ESSENTIAL MCQs IN PEDIATRICS

PART V

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DEDICATION

In loving memory of the professors **(Mahjoub Naddawi, Ibraheem Al-Nasir, Najla Ayub, and Najm Aldeen Rouznamaji)** who shaped our minds and hearts to be in love with pediatrics as a branch of medicine.

This book is dedicated to the cherished memory of those professors who touched our lives in profound ways. They were more than educators; they were beacons of pediatrics. Though they are no longer with us, their impact lives on through the countless students whose lives they framed.

Their way of teaching and mentorship continue to guide and shape our understanding of pediatrics, fuelling our passion for providing exceptional care to the sick children.

We are forever grateful for the knowledge, wisdom, and inspiration they bestowed upon us.

Each page of this book represents a witness to their enduring legacy.

Special thanks are extended to **Sumaya Alhameed** for her help in designing the cover of this book.

With respect and gratitude

ZUHAIR ALMUSAWI

PREFACE

This book takes readers on a journey into child health, unleashing the wonders of pediatrics. As you navigate through the pages of this book will embark on the captivating world of pediatrics, where every question brings forth new discoveries, challenges, or even opportunities to improve the lives of our precious little ones. It merges the scientific knowledge, clinical expertise, and a deep understanding of the unique needs of the undergraduates and postgraduate students studying pediatrics.

Within these pages, you will delve into the multifaceted world of pediatric MCQs, exploring the wide range of topics that span from the earliest moments of life to the transformative years of adolescence, unravelling the mysteries of different MCQs.

This book serves as a guide for the undergraduates and postgraduate students. It is a testament to the dedication and tireless efforts of authors who have worked tirelessly to nurture the field of pediatrics by more relevant MCQs.

You will encounter carefully crafted multiple-choice questions covering a wide range of pediatric topics. Each question is designed to test your knowledge, critical thinking abilities, and clinical decision-making skills. As you progress through the chapters, you will have the opportunity to explore various aspects of pediatrics, including development, neonatology, general pediatrics, and different pediatric subspecialties.

The MCQs are inspired by the clinical expertise of the authors, to offer a masterpiece that stands the test of time as a cornerstone in the field of pediatrics.

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SECTION 1

THE PROFESSION OF PEDIATRICS

AHMED TAWFEQ

QUESTIONS

1. The statement that explaining the term “integrity” in profession of pediatrician is
   A. awareness of situations that may result in conflict of interest or that may result in personal gain at the expense of the best interest of the patient
   B. accountability to one’s patients, their families, society, and the medical community to ensure that all needs are addressed
   C. treat all persons with respect and regard for their individual worth and dignity; be aware of emotional, personal, family, and cultural influences on a patient’s well-being
   D. listen attentively, respond humanely to patient/family concerns

2. In a case scenario when you decide to do an urgent surgery to a school age child had a life-threatening condition and in absence of his parents for a reason or another.
This ethical principle is called
   A. beneficence
   B. non maleficence
   C. justice
   D. paternalism

3. As ethical principle obtaining assent in older children means that
   A. giving full consent for the wanted procedure
   B. involving in decision-making with information appropriate to the child’s capacity to understand
   C. sharing consent between parents and the child
   D. sparing out the child opinion

4. Parents who had a positive family history of AD polycystic kidney disease are requesting a genetic testing for their 6-month-old child who’s free of symptoms.
Of the following, the **BEST** response would be
   A. offering the genetic testing
   B. deferring the genetic testing
   C. referring the child to a nephrologist
   D. waiting till the child can give informed consent

5. A parent with seriously ill 6-month-old infant offered by licensed researcher to participate in treatment trial of new medication for their child disease.
Of the following, the **MOST** appropriate action would be
   A. allow it within the local government legislations
   B. defer the participation
   C. allow to use the established medications only
   D. notify as fraud

6. Which of the following category of palliative care can be applied in cystic fibrosis patient?
   A. When a cure is possible but unlikely
   B. Long-term treatment with a goal of maintaining quality of life
   C. Treatment that is exclusively palliative after the diagnosis of a progressive condition is made
   D. When treatments are available for severe, non-progressive disabilities

7. Organ donation in pediatrics can occur only after
   A. fulfilling criteria for brain death
   B. fulfilling criteria for circulatory death
   C. any one of the above can be applicable
   D. none is acceptable any more in recent legislations

8. What is major cause for death in children 1-4 years old in US?
   A. Unintentional injuries
   B. Congenital anomalies
   C. Malignant neoplasms
   D. Influenza/pneumonia

9. Which of the following health challenges in adolescents is **MOST** strikingly increased in last few years?
   A. Alcohol consumption
   B. Drug abuse
   C. Vaping
   D. HIV infection

10. You are explaining the paradigm "changing morbidity" to the postgraduate pediatrician in the context of increase in mental illnesses in teenage group.
Of the following, the **MOST** accurate statement would be
   A. About 2% of children < 10 years may experience mental illness
   B. About 5% of children between 10-15 years may have mental health condition
   C. Only 10% of children of all age groups have suffered mental problem
   D. About 20% of children aged 13-18 years old has a mental health condition

SECTION 1

THE PROFESSION OF PEDIATRICS

AHMED TAWFEQ

ANSWERS

1.**(A)** B is referring to Reliability/responsibility, C is referring to respect for others, While D is referring to compassion/empathy, all of these terms under the tile of professionalism.

2.**(D)** Beneficence (doing good), nonmaleficence (doing no harm or as little harm as possible), and justice (the values involved in the equal distribution of goods, services, benefits, and burdens to the individual, family, or society). Paternalism challenges the principle of autonomy and involves the clinician deciding what is best for the patient. Under certain circumstances (e.g., when a patient has a life-threatening medical condition or a significant psychiatric disorder and is threatening self or others), paternalism is an accepted alternative to autonomy.

3.**(B)** Infants and young children do not have the capacity for making medical decisions, thus decision-making falls onto their parents. Parental decision-making should be guided by what is in the best interest of the child. In certain older children, assent can be obtained for medical decision-making. Obtaining the assent of a child is the process in which the child is involved in decision-making with information appropriate to the child's capacity to understand.

4.**(D)** For their own purposes, parents may pressure the pediatrician to order genetic tests when the child is still young. Testing for (polycystic kidney disease; Huntington disease; certain cancers, such as breast cancer in some ethnic populations; and hemochromatosis) should be delayed until the child has the capacity for informed consent or assent and is competent to make decisions, unless there is a direct benefit to the child at the time of testing.

5.**(A)** Many parents with seriously ill children hope that the research protocol will have a direct benefit for their child. The researcher should be sensitive and compassionate in explaining to the parents that research does not guarantee a (better) treatment. Special federal regulations have been developed to protect child and adolescent participants in human investigation, these regulations provide additional safeguards beyond those for adult participants while still providing the opportunity for children to benefit from the scientific advances of research.

6.**(B)** The death of a child is one of life's most difficult experiences. The palliative care approach is defined as patient- and family centered care that optimizes quality of life by anticipating, preventing, and treating suffering. This approach should be instituted when medical diagnosis, intervention, and treatment cannot reasonably be expected to affect the imminence of death. Examples for A is cancer, B is cystic fibrosis, C trisomy 13, D severe spastic quadriparesis with difficulty in controlling symptoms.

7.**(C)** Organ donation can occur one of two ways: after fulfilling criteria for neurologic (brain) death or through a process of donation after circulatory death (DCD). DCD has only recently gained acceptance in pediatrics. Organ donation and organ transplantation can provide significant life-extending benefits to a child who has a failing organ and is awaiting transplant, while at the same time place a high emotional impact on the donor family after the loss of a child.

8.**(A)** The overall causes of death in all children (1–24 years of age) in the United States in 2018, in order of frequency, were unintentional injuries (accidents), congenital anomalies, suicide, homicide, and malignant neoplasms.

9.**(C)** There is increasing substance use and abuse among U.S. adolescents. Fourteen percent of high school students report using some illicit or injection drugs, and 14% report using non-prescription opioids, both placing youth at risk for overdose and HIV infection. In 2017, 30% of surveyed high school students drank alcohol, 14% binge drank, and 17% rode in the car with someone who had been drinking. Tobacco cigarette use has declined among teens over the past decade, but the rate of e-cigarette use (or vaping) has increased from 1.5% in 2011 to 27.5% in 2019. Vaping has been proven to pose significant health risks to both users and nonusers.

10.**(D)** An estimation 1 in 5 children, ages 13–18 years, has a mental health condition. Half of all lifetime cases of mental illness begin by age 14 years. The average delay between onset of symptoms and intervention is 8–10 years. Suicide is the second leading cause of death for children ages 10–24 years, making early recognition of mental illness paramount.

SECTION 2

GROWTH AND DEVELOPMENT

MOHAIMEN ALKABI

QUESTIONS

1. What is the daily weight gain in the first 3–4 months of life?
   A. 10–20 gm
   B. 20–30 gm
   C. 30–40 gm
   D. 40–50 gm

2. What is the average length of a 1-year-old infant?
   A. 10 inches
   B. 20 inches
   C. 30 inches
   D. 40 inches

3. What is the average head circumference of a 3-month-old infant?
   A. 35 cm
   B. 38 cm
   C. 41 cm
   D. 44 cm

4. The mother of a 6-year-old boy is seeking your advice about the readiness of her boy to attend school.
Which of the following behavior is indicative of his readiness for school?
   A. Dresses and undresses with assistance
   B. Count to 4
   C. Knowledge of colors
   D. Dry at night

5. Which of the following is the **MOST** appropriate initial method to assess the development of children more than 6 years of age?
   A. Denver Developmental Screening Test II
   B. Ages and Stages Questionnaires
   C. Parents' Evaluation of Developmental Status
   D. School performance

6. The American Academy of Pediatrics recommends that all infants are screened for anemia at the age of
   A. 6 months
   B. 9 months
   C. 12 months
   D. 15 months

7. Low birth weight or premature infants are screened for anemia at birth and again at the age of

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

8. All adolescents should start annual screening for depression at the age of

A. 11 years
B. 13 years
C. 15 years
D. 17 years

9. Which is the **BEST** car safety issues for infant and toddler less than 2 years old?

A. Rear-facing safety seat
B. Forward-facing car seat
C. Lap and shoulder seat belts
D. Belt-positioning booster seat

10. Salma sits unsupported on the examination table, bangs two blocks together, say mama and dada in nonspecific way, and wave bye-bye.
What is the expected age for Salma?

A. 6 months
B. 9 months
C. 12 months
D. 15 months

11. Ahmed walks few steps, put block in cup, drink from cup, and say mama, dada in a specific way.
What is the expected age for Ahmed?

A. 9 months
B. 12 months
C. 15 months
D. 18 months

12. You observe Layla in pediatric inpatient ward. She runs toward her mother, kicks the ball and feeds the doll.
What is the expected age for Layla?

A. 12 months
B. 15 months
C. 18 months
D. 24 months

13. Suha walks backward, uses spoon and fork, scribbles, and follows a command.
What is the expected age for Suha?

A. 12 months

B. 15 months
C. 18 months
D. 24 months

14. Ali hops on one foot, brushes teeth without help, dresses alone, and names the colors.
What is the expected age for Ali?
A. 24 months
B. 30 months
C. 36 months
D. 48 months

15. At which age, the child begins to understand right and left?
A. 3 years
B. 4 years
C. 5 years
D. 6 years

16. At which age the child can understand the opposites?
A. 3 years
B. 4 years
C. 5 years
D. 6 years

17. The child can understand the concept of today, tomorrow and yesterday by the age of
A. 24 months
B. 36 months
C. 48 months
D. 6o months

18. At what age a child can skip?
A. 2 years
B. 3 years
C. 4 years
D. 5 years

19. What is the **BEST** age for vision screening?
A. 2 years
B. 3 years
C. 4 years
D. 5 years

20. What is the **MOST** common cause of severe visual impairment in children?
A. Congenital cataract
B. Congenital glaucoma
C. Retinopathy of prematurity

D. Optic neuritis

21. What is the **MOST** common presentation of mild to moderate visual impairment in children?

A. Myopia
B. Hyperopia
C. Astigmatism
D. Amblyopia

22. What is the **MOST** common cause of mild to moderate hearing loss in children?

A. Chronic otitis media
B. Congenital rubella infection
C. Congenital cytomegalovirus
D. Kernicterus

23. Which of the following medications may cause sensorineural hearing loss in children?

A. Erythromycin
B. Cephalexin
C. Gentamicin
D. Ampicillin

24. Which of the following hearing tests is required for all newborn prior to nursery discharge?

A. Tympanometry
B. Pure-tone audiometry
C. Behavior audiology
D. Otoacoustic emissions

25. An evaluation for primary amenorrhea should be considered for any female adolescent who has not reached menarche by

A. 13 years
B. 14 years
C. 15 years
D. 16 years

26. Which of the following developmental milestone indicate communication disorder for a 2-year-old child?

A. The child is not using short sentences (e.g., “Daddy went bye-bye”)
B. The child has not begun to ask questions (using where, what, why)
C. The child does not point to body parts without gestural cues
D. The child’s speech is not understood by unfamiliar listeners

27. Which of the following is the **MOST** common cause of acquired cerebral palsy?

A. Near drowning
B. Stroke
C. Hypoxia

D. Meningitis

28. Which of the following is the **MOST** common form of cerebral palsy?
A. Spastic
B. Dyskinetic
C. Ataxic
D. Dystonic

29. Which of the following conditions presents with hypertonia and developmental delay?
A. Prader-Willi syndrome
B. Down syndrome,
C. Angelman syndrome
D. Trisomy 18

30. Which of the following conditions presents with macrocephaly and developmental delay?
A. Angelman syndrome
B. Cornelia de Lange syndrome
C. Fetal alcohol effects
D. Sotos syndrome

SECTION 2

GROWTH AND DEVELOPMENT

MOHAIMEN ALKABI

ANSWERS

1.**(B)** Daily weight gain: 20–30 g for first 3–4 months ,15–20 g for rest of the first year.

2.**(C)** The average length is 20 in. at birth and 30 in. at 1 year.

3.**(C)** The average HC is 35 cm at birth (13.5 in.). HC increases: 1 cm per month for first year (2 cm per month for first 3 months, then slower)

4.**(C)**

TABLE 7.1 Evaluating School Readiness

PHYSICIAN OBSERVATIONS (BEHAVIORS OBSERVED IN THE OFFICE)
Ease of separation of the child from the parent
Speech development and articulation
Understanding of and ability to follow complex directions
Specific pre-academic skills
Knowledge of colors
Counts to 10
Knows age, first and last name, address, and phone number
Ability to copy shapes
Motor skills
Stand on one foot, skip, and catch a bounced ball
Dresses and undresses without assistance
PARENT OBSERVATIONS (QUESTIONS ANSWERED BY HISTORY)
Does the child play well with other children?
Does the child separate well, such as a child playing in the backyard alone with occasional monitoring by the parent?
Does the child show interest in books, letters, and numbers?
Can the child sustain attention to quiet activities?
How frequent are toilet-training *accidents*?

5.**(D)**

6.**(C)** Infants are screened at birth and again at 4 months if there is a documented risk, such as low birthweight or prematurity. All infants are screened at 12 months of age because this is when a high incidence of iron deficiency is noted.

7.**(B)**

8.**(A)** All adolescents, starting at age 11, should have annual depression screening with a validated tool.

9.**(A)** Infants and toddlers should ride in a rear-facing safety seat until they are at least 2 years old. It is considered safest for children to remain rear-facing after 2 years old for as long as possible while within weight and height limits set by the safety seat manufacturer.

10.**(B)** 9 months
Gross motor: Pulls to stand, gets into sitting position
Fine motor: Starting to pincer grasp, Bangs two blocks together
Personal-social: Waves bye-bye, plays pat-a-cake
Language: Says Dada and Mama, but nonspecific, two-syllable sounds

11.**(B)** 12 months
Gross motor: Walks, stoops and stands
Fine motor: Puts block in cup
Personal-social: Drinks from a cup, Imitates others
Language: Says Mama and Dada, specific, says one to two other words

12.**(D)** 18 months
Gross motor: Runs
Fine motor: Stacks four blocks, kicks a ball
Personal-social: Removes garment "Feeds" doll
Language: Says at least six words

13.**(B)** 15 months
Gross motor: Walks backward, stoops and recovers
Fine motor: Scribbles, stacks two blocks
Personal-social: Uses spoon and fork, helps in housework
Language: Says three to six words, follows commands

14.**(D)** 4 years
Gross motor: Balances well on each foot, hops on one foot
Fine motor: Draws person with three parts
Personal-social: Brushes teeth without help, dresses without help
Language: Names colors

15.**(D)** The child does begin to understand right and left at 6 years.

16.**(B)** The child can understand the opposites at 5 years.

17.**(B)** The child understands concepts of tomorrow and yesterday by 3 years and begin understand today by 2 years.

18.**(D)**

19.**(B)** At 3 years of age, children are screened for vision for the first time if they are developmentally able to be tested. Many children at this age do not have the interactive language or interpersonal skills to perform a vision screen; these children should be re-examined within 6 months to ensure that their vision is normal.

20.**(C)** The most common cause of severe visual impairment in children is retinopathy of prematurity.

21.**(A)** Children with mild to moderate visual impairment usually have an uncorrected refractive error. The most common presentation is myopia or near sightedness.

22.**(A)** The most common cause of mild to moderate hearing loss in children is a conduction abnormality caused by acquired middle ear disease (acute and chronic otitis media).

23.**(C)**

24.**(D)** Universal screening of newborns is required prior to nursery discharge and includes the following:
Auditory brainstem response (ABR) measures how the brain responds to sound. Clicks or tones are played through soft earphones into the infant's ears. Three electrodes placed on the infant's head measure the brain's response.
Otoacoustic emissions measure sound waves produced in the inner ear. A tiny probe is placed just inside the infant's ear canal. It measures the response (echo) when clicks or tones are played into the infant's ears.

25.**(C)** An evaluation for primary amenorrhea should be considered for any female adolescent who has not reached menarche by 15 years or has not done so within 3 years of thelarche (beginning of breast development). Lack of breast development by age 13 years also should be evaluated.

26.**(C)** Clues to when a Child with a communication disorder needs help in 24–36 months.

- By 24 months, at least 50% of the child's speech is not understood by familiar listeners.
- By 24 months, the child does not point to body parts without gestural cues.
- By 24 months, the child is not combining words into phrases (e.g., "go bye-bye," "go car," "want cookie").
- By 30 months, the child does not show understanding of spatial concepts: on, in, under, front, and back.
- By 30 months, the child is not using short sentences (e.g., "Daddy went bye-bye").
- By 30 months, the child has not begun to ask questions (using *where, what, why*).
- By 36 months, the child's speech is not understood by unfamiliar listeners.

27.**(D)** Meningitis and head injury (accidental and nonaccidental) are the most common causes of acquired CP.

28.**(A)** Spastic cerebral palsy: the most common form of cerebral palsy, it accounts for 70–80% of cases. It results from injury to the upper motor neurons of the pyramidal tract.

29.**(D)** **Hypotonia** with developmental delay presented in Prader-Willi syndrome, Down syndrome, Angelman syndrome, gangliosidosis, early cerebral palsy. While **Hypertonia** is present in Neurodegenerative conditions involving white matter, cerebral palsy, trisomy 18 syndrome.

30.**(D)** **Macrocephaly** with developmental delay presented in Alexander syndrome, Sotos syndrome, gangliosidosis, hydrocephalus, mucopolysaccharidosis, subdural effusion. While **microcephaly** presented in malnutrition, Angelman syndrome, Cornelia de Lange syndrome, fetal alcohol effects.

SECTION 3

BEHAVIORAL DISORDERS

ZUHAIR ALMUSAWI

QUESTIONS

1. A 2-month-old otherwise healthy infant brought by his anxious worried mother because of episodes of paroxysmal uncontrollable crying which last more than 3 hours associated with facial grimacing, leg flexion, and passing flatus. The crying start and stop without obvious cause and cannot be prevented or resolved.
Of the following, the **MOST** likely cause is

A. shaken baby syndrome
B. infantile colic
C. parental guilt
D. multiple physician visits

2. A young irritable pharmacist brought her 3-month-old bottle fed infant with excessive crying more than four hours, 4-5 days per week. The baby was examined by many pediatricians and their physical examination reveals no organic cause for the infant's excessive crying.
The mother used many medications with no much benefit, and she is asking about the **BEST** of the following

A. diphenhydramine
B. simethicone
C. lactase
D. probiotics

3. A 40-year-old mother visited you with her 3-month-old breast-fed healthy-looking infant who does not allow her to sleep each night because of excessive crying.
She is asking about the **MOST** serious complication of this crying

A. temper tantrum
B. maternal depression
C. nonaccidental trauma
D. failure to thrive

4. A 3-year-old girl presented with brief episodes of crying, screaming, and head banging 3-4 times per day, which lasts for 2-4 minutes. The girl looks normal between the attacks.
Of the following, the **MOST** likely cause is

A. attention-deficit/hyperactivity disorder
B. anxiety
C. conduct disorder
D. normal temper tantrum

5. A 7-year-old boy brought by his mother with failing to pay close attention and appearing to not listen when spoken to directly, together with fidgety, restlessness, and frequent interruption during history taking. Physical and developmental examination are normal.
Of the following, the core of overall treatment of this boy is

A. methylphenidate
B. behavioral therapy
C. atomoxetine
D. relaxation techniques

6. A 7-year-old healthy looking boy presented by his mother because of daily nocturnal bed wetting. The boy has no daytime incontinence or any associated urinary tract or neurologic symptoms. The mother wants a rapid cure due to her anxiety and poor coping.
Of the following, the recommended **FIRST-LINE** therapy is
A. alarm conditioning therapy
B. tricyclic antidepressants
C. desmopressin
D. anticholinergic medication

7. A 3-year-old child presents with chronic constipation and soiling with history of delayed passage of meconium, clinical examination revealed an empty rectum and tight sphincter.
Of the following, the **MOST** likely diagnosis is
A. Hirschsprung disease
B. hypothyroidism
C. anal stenosis
D. developmental delay

8. The parents of a preschool boy reports that their child has frequent soiling of liquid stool, on abdominal examination, you feel impacted stool with firm packed stool in the rectum.
Which of the following is **SUPERIOR** maintenance therapy for this boy?
A. Polyethylene glycol
B. Lactulose
C. Magnesium hydroxide
D. Bisacodyl

9. A 5-year-old boy awakened abruptly two hours after sleep with agitation and loud scream that is unresponsive to caregivers' attempts to console, with no recollection of what happens.
What is the **MOST** likely sleep disorder in this child?
A. Sleep terror
B. Confusional arousal
C. Nightmare
D. Irregular sleep-wake pattern

10. Which of the following drugs is useful in treating insomnia in children with ADHD?
A. Diphenhydramine
B. Clonidine
C. Clonazepam
D. Melatonin

SECTION 3

BEHAVIORAL DISORDERS

ZUHAIR ALMUSAWI

ANSWERS

1.**(B)** Infantile colic is defined as episodes of uncontrollable crying or fussing in an otherwise healthy infant. Colicky crying is often described as paroxysmal and may be characterized by facial grimacing, leg flexion, and passing flatus. It has been associated with shaken baby syndrome, postpartum depression, parental guilt, and multiple physician visits.
Varying diagnostic criteria for the actual medical definition for colic exist. **Wessel rule of threes** (crying for >3 hours per day, at least 3 days per week, for >3 weeks) has been replaced by the 2016 ROME IV criteria, which include the following:
1. Age <5 months when the symptoms start and stop
2. Recurrent and prolonged periods of crying, fussing, or irritability that start and stop without obvious cause and cannot be prevented or resolved by caregivers
3. No evidence of poor weight gain, fever, or illness
4. Caregiver reports of crying/fussing for ≥3 hours per day on ≥3 days/week in a telephone or face-to-face interview
5. Total daily crying confirmed to be ≥3 hours when measured by at least one prospectively kept 24-hour diary

2.**(D)** Medications, including phenobarbital, diphenhydramine, alcohol, simethicone, dicyclomine, and lactase, have no benefit and may cause serious side effects. Some studies have suggested that probiotics may be useful. The use of probiotics such as lactobacillus reuteri has been shown to decrease crying time in the infant with colic without adverse effects and can be considered after careful evaluation, certainty of diagnosis, and close follow-up.

3.**(C)** There is no evidence that infants with colic have adverse long-term outcomes in health or temperament after the affected period. Similarly, infantile colic does not have untoward long-term effects on maternal mental health, as parental distress tends to resolve as colic subsides. The most serious complication of colic is non-accidental trauma.

4.**(D)** Temper tantrums are brief episodes of extreme or unpleasant behavior that appear disproportionate to the situation, typically in response to frustration or anger. The median duration of a tantrum is 3 minutes. Behaviors vary and may include whining, crying, screaming, stomping, flailing, head banging, hitting, throwing objects, and biting.

5.**(B)** Many of the symptoms of ADHD mimic typical findings of normal development. Symptoms of inattention (e.g., failing to pay close attention to details, appearing to not listen when spoken to directly, executive functioning skills' concern), hyperactivity (e.g., being fidgety or restless, leaving a seat when expected to remain seated), or impulsivity (e.g., blurting out and interruption) may indicate a diagnosis of ADHD.
Behavioral therapy is central to children of all ages with ADHD and is the core of overall treatment. Behavioral management includes establishment of structure, routine, consistency in adult responses to behaviors, and appropriate behavioral goals.

6.**(C)** Alarm therapy is more time-intensive with high attrition rates and takes longer to work, but overall, it has a superior response rate and a lower relapse rate and is especially helpful in situations where the patient and family are coping well and adhering to the routine required for success. Desmopressin may work more quickly, making it ideal for occasional use such as sleep-overs; it may also be indicated when a more rapid effect is needed due to poor coping or when families have difficulty adhering to the routine of an alarm. Some studies have shown that the addition of an anticholinergic medication to desmopressin may be more effective than desmopressin alone. Tricyclic antidepressants (most commonly imipramine) are reserved for refractory cases due to potential for serious toxicity (particularly cardiac and psychiatric) and should only be prescribed by a specialist.

7.**(A)** The differential diagnosis for functional constipation and soiling includes organic causes of constipation (e.g., neurogenic, anatomic, endocrinologic, gastrointestinal, and pharmacologic). For instance, a child with chronic constipation with or without soiling who had delayed passage of meconium and has an empty rectum and tight sphincter may have Hirschsprung disease. Chronic constipation may be a presenting sign of spinal cord abnormalities, such as a spinal cord tumor or a tethered cord. Physical examination findings of altered lower extremity reflexes, absent anal wink, or a sacral hairy tuft or sacral sinus may be a clue to these anomalies. Hypothyroidism can present with chronic constipation and typically is accompanied by poor linear growth. Anal stenosis may lead to chronic constipation, as can the use of opiates, phenothiazine, antidepressants, and anticholinergics. Developmental problems, including cognitive impairment and developmental delays, may be associated with chronic constipation.

8.**(A)** Studies have shown polyethylene glycol powder to be superior to older medications such as lactulose, both in terms of efficacy and compliance. It is well tolerated because the taste and texture are palatable.

9.**(A)** Parasomnias include sleepwalking, sleep terrors, and confusional arousals. These occur during NREM sleep and are more likely during the first third of the night. They are most common in preschool children and are likely to resolve with time and developmental maturation. Sleep terrors consist of an abrupt awakening with a loud scream or agitation that is unresponsive to caregivers' attempts to console. Sleep terrors are differentiated from nightmares, which occur later in the night and result from arousal in REM or dreaming sleep. Children typically remember their nightmares but have no recollection of sleep terrors. Confusional arousals are similar to sleep terrors but tend to be less dramatic and last longer.

10.**(D)** Rarely, children with insomnia are treated pharmacologically. There are no U.S. Food and Drug Administration– approved medications for treating insomnia in children, and clinical evidence is lacking for most medications with off label use. Melatonin has soporific properties useful in treating delayed sleep phase syndrome and has been found to be effective in both children with normal development and those with developmental delays, as well as in children with ADHD and ASD.

SECTION 4

PSYCHIATRIC DISORDERS

HAYDER ALMUSAWI

QUESTIONS

1. According to the American Academy of Paediatrics (AAP), the recommended screening age for autism is

A. 6 and 12 months
B. 12 and 18 months
C. 18 and 24 months
D. 24 and 30 months

2. The Gold standard psychologic measure commonly recommended to confirm a diagnosis of autism spectrum disorder is

A. Autism Diagnostic Observation Schedule–2nd Edition (ADOS-2)
B. Childhood Autism Rating Scale, 2nd Edition [CARS2]
C. Modified Checklist for Autism in Toddlers–Revised [M-CHAT-R]
D. Gilliam Autism Rating Scale, 3rd Edition [GARS3]

3. The first-line medication for sleep dysregulation in patient with autism spectrum disorder is

A. risperidone
B. naltrexone
C. clonidine
D. melatonin

4. The medication used to decrease self-injurious behaviour in patient with autism spectrum disorder is

A. guanfacine
B. naltrexone
C. clonidine
D. melatonin

5. The **MOST** common obsessions in children involve fears of

A. dirt
B. sleep
C. water
D. cars

6. A 7-year-old boy presented with history of crying, screaming and hitting his mother with his hand until mother has done his wishes, he pushes his mother to dress and undress him nearly 7-8 times until he feels dressed properly, the boy would be late for school as a consequence of his behaviour. In the school the boy avoids going to toilet because he is afraid about the arrangement of his clothes.
What is the **BEST** initial pharmacological therapy?

A. Risperidone

B. Naltrexone
C. Clonidine
D. Fluoxetine

7. Which of the following infections may account for childhood-onset obsessive-compulsive disorder?
A. Staphylococcus aureus infection
B. Streptococcal infection
C. Chlamydia trachomatis infection
D. Klebsiella pneumoniae infection

8. Major depressive disorder (MDD) requires symptoms including either depressed mood or loss of interest or pleasure in nearly all activities for a minimum of
A. 2 weeks
B. 4 weeks
C. 8 weeks
D. 12 weeks

9. Disruptive mood dysregulation disorder (DMDD) includes children ages 6–18 years who presents with a chronic pattern of severe irritability and behavioural dysregulation for
A. ≥3 months
B. ≥6 months
C. ≥12 months
D. ≥18 months

10. What is the only medication approved by the U.S. Food and Drug Administration (FDA) for treatment of depression in youth 8 years and older?
A. Escitalopram
B. Citalopram
C. Paroxetine
D. Fluoxetine

11. An antidepressant should be given an adequate trial before switching or discontinuing unless there are serious side effects, an adequate trial means
A. 2 weeks at therapeutic doses
B. 4 weeks at therapeutic doses
C. 6 weeks at therapeutic doses
D. 8 weeks at therapeutic doses

12. For a first episode of depression in children and adolescents, treatment continue after remission of symptoms for
A. 1–3 months
B. 3–6 months
C. 6–9 months
D. 9–12 months

13. What is the hallmark of mania?

A. Decreased need for sleep without fatigue
B. Inflated self-esteem
C. More talkative than usual
D. Flight of ideas

14. The oldest proven treatment for mania which has been used in acute episodes and as a maintenance treatment in children and adolescents is

A. lithium
B. carbamazepine
C. olanzapine
D. risperidone

15. An adolescent girl presented with recurrent attacks of sweating, palpitations, feeling of chocking, chest pain, and fear of dying lasting about 10 minutes then improves gradually after reaching ED and receiving oxygen. The girl had persistent worries about having another attack. On examination; she looks pale, PR 130 BPM, BP 110/70, temperature 37.1^0C, and SPO2 98 on room air. Blood glucose, BUN and serum electrolyte are normal, ECG, echocardiography, and chest radiography are normal, blood film shows mild microcytic, hypochromic anaemia.

Of the following, the **MOST** appropriate next step in her management is

A. reassurance of the family
B. referral to paediatric cardiologist
C. referral to paediatric psychiatrist
D. referral to paediatric haematologist

16. Which of the following can be used in the management of anxiety disorders, if autonomic symptoms are present?

A. Fluoxetine
B. Fluvoxamine
C. Clonazepam
D. Clonidine

SECTION 4

PSYCHIATRIC DISORDERS

HAYDER ALMUSAWI

ANSWERS

1.**(C)** The American Academy of Pediatrics (AAP) recommends screening for autism at 18 and 24 months of age.

2.**(A)** There are numerous screening measures that may be employed (e.g., Childhood Autism Rating Scale, 2nd Edition [CARS2], Modified Checklist for Autism in Toddlers–Revised [M-CHAT-R], Gilliam Autism Rating Scale, 3rd Edition [GARS3], and Screening Tool for Autism in Toddlers and Young Children [STAT]) to assist with appropriate referral for diagnostic evaluation. Gold standard psychologic measures such as the Autism Diagnostic Observation Schedule–2nd Edition (ADOS-2) and Autism Diagnostic Interview–Revised (ADI-R) are commonly recommended to confirm a diagnosis.

3.**(D)** α2 agonists (guanfacine, clonidine) are used for hyperactivity, aggression, and sleep dysregulation, although melatonin is first-line medication for sleep dysregulation.

4.**(B)** Naltrexone has been used to decrease self-injurious behaviour, presumably by blocking endogenous opioids.

5.**(A)** The most common obsessions in children involve fears of contamination, fears of dirt/germs, repeated doubts, orderliness or precision, and aggressive thoughts.

6.**(D)** obsessive-compulsive disorder (OCD) is characterized by obsessions, compulsions, or both in the absence of another psychiatric disorder that better explains the symptoms. Obsessions involve recurrent intrusive thoughts, images, or impulses. Compulsions are repetitive, non-gratifying behaviours that a person feels driven to perform in order to reduce or prevent distress or anxiety. In children, rituals or compulsive symptoms often predominate over worries or obsessions, and the child may attempt to neutralize an obsessive thought by performing compulsions. Symptoms may or may not be recognized as being excessive or unreasonable. Selective serotonin reuptake inhibitors (SSRIs) are recommended for higher severity of symptoms.

7.**(B)** Streptococcal infection causing inflammation in the basal ganglia may account for 10% of childhood-onset OCD and is part of a condition referred to as pediatric autoimmune neuropsychiatric disorders associated with streptococcal (PANDAS) infection.

8.**(A)** Major depressive disorder (MDD) requires a minimum of 2 weeks of symptoms including either depressed mood or loss of interest or pleasure in nearly all activities.

9.**(C)** Disruptive mood dysregulation disorder (DMDD) includes children ages 6–18 years who present with a chronic (≥12 months) pattern of severe irritability and behavioral dysregulation.

10.**(D)** Fluoxetine is the only medication approved by the U.S. Food and Drug Administration (FDA) for treatment in youth 8 years and older, and escitalopram is approved for youth 12 and older. However, many other off-label medications such as citalopram, paroxetine, and venlafaxine have positive clinical trial results as well.

11.**(C)**

12.**(C)** An antidepressant should be given an adequate trial (6 weeks at therapeutic doses) before switching or discontinuing unless there are serious side effects. For a first episode of depression in children and adolescents, treatment for 6–9 months after remission of symptoms is recommended. Patients with recurrent or persistent depression may need to take antidepressants for extended periods (years or even a lifetime).

13.**(A)** A decreased need for sleep without fatigue is a hallmark of mania. There are no other diagnoses where a child has a greatly decreased amount of total sleep (compared with age-appropriate norms) and is not fatigued.

14.**(A)**

15.**(C)** A panic attack is a sudden unexpected onset of intense fear associated with a feeling of impending doom in the absence of real danger and may occur in the context of any anxiety disorder. The individual must experience four or more symptoms (e.g., sweating, palpitations, feeling of choking, chest pain, trembling/shaking, chills, paresthesias, fear of dying, fear of losing control, derealization, dizziness, nausea, or shortness of breath) during the attack. At least 1 month of persistent worrying about having another panic attack and/or a significant behavioural change following an attack (e.g., attempting to avoid another attack, avoidance of triggers such as exercise) is required to make the diagnosis.

16.**(D)** α_2 agonists (guanfacine and clonidine) may be useful if autonomic symptoms are present.

SECTION 5

PSYCHOSOCIAL ISSUES

ZUHAIR ALMUSAWI

QUESTIONS

1. Which of the following suggests short stature with preserved weight?
   A. Endocrine etiology
   B. Long-standing malnutrition
   C. Chromosomal abnormalities
   D. Congenital infection

2. With the rapid reinstitution of feeding in infants with marasmus, fluid and electrolyte homeostasis may be lost, which can result in life-threatening cardiac, pulmonary, or neurologic problems.
Of the following, the **MOST** likely cause is
   A. syndrome of inappropriate ADH
   B. over hydration
   C. re-feeding syndrome
   D. increased metabolic demand

3. An 18-month-old boy presents by his step mother with multiple bruises of different colors on abdomen, ear, and neck. The baby looks healthy with history of upper respiratory infection before two weeks, there is no family history of bleeding tendency and his CBC is normal.
Of the following, the **MOST** likely cause for these lesions is
   A. hemophilia
   B. vitamin K deficiency
   C. physical abuse
   D. external trauma

4. The parents of a 3-month-old girl brought their baby complaining from irritability, lethargy, vomiting, and seizures. The father told you that his primiparous young wife became frustrated and intolerant of infant crying which was started from age of 3-weeks with no response to all soothing maneuvers and colic medications.
Of the following, the **MOST** appropriate first investigation is
   A. lumbar puncture
   B. CT brain
   C. blood sugar
   D. serum calcium

5. A house wife mother brought her 2-year-old boy complaining from hematuria for the last six months, she consulted many doctors and admitted to hospital twice, fully investigated with no improvement or definite diagnosis. On examination the baby looks healthy, while you are examining the abdomen, the baby passed urine which was clear, while the result of lab was RBC +++.
Of the following, the **MOST** like diagnosis is

A. benign familial hematuria
B. IgA nephropathy
C. hypercalciuria
D. Munchausen syndrome by proxy

6. In most cases, the diagnosis of sexual abuse is made by
A. history obtained from the child
B. complete forensic interview
C. children's disclosures
D. genital discharge

7. Gender constancy, the understanding that one is always a male or always a female, is typically achieved by the age of
A. 2 years
B. 4 years
C. 6 years
D. 8 years

8. Which of the following substances, if used in the perinatal period has been associated with prematurity, intracranial hemorrhages, and abruptio placentae?
A. Cigarette smoking
B. Cocaine
C. Opiates
D. Alcohol

9. The child's reaction to the divorce is influenced by the child's age and developmental level. Consequences of stress can include irritability and disturbance in sleep and feeding patterns in
A. toddlers
B. preschool children
C. school-age children
D. adolescents

10. What is the **MOST** common possible reaction of children to the death of a parent?
A. Sadness and a yearning
B. Decrease in academic functioning
C. Lack of enjoyment with activities
D. Changes in appetite and sleep

SECTION 5

PSYCHOSOCIAL ISSUES

ZUHAIR ALMUSAWI

ANSWERS

1.**(A)** FTT that is symmetric (proportional weight, height/length, and head circumference) suggests long-standing malnutrition, chromosomal abnormalities, congenital infection, or teratogenic exposures. Short stature with preserved
weight suggests an endocrine etiology.

2.**(C)** With the rapid reinstitution of feeding after starvation, fluid and electrolyte homeostasis may be lost. Changes in serum electrolyte concentrations and the associated complications are collectively termed the re-feeding syndrome. These changes typically affect phosphorus, potassium, calcium, and magnesium and can result in life-threatening cardiac, pulmonary, or neurologic problems. Infants and children with marasmus, kwashiorkor, and anorexia nervosa and those who experience prolonged fasting are at risk for re-feeding syndrome. Re-feeding syndrome can be avoided by slow institution of nutrition, close monitoring of serum electrolytes during the initial days of feeding, and prompt replacement of depleted electrolytes.

3.**(C)** Bruises in healthy children generally are distributed over bony prominences; bruises that occur in an unusual distribution, such as isolated to the torso, ears, or neck, should raise concern of physical abuse.

4.**(B)** Abusive head trauma is the leading cause of mortality and morbidity from physical abuse. Most victims are young; infants predominate. Shaking and/or blunt impact trauma cause injuries. Caregiver intolerance of infant crying is a common precipitator. Symptoms at the time of presentation to medical care can range from occult head trauma with no neurologic symptoms to irritability, lethargy, vomiting, seizures, apnea, and coma.

5.**(D)** Munchausen syndrome by proxy, factitious disorder by proxy, and caregiver-fabricated illness in a child, this form of child maltreatment involves a child receiving unnecessary and harmful or potentially harmful medical care at the instigation of a caregiver.

6.**(A)** In most cases, the diagnosis of sexual abuse is made by the history obtained from the child. A complete forensic interview at the physician's office is not needed. Children may make spontaneous disclosures to the physician, or physicians may evaluate patients prior to investigative interviews and require information for medical decision making.

7.**(C)** Typically, a child can identify oneself as a boy or a girl by 3 years of age and begins to demonstrate initial expressions of gender identity. Gender atypical behavior is common in young children and may be part of typical development. However, gender constancy, the understanding that one is always a male or always a female, is typically achieved by age 6 years.

8.**(B)** Cigarette smoking during pregnancy is associated with lower birthweight and increased child behavioral problems. Use of cocaine in the perinatal period has been associated with prematurity, intracranial hemorrhages, and abruptio placentae. Exposure to opiates in utero can result in prematurity and neonatal withdrawal syndrome.

9.**(A)** Consequences of stress in infants and toddlers can include irritability and disturbance in sleep and feeding patterns. Preschool children may have magical beliefs about cause and effects and an egocentric view of the world. They may believe that something they did caused the divorce, leading them to be particularly upset. School-age children have a concrete understanding of cause and effect; if something bad happened, they understand that something caused it to happen. Children at this age may still worry that something they did caused the divorce. Adolescents are developing a sense of autonomy, a sense of morality, and the capacity for intimacy, and divorce may lead them to question previously held beliefs. They may be concerned about what the divorce means for their future and whether they, too, will experience marital failure.

10.**(A)** There are many possible reactions of children to the death of a parent or close relative. Sadness and a yearning to be with the dead relative are common. Sometimes a child might express a wish to die and so visit the dead relative, but a plan or desire to commit suicide is uncommon and would need immediate evaluation. A decrease in academic functioning, lack of enjoyment with activities, and changes in appetite and sleep can occur.

