrepancy due to pelvic obliquity
D. Shoulder imbalance
E. Lateral shift of the trunk

28. What are the **MOST** common congenital abnormalities identified in children with congenital scoliosis?
A. Cardiac anomalies
B. Intraspinal anomalies
C. Skin tags
D. Genitourinary abnormalities
E. Hemangiomas

29. An adolescent male brought by his father because of kyphosis which can be corrected voluntarily, a standing lateral radiograph was ordered and showed an increase in kyphosis but no pathologic changes of the involved vertebrae. The patient and his father are concerned regarding the cosmetic appearance and ask for treatment.
Of the following, the **MAINSTAY** of treatment is
A. physical therapy
B. bracing
C. surgery
D. reassurance
E. core strengthening

30. A 14- year- old boy presents with hyperkyphosis of the thoracic spine associated with a sharp contour. The boy is unable to correct the deformity voluntarily. He has mild intermittent pain near the apex of the kyphosis with no other neurologic symptoms. Standing lateral radiograph showed wedging of the vertebrae at T6, T7, T8, and T9.
Of the following, the **MOST** likely diagnosis is

A. spondylolisthesis
B. Scheuermann disease
C. postural kyphosis
D. congenital kyphosis
E. spondylolysis

31. Which of the following inflammatory diseases is a common cause of back pain in pediatric age group?
A. Vertebral osteomyelitis
B. Transverse myelitis
C. Diskitis
D. Perinephric abscess
E. Pancreatitis

32. A 14-year-old wrestler presents with low back pain radiate to the buttocks, exaggerated with spinal hyperextension. On examination there is spasm of the hamstring muscles with discomfort on palpation of the spinous process of the lower lumbar vertebrae but no any other neurologic symptoms.
Of the following, the **MOST** likely cause of this back pain is
A. spondylolisthesis
B. Scheuermann syndrome
C. spondylolysis
D. herniated disk
E. overuse syndrome

33. A 4-year-old boy presents with back pain, abdominal pain, fever, and malaise for the last 4 days. He refuses to walk, with local point tenderness over the middle back. When he asked to pick up an object from the ground, the pain increased in intensity.
Of the following, the **MOST** helpful diagnostic test is
A. plain radiograph
B. technetium bone scan
C. MRI
D. Blood culture
E. open biopsy of the disc space

34. What is the **MOST** common organism causing spine infections?
A. Kingella kingae

B. Group A streptococcus
C. Escherichia coli
D. Serratia marcescens.
E. Staphylococcus aureus

35. What is the **BEST** study to establish the diagnosis of a disc herniation?
A. Plain radiograph
B. Technetium bone scan
C. MRI
D. CT scan
E. US

36. A 5- year- old girl brought to hospital by her father complaining from recurrent attacks of urinary tract infections. On examination, a low posterior hairline, short neck, and decreased cervical range of motion were noticed. Radiographs of the cervical spine demonstrate congenital fusion and evidence of spinal instability
Of the following, the **MOST** likely diagnosis is
A. Goldenhar syndrome
B. Mohr syndrome
C. Klippel-Feil syndrome
D. Larsen syndrome
E. Ehlers-Danlos syndrome

37. Which of the following syndromes associated with polydactyly?
A. Apert syndrome
B. Holt-Oram syndrome
C. Trisomy 21
D. Laurence-Moon-Biedl syndrome
E. Ellis-van Creveld syndrome

38. What is the **MOST** commonly fractured bone of the lower limb in children?
A. Fibula
B. Tibia
C. Femur
D. Tibia and fibula
E. Calcaneus

39. A 2- year -old girl presents with a limp, refusal to bear weight, point tenderness at the middle area of tibia, and no history of trauma. Radiographs show no fracture and Inflammatory markers are negative.
Of the following, the **MOST** likely diagnosis is

A. osteomyelitis
B. toddler's fracture of the tibia
C. Osgood-Schlatter syndrome
D. green stick fracture of tibia
E. Tillaux fracture

40. What is the **MOST** common infecting organism in osteomyelitis among all age groups?

A. Group B streptococcus
B. Escherichia coli
C. Staphylococcus aureus
D. Group A streptococcus
E. Pseudomonas aeruginosa

41. Which of the following organisms is difficult to detect in osteomyelitis unless polymerase chain reaction (PCR) testing is used?

A. Kingella kingae
B. Streptococcus pneumoniae
C. Bartonella henselae
D. Group A streptococcus
E. Pseudomonas aeruginosa

42. Which antibiotic is recommended for treatment of osteomyelitis if pneumococcal isolates resistance to penicillin were recovered?

A. Clindamycin
B. Cefotaxime
C. Vancomycin
D. Cefazolin
E. piperacillin-tazobactam

43. What is the **MOST** common joint involved in hematogenous bacterial arthritis?

A. Knee
B. Hip

C. Ankle
D. Elbow
E. Wrist

44. What is the percent of positive blood cultures in cases of proven septic arthritis?

A. 10%
B. 20%
C. 30%
D. 40%
E. 50%

45. A 2-year-old child presents with fever, pain, swelling, erythema of knee joint, with refusal to walk for the last 3 days. Lab. investigations revealed normal leukocyte counts, slightly raised ESR, normal CRP. Plain films showed widening of the joint capsule while ultrasonography detected joint effusion.
Of the following, the **MOST** likely diagnosis is

A. proximal tibial osteomyelitis
B. pauciarticular rheumatoid arthritis
C. Lyme arthritis
D. septic arthritis
E. pyomyositis

46. Which of the following structural or acquired heart disease can participate in sports?

A. Hypertrophic cardiomyopathy (symptomatic)
B. Dilated cardiomyopathy (symptomatic)
C. Mitral valve prolapse
D. Coronary artery anomalies
E. Acute rheumatic fever with carditis

47. Which of the following cardiovascular diseases can result in sudden death during sports?

A. Hypertrophic cardiomyopathy
B. Malignant ventricular arrhythmias
C. Hypertension
D. Congenital heart disease
E. Symptomatic Wolff- Parkinson-White syndrome

48. What is the definition of sprain?
    A. Injury to a muscle or tendon
    B. Crush injury to any soft tissue
    C. Injury to a ligament or joint capsule
    D. Repetitive microtrauma that exceeds the body's rate of repair
    E. Evidence of microscopic trauma to tendons

49. Why children are more vulnerable to heat illness than adults?
    A. They have greater ratio of surface area to body mass
    B. They produce lesser heat per kilogram of body weight during activity.
    C. The sweat rate is greater in children
    D. The temperature at which sweating occurs is lower
    E. Children have a sensitive response

50. What is the character of the skin in heat stroke?
    A. Sweating skin
    B. Profuse sweating skin
    C. Hot dry skin
    D. Hot moist skin
    E. Flushed skin

51. Treatment of heat exhaustion includes moving to a cool environment, cooling the body with fans, removing excess clothing, and placing ice over the
    A. head
    B. groin and axillae
    C. dorsum of hands and feet
    D. chest and abdomen
    E. whole body

52. Which of the following common performance-enhancing substances is allowed by international olympic committee?
    A. Anabolic–androgenic steroids
    B. Creatine
    C. Human growth hormone
    D. Amphetamines/stimulants
    E. Erythropoietin/blood doping

53. Testicular atrophy is a major adverse effect of which of the following performance-enhancing substances?

A. Anabolic–androgenic steroids
B. Creatine
C. Human growth hormone
D. Amphetamines/stimulants
E. Erythropoietin/blood doping

54. A 3-year-old child brought by his worried mother because he looks short while his parents look normal height, he started walking alone after 21 months, his head is large with normal-sized trunk and short extremities mainly the proximal segments. He has normal Intelligence.
Of the following, the **MOS**T likely diagnosis is

A. thanatophoric dysplasia
B. achondroplasia
C. hypochondroplasia
D. Jansen Metaphyseal Dysplasia
E. Schmid metaphyseal dysplasia

55. A 9-year-old boy brought by his father due to poor school performance. On examination, he looks a little bit short stature affecting the limbs with a stocky build and slight frontal bossing of the head.
Of the following, the **MOST** likely diagnosis is

A. thanatophoric dysplasia
B. achondroplasia
C. hypochondroplasia
D. Jansen Metaphyseal Dysplasia
E. Schmid metaphyseal dysplasia

56. A 3-week-old boy with severe shortening of limb, clubfoot, associated with an unusual facial appearance. His serum calcium is 14mg/dL. Radiographs show short tubular bones with characteristic metaphyseal abnormalities that include flaring, irregular mineralization, fragmentation, and widening of the physeal space.
Of the following, the **MOST** likely diagnosis is

A. thanatophoric dysplasia
B. achondroplasia
C. hypochondroplasia

D. Jansen Metaphyseal Dysplasia
E. Schmid metaphyseal dysplasia

57. In which of the following conditions, the anterior fontanelle is wide and might remain open?
A. Rickets
B. Down syndrome
C. Hypochondroplasia
D. Cleidocranial dysplasia
E. Campomelic dysplasia

58. A 6-month-old boy presents with macrocephaly, hepatosplenomegaly, deafness, blindness, and severe anemia. Radiographs reveal diffuse bone sclerosis and blood test showed hypocalcemia. Most of the bone manifestations in this disease can be prevented or reversed by
A. hematopoietic stem cell transplantation
B. Calcitriol
C. interferon-γ
D. blood transfusions
E. vitamin D and calcium supplements

59. A 2-year-old boy present with short limbs, postaxial polydactyly of the hands, nail dysplasia and dental anomalies, skeletal radiographs reveal short tubular bones with clubbed ends, echo revealed atrial septal defects
Of the following, the **MOST** likely diagnosis is
A. Asphyxiating thoracic dystrophy
B. Ellis–van Creveld syndrome
C. Metatropic dysplasia
D. thanatophoric dysplasia
E. achondroplasia

60. Os calcaneus is the affected region in which of the following Juvenile Osteochondroses?
A. Legg-Calvé-Perthes disease
B. Osgood-Schlatter disease
C. Sever disease
D. Freiberg disease
E. Scheuermann disease

61. A 4-month-old boy brought by his mother with painful mandibular swelling for one month duration, the swelling is wood-like in consistency with minimal warmth or redness and no suppuration, the baby is irritable, feverish, and poor feeder. Laboratory investigations display elevated erythrocyte sedimentation rate, thrombocytosis, anemia and increased serum alkaline phosphatase, radiographic features shows soft tissue swelling, calcification, and cortical hyperostosis of mandible.
Of the following, the **MOST** likely diagnosis is

A. vitamin A intoxication
B. neuroblastoma
C. Caffey Disease
D. osteoid osteoma
E. scurvy

62. A 6- year- old child presents with short stature, blue scleral, blue-gray teeth that break easily, hearing loss, and kyphoscoliosis. Radiographs show Lower long bones osteoporosis, with metaphyseal flaring, and “popcorn” formation at growth plates.
Of the following, the **MOST** likely diagnosis is

A. Osteogenesis Imperfecta Type I
B. Osteogenesis Imperfecta Type II
C. Osteogenesis Imperfecta Type III
D. Osteogenesis Imperfecta Type IV
E. Osteogenesis Imperfecta Type V

63. What is the **MOST** obvious skeletal manifestation of Marfan syndrome (MFS)?

A. Pectus carinatum
B. Dolichostenomelia
C. Pes planus
D. Arachnodactyly
E. Amptodactyly

64. What is the leading cause of morbidity and mortality in young children with Marfan syndrome (MFS)?

A. Supraventricular arrhythmias
B. Mitral valve dysfunction
C. Ventricular dysrhythmias

D. Prolonged QT interval.
E. Dilated cardiomyopathy

65. What is the **MOST** common ocular manifestation in Marfan syndrome (MFS)?

A. Severe myopia
B. Flat cornea
C. Ectopia lentis
D. Hypoplastic iris
E. ciliary muscle hypoplasia

66. A 3-month-old infant presents with irritability, and failure to thrive. Laboratory results revealed hypercalcemia, hypercalciuria, and low alkaline phosphatase, ultrasonography shows nephrocalcinosis, X-rays reveal irregular ossification, punched-out areas, and metaphyseal cupping.
Of the following, the **MOST** likely diagnosis is

A. infantile hypophosphatasia
B. perinatal hypophosphatasia
C. hypophosphatasia tarda
D. pseudohypophosphatasia
E. metaphyseal Dysostosis

67. A 4-year-old child presents with skeletal pain, muscle weakness, premature exfoliation of primary teeth, mild skeletal deformities, and short stature. X-rays of long bones have characteristic “tongues” of radiolucency.
Of the following, the **MOST** likely diagnosis is

A. infantile hypophosphatasia
B. perinatal hypophosphatasia
C. hypophosphatasia tarda
D. pseudohypophosphatasia
E. metaphyseal Dysostosis

68. An 18-month-old child found to have high serum alkaline phosphatase 5,000 IU/L, which is detected as an incidental finding during screening laboratory evaluations for bow legs. There are no other clinical or laboratory signs of hepatic or bone disease. Laboratory assessments of: calcium, phosphorus, Cr, AST, ALT, GGT, bilirubin, PTH, and 25-hydroxyvitamin D are normal.
Of the following, the **MOST** likely diagnosis is

A. familial hyperphosphatasia
B. juvenile paget disease
C. familial expansile osteolysis
D. autosomal dominant hypophosphatemic rickets
E. transient hyperphosphatasia

Chapter 31

Bone and Joint Disorders Answers

ZUHAIR M. ALMUSAWI

1.(B) The head is disproportionately large at birth, and the ratio of head height to total height is approximately 1:4 at birth, which changes to 1:7.5 at skeletal maturity.

2.(C) By age 5 yr, birth height usually doubles and the child is approximately 60% of adult height. The child is approximately 80% of final height at 9 yr. During puberty, the standing height increases by approximately 1 cm/mo.

3.(E)

Painful limp

- ✓ Septic arthritis and osteomyelitis
- ✓ Transient synovitis
- ✓ Occult trauma (“toddler's fracture”)
- ✓ Intervertebral diskitis
- ✓ Malignancy
- ✓ Abuse

Painless limp

- ✓ Developmental dysplasia of the hip
- ✓ Neuromuscular disorder
- ✓ Polio
- ✓ Cerebral palsy
- ✓ Lower extremity length inequality

4.(A) A bone scan displays physiologic information rather than pure anatomy and relies on the emission of energy from the nucleotide injected into the patient. Indications include early septic arthritis, osteomyelitis, avascular necrosis, tumors (osteoid osteoma), metastatic lesions, occult and stress fractures, and cases of child abuse.

5.(B) When comparing risk for developmental dysplasia of the hip (DDH) with other congenital foot deformities, congenital calcaneovalgus has the highest association, with 19.4% of patients having coexisting DDH.

6.(A) Flatfoot is a common diagnosis; it has been estimated that up to 23% of the public may be affected, depending on the diagnostic criteria. Although

hypermobile or flexible pes planus represents a common source of concern for parents, these children are rarely symptomatic. Flatfeet are common in neonates and toddlers and are associated with physiologic ligamentous laxity. Improvement may be seen when the longitudinal arch develops between 5 and 10 yr of age.

7.(C) Although a unilateral cavus foot is most likely to result from an occult intraspinal anomaly, bilateral involvement usually suggests an underlying nerve or muscle disease.

8.(D) Calcaneal apophysitis (Sever disease) is the most common cause of heel pain in children; treatment includes activity modification, nonsteroidal antiinflammatory medications, heel cord stretching exercises, and heel cushions or arch supports.

9.(D) The extra digit may be either rudimentary or well formed, and plain radiographs of the foot help define the anatomy and evaluate any coexisting bony anomalies. Treatment is indicated for cosmesis and to allow for fitting with standard shoes. This involves surgical removal of the extra digit, and the procedure is generally performed between 9 and 12 mo of age. Rudimentary digits may be surgically excised earlier, but should not be “tied off.”

10.(B) A hammer toe involves flexion at the proximal IP (PIP) joint with or without the distal IP (DIP) joint, and the MTP joint may be hyperextended. This deformity may be distinguished from a curly toe by the absence of rotation. The 2nd toe is most commonly involved, and a painful callus may develop over the dorsum of the toe where it rubs on the shoe.

11. (A) Differential Diagnosis of Foot Pain According to Age

6-12 yr

Poorly fitting shoes
Trauma (fracture, sprain)
Juvenile idiopathic arthritis (enthesopathy)
Puncture wound
Sever disease (calcaneal apophysitis)
Accessory tarsal navicular bone
Hypermobile flatfoot
Tarsal coalition
Oncologic (Ewing sarcoma, leukemia)

12.(D) Medial (internal) tibial torsion manifests with in-toeing gait. It is commonly associated with congenital metatarsus varus, genu valgum, or femoral anteversion. This condition is usually seen during the second year of

life. It is often noticed after the child begins to walk independently. Many parents are concerned with a “bowed” appearance of the legs.

13.(D) Physiologic bowleg is a common torsional combination that is secondary to normal in utero positioning. Spontaneous resolution with normal growth and development can be anticipated. Persistence of varus beyond 2 yr of age may be pathologic.

14.(A) Secondary forms of osteoporosis include various neuromuscular disorders, chronic illness, endocrine disorders, and drug-induced and inborn errors of metabolism, including lysinuric protein intolerance and Gaucher disease.

15.(B) The appearance of symmetric bilateral genu valgum most pronounced around age 4 is part of the normal physiologic process of leg development. However, variation of up to 15 degrees of valgus is possible until 6 yr of age. The majority of physiologic valgus has a good chance of correction until this age.

16.(C)

17.(C) Ultrasound can be used to confirm a simple cystic lesion in the expected anatomical location and is often the only diagnostic test necessary to confirm the diagnosis with these reassuring findings.

18.(A) In most cases, reassurance is all that is needed for popliteal cysts because they often resolve spontaneously.

19.(B) Localized soft tissue swelling, along with an eventual firm and fixed increased prominence at the tibial tubercle, may occur with OS disease and may also be part of the initial complaint. There is typically no acute traumatic inciting event.

20.(D) Pain beneath or near the patella is the most common symptom. Bent knee activities, such as walking up and down stairs put the patella under high compressive loads and tend to aggravate the pain. Squatting, running, and other vigorous physical activities also exacerbate the anterior knee pain. Sitting in a flexed knee position for an extended period of time, the so-called theater sign, is another common complaint.

21.(E) A positive family history for DDH is found in 12–33% of affected patients. DDH is more common among female patients (80%), which are thought to be because of the greater susceptibility of female fetuses to maternal hormones, such as relaxin, which increases ligamentous laxity. Although only 2–3% of all babies are born in breech presentation, the incidence of DDH in these patients is 16–25%. Any condition that leads to a tighter intrauterine space and, consequently, less room for normal fetal motion may be associated with DDH.

These conditions include oligohydramnios, large birth weight, and first pregnancy.

22.(A) As the baby enters the 2nd and 3rd mo of life, the soft tissues begin to tighten, and the Ortolani and Barlow tests are no longer reliable. Limitation of abduction is the most reliable sign of a dislocated hip in this age group.

23.(C) Excessive lordosis, which develops secondary to altered hip mechanics, is common and is often the presenting complaint.

24.(A) Newborns hips that are Barlow-positive (reduced but dislocatable) or Ortolani positive (dislocated but reducible) should generally be treated with a Pavlik harness as soon as the diagnosis is made.

25.(A) Although transient synovitis can occur in all age groups. Approximately symptoms often develop acutely and usually consist of pain in the groin, anterior thigh, or knee, which may be referred from the hip.

The hip is not held flexed, abducted, or laterally rotated unless a significant effusion is present. They are often afebrile or have a lowgrade fever <38°C (100.4°F).

26.(D) Several classic radiographic signs have been reported that describe a "head at risk" for severe deformity. Lateral extrusion of the epiphysis, a horizontal physis, calcification lateral to the epiphysis, subluxation of the hip, and a radiolucent horizontal V in the lateral aspect of the physis (Gage's sign) are all associated with a poor prognosis.