SECTION 6

NUTRITION

MOHAIMEN ALKABI

QUESTIONS

1. A 25-day-old boy brought to your clinic by his mother with excessive crying, her mother stated that her milk is insufficient.
Which of the following is the **MOST** objective indicator of adequate milk intake?

A. Voiding pattern
B. Stool pattern
C. Weight gain rate
D. Frequency of feeding

2. Breastfed infants have reduced risk of

A. vitamin D deficiency
B. neonatal jaundice
C. severe lower respiratory disease
D. acute otitis externa

3. Which of the following maternal infections is considered a permanent contraindication for breast feeding in developed countries?

A. HIV
B. Active tuberculosis
C. Syphilis
D. Varicella

4. Which of the following conditions is more often in breast-fed infants than in formula-fed infants?

A. Jaundice
B. Intestinal bleeding
C. Diarrhea
D. Otitis media

5. Which of the following breast milk compositions is absent after freezing and pasteurizing?

A. IgA and SIgA
B. Lactoferrin
C. Lysozyme
D. Lipase

6. Which of the following components is absent in formula milk?

A. Vitamin D
B. Monoglycerides
C. Linoleic acid
D. Vitamin A

7. The mother of healthy full term exclusively breastfed newborn asks you about the time of starting iron supplement for her baby.
Of the following, the **BEST** response is at

A. birth
B. 2 months of age
C. 4 months of age
D. 6 months of age

8. At what age the infant can take fruit juice?

A. 4 months
B. 6 months
C. 9 months
D. 12 months

9. Children with eczema may be at a higher risk of developing peanut allergy. These infants should be introduced to peanut-containing products around

A. 4-6 month
B. 9-12 month
C. 12-15 month
D. 18 -24month

10. Which of the following associations is **CORRECT**?

A. Cushing syndrome --- Ovarian dysgenesis
B. Bardet-Biedl syndrome --- hypogonadism
C. Cohen syndrome --- syndactyly
D. Carpenter syndrome --- retinitis pigmentosa

11. A 2-year-old girl presented with intellectual disability, retinitis pigmentosa, diabetes mellitus, hearing loss, and hypogonadism.
Of the following, the **MOST** likely diagnosis is

A. Alstrom syndrome
B. Bardet-Biedl syndrome
C. Biemond syndrome
D. Fröhlich syndrome

12. Obesity is diagnosed in children younger than 2 years by

A. body mass index
B. weight for age
C. weight for length
D. midarm circumference

13. Obese children, ages 6–11 years, might be encouraged to modify their eating habits for gradual weight loss/month of no more than

A. 0.5 kg
B. 1 kg
C. 1.5 kg
D. 2 kg

14. Orlistat may be a safe and effective adjunct to dietary and behavioral modifications in the treatment of obesity for patients older than

A. 6 years
B. 8 years
C. 10 years
D. 12 years

15. Which of the following correlations between drug and vitamin deficiency is **TRUE**?

A. Sulfonamides --- Vitamin E
B. Phenobarbital --- Vitamin D
C. Antacids --- Vitamin A
D. Digitalis --- Vitamin K

16. You are a volunteer with a group of medical students in a refugee camp. You asked to examine an apathetic child who refuses to eat. Physical examination reveals thin hair and skin. The child also has edema of the face, hands, and legs.
Of the following, the **MOST** likely diagnosis is

A. vitamin B1 deficiency
B. niacin deficiency
C. marasmus
D. kwashiorkor

17. Which of the following is a major clinical manifestation of kwashiorkor?

A. Body weight is near normal for age
B. Edema
C. Pellagroid
D. Enlarged parotid glands

18. Which of the following skin manifestations appear in the most severe form of kwashiorkor?

A. Flag sign
B. Pellagroid
C. Flaky paint
D. Angular cheilosis

19. Which of the following clinical manifestations signify a severe and life-threatening malnutrition?

A. Hyperthermia
B. Edema
C. Bradycardia
D. Muscle wasting

20. A child presented with weakness, lassitude, dermatitis, photosensitivity, mucositis, diarrhea, vomiting, and dysphagia.
Which of following is the **MOST** likely diagnosis?

A. Ariboflavinosis
B. Pellagra

C. Wet beriberi
D. Scurvy

21. A 3-year-old girl presented with vomiting, diarrhea, failure to thrive, listlessness, and hyperirritability. Laboratory data show HCT 28%, hypochromic microcytic anemia. She had pulmonary tuberculosis and on treatment for 9 months.
Which of the following is the **BEST** treatment for anemia?

A. Pyridoxine
B. Folic acid
C. Cyanocobalamin
D. Ascorbic acid

22. Which of the following is the earliest manifestation of vitamin A deficiency?

A. Night Blindness
B. Xerosis of conjunctiva
C. Keratomalacia
D. Corneal ulceration

23. A 7-year-old boy presented with vomiting, generalized body weakness, headache, and blurred vision. On examination; he had convergent squint of the right eye and bilateral papilledema. The pulse rate and blood pressure were 60 beats/min and 70/50 mmHg respectively.
Which of the following is the **MOST** likely diagnosis?

A. Hypervitaminosis A
B. Hypervitaminosis D
C. Hypervitaminosis B6
D. Hypervitaminosis C

24. A 9-year-old girl with cystic fibrosis presented with difficulty walking, unsteady gait, and visual problem. Physical examination revealed loss of deep tendon reflexes and ophthalmoplegia.
Of the following, the **BEST** treatment is

A. tocopherol
B. vitamin E
C. ergocalciferol
D. cyanocobalamin

25. Which of the following is the **BEST** measure of vitamin D status?

A. 25(OH)D
B. 1,25(OH2) D
C. 24,25(OH2) D
D. Calcium, phosphorus, and alkaline phosphate

26. Breast-fed infants born of mothers with adequate vitamin D stores usually maintain adequate serum vitamin D levels for at least

A. 1 month
B. 2 months

C. 3 months
D. 4 months

27. A 28-week- preterm infant is now 6 weeks old and weighs 1,750 g. Physical examination reveals pallor and jaundice. Laboratory data shows Hb 8.1 g/dl, reticulocyte 8%, and platelet count $650*10^9$/L.
Which of the following is the **MOST** likely diagnosis?

A. Vitamin E deficiency
B. Anemia of prematurity
C. Vitamin C deficiency
D. Vitamin K deficiency

28. A 6-month-old boy presented with pallor for the last 3 months. The baby on breast feeding for the first 2 months and then shifted to goat milk.
Which of the following mineral or vitamins deficiency is the underlying cause of pallor?

A. Vitamin B_{12}
B. Folic acid
C. Iron
D. Zinc

29. Which of the following is the **MOST** likely cause of anemia in a 9-month-old breast fed boy to vegan mother who was supplied with iron since the age of 4 months?

A. Folate deficiency
B. Iron deficiency
C. Vitamin B_{12} deficiency
D. Vitamin E deficiency

30. Which of the following enhance non-heme iron absorption from gastrointestinal tract?

A. Bran
B. Polyphenols
C. Phytic acid
D. Fish

31. Oral treatment for a child with iron-deficiency anemia should be continued for

A. 2 months
B. 3 months
C. 4 months
D. 5 months

32. Which of the following trace elements deficiency causes hypercholesterolemia, weight loss, and decreased clotting proteins?

A. Zinc
B. Copper
C. Cobalt
D. Manganese

33. A 4-year-old boy presented with nausea, diarrhea, neurologic manifestations, nail and hair changes, and garlic odor.
Of the following, the **MOST** likely cause is

A. zinc deficiency
B. molybdenum deficiency
C. selenium toxicity
D. copper toxicity

34. Which of the following body organs is **MOST** likely affected by copper toxicity?

A. Liver
B. Kidney
C. Pancreas
D. Lung

35. A 10-month-old infant presented with an erythematous scaly skin lesion on the distal extremities, hands, feet, perioral and perineal areas, diarrhea, and recurrent pneumonia.
Which of the following is the **MOST** likely diagnosis?

A. Zinc deficiency
B. Scurvy
C. Thiamin deficiency
D. Vitamin A deficiency

SECTION 6

NUTRITION

MOHAIMEN ALKABI

ANSWERS

1.**(C)** Adequacy of milk intake can be assessed by voiding and stooling patterns of the infant. A well-hydrated infant void 6–8 times a day. Each void should soak a diaper, and urine should be colorless. By 5–7 days, loose yellow stools should be passed approximately four times a day. Rate of weight gain provides the most objective indicator of adequate milk intake. Total weight loss after birth should not exceed 7%, and birth weight should be regained by 10 days. The mean feeding frequency during the early weeks postpartum is 8–12 times per day.

2.**(C)** Benefits for infants who are breastfed have reduced risks of:

- Asthma.
- Obesity.
- Type 1 diabetes.
- Severe lower respiratory disease.
- Acute otitis media (ear infections).
- Sudden infant death syndrome (SIDS).
- Gastrointestinal infections (diarrhea/vomiting).
- Necrotizing enterocolitis (NEC) for preterm infants.

3.**(A)** Maternal infection with HIV is considered a contraindication for breast-feeding in developed countries. When the mother has active tuberculosis, syphilis, or varicella, restarting breast-feeding may be considered after therapy is initiated. If a woman has herpetic lesions on her breast, nursing and contact with the infant on that breast should be avoided. Women with genital herpes can breast-feed. Proper handwashing procedures should be stressed.

4.**(A)** In the newborn period, elevated concentrations of serum bilirubin are present more often in breast-fed infants than in formula-fed infants.

5.**(D)** The composition of human milk that change after pasteurization and freezing.

- Protein is reduced
- IgA and SIgA are reduced by 30%
- IgG is reduced by 30%
- Lactoferrin is reduced by 30%
- Lysozyme is reduced by 25%
- Lipases are absent

6.**(D)**

TABLE **27.2** Composition of Breast Milk, Breast Milk After Freezing and Pasteurization, and Representative Infant Formulas

COMPONENT	BREAST MILK	BREAST MILK AFTER FREEZING AND PASTEURIZING	STANDARD FORMULA	SOY FORMULA	HYPOALLERGENIC FORMULA
Protein	1.1/dL	Reduced	1.5/dL	1.7/dL	1.9/dL
Fat	4.0/dL	4.0/dL	3.6/dL	3.6/dL	3.8–3.3/dL
Carbohydrate	7.2/dL	7.2/dL	6.9–7.2/dL	6.8/dL	6.9–7.3/dL
Calcium	290 mg/L	290 mg/L	420–550 mg/L	700 mg/L	635–777 mg/L
Phosphorus	140 mg/L	140 mg/L	280–390 mg/L	500 mg/L	420–500 mg/L
Sodium	8.0 mg/L	8.0 mg/L	6.5–8.3 mg/L	13 mg/L	14 mg/L
Vitamin D	Variable	Variable	400/dL	400/dL	400/dL
Vitamin A	100%	100%	—	—	—
Osmolality	253 mOsm/L	253 mOsm/L	230 mOsm/L	200–220 mOsm/L	290 mOSm/L
Renal solute load	75 mOsm/L	75 mOsm/L	100–126 mOsm/L	126–150 mOsm/L	125–175 mOSm/L
IgA and SIgA	Present	Reduced 30%	0	0	0
IgM	Present	Present	0	0	0
IgG	Present	Reduced 30%	0	0	0
Lactoferrin	Present	Reduced 30%	0	0	0
Lysozyme	Present	Reduced 25%	0	0	0
Lipases	Present	0	0	0	0
Monoglycerides	Present	Present	Added to some formulas	Added to some formulas	Added to some formulas
Free fatty acids	Present	Present	Added to some formulas	Added to some formulas	Added to some formulas
Linoleic acid	Present	Present	Added to some formulas	Added to some formulas	Added to some formulas
α-linoleic acid	Present	Present	Added to some formulas	Added to some formulas	Added to some formulas
Bifidus factor	Present	Present	—	—	—
Oligosaccharides	Present	Present	—	—	—

7.**(C)**

8.**(D)** Infants do not need juices, but if juice is given, it should be started only after 12 months of age, given in a cup (as opposed to a bottle), and limited to 4 oz of 100% natural unsweetened juice (for toddlers age 1–3 yr) daily; juices should also be offered only with meals or snacks. Excess juices may reduce appetite for other more nutritious foods and cause diaper rash, diarrhea, and weight gain.

9.**(A)** Solid cereals are usually introduced first. Children with eczema may be at a higher risk of developing peanut allergy. These infants should be introduced to peanut-containing products around 4–6 months of age in order to prevent peanut allergies in the future. Cow's milk is not recommended before 12 months of age; parents may provide processed dairy products such as cheese and yogurt before 1 year of age. All foods with the potential to obstruct the young infant's main airway should be avoided in general until 4 years of age or older.

10.**(B)**

11.**(A)**

12.**(C)** BMI is a convenient screening tool that correlates fairly strongly with body fatness in children and adults. BMI age-specific and gender-specific percentile curves (for 2- to 20-

year-olds) allow an assessment of BMI percentile. For children younger than 2 years of age, weight-for-length measurements greater than the 95th percentile may indicate overweight and warrant further assessment.

13.**(A)** Obese children, ages 6–11 years, might be encouraged to modify their eating habits for gradual weight loss of no more than 1 lb (or ~0.5 kg) a month. Older children and adolescents who are obese or severely obese might be encouraged to modify their eating habits to aim for weight loss of up to 2 lb (or ~1 kg) a week.

14.**(B)**

15.**(B)**

16.**(D)** Physical examination of kwashiorkor reveals a relative maintenance of subcutaneous adipose tissue and a marked atrophy of muscle mass. Edema varies from a minor pitting of the dorsum of the foot to generalized edema with involvement of the eyelids and scrotum. The hair is sparse, easily plucked, and appears dull brown, red, or yellow-white.

17.**(A)**

18.**(C)** Skin changes are common and range from hyper pigmented hyperkeratosis to an erythematous macular rash (pellagroid) on the trunk and extremities. In the most severe form of kwashiorkor, a superficial desquamation occurs over pressure surfaces ("flaky paint" rash).

19.**(C)** Bradycardia and hypothermia signify severe and life-threatening malnutrition.

20.**(B)** Pellagra, or niacin-deficiency disease, is characterized by weakness, lassitude, dermatitis, photosensitivity, inflammation of mucous membranes, diarrhea, vomiting, dysphagia, and, in severe cases, dementia.

21.**(A)** Dietary deprivation or malabsorption of vitamin B_6 in children results in hypochromic microcytic anemia, vomiting, diarrhea, failure to thrive, listlessness, hyperirritability, and seizures. Children receiving isoniazid or penicillamine may require additional vitamin B_6 because the drug binds to the vitamin.

22.**(A)** The earliest symptom is night blindness, which is followed by xerosis of the conjunctiva and cornea. Untreated, xerophthalmia can result in ulceration, necrosis, keratomalacia, and a permanent corneal scar.

23.**(A)** Hypervitaminosis A also has serious sequelae, including headaches, pseudotumor cerebri, hepatotoxicity, and teratogenicity.

24.**(B)** Vitamin E deficiency occurs in children with fat malabsorption secondary to liver disease, untreated celiac disease, cystic fibrosis, and abetalipoproteinemia. In these children, without vitamin E supplementation, a syndrome of progressive sensory and motor neuropathy develops, ataxia and ophthalmoplegia.

25.**(A)**

26.**(B)**

27.**(A)** Deficient preterm infants (of vitamin E) at 1–2 months of age have hemolytic anemia characterized by an elevated reticulocyte count, an increased sensitivity of the erythrocytes to hemolysis in hydrogen peroxide, peripheral edema, and thrombocytosis.

28.**(B)** Goat's milk are low in folate.

29.**(C)** Exclusively breast-fed infants ingest adequate vitamin B_{12} unless the mother is a strict vegetarian without supplementation.

30.**(D)** The absorption of non-heme iron is influenced by the composition of consumed foods. Enhancers of non-heme iron absorption are ascorbic acid, meat, fish, and poultry. Inhibitors are bran, polyphenols (including the tannates in tea), and phytic acid, a compound found in legumes and whole grains.

31.**(D)** Treatment of iron-deficiency anemia includes changes in the diet to provide adequate iron and the administration of 3–6 mg iron/kg per 24 hours (as ferrous sulfate) divided tid. Reticulocytosis is noted within 3–7 days of starting treatment. Oral treatment should be continued for 5 months.

32.**(D)**

33.**(C)**

34.**(A)** patient with excess copper:
Acute: nausea, emesis, abdominal pain, coma, and hepatic necrosis.
Chronic toxicity (liver and brain injury) occurs in Wilson disease and secondary to excess intake.

35.**(A)** Clinical manifestations of mild zinc deficiency include anorexia, growth faltering, and immune impairment. Moderately severe manifestations include delayed sexual maturation, rough skin, and hepatosplenomegaly. The signs of severe deficiency include acral and periorificial erythematous scaling dermatitis, growth and immune impairment, diarrhea, mood changes, alopecia, night blindness, and photophobia.

SECTION 7

FLUIDS AND ELECTROLYTES

KHALID ALAARAJI

QUESTIONS

1. As a senior house officer, you have been asked to arrange the amount of maintenance intravenous fluid over 24 hours for a 15-year-old girl, weighing 65 kg postoperatively. You prescribed IV glucose saline for her.
The volume of fluid per day for this girl should not exceed

A. 2000 ml
B. 2400 ml
C. 2800 ml
D. 3200 ml

2. A 5-year-old boy admitted to emergency department complaining from repeated vomiting for 48 hours, his UOP is adequate.
Of the following, the **BEST** type of fluid for this child is

A. 5% dextrose in ¼ normal saline + 20mEq/L potassium chloride
B. 5% dextrose in ¼ normal saline + 10mEq/L potassium chloride
C. Ringer lactate + 10mEq/L potassium chloride
D. normal saline + 10mEq/L potassium chloride

3. A 6-year-old boy admitted to emergency department for management of severe respiratory distress secondary to pneumonia with huge pleural effusion. Chest tube inserted.
Which is the **BEST** type of replacement fluid for this child?

A. 5% dextrose in ¼ normal saline
B. Normal saline
C. 5% dextrose
D. 2.5% dextrose in ½ normal saline

4. A 3-year-old child, a known case of ITP diagnosed before 10 days on prednisolone with good response, admitted to emergency department with vomiting and diarrhea. Clinical assessment reveals evidence of moderate dehydration. Laboratory findings: Hb; 14 g/dL, WBC count; 18,000/µL, Platelet count 160/µL, blood urea; 90 mg/dl, and serum creatinine; 0.6 mg/dL.
What is the **MOST** likely explanation of this disproportionately high blood urea level?

A. Development of HUS
B. Steroid therapy
C. Toxin of causative organism
D. Renal hypoperfusion

5. A 9-month-old infant is brought to the emergency department after 5 days of vomiting and diarrhea. Heart rate; 110 beats/min, respiratory rate; 28 breaths/min. and temperature;

38^0C. The boy looks lethargic with dry mucous membranes, and slightly depressed fontanelle with decreased urine output.
What is the percentage of dehydration in this infant?

A. 3%
B. 6%
C. 10%
D. 15%

6. A 2-year-old boy is brought to the emergency department with diarrhea and vomiting for 3 days and decreased urine output for last 24 hours. On examination, temperature; 37.5^0 C, heart rate; 150 beats/min, respiratory rate; 32 breaths/min, and blood pressure; 80/40 mm Hg. The boy looks lethargic with dry mucous membranes and prolonged capillary refill time (>3seconds).
Of the following, the **MOST** appropriate bolus IV fluid therapy is

A. 5% Dextrose + ½ normal saline, 20 mL/kg
B. 5% dextrose, 20 ml/kg
C. Ringer lactate, 10 mL/kg
D. 0.9% normal saline, 20 ml/kg

7. A 9-month-old infant is brought to the ER with diarrhea and vomiting for the last 4 days. He is formula fed baby. The boy is lethargic and irritable, dry mucous membranes, prolonged capillary refill time (>3seconds), and sunken fontanelle with normal urine output. Heart rate (140 beats/min), respiratory rate (28 breaths/min), and blood pressure is 78/44 mmHg. Laboratory results; Blood urea (30 mg/dL), serum creatinine (0.8 mg/dL), serum sodium (168 mEq/L), serum potassium (4 mEq/L), and serum glucose 95 mg/dL.
Of the following, the **NEXT** step in management of this infant is to

A. start with ORS
B. restore intravascular volume
C. correct the serum sodium within 24hr
D. start with 1.25-1.5 times of maintenance fluid

8. A 6-month-old infant admitted to the emergency department with seizure. His mother reported that, he had vomiting and diarrhea over the past 4 days. He received an excess amount of water and diluted formula. On examination, temperature (37.5^0C), heart rate (150 beats/min), respiratory rate (32 breaths/min) and blood pressure (80/40 mm Hg). The boy looks lethargic with dry mucous membranes, prolonged capillary refill time, depressed fontanelle, and decreased urine output. Laboratory result; blood urea 68 mg/dL, serum creatinine 0.8 mg/dL, serum sodium 118mEq/L, and serum potassium 3.4 mEq/L.
Of the following, the **MOST** appropriate first line management is to

A. restore intravascular volume with 0.9 normal saline
B. correct the serum sodium within 24hr
C. use IV 3% hypertonic saline
D. give IV diazepam

9. A 9-year-old boy was admitted to intensive care unit before 7 days due to road traffic accident with disturbed level of consciousness who got full improvement after 24hr. Over past 48 hours, the boy complains from nausea and headache. On examination; heart rate

(78 beats/min), respiratory rate (22 breaths/min), blood pressure (100/70 mm Hg), temperature (37^0C), and there is no neck stiffness or any abnormal signs. Lab results reveal Hb; 11 g/dL, WBC count; 14,000/µL, platelet count 160/µL, blood urea; 20 mg/dL, serum creatinine; 0.9 mg/dL, serum sodium; 121 mEq/L, serum potassium; 4.1 mEq/L, blood glucose; 98 mg/dL, serum osmolality; 260 mOsm/kg, urine specific gravity; 1.020, and urine sodium; 50 mEq/L.
Of the following, the **MOST** appropriate treatment is

A. water restriction
B. isotonic saline o.9%
C. salt and water restriction
D. hypertonic saline 3%

10. Pseudohyponatremia is a laboratory artifact that is present when the plasma contains high concentrations of

A. lipid
B. calcium
C. urea
D. chloride

11. Which of the following elevated blood indices causes spurious hypokalemia?

A. Hemoglobin
B. Platelet count
C. White blood cell count
D. Red blood cell count

12. The combination of hypokalemia and metabolic acidosis is characteristic of

A. gastric losses
B. diuretics use
C. diarrhea
D. Bartter syndrome

13. At what age, the ratio of the ICF volume to the ECF volume approaches adult level?

A. 1 year
B. 4 years
C. 6 years
D. 10 years

14. A 7-month-old infant evaluated for failure to thrive; investigation reveals serum Na+ (130 mEq/L), serum K+ (2.8 mEq/L), serum Cl^- (88 mEq/L), BUN (28 mg/dl), and serum creatinine (0.8 mg/dl). Blood gas analysis showed pH (7.52), $HCO3^-$ (36 mEq/L), and PCO2 (44 mm Hg).
Of the following, the **MOST** helpful test for the diagnosis of this condition is to measure

A. anion gap
B. 17-hydroxy progesterone
C. urine chloride
D. serum calcium

15. The dominant cation in intracellular fluid (ICF) is
    A. sodium
    B. chloride
    C. potassium
    D. phosphate

16. How much is the maintenance fluid requirement for a 3-year-old child, whose body weight is 14 kg?
    A. 800mL
    B. 1,000mL
    C. 1,200mL
    D. 1,400mL

17. Which of the following urinary findings may be seen in children with significant dehydration?
    A. RBC casts
    B. Hyaline casts
    C. WBC casts
    D. Proteinuria (150–200mg/dL)

18. An 11-month-old infant is brought to the emergency department after 2 days of vomiting and diarrhea. Body weight; 9 kg, Heart rate; 135 beats/min, respiratory rate; 33 breaths/min. and temperature; 38^0C. The boy looks lethargic with dry mucous membranes, slightly prolonged capillary refill time, and slightly depressed fontanelle with decreased urine output.
How much is the deficit fluid volume for this child?
    A. 270 ml
    B. 450 ml
    C. 900 ml
    D. 1350 ml

19. Which of the following characterize proximal RTA from distal RTA?
    A. Acidosis
    B. Failure to thrive
    C. Ability to acidify the urine
    D. Hypokalemia

20. The amount of oral rehydration solution (ORS) that should be given within 4 hours to patients with moderate dehydration is
    A. 25 mL/kg
    B. 50 mL/kg
    C. 75 mL/kg
    D. 100 mL/kg

21. To minimize the risk of cerebral edema during correction of hypernatremic dehydration, the serum sodium concentration should not be decreased more than
    A. 10 mEq/L/24hours

B. 12 mEq/L/24hours
C. 14 mEq/L/24hours
D. 16 mEq/L/24hours

22. Which of the following is a complication of rapid correction of hyponatremia (>10 mEq/L/24 hr)?
A. Cerebral edema
B. Central pontine myelinolysis
C. Brain herniation
D. Brain hemorrhage

23. A 3-month-old girl presented with mild upper respiratory tract infection develped sudden onset of rapid deep breathing and mild dehydration; serum Na^+ (143 mEq/L), serum K^+ (5.5 mEq/L), serum Cl^- (92 mEq/L), pH (7.11), $HCO3^-$ (10 mEq/L), lactate 4 mmol/L, and PCO2 (23 mm Hg).
Of the following, the **MOST** appropriate next step is to
A. calculate anion gap
B. measure urine chloride
C. send for CXR
D. send for renal function test

24. What is the **MOST** concerning complication of long-term parenteral nutrition?
A. Phlebitis
B. Cholestatic liver disease
C. Bowel ischemia
D. Sepsis

25. What is the initial concentration of dextrose in central parenteral nutrition (PN)?
A. 10%
B. 20%
C. 30%
D. 40%

26. Which of the following medications may cause Fanconi syndrome?
A. Valproate
B. Carbamazepine
C. Levetiracetam
D. Phenobarbital

27. Maintenance solutions are usually D5 in normal saline plus 20 mEq/L KCl. The role of glucose in maintenance fluid is to provide approximately
A. 10% of normal caloric needs
B. 20% of normal caloric needs
C. 30% of normal caloric needs
D. 40% of normal caloric needs

28. A 6-month-old infant is evaluated for failure to thrive and frequent hospital admissions due to repeated vomiting, dehydration, and increased urine output; pH (7.52), $HCO3^-$ (40 mEq/L), PCO2 (40 mm Hg), serum K^+ (2mmol/L), serum sodium (135 mmol/L), serum chloride (90 mmol/L), with normal serum magnesium and increase urinary chloride.
Of the following, the **MOST** likely diagnosis is

A. Gitelman syndrome
B. Liddle syndrome
C. 17-Hydroxylase deficiency
D. Bartter syndrome

29. Which of the following is a cause of normal anion gap metabolic acidosis?

A. Diarrhea
B. Salicylate intoxication
C. Ketoacidosis
D. Chronic renal failure

.

30. Which of the following conditions may decrease the anion gap?

A. Hypocalcemia
B. Hypokalemia
C. Hypoalbuminemia
D. Hypomagnesemia

31. The appropriate respiratory compensation for acute metabolic acidosis or alkalosis is completed within

A. 1 day
B. 2 days
C. 3 days
D. 4 days

32. During metabolic alkalosis, as expected compensatory mechanism, the PCO_2 increases for each 10 mEq/L increase in the serum [HCO3 −] by

A. 4 mm Hg
B. 7 mm Hg
C. 10 mm Hg
D. 14 mm Hg

33. Which of the following is the **MOST** common indication for long-term parenteral nutrition?

A. Prematurity
B. Trauma
C. Burns
D. Short bowel syndrome

SECTION 7

FLUIDS AND ELECTROLYTES

KHALID ALAARAJI

ANSWERS

1.**(B)** The maximum total fluid per day is normally 2,400mL.

2.**(D)**

- ✓ Replacement of ongoing gastric losses (normal saline + 10meq/L potassium chloride)
- ✓ Replacement of ongoing stool losses (5% dextrose in ¼ normal saline + 20mEq/L sodium bicarbonate + 20mEq/L potassium chloride)

3.**(B)** Output from surgical drains and chest tubes, when significant, should be measured and replaced. Third space losses manifest with edema and ascites and are due to a shift of fluid from the intravascular space into the interstitial space. Third space losses cannot be quantitated. Nonetheless, these losses can be large and lead to intravascular volume depletion, despite weight gain from edema or ascites. Replacement of third space fluid is empirical but should be anticipated in patients who are at risk, such as children who have sepsis, shock, burns, or abdominal surgery. Third space losses and chest tube output are isotonic and usually require replacement with an isotonic fluid, such as normal saline (NS) or Ringer lactate.

4.**(B)** Serum blood urea nitrogen (BUN) and creatinine concentrations are useful in assessing a child with dehydration. Volume depletion without renal insufficiency may cause a disproportionate increase in the BUN, with little or no change in the creatinine concentration. This is secondary to increased passive reabsorption of urea in the proximal tubule caused by appropriate renal conservation of sodium and water. This increase in the BUN may be absent or blunted in a child with poor protein intake because urea production depends on protein degradation. Conversely, the BUN may be disproportionately increased in a child with increased urea production, as occurs in a child with a gastrointestinal bleed or a child who is receiving glucocorticoids. A significant elevation of the creatinine concentration suggests renal injury.

5.**(C)**

Assessment of Degree of Dehydration			
	MILD	MODERATE	SEVERE
Infant	5%	10%	15%
Adolescent	3%	6%	9%

6.**(D)** The child with dehydration requires acute intervention to ensure that there is adequate tissue perfusion. This resuscitation phase requires rapid restoration of the circulating intravascular volume, which should be done with an isotonic solution, such as NS or Ringer lactate. Blood is an appropriate fluid choice for a child with acute blood loss but is not always available at the start of fluid resuscitation. The child is given a fluid bolus, usually 20mL/kg of the isotonic solution, over about 20minutes. A child with severe dehydration

may require multiple fluid boluses and may need to receive fluid at a faster rate. The initial resuscitation and rehydration are complete when signs of intravascular volume depletion resolve. The child typically becomes more alert and has a lower heart rate, normal blood pressure, and improved perfusion.

7.**(B)**

- Restore intravascular volume: Normal saline: 20 mL/kg over 20 min (repeat until intravascular volume restored).
- Determine time for correction based on initial sodium concentration
  [Na] 145-157 mEq/L: 24 hrs
  [Na] 158-170 mEq/L: 48 hrs
  [Na] 171-183 mEq/L: 72 hrs
  [Na] 184-196 mEq/L: 84 hrs
- Administer fluid at a constant rate over the time for correction; Typical fluid: D5 1/2 normal saline (with 20 mEq/L KCl unless contraindicated) & Typical rate: 1.25-1.5 times maintenance.

8.**(C)** Emergency treatment of symptomatic hyponatremia, such as seizures, uses IV hypertonic saline to rapidly increase the serum sodium concentration, which leads to a decrease in brain edema. One milliliter per kilogram of 3% sodium chloride increases the serum sodium by approximately 1mEq/L. A child often improves after receiving 4–6mL/kg of 3% sodium chloride.

9.**(A)** Patients with hyponatremia and no evidence of volume overload or volume depletion have euvolemic hyponatremia. These patients typically have an excess of total body water and a slight decrease in total body sodium. Some of these patients have an increase in weight, implying that they are volume overloaded. Nevertheless, they usually appear normal or have only subtle signs of fluid overload. In the syndrome of inappropriate ADH (SIADH), there is secretion of ADH that is not inhibited by either low serum osmolality or expanded intravascular volume. Retention of water causes hyponatremia, and the expansion of the intravascular volume results in an increase in renal sodium excretion. SIADH is associated with physiologic stress, pneumonia, mechanical ventilation, meningitis, and other central nervous system disorders (trauma). Ectopic (tumor) production of ADH is rare in children. SIADH should be distinguished from cerebral salt wasting, which is a form of hypovolemic hyponatremia often associated with tuberculosis meningitis or intracranial hemorrhage. Infants can develop euvolemic hyponatremia as a result of excessive water consumption or inappropriately diluted formula.

10.**(A)** Pseudohyponatremia is a laboratory artifact that is present when the plasma contains high concentrations of protein or lipid. It does not occur when a direct ion-selective electrode determines the sodium concentration, a technique that is increasingly used in clinical laboratories. In true hyponatremia, the measured osmolality is low, whereas it is normal in pseudohyponatremia.

11.**(C)** Hypokalemia is common in children, with most cases related to gastroenteritis. Spurious hypokalemia occurs in patients with leukemia and elevated white blood cell

counts, if plasma for analysis is left at room temperature, permitting the white blood cells to take up potassium from the plasma.

12.**(C)** The combination of hypokalemia and metabolic acidosis is characteristic of diarrhea, and proximal or distal renal tubular acidosis. A concurrent metabolic alkalosis is characteristic of gastric losses, aldosterone excess, diuretics, Bartter syndrome, or Gitelman syndrome. Diarrhea has a high concentration of potassium, and the resulting hypokalemia is usually associated with a metabolic acidosis secondary to stool losses of bicarbonate. With emesis or nasogastric suction, there is gastric loss of potassium, but this is fairly minimal given the low potassium content of gastric fluid (~10mEq/L). More important is the gastric loss of hydrochloride, leading to a metabolic alkalosis and a state of volume depletion. Metabolic alkalosis and volume depletion increase urinary losses of potassium.

13.**(A)** TBW has two main compartments: intracellular fluid (ICF) and extracellular fluid (ECF). In the fetus and newborn, the ECF volume is larger than the ICF volume. The normal postnatal diuresis causes an immediate decrease in the ECF volume, which is then followed by continued expansion of the ICF volume because of cellular growth. By 1 year of age, the ratio of the ICF volume to the ECF volume approaches adult levels. The ECF volume is 20–25% of body weight, and the ICF volume is 30–40% of body weight. With puberty, the increased muscle mass of males results in a higher ICF volume than in females.

14.**(C)** The causes of a metabolic alkalosis are divided into two categories based on the urinary chloride.

- The alkalosis in patients with a low urinary chloride is maintained by volume depletion. They are called chloride responsive because volume repletion with fluid containing sodium chloride and potassium chloride is necessary to correct the metabolic alkalosis.
- The chloride-resistant causes of metabolic alkalosis can be subdivided based on blood pressure. Patients with the rare disorders that cause metabolic alkalosis and hypertension either have increased aldosterone or act as if they have increased aldosterone. Patients with Bartter syndrome or Gitelman syndrome have metabolic alkalosis, hypokalemia, and normal blood pressure secondary to renal tubular defects, which cause continuous urinary losses of chloride.

15.**(C)** The composition of solutes in the ICF and ECF is different. Sodium and chloride are the dominant cation and anion in the ECF. Potassium is the most abundant cation in the ICF, and proteins, organic anions, and phosphate are the most plentiful anions in the ICF. The dissimilarity between the anions in the ICF and the ECF is determined largely by the presence of intracellular molecules that do not cross the cell membrane, the barrier separating the ECF and the ICF. In contrast, the difference in the distribution of cations—sodium and potassium—is due to the activity of the Na+, K+-ATPase pump, which extrudes sodium from cells in exchange for potassium.

16.**(C)**

Body Weight Method for Calculating Maintenance Fluid Volume and Rate		
BODY WEIGHT (kg)	VOLUME PER DAY	HOURLY RATE
0–10	100mL/kg	4mL/kg/hr
11–20	1,000mL + 50mL/kg for each 1kg >10kg	40mL/hr + 2mL/kg/hr × (wt−10)
>20	1,500mL + 20mL/kg for each 1kg >20kg*	60mL/hr + 1mL/kg/hr × (wt−20)†

17.**(B)** The urine specific gravity is usually elevated (≥1.025) in cases of significant dehydration but decreases after rehydration. With dehydration, a urinalysis may show hyaline and granular casts, a few white blood cells and red blood cells, and 30–100mg/dL of proteinuria. These findings usually are not associated with significant renal pathology, and they remit with therapy. Hemoconcentration from dehydration increases the hematocrit and hemoglobin.

18.**(C)** Calculation of Fluid Deficit: A child with dehydration has lost water; there is usually a concurrent loss of sodium and potassium. The fluid deficit is the percentage of dehydration multiplied by the patient's weight (for a 9-kg child with moderate dehydration (10%) of 9 kg = 900 ml deficit).

19.**(C)** Patients with distal RTA (dRTA) cannot acidify their urine and can have a urine pH greater than 5.5, despite metabolic acidosis, while the ability to acidify the urine is intact in proximal RTA, and untreated patients have a urine pH less than 5.5.

20.**(D)** As a guideline for oral rehydration, 50mL/kg of the ORS should be given within 4 hours to patients with mild dehydration, and 100mL/kg should be given over 4 hours to patients with moderate dehydration.

21.**(B)**

22.**(B)**

23.**(A)** The plasma anion gap is useful for evaluating patients with metabolic acidosis. It divides patients into two diagnostic groups: normal anion gap and increased anion gap. The following formula determines the anion gap: Anion Gap = $Na^+ - (Cl^- + HCO_3^-)$. A normal anion gap is 9 ± 4 mEq/L.

24.**(B)** There are many potential complications of PN. Central venous lines are associated with complications during insertion (pneumothorax or bleeding) and long-term issues (thrombosis). Catheter-related sepsis, most commonly due to coagulase-negative staphylococci, is common and, on occasion, necessitates catheter removal. Other potential pathogens are Staphylococcus aureus, gram-negative bacilli, and fungi. Electrolyte abnormalities, nutritional deficiencies, hyperglycemia, and complications from excessive protein intake (azotemia or hyperammonemia) can be detected with careful monitoring.

The most concerning complication of long-term parenteral nutrition is cholestatic liver disease, which can lead to jaundice, cirrhosis, and, occasionally, liver failure. Current PN decreases the risk of liver disease by including reduced amounts of hepatotoxic amino acids and possibly the use of fish-based lipids. The best preventive strategy is early use of the gastrointestinal tract, even if only trophic feeds are tolerated.

25.**(B)**

- The dextrose concentration of peripheral PN is typically 10–12%, whereas central PN has a concentration of about 20%, although it may be increased to 25–30% in patients who are fluid restricted. To avoid hyperglycemia, the dextrose delivery is increased gradually when starting PN.
- Protein delivery in PN is via amino acids in the dextrose solution. The goal is 0.8–2 g protein/kg per 24hours for older children, 1.5–3 g/kg per 24hours for full-term and older infants, and 2.5–3.5 g/kg per 24hours for preterm infants.
- The 20% lipid emulsion provides essential fatty acids and calories. The lipid emulsion is started at a rate of 0.5–1g/kg per 24hours, gradually increasing the rate so that the patient receives adequate calories; this typically requires 2.5–3.5g/kg per 24hours
- The electrolyte and mineral composition of PN depends on the age and the underlying illness.

26.**(A)** Medications, such as ifosfamide, deferasirox, or valproate, may cause Fanconi syndrome.

27.**(B)** The glucose in maintenance fluids provides approximately 20% of normal caloric needs to prevent the development of starvation ketoacidosis and diminish the protein degradation that would occur if the patient received no calories, but maintenance fluids are not a long-term replacement for nutrition.

28.**(D)** In Bartter syndrome there are severe hypokalemia (usually <2mmol/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.

29.**(A)** The plasma anion gap divides patients with metabolic acidosis into two categories: normal anion gap and increased anion gap.
• Normal anion gap metabolic acidosis occurs in the setting of diarrhea and renal tubular acidosis.
• Increased anion gap metabolic acidosis occurs with lactic acidosis, ketoacidosis, chronic renal failure, and toxic ingestions.

30.**(C)** The anion gap should not be interpreted in dogmatic isolation; consideration of other laboratory abnormalities and the clinical history improves its diagnostic utility. A decrease in the albumin concentration of 1 g/dL decreases the anion gap by roughly 4 mEq/L. Similarly, albeit less commonly, an increase in unmeasured cations, such as calcium, potassium, or magnesium, decreases the anion gap. Conversely, a decrease in unmeasured cations is a rare cause of an increased anion gap. Because of these variables, the broad range of a normal anion gap and other factors, the presence of a normal or increased anion gap is not

always reliable in differentiating the causes of a metabolic acidosis, especially when the metabolic acidosis is mild.

31.**(A)** During a simple metabolic disorder, there is **respiratory** compensation; the Pco_2 decreases during metabolic acidosis and increases during metabolic alkalosis. With metabolic acidosis, the decrease in the pH increases the ventilatory drive, causing a decrease in the Pco_2. The fall in the CO_2 concentration leads to an increase in the pH. This appropriate respiratory compensation for a metabolic process happens quickly and is complete within 12–24hours.
During a primary respiratory process, there is metabolic compensation mediated by the **kidneys**. The kidneys respond to a respiratory acidosis by increasing hydrogen ion excretion, increasing bicarbonate generation, and raising the serum bicarbonate concentration. The kidneys increase bicarbonate excretion to compensate for a respiratory alkalosis; the serum bicarbonate concentration decreases. In contrast to a rapid respiratory compensation, it takes 3–4 days for the kidneys to complete appropriate metabolic compensation.

32.**(B)** While in metabolic acidosis, the decrease in the pH increases the ventilatory drive, causing a decrease in the Pco2. The fall in the CO2 concentration leads to an increase in the pH
Expected compensation $PCO_2 = 1.5 \times [HCO_3^-] + 8 \pm 2$.

33.**(D)** Acute PN is frequently given in an intensive care unit when there is poor tolerance of enteral feeds, potentially secondary to a transient ileus, concerns regarding bowel ischemia, or the risk of aspiration pneumonia. Short bowel syndrome is the most common indication for long-term PN; it may be caused by a congenital gastrointestinal anomaly or acquired after necrotizing enterocolitis.

- ACUTE; Prematurity, Trauma, Burns, Bowel surgery, Multiorgan system failure, Bone marrow transplantation, and Malignancy.
- CHRONIC; Short bowel, Intractable diarrhea syndromes, Intestinal pseudo-obstruction, Inflammatory bowel disease, and Immunodeficiency.