27.(B) The earliest abnormality noted on physical exam in patients with scoliosis is asymmetry of the posterior chest wall on forward bending. This test, called the Adams forward-bending test, is performed by placing a scoliometer at the apex of the deformity with the patient bending 45 degrees forward. An inclination measuring 7 degrees or more has been suggested as the cutoff for orthopedic referral.

28.(D) Genitourinary abnormalities are identified in 20–40% of children with congenital scoliosis and include unilateral renal agenesis, ureteral duplication, horseshoe kidney, and genital anomalies.

29.(D) Postural kyphosis is a common cosmetic concern and is most often recognized by parents or peers. The mainstay of treatment is reassurance.

30.(B) Scheuermann disease is the most common form of structural hyperkyphosis and is defined by wedging of >5 degrees of three or more consecutive vertebral bodies at the apex of the deformity on a lateral radiograph. In addition, the apex of the thoracic kyphosis is lower than expected.

31.(C) Differential Diagnosis of Back Pain

Inflammatory Diseases

- ✓ Diskitis*
- ✓ Vertebral osteomyelitis (pyogenic, tuberculosis)
- ✓ Spinal epidural abscess
- ✓ Transverse myelitis
- ✓ Pyelonephritis*
- ✓ Perinephric abscess
- ✓ Pancreatitis
- ✓ Paraspinal muscle abscess, myositis
- ✓ Psoas abscess
- ✓ Endocarditis
- ✓ Pelvic osteomyelitis or myositis
- ✓ Pelvic inflammatory disease

32.(C) Spondylolysis is common in athletes who engage in repetitive spinal hyperextension, especially gymnasts, football interior linemen, weight lifters, and wrestlers. Approximately 4–8% of the entire pediatric population is affected, making it the most common cause of back pain in adolescents when a diagnosis can be established.

33.(C) A high index of suspicion is required to establish the diagnosis of infectious spondylitis. The diagnosis may be established earlier using either a technetium bone scan or MRI; MRI is the most sensitive and specific imaging to diagnose osteomyelitis and to identify abscesses and/or neural compression.

34.(E) Staphylococcus aureus is the most common organism causing spine infections. Other organisms include Kingella kingae, and less often group A streptococcus, and Escherichia coli. Rare causes of vertebral bone infection include tuberculosis, Serratia marcescens, brucellosis, and cat-scratch disease.

Blood cultures have a sensitivity of only 30%. Percutaneous or open biopsy of the disc space is positive only 50–85% of the time; PCR is indicated for the diagnosis of Kingella.

35.(C) MRI is the best study to establish the diagnosis of a disc herniation. CT is especially helpful to visualize a partially ossified fragment associated with a slipped apophysis.

36.(C) Klippel-Feil syndrome (KFS) includes the classic triad of a low posterior hairline, short neck, and decreased cervical range of motion.

37.(E)

Syndromes Associated With Polydactyly

- ✓ Carpenter syndrome

- ✓ Ellis-van Creveld syndrome
- ✓ Meckel-Gruber syndrome
- ✓ Polysyndactyly
- ✓ Trisomy 13
- ✓ Orofaciodigital syndrome
- ✓ Rubinstein-Taybi syndrome

Syndromes Associated With Syndactyly

- ✓ Apert syndrome
- ✓ Carpenter syndrome
- ✓ de Lange syndrome
- ✓ Holt-Oram syndrome
- ✓ Orofaciodigital syndrome
- ✓ Polysyndactyly
- ✓ Trisomy 21
- ✓ Fetal hydantoin syndrome
- ✓ Laurence-Moon-Biedl syndrome
- ✓ Fanconi pancytopenia
- ✓ Trisomy 13
- ✓ Trisomy 18

38.(B)

39.(B) Toddler fractures occur in young ambulatory children. The age range for this fracture is typically from around 1-4 yr. The injury often occurs after a seemingly harmless twist or fall and is often unwitnessed. The radiographs may show no fracture; the diagnosis is made by physical examination. The classic symptom is refusal to bear weight, which can manifest as pulling up the affected extremity or florid display of protest. The other common sign is point tenderness at the fracture site. The AP and lateral views of the tibia-fibula might show a nondisplaced spiral fracture of the distal tibial metaphysis. An oblique view is often helpful because the fracture line may be visible in only one of the three views. Often the fracture line is not visualized until 2-3 wk later, when periosteal reaction and resorption at fracture site allow better visualization. Inflammatory markers may be ordered to rule out infectious processes if the diagnosis is in doubt.

40.(C) Staphylococcus aureus is the most common infecting organism in osteomyelitis among all age groups, including newborns.

41.(A) Kingella kingae is the second most common cause of osteomyelitis in children younger than 4 yr of age. The organism is established as a cause of osteomyelitis, spondylodiskitis, and septic arthritis in this age group, especially

when there is a subacute presentation. K. kingae can be difficult to detect unless polymerase chain reaction (PCR) testing is used.

42.(B) Cefotaxime or ceftriaxone is recommended for pneumococcal isolates with resistance to penicillin and for most Salmonella spp.

43.(A) Joints of the lower extremity constitute 75% of all cases of septic arthritis (Knee ~35, Hip ~25, Ankle ~10). The elbow, wrist, and shoulder joints are involved in approximately 25% of cases, and small joints are uncommonly infected, except in gonococcal arthritis.

44.(B) Blood cultures should be performed in all cases of suspected septic arthritis but are positive in 20% or fewer cases of proven or probable septic arthritis. Cervical, anal, and throat cultures should be obtained when gonococcus is suspected.

45.(D) The signs and symptoms of septic arthritis depend on the age of the patient. Early signs and symptoms may be subtle, particularly in neonates. Older infants and children might have fever and pain, with localizing signs such as swelling, erythema, and warmth of the affected joint. With involvement of joints of the pelvis and lower extremities, limp or refusal to walk often occurs. Plain films can suggest the diagnosis of septic arthritis by showing: widening of the joint capsule, soft tissue edema, and obliteration of normal fat lines. Ultrasonography is particularly helpful in detecting joint effusion and fluid collection in the soft tissue and subperiosteal regions.

46.(C)

47.(A) Sudden death during sports can result from undetected cardiac disease such as hypertrophic or other cardiomyopathies, anomalous coronary vessels, or a ruptured aorta in Marfan syndrome. In many cases, the underlying heart disease is not suspected, and death is the first sign of heart disease.

48.(C)

- A sprain is an injury to a ligament or joint capsule.
- A strain is an injury to a muscle or tendon.
- A contusion is a crush injury to any soft tissue.
- Overuse injuries are caused by repetitive microtrauma that exceeds the body's rate of repair.
- Tendinosis is evidence of microscopic trauma to tendons.

49.(A) Children are more vulnerable to heat illness than adults because they have greater ratio of surface area to body mass and produce greater heat per kilogram of body weight during activity. The sweat rate is lower in children, and the temperature at which sweating occurs is higher. Children can take longer to acclimatize to warmer, more humid environments. Children also have a blunted

thirst response compared with adults and might not consume enough fluid during exercise to prevent dehydration.

50.(C) Heat Stroke: MEDICAL EMERGENCY

- ✓ Core temperature of 40°C (104°F) or higher
- ✓ Hot dry skin
- ✓ Multiple system failure
- ✓ Delirium
- ✓ Convulsions
- ✓ Abnormal vital signs

51.(B)

52.(B) Creatine increase in strength, power output, sprint performance, total work to fatigue, peak force/power; decrease lactate threshold; increase weight and lean body mass.

53.(A) Anabolic–androgenic steroids

MAJOR ADVERSE EFFECTS

Testicular atrophy, CV disease, atherosclerosis, myocardial disease, liver dysfunction, cancer

MINOR ADVERSE EFFECTS

Acne, gynecomastia

54.(B) Infants usually exhibit delayed motor milestones, often not walking alone until18-24 mo. This is because of hypotonia and mechanical difficulty balancing the large head on a normal-sized trunk and short extremities. Intelligence is normal unless central nervous system complications develop. As the child begins to walk, the gibbus usually gives way to an exaggerated lumbar lordosis.

55.(C) Hypochondroplasia resembles achondroplasia but is milder. Usually, it is not apparent until childhood, when mild short stature affecting the limbs becomes evident. Children have a stocky build and slight frontal bossing of the head. Learning disabilities may be more common in this condition. Radiographic changes are mild and consistent with the mild achondroplastic phenotype. Complications are rare; in some patients the condition is never diagnosed. Adult heights range from 116 to 146 cm. Recombinant growth hormone therapy may enhance growth and improve body disproportion.

56.(D) Jansen metaphyseal chondrodysplasia is a rare, dominantly inherited chondrodysplasia characterized by severe shortening of limbs associated with an unusual facial appearance. Sometimes it is accompanied by clubfoot and hypercalcemia with serum calcium values of 13-15 mg/dL.

57.(D) Cleidocranial dysplasia might be recognized in infants because of drooping shoulders, open fontanelles, and prominent forehead.
The shoulders of patients with Cleidocranial dysplasia can meet in the midline. Radiographs reveal hypoplastic or absent clavicles, delayed ossification of the cranial bones with multiple ossification centers (wormian bones), and delayed ossification of pelvic bones. The anterior fontanelle is wide and might remain open.
58.(A) Most of the bone manifestations in severe osteopetrosis due to intrinsic osteoclast defects can be prevented or reversed by hematopoietic stem cell transplantation (HSCT), if carried out before development of irreversible secondary complications, such as visual impairment.
59.(B) The Ellis–van Creveld syndrome, also known as chondroectodermal dysplasia, is a skeletal and an ectodermal dysplasia. The skeletal dysplasia presents at birth with short limbs, especially the middle and distal segments, accompanied by postaxial polydactyly of the hands and sometimes of the feet. Nail dysplasia and dental anomalies (including neonatal, absent, premature loss of teeth, and upper lip defects) constitute the ectodermal dysplasia. Common manifestations also include atrial septal defects and other congenital heart defects.
60. (C)
Legg-Calvé-Perthes disease = Capital femoral epiphysis
Osgood-Schlatter disease = Tibial tubercle
Sever disease = Os calcaneus
Freiberg disease = Head of second metatarsal
Scheuermann disease = Vertebral bodies
Blount disease = Medial aspect of proximal tibial epiphysis
Osteochondritis dissecans = Subchondral regions of knee, hip, elbow, and ankle
61.(C) Prenatal and more often postnatal onsets have been described. Prenatal onset may be mild (autosomal dominant) or severe (autosomal recessive). Severe prenatal disease is characterized by typical bone lesions, polyhydramnios, hydrops fetalis, severe respiratory distress, prematurity, and high mortality. Onset in infancy (younger than 6 mo; average: 10 wk) is most common; manifestations include the sudden onset of irritability, swelling of contiguous soft tissue that precedes the cortical thickening of the underlying bones, fever, and anorexia. The swelling is painful with a wood-like induration but with minimal warmth or redness; suppuration is absent. There are unpredictable remissions and relapses; an episode can last 2 wk to 3 mo. The

most common bones involved include the mandible (75%), the clavicle, and the ulna.

62.(C) OI type III is the most severe nonlethal form of OI and results in significant physical disability. Birthweight and length are often low normal. Fractures usually occur in utero. There is relative macrocephaly and triangular facies. Postnatally, fractures occur from inconsequential trauma and heal with deformity. Disorganization of the bone matrix results in a "popcorn" appearance at the metaphyses. The rib cage has flaring at the base, and pectal deformity is frequent. Virtually all type III patients have scoliosis and vertebral compression. Growth falls below the curve by the 1st yr; all type III patients have extreme short stature. Scleral hue ranges from white to blue. Dentinogenesis imperfecta, hearing loss, and kyphoscoliosis may be present or develop over time.

63.(B) Overgrowth of the long bones (dolichostenomelia) is often the most obvious manifestation of MFS and may produce a reduced upper segment to lower segment ratio (UL/LS) or an arm span to height ratio >1.05 times.

64.(B) Insufficiency of the mitral valve can lead to heart failure, pulmonary hypertension, and death in infancy; this manifestation is the leading cause of morbidity and mortality in young children with the disorder.

65.(C) Dislocation of the ocular lens (ectopia lentis) occurs in around 60–70% of patients, although it is not unique to the disorder. Other ocular manifestations include early and severe myopia, flat cornea, increased axial length of the globe, hypoplastic iris, and ciliary muscle hypoplasia, causing decreased miosis. Patients are also predisposed to retinal detachment, early cataracts, and glaucoma.

66.(A) Infantile hypophosphatasia is next on the continuum. These infants present prior to 6 months of age with hypercalcemia/ hypercalciuria (leading to nephrocalcinosis), premature cranial suture fusion (that can lead to increased intracranial pressure), irritability, and failure to thrive. X-rays reveal irregular ossification, punched-out areas, and metaphyseal cupping. Prior to the availability of enzyme replacement therapy with asfotase alfa, mortality was estimated at 50%; survivors had significant disability.

67.(C) Hypophosphatasia tarda present with premature exfoliation of primary teeth (with the root intact due to poorly mineralized dental cementum), mild skeletal deformities, fracture, and variable short stature. Some children have skeletal pain and muscle weakness. Long bones can have characteristic "tongues" of radiolucency.

68.(E) In children between 6 mo and 2 yr of age, increases in alkaline phosphatase are often due to transient hyperphosphatasia . This is usually detected as an incidental finding during screening laboratory evaluations or evaluations performed to assess a specific complaint. Serum alkaline phosphatase values as high as 3,000-6,000 IU/L may be encountered. Liver and bone isoenzyme fractions are both elevated; there are no other clinical or laboratory signs of hepatic or bone disease. Diagnosis can be confirmed by a careful clinical history plus laboratory assessments of: calcium, phosphorus, Cr, AST, ALT, GGT, bilirubin, PTH, and 25-hydroxyvitamin D. The cause may be related to excess sialylation of alkaline phosphatase, which slows clearance. Alkaline phosphatase should be followed serially (every 2-3 mo) until resolution is documented. Resolution usually occurs within 4-6 mo.

Chapter 32

Rehabilitation Medicine Questions

AQEEL MAHDI

1. What is the recommended duration of prophylactic anti-epileptic medications in seizures associated with severe traumatic brain injury?

A. No prophylaxis
B. 1 week
C. 2 weeks
D. 6 weeks
E. 12 weeks

2. You examine a 10-year-old boy with history of traumatic brain injury due to car accident. He had hyperthermia, tachycardia, tachypnea, diaphoresis, and increased muscle tone.
Of the following, the **MOST** likely diagnosis is

A. paroxysmal sympathetic hyperactivity
B. pulmonary embolism
C. sepsis
D. elevated intracranial pressure
E. acute painful episode

3. Which of the following benzodiazepines is **MOST** commonly used to treat spasticity in a 4-year-old child?

A. Diazepam
B. Clonazepam
C. Lorazepam
D. Chlordiazepoxide
E. Midazolam

4. A family consults you about the **MOST** appropriate treatment for their 10-year-old boy who is a known case of cerebral palsy with severe spasticity of upper and lower limbs refractory to oral medication associated with excessive drooling of saliva.

Of the following, the **MOST** appropriate answer is

A. intramuscular botulinum toxin
B. phenol neurolysis
C. intrathecal baclofen
D. selective dorsal rhizotomy
E. Hemispherectomy

5. Which of the following is the **MOST** common neuroendocrine disorder following traumatic brain injury TBI?

A. Growth hormone deficiency
B. Cerebral salt wasting
C. Hyperpituitarism
D. Diabetes insipidus
E. Syndrome of inappropriate secretion of antidiuretic hormone

6. Which of the following oral antispasmodic medications is a preferred agent in the treatment of spasticity of spinal origin?

A. Baclofen
B. Diazepam
C. Clonidine
D. Dantrolene sodium
E. Tizanidine

7. Which of the following oral antispasmodic medications is acting by peripheral calcium blocking?

A. Baclofen
B. Diazepam
C. Clonidine
D. Dantrolene sodium
E. Tizanidine

8. Which of the following cervical spinal cord injuries has the best functional outcome?

A. C4
B. C5
C. C6
D. C7
E. C8

9. A 9-year-old boy presents with pounding frontal headache. Examination shows flushing of the face, blood Pressure 135/70, heart rate 60 bpm, and distended bladder. He had a history of spinal cord injury involving the upper thoracic vertebrae.
Of the following, the **MOST** likely diagnosis is

A. migraine
B. essential hypertension
C. pheochromocytoma
D. intracranial hypertension
E. autonomic dysreflexia

10. Which of the following factors considered protective against development of birth brachial plexus palsy?

A. Multiparous mothers
B. Mothers with excessive weight gain
C. Diabetic mothers
D. Cephalic presentation
E. Twins

11. Which of the following types of birth brachial plexus injury is the **LEAST** severe form?

A. Neurapraxia
B. Neurotmesis
C. Axonotmesis
D. Avulsion
E. Rupture

12. What is the recommended time for surgical intervention in patients with a complete brachial plexus palsy?

A. As soon as possible
B. 3 months
C. 6 months
D. 1 year
E. 2 years

13.Which of the following neurological level of meningomyelocele carry the highest risk of developing scoliosis?

A. Thoracic

B. Upper lumbar (L1-L2)
C. Midlumbar (L3)
D. Lower lumbar (L4-L5)
E. Sacral

14. Which of the following neurological level of meningomyelocele carry the highest risk of developing hip dislocation?
A. Thoracic
B. Upper lumbar (L1-L2)
C. Midlumbar (L3)
D. Lower lumbar (L4-L5)
E. Sacral

15. Which of the following is the mainstay of management in patients with meningomyelocele and neurogenic bladder?
A. Clean intermittent catheterization
B. Prophylactic antibiotics
C. Oxybutynin
D. Bladder augmentation
E. Urethral surgeries

16. Which of the following spinal cord segment of meningomyelocele will have minimal motor dysfunction?
A. L1-L2
B. L3-L4
C. L5
D. S1
E. S2-S3

17. What is the minimal age limit where a manual and electric power wheelchair can be used by children?
A. 2 years
B. 3 years
C. 4 years
D. 5 years
E. 6 years

18. Which of the following is the **MOST** common and long-lasting sequelae of traumatic brain injury?

A. Cognitive-behavioral disorders
B. Syndrome of inappropriate secretion of antidiuretic hormone
C. Posttraumatic seizure
D. Paroxysmal sympathetic hyperactivity
E. Spasticity

Chapter 32

Rehabilitation Medicine Answers

AQEEL MAHDI

1.(B) Prophylactic treatment with an anti-epileptic medication for 7 days after a traumatic brain injury is commonly prescribed. However, treatment with an anti-epileptic medication beyond 1 week offers no further benefit as a prophylactic agent.

2.(A) Paroxysmal sympathetic hyperactivity PSH is thought to be due to disruption of the inhibitory function of the mesencephalon on the diencephalon. Some drug-related symptoms may mimic the features of PSH for example, haloperidol and chlorpromazine may cause neuroleptic malignant syndrome, phenytoin may precipitate a fever, and cimetidine may produce extrapyramidal symptoms.

3.(A) Diazepam is the most commonly used medication to treat spasticity because of its long half-life and need for less-frequent administration. In children <2 yr of age, clonazepam is a good option because of the availability of a liquid formulation and dosing guidelines.

4.(C) Intrathecal baclofen (ITB) is highly effective in treating severe spasticity. ITB is delivered to the intrathecal space via a surgically implanted infusion pump and catheter. This method of delivery confers an advantage over enteral baclofen, in that central nervous system depressive effects are minimized and dosages can be titrated to functional effect.

5.(A) Deficiencies of growth hormone and gonadotropin are the most common disorders following TBI, resulting in growth retardation and precocious puberty respectively. About 8% of children with severe TBI sustain chronic pituitary dysfunction.

6.(A) Baclofen exerts an inhibitory effect on both monosynaptic and polysynaptic spinal reflexes. Unfortunately, supraspinal receptor sites also exist, resulting in sedation, which is common to all GABAergic medications.

7.(D)

- ✓ Baclofen: centrally acting, structural analog of GABA. Bind to GABAB receptors causing presynaptic inhibition of mono/ polysynaptic spinal reflexes.