SECTION 8

THE ACUTELY ILL OR INJURED CHILD

AHMED TAWFEQ

QUESTIONS

1. The first step in the focused examination of an acutely ill child is assessment of
   A. responsiveness
   B. airway
   C. breathing
   D. circulation

2. During the initial rapid assessment of an acutely ill child, the diagnostic evaluation often is started with pulse oximetry and
   A. capillary blood gas analysis
   B. serum electrolytes
   C. Doppler study of carotid arteries
   D. measurement of glucose level

3. In a collapsed child, chest compressions should be started if
   A. heart rate less than 70/min
   B. poor systemic perfusion
   C. no palpable pulse
   D. undetected blood pressure

4. The drug of choice for supraventricular tachycardia is
   A. atropine
   B. amiodarone
   C. digoxin
   D. adenosine

5. Which of the following is an indication for using calcium chloride infusion?
   A. Calcium channel blocker overdose
   B. Hypomagnesemia
   C. Hypokalemia
   D. Hyperuricemia

6. The side effect of using sodium bicarbonate may include
   A. hyponatremia
   B. hyperkalemia
   C. shifting the oxyhemoglobin curve to the left
   D. hypo-osmolality

7. What is the **MOST** serious side effect of extravasation of calcium infusion?
   A. Soft-tissue necrosis
   B. Erythema

C. Phlebitis
D. Calcinosis cutis

8. Defibrillation is usually used for
A. ventricular tachycardia
B. ventricular fibrillation
C. supraventricular tachycardia
D. atrial fibrillation

9. The cardioversion is usually used for
A. ventricular tachycardia
B. ventricular fibrillation
C. supraventricular tachycardia
D. atrial fibrillation

10. Which of the following conditions can cause hypoxic respiratory failure?
A. Aspiration pneumonia
B. Croup
C. Vocal cord paralysis
D. Post-extubation edema

11. Which of the following conditions can cause hypercarbic respiratory failure?
A. Acute bronchiolitis
B. Pediatric acute respiratory distress syndrome
C. Opioid's intoxication
D. Interstitial lung disease

12. A 4-month-old baby girl who had recently received her vaccination in the health center was complaining of high-grade fever, decreased appetite with local swelling at vaccination site for 2 days prior to admission. Investigations revealed leukocytosis with absolute neutrophilia, and increase markers of inflammation. Empiric antibiotic started but the fever continues. blood and urine cultures on day 4 of admission shows growth of E-coli. The antibiotics had changed according to sensitivity test. Mother mentioned that the baby sometimes not cope with breast feeding and near to chock. Meanwhile the baby starts to have tachypnea and tachycardia, CXR shows new infiltrate bilaterally with no cardiomegaly, but she was maintaining saturation above 92% on 2 liters of nasal O2.
Of the following, the **MOST** likely diagnosis is
A. uro-sepsis
B. aspiration pneumonia
C. vaccine related complications
D. pediatric ARDS

13. Which of the following is the **MOST** common cause of acute respiratory failure in children?
A. Bronchiolitis
B. Pneumonia
C. Upper airway obstruction

D. Pediatric ARDS

14. Which of the following meet the biochemical definition of childhood acute respiratory failure in arterial blood gas analysis?
A. Paco2 > 50mm
B. Paco2 > 60mm
C. Pao2 < 70mm
D. Pao2 < 90mm

15. What is the major complication of hypoxic respiratory failure?
A. Pneumomediastinum
B. Pneumothorax
C. Multiple organ dysfunction
D. Chronic fibrotic lung diseases

16. Which of the following can cause hypovolemic shock?
A. Sepsis
B. Anaphylaxis
C. Drug intoxication
D. Diabetes mellitus

17. Which of the following can cause distributive shock?
A. Congenital heart disease
B. Sepsis
C. Arrhythmia
D. Metabolic derangements

18. In untreated tension pneumothorax, the resultant shock is
A. cardiogenic shock
B. dissociative shock
C. distributive shock
D. obstructive shock

19. When the tissue perfusion is normal, but cells are unable to utilize oxygen because the hemoglobin has an abnormal affinity for oxygen, preventing its release to the tissues. This condition can cause
A. cardiogenic shock
B. dissociative shock
C. distributive shock
D. hypovolemic shock

20. Of the following clinical signs, the **MOST** important feature that present only in distributive shock is
A. tachycardia
B. bounding pulse
C. hypotension
D. decreased mental status

21. A 4-year-old boy presented to ER with severe pallor, tea color urine, and tinge of jaundice, his heart rate was 154 b/min., B.P 64/42 mmHg. He has weak pulse, and his capillary refill > 3 seconds. You send for cross matching and other related investigations. You gave a bolus of saline. There is improvement in vital signs but still not normalized.
Of the following, the **NEXT** step is to

A. give another bolus of normal saline
B. give a 15ml/kg of fresh frozen plasma
C. give a shot of PRBC transfusion
D. start serial measures of PCV

22. You are team leader in ER when you received acutely ill 12-month-old child who has pallor, weak peripheral pulses, with poor capillary refill. His HR is 180 bpm, and RR 60/min.
You already started supportive care and fluid therapy. You have noticed increasing dyspnea after 2nd shot of IV fluid and you had heard some rales over the back of the chest with hepatomegaly.
Of the following, the **MOST** important next action is to

A. stop further shots of IV fluids
B. start dopamine
C. give diuretics
D. do endotracheal Intubation

23. After resuscitation and initial stabilization of a drowning victim, the first investigation to be requested is

A. renal function
B. electrolytes
C. arterial Blood gas analysis
D. liver enzymes

24. You had attained a resuscitation of "unwitnessed" drowning victim.
He seems to be unresponsive.
Of the following, the **MOST** important next step is to

A. contact emergency medical service
B. start chest compression and breathing cycles
C. give 2 rescue breath
D. stabilize cervical spine

25. An 18-month-old toddler who was a victim of drowning in the bathtub for about 5 minutes. Mother had started resuscitation at home. She said he showed a response after few minutes when he opened his eyes and started to breath. On arrival to ER he was conscious, responsive, his temperature 36.5 cellules. He was having tachypnea, tachycardia, increased work of breathing, and decreased breath sounds with crackles over the lung fields. His blood pressure was normal and his O2 saturation is maintained with 2-3L of nasal O2.
Of the following, the **MOST** important next step is to

A. keep him in ER with close monitoring
B. start diuretics therapy

C. transfer him to intensive care unit
D. start to wean O2 slowly

26. The mother in the above scenario is asking about the prognosis of her child.
The **BEST** response to her would be
A. he would survive but with minimal CNS morbidity
B. he would survive but with moderate CNS morbidity
C. he may suffer mild chronic lung disease
D. he will likely to survive with intact neurologic function

27. A 3-year-old boy who survived from drowning, admitted to intensive care unit because of respiratory compromise. On day 3 of admission, he developed a pneumonic patch.
Of the following, the **BEST** choice of antibiotics should include
A. ampicillin
B. amoxicillin-clavulanate
C. ticarcillin
D. third generation cephalosporin

28. What is the **MOST** common cause of fatal childhood poisonings?
A. Analgesics
B. Hydrocarbons
C. Organophosphorus
D. Narcotics

29. Which of the following strategies is contraindicated with hydrocarbons ingestion?
A. Watchful waiting
B. Antibiotics
C. Gastric lavage
D. Steroid

30. A 3-year-old boy brought to ER. He was found comatose in the garden with evidence of hypersalivation, diaphoresis, and lacrimation. On examination he was found to have miosis, bradycardia, dyspnea, and fasciculations. You suspect intoxication.
Of the following, the **MOST** likely cause of this toxic syndrome is
A. amphetamines
B. tricyclic antidepressants
C. antihistamines
D. organophosphates

31. A 4-year-old boy brought to ER comatose. He developed a brief myoclonic seizure. His pupils were dilated but responsive to light, lower limbs show hyperreflexia. His blood pressure was on low side, his ECG showed prolonged QT interval. You suspect intoxication
Of the following, the **MOST** likely cause is
A. amphetamine
B. organophosphates
C. cyclic antidepressants
D. narcotics

32. In which of the following intoxications, a single-dose of activated charcoal can be the initial therapy?

A. Kerosene
B. Tricyclic antidepressant
C. Arsenic
D. Lead

33. In which of the following intoxications, multiple-doses activated charcoal can be the initial therapy?

A. Carbamazepine
B. Salicylate
C. Acetaminophen
D. Iron

34. A 3-year-old boy brought to ER with history of ingestion of many tablets of his mother antihypertensive medication (metoprolol) about 45 minutes before arrival. The family tried to induce vomiting at home. You have given him a single dose of activated charcoal, IV fluid and kept under close observation. The patient started to have bradycardia and hypotension. Of the following, the **MOST** important next step in management is to give

A. 10% dextrose bolus dose
B. atropine
C. glucagon
D. pralidoxime

35. What is the specific antidote for benzodiazepine intoxication?

A. N-acetylcysteine
B. Flumazenil
C. Deferoxamine
D. Naloxone

36. A 3-month-old baby girl who had referred for hearing assessment.
Of the following, what is the **BEST** choice for sedation in this procedure?

A. Midazolam
B. Chloral hydrate
C. Propofol
D. Ketamine

SECTION 8

THE ACUTELY ILL OR INJURED CHILD

AHMED TAWFEQ

ANSWERS

1.**(A)** Initial examination must focus on the responsiveness of a patient followed by assessment of the airway, breathing, and circulation.

2.**(D)**. Bedside measurement of glucose levels is important in any child with altered mental status or at risk for inadequate glycogen stores (infants, malnourished patients). After resuscitation measures, further diagnostic tests and imaging are often necessary. Hypoglycemia is common in infants and children who sustain cardiac arrest. Blood glucose should be checked, and hypoglycemia should be promptly treated with glucose.

3.**(C)** Chest compressions should be initiated if a pulse cannot be palpated or if the heart rate is less than 60 beats/min with concurrent signs of poor systemic perfusion. A and B is true if presented together.

4.**(D)**. Adenosine uses in

I. Paroxysmal SVT: Slows conduction through AV node and interrupts AV reentry pathways, which restore normal sinus symptoms.
II. Stress testing: A2A adenosine receptor agonist; activation of the A2A adenosine receptor produces coronary vasodilation and increases coronary blood flow.

- ✓ Half-life: <10 sec.
- ✓ Given as a rapid injection (1-3 sec) by peripheral IV route directly into vein or into IV line close (proximal) to patient and is followed by rapid NS flush after each injection (20 mL for adults, 5 mL or more for pediatrics).
- ✓ Place patient in mild reverse Trendelenburg position before giving drug.

5.**(A)**. Hypocalcemia, calcium channel blocker overdose, hypermagnesemia, hyperkalemia are indications for use of calcium chloride (10%), IV/IO: 20mg/kg; maximum single dose 2g; to be administered slowly.

6.**(C)** The routine use of sodium bicarbonate is currently not recommended. Sodium bicarbonate may be judiciously used to treat toxidromes or hyperkalemic arrest; however, oxygen delivery and elimination of CO2 must be established first. Side effects include hypernatremia, hyperosmolality, hypokalemia, metabolic alkalosis (shifting the oxyhemoglobin curve to the left and impairing tissue oxygen delivery), reduced ionized calcium level, and impaired cardiac function.

7.**(A)**

8.**(B)** Prompt electrical defibrillation is indicated when ventricular fibrillation or pulseless ventricular tachycardia is noted. CPR should continue until immediately before defibrillation and resume immediately afterward, minimizing interruptions in compressions. If a second attempt at defibrillation is necessary, it should be followed by a dose of epinephrine. Children failing two episodes of defibrillation may benefit from administration of amiodarone or lidocaine. Pulse ventricular tachycardia benefited from amiodarone.

9.**(C)**. Defibrillation should be distinguished from cardioversion of supraventricular tachycardias (which can also cause cardiovascular compromise). Cardioversion requires a lower starting dose and synchronization of the discharge to the electrocardiogram to prevent discharging during a susceptible period, which may convert supraventricular tachycardia to ventricular tachycardia or fibrillation.

10.**(A)** Hypoxic respiratory failure resulting from impairment of alveolar-capillary function is seen in ARDS; cardiogenic pulmonary edema; interstitial lung disease; aspiration pneumonia; bronchiolitis; bacterial, fungal, or viral pneumonia; and sepsis. It also can be due to intracardiac or intrapulmonary shunting seen with atelectasis and pulmonary embolism. B; C and D causing hypercarbic respiratory failure. This type of respiratory failure if not treated adequately can cause hypercarbia ultimately due to complications (mixed form).

11.**(C)** Hypercarbic respiratory failure can occur when the respiratory center fails as a result of drug use (opioids, barbiturates, anesthetic agents), neurologic or neuromuscular junction abnormalities (cervical spine trauma, demyelinating diseases, anterior horn cell disease, botulism), chest wall injuries, or diseases that cause increased resistance to airflow (croup, vocal cord paralysis, post-extubation edema).

12.**(D)** Pediatric (ARDS) is a non-cardiogenic pulmonary edema which can cause respiratory failure. Pediatric ARDS, and its severity, is defined by (1) timing: onset within 7 days of known clinical insult; (2) respiratory failure not fully explained by cardiac failure or fluid overload; (3) chest imaging findings of new infiltrate(s) consistent with acute pulmonary parenchymal disease; and (4) impairment in oxygenation. Pediatric ARDS can be triggered by a variety of insults including sepsis, pneumonia, shock, burns, or traumatic injury, all resulting in inflammation and increased vascular permeability leading to pulmonary edema. Numerous mediators of inflammation (tumor necrosis factor, interferon-γ, nuclear factor κB, and adhesion molecules) as well as surfactant action may be involved in the development of ARDS.

13.**(A)** Acute respiratory failure is frequently caused by bronchiolitis, often caused by respiratory syncytial virus (RSV), asthma, pneumonia, upper airway obstruction, and systemic inflammation resulting in ARDS. Notably, bronchiolitis and asthma are the most

common causes of acute respiratory failure in pediatric patients. RSV infects 90% of children within the first 2 years of life and frequently reinfects older children and adults.

14.**(A)** Acute respiratory failure occurs when the pulmonary system is unable to maintain adequate gas exchange to meet metabolic demands. The resulting failure can be classified as hypercarbic (Paco2 > 50mm Hg in previously healthy children), hypoxemic (Pao2 < 60mm Hg in previously healthy children without an intracardiac shunt), or both.

15.**(C)** The major complication of hypoxic respiratory failure is the development of organ dysfunction. Multiple organ dysfunction includes the development of two or more of the following: respiratory failure, cardiac failure, renal insufficiency/ failure, gastrointestinal or hepatic insufficiency, disseminated intravascular coagulation, and hypoxic-ischemic brain injury. Mortality rates increase with increasing numbers of involved organs. Complications associated with mechanical ventilation include pressure-related and volume-related lung injury. Both overdistention and insufficient lung distention (loss of functional residual capacity) are associated with lung injury. Pneumomediastinum and pneumothorax are potential complications of both the disease process and iatrogenic overdistention. Inflammatory mediators may play a role in the development of chronic fibrotic lung diseases in ventilated patients.

16.**(D)** Acute hypovolemia is the most common cause of shock in children. It results from loss of fluid from the intravascular space secondary to inadequate intake or excessive losses (vomiting and diarrhea, blood loss, capillary leak syndromes, or pathologic renal fluid losses) while A, B, and C are examples of distributive shock. All forms of shock produce evidence that tissue perfusion and/ or oxygenation are insufficient (increased heart rate, abnormal blood pressure, alterations of peripheral pulses). The etiology of shock may alter the initial presentation of these signs and symptoms.

17.**(B)** Distributive shock results from a relative inadequacy of intravascular volume caused by venous or arterial vasodilation. This maldistribution of blood flow can result in profound inadequacies in tissue perfusion, even in the presence of a normal or high cardiac output. Septic shock is the most common type of distributive shock in children. Other causes include anaphylaxis, neurologic injury, and ingestion of certain drugs or poisons.

18.**(D)** Etiologies of obstructive shock include both congenital lesions (such as coarctation of the aorta, interrupted aortic arch, and severe aortic valvular stenosis) and acquired diseases (tension pneumothorax; cardiac tamponade; Budd chiari syndrome; pulmonary embolism; abdominal compartment syndrome; constrictive pericarditis; superior vena cava syndrome).

19.**(B)** Etiologies of obstructive shock include both congenital lesions (such as coarctation of the aorta, interrupted aortic arch, and severe aortic valvular stenosis) and acquired diseases (tension pneumothorax; cardiac tamponade; Budd chiari syndrome; pulmonary embolism; abdominal compartment syndrome; constrictive pericarditis; superior vena cava syndrome).

20.**(B)** In hypovolemic shock the signs of sympathoadrenal activity (tachycardia, vasoconstriction), clinical manifestations include signs of dehydration (dry mucous membranes, decreased urine output) or blood loss (pallor). Patients with distributive shock usually have tachycardia and alterations of peripheral perfusion. In early stages, when cytokine release results in vasodilation, pulses may be bounding and vital organ function may be maintained (an alert patient, with rapid capillary refill and some urine output in warm shock). As the disease progresses untreated, extremities become cool and mottled with a delayed capillary refill time. At this stage, the patient has hypotension and vasoconstriction. In distributive shock the mental status is usually preserved.

21.**(C)** In case of hypovolemic shock, Isotonic crystalloids (normal saline or a balanced crystalloid solution such as lactated Ringer or Plasma-Lyte) are the initial fluids of choice. A bolus of 10–20mL/kg should be delivered in monitored conditions. Improvement, but not correction, after an initial bolus should prompt repeated boluses until circulation has been re-established. Because most children with shock have noncardiac causes, fluid administration of this magnitude is well tolerated. If hemorrhage is known or highly suspected, administration of packed red blood cells is appropriate. Monitoring for deteriorating physiologic status during fluid resuscitation (increase in heart rate or decrease in blood pressure) identifies children who may have diminished cardiac function.

22.**(A)** Both A and B are true but you should do A then B. Cardiogenic shock results when the myocardium is unable to supply the cardiac output required to support tissue perfusion and organ function. Because of this self-perpetuating cycle, heart failure progressing to death may be rapid. Patients with cardiogenic shock have tachycardia and tachypnea. The liver is usually enlarged, a gallop is often present, and jugular venous distention may be noted. Because renal blood flow is poor, sodium and water are retained, resulting in anasarca. Therapy may be initiated with dopamine at 5–20µg/kg per minute; however, epinephrine or norepinephrine may be preferable in patients with decompensated shock.

23.**(C)** After resuscitation, arterial blood gas measurement assists in assessing pulmonary gas exchange. A chemistry profile may reveal elevated liver enzymes if hypoxemia and ischemia were of long duration and provide baseline renal function. Electrolytes are often obtained, although alterations of serum electrolytes are minimal, even in freshwater drowning.

24.**(D)** Resuscitation of a drowning victim includes the basic ABCs. Victims of unwitnessed drowning require stabilization of the cervical spine because of the possibility of a fall or diving injury. The usual steps is to do; D, C, B, and A whenever possible.

25.**(C)** Those children begin breathing spontaneously and awaken before arrival at an emergency department and does not have significant pulmonary compromise still require careful observation for pulmonary complications over the subsequent 4–6 hours. Children who have evidence of lung injury, cardiovascular compromise, or neurologic compromise

should be monitored in an intensive care unit. Pulmonary dysfunction often results in hypoxemia. Oxygen supplementation should be implemented to maintain normal oxygen saturations. Mechanical ventilation may be needed in patients with significant pulmonary or neurologic dysfunction. Cardiovascular compromise is often the result of impaired cardiac contractility secondary to hypoxic-ischemic injury (can cause ARDS).

26.**(D)** The outcome of drowning is determined by the success of immediate resuscitation efforts and the severity of hypoxic-ischemic injury to the brain. Patients who have regained consciousness on arrival at the hospital will likely survive with intact neurologic function. Unfavorable prognostic markers include submersion lasting longer than 5 minutes, delayed resuscitation, the need for continued cardiopulmonary resuscitation (CPR) at the hospital, Glasgow Coma Scale of 5 or less, fixed and dilated pupils, seizures, and coma for more than 72 hours.

27.**(D)** Prophylactic antibiotics have not been shown to be beneficial, but in patients who develop delayed pulmonary infection, broad antibiotic coverage should include Aeromonas species. Aeromonas species are sensitive to tetracycline, chloramphenicol, trimethoprim-sulfamethoxazole, third-generation cephalosporins, aminoglycosides (except streptomycin) and fluoroquinolones**.** Most species are resistant to ampicillin, ticarcillin, amoxicillin-clavulanate and first-generation cephalosporins.

28.**(A)** The most common agents ingested by young children include cosmetics, personal care products, cleaning solutions, and analgesics. In 2018, fatal childhood poisonings were most commonly caused by analgesics, fumes/gases/vapors, antihistamines, and cardiovascular medications.

29.**(C)** Hydrocarbon ingestion occasionally may result in systemic toxicity, but more often it leads to pulmonary toxicity. Hydrocarbons with low viscosity, low surface tension, and high volatility pose the greatest risk of producing aspiration pneumonia; however, when swallowed, they pose no risk unless emesis is induced. Emesis or lavage should not be initiated in a child who has ingested volatile hydrocarbons.

30.**(D)** Same signs and symptoms also can be seen in nicotine poisoning.

31.**(C)** C, may present as coma, convulsions, mydriasis, hyperreflexia, arrhythmia (prolonged Q-T interval), cardiac arrest, and shock. A, may present as tachycardia, hypertension, hyperthermia, psychosis and paranoia, seizures, mydriasis, diaphoresis, piloerection, and aggressive behavior. D, may present as coma, respiratory depression, hypotension, pinpoint pupils, bradycardia.

32.**(B)** Single-dose activated charcoal decreases drug absorption when used within 1 hour of ingestion; however, it has not been shown to improve clinical outcome. Thus, it should be used selectively in the management of a poisoned patient. Charcoal is ineffective against caustic or corrosive agents, hydrocarbons, heavy metals (arsenic, lead, mercury, iron, lithium), glycols, and water-insoluble compounds.

33.**(A)** Multiple-dose activated charcoal should be considered only if a patient has ingested a life-threatening amount of carbamazepine, dapsone, phenobarbital, quinine, or theophylline.

34.**(C)** Clinical presentation of beta-blocker overdose: bradycardia, hypotension, arrhythmias, and hypoglycemia. Therapies include beta-agonists, glucagon, and phosphodiesterase inhibitors**.** However, in beta-blocker poisoning where symptomatic bradycardia and hypotension are present, high-dose glucagon is considered the first-line antidote.

35.**(B)** A is for paracetamol (acetaminophen), C for iron, and D for opioid.

36.**(B)** The best choice for sedation is chloral hydrate and dexmedetomidine, both cause no respiratory depression. A is causing anxiolytics, sedation, muscle relaxation, amnesia, C causes rapid-onset sedation for induction and maintenance of anesthesia and also used in refractory seizures. D causes anesthesia, analgesia, amnesia and have a lot of side effects.

SECTION 9

HUMAN GENETICS AND DYSMORPHOLOGY

QAHTAN ALOBAIDY

QUESTIONS

1. How many generations of family members should be at least represented in drawing a pedigree?

A. one
B. two
C. three
D. four

2. Parents of a 4-year-old boy, seeking advice of their son about his short stature. On examination you find macrocephaly, a flat midface with a prominent forehead, and rhizomelic limbs.
What is the **MOST** likely mode of inheritance of this disorder?

A. Autosomal dominant
B. Autosomal recessive
C. X-Linked dominant
D. X-Linked recessive

3. You are the responsible about the genetic counseling of sickle cell disease in hematological unit. Which of the following rules is a feature of this disease inheritance?

A. Trait appears in every generation
B. Males and females are likely to be affected equally
C. Incidence of the trait is higher in males than in females
D. Each child of an affected parent has a 1-in-2 chance of being affected

4. Which of the following is inherited as an autosomal recessive disorder?

A. Achondroplasia
B. Neurofibromatosis Type 1
C. Marfan Syndrome
D. Friedreich ataxia

5. In the outpatient clinic for rare diseases, parents seeking advice for their son who had skeletal deformity in form of a tall, thin body habitus, long thin fingers and toes, scoliosis, pes planus, and joint laxity. Eye findings include dislocation of the lens, severe myopia, flat lens, and hypoplastic iris. Cardiac findings include aortic insufficiency and dilatation of the aortic root.
Which of the following is the **MOST** likely mode of inheritance of this disorder?

A. Autosomal dominant
B. Autosomal recessive
C. X-Linked dominant
D. X-Linked recessive

6. The mother of a baby recently diagnosed as phenylketonuria is asking about the chance of having another sibling with the same condition.
The **BEST** answer is

A. 5 %
B. 25%
C. 50%
D. 100%

7. A family with neurofibromatosis type 1, asking about the manner of inheritance of such disorder?
Which of the following rules of this inheritance should be mentioned?

A. Trait appears in every generation
B. On average, 25% of siblings of the proband are affected
C. Rare traits are likely to be associated with parental consanguinity
D. Traits generally involve mutations in genes that code for enzymes

8. An affected father transmits the mutation to all his daughters, who are carriers, but not to his sons.
Which of the following disorders follow such rules of inheritance?

A. Rett syndrome
B. Incontinentia pigmenti
C. Hypophosphatemic rickets
D. Duchenne muscular dystrophy

9. A 2-year-old girl who was developmentally normal, presented with microcephaly, developmental regression, and seizure disorder late in the first year. On examination; you noticed, she is adopting a repetitive handwashing movement and autistic behaviors.
Which of the following is the **MOST** likely diagnosis?

A. Cri du chat syndrome
B. Untreated phenylketonuria
C. Smith-Lemli-Opitz syndrome
D. Rett syndrome

10. What is the mode of inheritance of myelomeningocele?

A. Autosomal dominant
B. Mitochondrial Inheritance
C. Polygenic inheritance
D. X-Linked recessive

11. A pregnant mother, her first baby underwent surgery for hypertrophic pyloric stenosis, asking about the recurrence risk for such condition in her present male fetus.
Which of the following is the **BEST** answer?

A. 5%
B. 25%
C. 50%
D. 75%

12. Which of the following inheritance pattern is affected by consanguinity?

A. Autosomal recessive
B. Mitochondrial Inheritance
C. Polygenic inheritance
D. X-Linked recessive

13. Which of the following disorders should be screened in people whose ancestors originated in the Mediterranean basin?

A. Thalassemia
B. Cystic fibrosis
C. Fanconi anemia
D. Sickle cell disease

14. Which of the following disorders is **MOST** likely to occur in fetus of pregnant women with low levels of serum α-fetoprotein?

A. Omphalocele
B. Gastroschisis
C. Autosomal trisomy
D. Neural tube defect

15. A quad screen (conjugated estriol (uE3), inhibin A, and human chorionic gonadotropin (HCG) with maternal serum α-fetoprotein) correctly identifies about 80% of women who are carrying a baby with

A. trisomy 13
B. trisomy 16
C. trisomy 18
D. trisomy 21

16. A worried healthy parents whose son had hereditary angioedema, consult you about the risk of recurrence of such condition in their progeny.
Which of the following is the **BEST** answer?

A. High
B. Moderate
C. Low
D. Extremely low

17. A worried parents whose son suspected to be a case of Prader-Willi syndrome, asking how to confirm such condition.
Which of the following is the **BEST** genetic study to confirm the diagnosis?

A. Exome Sequencing
B. Whole Genome Sequencing
C. Fluorescent in Situ Hybridization
D. Microarray Comparative Genomic Hybridization

18. How much is the risk of fetal loses in women with positive quad screen?

A. 20%
B. 40%

C. 60%
D. 80%

19.Which of the following is the **MOST** likely cytogenetic finding in a mother of Down syndrome in which the age does not seem to be a risk factor?

A. Mosaicism
B. Nondisjunction
C. Trisomy rescue
D. Robertsonian translocation

20. What is the **MOST** common congenital heart diease in children with Down syndrome?

A. Atrioventricular septal defect
B. Ventricular septal defect
C. Atrial septal defects
D. Patent ductus arteriosus

21. Which of the following type of malignancies is likely to occur in an 18-month-old child with Down syndrome?

A. Acute lymphoblastic leukemia
B. Chronic lymphoblastic leukemia
C. Acute megakaryoblastic leukemia
D. Chronic megakaryoblastic leukemia

22. On examination of a male neonate in the delivery room; you find that he has polydactyly, cutis aplasia, cleft palate and microphthalmia.
Which of the following is the **MOST** likely diagnosis?

A. trisomy 13
B. trisomy 16
C. trisomy 18
D. trisomy 21

23. The healthcare provider referred a boy with learning difficulties, to be seen by a pediatrician, who found that the baby has moderate delay in his cognitive development, broad forehead, puffiness around the eyes, flat bridge of the nose, full cheeks, and a small chin.
Which of the following is the **MOST** likely diagnosis?

A. Noonan syndrome
B. Klinefelter syndrome
C. Williams syndrome
D. Cri du Chat syndrome

24. A pregnant mother consulted you about her last sonographic findings that showed an increased amount of amniotic fluid.
Which of the following system anomalies may be associated with such finding?

A. Central nervous system
B. Urinary tract system
C. Respiratory system

D. Musculoskeletal system

25. Which of the following conditions is associated with advanced paternal age?
    A. Achondroplasia
    B. Cri du Chat syndrome
    C. Trisomy 18
    D. Trisomy 13

26. Which of the following is the **BEST** time for ultrasonic screening of fetal malformations?
    A. First trimester
    B. Second trimester
    C. Third trimester
    D. At any time

SECTION 9

HUMAN GENETICS AND DYSMORPHOLOGY

QAHTAN ALOBAIDY

ANSWERS

1.**(C)**

2.**(A)** Achondroplasia caused by a defect in cartilage-derived bone, ACH is the most common skeletal dysplasia in humans. The bony abnormalities lead to short stature, macrocephaly, a flat midface with a prominent forehead, and rhizomelic ("root of the limb") shortening of the limbs. The disorder occurs in approximately 1 in 25,000 births.

3.**(B)** Rules of autosomal recessive inheritance

- Trait appears in siblings, not in their parents or their offspring
- On average, 25% of siblings of the proband are affected (at the time of conception, each sibling has a 25% chance of being affected)
- A normal sibling of an affected individual has a two-thirds chance of being a carrier (heterozygote)
- Males and females are likely to be affected equally.
- Rare traits are likely to be associated with parental consanguinity.
- Traits generally involve mutations in genes that code for enzymes (e.g., phenylalanine hydroxylase–deficient in PKU) and are associated with serious illness and shortened life span.

4.**(D)** Other distractors are inherited as autosomal dominant.

5.**(A)** Marfan syndrome a condition that occurs in approximately 1 in 10,000 individuals, Marfan syndrome shows pleiotropy, a condition in which abnormalities in multiple organ systems are caused by a pathologic variant in a single gene. Marfan syndrome is caused by a pathologic variant in FBN1, the gene for fibrillin (a connective tissue protein). Clinical symptoms mainly involve three systems: skeletal, ophthalmologic, and cardiac.

6.**(B)** Children affected with autosomal recessive disorders are usually born to unaffected parents, each of whom carries one copy of the variant. If both members of a couple are carriers (or heterozygotes) for this variant, each of their offspring has a 25% chance of being affected.

7.**(A)** Rules of autosomal dominant inheritance

- Trait appears in every generation.
- Each child of an affected parent has a 1-in-2 chance of being affected.
- Males and females are equally affected.
- Male-to-male transmission occurs.

- Traits generally involve mutations in genes that code for regulatory or structural proteins (collagen).

8.**(D)** X-Linked Recessive Inheritance: most disorders involving the X chromosome show recessive inheritance. With only one copy of the X chromosome, males are more likely to manifest these diseases than females. Each son born to a female carrier of an X-linked recessive trait has a 50% chance of inheriting the trait, but none of her daughters would be expected to be affected (each daughter has a 50% chance of being a carrier).

9.**(D)** Only a few examples of X-linked dominantly inherited disorders have been described. Both males and females are affected by this group of disorders, but females have less severe symptoms due to X chromosome inactivation. This is the case for X-linked Rett syndrome. Rett syndrome, caused by variants in the MECP2 gene, females are normal at birth but later in the first year of life develop microcephaly, developmental regression, and often a seizure disorder. Females are often diagnosed with autism and, by 2 years of age, adopt a handwashing posture that causes them to lose all purposeful hand movements.

10.**(C)** Multifactorial disorders also known as polygenic inheritance, multifactorially inherited disorders result from the interplay of genetic and environmental factors.

11.**(A)** Hypertrophic pyloric stenosis occurring in about 1 in 300 children, hypertrophic pyloric stenosis is five times more likely to occur in males than in females. When a child with HPS is born, the recurrence risk in future progeny is 5–10% for males and 1.5–2% for females.

12.**(A)** In some cultures, it is common, even desirable, for relatives to marry. This relatedness or consanguinity increases the likelihood that offspring may be born with a rare autosomal recessive (AR) condition, as both parents may be carriers of the same pathologic gene. Generally, the closer the relation between the partners, the greater the chance that the couple shares one or more mutated genes in common, increasing the risk that offspring will have an AR disorder.

13.**(A)** People of Ashkenazi Jewish background may choose to be screened for heterozygosity for an expanding panel of AR disorders, including Tay-Sachs and Niemann-Pick diseases, Bloom and Gaucher syndromes, Canavan disease, cystic fibrosis, Fanconi anemia, and familial dysautonomia. Individuals from Western and sub-Saharan Africa may choose to be screened for sickle cell disease. People whose ancestors originated in the Mediterranean basin may be screened for thalassemia.

14.**(C)** Elevated maternal serum α-fetoprotein (MS AFP) is used to identify pregnancies in which the fetus is affected with neural tube defects, omphalocele, or gastroschisis. An additional association between low levels of MS AFP and fetal aneuploidy is known. Approximately 50% of fetuses with autosomal trisomies (trisomy 21 [Down syndrome], trisomy 18, trisomy 13) can be detected by low levels of MS AFP.

15.**(D)** The quad screen is performed in the second trimester. During the first trimester, measurement of the fetal nuchal fold by sonogram can be used to identify risk for

aneuploidy; increase in the nuchal thickness is a marker not only for fetal chromosomal anomalies but for certain genetic and structural abnormalities as well. The detection rate for aneuploidy using this technique alone is 70%. The addition of testing for abnormalities in free β-HCG and PAPP-A (pregnancy-associated plasma protein) increases first trimester screening to a detection rate of almost 90%. However, positive predictive values remain low at less than 5%.

16.**(D)** When a child is affected with the new onset of an autosomal dominant disorder, it is necessary to closely examine the parents for the presence of manifestations. If the parents are unaffected, the child's condition is most likely the result of a new mutation, in which case the risk of recurrence is extremely low (although not 0, because of the possibility of gonadal mosaicism in one of the parents).

17.**(C)** Fluorescent in situ hybridization (FISH) allows the identification of the presence, absence, or rearrangement of a specific region of DNA. This technique is useful in Prader-Willi and Angelman syndromes, in which a deletion in a segment of 15q11.2 occurs, in Williams syndrome, known to be associated with a deletion in 7q11.2, in velocardiofacial (DiGeorge) syndrome, which is associated with a deletion of 22q11.2, and in other disorders in which a small duplication or deletion is known to occur.

18.**(D)** The fetal loss rate for Down syndrome, the most viable of the autosomal aneuploidies, approaches 80%.

19.**(D)** Translocation Down syndrome refers to the type of Down syndrome that is caused by rearranged chromosome material. In this case, there are three 21 chromosomes, just like there are in trisomy 21, but one of the 21 chromosomes is attached to another chromosome, instead of being separate.

20.**(A)** Almost half of all children with Down syndrome have congenital heart disease, including atrioventricular canal, ventriculoseptal or atrioseptal defects, and valvular disease.

21.**(C)** Children with Down syndrome (DS) also have an increased risk of leukemia, with a 10- to 20-fold increase in risk compared with individuals without DS. In children with DS who are younger than 2 years of age, the type is generally acute megakaryoblastic leukemia; in individuals older than 3 years of age, the types of leukemia are similar to those of other children, with acute lymphoblastic leukemia being the predominant type.

22.**(A)** Many infants with trisomy 13 have a punched-out scalp lesion over the occiput called aplasia cutis congenita; when seen in conjunction with polydactyly and some or all the facial features, this finding is essentially pathognomonic for the diagnosis of trisomy 13.

23.**(C)** Individuals with Williams syndrome often have a striking personality. Loquacious and gregarious, they are often described as having a cocktail party personality. However, at least 10% of children with Williams syndrome have features of autism spectrum disorder.

24.**(A)** An increased amount of amniotic fluid may be associated with intestinal obstruction or a central nervous system anomaly that leads to poor swallowing. A decreased amount of

fluid may be the result of a chronic amniotic fluid leak or point to a urinary tract abnormality that results in a failure to produce urine.

25.**(A)** As a woman gets older, there is increased risk of nondisjunction leading to trisomies. Advanced paternal age may be associated with the risk of a new mutation leading to an autosomal dominant trait. paternal age increases the frequency of congenital diseases such as heart malformations as well as oral, palate and lip cleft. Moreover, mental disorders (autism, schizophrenia, bipolar disorder, low IQ level as well as ADHD) also occur more frequently in advanced father's age.

26.**(B)**

SECTION 10

METABOLIC DISORDERS

QAHTAN ALOBAIDY

QUESTIONS

1. Inborn errors of metabolism in newborn can mimic
   A. early- onset sepsis
   B. late-onset sepsis
   C. duct dependent congenital heart disease
   D. TOURCH infections

2. Clinical hyperammonemia occurs in patients with defect in the urea cycle when the ingested diet contains increased amount of
   A. protein
   B. carbohydrate
   C. fat
   D. glucose

3. Which of the following metabolic disorders presents with hepatomegaly in infancy?
   A. Tyrosinemia type 1
   B. Propionic acidemia
   C. Mucopolysaccharidosis I
   D. Congenital disorders of glycosylation

4. Ketotic hypoglycemia is a condition occurring in an otherwise healthy children with impaired tolerance for fasting.
It first appears in the
   A. first six months of life
   B. second six months of life
   C. second year of life
   D. third year of life

5. Which of the following metabolic disorders is presented with permanent ketosis?
   A. Methylmalonic acidemia
   B. Pyruvate carboxylase deficiency
   C. Succinyl-CoA:3 ketoacid CoA transferase (SCOT) deficiency
   D. Medium-chain acyl-CoA dehydrogenase deficiency (MCAD)

6. Which of the following inborn errors of metabolism can cause thrombocytopenia and neutropenia?
   A. Galactosemia
   B. Organic acidurias
   C. Pyruvate dehydrogenase deficiency
   D. Respiratory chain (mitochondrial) disorders

7. Orotic acid in urine is ordered as the initial diagnostic test for evaluation of which of the following inborn errors of metabolism?

A. Propionic acidemia
B. Urea cycle disorders
C. Fatty acid oxidation disorders
D. Hereditary fructose intolerance

8. Acylglycines in urine is ordered as the initial diagnostic test for evaluation of which of the following inborn errors of metabolism?

A. Propionic acidemia
B. Urea cycle disorders
C. Fatty acid oxidation disorders
D. Hereditary fructose intolerance

9. A 9-month-old boy presented with difficult feeding, hypotonia, and difficulty of breathing. During examination; you noticed frequent hiccup, wandering eye movements, and abnormal jerky movements.
Which of the following profiles is **MOST** useful in diagnosing such a case?

A. Plasma amino acids
B. Urine amino acids
C. CSF amino acids
D. Plasma acylcarnitine

10. A 2-month-old infant presented with hypotonia, feeding difficulties, macroglossia and hepatomegaly. ECHO study shows hypertrophic cardiomyopathy while ECG shows short P-R interval.
Which of the following is the **MOST** likely diagnosis?

A. Type 1a: von Gierke
B. Type II: Pompe
C. Type III: Forbes
D. Type IV: Andersen

11. A 3-month-old baby boy presented with recurrent attacks of hypoglycemia. Examination shows hypotonia and hepatomegaly. Lab data reveals hyperlipidemia, and hypertriglyceridemia.
Which of the following diseases is the **MOST** likely diagnosis?

A. von Gierke
B. Pompe
C. Forbes
D. Andersen

12. Which of the following glycogen storage diseases is X- linked inheritance?

A. Type I
B. Type II
C. Type III
D. Type VIII

13. Neutropenia is typically seen in which of the following glycogen storage diseases?

A. Type 1b
B. Type II: Pompe
C. Type III: Forbes
D. Type IV: Andersen

14. A neonate presented with recurrent attacks of hypoglycemia, vomiting, and diarrhea. Examination shows jaundice, hepatomegaly, and cataracts. Lab investigations reveals glycosuria, aminoaciduria with a positive reaction for reducing substances in the urine, and a normal anion-gap hyperchloremic metabolic acidosis.
Which of the following is the **MOST** likely diagnosis?

A. Fructosuria
B. Galactosemia
C. Galactokinase deficiency
D. Hereditary fructose intolerance

15. A 7-day-old girl admitted to the pediatric metabolism clinic, she was born at 40 weeks of gestation with a 2300 Gr birth weight and hospitalized for 2 days. Her parents are first degree cousins, the mother had two miscarriages. Physical examination revealed microcephaly, mild facial dysmorphism, and cardiac murmur.
Which of the following is the treatment of choice in classic form of this disease?

A. Sapropterin
B. Diet restricted phenylalanine
C. Diet free phenylalanine
D. Enzyme replacement therapy

16. Which of the following metabolic disorders has an increased concentration of urine succinyl acetone?

A. Phenylketonuria
B. Tyrosinemia
C. Homocystinuria
D. Maple syrup urine disease

17. A 9-year-old male presented with arterial thromboses, developmental delay, dislocated ocular lenses; long, slender extremities; malar flushing; and livedo reticularis.
Which of the following is the **MOST** likely diagnosis?

A. Homocystinuria
B. Marfan syndrome
C. Maple syrup urine disease
D. Argininosuccinate lyase deficiency

18. A 3-week-old female presented to emergency ward with poor feeding, vomiting, tachypnea with profound depression of the central nervous system, associated with alternating hypotonia and hypertonia, opisthotonos, and seizures. Laboratory data reveals hypoglycemia and metabolic acidosis, while amino acids measurements showed high increase in plasma leucine with less increase in isoleucine, and valine concentrations.
Which of the following is the **MOST** likely diagnosis?