- ✓ Diazepam: Centrally acting, binds to GABAA receptors mediating presynaptic inhibition in brain stem reticular formation and spinal polysynaptic pathways.
- ✓ Clonidine: Centrally acting mixed alpha adrenoceptor agonist.
- ✓ Dantrolene sodium: Peripheral action, blocking release of calcium from sarcoplasmic reticulum with uncoupling of nerve excitation and skeletal muscle contraction.
- ✓ Tizanidine: Centrally acting, alpha-2 adrenoceptor agonist activity at both spinal and supraspinal sites.

8.(E) At C8 level patient can do feeding, grooming, upper extremity dressing, lower extremity dressing, bathing, bed mobility, weight shifts, transfer independently.

9.(E) Children with neurologic levels of injury at T6 or above are at particular risk for interruption and decentralization of the autonomic nervous system. Autonomic dysreflexia AD is a sustained sympathetic response as a result of a noxious stimulus below the level of injury. Symptoms resulting from AD typically include hypertension, bradycardia, headache, and flushing of the skin above the level of injury, although vague symptoms such as fatigue, irritability, or crying may be the presenting symptoms in younger patients. Noxious stimuli are most often localized to bladder or rectal distention, but may include a number of other causes.

10.(E) Risk factors for birth brachial plexus injury include prior infants with birth brachial plexus palsy BBPP, shoulder dystocia, birth weight >4 kg, multiparous mothers, mothers with excessive weight gain, and diabetic mothers. Delivering twins or triplets, as well as cesarean sections, have been described as protective from BBPP.

11.(A) Nerve injuries include neurapraxia, neurotmesis, and axonotmesis. Neurapraxia is the least severe of these types and is a reversible loss of nerve conduction. This type will recover. Neurotmesis is the most severe and is a total and complete disruption of the nerve. An avulsion describes a neurotmesis of a preganglionic lesion, and a rupture describes the same event in a postganglionic lesion. Axonotmesis is the intermediate form and the most difficult to delineate.

12.(B) Infants who do not show satisfactory improvement in muscle strength are candidates for surgical intervention. Classically the lack of elbow flexion to 3/5 or greater strength by 3 mo of age merits referral for nerve surgery. The specific criteria and timing remain under debate. Those with a complete brachial plexus palsy with a flaccid arm and lack of sensation are under

consideration for surgery at 3 mo of age, and those with upper-plexus involvement are considered between 3 and 6 (or even 9) mo of age.

13.(A) The development of scoliosis has an association with the neurologic level. Children with thoracic level defects have an 80–100% risk, whereas those with a sacral level are at very low risk.

14.(C) The development of the hip is also influenced by neurologic level. The risk for dislocation is highest for those with lesions at the L3 level, followed by L1-L2. Unilateral hip dislocations should be fixed surgically, as they may result in pelvic obliquity and problems with sitting, whereas bilateral dislocations generally do not require interventions.

15.(A) The goals of treatment interventions are to protect kidney function and achieve social continence. The introduction of clean intermittent catheterization is the mainstay of management.

16.(E)

17.(A) Children as young as age 2 year can self-propel a manual wheelchair and operate a power wheelchair.

18.(A) Cognitive and behavioral impairments (poor memory-learning and executive skills, hyperactivity, depression, awareness deficits) are the most common and long-lasting sequelae of TBI. These deficits can inhibit successful school re-entry and participation in social activities.

Chapter 33

Environmental health Questions

HAIDAR A. N. ABOOD

1. Which of the following conditions is linked to environmental mycotoxin exposures?

A. Cerebral palsy
B. Neural tube defects
C. Thyroid cancer
D. Kidney stones
E. Chloracne

2. A 6-year-old boy presents to the E/D with cyanosis, headache, dizziness and generalized weakness two hours following eating excessive amount of cured meat. He vomited twice during transport to the hospital. Physical examination shows HR 120 bpm, RR 30 bpm, SpO_2 85%. ECG and CXR are normal. An arterial blood sample (obtained for ABG analysis) shows chocolate brown-colored blood. Ingestion of contaminated food is suspected.
Of the following, the **MOST** likely food contaminant is

A. nitrite
B. antimony
C. arsenic
D. mercury
E. copper

3. Which of the following imaging tests has the **HIGHEST** average radiation doses?

A. Computed tomography: brain
B. Computed tomography: chest
C. Computed tomography: abdomen/pelvis
D. Nuclear medicine (^{99m}Tc methylene diphosphonate–Bone)
E. Positron emission tomography (^{18}F-FDG; whole body)

4. Which of the following is the **MOST** radiosensitive tissue?

A. Brain
B. Skin
C. Lung
D. Liver
E. Gonads

5. Which of the following age groups is **MOST** sensitive to radiation-induced carcinogenesis?

F. Neonates
G. Infants
H. Toddlers
I. Preschool children
J. School children

6. Radiation induced cell injury results primarily from damage to

A. cell membrane
B. cytoplasmic reticulum
C. RNA
D. DNA
E. ribosomes

7. Which of the following effects of radiation therapy is the **MOST** severe acute reaction?

A. Pneumonitis
B. Dermatitis
C. Mucositis
D. Esophagitis
E. Cerebral edema

8. Which of the following primary malignancies has the **HIGHEST** cumulative incidence of a second neoplasm?

A. Soft tissue sarcoma
B. CNS cancers
C. Leukemia
D. Hodgkin disease
E. Non-Hodgkin disease lymphoma

9. After acute penetrating whole-body irradiation, the expected outcome depends on absolute lymphocyte count (ALC).
Which of the following ALC ranges determined within first 48 hours after exposure is associated with severe injury and fair prognosis?

A. 1,000-3,000
B. 1,000-1,500
C. 500-1,000
D. 100-500
E. <100

10. What is the **MOST** common site for accidental localized irradiation injuries?

A. Face
B. Hand
C. Foot
D. Thigh
E. Buttocks

11. Which of the following radiopharmaceutical agents requires complete cessation of breast feeding if administered to a lactating mother?

A. ^{131}-I Sodium iodide
B. ^{99m}Tc pertechnetate
C. ^{123}-I Sodium iodide
D. ^{99m}-Tc MDP
E. ^{51}Cr EDTA

12. A 2-month-old boy was breast fed during the last night by his mother who received ^{131}-I Sodium iodide before 4 days as a part of investigations for her thyroid nodule. The mother was advised previously to stop breast feeding but she believed that 3 days would be enough to clean her body.
Of the following, the **MOST** appropriate treatment is

A. no treatment is required
B. dilution therapy with forcing fluids
C. blocking therapy with potassium iodine
D. chelation therapy with calcium diethylene triamine pentaacetic acid
E. removal treatment with prussian blue

13. Which of the following chemical pollutants is associated with increased risk of sudden infant death syndrome?

A. Benzene
B. Environmental tobacco smoke
C. Lead
D. Methyl mercury
E. Organophosphate insecticides

14. Which of the following is the principal source of air pollution?
A. Agricultural use of pesticides
B. Industrial waste
C. Military sources
D. Natural sources
E. Fuel combustion

15. Which of the following chronic toxic effects is caused by organophosphate insecticides?
A. Cancer
B. Hormonal disruption
C. Reproductive impairment
D. Polyneuropathy
E. Pulmonary fibrosis

16. Children suffering from more than simple urticaria (e.g., wheezing, evidence of laryngeal edema or cardiovascular instability) following hymenoptera (including the stinging ants, bees, and wasps) envenomation are at high risk for progressing to systemic anaphylaxis with future stings.
Of the following, the **MOST** appropriate treatment that can reduce this risk is
A. Antihistamines
B. Regular epinephrine
C. Immunotherapy
D. Systemic corticosteroids
E. Leukotriene modifiers

17. Which of the following insects regularly causes fatalities in Middle East children?
A. Ants
B. Bees
C. Spiders
D. Scorpions

E. Wasps

18. Tobacco smoke exposure can be considered a modifiable risk factor for children with

A. sickle cell disease
B. immune thrombocytopenia
C. type I diabetes mellitus
D. hypothyroidism
E. rickets

19. Which of the following is the **MOST** appropriate method to treat children's second-hand tobacco smoke (SHS) exposure?

A. Parents smoking outside the home
B. helping parents quit smoking by the 5 A's method
C. wearing a smoking jacket
D. helping parents quit smoking by the Ask, Advise, Refer model
E. helping parents quit smoking by the CEASE program

20. Which of the following heavy metals has the **MOST** prevalent human exposure and requires a screening program?

A. Lead
B. Mercury
C. Arsenic
D. Cadmium
E. Thallium

21. Which of the following is **MOST** commonly required for the treatment of scorpion stings in children?

A. Intravenous opioids
B. Oral pain killers
C. Intravenous benzodiazepines
D. Scorpion antivenom
E. Local anesthesia

22. Which of the following is the classical presentation after toxic exposure to arsine gas?

A. Encephalopathy
B. Acute hemolysis

C. Cardiac arrhythmias
D. Acute tubular necrosis
E. Hemorrhagic gastroenteritis

23. A 5-year-old girl lives in an industrial area of glass refining, presents with chronic cough and mild diarrhea for the last two months. She also had prolonged fatigue, malaise, and weight loss. On examination she has peripheral sensorimotor neuropathy, mild alopecia, oral ulceration, generalized pruritic macular rash and transverse white striae on the nails. CBC shows leukopenia, anemia, and thrombocytopenia. Environmental exposure to heavy metals is suspected.
Of the following, the **MOST** likely offender is

A. lead
B. mercury
C. arsenic
D. cadmium
E. thallium

24. Definitive diagnosis of acute arsenic intoxication depends on quantitative analysis of arsenic in

A. blood
B. urine
C. saliva
D. hair
E. nail

25. Which of the following routes of exposure is usually associated with elemental mercury poisoning?

A. Oral
B. Dermal
C. Inhalation
D. Parenteral
E. Sublingual

26. What is the heavy metal responsible for the toxic effects of ingestion of a button battery?

A. Organic mercury
B. Organic lead

C. Arsenic salts
D. Inorganic mercury salts
E. Inorganic lead

27. A 6-year-old boy presents to the hospital with tremor, neuropsychiatric disturbances (emotional lability, delirium, and impaired memory), and gingivostomatitis. His father (a worker in the national batteries factory) describes the tremor as a fine intention tremor of the fingers that is abolished during sleep. On examination there is mixed sensorimotor neuropathy and visual disturbances. Chronic exposure to a heavy metal is suspected.
Of the following, the **MOST** likely offender is