A. Ornithine transcarbamylase (OTC) deficiency
B. Argininosuccinate lyase (ASL) deficiency
C. Maple syrup urine disease (MSUD)
D. Propionic acidemia

19. Which of the following urea cycle disorders is inherited as X-linked pattern?
A. N-acetylglutamate synthase (NAGS) deficiency
B. Carbamoyl phosphate synthetase 1 (CPS1) deficiency
C. Ornithine transcarbamylase (OTC) deficiency
D. Argininosuccinate lyase (ASL) deficiency

20. Which of the following urea cycle disorders cause trichorrhexis nodosa (coarse brittle hair)?
A. N-acetylglutamate synthase (NAGS) deficiency
B. Carbamoyl phosphate synthetase 1 (CPS1) deficiency
C. Ornithine transcarbamylase (OTC) deficiency
D. Argininosuccinate lyase (ASL) deficiency

21. Which of the following disorders of amino acid transport is treated by penicillamine?
A. Cystinuria
B. Hartnup syndrome
C. Homocystinuria
D. Tyrosinemia

22. Which of the following is the **BEST** line of management when ammonia level is (>1,000 µM)?
A. Sodium benzoate
B. Sodium phenylacetate
C. Phenylbutyrate
D. Hemodialysis

23. A 5-month-baby boy presented with poor feeding, vomiting, seizures, and lack of energy. After few hours, patient became comatose. A characteristic sweaty feet odor is noticed during his examination.
Which of the following is the **MOST** likely diagnosis?
A. Propionic acidemia
B. Isovaleric acidemia
C. Biotin deficiency
D. Methylmalonic acidemia

24. An 8-month-old boy presented with history of fever, convulsions, dystonic posturing, altered sensorium, and loss of motor and mental milestones for 1 month. Examination of the head showed macrocephaly, magnetic resonance imaging of the brain revealed frontoparietal atrophy, "bat-wing appearance,". Carnitine and acylcarnitine profile revealed low total carnitine, with normal levels of plasma amino acids. Urine gas chromatography mass spectrometry showed an elevated level of ketones.
Which of the following is the **MOST** likely diagnosis?

A. Glutaric acidemia I
B. Isovaleric acidemia
C. Propionic acidemia
D. Methylmalonic acidemia

25. An infant presented with seizures and developmental delay. On examination; hypotonia, deafness, vision loss, alopecia and skin rash. Investigations results revealed metabolic acidosis and hyperammonemia.
Which of the following is the **MOST** likely diagnosis?
A. Biotinidase deficiency
B. Isovaleric acidemia
C. Propionic acidemia
D. Methylmalonic acidemia

26. A 23-month-old baby girl brought for resuscitation by an ambulance with a 24-hour history of being unwell, 18-hour history of persistent vomiting and progressively worsening lethargy. On examination, she was lethargic with marked cold extremities and hypothermia. Urgent capillary blood glucose was 1 mmol/L. Venous blood gas on presentation revealed (pH 7.28, partial pressure of carbon dioxide (pCO2) 5.3, partial pressure of oxygen (pO2) 5.8, bicarbonate 18, base excess 7.9, lactate 5), and elevated liver enzymes with significant carnitine depletion.
Which of the following is the **MOST** likely diagnosis?
A. Medium-chain Acyl-CoA dehydrogenase (MCAD)
B. Long-chain hydroxyacyl-CoA dehydrogenase (LCHAD)
C. Very long-chain Acyl-CoA dehydrogenase (VLCAD)
D. 3-Hydroxy-3-methylglutaryl-CoA lyase deficiency

27. A 14-day-old infant presented with nonketotic hypoglycemia, metabolic acidosis, hepatomegaly, cardiomegaly, and the odor of sweaty feet; Radiological findings show cardiomegaly and renal cysts.
Which of the following is the **MOST** likely diagnosis?
A. Glutaric aciduria type I
B. Glutaric aciduria type II
C. Carnitine deficiency
D. Biotinidase deficiency

28. Which of the following drugs can cause secondary carnitine deficiency?
A. Carbamazepine
B. Clobazam
C. Lamotrigine
D. Sodium valproate

29. A 7-month-old male presented with history of recurrent fits and failure to thrive. On examination: hypotonia, high foreheads, flat orbital ridges, widely open fontanels, and hepatomegaly. Plain X-ray revealed bony stippling of the long bones.
Which of the following is the **MOST** likely diagnosis?
A. Zellweger syndrome

B. Adrenoleukodystrophy
C. Infantile Refsum disease
D. Mucopolysaccharidosis type 1

30. A 4-year-old boy presented to endocrinology unit as short stature, dysmorphic features, and hepatosplenomegaly; skeletal survey showed dysostosis multiplex. Ophthalmological examination shows clear cornea with retinal degeneration and papilledema. Blood film shows WBC Alder-Reilly bodies.
Which of the following is the **MOST** likely diagnosis?
A. MPS I (Hurler)
B. MPS II (Hunter)
C. MPS III (Sanfilippo)
D. MPS IV (Morquiro)

31. A 7-month-old male had regular visits to neurological unit with profound loss of CNS function. Physical examination revealed hepatosplenomegaly and radiological bony stippling of the long bones. Ophthalmological examination showed clear cornea with cherry red spots.
Which of the following is the **MOST** likely diagnosis?
A. Glucosylceramide lipidosis (Gaucher 1)
B. Glucosylceramide lipidosis 2 (Gaucher 2)
C. Sphingomyelin lipidosis A (Niemann Pick A)
D. Sphingomyelin lipidosis B (Niemann Pick B)

32. Which of the following lipidoses is positive for skeletal finding (dysostosis multiplex) which simulate that found in mucopolysaccharidosis?
A. Fabry disease
B. Metachromatic leukodystrophy
C. GM2 gangliosidosis (Tay-Sachs)
D. Generalized gangliosidosis (infantile) (GM1)

33. Which of the following diseases has normal size liver and spleen in clinical examination?
A. Wolman disease
B. Sialidosis I (formerly ML I)
C. Multiple sulfatase deficiency
D. Generalized gangliosidosis (infantile) (GM1

34. Which of the following diseases has hepatosplenomegaly in clinical examination?
A. Krabbe disease
B. Metachromatic leukodystrophy
C. ML III (pseudo-Hurler polydystrophy)
D. Glucosylceramide lipidosis (Gaucher 1)

35. Which of the following amino acids is a proven therapy for preventing stroke like episodes in MELAS (mitochondrial encephalopathy, lactic acidosis, and stroke like episodes)?
A. Histidine

B. Leucine
C. Arginine
D. Methionine

SECTION 10

METABOLIC DISORDERS

QAHTAN ALOBAIDY

ANSWERS

1.**(B)** Symptoms for inborn errors of metabolism of substrate and intermediary metabolism develop once a significant amount of toxic metabolites accumulate following the initiation of feeding and may include the following: poor feeding, vomiting, diarrhea, and/or dehydration; temperature instability; tachypnea; apnea.

2.**(A)** Infants who are affected by defects in the urea cycle may continue to do well while receiving the low-protein intake of breast milk, developing clinical hyperammonemia when dietary protein is increased, or when catabolic stress occurs.

3.**(C)** Inborn errors of metabolism presenting with hepatomegaly or hepatic dysfunction in Infants are: GSD I, GSD III, Mucopolysaccharidosis I and II, Gaucher and Niemann-Pick diseases.

4.**(C)** Ketotic hypoglycemia first appears in the second year of life, occurs in otherwise healthy children, and typically resolves spontaneously by adolescence.

5.**(C)** In disorders of ketone utilization which frequently present in the context of fasting, infection with fever, or decreased intake secondary to vomiting and diarrhea, hypoglycemia may be profound; the ketosis resolves slowly. While permanent ketosis is pathognomonic of succinyl-CoA:3 ketoacid CoA transferase (SCOT) deficiency.

6.**(B)** Many organic acid disorders present in the neonatal period. Typically, an affected newborn appears normal for the first days of life, but then may develop vomiting, poor feeding, failure to thrive, hypoglycemia, hyperammonemia, seizures, hypotonia and lethargy, progressing to coma.

7.**(B)**

8.**(C)**

9.**(C)** The CSF amino acid profile is most useful in diagnosing errors of amino acid metabolism such as nonketotic hyperglycinemia and serine biosynthesis defects.

10.**(B)** Type II: Pompe, effected enzyme is lysosomal α-glucosidase, organs affected including all; notably striated muscle, nerve cells. Prognosis very poor in neonatal form; death in the first year of life is usual; variants exist; therapy with recombinant human α-glucosidase.

11.**(A)** Type 1a: von Gierke the effected enzyme is Glucose-6- phosphatase.
Clinical syndrome: Hypoglycemia, lactic acidosis, ketosis, hepatomegaly, hypotonia, slow growth, diarrhea, bleeding disorder, gout, hyperlipidemia, hypertriglyceridemia, hyperuricemia, and xanthomas.

12.**(D)** Except for one form of hepatic phosphorylase kinase, which is X linked, glycogen storage diseases are autosomal recessive.

13.**(A)** As type 1a, but in addition have clinically significant neutropenia, periodontal disease, and inflammatory bowel disease symptoms.

14.**(B)** Galactosemia is an autosomal recessive disease caused by deficiency of galactose-1-phosphate uridyl transferase. Clinical manifestations are most striking in a neonate who, when fed breast or cow's milk, generally exhibits evidence of liver failure (hepatomegaly, hyperbilirubinemia, disorders of coagulation, hypoglycemia), disordered renal tubular function (acidosis, glycosuria, aminoaciduria), food intolerance (vomiting, diarrhea, poor feeding), neurologic changes (lethargy, irritability, seizures), and cataracts.

15.**(B)** Treatment aims to maintain plasma phenylalanine values in the therapeutic range of 120–360mM using a diet specifically restricted in phenylalanine but otherwise nutritionally complete. Treatment for life is recommended to reduce the risks of long-term neuropsychiatric problems and reduce the risk of maternal PKU syndrome. Outcome of treatment in classic PKU is excellent. Treatment with modified preparation of tetrahydrobiopterin (sapropterin) shows good response in individuals with mild to moderate PKU and in 10% of individuals with classic PKU. Enzyme replacement therapy has shown effectiveness and is FDA approved for adults with PKU.

16.**(B)** Quantitative measurement of plasma tyrosine and blood or urine succinyl acetone is performed after a positive neonatal screen. The diagnosis of tyrosinemia I is confirmed by an increased concentration of succinyl acetone; DNA testing is available.

17.**(A)** An excess of homocysteine produces a slowly evolving clinical syndrome that includes dislocated ocular lenses; long, slender extremities; malar flushing; and livedo reticularis. Arachnodactyly, scoliosis, pectus excavatum or carinatum, and genu valgum are skeletal features that mimic Marfan syndrome. Intellectual disability, psychiatric illness, or both may be present. Major arterial or venous thromboses are a constant threat.

18.**(C)** Although MSUD has intermittent-onset and late-onset forms, clinical manifestations of the classic form typically occur within 1–4 weeks of birth. Poor feeding, vomiting, and tachypnea commonly are noted, but the hallmark of the disease is profound depression of the central nervous system, associated with alternating hypotonia and hypertonia (extensor spasms, "fencing" or "bicycling"), opisthotonos, and seizures. Hyperleucinosis is the cause of the central nervous system depression and may cause acute cerebral edema. Decompensation during illness may be rapid and severe. The urine may have the odor of maple syrup.

19.**(C)** OTC deficiency is X linked. Pathogenic variants range from whole gene deletions to single nucleotide substitutions.

20.**(D)** ASL deficiency is autosomal recessive, and most children are detected as a result of an elevated citrulline on newborn screening. In addition to hyperammonemia the disorder is

characterized by hepatitis, cirrhosis, neurocognitive deficiencies, hypertension, and trichorrhexis nodosa (coarse brittle hair).

21.**(A)** Treatment is based on increasing the solubility of cystine by alkalinizing the urine with sodium bicarbonate or sodium citrate and by complexing it with compounds such as penicillamine.

22.**(D)** Ammonia can be eliminated by use of the alternate pathway agents, sodium benzoate and sodium phenylacetate, which are excreted in the urine as conjugates of glycine and glutamine. Arginine, which is usually deficient, is supplied. When ammonia levels are very high (>1,000 μM) or refractory to therapy, direct removal of ammonia using hemodialysis or hemofiltration, but not peritoneal dialysis (clearance of ammonia is substantially lower), is required.

23.**(B)** The strong odor of isovaleric acid results in sweaty feet or rancid cheese odor in untreated infants, best appreciated in body sweat or cerumen.

24.**(A)** Clinical manifestations include macrocephaly (75% of affected individuals), which may be present at birth. Before the advent of newborn screening, more than 70% of children had metabolic stroke like episodes associated with infarction of the basal ganglia and dystonia which characteristically develops after an episode of intercurrent illness or stress but may occur spontaneously.

25.**(A)** Clinical manifestations of biotin deficiency vary greatly (seizures, hypotonia, sensorineural deafness, vision loss, alopecia, skin rash, metabolic acidosis, hyperammonemia, immune deficits, ataxia, developmental delay) and depend on which enzymes in which tissues have the most biotin depletion.

26.**(A)** MCAD deficiency is the most common inborn error of β-oxidation. Hypoketotic hypoglycemia is a common manifestation, as is a Reye syndrome–like illness with hypoglycemia and elevated liver enzymes. Fatty infiltration of the liver also occurs. True hepatic failure is rare. Episodes may be recurrent in the patient or the family. Sudden infant death syndrome is reported in infants with MCAD deficiency, perhaps related to hypoglycemia.

27.**(B)** In Glutaric aciduria type II when the enzyme essentially is nonfunctional, congenital anomalies are common, including renal cysts, facial abnormalities, rocker-bottom feet, and hypospadias. Severely affected infants have nonketotic hypoglycemia, metabolic acidosis, hepatomegaly, cardiomegaly, and the odor of sweaty feet soon after birth; these infants may die within the neonatal period. Less severely affected infants may have a more episodic, Reye syndrome–like illness. Skeletal and cardiac myopathy with rhabdomyolysis can be prominent in this complex, multisystemic disease. Onset in later childhood may be marked by recurrent hypoglycemia and myopathy.

28.**(D)** Carnitine deficiency is either primary (caused by failure of intake, synthesis, or transport of carnitine) or secondary (caused by the excretion of excessive amounts of carnitine as carnityl esters in patients with other inborn errors of metabolism, treatment

with drugs that complex carnitine such as valproic acid, or as a result of renal replacement therapy).

29.**(A)** Affected infants with Zellweger syndrome have high foreheads, flat orbital ridges, widely open fontanels, hepatomegaly, bony stippling of the long bones, and hypotonia. Other anomalies are common. Failure to thrive, seizures, and nystagmus develop early, and death occurs within the first year.

30.**(B)**

31.**(C)**

32.**(D)**

33.**(B)**

34.**(D)**

35.**(C)** Specific treatment is limited for most mitochondrial defects. Providing adequate calories with intravenous dextrose during acute illness is essential. Vitamin cofactors for the respiratory chain, such as riboflavin and pharmaceutical forms of coenzyme Q, are often used, but evidence for their efficacy alone or in combination is poor. Proven therapies exist for some disorders, such as arginine supplementation for preventing stroke like episodes in MELAS and idebenone for vision loss in LHON (Leber hereditary optic neuropathy).

SECTION 11

FETAL AND NEONATAL MEDICINE

HAYDER ALMUSAWI

QUESTIONS

1. An18-day-old girl presented with greenish vomitus twice at morning. On examination the infant is irritable, and flexes her legs on her abdomen.
Of the following, the **MOST** likely cause of emesis is

A. lactase deficiency
B. over feeding
C. pyloric stenosis
D. small bowel obstruction

2. Your junior house officer is asking you to interpret a lab result of neonatal screening program for one of the new-borns who is visiting the well-baby clinic. You noticed that his T4 below the lower normal limit while TSH was normal.
Of the following, the **BEST** response is to

A. start low dose thyroxine treatment
B. repeat the test
C. consider the test as normal
D. order a sonogram of neck

3. On neonatal screening program, a term newborn male weighing 3.1 kg found to have an elevated leucine level and positive urine ketones.
What is your appropriate **NEXT** step?

A. Repeat the test
B. Order urine for reducing substance
C. Immediate neonatal ICU care
D. Perform genital examination

4. Which of the following malformations may be associated with spontaneous pneumothorax?

A. Gastrointestinal malformations
B. Neural tube defect
C. Cardiac malformations
D. Renal malformations

5. You are examining a 72- hour-old male neonate, he is on exclusive breastfeeding and has jaundice on his face and chest. TSB is 12 mg/dL, the mother is worried and asking about a rapid remedy.
Of the following, the **BEST** response is to

A. breastfeed the infant every 2 hours
B. give the baby glucose water with breast milk
C. obtain a follow-up visit within 24 hours
D. start phototherapy

6. You are called to evaluate a cyanotic male newborn who is breathing comfortably with oxygen nasal prong, His HR 140 beats/min, RR 50 breaths/min, and temperature of 37°C. Auscultation reveals clear lung fields with soft systolic ejection murmur over the left sternal border. Echocardiography shows elevated pulmonary artery pressure and right-to-left shunting through PDA.
Of the following, the **MOST** likely diagnosis is

A. persistent pulmonary hypertension
B. total anomalous pulmonary venous return
C. transposition of the great vessels
D. tricuspid atresia

7. A 6-week-old boy infant who was born at 34 weeks' gestation brought by his mother for check-up. The baby is breastfed and gaining weight appropriately. He looks well apart from mild pallor and his hematocrit is 30%.
Of the following, the **MOST** appropriate action is

A. erythropoietin therapy
B. folic acid supplementation
C. oral iron supplementation
D. reassurance

8. A full term neonate is noted to be plethoric; his haemoglobin is 24 g/dL and hematocrit 74% on a venous sample.
Of the following, the **MOST** likely complication is

A. hyperbilirubinemia
B. hypoglycemia
C. hypocalcemia
D. stroke

9. A newly delivered full-term baby presented to you with cyanosis, apnea and bradycardia; after initial resuscitation the baby still cyanotic, hypoventilated and his heart rate less than 100 beat/min.
Of the following, the **NEXT** step in management is

A. intravenous epinephrine
B. endotracheal intubation
C. external cardiac massage
D. positive pressure ventilation

10. In the delivery room you received a newborn boy with pink body, blue extremities, weak cry, extremity flexion, and HR 95 beat per minute.
How much is his APGAR score?

A. 4
B. 5
C. 6
D. 7

11. You have been called to evaluate a 33 weeks gestational age male who developed recurrent attacks of apnoea. On examination the neonate is hypothermic, pale, with

distended abdomen and abdominal wall erythema. The nurse told you, the baby has vomiting and diarrhoea as well.
What is the **MOST** likely diagnosis?

A. Septicaemia
B. Apnea of prematurity
C. Necrotizing enterocolitis
D. Acute gastroenteritis

12. Which of the following scenarios calls for urgent surgical intervention in a newborn diagnosed with NEC?

A. A 2 kg term male with sonographic suggestion of thickening of bowel loops
B. A 1 and half kg premature female with abdominal CT suggestive of pneumatosis intestinalis
C. A 2 kg term female with plain film showing multiple air fluid level
D. A 1.5 kg premature male with plain film suggestive of pneumoperitoneum

13. What is the 1st step in the management of neonate with NEC?

A. Gastrointestinal decompression with nasogastric suction
B. Surgical consultation
C. Total parenteral nutrition
D. Systemic broad-spectrum antibiotics

14. You are called to evaluate a 48 hours preterm neonate in NICU who developed sudden attack of apnoea. Physical examination reveals pale, unconscious baby with bulging fontanel and cutaneous mottling.
Of the following, the **MOST** likely cause of apnoea is

A. subdural haemorrhage
B. subarachnoid haemorrhage
C. intraventricular haemorrhage
D. intraparenchymal haemorrhage

15. Neonatal seizures noted in the delivery room often are caused by

A. hypoglycaemia
B. hypocalcemia
C. intraventricular haemorrhage
D. severe anoxia

16. A 2-month-old infant presented with history of repeated vomiting followed by recurrent attacks of seizures. On examination, the baby looks drowsy, pale, with a temperature of 37.2^0C, RBS 79 mg/dl. His OFC 43 cm. The infant was a product of forceps delivery due to cephalopelvic disproportion.
What is the **MOST** likely cause of the infant condition?

A. Subdural haemorrhage
B. Subarachnoid haemorrhage
C. Intraventricular haemorrhage
D. Intraparenchymal haemorrhage

17. Maternal humoral immunity may protect the fetus against some neonatal pathogens, such as

A. group B streptococci
B. Escherichia coli
C. Haemophilus influenzae
D. Klebsiella

18. You are called to evaluate a 2- hour-old preterm male neonate who is grunting with tachypnea and cyanosis, the baby was a product of normal vaginal delivery after 32 hours ruptured membrane, the mother had fever and leucocytosis during delivery.
What is the **MOST** likely cause of his respiratory distress?

A. Respiratory distress syndrome
B. Congenital heart disease
C. Early onset sepsis
D. Sever hypoxia

19. Your junior house officer is asking you about the type and duration of treatment of neonatal meningitis.
Of the following, the **MOST** appropriate answer is

A. ampicillin and gentamycin for 10–14 days
B. ampicillin and ceftriaxone for 10–14 days
C. ampicillin and gentamycin for 21days
D. ampicillin and ceftriaxone for 21 days

20. Which of the following infections acquired during pregnancy result in fever, jaundice, hepatosplenomegaly, hydrocephalus, intracranial calcifications and chorioretinitis?

A. Toxoplasma gondii
B. Rubella virus
C. Herpes simplex type 2
D. CMV

21. Toxoplasmosis in a 27-day-old neonate can best be confirmed by

A. IgM antibodies against toxoplasma in the mother
B. IgG antibodies against toxoplasma in the mother
C. IgM antibodies against toxoplasma in the neonate
D. IgG antibodies against toxoplasma in the neonate

22. A medical student is asking you about the lines of management of asymptomatic patient with congenital toxoplasmosis.
Of the following, the recommended guideline is

A. watchful waiting
B. follow up every 3 months till 2 years
C. pyrimethamine with sulfadiazine up to 1 year
D. start pyrimethamine when the symptoms appear

23. Which of the following congenital infections is the leading cause of sensorineural hearing loss?

A. Toxoplasma gondii
B. Rubella virus
C. Herpes simplex type 2
D. CMV

24. A 3-week-old neonate presented with history of prolonged neonatal jaundice and poor feeding. On examination; he has cataract, purpuric skin lesions, hepatosplenomegaly, and systolic murmur.
Which of the following antibodies are likely to be detected in the blood of this neonate?

A. IgM antibodies against toxoplasma
B. IgM antibodies against rubella
C. IgM antibodies against CMV
D. IgM antibodies against herpes simplex

25. The mother of a neonate with congenital rubella infection asks you about the need for isolation of her baby.
What is the **MOST** appropriate answer?

A. No need for isolation
B. Need isolation up to 3 months
C. Only the respiratory secretions transmit the virus
D. Need isolation while in hospital

26. You are asked to evaluate a 14-day-old neonate weighing 1900 gm presented with microcephaly, chorioretinitis, and hepatosplenomegaly. Skull radiology shows periventricular calcification and his blood film reveals thrombocytopenia.
Of the following, the **MOST** likely cause is

A. Toxoplasma gondii
B. Rubella virus
C. Herpes simplex type 2
D. CMV

27. A young mother with CMV infection asks you about the risk of breast feeding for her baby.
What is the **MOST** appropriate answer?

A. Mother's milk is not associated with newborn illness
B. Breast feeding is contraindicated in developed country
C. Mother's milk is safe after pasteurization
D. Breast feeding associated with 20% risk of transmitting infection

SECTION 11

FETAL AND NEONATAL MEDICINE

HAYDER ALMUSAWI

ANSWERS

1.**(D)**

2.**(B)** If ↓ T4 and ↑ TSH we repeat blood specimen or thyroid function testing and begin thyroxine treatment.

3.**(C)** ↑ Leucine level in new born screening test suggest either Maple syrup urine disease or transient elevation of leucine so requiring clinical evaluation including urine for ketones, acid-base status, amino acid studies, immediate neonatal ICU care if urine ketones positive.

4.**(D)** Spontaneous pneumothorax is seen in fewer than 1% of deliveries and may be associated with renal malformations.

5.**(C)** The clinical pattern of physiologic jaundice in term infants includes a peak indirect-reacting bilirubin level of no more than 12 mg/dL on day 3 of life. Jaundice is unphysiological or pathologic if it is clinically evident on the first day of life, if the bilirubin level increases more than 0.5 mg/dL/hour, if the peak bilirubin is greater than 13 mg/Dl in term infants, if the direct bilirubin fraction is greater than 1.5 mg/dL, or if hepatosplenomegaly and anemia are present.

6.**(A)** The diagnosis is confirmed by echocardiographic examination, which shows elevated pulmonary artery pressures and sites of right-to-left shunting. Significant right-to-left shunting through a patent foramen ovale, through a PDA, and through intrapulmonary channels is characteristic of PPHN. Echocardiography also rules out structural congenital heart disease and transient myocardial dysfunction.

7.**(D)** The physiologic anemia noted at 2–3 months of age in term infants and at 1–2 months of age in preterm infants is a normal process that does not result in signs of illness and does not require any treatment. It is a physiologic condition believed to be related to several factors, including increased tissue oxygenation experienced at birth, shortened RBC life span, and low erythropoietin levels.

8.**(A)** Hyperbilirubinemia may reflect the poor hepatic circulation or the increased amount of haemoglobin that is being broken down into bilirubin.

9.**(D)** If the neonate is apneic or hypoventilating and remains cyanotic, artificial ventilation should be initiated. If the infant does not revive, an endotracheal tube should be placed. An adequate response to ventilation includes good chest rise, return of breath sounds, well-oxygenated color, heart rate returning to the normal range (120–160 beats/minute), normal end-tidal carbon dioxide, and, later, increased muscle activity and wakefulness.

10.**(B)**

TABLE 58.7 Apgar Score			
	POINTS		
SIGNS	0	1	2
Heart rate	0	<100/min	>100/min
Respiration	None	Weak cry	Vigorous cry
Muscle tone	None	Some extremity flexion	Arms, legs well flexed
Reflex irritability	None	Some motion	Cry, withdrawal
Color of body	Blue	Pink body, blue extremities	Pink all over

At intervals of 1 and 5 minutes after birth, each of the five physiologic parameters is observed or elicited by a qualified examiner. Full-term infants with a normal cardiopulmonary adaptation should score 8–9 at 1 and 5 minutes. Apgar scores of 4–7 warrant close attention to determine whether the infant's status will improve and to ascertain whether any pathologic condition is contributing to the low Apgar score.

11.**(C)**

12.**(D)**

13.**(A)** Early clinical signs of NEC include abdominal distention, feeding intolerance/increased gastric residuals, emesis, rectal bleeding, and occasional diarrhea. As the disease progresses, patients may develop marked abdominal distention, bilious emesis, ascites, abdominal wall erythema, lethargy, temperature instability, increased episodes of apnea/bradycardia, disseminated intravascular coagulation, and shock. With abdominal perforation, the abdomen may develop a bluish discoloration. Radiographic imaging is essential to the diagnosis of NEC. The earliest radiographic finding is intestinal ileus, often associated with thickening of the bowel loops and air-fluid levels. The pathognomonic radiographic finding is pneumatosis intestinalis. Radiographic findings also may include a fixed or persistent dilated loop of bowel, intrahepatic venous gas, and pneumoperitoneum seen with bowel perforation. The management of NEC includes the discontinuation of enteral feedings, gastrointestinal decompression with nasogastric suction, fluid and electrolyte replacement, total parenteral nutrition, and systemic broad-spectrum antibiotics. When the diagnosis of NEC is made, consultation with a pediatric surgeon should be obtained. Even with aggressive and appropriate medical management, 25–50% of infants with NEC require surgical intervention.

14.**(C)** Intraventricular hemorrhage (IVH) is a common cause of seizures in premature infants and often occurs between 1 and 3 days of age. Seizures with IVH are associated with a bulging fontanel, hemorrhagic spinal fluid, anemia, lethargy, and coma.

15.**(D)** Seizures caused by hypoglycemia often occur when blood glucose levels decline to the lowest postnatal value (at 1–2 hours of age or after 24–48 hours of poor nutritional intake). Seizures caused by hypocalcemia and hypomagnesemia develop in high-risk infants and respond well to therapy with calcium, magnesium, or both. Seizures noted in the delivery room often are caused by direct injection of local anesthetic agents into the fetal scalp (associated with transient bradycardia and fixed dilated pupils), severe anoxia, or congenital brain malformation.

16.**(A)** Subdural hemorrhages are seen in association with birth trauma, cephalopelvic disproportion, forceps delivery, large for gestational age infants, skull fractures, and postnatal head trauma. The subdural hematoma does not always cause symptoms immediately after birth. Anemia, vomiting, seizures, and macrocephaly may occur in an infant who is 1–2 months of age. The treatment of all symptomatic subdural hematomas is surgical evacuation.

17.**(A)** Maternal humoral immunity may protect the fetus against some neonatal pathogens, such as group B streptococci and HSV.

18.**(C)** Early-onset sepsis often begins in utero and usually is a result of infection caused by the bacteria in the mother's genitourinary tract. Organisms related to this sepsis include group B streptococci, E. coli, Klebsiella, L. monocytogenes, and nontypeable H. influenzae. Most infected infants are premature and show nonspecific cardiorespiratory signs, such as grunting, tachypnea, and cyanosis at birth. Risk factors for early-onset sepsis include vaginal colonization with group B streptococci, prolonged rupture of the membranes (>24 hours), amnionitis, maternal fever or leukocytosis, fetal tachycardia, and preterm birth. Black race and male sex are unexplained additional risk factors for neonatal sepsis.

19.**(C)** The mainstay of treatment for sepsis and meningitis is immediate antibiotic therapy. A combination of ampicillin and an aminoglycoside (usually gentamicin) for 10–14 days is effective treatment against most organisms responsible for early-onset sepsis. The combination of ampicillin and cefepime also is proposed as an alternative method of treatment. If meningitis is present, the treatment should be extended to 21 days or 14 days after a negative result from a CSF culture.

20.**(A)**

21.**(C)**

22.**(C)** The classic findings of hydrocephalus, chorioretinitis, and intracerebral calcifications suggest the diagnosis of congenital toxoplasmosis. Serologic tests are the primary means of diagnosis. IgGspecific antibodies achieve a peak concentration 1–2 months after infection and remain positive indefinitely. For infants with seroconversion or a fourfold increase in IgG titers, specific IgM antibody determinations should be performed to confirm disease. For symptomatic and asymptomatic congenital infections, initial therapy should include pyrimethamine (supplemented with folic acid) combined with sulfadiazine. Duration of therapy is often prolonged, even up to 1 year.

23.**(D)** CMV is the most common congenital infection and the leading cause of sensorineural hearing loss, intellectual disability, retinal disease, and cerebral palsy.

24.**(B)**

25.**(D)** The most common characteristic abnormalities associated with congenital rubella include ophthalmologic (cataracts, retinopathy, and glaucoma), cardiac (patent ductus arteriosus and peripheral pulmonary artery stenosis), auditory (sensorineural hearing loss), and neurologic (behavioral disorders, meningoencephalitis, and developmental delay) conditions. In addition, infants can present with growth retardation, hepatosplenomegaly, early-onset jaundice, thrombocytopenia, radiolucent bone disease, and purpuric skin lesions ("blueberry muffin" appearance from dermal erythropoiesis). Detection of rubella-specific IgM antibody usually indicates recent infection. Measurement of rubella-specific IgG over several months can be confirmatory. Rubella virus can be isolated from blood, urine, CSF, and throat swab specimens. Infants with congenital rubella are chronically and persistently infected and tend to shed live virus in urine, stools, and respiratory secretions for 1 year. Infants should be isolated while in the hospital and kept away from susceptible pregnant women when sent home.

26.**(D)**

27.**(A)** More than 90% of infants who have congenital CMV infection exhibit no clinical evidence of disease at birth. Approximately 10% of infected infants are small for gestational age and have symptoms at birth. Findings include microcephaly, thrombocytopenia, hepatosplenomegaly, hepatitis, intracranial calcifications, chorioretinitis, and hearing abnormalities. Some infants can present with a blueberry muffin appearance as the result of dermal erythropoiesis. Skull films may reveal periventricular calcifications. An additional 10% of infected infants may not present until later in infancy or early childhood, when they are found to have sensorineural hearing loss and developmental delays. Perinatal CMV infection acquired during birth or from mother's milk is not associated with newborn illness or CNS sequelae.

SECTION 12

ADOLESCENT MEDICINE

AHMED TAWFEQ

QUESTIONS

1. A 14-year-old girl referred to endocrinology clinic for evaluation of short stature and amenorrhea, she had history of balloon dilatation of coarctation of aorta during infancy, after which the child lead normal life. Examination shows only minor findings like high, narrow roof of the mouth and Low hairline at the back of the head.
Of the following, the **MOST** likely hormonal finding is

A. high FSH
B. low FSH
C. high LH
D. low LH

2. A 16-year-old girl who had referred to endocrinology clinic for evaluation of primary amenorrhea. There was no complaint apart from school difficulties in the last few years. Examination shows dry pale skin with puffy eyes.
Of the followings, the **MOST** useful laboratory investigation would be

A. FSH
B. LH
C. TSH
D. Blood urea and creatinine

3. A 16-year-old girl presented to endocrinology clinic with complaint of (no menses for the last 3 months). Examination shows well grown girl with normal signs of puberty. She reported that in the last 6 months she had participated in competitive sport activity in the school.
Of the following, the expected hormonal change would be

A. high FSH
B. low LH
C. high androgens
D. low TSH

4. A 16-year-old girl referred to endocrinology clinic with gaining weight, disturbed menstrual cycle, acne and hirsutism. Her hormonal assay was normal apart from low level of LH, and high level of androgens.
Of the following, the **MOST** likely diagnosis is

A. polycystic ovary disease
B. adrenal tumor
C. late-onset congenital adrenal hyperplasia
D. hypothyroidism

5. A 16-year-old girl presented with weight loss of about 30% of ideal weight and secondary amenorrhea. She is always concern about her weight and consider herself as an (obese girl).

Physical examination shows fine hair on the face and trunk, rough and scaly skin, bradycardia, hypothermia, erosion of tooth enamel, and acrocyanosis of the hands and feet. Of the following, the **MOST** likely diagnosis is

A. hypothyroidism
B. obsessive-compulsive disorder
C. bulimia nervosa
D. anorexia nervosa

6. A 15-year-old girl presented with history of significant weight loss, and she is much concerned about consumption of food supplements, she dislikes many types of food and sometimes using feeding tube for eating. Her past medical history reveals mild type of autism. She looks pale and malnourished; her vitals were normal.
Of the following, the **MOST** likely diagnosis is

A. bulimia nervosa
B. anorexia nervosa
C. avoidant restrictive food intake disorder
D. obsessive-compulsive disorder

7. Regarding girls, the first external manifestation of puberty is

A. breast development
B. growth of pubic hair
C. increase in height velocity
D. menstruation

8. Which of the following indicate an early stage of puberty in boys?

A. Deepening of the voice
B. Pubic hair
C. Axillary hair
D. Increase in height velocity

9. The street name of (cannabis, weed or joints) refer to which substance abuse?

A. LSD
B. Marijuana
C. MDMA
D. Phencyclidine

10. Which of the following substances that abused by adolescent is a hallucinogenic agent?

A. Cocaine
B. Amphetamines
C. Codeine
D. Flunitrazepam

SECTION 12

ADOLESCENT MEDICINE

AHMED TAWFEQ

ANSWERS

1.**(A)** Primary amenorrhea is defined as the absence of menstruation by 15 years of age in the presence of secondary sexual characteristics, 13 years of age in the absence of secondary sexual characteristics, or no onset of menses within 3–4 years of thelarche. Features that should prompt earlier evaluation include signs of hirsutism, a history of excessive exercise or an eating disorder, or absence of menses at any age with symptoms suggestive of an outflow tract obstruction (e.g., cyclic abdominal pain). Endocrine evaluation is indicated for girls with primary amenorrhea without secondary sexual characteristics. An elevated follicle-stimulating hormone (FSH) over 25 IU/L on two occasions 4 weeks apart indicates hypergonadotropic hypogonadism, which may reflect ovarian dysgenesis/agenesis and warrants a karyotype looking for Turner syndrome.

2.**(C)** All are true in evaluation of primary amenorrhea. A and B are important for subtyping to hyper or hypo gonadotrophic hypogonadism, C goes with the findings in the scenario. D is unlikely. Hypothyroidism is a common cause of menstrual dysfunction. Prolactinoma, although rare, must be ruled out.

3.**(B)** Secondary amenorrhea refers to the cessation of menses for more than 3 consecutive months in girls who previously had regular menses, or greater than 6 months in girls who had irregular menses. The most common causes of secondary amenorrhea are pregnancy, anorexia/stress (low luteinizing hormone [LH], FSH, and estradiol), and polycystic ovary syndrome (PCOS).

4.**(A)** In PCOS, there may be symptoms of androgen excess, such as acne and hirsutism, weight gain, and, with insulin resistance, acanthosis. If hirsutism or virilization is present, free and total testosterone, androstenedione, and didehydroepiandersterone sulfate (DHEAS) should be measured to rule out ovarian or adrenal tumors. Weight gain is against adrenal tumors. A normal 17-hydroxyprogesterone level rules out late-onset congenital adrenal hyperplasia.

5.**(D)** In addition patients with anorexia can presented with wearing oversized, layered clothing to hide appearance or, conversely, tight-fitting clothing to exaggerate appearance.

6.**(C)** Avoidant Restrictive Food Intake Disorder (ARFID) is a newly categorized eating disorder. ARFID is similar to anorexia in that both disorders involve limitations in the amount and/or types of food consumed, but unlike anorexia, ARFID does not involve any distress about body shape or size, or fears of fatness. Although many children go through phases of picky or selective eating, a person with ARFID does not consume enough calories to grow and develop properly and, in adults, to maintain basic body function. In children, this results in stalled weight gain and vertical growth; in adults, this results in weight loss.

ARFID can also result in problems at school or work, due to difficulties eating with others and extended times needed to eat.

7.**(A)** It's started by A then B, C and lastly by D.

8.**(A)** Testicular enlargement (≥2.5 cm) corresponds to Tanner stages 1 to 2 for boys. Testicular enlargement is followed by pubic hair development at the base of the penis (adrenarche) and then axillary hair within the year. The growth spurt is a relatively late event; it can occur from 10½ to 16 years of age. Deepening of the voice, the appearance of facial hair, and acne indicate the early stages of puberty.

9.**(B)** A street name is (acid), C is (ecstasy, molly) and D is (PCP, angel dust).

10.**(C)** A and B are stimulant agents while D is a tranquilizer.

SECTION 13

IMMUNOLOGY

MOHAIMEN ALKABI

QUESTIONS

1. The adaptive immune system consists of
   A. neutrophils
   B. monocytes/macrophages
   C. natural killer (NK) cells
   D. T and B lymphocytes

2. Which of the following immune deficiency disorders presents in the first months of life?
   A. Congenital agammaglobulinemia
   B. Severe combined immunodeficiency (SCID)
   C. Common variable immunodeficiency
   D. Leukocyte adhesion deficiency (LAD)

3. You are evaluating a 10-year-old girl who has otitis media infection. She has had four episode of pneumonias and two of otitis media in the last 2 years.
Which of the following diagnostic studies is important?
   A. Lymphocyte count and morphology
   B. C1-inhibitor level and function
   C. Quantitative immunoglobulin levels
   D. Delayed hypersensitivity skin tests

4. The preferred screening test for patients with complement defects is
   A. C 3
   B. C 4
   C. C 2
   D. CH50

5. A 13-month-old boy presented with fever, vomiting, fit, and bulging fontanelle. He had 3 episodes of otitis media. The mother reports that he had a brother who died because of bacterial infection, when he was one year old. Physical examination reveals absence of tonsillar tissue.
Of the following, the **MOST** likely explanation for these findings is
   A. agammaglobulinemia
   B. common variable immune deficiency
   C. transient hypogammaglobinemia
   D. severe combined immune deficiency

6. A 14-year-old presents with coughing, rhinorrhea, sneezing, respiratory distress, and fever (up to 39.5 c). Past medical history reported recurrent attack of pneumonia, sinusitis and immune thrombocytopenia. Physical examination reveals congested tonsil.
Of the following, the **MOST** likely diagnosis is

A. chronic granulomatous disease
B. severe combined immunodeficiency
C. X-linked agammaglobulinemia
D. common variable immune deficiency

7. You are evaluating a 3-year-old boy in the emergency department for anaphylaxes after IVIG administration. Past medical history was normal.
Which of the following is the **MOST** likely diagnosis?

A. HIV
B. IgM deficiency
C. IgG deficiency
D. Selective IgA deficiency

8. A 6-month-old male infant presented with history of diarrhea for the last 3 weeks and chronic rash in oral and perineal area. Past medical history reveals two episodes of pneumonia. Physical examination reveals failure to thrive, oral and perineal candida infections.
Of the following, the **MOST** likely diagnosis is

A. severe combined immunodeficiency (SCID)
B. chronic granulomatous disease
C. common variable immunodeficiency
D. hyper IgM syndrome

9. A 14-month-old boy brought to ER with tetanic seizure. His parents state that their son's has recurrent attack of tetany since birth. Past medical history reveals frequent attacks of otitis media, and one attack of pneumonia requiring hospitalization last month. During your review of the laboratory studies, you noticed that the infant has hypocalcemia.
Of the following, the **MOST** likely diagnosis is

A. Bruton (X-linked) agammaglobulinemia
B. chronic granulomatous disease
C. DiGeorge syndrome
D. Wiskott-Aldrich syndrome

10. A 6-year-old boy is referred to you for evaluation of recurrent sinopulmonary infections. Past medical history revealed severe lower respiratory tract infections in last year with diagnosis of P. jirovecii pneumonia. Serum IgA and IgG were undetected while IgM was normal.
Which of the following is the **MOST** likely diagnosis?

A. Common variable immunodeficiency
B. Hyper IgM syndrome

C. IgG subclass deficiency
D. Specific antibody deficiency syndrome

11. A 7-month- old boy presented with erythroderma, desquamation, chronic diarrhea, failure to thrive, lymphadenopathy, eosinophilia, hepatosplenomegaly, and elevated serum IgE.
Of the following, the **MOST** likely diagnosis is
A. Omenn syndrome
B. bare lymphocyte syndrome
C. reticular dysgenesis
D. Wiskot Aldrich syndrome

12. A 5-month-old girl presented with recurrent oral thrush, failure to thrive, and two episodes of otitis media. Laboratory evaluation reveals undetected IgG, IgA, IgM and total lack of both lymphocytes and granulocytes in their peripheral blood.
Of the following, the **MOST** likely diagnosis is
A. bare lymphocyte syndrome
B. reticular dysgenesis
C. agammaglobulinemia
D. common variable immunodeficiency

13. A 12-month-old boy presented with severe eczema since birth. His parents state that their son's eczema started soon after birth and has not responded to topical steroids. The past medical history reveals that he had frequent otitis media and pneumonias. Further evaluation reveals thrombocytopenia and elevated levels of IgE and IgA, with decreased IgM.
Of the following, the **MOST** likely diagnosis is
A. Bruton (X-linked) agammaglobulinemia
B. chronic granulomatous disease
C. DiGeorge syndrome
D. Wiskott-Aldrich syndrome

14. An 8-year-old boy is brought to your office for evaluation of failure to thrive and persistent eczema. The child had several episodes of pneumonia, skin abscesses and recurrent fractures. Physical examination reveals coarse face and failure of shedding primary teeth. His serum IgE level is very high.
Of the following, the **MOST** likely CBC finding is
A. small platelets
B. neutrophilia
C. lymphopenia
D. eosinophilia

15. Which of the following pathogens is a life threating infection in X-linked lymphoproliferative disease?
A. Epstein-Barr virus

B. Cytomegalviruse
C. HIV
D. Human polio virus

16. A 4-year-old boy presents for evaluation of unsteady gait and frequent falls. He had history of recurrent episodes of otitis media and sinopulmonary infection. Physical examination reveals oculocutaneous telangiectasia.
Which of the following is **MORE** likely to occur in this child?

A. Precious puberty
B. Leukemia
C. Autoimmune disease
D. Metaphyseal dysplasia

17. Which of the following immune deficiency disorders presents with autoimmune endocrinopathy?

A. Hyper IgM syndrome
B. Chronic mucocutaneous candidiasis
C. Nijmegen breakage syndrome
D. X-linked lymphoproliferative disease

18. A 6-year-old girl presented for evaluation of recurrent oral ulcers, joints pain, lymphadenopathy, and pharyngitis. The mother reported that, for the last year, the child had episodes of fever every 3-4 weeks that lasts for 5 days and resolved.
Which of the following is the **MOST** likely diagnosis?

A. Chronic granulomatous disease
B. Myeloperoxidase deficiency
C. Congenital neutropenia
D. Cyclic neutropenia

19. What is the hallmark of leukocyte adhesion deficiency type 1?

A. Delay separation of umbilical cord
B. Recurrent lung abscess
C. Absence of pus at site of infection
D. Neutrophil count usually is greater than 20,000/mm3

20. A 3-year-old boy presented with obstructive uropathy and two episodes of lung abscess. A physical examination reveals lymphadenopathy, hepatosplenomegaly, dermatitis, and failure to thrive. Laboratory data shows hypergammaglobulinemia.
Of the following, the **MOST** likely diagnosis is

A. Gaucher disease
B. acute leukemia
C. leukocyte adhesion deficiency II
D. chronic granulomatous disease

21. A 4-year-old girl presents with recurrent episodes of non-pruritic facial, lips, and periorbital swelling lasting 48–72 hours that occur spontaneously or after minor trauma.

The last episode associated with stridor and respiratory distress which does not respond to steroid, the mother noticed her father had similar episode associated with abdominal pain. Of the following, the **MOST** likely cause is

A. deficiency of C1-inhibitor
B. C1q, C1r deficiency
C. properdin deficiency
D. factor h deficiency

22. Which of the following complement deficiencies associated with recurrent Neisseria infections?

A. C1-inhibitor deficiency
B. C2 deficiency
C. C3 deficiency
D. C4 deficiency

SECTION 13

IMMUNOLOGY

MOHAIMEN ALKABI

ANSWERS

1.**(D)** The innate immune system includes soluble factors, including acute-phase proteins, cytokines, chemokines, and complement, as well as cellular components, including neutrophils, monocytes/macrophages, innate lymphoid cells, and natural killer (NK) cells. The adaptive immune system consists of T and B lymphocytes and their effector molecules.

2.**(D)** Age of onset of symptoms can be helpful in defining an immune deficiency, although significant variability occurs. Neutrophil defects (e.g., congenital neutropenia, LAD, CGD) typically present in the first several months of life. Antibody defects (e.g., congenital agammaglobulinemia) and severe T-cell defects (e.g., severe combined immunodeficiency [SCID]) typically present after 3 months of life after maternal antibody levels have waned. Presentation with symptoms of an antibody deficiency later in life (adolescence or adulthood) suggests common variable immunodeficiency (CVID) rather than congenital agammaglobulinemia, although milder phenotypes of primary immunodeficiency disease may not present until later in life.

3.**(C)** Quantitative immunoglobulin levels: IgG, IgA, IgM, IgE, isohemagglutinin titers (anti-A, anti-B, measures IgM function) is essential for patients with B-cell defects.

4.**(D)**

5.**(A)** Agammaglobulinemia usually present during the first 6–12 months of life with recurrent infections, sinusitis, pneumonia, meningitis (encapsulated bacteria, enteroviruses, atypical bacteria).

6.**(D)** Common variable immunodeficiency (CVID) usually present in the second and third decades of life with sinusitis, bronchitis, bronchiectasis, pneumonia, interstitial lung disease, chronic diarrhea, autoimmune cytopenias, and lymphomas.

7.**(D)**

8.**(A)** Clinical manifestations of SCID include failure to thrive, severe bacterial infections, chronic candidiasis and other fungal infections, chronic viral infections, infection with Pneumocystis jirovecii and other opportunistic organisms, and intractable diarrhea.

9.**(C)** DiGeorge syndrome has deletion on chromosome 22q11.2. DiGeorge syndrome is classically characterized by hypocalcemic (hypoparathyroidism) tetany, conotruncal and aortic arch anomalies, and increased infections. The diagnosis is established by fluorescent in situ hybridization, chromosomal microarray studies, or a polymerase chain reaction with a DNA probe to detect the chromosomal deletion.

10.**(B)** Hyper-IgM syndrome is characterized by a failure of immunoglobulin isotype switching from IgM and IgD to IgG, IgA, or IgE, and a lack of memory responses. Affected patients have normal or elevated serum levels of IgM with low or absent levels of IgG, IgA, and IgE. All patients with hyper-IgM syndrome have increased susceptibility to sinopulmonary infections, whereas patients with defects in CD40 ligand or CD40 are susceptible to opportunistic infections, such as P. jiroveci (carinii) and Cryptosporidium parvum.

11.**(A)** Omenn syndrome presents with skin and pulmonary abscesses, fungal infections, eczema, elevated IgE, coarse facial features, failure to shed primary teeth, and frequent fractures.

12.**(B)** Reticular dysgenesis presents with recurrent oral thrush, sinopulmonary infection, recurrent otitis media, failure to thrive, agamaglubinemia, alymphocytosis, and agranulocytosis.

13.**(D)** Wiskott-Aldrich syndrome is an X-linked disorder characterized by thrombocytopenia, eczema, defects in cell-mediated and humoral immunity, and a predisposition to lymphoproliferative disease. Wiskott-Aldrich syndrome is caused by mutations of the *WAS* gene coding for the Wiskott-Aldrich syndrome protein (WASP), which is expressed in lymphocytes, platelets, and monocytes. Deficiency of this protein results in elevated levels of IgE and IgA, decreased IgM, poor responses to polysaccharide antigens, waning T-cell function, and profound thrombocytopenia. Opportunistic infections and autoimmune cytopenias become problematic in older children.

14.**(D)** Hyper-IgE syndrome is characterized by markedly elevated serum IgE levels, a rash that resembles atopic dermatitis, eosinophilia, and staphylococcal abscesses of the skin, lungs, joints, and viscera. Infections with H. influenza type b, Candida, and Aspergillus also may occur. These patients have coarse facial features, joint laxity, osteopenia with fractures, and may have giant pneumatoceles in the lungs after staphylococcal pneumonias. Although serum IgG, IgA, and IgM levels are normal, humoral immune responses to specific antigens are reduced, as is cell-mediated immunity.

15.**(A)**

16.**(B)** Ataxia telangiectasia cells are exquisitely sensitive to irradiation. Malignancies including leukemia or lymphoma can occur. Diabetes also may be present, and sexual maturation is delayed.

17.**(B)** Chronic mucocutaneous candidiasis (autoimmune polyendocrinopathy- candidiasis-ectodermal dystrophy [APECED]) is characterized by chronic or recurrent candida infections of the mucous membranes, skin, and nails.

18.**(D)** Cyclic neutropenia is a stem cell disorder in which all marrow elements cycle resulting in transient neutropenia. It may be transmitted as an autosomal dominant, recessive, or sporadic disorder. Because of the short half-life of neutrophils in the blood (6–7 hours) compared to platelets (10days) and red blood cells (120 days), neutropenia is the only

clinically significant abnormality. The usual cycle is 21 days, with neutropenia lasting 4–6 days, accompanied by monocytosis and often by eosinophilia.

19.**(C)** A hallmark of defects in neutrophil migration is the absence of pus at sites of infection.

20.**(D)** Patients with **CGD** characteristically have lymphadenopathy, hypergammaglobulinemia, hepatosplenomegaly, dermatitis, failure to thrive, anemia, chronic diarrhea, and abscesses. Infections occur in the lungs, the middle ear, gastrointestinal tract, skin, urinary tract, lymph nodes, liver, and bones. Granulomas are prominent and may obstruct the pylorus or ureters.

21.**(A)** Congenital deficiency of C1-inhibitor results in hereditary angioedema, characterized by recurrent episodes of no pruritic angioedema lasting 48–72 hours that occur spontaneously or after minor trauma, stress, or anxiety. Intestinal edema can cause acute abdominal pain; edema of the upper airway can be life threatening and may necessitate emergency tracheostomy. Importantly, the angioedema in this disorder is not responsive to corticosteroids, and this feature is useful in distinguishing from more common forms of idiopathic angioedema. Hereditary angioedema is inherited as an autosomal dominant disease.

22.**(C)**

SECTION 14

ALLERGY

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QUESTIONS

1. You are discussing with medical students the types of hypersensitivity disorders of the immune system which are classified into four groups, based on the mechanism of tissue inflammation.
Which of the following disorders is an example of type II hypersensitivity reaction?

A. Urticaria
B. Contact dermatitis
C. Hypersensitivity pneumonitis
D. Goodpasture syndrome

2. Which of the following immunoglobulins is responsible for type I hypersensitivity reaction?

A. IgA
B. IgG
C. IgM
D. IgE

3. Tuberculin skin test is an example of which type of hypersensitivity reactions?

A. I
B. II
C. III
D. IV

4. You are evaluating a 10-year-old asthmatic child for possible house dust mite allergy as a cause for his poorly controlled asthma.
Of the following, the **MOST** sensitive method for detecting the presence of allergen-specific house dust mite is

A. properly performed skin tests
B. enzyme-linked immunosorbent assay (ELISA)antigen-specific IgE
C. high serum IgE level
D. serum eosinophilia

5. You prescribed amoxicillin syrup to a 4-year-old girl to treat a lower respiratory tract infection; the mother is concerned about possible penicillin allergy.
Of the following, the **BEST** statement to address mother's fears is

A. penicillin allergy should be evaluated when the individual is well
B. penicillin allergy should be evaluated only when the individual need penicillin treatment
C. treatment with penicillin should be avoided in such patient
D. there is no reliable way of detecting penicillin allergy

6. A 6-year-old boy presented with clear thin rhinorrhea, nasal congestion, sneezing paroxysms, and pruritus of the eyes, nose, ears, and palate. Physical examination is unremarkable apart from bilateral clear nasal discharge. His symptoms happen mainly after returning from his swimming lessons.
Of the following, the **MOST** likely diagnosis is

A. rhinosinusitis
B. seasonal allergic rhinitis
C. perennial allergic rhinitis
D. noninfectious rhinitis

7. Which of the following is the **MOST** frequently used drug for treatment of allergic rhinitis?

A. Intranasal corticosteroids
B. Antihistamines
C. Decongestants
D. Nasal anticholinergic

8. Which of the following drugs will cause false negative skin prick test?

A. Systemic Corticosteroid
B. Inhaled Corticosteroid
C. Antihistamine
D. β_2 antagonist

9. What is the **MOST** common cause of food-induced eczematous reactions?

A. Milk
B. Egg
C. Nut
D. Fish

10. You are examining a 1 -year- old boy with atopic dermatitis with recent change of his skin lesion as pustular lesions with crusting and honey-colored exudate involving his face.
Of the following, the **MOST** likely causative microorganism is

A. Staphylococcus aureus
B. Staphylococcus epidermdis
C. Streptococcus viridans
D. Herpes simplex

11. Which of the following areas of the skin is spared in infant with atopic dermatitis?

A. Scalp.
B. Cheeks
C. Extensor surfaces of the extremities
D. Diaper area

12. A mother bought her 11–month-old boy with history of atopic dermatitis, recurrent otitis media. His CBC shows thrombocytopenia and small-sized platelets.
Of the following, the **MOST** likely diagnosis is

A. Langerhans cell histiocytosis
B. Wiskott-Aldrich syndrome

C. Dermatitis herpetiformis
D. Immune thrombocytopenic purpura

13. You are asked by the parents of an infant with atopic dermatitis about the prognosis of their infant condition.
Of the following, the **BEST** statement is
A. atopic dermatitis is a chronic, relapsing skin disorder with no remission
B. atopic dermatitis is a curable disease with good treatment protocol
C. symptoms become more severe in two thirds of children
D. complete remission occurs in approximately 20% of the cases

14. A 10 -year -old boy presented with history of recurrent attacks of swelling that involves the face, peripheral extremities, genitalia, and oropharynx without urticarial lesion. Episodes are often triggered by trauma. Investigation shows decreased level of C4 and C1 inhibitor.
Of the following, the **MOST** likely diagnosis is
A. type I hereditary angioedema
B. idiopathic angioedema
C. type II Hereditary angioedema
D. vasculitis

15. Which of the following is the **MOST** common cause of acute urticaria in children?
A. Food
B. Medication
C. Insect sting
D. Infection

16. A 13–year-old boy presented with history of acute urticarial rash, swelling that involves his lips and tongue, and hypotension.
Which of the following tests may help you to reach the diagnosis?
A. Serum Histamine
B. Serum tryptase
C. Serum interleukin-4
D. Serum interleukin-5

17. A 12-year-old boy had history of episodic urticaria from early infancy with sensorineural deafness that progressively became worse and he got amyloidosis, arthralgias, and skeletal abnormalities.
Of the following, the **MOST** likely diagnosis is
A. mastocytosis
B. Muckle-Wells syndrome
C. Schnitzler syndrome
D. urticarial vasculitis

18. Which of the following is the **MOST** common type of physical urticaria?
A. Dermatographism
B. Cholinergic urticarial

C. Cold urticarial
D. Exercise-induced anaphylaxis

19. A school age child got stung in his hand by a yellow jacket, after 24 hours, a large local painful erythematous swelling developed.
Of the following, the **BEST** treatment option is
A. oral antihistamine
B. oral corticosteroid
C. oral antibiotics
D. oral analgesic

20. A mother of a 10-year-old boy is concerned about his seafood allergy (is my child going to outgrow this)?
Which of the following would be your appropriate answer?
A. For patient without asthma, 85% of them develop tolerance by 3 years old
B. For patient with asthma, 21% of patients have persistent allergy at age 16 years
C. Nine percent of patients will develop oral tolerance
D. Allergy will continue to be a lifelong affliction

21. You are called for examination of a 10-year-old boy with history of intensely pruritic vesicular rash on extensor surfaces and buttocks with previous history of chronic diarrhea, malabsorption, and abdominal distention. His weight is below 3rd centile. Skin biopsy shows IgA granule deposits at the dermal-epidermal junction.
Of the following, the **MOST** likely diagnosis is
A. atopic dermatitis
B. bullous pemphigoid
C. linear IgA bullous dermatosis
D. dermatitis herpetiformis

22. A pharmacist mother consults you about her 10-year-old boy with poorly controlled asthma. He is treated with formoterol-budesonide (6.5/500 mg) every 12 hour, his adherence and technique were satisfactory. Total serum IgE was 400 IU with positive skin prick test for house dust mite.
Which of the following biological agents would be appropriate for this boy?
A. Dupilumab
B. Omalizumab
C. Benralizumab
D. Mepolizumab

23. You are consulted to examine a 6-year-old asthmatic boy for follow up visit. He had a day time symptom occurring more than twice per week.
Of the following, the **PREFERRED** step of treatment would be
A. short acting B- agonist as needed
B. low dose ICS
C. montelukast
D. theophylline

24. You are discussing with medical students the value of asthma predictive index (API) for diagnosis of asthma in children below 5 years old.
Which of the following is a major criterion of modified API?

A. Inhalant allergen sensitization
B. Food allergen sensitization
C. Allergic rhinitis
D. Eosinophils ≥4%

25. In children younger than 3 years of age, the strongest predictor for wheezing to continue into persistent asthma is

A. parental asthma
B. severe bronchiolitis (i.e., requiring hospitalization)
C. atopic dermatiis
D. peripheral blood eosinophilia >4%

26. A mother brought her 3-year-old child with history of recurrent wheeze and frequent visits to emergency department; you suspect asthma.
Which of the following may aids your diagnosis?

A. Perform spirometry test
B. Measure peak expiratory flow
C. Methacholine challenge test
D. Therapeutic trial of controller medications

27. You are examining an 8-year- old child with acute asthmatic attack who had bilateral wheeze with signs of respiratory distress, his respiratory rate 40 bpm, SpO2 88% and temperature was 40^0C. The house officer suggests CXR.
Which of the following is an indication for his suggestion?

A. Moderate- severe exacerbation
B. Before starting systemic corticosteroid
C. His high temperature
D. His low SpO2

28. Which of the following is the **MOST** common chronic childhood illness?

A. Asthma
B. Diabetes mellitus
C. Celiac disease
D. Epilepsy

29. Exhaled nitric oxide (FeNO) is useful for diagnosis of

A. allergic rhinitis
B. asthma
C. cystic fibrosis
D. primary ciliary dyskinesia

30. Which of the following is an indication of CXR in acute asthma exacerbation?

A. First episode
B. SPo2 < 90%

C. Patient requiring systemic steroid
D. Severe episode

31. Which of the following will reduce local side effects of inhaled corticosteroids in asthmatic patients?
A. Using LABA with ICS
B. Using dry powder preparation of ICS
C. Rinsing the mouth with water after using ICS
D. Using low potency ICS

32. Which of the following β_2 agonist can be given once daily in asthmatic patients?
A. Salbutamol
B. Salmeterol
C. Formoterol
D. Vilanterol

33. Which of the following Leukotriene modifiers (daily oral asthma controller medications) act as leukotriene synthesis inhibitors?
A. Montelukast
B. Zafirlukast
C. Zileuton
D. Pranlukast

34. Which of the following Leukotriene modifiers is **MOST** commonly used in children?
A. Montelukast
B. Zafirlukast
C. Zileuton
D. Pranlukast

35. Which of the following is the **MOST** common complication of intranasal corticosteroids use?
A. Nasal bleeding
B. Septal perforation
C. Adrenal suppression
D. Sneezing

36. Which of the following drugs is considered as first-generation antihistamines?
A. Hydroxyzine
B. Cetirizine
C. Desloratadine
D. Fexofenadine

37. Which of the following is an example of non-immune mediated adverse drug reaction?
A. Hemolytic anemia from cephalosporins
B. Contact dermatitis from topical antihistamine
C. Stevens-Johnson syndrome from antiepileptic
D. Hemolytic anemia in a patient with G6PD deficiency after primaquine therapy

38. Which of the following is the **MOST** common cause of chronic urticaria in children?
    A. Drugs
    B. Insect allergy
    C. Food allergy
    D. Idiopathic

39. Serum sickness is an example of which type of hypersensitivity reaction?
    A. I
    B. II
    C. III
    D. IV

40. Which of the following substances can cause anaphylactoid reaction?
    A. Latex
    B. Allergen immunotherapy
    C. Biologic agent
    D. Radiocontrast media

41. The risk of having an allergic reaction to a cephalosporin in a child who has reacted positively to penicillin skin testing is
    A. < 2%
    B. > 10%
    C. < 30%
    D. > 50%

SECTION 14

ALLERGY

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ANSWERS

1.**(D)**

2.**(D)**

3.**(D)**

TABLE 77.1 Gell and Coombs Classification of Hypersensitivity Disorders

TYPE	INTERVAL BETWEEN EXPOSURE AND REACTION	EFFECTOR MOLECULE	TARGET OR ANTIGEN	EXAMPLES OF MEDIATORS	EXAMPLES
I Immediate Late phase	<30 min 2–12 hr	IgE	Pollens, food, venom, drugs	Histamine, tryptase, leukotrienes, prostaglandins, platelet-activating factor	Anaphylaxis, urticaria, allergic rhinitis, allergic asthma
II Cytotoxic antibody	Variable (min–hr)	IgM, IgG, IgA	Red blood cells, platelets	Complement	Hemolytic anemia, thrombocytopenia, Goodpasture syndrome
III Immune complex	1–3 wk after drug exposure	Antigen-antibody complexes	Blood vessels, liver, spleen, kidney, lung	Complement, anaphylatoxin	Serum sickness, hypersensitivity pneumonitis
IV Delayed type	2–7 days after drug exposure	Lymphocytes	*Mycobacterium tuberculosis*, chemicals	Cytokines (IFN-γ, TNFα, GM-CSF)	TB skin test reactions, contact dermatitis, graft-vs-host disease

GM-CSF, Granulocyte-macrophage colony-stimulating factor; *IFN-γ*, interferon-γ; *TB*, tuberculosis; *TNFα*, tumor necrosis factor-α.

4.**(A)** There are two methods for identifying allergen-specific IgE: In vivo skin testing and in vitro serum testing. Properly performed skin tests are the most sensitive method for detecting the presence of allergen-specific IgE. In vitro serum testing, such as immunoassays like the enzyme-linked immunosorbent assay (ELISA), measures levels of antigen-specific IgE. Many allergists and laboratories regard the Immuno CAP System (Thermo Fisher Scientific, Waltham, MA) as the method of choice. This method uses a solid phase and shows higher sensitivity, specificity, and reproducibility. The assay uses a quantitative fluorescent immunoassay (FEIA) that is more sensitive than other methods.

5.**(A)** The broadest experience with managing adverse drug reactions is with penicillin. Penicillin allergy should be evaluated when the individual is well and not in acute need of treatment. Penicillin skin testing is helpful for IgE-mediated reactions because of its negative predictive value; only 1–3% of patients with negative skin tests have a reaction, which is mild, when re-exposed to penicillin. Skin testing for penicillin should be performed using the major determinant, penicilloyl polylysine (available as Pre-Pen [ALK-Abelló, Inc, Hørsholm, Denmark]), and minor determinants, which include penicillin G, penicilloate, and penilloate. Skin testing to penicillin does not predict non–IgE-mediated reactions. For patients with a history consistent with serum sickness or desquamative-type reactions, skin testing should not be performed, and penicillin should be avoided indefinitely.

6.**(D)** Nonallergic, noninfectious rhinitis (formerly known as vasomotor rhinitis) can manifest as rhinorrhea and sneezing in children with profuse clear nasal discharge. Exposure to irritants, such as cigarette smoke and dust, and strong fumes and odors, such as perfumes and chlorine in swimming pools, can trigger these nasal symptoms.

7.**(B)** Antihistamines are the medications used most frequently to treat allergic rhinitis. They are useful in treating rhinorrhea, sneezing, nasal itching, and ocular itching but are less helpful in treating nasal congestion.

8.**(C)**

TABLE **77.4** Determination of Allergen-Specific IgF by Skin Testing vs In Vitro Testing

VARIABLE	SKIN TEST*	sIgE ASSAY
Risk of allergic reaction	Yes (especially ID)	No
Relative sensitivity	High	High
Affected by antihistamines	Yes	No
Affected by corticosteroids	Usually not	No
Affected by extensive dermatitis or dermographism	Yes	No
Broad selection of antigens	Fewer	Yes
Immediate results	Yes	No
Expensive	No	Yes
Lability of allergens	Yes	No
Results evident to patient	Yes	No

**Skin testing may be the prick test or intradermal (ID) injection.*
From Bunyavanich S, Kattan J, Sicherer SH. Diagnosis of allergic disease. In Kliegman RM, St Geme J, eds. Nelson Textbook of Pediatrics. *21st ed. Philadelphia: Elsevier; 2020 [table 167.3].*

9.**(B)** Egg allergy is the most common cause of food-induced eczematous reactions.

10.**(A)** Defective cell-mediated immunity leads to increased susceptibility to many bacterial, viral, and fungal infections of the skin. More than 90% of patients with atopic dermatitis have colonization of lesioned skin with Staphylococcus aureus (S. aureus), and more than 75% of patients have colonization of uninvolved skin. Colonization and infection by S. aureus are associated with disease severity. S. aureus secretes exotoxins that act as superantigens, stimulating T cells and increasing IgE production.

11.**(D)** The clinical manifestations of atopic dermatitis vary with age. In infants, atopic dermatitis involves the face, scalp, cheeks, and extensor surfaces of the extremities. The

diaper area is spared. In older children, the rash localizes to the antecubital and popliteal fossae, head, and neck. In adolescents and adults, lichenified plaques are seen in flexural areas and head and neck regions.

12.**(B)** Many conditions share signs and symptoms of atopic dermatitis. Infants presenting in the first year of life with failure to thrive, recurrent skin or systemic infections, and scaling, erythematous rash should be evaluated for immunodeficiency disorders. Wiskott-Aldrich syndrome is an X-linked recessive syndrome characterized by atopic dermatitis, thrombocytopenia, small-sized platelets, and recurrent infections. Langerhans cell histiocytosis is characterized by hemorrhagic or petechial lesions.

13.**(D)** Atopic dermatitis is a chronic, relapsing skin disorder that tends to be more severe and prominent in young children. Symptoms become less severe in two thirds of children, with complete remission for approximately 20%.

14.**(A)** Patients with HAE rarely have urticaria associated with angioedema, and the swelling is not relieved with antihistamines or oral corticosteroids. Most patients (85%) have type I disease, which is due to decreased production of C1-esterase inhibitor. A minority of patients (15%) have type II disease, which is due to production of dysfunctional C1-esterase inhibitor

TABLE **81.3** Complement Evaluation of Patients with Recurrent Angioedema

ASSAY	IDIOPATHIC ANGIOEDEMA	TYPE I HEREDITARY ANGIOEDEMA	TYPE II HEREDITARY ANGIOEDEMA	HEREDITARY ANGIOEDEMA WITH NORMAL C1 INHIBITOR	ACQUIRED C1-ESTERASE INHIBITOR DEFICIENCY	VASCULITIS
C4	Normal	Low	Low	Normal	Low	Low or normal
C1-esterase inhibitor level	Normal	Low	Normal	Normal	Low	Normal
C1-esterase inhibitor function	Normal	Low	Low	Normal	Low	Normal
C1q	Normal	Normal	Normal	Normal	Low	Low or normal

From Zuraw BL. Urticaria and angioedema. In: Leung DYM, Szefler SJ, Bonilla FA, et al., eds. Pediatric Allergy: Principles and Practice. *3rd ed. Philadelphia: Elsevier; 2016:463.*

15.**(D)** In the pediatric population, viral illnesses are responsible for the majority of acute urticaria.

16.**(B)** Measurement of the mast cell mediators, histamine and tryptase, may be helpful when the diagnosis of anaphylaxis is in question. A tryptase level is a more useful test because histamine is released quickly, has a very short half-life, and is often difficult to detect in the serum. Serum tryptase levels peak 1–1.5hours after anaphylaxis. Elevated levels may be helpful in establishing the diagnosis, but normal tryptase levels do not rule out the diagnosis. It is best to measure a serum tryptase level 1–2 hours after the onset of symptoms. It also can be measured retrospectively on stored serum that is less than 2 days old.

17.**(B)** The diagnosis of urticaria and angioedema is straight forward; finding the etiology may be more difficult. Other dermatologic conditions can mimic urticaria. Erythema

multiforme has target-shaped, erythematous, macular, or papular lesions that may look similar to urticaria, but the lesions are fixed and last for several days. Other dermatologic diseases such as dermatitis herpetiformis and bullous pemphigoid are quite pruritic, and early on, the lesions may resemble urticaria. Mastocytosis is characterized by mast cell infiltration of various organs, including the skin. Some patients have skin lesions similar in appearance to urticaria rather than the classic urticaria pigmentosa. Urticaria pigmentosa appears as hyperpigmented, red-brown macules that may coalesce. When these lesions are stroked, they urticate, which is called the Darier sign. Muckle-Wells syndrome. It is an autosomal dominant autoinflammatory disorder characterized by episodic urticaria presenting in infancy with sensorineural deafness, amyloidosis, arthralgias, and skeletal abnormalities. Also rare, Schnitzler syndrome is characterized by chronic urticaria, macroglobulinemia, bone pain, anemia, fever, fatigue, and weight loss. Urticarial vasculitis is a small vessel vasculitis with histologic features of a leukocytoclastic response. The main distinguishing feature is that the lesions last longer than 24hours, may be tender, and leave behind skin pigmentation. Skin biopsy is required for definitive diagnosis.

18.**(A)** The most common physical urticaria is dermatographism, affecting 2–5% of persons. It is characterized by an urticarial reaction localized to the site of skin trauma. It has been suggested that trauma induces an IgE-mediated reaction causing histamine to be released from the mast cells.

19.**(B)** Large local reactions to insect stings are more common in children, with an estimated incidence of 20% for children and 10% for adults. Occasionally, large local reactions may be mistaken for cellulitis. Infection is unlikely if the reaction occurs within 24–48 hours after a sting. Treatment is with an oral corticosteroid for 4–5 days rather than oral antibiotics

20.**(D)** For cow's milk–allergic patients who have multiple food allergies, asthma, and allergic rhinitis, 21% of patients have persistent milk allergy at age 16 years; 85% of children who do not have additional atopic disease develop cow's milk tolerance by 3 years of age. Similarly, 66% of children with egg allergy but no other atopic diseases develop tolerance by 5 years of age; 32% of those with additional atopic disease will still be sensitive to egg by age 16 years. The natural histories of soy and wheat allergy are similar: oral tolerance in 25–29% by age 4 years, 45–56% by age 6 or 8 years, and 65–69% by age 10 or 12 years. Twenty percent of children with peanut allergy outgrow their allergy. Nine percent of tree nut allergic patients will develop oral tolerance. Food allergies to seafood continue to be a lifelong affliction.

21. **(D)** Patients with dermatitis herpetiformis had intensely pruritic vesicular rash on extensor surfaces and buttocks. Biopsy diagnostic, shows IgA granule deposits at the dermal epidermal junction; resolves with dietary gluten avoidance

22.**(D)**

TABLE **78.4** List of U.S. Food and Drug Administration–Approved Biologic Medications for the Treatment of Asthma in Children and Their Characteristics

NAME	INDICATION	MECHANISM OF ACTION	DOSE	MOST COMMON SIDE EFFECTS	ADMINISTRATION CONSIDERATIONS	OTHER
Benralizumab (Fasenra)	• Add-on controller therapy for severe eosinophilic asthma >12yr of age	• IL-5 receptor (at the α-subunit) blocker • Induces apoptosis of eosinophils through ADCC*	• 30mg SC every 4wk x 3 • Then every 8wk	• Injection site reactions • Headache • Pharyngitis	• Treat preexisting helminth infections • 30-min observation from first dose	• Improved efficacy if AEC‡ >150 cells/μL
Dupilumab (Dupixent)	• Add-on controller therapy for moderate to severe persistent asthma eosinophilic phenotype or oral corticosteroid dependent >12yr of age	• IL-4 receptor blocker (at α subunit) preventing IL-4 and IL-13 signaling	• 400mg SC once then 200mg SC every 2wk OR • 600mg SC once then 300mg every 2wk • With concomitant moderate to severe atopic dermatitis, 600mg once then 300mg every 2wk	• Injection site reactions • Eosinophilia • Oropharyngeal pain	• Avoid live vaccines • Treat preexisting helminth infections • Option for self-administration	• Improved efficacy if AEC >150 cells/μL
Mepolizumab (Nucala)	• Add-on controller therapy for severe persistent asthma eosinophilic phenotype >6yr of age	• IL5 antagonist preventing binding of IL-5 receptor on eosinophils	• Patients >12yr of age: 100mg SC every 4wk • Patients 6–11yr of age SC 40mg every 4wk	• Injection site reactions • Herpes zoster • Headache • Back pain • Fatigue	• Zoster vaccination • Treat preexisting helminth infections • 30-min observation from first dose	• AEC >150 in last 6wk OR • AEC 300 in last 12 months
Omalizumab (Xolair)	• Moderate to severe persistent asthma aged >6yr of age with sensitivity to perennial aeroallergens	• Binds free IgE preventing IgE binding to high affinity IgE receptor on mast cells and basophils minimizing release of chemical mediators involved in allergic inflammatory cascade	• Dependent on age, weight, and total IgE level • Administered SC every 2–4 wk	• Anaphylaxis • Injection site reactions • Arthralgia	• Patients should have autoinjectable epinephrine at time of injection • 90-min wait after first 3 injections, then 30-min wait with subsequent doses • Equipment available to treat anaphylaxis	• Total IgE >30ku/L AND • Positive skin prick test or serum-specific IgE to perennial aeroallergen

*Antibody-dependent cell-mediated cytotoxicity.
AEC, Absolute blood eosinophil count; IL, interleukin; SC, subcutaneous.

23.**(B)** To determine whether a child is having more persistent asthma, using the Rules of Two is helpful: daytime symptoms occurring two or more times per week or nocturnal awakenings two or more times per month implies a need for daily anti-inflammatory medication. Inhaled corticosteroids are the preferred initial long-term control therapy for children of all ages.

24.**(A)** Daily long-term control therapy is recommended for infants and young children 0–4 years of age that had four or more episodes of wheezing in the previous year that lasted more than 1 day, affected sleep, and who have a positive asthma predictive index.

TABLE **78.5** Modified Asthma Predictive Index for Children

At least four wheezing episodes *plus*:	
One major criteria	Or two minor criteria
Parental asthma	Allergic rhinitis
Eczema	Wheezing apart from colds
Inhalant allergen sensitization	Eosinophils ≥4%
	Food allergen sensitization

From Liu AH, Martinez FD. Natural history of allergic diseases and asthma. In: Leung DYM, Szefler SJ, Bonilla FA, et al., eds. Pediatric Allergy: Principles and Practice. 3rd ed. Philadelphia: Elsevier; 2016 (fig. 2.4).

25.**(C)** for some children, symptoms of wheezing with respiratory infections subside in the preschool years, whereas others have more persistent asthma symptoms. Prognostic indicators for children younger than 3 years of age who are at risk for persistent asthma are known as the Modified Asthma Predictive Index for children. Atopy is the strongest predictor for wheezing continuing into persistent asthma.

26.**(D)** Children older than 5 years of age can usually perform spirometry maneuvers. Peak expiratory flow offers a simple, inexpensive way to measure asthma control at home; variability in predicted peak flow reference values make spirometry preferred over peak flow measures in the diagnosis of asthma. For younger children who cannot perform spirometry maneuvers or peak flow, a therapeutic trial of controller medications aids in the diagnosis of asthma.

27.**(C)** A chest x-ray should be performed with the first episode of asthma or with recurrent episodes of undiagnosed cough or wheeze to exclude anatomic abnormalities. Repeat chest x-rays are not needed with new episodes unless there is fever (suggesting pneumonia) or localized findings on physical examination.

28.**(A)** Asthma is the most common chronic disease of childhood in industrialized countries, affecting over 5 million children younger than 18 years of age in the United States (7.5% of children). In adults, females are more likely than males to have asthma, but in children the reverse is true.

29.**(B)** In asthmatic patients, measurement of a biomarker, exhaled nitric oxide (FeNO), reflects eosinophilic airway inflammation and provides information on those patients who may better respond to corticosteroid treatment, aids in monitoring response to therapy and dose optimization, and detects non adherence to corticosteroid therapy.

30.**(A)** Chest x-ray should be performed with the first episode of asthma or with recurrent episodes of undiagnosed cough or wheeze to exclude anatomic abnormalities. Repeat chest x-rays are not needed with new episodes unless there is fever (suggesting pneumonia) or localized findings on physical examination.

31.**(C)** Rinsing the mouth after inhalation and using large volume spacers help lessen the local adverse effects of dysphonia and candidiasis and decrease systemic absorption from the gastrointestinal tract.

32.**(D)** Vilanterol, an ultralong-acting β2 agonist, is approved in patients aged 18 years and older and has the advantage of once-daily dosing.

33.**(C)** Two classes of leukotriene modifiers include cysteinyl leukotriene receptor antagonists (zafirlukast and montelukast) and leukotriene synthesis inhibitors (zileuton).

34.**(A)** Montelukast is dosed once daily at night as 4-mg granules or chewable tablets (for children 6 months to 5 years of age), 5-mg chewable tablets (6–14 years of age), and 10-mg tablets (≥15 years of age).

35.**(D)** The most common adverse effects include local irritation, burning, and sneezing, which occur in 10% of patients. Nasal bleeding from improper technique (spraying the nasal septum) can occur. Rare cases of nasal septal perforation have been reported.

36.**(A)** First-generation antihistamines, such as diphenhydramine and hydroxyzine, easily cross the blood-brain barrier with sedation as the most common reported adverse effect. Second-generation antihistamines such as cetirizine, desloratadine, fexofenadine, levocetirizine, and loratadine, are less likely to cross the blood-brain barrier resulting in less sedation.

37.**(D)** A, B and C are examples of immunological adverse drug reaction while D is example of unpredicted non- immunological adverse drug reaction.

38.**(D)** A causal agent is more likely to be found in acute urticaria such as a food, medication, insect sting, infection, blood transfusion, or contact agent. With chronic urticaria, an etiology is less likely to be found, and most cases are idiopathic.

39.**(C)** Serum sickness is the prototypical type III reaction where antibody binds to antigen forming immune complexes that can cause vascular injury or end-organ damage. Symptoms classically are fever, arthralgias, lymphadenopathy, and rash.

40.**(D)** Anaphylaxis is mediated by IgE, whereas anaphylactoid reactions result from mechanisms that are due to nonimmunologic mechanisms. Both reactions are acute, severe, and can be life threatening due to massive release of inflammatory mediators.
Latex allergen immunotherapy and challenge tests Biologic agent cause IgE-mediated anaphylaxis while radiocontrast media causes anaphylactoid reaction.

41.**(A)** The risk of having an allergic reaction to a cephalosporin in a child who has reacted positively to penicillin skin testing is less than 2%. It is believed that the first-generation cephalosporins are more likely than second-generation or third generation cephalosporins to be cross reactive. This is due to the chemical similarity of side chains of the β-lactam ring between penicillin and first-generation cephalosporins.

SECTION 15

RHEUMATIC DISEASES OF CHILDHOOD

GHASSAN SAMI

QUESTIONS

1. You are leading a group session for medical college students. One of the students is presenting a case of an 11-month-old infant presented with fever, rash, non-purulent conjunctivitis, hands and foot swelling, and perineal scaling. You ask the student to describe the nature of fever.
The pattern of fever that **BEST** fits this infant's illness is

A. sustained
B. relapsing
C. hectic
D. regularly patterned

2. Rheumatological diseases may be presented with symptoms of localized inflammation that vary depending on the involved sites like myositis (inflammation of the muscle) or vasculitis (inflammation of blood vessels).
In this regard, the term "Enthesitis" means the inflammation of

A. lining of pleura, pericardium, or peritoneum
B. synovium
C. buccal mucosa
D. insertion of ligament to a bone

3. You are seeing a 4-year-old boy brought by his mother with a complaint of swollen knees with pain of two days duration. His condition is associated with purpuric rash that extends from his legs up to his buttocks. He also complains of some abdominal pain with tenderness. His vitals are normal and his initial laboratory investigations including CBC and serum electrolytes revealed nothing significant but his urinalysis shows moderate RBCs and 3+ protein. The mother is wondering what could be the cause behind her son's illness.
The answer for the mother's question is that

A. it is of unknown cause
B. it is most often a complication of "strep" infection
C. it is associated with viral illness
D. it is a sex chromosome linked disease

4. During the physical examination of an 8-year-old male child, you found palpable rash extending over his arms, legs and buttocks; the trunk is spared. The rash is not fading when applying pressure on it. There are few ecchymotic areas and there is scrotal swelling.
Of the following, the manifestation that is **MOST** encountered in this patient is

A. arthritis
B. gastrointestinal involvement
C. orchitis
D. renal involvement

5. A 13-year-old boy is brought to your clinic with complaint of intermittent abdominal pain for 2 days. He also notes that faint rash had developed on his legs over the last 24 hours. He has a history of a flu-like illness a few weeks ago. On examination, he is afebrile, and he has mild abdominal tenderness. There are few palpable, non-blanching lesions visible on his thighs bilaterally.
Of the following, the **MOST** important laboratory test to be included in your initial workup is

A. CRP
B. platelet count
C. urinalysis
D. abdominal ultrasound

6. You are explaining to a resident working with you in the emergency department about how to make the diagnosis of Henoch-Schönlein purpura (IgAV) for a child presented to ER with history of purpuric rash, abdominal pain and arthralgia. You clarify to her that the diagnosis of IgAV is based on the presence of two of four criteria, which provides high sensitivity and specificity for the disease.
Of the following, the definition which is considered a true criterion to be included in the diagnosis of IgAV is

A. microscopic hematuria
B. diffuse abdominal pain
C. lower limbs joint pain or inflammation
D. age <5 yr at onset of symptoms

7. You are following a patient who was diagnosed as a case of Henoch-Schönlein purpura 3 weeks ago. Prednisolone was prescribed for him for the complaint of severe abdominal pain. He is now in his second week of a 4-week-tapering plan of steroids. His mother reported that he was doing well until recently when he begins to complain of recurrent abdominal pain. The patient denied any change in bowel habits, stool or urine color and there is no vomiting. On examination, the patient looks well, alert, and when asked to localize the pain, he passed his hand over the whole abdomen which was mildly tender to touch.
The **BEST** action for the management of this patient's complaint is to

A. stop steroid immediately and switch to non-steroidal anti-inflammatory drugs
B. send for ultrasonic abdominal examination to exclude intussusception
C. add H2-receptors antagonist or proton pump inhibitor to his treatment
D. extend the course of steroid for another 2 weeks

8. You are counseling the parents of a 5-year-old boy with IgA vasculitis who presented with skin rash and arthritis for the last 1 week. There was no abdominal or renal involvement. He was kept on symptomatic treatment for the joint pain with nonsteroidal anti-inflammatory medications.
Which of the following is **MOST** appropriate regarding the course of his illness?