A. lead
B. arsenic
C. aluminum
D. copper
E. mercury

28. A 3-year-old girl presents to the E/D two hours following ingestion of a hair-removal powder frequently used as folk remedy by her grandmother. The girl vomited twice before hospital arrival and the parents describe a history of convulsion during transport. On arrival she is semiconscious and crying while holding her stomach with bloody diarrhea and moderate dehydration. ECG reveals QT interval prolongation and polymorphous ventricular tachycardia, CXR shows diffuse pulmonary edema, while plain abdominal X-ray is normal.
After stabilizing the patient, the **MOST** appropriate next step management is

A. collecting urine for 24 hours and send for arsenic level
B. performing gastric lavage
C. administration of activated charcoal
D. whole-bowel irrigation
E. starting chelation therapy with dimercaprol

29. Which of the following chelators is preferred for chronic inorganic mercury poisoning?

A. Succimer (DMSA)
B. Dimercaprol (BAL)
C. D-Penicillamine
D. N-acetyl-D,L-penicillamine
E. Calcium disodium edetate ($CaNa_2EDTA$)

30. Which of the following is the **STANDARD** specimen for estimating lead toxicity?

A. urine
B. blood
C. saliva
D. hair
E. nails

31. A 9-year-old boy presented to the ER two hours after a rattle snake bite in his right leg. His leg was progressively swollen, tender, and erythematous with localized area of ecchymosis over the bitten site. Few minutes following removing the field-placed tourniquet, the boy developed severe epistaxis requiring nasal packing. Investigations revealed WBC 6000/mm^3, platelet 40,000/mm^3, INR 2.5, fibrinogen 60 mg/dL, and activated partial thromboplastin time (aPTT) 40 sec. Following these finding, Crotalinae polyvalent immune Fab antivenom was administered

Of the following, the **MAIN** indication for antivenin administration is

A. low platelet count
B. high INR
C. epistaxis
D. high aPTT
E. low fibrinogen level

32. Which of the following is the **MOST** common pathway for lead to enter the body?

A. Drinking contaminated water
B. Eating contaminated food
C. Cutaneous contamination with inorganic lead compounds
D. Nonnutritive hand-to-mouth activity of young children
E. Cutaneous contamination with organic lead compounds

33. Which of the following factors can enhance GI lead absorption?

A. Ingestion of large paint chips
B. Ingestion of lead with meal
C. Alkaline medium
D. Presence of calcium in meal
E. Iron deficiency

34. Which of the following is the **MOST** effective first aid measure after snake envenomation?

A. Local application of ice
B. Application of proximal tourniquets in the bitten limb
C. Immobilization of the bitten limb
D. Incision and squeezing of the wound
E. Wound suction

35. A 5-year-old boy lived in an old house presents with chronic constipation associated with headache and lethargy. The boy had two attacks of generalized seizures last week. On examination he looks pale and confused, ophthalmoscopy reveals papilledema. Serum ferritin 12 ng/mL, Hb 9 g/dL, Erythrocyte protoporphyrin level 40 µg/dL and radiographs of long bones show dense bands at the metaphyses. Chronic exposure to a heavy metal is suspected.
Of the following, the **MOST** likely offender is

A. lead
B. arsenic
C. aluminum
D. cadmium
E. mercury

36. Which of the following is the earliest manifestation of neurotoxicity after neurotoxic snake envenomation?

A. Upper limb weakness
B. Ptosis
C. Lower limb weakness
D. Respiratory muscles weakness
E. Flaccid paralysis

37. Which of the following drug combinations is indicated for children with lead encephalopathy?

A. $CaNa_2$ EDTA plus DMSA
B. $CaNa_2$ EDTA plus BAL
C. DMSA plus BAL
D. D-Penicillamine plus DMSA
E. D-Penicillamine plus BAL

38. Which of the following mushroom species is the **MOST** fatal?

A. Gyromitra
B. Boletus
C. Cortinarius
D. Amanita
E. Psilocybe

39. A 3-year-old boy presents to the E/D with delirium for the last 30 minutes. The boy was completely healthy before one hour when he ingests a wild mushroom from the adjacent garden. Physical examination shows plethoric face, dilated pupils, hot and dry skin, HR 150 bpm, RR 24 bpm, and temp 38 ˚C.
Of the following, the **MOST** appropriate management is

F. administration of activated charcoal
G. performing gastric lavage
H. whole-bowel irrigation
I. administration of cathartics
J. induction of emesis

40. A 13-year-old boy presents to the E/D with generalized erythema and shortness of breath, one hour after having dinner in a seafood restaurant. The symptoms started 20 min after eating a fish dish as abdominal pain and diarrhea associated with oral numbness. On examination there is generalized urticarial rash, diaphoresis, facial swelling, and tachycardia.
Of the following the **MOST** likely diagnosis is

A. shellfish poisoning
B. scombroid fish poisoning
C. ciguatera fish poisoning
D. pufferfish poisoning
E. Rudderfish poisoning

41. What is the classical toxic effect of illegal addition of melamine to infant formula?

A. Encephalopathy
B. Gall stone formation
C. Chemical hepatitis
D. Renal stone formation
E. Hemorrhagic gastroenteritis

42. In chemical and biologic terrorism, patients can be classified as having primarily respiratory, neuromuscular, or dermatologic manifestations. In this regard, which of the following agents has prominent dermatologic findings?

A. Mustard
B. Anthrax
C. Plague
D. Chlorine
E. Phosgene

43. The nerve agents (tabun, sarin, soman, and VX) antidote kits consisting of prefilled autoinjectors designed for the rapid administration of

A. Pralidoxime and pyridostigmine
B. Pralidoxime and diazepam
C. Atropine and pralidoxime
D. Atropine and diazepam
E. Atropine, pralidoxime, and diazepam

44. What is the **MAJOR** presentation of envenomations due to snakes, spiders, scorpions, and other venomous animals?

A. Acute compartment syndrome
B. Coagulopathy
C. Anaphylaxis
D. Localized pain and swelling
E. Infected wounds

45. A 10-year-old boy presents to the E/D with large lacerated wound in his left thigh due to dog bite. After appropriate material has been obtained for culture, the wound is anesthetized, cleaned, and prepared for vigorous irrigation.
Of the following, the **MOST** appropriate irrigation method is

A. copious irrigation with sterile saline
B. Irrigation with antibiotic-containing solutions
C. irrigation with catheter
D. blunt-tipped needle irrigation
E. high-pressure irrigation

46. Which of the following antimicrobials can be considered as **FIRST CHOICE** for the treatment of most infected wounds caused by animal and human bites?

A. Clindamycin

B. Trimethoprim/sulfamethoxazole
C. Ciprofloxacin
D. Cefotaxime
E. Amoxicillin/clavulanic acid

47. Which of the following is the drug of **CHOICE** for the treatment of rat bite fever?

A. Penicillin G
B. Doxycycline
C. Gentamicin
D. Streptomycin
F. Clindamycin

Chapter 33

Environmental health Answers

HAIDAR A. N. ABOOD

1.(B) Exposures to mycotoxins have been linked to at least 2 conditions that affect children: neural tube defects and acute pulmonary hemorrhage. Other conditions are linked to other environmental exposure; cerebral palsy to methylmercury, thyroid cancer to radiation, kidney stones to melamine, and chloracne to dioxin.

2.(A) Nitrates and nitrites not only help kill bacteria, but also produce a characteristic flavor and give meat a pink or red color. Nitrite salts are most often used in curing meat. Nitrite poisoning usually occurred 1–2 hr following eating cured meals, any contaminated foods with nitrates, or spinach exposed to excessive nitrification. Signs and symptoms include nausea, vomiting, cyanosis, headache, dizziness, weakness, loss of consciousness, chocolate brown–colored blood. It is usually self-limited. Treatment is supportive care; methylene blue may be required to correct methemoglobinemia.

3.(E) Average radiation doses (mSv) by Imaging test for pediatric population:

- ✓ Interventional fluoroscopy: AP & Lat abdomen _____(0.2-1.1 mSv/min)
- ✓ Interventional fluoroscopy: head _____(0.02-0.08 mSv/min)
- ✓ Interventional fluoroscopy: cardiac _____(0.1-1 mSv/min)
- ✓ Digital radiography: 2 view chest _____(0.04-0.06)
- ✓ Digital radiography: 2 view abdomen _____(0.1-0.4)
- ✓ Computed tomography: brain _____(0.8-4)
- ✓ Computed tomography: chest _____(1-4)
- ✓ Computed tomography: abdomen/pelvis _____(2-7)
- ✓ Nuclear medicine (^{99m}Tc methylene diphosphonate–Bone) (5-7)
- ✓ Positron emission tomography (^{18}F-FDG; whole body) ____(3-15)

There is clear evidence of radiation-induced cancer risk for whole body exposure >100 mSv, whereas no direct epidemiological data supports exposure

<10 mSv. What is unclear is the cancer risk for calculated effective dose levels between 10 and 100 mSv.

4.(C) Tissue radiosensitive weighting factors (W_T) according to international commission on radiological protection report 103:

- ✓ Red bone-marrow, colon, lung, stomach, breast, remainder tissues (0.12)
- ✓ Gonads (0.08)
- ✓ Bladder, oesophagus, liver, thyroid (0.04)
- ✓ Bone surface, brain, salivary glands, skin (0.01)

5.(A) Compared with middle-aged adults, children are generally are 2 times more sensitive to radiation-induced carcinogenesis, and neonates are more sensitive than older children.

6.(D)

7.(A) One of the most severe acute reactions is pneumonitis. It can be manifest within 24 hr of irradiation when there is an exudation of proteinaceous material into the alveoli and intraalveolar edema. Most often, radiation pneumonitis begins 2-6 mo after the beginning of radiation with a clinical presentation of fever, cough, congestion, and pleuritic pain.