A. The rash can wax and wane for 1 year after the initial episode
B. The arthritis may leave permanent joint damage
C. Renal failure is a common complication
D. Long-term renal complications are unrelated to the degree of renal involvement initially

9. Upon discharge of an infant with Kawasaki disease with normal initial echocardiographic assessment, the second one is advised after

A. 2 weeks
B. 4 weeks
C. 6 weeks
D. 12 weeks

10. The clinical features of Kawasaki disease include cervical lymphadenopathy. For the lymphadenopathy to meet the epidemiologic case definition as a clinical criterion for the diagnosis of Kawasaki disease, it should be

A. multiple
B. tender
C. bilateral
D. more than 1.5 cm in diameter

11. You are evaluating a 7-year-old child presents with history of high-grade fever of 6 days duration associated with non-purulent conjunctivitis, strawberry tongue, maculopapular rash on the trunk and extremities and abdominal pain. Laboratory tests results show:
WBC 23.0 $\times 10^9$/l, Neutrophils 89%, Lymphocytes 9%, Hb 10.2 g/dl, Platelet count 70 $\times 10^9$/l, CRP 200 mg/l, Urinalysis: RBC 3-4, pus 1+/HPF, SARS-CoV-2 IgG positive, PCR for SARS-CoV-2 negative
Of the following, the **MOST** likely diagnosis is

A. scarlet fever
B. Adenovirus infection
C. multisystem inflammatory disease–children
D. Kawasaki disease

12. What is the **MOST** common chronic rheumatologic disease in childhood?

A. Henoch-Schönlein purpura
B. Juvenile idiopathic arthritis
C. Rheumatic fever
D. Systemic lupus erythematosus

13. In which of the following groups of juvenile arthritis, males are affected more than females?

A. Oligoarticular juvenile idiopathic arthritis
B. Polyarticular juvenile idiopathic arthritis
C. Systemic juvenile idiopathic arthritis
D. Enthesitis related arthritis

14. A 4-year-old female child was referred to you by a general practitioner for the complaint of recurrent leg pain for the last 7 months. The parents have sought medical advice from a number of doctors who mostly attributed the cause of pain to trauma or considered it as growing pain. Pain killers were prescribed in many occasions that showed temporary response. The girl has never complained of fever, skin rash or loss of weight. On examination, she looks well, not pale, body temperature is 37.2°C. There is bilateral palpable, non-tender, movable submandibular lymph nodes, with a largest diameter of 1

cm. While examining for lymph nodes, you noticed that there is tenderness over her left temporomandibular joint. Her knees and left ankle joint are swollen, and tender.
Of the following, the **MOST** likely diagnosis is

A. rheumatic fever
B. systemic lupus erythematosus
C. juvenile idiopathic arthritis
D. acute leukemia

15. A concerned colleague is consulting you about the risk of her niece to develop uveitis. The young girl is 5-year-old, who has been recently diagnosed as a case of juvenile idiopathic arthritis (JIA) and is scheduled for ophthalmological screening.
You explain to your colleague that the risk of uveitis is **HIGHEST** when the girl has

A. oligoarticular JIA with positive antinuclear antibodies
B. polyarticular JIA with positive antinuclear antibodies
C. polyarticular JIA with negative antinuclear antibodies
D. oligoarticular JIA with negative antinuclear antibodies

16. A 10-year-old girl was brought to the hospital by her parents. The girl was enjoying good health until about 6 months ago when she started to have “growing pains”, feeling stiff and uncomfortable in the morning along with periodic ache and pain in her knees, and hands. After few months of this, the parents noticed that her knees and wrists were often swollen and warm, and she was sometimes limping. She could not open a bottle of water, was having difficulty sitting on the floor, and was often unable to fully participate in physical activities at school.
On physical examination, she looks pale and frail, her weight is 20 kg (5^{th} percentile), height is 122cm (25^{th} percentile), temperature is 38°C, had swelling in both her knees, right ankle, and both wrists. Other parts of examination were normal. Her laboratory investigations are shown: Hb 9.2 g/L, WBC 12.2 $\times 10^9$/L, Platelet 430 $\times 10^9$/L, Blood film: normochromic, normocytic anemia, Rheumatoid factor negative, ESR 66 mm/hr, CRP 28 mg/L, X-rays showed no signs of fracture or erosions.
Of the following, the **MOST** likely diagnosis is

A. oligoarticular juvenile idiopathic arthritis
B. polyarticular juvenile idiopathic arthritis
C. systemic juvenile idiopathic arthritis
D. early-onset adult rheumatoid arthritis

16. Making comparison between the group of Enthesitis-Related Arthritides, which of the following has more male gender predominance?

A. juvenile ankylosing spondylitis
B. psoriatic arthritis
C. reactive arthritis
D. arthritis of inflammatory bowel disease

17. A 14-year-old boy presented to a rheumatology clinic with persistent arthritis involving the knees, ankles, wrists, first metatarsophalangeal joints of both feet, and severe pain in both feet at the Achilles tendon attachments to the calcaneus. He also complains of back pain in lumbar area and sacroiliac joint pain that alternated between the right and left sides.

He had early-morning stiffness but he usually felt better during the day. During the previous year he had been treated with diclofenac and naproxen with no much improvement.
The patient's symptoms had started at the age of 8 years with oligoarthritis involving the knees, and left ankle; 8 months later, he started to complain of recurrent episodes of arthritis and heel pain for several months, leaving him unable to attend school most days. He was treated successfully with NSAIDs, antibiotics and intra-articular glucocorticoid injection. He became asymptomatic between the ages of 11 and 13 years, and he did not require any treatment at that time. The patient is HLA-B27-positive.
Of the following, the **BEST** treatment for the patient's current illness is

A. sulfasalazine
B. methotrexate
C. prednisolone
D. infliximab

18. You are seeing a 4-year-old female who has been limping with swelling of her right knee for 2 weeks. Her parents note that she cannot fully extend her right knee. She sometimes does not want to walk especially in the morning. Her past medical history is unremarkable. Physical examination demonstrates no fever, and swelling of the right knee. There is no increased heat or pain upon touching the joint. She appears unconcerned about her limp and swelling.
Labs results:
WBC; 10,600 × 10^9/L with 43% neutrophils, 50% lymphocytes, 8% monocytes.
Hb; 11g/dL
Platelets; 322 × 10^9/L
CRP; 20mg/L
Urinalysis; normal
Rheumatoid factor; negative
ANA; 1:40
X-ray of bone; normal
Arthrocentesis was performed and the results of synovial fluid analysis showed WBC 23,000 cell/ml, neutrophil 70%, lymphocytes 30%, negative gram stain, and negative culture.
Of the following, the **MOST** likely diagnosis is

A. systemic lupus erythematosus
B. septic arthritis
C. juvenile idiopathic arthritis
D. reactive arthritis

19. You are following up a 9-year-old girl who was diagnosed as case of polyarticular juvenile idiopathic arthritis before 2 months. The girl was well until about 8 months ago when she started to have pain in her knees, ankles and hands which gradually progress to become a confining arthritis. You initiated treatment with ibuprofen without much improvement so you decided later to add naproxen.
Today, she is still complaining of pain in her knees and ankles that limits her daily activity and prevents her from sharing physical activity with other peers.
Of the following, the **NEXT** drug of choice that should be used for this girl's poorly responding arthritis is

A. prednisolone

B. methotrexate
C. infliximab
D. tocilizumab

20. What is the preferred route for administration of methotrexate in patients with juvenile idiopathic arthritis?
A. orally
B. subcutaneously
C. intramuscularly
D. intravenously

21. A 10-year-old female, who is a known case of systemic onset juvenile rheumatoid arthritis, was doing well for the past 2 years on both naproxen and methotrexate.
During the last 3 weeks, she developed high spiking fever, associated with pallor, gum bleeding, petechiae, joint pain and swelling, cervical lymphadenopathy, and hepatosplenomegaly. Her laboratory results are as follows:
CBC: Hb 8.4g/dL, WBC 9.300 x 10^9/L, platelet 85,000 x 10^9/L
Liver Function test: AST 80 IU/L, ALT 65 IU/L, Alkaline Phosphatase 286 IU/L
Serum ferritin >3000 ng/mL
PT 16.5 sec, PTT 26 sec, INR 1.31, fibrinogen 182 mg/dL (normal 230-510 mg/dL)
Of the following, the **MOST** likely explanation for her condition is
A. flare of her disease
B. adverse effect of methotrexate
C. macrophage activation syndrome
D. autoimmune hepatitis

22. During a clinical session with students of medical school, you are discussing a case of a 6-year-old male who has been diagnosed as a case of SLE. A wondering student asks if SLE in children is similarly prevalent in females as it is in adults.
Of the following, the **BEST** answer for this inquiry is
A. Yes, in children, SLE similarly affects females more
B. No, there is an equal gender distribution of SLE in children
C. No, in children, SLE affects primarily males
D. Actually, it is an extremely rare disease in children and no enough researches are available to determine the gender distribution

23. Of the following clinical manifestation of systemic lupus erythematosus, the one that can cause permanent local damage is
A. malar rash
B. photo sensitivity
C. discoid lupus
D. arthritis

24. Which of the following groups' lymphadenopathy is particularly sensitive indicator of the systemic lupus erythematosus activity?
A. Anterior cervical
B. Posterior cervical

C. Axillary
D. Epitrochlear

25. A 15-year-old adolescent girl presented to the hospital complaining of fever, progressive shortness of breath, and chest pain over the past month. Additional complaints included, oral ulcers, and bilateral knee joints pain. Her past medical history is insignificant.
Physical examination shows a raised erythematous rash on the cheeks, and leg edema. Her initial laboratory values were notable for hemoglobin of 9.0 g/dL, and platelet count 100,000/µL, with 3+ protein and 3+ hemoglobin on urinalysis.
What is the **MOST** likely diagnosis?

A. Systemic lupus erythematosus
B. Systemic Juvenile Idiopathic Arthritis
C. Behçet's disease
D. Dermatomyositis

26. A 14 years old male, was presented to your practice complaining of significant fatigue and malaise, low-grade fever, swelling and painful movement of both hands and fingers, and weight loss over the past 3 months. On physical examination, the patient looks ill, pale, not jaundiced, there is no rash, but he has 2 coin-sized thick scaly, red patches of hair loss on the scalp. There is bilateral leg edema. His initial laboratory tests are shown
Hb 8.7 gm/dL
WBC 3.4 x 10^9/L, N 56%, L 39%
Platelet 90 x 10^9/L
Urinalysis proteinuria 3+, RBC cast 2+, granular cast 2+
The type of antibodies **MOSTLY** to be elicited in this patient are

A. Anti-double-stranded DNA and anti-smith
B. Anti-nuclear antibody and myositis-specific antibodies
C. Anti-phospholipids antibody, anti-Ro and anti-La
D. Anti-nuclear antibody and Anti-double-stranded DNA

27. During the clinical case conference in your hospital, you are reviewing the laboratory results of a patient who have just been diagnosed as a case of systemic lupus erythematosus. Some of the results are shown
Hb 9.1 gm/dL
WBC 4,000 x 10^9/L
Platelet 111 x 10^9/L
Total serum protein 7.9 g/dL, albumin 3.4 g/dL (3.5-5 g/dL), globulin 4.5 g/dL (2-3.5 g/dL)
TSB 1.2 mg/dL, ALT 45 U/L, AST 50 U/L
Urinalysis proteinuria 1+, RBC 3+
One of your colleagues asks about the best explanation for elevated globulin fraction in this patient. The answer is:

A. It reflects the onset of liver involvement
B. It is due to excessive antibody production
C. A compensatory mechanism for the lost albumin in renal disease
D. Rise in the total globulin is caused by increase serum level of free transferrin

28. A 13-year-old boy was admitted to the hospital for daily fever with the highest temperature of 39.2°C over the last 10 days. This boy also manifested rash, non-exudative conjunctivitis, and knee and ankle arthralgia, without chills, cough, vomiting, abdominal pain, diarrhea, or convulsion. Physical examination was notable for tachycardia (heart rate 157 beats/min), erythema on his face and hepatosplenomegaly. The initial lab tests showed: WBC 3.8 × 10^9/L, neutrophil% 77.7%, hemoglobin 9.4 g/dL, platelet 322 × 10^9/L; CRP 82 mg/L; ESR 129 mm/h. The results of liver function, renal function, coagulation function, and urinalysis, were all normal. Radiographs of the knees, ankles and hands were normal. The result of echocardiogram implicated coronary artery dilation and aortic regurgitation.
Further laboratory tests showed: Coomb's test (+), decreased C3 and C4, ANA (+), ds-DNA (+).
What is the **BEST** initial treatment for this patient?

A. Methylprednisolone
B. Intravenous immunoglobulin
C. Nonsteroidal antiinflammatory drugs
D. Methotrexate

29. A 15-year-old adolescent girl presented to the hospital complaining of fatigue, difficult breathing, malar rash, mouth ulceration and bilateral leg edema. She is a known case of SLE since she was 10, and she was treated initially with hydroxychloroquine and prednisolone for a period of 2 years. Subsequently, she was maintained only on hydroxychloroquine. In the hospital, she was found to have proteinuria. Her eGFR was 69 mL/min/1.73m^2. Inflammatory markers were only slightly elevated but there was evidence of SLE serological activity (high anti-dsDNA; low C3). She underwent a kidney biopsy which confirmed the lupus nephritis diagnosis.
She received 3 days of pulse methylprednisolone, followed by oral prednisone daily. She was also started on monthly intravenous cyclophosphamide. After 6 months of therapy, her condition did not improve significantly.
What is the **BEST** next therapeutic choice?

A. Renal transplant
B. Methotrexate
C. Mycophenolate mofetil
D. Azathioprine

30. You are attending a rheumatology clinic as part of your clinical attachment training program. An adolescent girl has just entered the room accompanied by her mother. You noticed that the girl has a bilateral blue-violet discoloration surrounding her eyes with slight periorbital edema. The consultant rheumatologist is explaining that this patient is a newly diagnosed case of juvenile dermatomyositis and that this discoloration is one of the characteristic skin manifestations of the disease.
The term that **BEST** describes this girl's skin manifestation is

A. photosensitivity
B. Gottron papules
C. Shawl sign
D. heliotrope rash

31. A previously well 8–year–old girl presented to the hospital with an 8-week history of recurrent fever, followed by a skin rash, and weakness of all limbs. The fever was of low grade and was treated with antibiotics and antipyretics without improvement. The rash was maculopapular and itchy and started on the upper extremities (predominantly on the knuckles) and progressed to involve the thighs. She developed a gradual and symmetrical limb weakness. She was unable to comb her hair, climb stairs, or get up from squatting position. Weakness of limbs was accompanied by muscle and joint pain. There was a reported nasal tone to her voice in the course of illness, but no history of dysphagia.
On physical examination she was afebrile, had scaly, red macules located on both knees, some coalesced to form patches on the knuckles. There was no joint swelling. Her muscle bulk was normal but she had generalized tenderness in all muscles and a reduced power of the upper and lower limbs with a positive Gowers' sign. Laboratory investigations revealed elevated serum levels of creatine kinase, LDH, AST, CRP, and ESR.
Which of the following is **TRUE** regarding this girl's illness?

A. The muscle weakness primarily affects the distal muscles
B. Wearing sun block and refrain from prolonged sun exposure prevent exacerbation of muscle disease
C. Vitamin D and calcium supplements should be avoided because of the high risk of calcium deposition in subcutaneous tissue
D. The most serious complication is cardiomyopathy

32. A mother brings her 3-year-old son to your clinic with the complaint of leg pain that occurred frequently during the last few weeks. The mother describes recurring incidents of excessive crying due to leg pain occur mainly during the evening and sometimes awaken him from sleep. She managed to relief the pain with leg massage but she is concerned that he might need medical attention. The child has no history of trauma, fever, leg swelling, or rash. On examination, he looks well, afebrile, not pale, has no lymphadenopathy, nor organomegaly, and the examination of his legs revealed nothing apart from few bruises over his shin.
Of the following, the investigation that would reasonably be ordered for this child is

A. complete blood count
B. X-ray of the legs
C. serum calcium
D. serum zinc

33. During a well-child visit for a 10-year-old boy, you noticed that he has flat foot. The child denies any history of recurrent leg or back pain.
What is the **BEST** management for this child?

A. Refer him to orthopedician for surgical treatment
B. Medial arch supports are enough to prevent complications
C. No treatment is required for the time being
D. Swapping shoes may correct the defect

SECTION 15

RHEUMATIC DISEASES OF CHILDHOOD

GHASSAN SAMI

ANSWERS

1.**(C)** Fever generally classified into the following:

- Intermittant (fluctuating between normal temperatures and fever levels over the course of the day)
- Remittant (temperature fluctuates, but though it falls, it never falls back to normal)
  In these two patterns the amplitude of temperature change is more than 0.3°C and less than 1.4°C.
- Hectic (either intermittant or remittant when the difference between peak and trough temperature is 1.4°C or more)
- Continous is a pattern in which there is little change (0.3°C or less) in the elevated temperature during a 24-hour period
- Relapsing (a variant of the intermittent pattern, fever spikes are separated by days or weeks of intervening normal temperature.)

A hectic fever, without periodicity or pattern, is commonly found in vasculitides such as Kawasaki disease but also occurs in children with underlying infection.

2.**(D)** Symptoms of localized inflammation vary depending on the involved site. Arthritis, or inflammation of the synovium (synovitis), leads to joint pain, swelling, decreased range of motion, and functional limitations. Enthesitis is inflammation at the insertion of a ligament to a bone. Serositis, inflammation of serosal lining such as pleuritis, pericarditis, or peritonitis, gives rise to chest pain, shortness of breath, or abdominal pain.

3.**(A)** Henoch-Schönlein purpura (*aka* immunoglobulin A vasculitis [IgAV]) is a vasculitis of unknown etiology. The immune complexes associated with IgAV are predominantly composed of IgA, suggesting a hypersensitivity process. IgAV is slightly more common in males than females.

4.**(A)** Arthritis occurs in 80% of patients with HSP and is most common in the lower extremities. Gastrointestinal involvement occurs in about one half of affected children and most typically presents as mild to moderate crampy abdominal pain. One third of children with HPS develop renal involvement, which can be acute or chronic.
The hallmark of HSP is palpable purpura which can be accompanied by edema, particularly of the calves and dorsum of the feet, scalp, and scrotum or labia. It occasionally is associated with encephalopathy, pancreatitis, and orchitis.

5.**(B)** The platelet count is the most important test, because IgAV (immunoglobulin A vasculitis, *aka* Henoch-Schönlein purpura) is characterized by nonthrombocytopenic purpura with a normal, or even high, platelet count, differentiating IgAV from other causes of purpura that are associated with thrombocytopenia such as autoimmune thrombocytopenia, systemic lupus erythematosus, or leukemia.

6.**(B)** The diagnosis of IgAV is based on the presence of two of four criteria, which provides 87.1% sensitivity and 87.7% specificity for the disease. These criteria are

- Palpable purpura (Raised, palpable hemorrhagic skin lesions in the absence of thrombocytopenia)
- Bowel angina (Diffuse abdominal pain or the diagnosis of bowel ischemia)
- Diagnostic biopsy (Histologic changes showing granulocytes in the walls of arterioles or venules; IgA deposits in vessel wall)
- Pediatric age group (Age <20 yr at onset of symptoms)

7.**(D)** A typical dosing regimen is prednisone (1–2 mg/kg/day, maximum 60 mg/day) with a gradual taper plan over 4–6 weeks. Recurrence of abdominal pain and other symptoms as corticosteroids are weaned necessitate the longer course of treatment.

8.**(A)** The arthritis of IgAV does not leave any permanent joint damage. Renal involvement rarely may lead to renal failure. Patients with renal disease (elevated blood urea nitrogen, persistent high-grade proteinuria) are at highest risk for long-term complications such as hypertension or renal insufficiency, particularly if the initial course was marked by significant nephritis.

9.**(A)** Echocardiography should be performed at diagnosis and again after 1-2 wk of illness. If the results are normal, a repeat study should be performed 6-8 wk after onset of illness. If results of either of the initial studies are abnormal, more frequent echocardiography or other studies may be necessary. In patients without CCA at any time during the illness, echocardiography and a lipid profile are recommended 1 yr later.

10.**(D)**

11.**(C)** Multisystem inflammatory disease–children (MIS-C) is a complication of COVID-19 characterized by fever, rash, non- purulent conjunctivitis, gastrointestinal symptoms, cardiac involvement, shock, marked inflammation, and a positive serologic test for SARS-CoV-2 (often with a negative PCR for the virus). In the absence of a positive serologic test, it is often difficult to distinguish from KD.

12.**(B)**

13.**(D)**

14.**(C)** The child in the vignette has a clinical set of finding suggestive for the diagnosis of oligoarticular juvenile idiopathic arthritis which is the most common form of JIA. The arthritis is found in medium-sized to large joints; the knee is the most common joint involved, followed by the ankle and the wrist. Temporomandibular joint involvement occurs in more than 75% of patients. Children with oligoarticular JIA may be otherwise well without any evidence of systemic inflammation. In JIA, the onset of the arthritis is slow and the actual joint swelling is often noticed acutely by the child or parent, such as after an accident or fall, and can be confused with trauma.

15.**(A)** All children with chronic arthritis are at risk for chronic iridocyclitis or uveitis. The presence of a positive antinuclear antibody identifies children with arthritis who are at higher risk for chronic uveitis. Although all children with JIA are at increased risk, the subgroup of children, particularly young girls, with oligoarticular JIA and a positive antinuclear antibody are at highest risk, with an incidence of uveitis of 80%.

16.**(B)** Polyarticular JIA describes children with arthritis in five or more joints within the first 6 months of diagnosis. In contrast to oligoarticular JIA, children with polyarticular disease can present with evidence of systemic inflammation, including malaise, low-grade fever, growth retardation, anemia of chronic disease, and elevated markers of inflammation. The presence of a positive rheumatoid factor or anticyclic citrullinated protein (anti-CCP) antibody most likely represents a subgroup with true adult rheumatoid arthritis. Systemic Juvenile Idiopathic Arthritis does not present with onset of arthritis but rather with preceding systemic inflammation manifests with a typical recurring, spiking fever, accompanied by a rash.

16.**(A)**

17.**(D)** This patient has a clinical set of features that met the diagnostic criteria of juvenile ankylosing spondylitis which is categorized as one of the enthesitis-related arthritis. This group of diseases can present with peripheral arthritis and can be initially classified in other subgroups. It is only later, when the patient develops evidence of sacroiliac arthritis, psoriasis, or gastrointestinal disease, that the diagnosis becomes clear. Other important features of this group include the frequent presence of HLA-B27 and the need for earlier treatment with TNF blockers (e.g., Infliximab) as axial disease can have a poor response to conventional disease-modifying antirheumatic drugs (DMARDs) like methotrexate.

18.**(C)** Patients with SLE can present either in an abrupt fashion with fulminant disease or in an indolent manner. Nonspecific symptoms are common and may include significant fatigue and malaise, low-grade fever, and weight loss. The arthritis typically involves the small joints of the hands. In septic arthritis, leukocytosis and elevated CRP are common. Arthrocentesis is important to distinguish among the causes of arthritis (see table below). Reactive arthritis is typically symmetric and polyarticular, and it usually involves the large joints, especially the hips. Patients may have had a preceding episode of gastroenteritis or urethritis.

Synovial Fluid Findings in Various Joint Diseases		
CONDITION	WBC COUNT (mm^3/L)	PMNS (%)
Systemic lupus erythematosus	0–9,000 (3,000)	<20
Juvenile idiopathic arthritis, reactive arthritis, inflammatory bowel disease	250–80,000 (19,000)	>70
Infectious pyogenic infection	10,000–250,000 (80,000)	>90

19.**(B)** Systemic corticosteroid medications should be avoided in all but the most extreme circumstances, such as for severe systemic JIA with internal organ involvement or for significant active arthritis leading to functional disability. Methotrexate has become the

drug of choice for polyarticular and systemic JIA, which may not respond to baseline agents alone.

20.**(B)**

21.**(C)** A very serious complication of systemic JIA, macrophage activation syndrome (MAS), has received increasing recognition. Occurring in more than 10% of systemic JIA patients. The patient develops high fevers, hepatosplenomegaly, lymphadenopathy, and a bleeding diathesis. Laboratory studies reveal pancytopenia, elevated ferritin, transaminases, and triglycerides, as well as elevated soluble CD25. There is evidence of disseminated intravascular coagulation with low fibrinogen, and increased D-dimer. Bone marrow sampling may identify mature macrophages displaying hemophagocytic activity.
22.**(B)** Although there is a female predominance of this disease in adolescence and adulthood, there is an equal gender distribution in children.

23.**(C)** Both malar rashes and photosensitivity improve with appropriate therapy. The rash of discoid lupus, by contrast, results in permanent scarring and loss of pigmentation in the affected area. If discoid lupus occurs in the scalp, permanent alopecia ensues because of loss of hair follicles. The arthritis is non-erosive and rarely deforming.

24.**(C)**

25.**(A)** Criteria have been developed for the diagnosis of SLE. This patient has mouth ulcers, arthritis, serositis, malar rash, nephritis, and hematologic disorders. The presence of 4 of 11 of these criteria has 98% sensitivity and 97% specificity for SLE. Behçet disease is a rare vasculitic disorder that is characterized by a triple-symptom complex of recurrent oral aphthous ulcers, genital ulcers, and uveitis. Systemic Juvenile Idiopathic Arthritis is characterized by the occurrence of chronic arthritis, intermittently high spiking fever, evanescent rash (lasting a few hours), macular and salmon colored on the trunk and extremities during fever episodes, hepatomegaly and/or splenomegaly, lymphadenopathy, and serositis. Dermatomyositis tends to present in a slow, progressive fashion, with insidious onset of fatigue, malaise, and progressive muscle weakness, accompanied by low-grade fevers and rash.

26.**(D)** the boy in the vignette has SLE manifested by arthritis involving small joints of both hands, discoid lupus leading to loss of hair, hematologic abnormalities, and renal disease. Although nonspecific, a positive antinuclear antibody is found in more than 99% of patients with SLE. Antibodies to double-stranded DNA are present in most patients (70%) with SLE and are found almost exclusively in the disease. Antibodies directed against Smith are specific to SLE but found in only approximately 30% of persons with SLE. Antibodies to Ro (SSA) and La (SSB) can also be found in patients with SLE (15%) but also occur in patients with Sjögren syndrome.

27.**(B)** Excessive antibody production can lead to polyclonal hypergammopathy, with an elevated globulin fraction in the serum.

28.**(A)** Among the less specific diagnostic manifestations of SLE are fever, arthralgia, hepatosplenomegaly, episcleritis (localized or diffused redness without discharge), endocarditis, and myocarditis. Excessive circulating antibodies and immune complexes also lead to the consumption of complement proteins, with low levels of C3 and C4. Corticosteroids have been the mainstay of treatment for SLE for decades. Initial use of pulse methylprednisolone and high-dose oral prednisone frequently is required, followed by cautious tapering to minimize recurrence of symptoms. For patients who are not able to tolerate the tapering of corticosteroids, the use of steroid-sparing agents, such as methotrexate may be indicated. IVIG is the mainstay of therapy for KD. Splenomegaly and hypocomplementemia are not known clinical features to occur in Kawasaki disease. NSAIDs are often the first choice in the treatment of JIA. sJIA is characterized by arthritis, fever, rash, hepatosplenomegaly, lymphadenopathy, serositis (pericarditis) and leukocytosis. For patients who are not able to tolerate the tapering of corticosteroids, the use of steroid-sparing agents, such as methotrexate may be indicated.

29.**(C)** Azathioprine and mycophenolate mofetil have roles in the treatment of lupus nephritis due to the improved side effect profile compared to cyclophosphamide.
The childhood Arthritis Rheumatology Research Alliance (CARRA) consensus treatment plan for induction therapy of newly diagnosed lupus nephritis which advise 6 mo of induction therapy with either cyclophosphamide or mycophenolate mofetil (MMF) used in combination with a standardized glucocorticoid regimen. For patients who fail to achieve a partial response in 6 mo, it is appropriate to switch agents. Maintenance therapy consist of cyclophosphamide, MMF, or azathioprine.

30.**(D)** A classic JDM (photosensitivity) rash occurs on the face and across the cheeks but erythema also can be found on the shoulders and back (shawl sign). Patients may have heliotrope discoloration of the eyelids. Scaly, red plaques (Gottron papules) classically are found across the knuckles but can be found on the extensor surfaces of any joint.

31.**(B)** The muscle disease of JDM primarily affects the proximal muscles, particularly the hip and shoulder girdles. Exposure to the sun worsens the cutaneous manifestations and exacerbates the muscle disease; sunlight may lead to flare. Patients should be advised to wear sun block and refrain from prolonged sun exposure. Accordingly, supplementation with calcium and active forms of vitamin D is also indicated. The most serious complication of JDM is the development of calcinosis. Dystrophic calcification can occur in the skin and soft tissues in any area of the body.

32.**(A)** The diagnosis of growing pains is based on a typical history and a normal physical examination. It is important to consider leukemia as a cause of nocturnal leg pain in children of this age group, so it may be prudent to document a normal complete blood count.

33.**(C)** The treatment of hypermobility consists of reassurance and regular stretching. Arch supports can be helpful in children with symptomatic pes planus but are not indicated in the absence of symptoms. Benign hypermobility tends to improve with increasing age and is not associated with long-term complications.

SECTION 16

INFECTIOUS DISEASES

ZUHAIR ALMUSAWI

QUESTIONS

1. A young mother brought her 9-month-old breast fed baby to primary health center to receive his second dose Rota vaccine, the baby has an upper respiratory tract infection with low grade fever, the baby received his first dose at age of 13 weeks.
Which of the following is correct?

A. The baby can take the vaccine now
B. The baby should not take the vaccine
C. The baby can take the vaccine after subsidence of fever
D. The mother should not breast feed the baby one hour after vaccination

2. A 2-year-old child presented with cough, conjunctivitis, and fever for 3 days followed by macular rash that spreads over most of the body over 24 hours, together with prominent cervical nodes and abdominal pain. On examination the baby has bulging red tympanic membranes of both ears with bilateral fine rales on chest auscultation.
Of the following, the **MOST** likely diagnosis is

A. measles
B. adenoviral infection
C. scarlet fever
D. Kawasaki disease

3. A 3-year-old child presented with conjunctivitis, anorexia, malaise, and low-grade fever accompanied by an erythematous, maculopapular, discrete rash on the face and spreads to the body. On examination, he has retroauricular and posterior occipital lymphadenopathy with rose-colored spots on the soft palate.
Of the following, the **MOST** likely diagnosis is

A. measles
B. enteroviral infection
C. scarlet fever
D. rubella

4. A 15-month-old boy suddenly developed high fever followed by seizure. He is alert, behave normally without significant physical findings, PCR detect HHV-6 in his blood.
Of the following, the **MOST** appropriate treatment is

A. ganciclovir
B. foscarnet
C. adequate hydration and antipyretics
D. vancomycin and ceftriaxone

5. An 8-year-old boy with sickle cell disease developed low-grade fever, malaise, myalgia, mild conjunctivitis, and headache followed by red check with circumoral pallor. An erythematous symmetric, pruritic, maculopapular, truncal rash appears 2 days later with

progressive pallor, diarrhea, and cough. His CBC reveals reticulocyte count 1, hemoglobin 6 gm/dl, with neutropenia.
Of the following, the **MOST** appropriate management is

A. intravenous immunoglobulin
B. blood transfusion
C. intravenous immunoglobulin and blood transfusion
D. hydroxyurea

6. A 10-year-old boy presented with right facial paralysis and ear canal vesicles. Which of the following you recommend for this boy?

A. Oral famciclovir
B. Oral valacyclovir
C. Oral prednisolone
D. Symptomatic therapy

7. A 4-year-old boy presented with fever, malaise, and anorexia followed next day by pruritic small red papules that rapidly progress to non-umbilicated, oval, teardrop-like vesicles beginning on the trunk followed by the head and face, together with generalized lymphadenopathy.
Of the following, the **MOST** appropriate treatment is

A. symptomatic therapy
B. oral acyclovir
C. oral antibiotics
D. Varicella-zoster immunoglobulin

8. A 35-year-old mother developed primary varicella 2 days before normal vaginal delivery, her newborn is healthy looking full-term male baby.
What is your **NEXT** step for this baby?

A. Keep him in the hospital for close observation
B. Start varicella-zoster immunoglobulin (VZIG) as soon as possible
C. Start iv acyclovir immediately
D. Send for serum varicella PCR of the baby

9. A 5-year-old toxic appearing, feverish child presented with rapidly advancing tender, bright red lesion with sharp margins on his right hand, with enlargement of right axillary lymph nodes.
Of the following, the **MOST** likely cause is

A. group A streptococcus
B. Staphylococcus aureus
C. Pseudomonas aeruginosa
D. Aspergillus

10. A 3-year-old boy presented with well-demarcated, very tender, marked perianal erythema extending 2 cm from the anus, with anal pruritus and
painful defecation, sometimes with blood-streaked stools.
Of the following, the **MOST** likely diagnosis is

A. candidiasis

B. pinworm infection
C. anal fissure
D. perianal dermatitis

11. A 3-year-old healthy looking child presented with 3 small (3 mm), pearly flesh-colored, nontender, dome-shaped papules with central umbilication in the neck.
Which of the following treatment is recommended for such lesions?
A. No specific treatment
B. Cryotherapy
C. Topical 0.9% cantharidin
D. Removal by curettage

12. A 3-year-old boy brought by his mother complaining from fever, fatigue and malaise. On examination, he has bilateral anterior and posterior cervical and submandibular lymphadenopathy with hepatosplenomgaly. The pharynx shows enlarged tonsils with exudate and an enanthem with pharyngeal petechiae.
Of the following, the **MOST** reliable test for diagnosis of this disease is
A. polymerase chain reaction
B. complete blood count
C. immunoglobulin M antiviral capsid antigen
D. heterophile antibody

13. Which of the following is associated with poor prognosis in bacterial meningitis?
A. School age
B. Short duration of illness
C. Coma at presentation
D. High CSF white blood cell count

14. Extreme elevation of protein and reduction of glucose in CSF suggest
A. meningeal carcinomatosis
B. bacterial meningitis
C. autoimmune encephalitis
D. acute disseminated encephalomyelitis (ADEM)

15. A 3-year-old boy presented with low grade fever, nasal congestion, rhinorrhea, sore throat, and occasional nonproductive cough. Examination of the nasal mucosa reveals swollen, erythematous nasal turbinates.
Of the following, the **MOST** specific management for this boy is
A. antihistamines
B. cough suppressants
C. symptomatic therapy
D. vitamin C

16. Which of the following viruses cause acute pharyngitis as a prominent symptom?
A. Rhino virus
B. Adeno virus
C. Parainfluenza virus

D. Respiratory syncytial virus

17. A young mother brought her 6-year-old boy suffering from cough, sore throat, dysphagia, abdominal pain, and high fever. On examination the pharynx was red, the tonsils are enlarged, with petechiae on the soft palate and posterior pharynx. His anterior cervical lymph nodes were enlarged and tender to touch.
Of the following, the **MOST** helpful diagnostic test to guide your treatment is

A. white blood cell count
B. erythrocyte sedimentation rate
C. C-reactive protein
D. rapid streptococcal antigen test

18. A 4-year-old boy brought by his mother suffering from recurrent attacks of fever, aphthous stomatitis, pharyngitis, and cervical adenitis. Episodes last approximately 5 days, with a mean of 28 days between episodes.
Of the following, the **BEST** recommended management for this child is

A. oral prednisone
B. nonsteroidal anti-inflammatory drugs
C. intramuscular benzathine penicillin 600,000 U monthly till school entrance
D. oral amoxicillin as a single daily dose (50 mg/kg) for 10 days

19. Which of the following regimes is recommended to treat group A streptococcal pharyngitis in a 40 kg weighing boy allergic to penicillin?

A. Azithromycin 250 mg daily for 5 days
B. Cefixime 400 mg daily for 10 days
C. Cephalexin 500 mg BID for10 days
D. Clindamycin 300 mg/dose TID for 5 days

20. A 5-year-old child presented with persistent rhinorrhea, nasal congestion, and cough, especially at night for the last 14 days, associated with halitosis, headache,
facial swelling and tenderness.
Of the following, the **FIRST-LINE** therapy for this child is

A. amoxicillin-clavulanate for 7 days
B. amoxicillin for 10–14 days
C. clindamycin for 10 days
D. ceftriaxone daily for 5 days

21. An 11-month-old infant brought by his mother complaining from common cold followed by repeated vomiting, high fever, irritability, and poor feeding. After full examination you found an erythematous, opaque, bulging tympanic membrane.
Of the following, the recommended **NEXT** step treatment is

A. amoxicillin-clavulanate for 10 days
B. cefdinir for 7 days
C. ceftriaxone 3 days
D. azithromycin for 5 days

22. A 7-year-old boy presented with aural pain and tenderness with movement of the pinna, and with chewing. Inspection of the auditory canal revealed mild erythema with scant discharge.
Of the following, the **MOST** likely diagnosis is

A. otitis externa
B. otitis media with tympanic perforation
C. mastoiditis
D. cholesteatoma

23. A 2-year-old boy awakened at midnight with harsh cough, hoarseness, inspiratory stridor, low-grade fever, and respiratory distress (labored
breathing, suprasternal, intercostal, and subcostal retractions).
Which of the following is the first line management for this boy?

A. Oral or intramuscular dexamethasone once
B. Prednisolone orally for 3 days
C. Aerosolized racemic epinephrine
D. Oxygen therapy

24. A 3-week-old neonate presented with cough and repeated apneic attacks; his mother has persistent, nonproductive cough that begins as a nonspecific upper respiratory tract infection for the last 3- weeks before delivery.
Of the following, the **MOST** likely etiological agent of the neonate condition is

A. Bordetella pertussis
B. Mycoplasma hominis
C. Chlamydophila pneumoniae
D. Respiratory syncytial virus

25. A 6-month-old infant presented with cough and rhinorrhea which progresses over few days to noisy, raspy breathing and audible wheezing, associated with a low-grade fever and irritability. Physical examination revealed diffuse wheezes and crackles with prolongation of the expiratory phase, nasal flaring, intercostal and suprasternal retractions.
Which of the following tests is adequate for management of this infant?

A. Complete blood count
B. Pulse oximetry
C. Polymerase chain reaction for RSV
D. Chest radiograph

26. What is the **MOST** common cause of afebrile pneumonia in a 6-week-old infant?

A. Respiratory syncytial virus
B. Human metapneumovirus
C. Chlamydia trachomatis
D. Ureaplasma urealyticum

27. Which of the following agents of pneumonia is characterized by mild eosinophilia?

A. Aspergillosis
B. Pneumocystis jirovecii
C. Burkholderia cepacia

D. Chlamydia trachomatis

28. Approximately 8–10% of infective endocarditis in children occurs without structural heart disease (normal native valve) or other obvious risk factors.
What is the **MOST** likely infective organism in these cases?

A. Viridans Streptococcus groups
B. Staphylococcus aureus
C. Enterococcus
D. Coxiella burnetii

29. Which of the following viruses occurs in people of all ages, year-round, and is the **MOST** common cause of outbreaks of acute gastroenteritis?

A. Rotavirus
B. Astroviruses
C. Sapovirus
D. Norovirus

30. An 18-month-old boy presents with insidious onset of progressive anorexia, nausea, gaseousness, abdominal distention, watery diarrhea, and weight loss.
Of the following, the **MOST** likely cause is

A. Campylobacter jejuni
B. Yersinia enterocolitica
C. G. lamblia
D. Cryptosporidium

31. Which of the following is the diagnostic test of choice in giardiasis?

A. Microscopical stool examination
B. Nucleic acid amplification test (NAAT)
C. Duedenal aspirate
D. Serologic tests

32. Which of the following organisms can form Intestinal abscesses leading to intestinal perforation?

A. Shigella
B. Campylobacter jejuni
C. Clostridium difficile
D. Enterohemorrhagic (EHEC)

33. Which of the following is a good predictor of severe hepatocellular injury and progression to fulminant hepatic failure?

A. Alanine aminotransferase (ALT)
B. Aspartate aminotransferase (AST)
C. Total and direct (conjugated) bilirubin levels
D. Prothrombin time

34. A 9-month-old infant exposed to his 9-year-old brother with hepatitis A, 3 days ago, the mother is worried and consulted you for advice.

What is the **MOST** appropriate advice?

A. A single dose of HAV vaccine
B. Immunoglobulin given intramuscularly (IGIM)
C. IGIM along with the HAV vaccine
D. Good hygienic practices

35. A young primigravida HBsAg-positive mother gave birth to a full-term male newborn via C/S. The mother is asking how to protect her baby?

A. The baby should receive HBV vaccine immediately after birth.
B. The baby should receive HBV vaccine and hepatitis B immunoglobulin within 12 hours of birth.
C. The baby should receive HBIG as soon as possible.
D. The baby should take his usual scheduled vaccinations.

36. At what age, children begin to show the classic signs of UTI?

A. 6 months
B. 1 year
C. 2 years
D. 5 years

37. Which of the following imaging studies is **MOST** convenient to detect vesicoureteral reflux?

A. Ultrasonography
B. Voiding cystourethrogram (VCUG)
C. technetium-99m dimercaptosuccinic acid (DMSA)
D. Renal MRI

38. A 12-year-old girl presented with minimal, clear, thin vaginal discharge without pruritus or local signs of inflammation, the girl has no menarche yet, no pathogenic organisms on vaginal discharge culture.
Of the following, the **MOST** appropriate treatment is

A. improved hygiene
B. metronidazole
C. topical antifungal
D. reassurance

39. Which of the following is the recommended treatment for uncomplicated gonococcal infection of urethra in a 16-year-adolescent boy weighing 60 kg?

A. Ceftriaxone
B. Ciprofloxacin
C. Erythromycin
D. Azithromycin

40. A 15-year-old girl developed headache, fever, malaise, dysuria followed by multiple painful, grouped vesicles on external genitalia with regional lymphadenopathy.
Which of the following treatments is effective in reducing the severity and duration of symptoms?

A. Doxycycline
B. Azithromycin
C. Antifungal
D. Valacyclovir

41. A 5-year-old boy brought to emergency department suffering from Fever, anorexia, irritability, and lethargy. On examination, he has focal pain with exquisite point tenderness over the right upper leg associated with warmth, erythema, and swelling.
Of the following, the diagnostic study of choice for this boy is
A. magnetic resonance imaging
B. blood culture
C. direct metaphyseal needle aspiration
D. plain radiograph

42. A 4-year-old boy presented with fever, anorexia, irritability, and right upper thigh pain of two days duration. The affected area looks warm, red, with exquisite point tenderness over the bone. Magnetic resonance imaging (MRI) shows evidence of inflammatory marrow changes. Direct subperiosteal needle aspiration culture is negative.
Of the following, the **MOST** likely causative organism is
A. Staphylococcus aureus
B. Streptococcus pneumoniae
C. Kingella kingae
D. Haemophilus influenzae type b

43. Which of the following is the initial diagnostic procedure of choice for evaluating suppurative infections of the hip?
A. Plain radiograph
B. Ultrasound
C. Magnetic resonance imaging
D. CT scan

44.A 26-hour-newborn presented with copious purulent ocular discharge with matted eyelids. Physical examination reveals chemosis, injection of the conjunctivae, and edema of the eyelids.
Of the following, the **MOST** likely organism is
A. Chlamydia trachomatis
B. Streptococcus pneumoniae
C. Moraxella catarrhalis
D. Neisseria gonorrhoeae

45. Hyperpurulent conjunctivitis characterized by reaccumulation of purulent discharge within minutes is characteristic of infection with
A. Neisseria gonorrhoeae
B. Staphylococcus
C. Pseudomonas
D. E. coli

46. A 10-year-old girl with thalassemia diagnosed at one year, she is on regular blood transfusion and deferoxamine for iron overload.
Which of the following pathogens is more likely to be associated with her condition?

A. Streptococcus pneumonia
B. Pneumocystis jirovecii
C. Aspergillus sp.
D. Yersinia enterocolitica

47. A 5-year-old boy presented with high fever, rigors, sweats, and headache that occur every 48 hours for the last 10 days, the boy looks pale with decreased level of consciousness, his spleen is 10 cm below costal margin.
Of the following, the **MOST** likely diagnosis is

A. typhoid fever
B. brucellosis
C. kala-azar
D. malaria

48. Which of the following is the major manifestation of hookworm infestation?

A. anemia
B. abdominal pain
C. indigestion
D. diarrhea

49. A 5-year-old boy brought to emergency department complaining from cough, blood-stained sputum, eosinophilia, and infiltrates on chest x-ray film. The boy also has abdominal pain and distention with recent onset of jaundice.
Of the following, the **MOST** likely cause is

A. Ancylostoma duodenale
B. Trichuris trichiura
C. Ascaris lumbricoides
D. Strongyloides stercoralis

50. After how long of starting effective treatment, most infectious TB patients become noninfectious?

A. within 1 week
B. within 2 weeks
C. within 4 weeks
D. within 6 weeks

51. A 16-year-old boy presented with productive cough, hemoptysis, fever, night sweats, malaise, and weight loss. CXR reveals right upper lobe infiltrate with cavity formation. The tuberculin skin test (TST) and interferon-γ release assay (IGRA) are positive.
Which of the following is the **BEST** clinical description?

A. Latent tuberculosis
B. Primary TB
C. Reactivation pulmonary tuberculosis
D. Miliary tuberculosis

52. Which of the following is **TRUE** regarding IGRA test?

A. It is more sensitive than Mantoux test
B. It is unaffected by prior BCG vaccination
C. It requires two visits to complete
D. It is the recommended for persons older than 6 years of age

53. Which of the following manifestations are more common in children than adults with HIV infection?

A. Chronic parotid swelling
B. Weight loss
C. Lymphadenopathy
D. Maculopapular rash

54. After what age, persistence of a positive HIV antibody test indicates HIV infection.?

A. 6 months
B. 12 months
C. 18 months
D. 24 months

INFECTIOUS DISEASES

ZUHAIR ALMUSAWI

ANSWERS

1.**(B)** Rotavirus vaccine, maximum age for first dose is 14 weeks and 6 days, while maximum age for final dose is 8 months.

2.**(A)** Measles infection is divided into four phases: incubation, prodromal (catarrhal), exanthematous (rash), and recovery. The manifestations of the 3-day prodromal period are cough, coryza, conjunctivitis, and the pathognomonic **Koplik spots.** The conjunctiva may reveal a characteristic transverse line of inflammation along the eyelid margin **(Stimson line)**. The classic symptoms of cough, coryza, and conjunctivitis continue to occur during the secondary viremia of the exanthematous phase, which often is accompanied by high fever (40–40.5°C). The macular rash begins on the head (often above the hairline) and spreads over most of the body in a cephalad-to-caudal pattern over 24 hours. Areas of the rash are often confluent. Generalized lymphadenopathy may be noted with the rash, with cervical nodes being most prominent. Otitis media, pneumonia, and diarrhea are more common in infants.

3.**(D)** The incubation period for postnatal rubella is typically 16–18 days. The mild catarrhal symptoms of the prodromal phase of rubella may go unnoticed. The characteristic signs of rubella are retroauricular, posterior cervical, and posterior occipital lymphadenopathy accompanied by an erythematous, maculopapular, discrete rash. The rash begins on the face and spreads to the body, lasting for 3 days, and is less prominent than that of measles. Rose-colored spots on the soft palate, known as **Forchheimer spots**, develop in 20% of patients and may appear before the rash. Other manifestations of rubella include mild pharyngitis, conjunctivitis, anorexia, headache, malaise, and low-grade fever. Polyarthritis, usually of the hands, may occur, especially among adult females, but usually resolves without sequelae. Paresthesias and tendinitis may occur.

4.**(C)** There is no specific therapy for roseola. Routine supportive care includes maintaining adequate hydration and antipyretics. In immunocompromised hosts, use of ganciclovir or foscarnet can be considered.

5.**(B)** There is no specific therapy for erythema infectiosum other than routine supportive care. Transfusions may be required for transient aplastic crisis. Intrauterine transfusion has been performed for hydrops fetalis associated with fetal parvovirus B19 infection. Intravenous immunoglobulin may be used for immunocompromised persons with severe anemia or chronic infection.

6.**(B)** Involvement of cranial nerve VII by herpes zoster may result in facial paralysis and ear canal vesicles (Ramsay Hunt syndrome). Oral famciclovir and valacyclovir have much greater oral bioavailability than acyclovir and are recommended for treatment of zoster in adults. Valacyclovir is approved for use in children 2 years and older, and acyclovir is available for use in all age groups.

7.**(A)** Symptomatic therapy of varicella includes non-aspirin antipyretics, cool baths, and careful hygiene. Routine oral administration of acyclovir is not recommended in otherwise healthy children with varicella.

8.**(B)** A severe form of neonatal varicella may develop in newborns of mothers with primary varicella (but not shingles) occurring 5 days before to 2 days after delivery. The fetus is exposed to a large inoculum of virus but is born before the maternal antibody response develops and can cross the placenta. These infants should be treated as soon as possible with varicella-zoster immunoglobulin (VZIG) or intravenous immunoglobulin if VZIG is unavailable, to attempt to prevent or ameliorate the infection.

9.**(A)** Erysipelas is a superficial variant of cellulitis usually caused by group A streptococcus that involves the dermis only. The rapidly advancing lesions are tender, bright red in appearance, and have sharp margins. The patients may appear toxic. Blood cultures are recommended for erysipelas.

10.**(D)** Perianal dermatitis (perianal streptococcal disease) is caused by group A streptococcus and is characterized by well-demarcated, very tender, marked perianal erythema extending 2 cm from the anus. Manifestations include anal pruritus and painful defecation, sometimes with blood-streaked stools.

11.**(A)** Molluscum contagiosum lesions are self-limited, resolving over months to years, and usually no specific treatment is recommended. Available treatment options are limited to destructive modalities, such as cryotherapy with topical liquid nitrogen, vesicant therapy with topical 0.9% cantharidin, or removal by curettage, and should be reserved for extensive disease.

12.**(C)** The most reliable test for diagnosis of acute EBV infection is the immunoglobulin M (IgM) antiviral capsid antigen. Heterophile antibody is also diagnostic but is not reliably positive in children younger than 4 years with infectious mononucleosis.

13.**(C)** Poor prognosis is associated with young age, long duration of illness before effective antibiotic therapy, seizures, coma at presentation, shock, low or absent CSF white blood cell count in the presence of visible bacteria on CSF Gram stain, and immunocompromised status.

14.**(A)** Extreme elevations of protein and reductions of glucose suggest tuberculosis, cryptococcal infection, or meningeal carcinomatosis.

15.**(C)** There is no specific therapy for URIs. Antibacterial therapy is not beneficial and instead may cause harm. Management consists of symptomatic therapies. Antihistamines and decongestants are not recommended for children younger than 6 years of age because of adverse effects and lack of benefits. Similarly, cough suppressants and expectorants have not been shown to be beneficial. Vitamin C and inhalation of warm, humidified air are no more effective than placebo. The benefit of zinc lozenges or sprays has been inconsistent in children.

16.**(B)** Many viruses cause acute pharyngitis. Some viruses, such as adenoviruses, are more likely than others to cause pharyngitis as a prominent symptom, whereas other viruses, such as rhinoviruses, are more likely to cause pharyngitis as a minor part of an illness that primarily features other symptoms, such as rhinorrhea or cough.

17.**(D)** The principal challenge is to distinguish pharyngitis caused by group A streptococcus from pharyngitis caused by viral organisms. A rapid streptococcal antigen test, a throat culture, or both should be performed to improve diagnostic precision and to identify children most likely to benefit from antibiotic therapy. The predictive values of white blood cell count, erythrocyte sedimentation rate, and C-reactive protein are not sufficient to distinguish streptococcal from non-streptococcal pharyngitis, and these tests are not routinely recommended.

18.**(A)** A syndrome of periodic fever, aphthous stomatitis, pharyngitis, and cervical adenitis (PFAPA) is a rare cause of recurrent fever in children. The fevers begin at a young age (usually <5 years). Episodes last approximately 5 days, with a mean of 28 days between episodes. Episodes are shorter with oral prednisone and do not respond to nonsteroidal anti-inflammatory drugs or antibiotics. Long-term sequelae do not develop.

19.**(C)** ***For persons allergic to penicillin:***
Cephalexin 25 mg/kg/dose BID, maximum dose 500 mg/dose ×10 days
Clindamycin 6–7 mg/kg/dose TID, maximum dose 300 mg/dose ×10 days
For persons allergic to β-lactams:
Azithromycin, children: 12 mg/kg (maximum: 500 mg/dose) on day 1 followed by 6 mg/kg/dose (maximum: 250 mg/dose) OD on days 2 through 5.

20.**(B)** Amoxicillin for 10–14 days can be used as first-line therapy of uncomplicated sinusitis in children. Broadening therapy to amoxicillin-clavulanate is appropriate if there is no clinical response to amoxicillin within 2–3 days, if risk factors for resistant organisms are present (antibiotic treatment in the preceding 1–3 months, daycare attendance), if there is chronic sinusitis, or if any amount of eye swelling is present.

21.**(C)** Intramuscular ceftriaxone is especially appropriate for children with vomiting that precludes oral treatment.

22.**(A)** Pain, tenderness, and aural discharge are the characteristic clinical findings of otitis externa. Fever is notably absent.

23.**(A)** Oral or intramuscular dexamethasone for children with viral croup reduces symptoms and the need for hospitalization and shortens hospital stays. Dexamethasone (0.6 mg/kg) may be given once intramuscularly or orally. Alternatively, prednisolone (2 mg/kg/day) may be given orally for 3 days. For significant airway compromise, administration of aerosolized racemic (D- and L-) epinephrine reduces subglottic edema by adrenergic vasoconstriction, temporarily producing marked clinical improvement. Sedatives should be used cautiously and in the intensive care unit only.

24.**(A)** Infants may not display the classic findings, and the first sign in the neonate may be apnea. Young infants are unlikely to have the classic whoop, more likely to have central nervous system damage as a result of hypoxia, and more likely to have secondary bacterial pneumonia. Adolescents and adults with pertussis usually present with a prolonged bronchitic illness with persistent, nonproductive cough that often begins as a nonspecific upper respiratory tract infection. In general, adolescents and adults do not have a whoop with the cough, although they may have severe paroxysms.

25.**(B)** Routine laboratory tests are not required to confirm the diagnosis. It is important to assess oxygenation in severe cases of bronchiolitis. Pulse oximetry is adequate for monitoring oxygen saturation.

26.**(C)** Chlamydia trachomatis and, less commonly, Mycoplasma hominis, Ureaplasma urealyticum, and cytomegalovirus (CMV) cause a similar respiratory syndrome in infants 2 weeks to 3 months of age, with subacute onset of an afebrile pneumonia; cough and hyperinflation are the predominant signs.

27.**(D)** Mild eosinophilia is characteristic of infant C. trachomatis pneumonia.

28.**(B)**

29.**(D)** Norovirus occurs in people of all ages, year-round, and is the most common cause of outbreaks of acute gastroenteritis because it is highly contagious.

30.**(C)** G. lamblia is transmitted through ingestion of cysts, either from contact with an infected individual or from food or water contaminated with infected feces. The organism adheres to the microvilli of the duodenal and jejunal epithelium. Insidious onset of progressive anorexia, nausea, gaseousness, abdominal distention, watery diarrhea, secondary lactose intolerance, and weight loss is characteristic of giardiasis.

31.**(B)** Giardiasis can be diagnosed by identifying trophozoites or cysts in stool, but this may require three specimens. More specific and sensitive fecal immunoassays and NAATs are the diagnostic tests of choice.

32.**(A)** Intestinal abscesses can form with Shigella, Yersinia, and Salmonella infections, leading to intestinal perforation, a life-threatening complication.

33.**(D)** Alanine aminotransferase (ALT) and aspartate aminotransferase (AST) levels are elevated and generally reflect the degree of parenchymal inflammation. Alkaline phosphatase and total and direct (conjugated) bilirubin levels indicate the degree of cholestasis, which results from hepatocellular and bile duct damage. The prothrombin time is a good predictor of severe hepatocellular injury and progression to fulminant hepatic failure.

34.**(B)** Unvaccinated household and sexual contacts of persons with HAV should receive postexposure prophylaxis as soon as possible and within 2 weeks of the last exposure. A single dose of HAV vaccine at the age-appropriate dose is preferred for persons 12 months

to 40 years of age. Immunoglobulin (0.1 mL/ kg) given intramuscularly (IGIM) is preferred for children under 12 months of age, persons over 40 years of age, and immunocompromised persons.

35.**(B)** Infants born to HBsAg-positive mothers should receive HBV vaccine and hepatitis B immunoglobulin (HBIG; 0.5 mL) within 12 hours of birth, with subsequent vaccine doses at 1 and 6 months of age followed by testing for HBsAg and anti-HBs at 9–15 months of age. Infants born to mothers whose HBsAg status is unknown should receive vaccine within 12 hours of birth. If maternal testing is positive for HBsAg, the infant should receive HBIG as soon as possible (no later than 1 week of age).

36.**(C)** Infants 1 month to 2 years of age may present with feeding problems, failure to thrive, diarrhea, vomiting, or unexplained fever. The symptoms may masquerade as gastrointestinal illness, with colic, irritability, and crying periods. At 2 years of age, children begin to show the classic signs of UTI, such as urgency, dysuria, frequency, and abdominal or back pain. The presence of UTI should be suspected in all infants and young children with unexplained fever and in patients of all ages with fever and congenital anomalies of the urinary tract.

37.**(B)** Voiding cystourethrogram (VCUG) is indicated if the ultrasound is abnormal (hydronephrosis, scarring, or other findings suggesting obstruction or congenital abnormality). Vesicoureteral reflux is the most common abnormality found and is ranked from grade I (ureter only) to grade V (complete gross dilation of the ureter and obliteration of caliceal and pelvic anatomy).

38.**(D)** Physiologic vaginal discharge or physiologic leukorrhea of desquamated vaginal cells and mucus occurs normally in females soon after birth, with discharge lasting about 1 week, and appears again at 6–12 months before menarche. The discharge is minimal, clear, and thin without pruritus or inflammation. No treatment is necessary.

39.**(A)** Increasing rates of resistance to fluoroquinolones limit treatment options. A single IM dose of ceftriaxone (500 mg for patients 45–150 kg and 1 g for patients >150 kg) is recommended for uncomplicated gonococcal infections of the cervix, urethra, and rectum.

40.**(D)** Oral acyclovir, famciclovir, and valacyclovir are effective treatments in reducing the severity and duration of symptoms in primary cases and may reduce recurrences. Once-daily suppressive therapy reduces the frequency of genital herpes recurrences by more than 75%, and lesions tend to be mild when they do occur.

41.**(A)** Magnetic resonance imaging (MRI) is sensitive to the inflammatory marrow changes even during the earliest stages of osteomyelitis and is the diagnostic study of choice.

42.**(C)** The use of polymerase chain reaction (PCR) testing reveals that a significant proportion of culture-negative osteomyelitis is due to Kingella kingae, particularly in children under 5 years of age.

43.**(B)** Ultrasound is especially helpful for identifying joint effusions and is the initial diagnostic procedure of choice for evaluating suppurative infections of the hip. Plain radiographs typically add little information to the physical findings. Radiographs may show joint capsule swelling, a widened joint space, and displacement of adjacent normal fat lines. Magnetic resonance imaging is helpful to confirm the diagnosis and exclude concurrent bone infection or deep abscesses.

44.**(D)** The timing and manifestations of neonatal conjunctivitis are helpful in identifying the cause. N. gonorrhoeae causes severe conjunctivitis with profuse purulent discharge 1–7 days after birth.

45.**(A)** Distinguishing bacterial from viral conjunctivitis by presentation and appearance is difficult. Vesicular lid lesions, if present, suggest HSV. Hyperpurulent conjunctivitis characterized by reaccumulation of purulent discharge within minutes is characteristic of N. gonorrhoeae infection.

46.**(D)** Rhizopus spp. and Yersinia enterocolitica are more likely to be associated with her condition.

47.**(D)** The most important aspect of diagnosing malaria in children is to consider the possibility of malaria in any child who has fever, chills, splenomegaly, anemia, or decreased level of consciousness with a history of recent travel or residence in an endemic area, regardless of the use of chemoprophylaxis.

48.**(A)** Symptoms of abdominal pain, anorexia, indigestion, fullness, and diarrhea occur with hookworm infestation. The major manifestation of infection is anemia.

49.**(C)** Manifestations may be the result of migration of the larvae to other sites of the body or the presence of adult worms in the intestine. Pulmonary ascariasis occurs as the larvae migrate through the lung, producing cough, blood-stained sputum, eosinophilia, and transient infiltrates on chest x-ray films. Adult larvae in the small intestine may cause abdominal pain and distention. Intestinal obstruction from adult worms can occur, particularly in children due to the small diameter of their intestinal lumens and their propensity to acquire large worm burdens. Migration of worms into the bile duct may rarely cause acute biliary obstruction.

50.**(B)** Most infectious patients become noninfectious within 2 weeks of starting effective treatment. Cavitary disease, nonadherence to treatment, and resistant infection increase the risk of transmission.

51.**(C)** Reactivation pulmonary tuberculosis, common in adolescents and typical in adults, usually is confined to apical segments of upper lobes or superior segments of lower lobes. There is usually little lymphadenopathy and no extrathoracic infection. This is a manifestation of a secondary expansion of infection at a site seeded years previously during primary infection. Advanced disease is associated with cavitation and endobronchial spread of bacilli. Symptoms include fever, night sweats, malaise, and weight loss. A productive cough and hemoptysis often herald cavitation and bronchial erosion.

52.**(B)** An IGRA is a whole blood test that measures INF-γ production by T lymphocytes in response to antigens from the M. tuberculosis complex, which includes M. bovis but not antigens found in the BCG vaccine. An IGRA is the recommended diagnostic test for persons older than 2–4 years of age in the United States. It has similar sensitivity to the TST but improved specificity because it is unaffected by prior BCG vaccination. It is also practically easier to obtain, as it requires only one visit to complete.

53.**(A)** Manifestations that are more common in children than adults with HIV infection include recurrent bacterial infections, lymphoid hyperplasia, chronic parotid swelling, lymphocytic interstitial pneumonitis, and earlier onset of progressive neurologic deterioration. Pulmonary manifestations of HIV infection are common and include Pneumocystis jirovecii pneumonia, which can present early in infancy as a primary pneumonia characterized by hypoxia, tachypnea, retractions, elevated serum lactate dehydrogenase, and fever.

54.**(C)** Loss of HIV antibody combined with negative HIV DNA PCR confirms absence of HIV infection. Persistence of a positive HIV antibody test after 18 months of age indicates HIV infection.

SECTION 17

THE DIGESTIVE SYSTEM

KHALID ALAARAJI

QUESTIONS

1. A 7-year-old female presented with recurrent, periumbilical, crampy, abdominal pain mainly at the morning for the last 3 months which interfere with school attendance. The pain is not related to specific meal, relieved by defecation, and associated with change in bowel. Of note, growth parameters are normal.
Of the following, the **MOST** likely diagnosis is

A. functional abdominal pain
B. irritable bowel syndrome
C. celiac disease
D. lactose intolerance

2. Regarding to above scenario, while a waiting for laboratory and ultrasound results.
Which of the following is the **BEST** action?

A. Avoid school attendance
B. Start amitriptyline
C. Start gluten free diet
D. Trial of 3-day lactose-free diet

3. A 4-year-old girl brought by anxious mother because of frequent soiled underwear with infrequent passage of large-caliber stools for last year.
Which of the following is the **MOST** likely finding on rectal examination?

A. Anteriorly displaced anus
B. Snug anal sphincter and empty, contracted rectum
C. Normal or reduced sphincter tone and dilated rectal vault
D. Lax sphincter tone

4. Which of the following medications is the **MOST** likely cause of dental caries?

A. Fluoride
B. Tetracycline
C. Phenytoin
D. Anticholinergic drugs

5. Which of the following is affected by pill ulcers?

A. Oral cavity
B. Esophagus
C. Stomach
D. Duodenum

6. Which of the following caustic products produce the worst esophageal injuries?

A. Bleach
B. Detergents

C. Granular products
D. Lye-based drain cleaners

7. A 3-week-old boy, product of uneventful pregnancy, presented with repeated bilious vomiting for the last 3 days, there is no history of polyhydramnios and no abdominal distention and pass meconium within the first 24hr of life.
Which of the following is the **BEST** method to confirm the diagnosis?

A. Plain abdominal x-ray
B. Abdominal ultrasound
C. Upper gastrointestinal series
D. Contrast enema

8. A 5-day-old boy with Down syndrome presented with repeated bilious vomiting for the last 2 days, slight abdominal distention and pass meconium within first 24hr after birth.
Of the following, the **MOST** likely diagnosis is

A. mid gut volvulus
B. duodenal atresia
C. pyloric atresia
D. Hirschsprung disease

9. Which of the following conditions is the **MOST** likely cause of substantial enamel erosion and caries?

A. Renal failure
B. Congenital syphilis
C. Crohn disease
D. Gastroesophageal reflux disease

10. A 5-day-old boy brought to ER because of maternal concerns of difficult stooling. He had uneventful perinatal period, started breastfeeding well, and passed urine within 1st 24hr after birth. There was delay in passage of meconium with progressive abdominal distention. Digital rectal examination reveals an empty rectum.
Of the following, the **BEST** diagnostic tool is

A. erect radiograph
B. barium enema
C. anorectal manometry
D. rectal biopsy

11. A 10-year-old girl with frequent visit for the last 3-month complaining from nausea, right upper quadrant abdominal pain, and frequent 4 – 6 time loose bloody bowel motion. She lost about 8 kg in the last six months. Lab data reveals hemoglobin: 8 g/dL, serum albumin: 2.5 g/dL, and negative stool culture.
Of the following, the **BEST** method for assessment is

A. upper endoscopy
B. colonoscopy
C. upper GI series with small bowel follow-through
D. computed tomography (CT) scanning

12. To prevent dental caries; The American Academy of Pediatrics recommends that primary care physicians apply fluoride varnish to the teeth of all children every

A. 3–6 months
B. 9–12 months
C. 15–18 months
D. 21–24 months

13. A 14-year-old girl complains from of recurrent abdominal pain, diarrhea, fatigue, and poor appetite associated with oral ulcers and joints pain. She lost 9 kg over the last 6 months. Rectal examination reveals an anal fissure with exudate. Lab results: WBC count 10,900/µL, hemoglobin 8.2 g/dL, platelet count 550 /µL, CRP 4.7 mg/dL, and ESR 107 mm/h.
Of the following, the **MOST** likely diagnosis is

A. Crohn disease
B. intestinal lymphoma
C. ulcerative colitis
D. celiac disease

14. What is the optimal age for surgical repair of cleft lip?

A. 3 months
B. 6 months
C. 9 months
D. 12 months

15. A previously healthy, 12-month-old boy presents because of mother concerns about his infrequent stooling, refusal to eat, protracted vomiting with episodes of excessive crying every 20 to 30 minutes. Temperature, 37.9°C; heart rate, 154 beats/min; respiratory rate, 35 breaths/min; and oxygen saturation, 98% on room air. On physical examination; slightly distended abdomen, dry mucous membranes, cool extremities with a capillary refill time of > 3 seconds.
Which of the following is the **MOST** appropriate action?

A. Surgical consultation
B. Fluid resuscitation
C. Abdominal ultrasound
D. Pneumatic reduction

16. A 13-year-old boy with diabetes mellitus complains from loose stools and intermittent abdominal pain over the past 4 months. Abdomen is distended and tympanic. Height is below 3rd percentile, weight at the 3rd percentile. One year ago, the adolescent's weight was at the 36th percentile
Of the following, the **MOST** likely diagnosis is

A. Celiac disease
B. Hypothyroidism
C. Crohn disease
D. Developmental lactose intolerance

17. Which of the following medications used during pregnancy increase the risk of cleft lip?

A. Valproic acid

B. Aspirin
C. Metformin
D. Methyldopa

18. A 3-week-old breast fed infant evaluated for prolonged neonatal jaundice. On examination; active, just palpable liver, ejection systolic murmur in pulmonary area, deep-set eyes, prominent forehead, and a pointed chin. lab results show: TSB; 8.8 mg/dl and conjugated bilirubin level; 5.6 mg/Dl, SGPT; 113 IU/L, and GGT is 310 IU/L.
Of the following, the **MOST** likely diagnosis is
A. congenital rubella
B. breast milk jaundice
C. Alagille syndrome
D. Galactosemia

19. A 15-year-old girl presented to outpatient clinic for evaluation of severe recurrent abdominal pain over the past 6 months. The episode, is characterized by periumbilical pain, nausea, vomiting, headache that interfere with daily activity. The girl is well nourished with normal growth parameters. The abdominal sonograph, celiac screen, liver and renal function tests, urinalysis were normal, and stool for Helicobacter pylori was negative.
Of the following, the **MOST** likely diagnosis is
A. abdominal migraine
B. Crohn's disease
C. cyclic vomiting syndrome
D. functional abdominal pain

20.At what age, most infants stop spitting up?
A. 3–6 months
B. 6–9 months
C. 9–12 months
D. 12–15 months

21. A 14-year-old boy presented with altered mental status, dark color urine and icterus. Over the past 4 months, mother noted a decline in school performance, worsening handwriting, and progressive fatigability. He had received blood transfusion once. On examination he was confused, pupils reactive, brisk deep tendon reflexes, jaundice, distended abdomen, and hepatosplenomegaly. Lab results show; Hb 8 g/dL, WBC count; 16,000/µL, Platelet count 140/µL, serum bilirubin 7.3 mg/dl, ALT 210 U/L, AST 340 U/L.
Of the following, the **NEXT** step to establish the diagnosis is
A. Combs test
B. serum ceruloplasmin
C. viral hepatitis screen
D. liver biopsy

22. What is the **MOST** common form of tracheoesophageal fistula (TEF)?
A. H-type TEF
B. Esophageal atresia with no TEF
C. Esophageal atresia with distal TEF

D. Esophageal atresia with proximal TEF

23. What is the **MOST** common associated defect with tracheoesophageal fistula (TEF)?
A. Renal anomalies
B. Vertebral anomalies
C. Cardiac anomalies
D. GI anomalies

24. A 12-year-old boy is evaluated for weight loss over the past 6 months, during this period, he has had difficulty with eating and swallowing associated with abdominal pain and vomiting always ended by food impaction which make him to visit emergency department. He is on controller therapy for asthma. On examination, he appears thin, wasted, with soft abdomen.
Of the following, the **MOST** likely diagnosis is
A. eosinophilic esophagitis
B. achalasia
C. gastroesophageal reflux disease
D. peptic ulcer disease

25. You are called to evaluate a newly delivered baby boy to a polyhydramnios mother. He has excessive oral secretions requiring frequent suction. Vital signs show a temperature of 37°C, heart rate of 120 beats/min, and a respiratory rate of 45 breaths/min, O2 saturation is 97% at room air. A chest radiograph is pending.
What is the **MOST** important next step to reach the diagnosis?
A. Chest x-ray with NG tube
B. Chest C-T scan
C. Barium study
D. Chest US

26. A 6-year-old girl is brought to medical advice for evaluation of frequent soiling with small amounts of offensive loose stool. She gives history of nonspecific infrequent periumbilical abdominal pain which interfere with eating. There is history of passage of hard stools every few days. Physical examination reveals soft abdomen with palpable hard mass in the left lower quadrant. Rectal examination reveals normal sphincter tone, dilated rectal vault, and hard stool mass.
Of the following, the **BEST** initial step of management is to
A. start oral polyethylene glycol
B. decrease milk consumption
C. increase fiber diet
D. use mineral oil enema

27. Which of the following objects is the **MOST** common cause of esophageal foreign body?
A. Coins
B. Small toys
C. Food items
D. Button batteries

28. A 4-week-old boy presented for evaluation of weight loss. His mother has had concern about his frequent, nonbilious, forceful vomiting. On examination he is thin, active, eager to feed, with soft, nontender abdomen, and decreased urine output. You send him for biochemistry study.
Of the following, the **EXPECTED** blood gas finding is

A. hypochloremic hypokalemic metabolic alkalosis
B. hypochloremic hyponatremic metabolic acidosis
C. hyperchloremic hyperkalemic metabolic alkalosis
D. hyperchloremic hypernatremic metabolic acidosis

29. A 2-year-old girl presented for evaluation of diarrhea that persist for about 8 weeks. The stool is loose and contains undigested food particles. There are no constitutional symptoms. She looks well hydrated, with normal growth parameters.
What the **BEST** line of management?

A. Gluten free diet
B. Lactose free formula
C. Reduce beverage intake
D. Zinc supplement

30. What is the preferred test to asses bile duct injury?

A. Alanine aminotransferase
B. Aspartate aminotransferase
C. γ-glutamyltransferase
D. Prothrombin time

31. An anxious mother brought her healthy looking 4-month-old breastfed male infant who had frequent effortless spit-up of milk. He is gaining weight adequately.
Of the following, the **BEST** line of management is

A. reassurance
B. PPI
C. AR formula
D. Referring to GI specialist

32. A 6-month-old boy is brought to health center for evaluation of vomiting since birth. The vomiting is forceful, nonbilious, and associated with arching trunk. He is on mixed feeding and his mother changes the formula without benefit. His growth chart shows that his weight has declined from the 50th to 25th percentile over the past 3 months.
Of the following, the **BEST** next step in evaluation is

A. abdominal ultrasonography
B. upper gastrointestinal series
C. 24-hour esophageal pH probe monitoring
D. upper endoscopy

33. Which of the following deciduous teeth is the first to erupt?

A. Lower central incisors
B. Upper central incisors
C. Lateral incisors

D. Canines

34. Referred pain is a painful sensation in a body region distant from the true source of pain. The pain that referred to the region below the right scapula is mostly from

A. stomach
B. liver
C. pancreas
D. gallbladder

35. A 13-year-old girl presents with a history of periumbilical pain for the last 6 months mainly at morning, not related to meal or defecation, sometime accompanied by nausea with a normal bowel motion, there is no other constitutional symptoms. The girl is well-nourished with normal growth parameters. All investigations regarding her condition were normal including; abdominal sonograph, celiac screen, liver and renal function tests, urinalysis, and stool for Helicobacter pylori.
Of the following, the **BEST** treatment plan for this patient is

A. amitriptyline
B. probiotics
C. peppermint oil
D. no medication

36. A 9-year-old previously healthy girl admitted to the emergency department complaining from disturbance in the mental status, excessive sleepiness and labored breathing over the past 12 hours. Her mother reports that she has had malaise, nausea, vomiting, and abdominal discomfort for the last 4 days. Examination shows soft abdomen with liver 2 cm below the costal margin, clear chest, regular heart rhythm. She is confused, her pupils reactive, and has brisk deep tendon reflexes. Heart rate; 88 beats/min, respiratory rate; 27 breaths/min, temp; 38°C, blood pressure; 95/40 mm Hg, and O_2 saturation; 97% on room air. Hb; 10 g/dL, WBC count; 21,000/µL, Platelet count 160/µL, blood urea; 20 mg/Dl, serum creatinine; 1.1 mg/dL, aspartate aminotransferase; 2,000 U/L, alanine aminotransferase 2,100 U/L.
Of the following, the **NEXT** step in management is to send for

A. CSF study
B. serum ammonia
C. brain MRI
D. PT, PTT, INR

37. Which of the following tests is **BEST** for diagnosis of acute pancreatitis?

A. Serum amylase
B. C-peptide
C. Serum lipase
D. Fecal elastase

38. A 5-year-old boy with nephrotic syndrome, on steroids for the last 2 months, brought to emergency department with severe abdominal pain which restrict his movement associated with loose non bloody bowel motion. He is afebrile, looks unwell, lying on left side and the abdomen is tense with rebound tenderness.

Of the following, the **MOST** likely causative organism is

A. Staphylococcal aureus
B. Campylobacter jejune
C. Streptococcus pneumoniae
D. Escherichia coli

THE DIGESTIVE SYSTEM

KHALID ALAARAJI

ANSWERS

1.**(B)** Children with functional abdominal pain characteristically have pain almost daily. The pain is not associated with meals or relieved by defecation and is often associated with a tendency toward anxiety and perfectionism. Symptoms are often exacerbated by stress at school or novel social situations. The pain often is worst in the morning and often prevents or delays children from attending school. IBS is a subset of functional abdominal pain, characterized by onset of pain at the time of a change in stool frequency or consistency, and relief of pain with defecation. Symptoms in IBS are linked to gut motility.

2.**(D)** While waiting for laboratory and ultrasound results, a 3-day trial of a lactose-free diet can evaluate for lactose intolerance. If tests are normal and no warning signs are present, testing should be stopped. If there are warning signs, progression of symptoms, or laboratory abnormalities that suggest a specific diagnosis, additional investigation may be warranted. A child who is repeatedly kept home from school because of pain receives reinforcement in the form of being excused from responsibilities and withdraws from full social functioning. This tends to both increase anxiety and prolong the course.

3.**(C) Functional constipation**
- History: No history of significant neonatal constipation, onset at potty training, large-caliber stools, retentive posturing, may have encopresis
- Examination: Normal or reduced sphincter tone, dilated rectal vault, fecal impaction, soiled underwear, palpable fecal mass in left lower quadrant
- Laboratory: No abnormalities, barium enema would show dilated distal bowel

4.**(D)** Medications taken for a variety of conditions may cause oral abnormalities. Drugs with anticholinergic properties diminish saliva production and increase the risk of dental caries and parotitis. Tetracyclines taken before the eruption of the permanent teeth stain the enamel. Excessive fluoride in vitamin preparations or in drinking water can result in mottled teeth. Gingival hypertrophy may be caused by cyclosporine, phenytoin, and calcium channel blockers.

5.**(B)** Pill ulcers occur when certain medications (tetracyclines and nonsteroidal antiinflammatory drugs [NSAIDs]) are swallowed without sufficient liquids, allowing prolonged direct contact of the pill with the esophageal mucosa. Pill injury causes severe chest pain and often prominent odynophagia and dysphagia.

6.**(D)** Lye-based drain cleaners, especially liquid products, cause the worst injuries because they are swallowed easily and liquefy tissue rapidly. Full-thickness burns can occur in seconds. Granular products are less likely to cause esophageal injury during accidental

exposures because they burn the tongue and lips and often are expelled before swallowing. Less caustic agents, such as bleach and detergents, cause less serious injury.

7.**(D)** About 60% of children with malrotation present with symptoms of bilious vomiting during the first month of life. The remaining 40% present later in infancy or childhood. Plain abdominal x-rays generally show evidence of obstruction. Abdominal ultrasound may show evidence of malrotation. An upper gastrointestinal (GI) series shows the absence of a typical duodenal "C-loop," with the duodenum instead remaining on the right side of the abdomen. Abnormal placement of the cecum on follow-through (or by contrast enema) confirms the diagnosis.

8.**(B)** Duodenal atresia is associated with other anomalies in more than half of infants. Down syndrome is the most common associated disorder, but it can also be associated with biliary, cardiac, renal, or vertebral anomalies. Jejunal and ileal atresia can be seen in meconium ileus secondary to cystic fibrosis.

9.**(D)** Gastroesophageal reflux disease (GERD) can lead to substantial enamel erosion and caries. Neonatal hyperbilirubinemia can result in discoloration of the deciduous teeth. Renal failure is associated with mottled enamel of the permanent teeth. Congenital syphilis causes marked abnormalities in the shape of teeth, especially incisors and molars. Celiac disease can result in enamel defects. Crohn disease is associated with oral aphthous ulcers. Abnormal pigmentation of the lips and buccal mucosa is seen with Peutz-Jeghers syndrome (freckling) and Addison disease (hyperpigmented macules). Candidiasis is seen commonly with immunodeficiency disorders and diabetes. Leukemic infiltrates result in gum hyperplasia and bleeding; treatment of neoplastic conditions can cause severe mucositis. Some tumors, including lymphoma, may present as mass lesions of the buccal cavity

10.**(D)** Hirschsprung disease is a motility defect caused by failure of ganglion cell precursors to migrate into the distal bowel during fetal life. A deep rectal biopsy specimen obtained surgically or by using a suction biopsy instrument is required for diagnosis. When no ganglion cells are shown in the submucosal plexus, accompanied by nerve trunk hyperplasia, the diagnosis is certain. Barium enema and anorectal manometry may be used before biopsy, but false-negative and false-positive results can occur. Therapy is surgical.

11.**(B)** In patients with suspected IBD, upper endoscopy and colonoscopy are recommended. **Colonoscopic** findings in UC include diffuse carpeting of the distal or entire colon with tiny ulcers and loss of haustral folds. Within the involved segment, no skip areas are present. In CD, ulcerations tend to be much larger and deeper with a linear, branching, or aphthous appearance; skip areas are usually present.
Upper endoscopy cannot evaluate the jejunum and ileum but is more sensitive than contrast studies in identifying proximal CD involvement.
Other methods to detect small bowel involvement include **video capsule endoscopy**; **upper GI series with small bowel follow-through**; **computed tomography (CT) scanning**, which can detect small bowel disease as well as abscesses; and **MR enterography**, which has the advantage of no radiation and good sensitivity for finding active bowel disease.

12.**(A)** The American Academy of Pediatrics recommends that primary care physicians apply fluoride varnish to the teeth of all children every 3–6 months, starting at the emergence of the first tooth. Fluoride varnish is a concentrated fluoride preparation that adheres to the teeth on contact with saliva. It is painted onto the teeth with a small brush, after first drying the tooth with a gauze pad, and has a prolonged cavity prevention effect.

13.**(A)** Clinical manifestations depend on the region of involvement. UC involves only the colon, whereas CD can include the entire gut from mouth to anus. Symptoms can be subtle in CD. Small bowel involvement in CD is associated with loss of appetite, crampy postprandial pain, poor growth, delayed puberty, fever, anemia, and lethargy. Symptoms may be present for some time before the diagnosis is made. Severe CD with fibrosis may cause partial or complete small bowel obstruction. Perineal abnormalities, including skin tags and fistulas, are another feature distinguishing CD from UC. Other extraintestinal manifestations of CD include arthritis, erythema nodosum, and uveitis.

14.**(A)** Surgical closure of the cleft lip is usually done by 3 months of age. Closure of the palate follows, usually before 1 year of age.

15.**(B)** Therapy of intussusception must begin with placement of an IV catheter and a nasogastric tube. Before radiologic intervention is attempted, the child in this scenario must have adequate fluid resuscitation to correct the often severe dehydration caused by vomiting and third space losses. Ultrasound may be performed before the fluid resuscitation is complete. Surgical consultation should be obtained early as the surgeon may prefer to be present during non-operative reduction. If pneumatic or hydrostatic reduction is successful, the child should be admitted to the hospital for overnight observation of possible recurrence (risk is 5–10%). If reduction is not complete, emergency surgery is required.

16.**(A)** Serologic markers include antiendomysial, deamidated antigliadin, and tissue transglutaminase IgA antibodies. Because IgA deficiency may be associated with celiac disease, total serum IgA also must be measured to document the accuracy of these tests. In the absence of IgA deficiency, either test yields a sensitivity and specificity of 95%. An endoscopic small bowel biopsy is essential to confirm the diagnosis and should be performed while the patient is still ingesting gluten.

17.**(A)** Environmental factors during gestation also increase risk, including drugs (phenytoin, valproic acid, thalidomide), maternal alcohol and tobacco use, dioxins and other herbicides, and possibly high altitude. Chromosomal and nonchromosomal syndromes are associated with clefting, as are specific genes in some families.

18.**(C)** Alagille syndrome is characterized by chronic cholestasis with the unique liver biopsy finding of paucity of bile ducts in the portal triads. Associated abnormalities in some (syndromic) types include peripheral pulmonic stenosis or other cardiac anomalies; hypertelorism; unusual facies with deep-set eyes, prominent forehead, and a pointed chin; butterfly vertebrae; and a defect of the ocular limbus (posterior embryotoxon).