8.(D) Radiation therapy increases the risk of second cancers in a dose-dependent manner for nongenetic neoplasms. Almost 70% of the second neoplasms are in the field of the original irradiation. Primary malignancies with the highest cumulative incidence of a second neoplasm in the order of frequency are Hodgkin disease (7.6), soft tissue sarcoma (4.0), cancers of bone (3.3), leukemia (2.1), central nervous system (CNS) cancers (2.1), and non-Hodgkin disease lymphoma (1.9). This reflects an overall standard incidence rate of 6.38%. The most prevalent second tumors are bone, breast, thyroid, and CNS lesions.

9.(C) Expected outcome based on absolute lymphocyte count after acute penetrating whole-body irradiation:

Minimal ALC within First 48 hr after exposure	**Prognosis**
✓ 1,000-3,000 (normal range)	No significant injury
✓ 1,000-1,500	Significant but probably nonlethal injury, good prognosis
✓ 500-1,000	Severe injury, fair prognosis
✓ 100-500	Very severe injury, poor prognosis
✓ <100	Lethal without compatible bone marrow donor

10.(B) The hand is the most common site for accidental localized irradiation injuries, usually as a result of picking up or playing with lost radiation sources. The second most common accidental site is the thigh and buttocks, predominantly from placing unsuspected highly radioactive sources in the pockets.

11.(A) Nuclear regulatory commission guidelines on breastfeeding during the period of a nuclear medicine examination recommended complete cessation of breast feeding if ^{131}-I Sodium iodide is administered to a lactating mother, and stopping breast feeding for 24 hr if ^{99m}Tc pertechnetate is used, while no interruption in breast feeding is required for other distracters.

12.(C) The most effective treatment of internal contamination with radionuclide requires knowledge of both the radionuclide and the chemical form. Treatment must be instituted quickly to be effective. Blocking therapy is the administration of potassium iodine or other stable iodine containing compounds to patients with known internal contamination with radioactive iodine. The stable iodine effectively blocks the thyroid, although its effectiveness decreases rapidly as time elapses after the contamination.

13.(B) There is increased risk of sudden infant death syndrome and asthma in infants exposed to air pollution and environmental tobacco smoke. Benzene is associated with childhood cancer, lead with neurobehavioral toxicity, while methyl mercury, and organophosphate insecticides are associated with developmental neurotoxicity.

14.(E) Fuel combustion is the principal source of air pollution. In high- and middle-income countries, combustion of fossil fuels (coal, oil, and gas) accounts for most air pollution. In low- and lower-middle-income countries, the major source is burning of biomass: wood, dung, straw, and charcoal. Coal is the single most highly polluting fossil fuel and also the most important source of the greenhouse gas emissions that drive global climate change.

15.(D) Pesticides can cause a range of chronic toxic effects that include: polyneuropathy and central nervous system dysfunction (organophosphates); hormonal disruption and reproductive impairment (DDT, kepone, dibromochloropropane); cancer (aldrin, dieldrin, chlorophenoxy herbicides [2,4,5-T]); and pulmonary fibrosis (paraquat).

16.(C) Immunotherapy reduces the risk of systemic anaphylaxis from future stings in high-risk patients from somewhere between 30% and 60% to <5%.

17.(D) Most scorpion envenomations occur in the southwestern United States, and fatalities are rare. In other regions of the world-especially Latin America,

Africa, the Middle East, and Asia-a number of scorpions regularly cause fatalities.

18.(A) Tobacco smoke exposure can be considered a modifiable risk factor for children with sickle cell disease and with CF. Children with sickle cell disease who are exposed to second-hand tobacco smoke have increased morbidity, specifically increased rates of Emergency Department visits and hospitalizations for vaso-occlusive crisis and acute chest syndrome. In addition, tobacco smoke exposure is also associated with pulmonary function abnormalities among children with sickle cell disease, independent of their baseline disease.

19.(E) The best method to treat children's SHS exposure is to eliminate this exposure by helping parents quit smoking. Methods to reduce exposure such as "smoking outside" or wearing a "smoking jacket" have not been shown to eliminate biochemically confirmed SHS exposure. The 5 A's method (Ask, Advise, Assess, Assist, and Arrange) was developed for adults. The pediatric model has been abbreviated to the Ask, Advise, and Refer. The Clinical Effort Against Secondhand Smoke Exposure (CEASE) is a program that trains pediatricians and their office staff to systematically provide cessation counseling and interventions to parents and other adults who smoke, while offering more assistance in quitting smoking than Ask/Advise/Refer.

20.(A) Lead, mercury, arsenic, and cadmium, four of the World Health Organization's (WHO) "Ten chemicals of greatest public health concern," are the heavy metals posing the greatest threats to humans. The most prevalent of these exposures is lead. It is estimated that 99% of lead-poisoned children are identified by screening procedures rather than through clinical recognition of lead related symptoms. Exposure to thallium is relatively rare.

21.(B) Most scorpion stings do not produce severe effects and require only wound care and orally administered pain medications. However, patients with more severe symptoms may require intravenous opioids for analgesia and benzodiazepines for severe muscle spasm or agitation. The antivenom is recommended for critically ill patients with neurotoxicity or other severe symptoms, including intractable pain that is not responsive to adequate doses of opioid analgesics.

22.(B) Arsine gas is colorless, odorless, nonirritating, and highly toxic. After exposure to arsine gas, absorbed arsine enters RBCs and is oxidized to arsenic dihydride and elemental arsenic. Complexing of these derivatives with red cell sulfhydryl groups results in cell membrane instability and massive hemolysis. Other forms of arsenic cause the other distractors.

23.(C) Subacute exposure to arsenic is characterized by prolonged fatigue, malaise, weight loss, headache, chronic encephalopathy, peripheral sensorimotor neuropathy, leukopenia, anemia, thrombocytopenia, chronic cough, and gastroenteritis. Mees lines (transverse white striae on the nails) become apparent 1-2 months after exposure in approximately 5% of patients. Dermatologic findings include alopecia, oral ulceration, peripheral edema, a pruritic macular rash, and desquamation. Chronic arsenic toxicity causes significant morbidity in children resulting in skin lesions, lung disease, and defect in intellectual function. Chronic exposure to low levels of arsenic is usually from environmental or occupational sources.

24.(B) Because urinary excretion of arsenic is intermittent, definitive diagnosis depends on a 24 hr urine collection. Concentrations greater than 50 µg/L in a 24 hr urine specimen are consistent with arsenic intoxication. Blood arsenic levels rarely are helpful because of their high variability and the rapid clearance of arsenic from the blood in acute poisonings. Elevated arsenic values in the hair or nails must be interpreted cautiously because of the possibility of external contamination.

25.(C) Mercury exists in three forms: elemental mercury, inorganic mercury salts, and organic mercury. Elemental mercury is present in thermometers, sphygmomanometers, barometers, batteries, and gold or silver smelting processes. Dental amalgams containing elemental mercury release trace amounts of mercury. Severe inhalation poisonings have resulted from attempts to separate gold from gold ore by heating mercury and forming a gold mercury amalgam. Inhaled elemental mercury vapor is 80% absorbed by the lungs and is distributed rapidly to the central nervous system because of its high lipid solubility. Elemental mercury liquid is poorly absorbed from the gastrointestinal tract, with less than 0.1% being absorbed.

26.(D) Acute ingestion of inorganic mercury salts (typically secondary to ingestion of a button battery) can manifest in a few hours as corrosive gastroenteritis, signified by metallic taste, oropharyngeal burns, nausea, hematemesis, severe abdominal pain, hematochezia, acute tubular necrosis, cardiovascular collapse, and death.

27.(E) Chronic inorganic mercury intoxication produces the classic triad consisting of tremor, neuropsychiatric disturbances, and gingivostomatitis. The syndrome may result from long-term exposure to elemental mercury, inorganic mercury salts, or certain organic mercury compounds, all of which may be metabolized to mercuric ions. The tremor starts as a fine intention tremor of the fingers that is abolished during sleep but that may later involve the face and

progress to choreoathetosis and spasmodic ballismus. Mixed sensorimotor neuropathy and visual disturbances may also be present. The neuropsychiatric disturbances include emotional lability, delirium, headaches, memory loss, insomnia, anorexia, and fatigue. Renal dysfunction ranges from asymptomatic proteinuria to nephrotic syndrome.

28.(E) Acute ingestion of inorganic arsenic and mercury salts results in hemorrhagic gastroenteritis, cardiovascular collapse, and multi-organ dysfunction. Because of the corrosive effects of these compounds, induced emesis is not recommended, and endoscopy may be considered before gastric lavage. Arsenic and mercury are not well adsorbed to activated charcoal. Whole-bowel irrigation is used to remove any radiopaque material remaining in the gastrointestinal tract. Chelation for acute arsenic and mercury poisoning is most effective when administered as soon as possible after the exposure. Chelation should be continued until 24 hr urinary arsenic or mercury levels return to normal (<50 µg/L for arsenic and <20 µg/L for mercury), the patient is symptom-free, or the remaining toxic effects are believed to be irreversible. Dimercaprol, also known as British antilewisite (BAL), is the chelator of choice for a patient who cannot tolerate oral therapy, as often is true for critically ill patients and after ingestion of the corrosive inorganic arsenic and mercury salts.

29.(A) Oral chelating agents are used to replace the painful BAL injections when the patient is stable enough to tolerate oral therapy and prolonged chelation is necessary. Succimer, also known as 2,3-dimercaptosuccinic acid (DMSA), is an orally administered water-soluble derivative of BAL. D-Penicillamine is an orally administered chelator that can be considered for less-severe mercury poisoning or as an adjunct to BAL therapy in arsenic poisoning, but its use is largely restricted because of the potential for significant leukopenia, thrombocytopenia, and proteinuria. A newer investigational analog, N-acetyl-D,L-penicillamine, is used with variable success in mercury poisoning. $CaNa_2EDTA$ is used for lead poisoning.

30.(B)

31.(C) Indications for antivenin administration in snake envenomation are:

- ✓ Evidence of clinically significant bleeding
- ✓ Platelet count below 25,000/mm^3
- ✓ International normalized ratio (INR) >3
- ✓ Activated partial thromboplastin time (aPTT) >50 sec
- ✓ Fibrinogen <50 mg/dL

If tourniquets are placed in the field, they should be cautiously removed after venous access is obtained due to possible adverse effects that may follow from a sudden release of venom into the systemic circulation.

32.(D) The nonnutritive hand-to-mouth activity of young children is the most common pathway for lead to enter the body. In most cases, lead is ingested, either as a component of dust licked off of surfaces or in swallowed paint chips, through water contaminated by its flow through lead pipes or brass fixtures, or from contaminated foods or liquids. Cutaneous contamination with inorganic lead compounds, such as those found in pigments, does not result in a substantial amount of absorption. Organic lead compounds, such as tetraethyl lead, may penetrate through skin, however.

33.(E)

34.(C) Prehospital care should focus on rapid transport to the emergency department while providing supportive care. Constrictive clothing, jewelry, and watches should be removed, and the injured body part should be immobilized in a position of function at the level of the heart. Many popularized field treatments for snake bites—such as tourniquets, ice, electric shock, incision, and suction—have proven ineffective or deleterious.

35.(A) Gastrointestinal symptoms of lead poisoning include anorexia, abdominal pain, vomiting, and constipation, often occurring and recurring over a period of weeks. Children with blood lead levels (BLLs) > 20 μg/dL are twice as likely to have gastrointestinal complaints as those with lower BLLs. Central nervous system symptoms are related to worsening cerebral edema and increased intracranial pressure. Headaches, change in mentation, lethargy, papilledema, seizures, and coma leading to death are rarely seen at levels lower than 100 μg/dL but have been reported in children with a BLL as low as 70 μg/dL. Erythrocyte protoporphyrin levels > 35 μg/dL are abnormal and are consistent with lead poisoning, iron deficiency, or recent inflammatory disease. Radiographs of long bones may show dense bands at the metaphyses. Most cases of anemia in lead-poisoned children are a result of other factors, such as iron deficiency and hemoglobinopathies.

36.(B) Following neurotoxic snake envenomation, manifestations of neurotoxicity generally are caused by curare-like blockade at the neuromuscular junction. Symptoms usually begin with cranial nerve palsies such as ptosis, dysarthria, and dysphagia and may progress to respiratory failure and complete paralysis.

37.(B) Children with BLLs (blood lead levels) of 44-70 μg/dL may be treated with a single drug, preferably DMSA. Those with BLLs of 70 μg/dL or greater require

two-drug treatment: $CaNa_2$ EDTA in combination with either DMSA or BAL for those without evidence of encephalopathy, or $CaNa_2$ EDTA and BAL for those with encephalopathy.

38.(D) Poisonings by species of Amanita (death cap mushroom) and Galerina account for 95% of the fatalities from mushroom intoxication.

39.(A) General management of mushroom ingestion:

- ✓ Determine history of ingestion
- ✓ Attempt to determine the possible syndromes
- ✓ Administer activated charcoal.
- ✓ If feasible and when indicated; identification of mushroom by mycologist.
- ✓ Try to perform a preliminary identification of mushroom and spores.
- ✓ Maintain supportive measures
- ✓ Avoid antispasmodics for gastrointestinal symptoms.
- ✓ Anticipate the clinical course.

40.(B) Scombroid (pseudoallergic) fish poisoning: Ingestion of members of the Scombridae families, including albacore, mackerel, tuna, bonita, and kingfish, have been linked to pseudoallergic fish poisoning. The bacterial transformation of histidine to histamine (if refrigeration is inadequate) is responsible for the clinical syndrome. Histidine is found in high concentrations in the flesh of scombroid fish. The onset of clinical manifestations is acute and occurs within 10 min to 2 hr of ingestion. The most common symptoms and signs are diarrhea, erythema, sweating, flushing, diaphoresis, urticaria, nausea, and headache. Abdominal pain, tachycardia, oral burning or numbness, dizziness, respiratory distress, hives, and facial swelling also occur. The illness is usually self-limited, terminating within 8-24 hr.

41.(D) Melamine contains 66% nitrogen by mass. The illegal addition of melamine to infant formula can give the formula a milky appearance and falsely raise the protein content as measured by nitrogen testing. Melamine, combined with cyanuric acid, forms cyanurate crystals in the kidneys. Along with protein, uric acid, and phosphate, melamine forms renal calculi.

42.(A) Following exposure to mustard (a blistering agent) during terrorist attacks, dermatologic findings are prominent, while for other distracters respiratory findings are prominent.

43.(C)

44.(D) Envenomations due to snakes, spiders, scorpions, and other venomous animals can cause significant morbidity and mortality, although the majority cause only localized pain and swelling.

45.(A) After appropriate material has been obtained for culture, the animal bite wound should be anesthetized, cleaned, and vigorously irrigated with copious amounts of sterile saline. Irrigation with antibiotic-containing solutions provides no advantage over irrigation with saline alone and may cause local irritation of the tissues. Puncture wounds should be thoroughly cleansed and gently irrigated with a catheter or blunt-tipped needle; high-pressure irrigation should not be employed. Avulsed or devitalized tissue should be debrided and any fluctuant areas incised and drained.

46.(E) The following antimicrobials can be considered for most terrestrial animal and human bites.

- ✓ First choice: Amoxicillin/clavulanic acid
- ✓ Penicillin allergy, the following regimens can be considered
  - Clindamycin plus ciprofloxacin or levofloxacin or trimethoprim/sulfamethoxazole
  - Doxycycline
  - Moxifloxacin
- ✓ In cases where intravenous antibiotics are deemed necessary, single antimicrobial choices can include ampicillin/sulbactam, cefoxitin, ertapenem, or moxifloxacin.

47.(A) Penicillin is the drug of choice for both forms of rat bite fever. Intravenous penicillin G or intramuscular penicillin G procaine is recommended for 7-10 days. Doxycycline, gentamicin, or streptomycin represents effective alternatives for penicillin allergic patients.

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Chapter 34

Laboratory Medicine Questions

HAIDAR A. N. ABOOD

1. Which of the following variables has no considerable effects when developing reference laboratory values for pediatric population?

A. Genetic composition
B. Physiologic development
C. Environmental influences
D. Subclinical disease
E. Socioeconomic status

2. Which of the following terms is a measure of the nearness of a test result to the actual value?

A. Precision
B. Accuracy
C. Sensitivity
D. Specificity
E. Reproducibility

3. Assume you have the results of PCR test for COVID-19; the number of positive tests is 95 collected from a 100 truly-infected children.
The sensitivity of the tests is

A. 5%
B. 15%
C. 75%
D. 85%
E. 95%

4. Which of the following diseases requires further confirmative investigations in later life following early diagnosis through neonatal screening programs?

A. Phenylketonuria
B. Galactosemia
C. Hypothyroidism

D. Cystic fibrosis
E. Sickle cell disease

5. Which of the following analytes is measured in serum?
A. Immunoglobulin E
B. Carbon dioxide, partial pressure
C. Antidiuretic hormone
D. Ammonia
E. Magnesium

6. What is the best analytical technique for neonatal screening program for inborn error of metabolism?
A. Enzyme-amplified immunoassay
B. Atomic absorption
C. High-performance liquid chromatography (HPLC)
D. Tandem mass spectrometry (MS/MS)
E. Gas chromatography mass spectrometry (GC/MS)

7. Which of the following analytes is measured in plasma?
A. Bilirubin
B. Aspartate transaminase
C. Albumin
D. C-reactive protein
E. Base excess

8. Assume you are evaluating the specificity of an immunoassay test for rapid diagnosis of COVID-19; the number of normal children tested is 150 and the test is positive in 30 children.
The specificity of the tests is
A. 30%
B. 50%
C. 70%
D. 80%
E. 90%

9. What is the best analytical technique for HIV screening?
A. Atomic absorption
B. ELISA

C. High-performance liquid chromatography (HPLC)
D. PCR
E. Flowcytometry

10. Which of the following analytes is measured in whole blood?
A. Iron
B. TSH
C. Uric acid
D. Lead
E. Potassium

Chapter 34

Laboratory Medicine Answers

HAIDAR A. N. ABOOD

1.(E) Normal values (reference intervals) are difficult to establish within the pediatric population. Differences in genetic composition, physiologic development, environmental influences, and subclinical disease are variables that need to be considered when developing reference intervals.

2.(B) Accuracy is a measure of the nearness of a test result to the actual value, whereas precision is a measure of the reproducibility (how close are repetitive measurements of a single sample) of a result. Sensitivity is ability to detect a disease and specificity is ability to define the absence of a disease.

3.(E) Sensitivity = number positive by test x 100 / total number positive

4.(D) The results of neonatal screening for cystic fibrosis show clear benefits associated with preclinical diagnosis, but some inherent difficulties associated with genetic screening for complex autosomal recessive diseases that are common and are caused by a large number of mutations (>1,500) of a single gene. The definitive diagnostic test for CF is the measurement of concentrations of chloride in sweat, a test that is not practical during the first week of life.

5.(A) Immunoglobulin E

Carbon dioxide, partial pressure (whole blood), Antidiuretic hormone (plasma), Ammonia (plasma), Magnesium (plasma).

6.(D) Tandem mass spectrometry (MS/MS) is a technically advanced method in which many compounds are initially fragmented and separated by molecular weight. Each compound is then fragmented again. Identification of compounds is based on characteristic fragments. The process requires approximately 2 min per sample and can detect 20 or more inborn errors of metabolism.

7.(C) Albumin

Bilirubin (serum), Aspartate transaminase (serum), C-reactive protein (serum), Base excess (whole blood).

8.(D) Specificity = number negative by test x 100 / total number without disease

9.(B)

10.(D) Lead.

Iron (plasma), TSH (serum), Uric acid (serum), Potassium (serum).

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ZUHAIR M. ALMUSAWI
CABP, IBCLC, FRCP Edin
PROFESSOR OF PEDIATRICS
COLLEGE OF MEDICINE / UNIVERSITY OF KERBALA
CONSULTANT PEDIATRICIAN
KARBALA TEACHING HOSPITAL FOR CHILDREN
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