19.**(A)** 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 1hour 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.

20.**(C)** Physiologic GER ("spitting up") is normal in infants. Nearly half of all infants are reported to spit up between 2 and 4 months of age. Infants who regurgitate meet the criteria for physiologic GER so long as they maintain adequate nutrition and have no signs of respiratory complications or esophagitis. Contributing factors of infantile GER include liquid diet; horizontal body position; short, narrow esophagus; small, noncompliant stomach; frequent, relatively large volume feedings; and an immature lower esophageal sphincter (LES). As infants grow, they spend more time upright, eat more solid foods, develop a longer and larger diameter esophagus, have a larger and more compliant stomach, and experience lower caloric needs per unit of body weight. As a result, most infants stop spitting up by 9–12 months of age.

21.**(B)** Wilson disease is characterized by abnormal storage of copper in the liver, leading to hepatocellular injury, CNS dysfunction, and hemolytic anemia. Clinical presentation varies but seldom occurs before age 3 years. Neurologic abnormalities may predominate, including tremor, decline in school performance, worsening handwriting, and psychiatric disturbances. Hemolytic anemia may be the first noted symptom. Hepatic presentations include appearance of jaundice, spider hemangiomas, portal hypertension and its consequences, and fulminant hepatic failure. The diagnosis is made by identifying low serum levels of ceruloplasmin, elevated 24-hour urine copper excretion, the presence of Kayser-Fleischer rings in the iris, evidence of hemolysis, and elevated hepatic copper content. In any single patient, one or more of these measures may be normal.

22.**(C)** The most common forms of TEF occur with esophageal atresia; the "H-type" TEF without atresia is uncommon, as is esophageal atresia without TEF.

- ✓ Esophageal atresia with distal TEF (85%)
- ✓ Esophageal atresia with no TEF (8%)
- ✓ H-type TEF (4%)
- ✓ Esophageal atresia with proximal TEF (2%)
- ✓ Esophageal atresia with proximal and distal TEF (1%)

23.**(B)** Associated defects include the VACTERL association—vertebral anomalies (70%), anal atresia (imperforate anus) (50%), cardiac anomalies (30%), TEF (70%), renal anomalies (50%), and limb anomalies (polydactyly, forearm defects, absent thumbs, syndactyly) (70%). A single-artery umbilical cord is often present. Infants with esophageal atresia have a history of polyhydramnios, exhibit drooling, and have mucus and saliva bubbling from the nose and mouth. Patients with a TEF are vulnerable to aspiration pneumonia. When TEF is suspected, the first feeding should be delayed until a diagnostic study is performed.

24.**(A)** Eosinophilic esophagitis this chronic immune-mediated disorder is characterized by infiltration of eosinophils into the mucosa of the esophagus. It is more common in males than females and in patients with history of atopy. In young children, it may present with oral aversion, vomiting, and failure to thrive. In school-age children, it may present with vague abdominal pain or vomiting. In adolescents and adults, it presents with dysphagia and food impactions. These symptoms are attributed to the inflammatory response in the esophagus leading to edema and poor esophageal motility.

25.**(A)** The simplest test for TEF is to gently attempt to place a 10F or larger tube via the mouth into the stomach. The passage of the tube is blocked at the level of the atresia. A chest x-ray reveals the tube coiled in the esophageal pouch. Air can be injected through the tube to outline the atretic pouch. Barium should not be used because of extreme risk of aspiration, but a tiny amount of dilute water-soluble contrast agent can be given carefully, then suctioned when the defect is clearly shown.

26.**(D)** Children with functional fecal retention often exhibit “retentive posturing” (standing or sitting with legs extended and stiff or crossed legs) and have associated fecal incontinence caused by leakage of retained stool (encopresis). Young children with painful defecation must have a prolonged course of stool softener therapy to alleviate fear of defecation. The child should be asked to sit on the toilet for a few minutes on awakening in the morning and immediately after meals, when the colon is most active and it is easiest to pass a stool. Use of a positive reinforcement system for taking medication and sitting on the toilet is helpful for younger children. The stool softener chosen should be non–habit forming, safe, and palatable. Polyethylene glycol and milk of magnesia are the most commonly used agents and because of prolonged fecal impaction perform a mineral oil enema.

27.**(A)** Young children often place nonfood items in the mouth. When these items are swallowed, they may become lodged in the esophagus at the thoracic inlet or at the LES. The most common objects are coins. Smaller coins may pass harmlessly into the stomach, where they rarely cause symptoms. Other common esophageal foreign bodies include food items, small toys or toy parts, button batteries, and other small household items. Children with a prior history of esophageal atresia or with poor motility secondary to GER or EoE are more prone to food impactions, which seldom occur in the normal esophagus.

28.**(A)** Pyloric stenosis occurs in the first 1–2 months of life and is characterized by steadily worsening, forceful vomiting that occurs immediately after feedings. A visibly distended

stomach, often with visible peristaltic waves, is often seen before vomiting. Pyloric stenosis is more common in male infants; the family history may be positive.

29.**(C)** A common cause of chronic loose stools in early childhood is functional diarrhea, commonly known as toddler's diarrhea. This condition is defined by frequent watery stools in the setting of normal growth and weight gain and is caused by excessive intake of sweetened liquids overwhelming the disaccharide absorptive capacity of the intestine. Diarrhea typically improves tremendously when the child's beverage intake is reduced or changed.

30.**(C)** Tests of liver dysfunction include total and direct bilirubin, alanine aminotransferase, aspartate aminotransferase for evidence of hepatocellular injury, and γ-glutamyltransferase or alkaline phosphatase for evidence of bile duct injury. Hepatic synthetic function can be assessed by coagulation factor levels, prothrombin time, and albumin level.

31.**(A)** A clinical diagnosis is often sufficient in children with classic effortless regurgitation and no complications (physiologic GER).

32.**(C)** A barium upper gastrointestinal (GI) series helps to rule out gastric outlet obstruction, malrotation, or other anatomic contributors to GER. Because of the brief nature of the examination, a negative barium study does not rule out GER, nor does it rule it in as it is normal to have some reflux into the esophagus many times per day. A 24- hour esophageal pH probe monitoring uses a pH electrode placed transnasally into the distal esophagus, with continuous recording of esophageal pH. Data typically are gathered for 24hours and analyzed for the number and temporal pattern of acid reflux events. Esophageal impedance monitoring records the migration of electrolyte-rich gastric fluid in the esophagus. Endoscopy is useful to evaluate for esophagitis, esophageal stricture, and anatomic abnormalities.

33.**(A)** The lower central incisors are typically the first to erupt, followed by the upper central incisors, lateral incisors, first molars, and bicuspids. Delayed eruption may occur in association with hypopituitarism, hypothyroidism, osteopetrosis, Gaucher disease, Down syndrome, cleidocranial dysplasia, and rickets. Deciduous teeth begin to be replaced by the permanent teeth at around age 6 years. The sequence of replacement is similar to that of the appearance of deciduous teeth.

34.**(D)** The location of referred pain is predictable based on the locus of visceral injury. Stomach pain is referred to the epigastric and retrosternal regions, and liver and pancreas pain is referred to the epigastric region. Gallbladder pain often is referred to the region below the right scapula. Somatic pathways stimulated by small bowel visceral afferents affect the periumbilical area, and colonic injury results in infraumbilical referred pain.

35.**(D)** Children with functional abdominal pain characteristically have pain almost daily. The pain is not associated with meals or relieved by defecation and is often associated with a

tendency toward anxiety and perfectionism. Symptoms are often exacerbated by stress at school or novel social situations. The pain often is worst in the morning and often prevents or delays children from attending school. To break the cycle of pain and disability, the child with functional pain must be assisted in returning to normal activities immediately. Instead of being sent home from school with stomachaches, a child may be allowed to take a short break from class until symptoms abate. The child and parents should be informed that pain is likely to be worse on the day the child returns to school as anxiety worsens dysmotility and enhances pain perception. Medications may be helpful but not shown to have significant benefit in children with functional abdominal pain.. Probiotics and peppermint oil can be beneficial in treating IBS. Dietary changes such as the low FODMAP (fermentable olio-di-mono-saccharides and polyols) diet (reducing highly fermentable sugars in the diet) can be beneficial in a subset of patients.

36.**(D)** Coagulation tests and serum albumin are used to follow hepatic synthetic function. These tests are confounded by administration of blood products and clotting factors. Vitamin K should be administered to maximize the liver's ability to synthesize factors II, VII, IX, and X. In addition to monitoring prothrombin time and partial thromboplastin time, many centers measure factor V serially as a sensitive index of hepatic synthetic function. The INR (international normalized ratio) reflects liver synthetic function (~ to prothrombin time) and predicts the severity of hepatic failure. Renal function tests, electrolytes, serum ammonia, blood counts, and urinalysis also should be followed. In the setting of acute liver failure, liver biopsy may be indicated to ascertain the nature and degree of injury and estimate the likelihood of recovery.

37.**(C)** As acute pancreatitis progresses, the amylase level tends to decline faster than lipase, making the latter a good choice for diagnostic testing late in the course of the disease. Fecal elastase is the preferred test for pancreatic insufficiency.

38.**(C)** Peritonitis is characterized on examination by marked abdominal tenderness. Rebound tenderness also generally is quite pronounced. The patient tends to move very little owing to intense peritoneal irritation and pain. Fever is not always present, and absence of fever should not be regarded as contradictory to the diagnosis. Patients who are taking corticosteroids for an underlying condition, such as nephrotic syndrome, are likely to have little fever and reduced tenderness. Spontaneous bacterial peritonitis is most commonly caused by Streptococcus pneumoniae or by Escherichia coli.

SECTION 18

THE RESPIRATORY SYSTEM

MARYAM Z ALMUSAWI

QUESTIONS

1. The lymphoid tissue that can obstruct airflow through the nasopharynx, if enlarged is called
   A. tonsils
   B. adenoids
   C. parapharyngeal lymph nodes
   D. retropharyngeal lymph nodes

2. What is the narrowest portion of the airway in children under 3 years of age?
   A. Vocal fold
   B. Nasopharynx
   C. Cricoid ring
   D. Glottis

3. The growth of new alveoli occurs during the first 2 years of life and is complete by
   A. 6 years of age
   B. 8 years of age
   C. 10 years of age
   D. 12 years of age

4. The volume of gas retained in the lung at the end of a relaxed exhalation is the
   A. functional residual capacity (FRC)
   B. residual volume (RV)
   C. tidal volume (TV)
   D. vital capacity (VC)

5. Which of the following disorders resulting in V/Q mismatching?
   A. Apnea
   B. Narcotic overdose
   C. Pulmonary fibrosis
   D. Pneumonia

6. A repetitive, staccato cough is usually seen in
   A. Chlamydial infections
   B. asthma
   C. gastroesophageal reflux
   D. pertussis

7. What is the definition of hyperpnea?
   A. Increased respiratory rate
   B. Increased depth of breathing and normal rate
   C. Increased rate with regular deep respiration

D. Rapid, deep respiration followed by apnea

8. Which of the following disease processes is characterized by increased vocal fremitus?
A. Consolidation
B. Pleural effusion
C. Atelectasis
D. Pneumothorax

9. Computed tomography (CT) of the chest is the imaging tool of choice for evaluating
A. great vessel anatomy
B. mediastinal lesions
C. parapneumonic effusion
D. bronchiectasis

10. Ultrasonography is the imaging procedure of choice for assessing
A. parapneumonic effusion
B. interstitial lung disease
C. bronchiectasis
D. mediastinal lesions

11. The PCO_2 in venous samples is higher than arterial PCO_2 by approximately
A. 4 mm Hg
B. 6 mm Hg
C. 8 mm Hg
D. 10 mm Hg

12. Lung volumes and airflow rates using spirometry are compared to predicted values of the patient that rely mostly on
A. age
B. gender
C. race
D. height

13. Rigid bronchoscopy is the method of choice for
A. identifying stenosis
B. identifying endobronchial lesions
C. removing foreign bodies from the airways
D. obtaining airway samples for culture

14. A 5-year-old boy presented with shortness of breath and wheezy chest, you suspect asthma and order a chest Xray.
What is the best indicator of hyperinflation in AP view for this patient?
A. Hyperlucent lungs
B. Viewing 7 posterior ribs
C. Viewing 5 anterior ribs
D. Flattened diaphragm

15. While you are giving a clinical session to the 6th grade medical students, one asked about the type of breathing for a 6-year-old boy admitted to emergency ward for treatment of meningitis describing his respiration as rapid, deep, and followed by apnea.
What is the **BEST** term for description of this type of breathing?

A. Biot respiration
B. Periodic breathing
C. Apneustic respiration
D. Cheyne- Stokes respiration

16. A 9 -month- boy with SMA type 1 presented with respiratory distress. You decide to put him on mechanical ventilator.
Which of the following gives good information about his ventilation and oxygenation?

A. Arterial blood gas analysis
B. Venous blood gas analysis
C. Capillary blood gas analysis
D. Pulse oximetry reading

17. A 9-year-old boy presented with history of night dry cough that disturb his sleep, increased with exercise, your examination revealed a well-nourished oriented boy, his respiratory system examination showed symmetrical chest movement, resonant percussion sounds, normal vesicular breathing, with end expiratory wheeze.
What is your **EXPECTED** spirometry finding?

A. FVC % predicted < 60 %
B. FVC/FEV1 ratio > 80 %
C. FEV 1% predicted improved >10% after bronchodilator inhalation
D. FEF 25-75% improved >25% after bronchodilator inhalation

18. Which method of oxygenation can give fixed FiO_2?

A. Nasal cannula
B. Face mask
C. Venturi mask
D. Non rebreather mask

19. What is the best delivery method of inhaled steroid for a 4-year-old asthmatic girl?

A. Dry powder inhaler
B. Nebulizer
C. Spacer with mouth piece
D. Spacer with facemask

20. A 7-month-boy brought to your clinic after visiting many doctors because of the child noisy breathing that started shortly after birth. On examination; the baby is thriving well, having stridor, mild suprasternal retraction and occasional cough, these findings became worse with crying and feeding.
Of the following, the **MOST** appropriate management is

A. reassurance
B. budesonide nebulizer
C. adrenaline nebulizer

D. systemic steroid

21. A 17-month-old girl with recurrent episodes of monophonic expiratory wheeze and barking cough that started since age of 4 months with poor response to asthma treatment, her mother report that she always had noisy breathing, no dysphagia, stridor or any added heart sounds.
Of the following, the **MOST** appropriate management is

A. chest CT scan
B. echocardiograph
C. flexible bronchoscope
D. upper GI barium study

22. What is the **MOST** common indication for tracheostomy in children?

A. Upper airway obstruction
B. Prolonged mechanical ventilation
C. Tracheobronchomalacia
D. Extensive Staphylococcal pneumonia

23.A 4-month-old Infant presented with respiratory difficulties within the first weeks of life in the form of profound loss of respiratory control during sleep leading to apnea, hypercarbia, and hypoxemia. The mother gave history of chronic constipation, abdominal distention, occasional vomiting, and failure to thrive.
Of the following, the **MOST** likely diagnosis is

A. Hirschsprung disease
B. Prader-Willi syndrome
C. Arnold-Chiari malformation
D. congenital central hypoventilation syndrome

24. An 18-month-old healthy looking boy suddenly developed cessation of breathing, syncope, brief tonic-clonic movements associated with cyanosis and loss of consciousness. This is the third attack for him during this month, these attacks occur after crying or agitation for any cause.
Of the following, the **MOST** likely diagnosis is

A. seizure disorder
B. congenital central hypoventilation syndrome
C. breath holding spells
D. hyper cyanotic spells

25. A 7-year-old obese boy is complaining from snoring, restless sleep, poor school performance, behaviour problems, morning headache, and mouth breathing.
Of the following, the **MOST** appropriate treatment is

A. tracheostomy
B. CPAP
C. supplemental oxygen
D. adenotonsillectomy

26. A 6-month-old healthy looking boy suddenly developed cyanosis, irregular breathing, floppiness, and lethargy lasted for about 30 seconds. Careful history and physical examination failed to show any evident cause.
Of the following, the **MOST** likely cause is

A. apparent life-threatening event (ALTE)
B. brief resolved unexplained event (BRUE)
C. gastroesophageal reflux
D. seizure disorder

27. Which of the following may be considered in the work up of low-risk infant with BRUE?

A. Metabolic panel
B. Complete blood count
C. EEG
D. Pertussis screening

28. Which of the following increases the risk of SIDS?

A. Female gender
B. Low birthweight infant
C. Alcoholic mother
D. Previous attack of BRUE

29. Which of the following is a relatively common cause of sudden unexpected infant death?

A. Laryngospasm
B. Seizure disorder
C. Hemosiderosis/pulmonary hemorrhage syndrome
D. Cardiac arrhythmia

30. What is the **MOST** common sub-acute upper airway obstruction in infants?

A. Chronic rhinitis
B. Laryngomalacia
C. Subglottic stenosis
D. Airway hemangioma

31. What is the **MOST** common sub-acute upper airway obstruction in toddlers?

A. Recurrent rhinitis
B. Hypertrophied tonsils and adenoids
C. Spasmodic croup
D. Laryngeal papillomatosis

32. Radiographic evaluation of a child with stridor is rarely helpful. Which of the following conditions may have steeple sign?

A. Epiglottitis
B. Bacterial tracheitis
C. Angioedema
D. Spasmodic croup

33. A 6-week-old infant presented with cyanosis, nasal discharge, significant respiratory distress, especially when feeding. The cyanosis is usually relieved by crying.

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

A. nasal polyps
B. nasal septal deviation
C. nasal septal hemangioma
D. choanal atresia

34. A 4-month-old healthy looking boy presented with inspiratory stridor loudest when the infant is feeding or active and decreases when the infant is relaxed or placed prone, or when the neck is flexed.
Of the following, the **MOST** appropriate therapy for this baby is

A. proton pump inhibitors
B. supraglottoplasty
C. tracheostomy
D. watchful waiting

35. A 4-year-old boy presented with snoring, mouth breathing, and recurrent otitis media.
Of the following, the **MOST** likely cause is

A. adenoidal hypertrophy
B. laryngeal hemangioma
C. nasal polyps
D. unilateral choanal atresia

36. What is the **MOST** common lower airway disease in children?

A. Cystic fibrosis
B. Asthma
C. Bronchiolitis
D. Bronchitis

37. What is the diagnostic procedure of choice for compression of the trachea by vascular structures?

A. Barium swallow
B. Bronchoscopy
C. Computed tomography (CT) angiogram
D. Video fluoroscopic swallowing study

38. A 5-year-old boy presented with impaired growth, early onset, year-round nasal congestion and wet cough, recurrent middle ear effusions with associated hearing loss, chronic sinusitis, and nasal polyps.
Of the following, the **MOST** sensitive and specific diagnostic test for this boy is

A. ultrastructural analysis of respiratory cilia
B. measurement of nasal nitric oxide
C. high-speed video microscopy
D. high-resolution chest CT scans

39. Which of the following may reduce the number of exacerbations per year in a child with primary ciliary dyskinesia?

A. Chest physiotherapy

B. Inhaled hypertonic saline
C. Low-dose macrolides
D. Prompt treatment of bacterial infections

40. Pulmonary arterial hypertension in children is defined as a mean pulmonary artery pressure of
A. 20 mm Hg or higher
B. 30 mm Hg or higher
C. 40 mm Hg or higher
D. 50 mm Hg or higher

41. Which of the following is a presenting symptom of corpulmonale?
A. Orthopnea
B. Unexplained syncope
C. Chronic cough
D. Excessive sweating

42. Which of the following investigations gives a definitive diagnosis of corpulmonale?
A. Echocardiography
B. Cardiac magnetic
C. Lung biopsy
D. Cardiac catheterization

43. A 3-year-old boy presented with recurrent attacks of profound respiratory symptoms, hypoxemia, diffuse infiltrates on radiographs, and minimal hemoptysis. Blood film showed iron-deficiency anemia, while bronchoalveolar lavage revealed hemosiderin-laden macrophages.
Of the following, the **MOST** likely diagnosis is
A. Wegener granulomatosis
B. pulmonary capillaritis
C. idiopathic pulmonary hemosiderosis
D. bronchopulmonary sequestration

44. Which of the following procedures provide accurate diagnosis of pulmonary embolism in children?
A. Ventilation-perfusion scans
B. Catheter pulmonary angiography
C. Doppler ultrasonography
D. CT pulmonary angiography

45. Which of the following causes false-negative results on sweat testing of cystic fibrosis?
A. Eczema
B. Hypothyroidism
C. Malnutrition
D. Edema

46. Which of the following is the standard diagnostic test for cystic fibrosis?

A. DNA analysis
B. Sweat chloride test
C. Immunoreactive trypsinogen (IRT) levels
D. Fecal elastase levels

47. Which of the following cystic fibrosis transmembrane conductance regulator (CFTR) modulators demonstrated improvement in lung function and symptom-related quality of life, and decreased sweat chloride in cystic fibrosis patients?
A. Tezacaftor-ivacaftor.
B. Elexacaftor-tezacaftor-ivacaftor
C. Ivacaftor
D. Ivacaftor and lumacaftor

48. Antibiotic therapy is important in controlling chronic infection in cystic fibrosis. Antibiotics are selected based on organisms identified by sputum culture. If patients are unable to provide sputum. Which of the following cultures can be used to direct antibiotic therapy?
A. Throat
B. Blood
C. Broncho-alveolar lavage
D. Fine needle aspirate

49. Distal intestinal obstruction syndrome in CF patients beyond the neonatal period may need to be treated with
A. oral laxatives (polyethylene glycol)
B. paraffin enema
C. high pancreatic enzyme dosage
D. dietary fiber

50. Which of the following imaging study is the **BEST** for differentiating air within the lung parenchyma (cystic lung disease) from air in the pleural space?
A. Upright chest radiographs
B. Computed tomography (CT) scans of chest
C. Ultrasonography of chest
D. MRI of chest

51. Which of the following pneumothoraces may not require intervention as they often resolve spontaneously?
A. <20% of thorax occupied with pleural air
B. <25% of thorax occupied with pleural air
C. <30% of thorax occupied with pleural air
D. <35% of thorax occupied with pleural air

52. What is the **MOST** common cause of parapneumonic effusions?
A. Hemophilus influenza
B. Streptococcus pneumoniae
C. Staphylococcus aureus

D. TB

53. What is the most common bacterial species encountered in the airways of patients with CF in all ages?
A. Haemophilus influenzae
B. Staphylococcus aureus
C. Pseudomonas aeruginosa
D. Burkholderia cepacia complex species

54. When do respiratory symptoms typically appear in individuals with CF?
A. During neonatal period
B. In first few years of life
C. In late childhood
D. During adolescence

55. What is the initial respiratory symptom of CF in young infants?
A. Productive cough
B. Wheezing
C. Shortness of breath
D. Chest pain

56. What is the first discernible change seen in chest radiographs in CF patients?
A. Cyst formation
B. Collapse
C. Hyperinflation
D. Consolidation

SECTION 18

THE RESPIRATORY SYSTEM

MARYAM Z ALMUSAWI

ANSWERS

1.**(B)** Enlarged lymphoid tissue can obstruct airflow through the nasopharynx (adenoids) or the posterior pharynx (tonsils).

2.**(C)** In children under 3 years of age, the cricoid ring is the narrowest portion of the airway. In older children and adults, it is the glottis.

3.**(B)** The growth of new alveoli occurs during the first 2 years of life and is complete by 8 years of age. After this time, lung volume increases primarily by increase in alveolar dimensions, with new alveoli rarely formed.

4.**(A)** Tidal volume (TV) is the amount of air inspired with each relaxed breath. The volume of gas retained in the lung at the end of a relaxed exhalation is the functional residual capacity (FRC). This gas volume maintains exchange of O2 between breaths. Total lung capacity (TLC) is the volume of gas in the lungs at the end of maximal inhalation, and residual volume (RV) is the volume of gas left in the lungs at the end of a maximal exhalation. Vital capacity (VC) is the maximal amount of air that can be forcibly expelled from the lungs and is the difference between TLC and RV.

5.**(D)**

TABLE 133.1 Causes of Hypoxemia

CAUSE	EXAMPLE(S)	Pao_2	$Paco_2$	Pao_2 IMPROVES WITH SUPPLEMENTAL OXYGEN
Ventilation-perfusion mismatch	Asthma Bronchopulmonary dysplasia Pneumonia Atelectasis	↓	Normal, ↓, or ↑	Yes
Hypoventilation	Apnea Narcotic overdose Neuromuscular disease	↓	↑	Yes
Extrapulmonary shunt	Cyanotic heart disease	↓	Normal or ↑	No
Intrapulmonary shunt	Pulmonary arteriovenous malformation Atelectasis	↓	Normal or ↑	No
Low F_{IO_2}	High altitude	↓	↓	Yes
Diffusion defect	Scleroderma Hepatopulmonary syndrome Pulmonary fibrosis	↓	Normal	Yes

F_{IO_2}, Fraction of inspired oxygen; $Paco_2$, arterial partial pressure of carbon dioxide; Pao_2, arterial partial pressure of oxygen.

6.**(A)** Night time cough is a hallmark of asthma and can also be caused by gastroesophageal reflux disease or postnasal drip. Cough with exercise is highly suggestive of exercise-induced asthma/bronchospasm. Habit cough can masquerade as organic disease but should disappear
when children are distracted or during sleep. Paroxysmal cough (especially associated with cyanosis) suggests pertussis, whereas a repetitive, staccato cough occurs in chlamydial infections in infants. A harsh, brassy, seal-like cough suggests croup, tracheomalacia, or

psychogenic (habit) cough. Sudden onset of cough after a choking episode suggests foreign body aspiration.

7.**(B)**

TABLE 133.3 Abnormal Breathing Patterns

PATTERN	FEATURES	CAUSES
Tachypnea	Respiratory rate > normal for age	Restrictive physiology, fever, increased metabolic demands, stress
Bradypnea	Respiratory rate < normal for age	Sleep, sedating medications, CNS injury, metabolic alkalosis
Hyperpnea	Increased depth of breathing, normal rate	Stress, exercise, metabolic acidosis
Periodic breathing	Brief pauses (<10 sec) followed by rapid, shallow breaths	Normal variant for preterm and term infants, prominent in sleep
Kussmaul respiration	Increased rate, increased tidal volume, regular deep respiration	Metabolic acidosis, especially diabetic ketoacidosis, uremia
Cheyne-Stokes respiration	Cyclic pattern of waxing and waning of depth of breathing interposed with apnea	CNS injury (brainstem), elevated intracranial pressure, heart failure, uremia
Biot respiration	Rapid, deep respiration followed by apnea	CNS injury or infection
Apneustic respiration	Long inspiration with short or staccato expiration	Brainstem lesion
Agonal respiration	Slow rate, variable tidal volume	Shock, sepsis, or asphyxia

CNS, Central nervous system.

8.**(A)**

TABLE 133.4 Physical Signs of Pulmonary Disease

DISEASE PROCESS	MEDIASTINAL DEVIATION	CHEST MOTION	VOCAL FREMITUS	PERCUSSION	BREATH SOUNDS	ADVENTITIOUS SOUNDS	VOICE SIGNS
Consolidation	No	Reduced over area	Increased	Dull	Bronchial or reduced	None or crackles	Egophony* Pectoriloquy†
Bronchospasm	No	Hyperexpansion with limited motion	Normal or decreased	Hyperresonant	Normal to decreased	Wheezes, crackles	Normal to decreased
Atelectasis	Shift toward affected side	Reduced over area	Decreased	Dull	Reduced	None or crackles	None
Pneumothorax	With tension: shift to the opposite side	Reduced over PMI	None	Resonant	None	None	None
Pleural effusion	If large: shift to opposite side	Reduced over area	None or reduced	Dull	None	Egophony Friction rub	Muffled
Interstitial process	No	Reduced	Normal to increased	Normal	Normal	Inspiratory crackles	None

9.**(D)** Computed tomography (CT) of the chest is the imaging tool of choice for evaluating masses, interstitial lung disease, and bronchiectasis, as well as delineating pleural from parenchymal lesions.

10.**(A)** Ultrasonography can be used to delineate some intrathoracic masses and is the imaging procedure of choice for assessing size and character of parapneumonic effusion/empyema. It is also useful for assessing diaphragmatic motion in small children.

11.**(B)** The partial pressure of carbon dioxide (Pco2) from a capillary sample is similar to that from arterial blood. The Pco_2 in venous samples is approximately 6 mm Hg higher than arterial Pco_2.

12.**(D)** During the forced expiratory manoeuvre, forced vital capacity (FVC), forced expired volume in the first second (FEV1), and forced expiratory flow (FEF) rates are measured. These are compared to predicted values based on patient age, gender, and race, but rely mostly on height.

13.**(C)** Flexible bronchoscopy is useful in identifying dynamic or static airway abnormalities (stenosis, malacia, endobronchial lesions, or excessive secretions) and to obtain airway samples for culture (bronchoalveolar lavage). Rigid bronchoscopy is the method of choice for removing foreign bodies from the airways and performing other interventions, such as airway dilation.

14.**(D)** flattened diaphragms and an increased AP diameter on lateral projection are better indicators of hyperinflation.

15.**(A)** Biot respiration is a rapid, deep respiration followed by apnea usually seen in CNS injury or Infection.

16.**(A)** A properly performed arterial blood gas analysis provides information about the effectiveness of both oxygenation and ventilation. Capillary or venous samples should not be used to assess oxygenation. The partial pressure of carbon dioxide (Pco2) from a capillary

sample is similar to that from arterial blood. The Pco2 in venous samples is approximately 6 mm Hg higher than arterial PCO_2.

17.**(D)** Spirometry can detect reversible airway obstruction characteristics of asthma when a significant improvement in FEV1 (>12%) or in FEF25–75% (>25%) following inhalation of a bronchodilator is measured.

18.**(C)**

19.**(D)**

20.**(A)** Laryngomalacia.

21.**(C)** Tracheobronchomalacia.

22.**(A)** Congenital or acquired upper airway obstruction is the most common indication for tracheostomy in children. In addition, if prolonged mechanical ventilation is required, elective tracheostomy can be performed to increase patient comfort and facilitate nursing care.

23.**(D)** Congenital central hypoventilation syndrome (CCHS) is a rare genetic disorder in which there is profound loss of respiratory control during sleep leading to primary central apnea, hypercarbia, and hypoxemia. Individuals with CCHS have a defect in the PHOX2B gene, which is necessary for autonomic nervous system development. Infants with CCHS typically have respiratory difficulties within the first several weeks of life although it can also present later in childhood. Some PHOX2B mutation types are associated with an increased risk of Hirschsprung disease and neural crest tumors (neuroblastoma).

24.**(C)** Breath holding spells are a form of apnea occurring when a young child is awake, usually triggered by emotional stress. Breath holding spells typically occur in children under 3 years
of age during periods of agitation or crying and can be associated with cyanosis and loss of consciousness. The return of spontaneous breathing usually occurs quickly. Most breath holding spells are benign and lessen over time, but seizures and disorders of central respiratory control should be considered.

25.**(B)** Targeted treatment of obstructive sleep apnea syndrome OSA starts with determining whether the child will benefit from adenotonsillectomy. If surgical intervention is not indicated or fails to alleviate the problem, then the use of continuous positive airway pressure (CPAP) or bilevel positive airway pressure (BiPAP) via mask interfaces can be used to distend the upper airway during sleep. This requires a well-fitted nasal mask, which may not be well tolerated in young children. Supplemental oxygen can blunt hypoxemia in milder cases of OSA but does not alter obstruction and sleep fragmentation. In extreme cases, especially those associated with craniofacial abnormalities or hypotonia, tracheostomy may be indicated.

26.**(B)** A brief resolved unexplained event (BRUE) is defined as a short (<1 minute), self-limited episode with one or more of the following features: (1) cyanosis or pallor, (2) absent or irregular breathing, (3) change in tone, or (4) altered level of responsiveness in an infant under 1 year of age. Importantly, BRUE differs from the previously used diagnostic term apparent life-threatening event (ALTE) in that it includes only episodes with cyanosis (not rubor) and/or altered responsiveness and does not include caregiver perception of the event as "life threatening." The label BRUE should be used only when no other explanation for the event is identified on detailed history and physical examination.

27.**(D)** Infants over 2 months of age and born at term or born at more than 32 weeks gestation and currently 45 weeks gestational age who experience a single, brief (<1 minute), self-limited (no cardiopulmonary resuscitation [CPR] required) BRUE are considered at low risk for undiagnosed serious condition or repeat event. An electrocardiogram and pertussis screening may be considered in this low-risk group. Additional diagnostic tests (e.g., metabolic panel, complete blood count, imaging, PSG, and EEG) and hospital admission for cardiorespiratory monitoring are not recommended.

28.**(B)** The risk of SIDS is higher in male, premature, and low birthweight infants; infants born to impoverished mothers; and mothers who smoke cigarettes or use illicit drugs. The risk of SIDS is increased three- to fivefold in siblings of infants who have died of SIDS and is highest during the winter. Importantly, there is no association between BRUE/ALTE and SIDS.

29.**(B)** Sudden unexpected infant death (SUID) is a term to describe any sudden and unexpected death occurring in the first year of life without an obvious cause before further investigation; this term includes those deaths caused by SIDS as well as those with subsequently identified explanation.

30.**(B)**

INFANT

- Chronic or recurrent rhinitis (infection, acid reflux, irritant)
- Laryngomalacia (most common)
- Subglottic stenosis (congenital or acquired [e.g., after intubation])
- Laryngeal web or cyst
- Laryngeal papillomatosis
- Airway hemangioma
- Vascular rings/slings

31.**(B)**

TODDLER

- Chronic or recurrent rhinitis (infection, allergy, irritant)
- Hypertrophied tonsils and adenoids (most common)
- Spasmodic croup
- Laryngeal papillomatosis
- Vascular rings/slings

32.**(C)**

Epiglottitis = Thumb sign
Bacterial tracheitis = Ragged tracheal border
Spasmodic croup = Often normal

33.**(D)** Choanal stenosis/atresia is a congenital problem presenting in the neonatal period. It may be bilateral or unilateral and is relatively rare. Neonates are generally obligate nose breathers, so obstruction of nasal passages can cause significant respiratory distress, especially when feeding. Crying by passes the obstruction because crying infants breathe through the mouth. Inability to easily pass a small catheter through the nostrils should raise the suspicion of choanal atresia. The diagnosis is confirmed by CT scan and by inspecting the area directly with a flexible nasopharyngoscope. An oral airway may be useful in the short term, but the definitive treatment is surgery.

34.**(D)** In most cases, no therapy is required for laryngomalacia. The infant should be observed closely during times of respiratory infection for evidence of respiratory compromise. Medical management for moderate to severe laryngomalacia can include acid suppression for those infants with suspected gastroesophageal reflux, feeding therapy, and optimization of nutritional status. Infants with severe laryngomalacia resulting in hypoventilation, hypoxia, or growth failure may benefit from a surgical procedure (e.g., supraglottoplasty) or, in extreme cases, a tracheostomy to bypass the upper airway.

35.**(A)** The signs of adenoidal and tonsillar hypertrophy are mouth breathing, snoring, and, in some patients, obstructive sleep apnea. The eustachian tubes enter the nasopharynx at the choanae and can be obstructed by enlarged adenoids, predisposing to recurrent or persistent otitis media.

36.**(B)**

37.**(C)** The diagnosis of vascular anomalies can often be made by a barium swallow, which identifies the esophageal compression. Bronchoscopy will identify a pulsatile compression of the airway, but the diagnostic procedures of choice are a computed tomography (CT) angiogram of the chest and great vessels or magnetic resonance angiogram (MRA).

38.**(B)** Primary ciliary dyskinesia PCD diagnosis has historically relied on a combination of clinical features and ultrastructural analysis of respiratory cilia by electron microscopy, obtained from scrapings/biopsy of nasal or airway epithelium. The measurement of nasal nitric oxide is the most sensitive and specific diagnostic test for PCD, and a low nasal nitric oxide level (<77 nL/min) is consistent with PCD. High-speed videomicroscopy assesses ciliary beat frequency and pattern but requires specialized equipment, and, currently, there is an absence of standardized reporting methods.

39.**(C)** Surveillance cultures help identify organisms involved and guide antibiotic therapy. Chest physiotherapy and prompt treatment of bacterial infections are helpful, but the course of the lower airway disease tends to be slowly progressive. Inhaled hypertonic saline improves cough clearance and has been shown to improve lung function. Anti-inflammatory antibiotics, such as low-dose macrolides, may reduce the number of exacerbations per year. Sinus surgical procedures are often done to manage chronic sinusitis, but their benefit is

questionable. Most children require placement of pressure equalization (PE) tubes for management of recurrent otitis media. Multidisciplinary care with pulmonology, otolaryngology, audiology, respiratory therapy, and nutrition is recommended.

40.**(A)** Pulmonary arterial hypertension is defined as a mean pulmonary artery pressure of 20 mm Hg or higher in children over 3 months of age at sea level.

41.**(B)** Exertional dyspnea and unexplained syncope are presenting symptoms. In addition to the other physical findings associated with pulmonary and cardiac diseases, an accentuated pulmonary component of the second heart sound may be heard.

42.**(D)** Definitive diagnosis of cor pulmonale is made by cardiac catheterization, but echocardiography may confirm the presence of right ventricular hypertrophy, ventricular dysfunction, interventricular septal flattening, and tricuspid insufficiency, which can be used to estimate the pulmonary artery pressures. Cardiac magnetic resonance imaging can be useful in diagnosis and to assess changes in ventricular function and size. Lung biopsy may be considered for children with PAH suspected of diffuse lung disease, pulmonary venoocclusive disease, or vasculitis.

43.**(C)** Idiopathic pulmonary hemosiderosis is a rare disorder characterized by recurrent alveolar bleeding, iron-deficiency anemia, and hemosiderin-laden macrophages in the lung, which can be identified microscopically with the use of special iron-staining techniques in bronchoalveolar lavage or lung biopsy specimens.

44.**(D)** CT pulmonary angiography (CTPA) is the current standard of care and provides accurate diagnosis with rapid turnaround time. CT also provides information on other potential causes of acute chest pain. Chest radiograph has limited utility, occasionally showing findings of PE or infarction, but is useful in evaluating other potential causes of chest pain. Ventilation-perfusion scans may reveal defects in perfusion without matching ventilation defects, but they are difficult to perform in young children. Catheter pulmonary angiography is no longer used for diagnosis and is used only for interventional management. Doppler or compression ultrasonography can be useful in assessing patients for lower extremity DVTs. Children with pulmonary embolism without an obvious cause should be evaluated for hypercoagulable states, the most common of which is factor V Leiden.

45.**(D)**

TABLE 137.3	Causes of False-Positive and False-Negative Results on Sweat Testing
FALSE POSITIVE	
Adrenal insufficiency Eczema Ectodermal dysplasia Nephrogenic diabetes insipidus Hypothyroidism Fucosidosis Mucopolysaccharidosis Dehydration Malnutrition Poor technique/inadequate sweat collection Type I glycogen storage disease Panhypopituitarism Pseudohypoaldosteronism Hypoparathyroidism Prostaglandin E_1 administration	
FALSE NEGATIVE	
Edema Poor technique/inadequate sweat collection	

46.**(B)** The altered chloride ion conductance in the sweat gland results in excessively high sweat sodium and chloride levels. This is the basis for the sweat chloride test, which is still the standard diagnostic test for this disorder. It is positive (elevated sweat chloride ≥60 mEq/L) in 99% of patients with CF.

47.**(B)** The triple-drug combination elexacaftor-tezacaftor-ivacaftor is an important new therapy for individuals who have at least one F508del mutation, which includes approximately 85% of people with CF. Elexacaftor-tezacaftor-ivacaftor demonstrated improvement in lung function and symptom-related quality of life, and decreased sweat chloride, compared to dual-therapy tezacaftor-ivacaftor. As of mid-2021, it is approved for adults and children 6 years of age and older.

48.**(A)** Antibiotic therapy is important in controlling chronic infection. Monitoring pulmonary bacterial flora via airway cultures and providing aggressive therapy with appropriate antibiotics (oral, aerosolized, and intravenous [IV]) help to slow the progression of lung disease. Patients often require 2- to 3-week courses of high-dose IV antibiotics and aggressive chest physiotherapy to treat pulmonary exacerbations. Antibiotics are selected based on organisms identified by sputum culture. If patients are unable to provide sputum, then a throat culture for CF pathogens can be used to direct therapy. Common infecting organisms include P. aeruginosa and S. aureus.

49.**(A)** Intestinal obstruction in CF patients beyond the neonatal period is often due to DIOS, which may need to be treated with courses of oral laxatives (polyethylene glycol) or, in more refractory cases, with gastrografin enemas. Pancreatic enzyme dosage adjustment, adequate hydration, and dietary fibre may help prevent recurrent episodes.

50.**(B)** When indicated, CT scans of the chest can be useful in quantifying the size of pneumothoraces, for differentiating air within the lung parenchyma (cystic lung disease) from air in the pleural space, and for identifying subpleural blebs that may be present in spontaneous recurrent pneumothoraces.

51.**(A)** Small pneumothoraces (<20% of thorax occupied with pleural air) may not require intervention as they often resolve spontaneously. Inhaling high concentrations of supplemental O2 may enhance reabsorption of pleural air by washing out nitrogen from the blood. Larger pneumothoraces and any tension pneumothorax require immediate drainage of the air, preferably via chest tube.

52.**(B)** Most parapneumonic effusions are due to pneumonia caused by Streptococcus pneumoniae, group A streptococci, or Staphylococcus aureus.

53.**(B)**

54.**(B)**

55.**(B)**

56.**(C)**

SECTION 19

THE CARDIOVASCULAR SYSTEM

AHMED TAWFEQ

QUESTIONS

1. A 5-year-old boy presented to OP clinic with history of fainting attacks. The mother describes the episode as sudden pallor and falling down, followed by loss of consciousness and abnormal movements of extremities that last for about 30 seconds. The child remains hot and sweaty for a while when he awakes. There is family history of congenital heart disease (a small VSD in older sibling which closed spontaneously), physical examination shows well grown child with normal vitals and cardiovascular examination, his ECG is also normal.

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

A. cardiac syncope
B. carotid sinus syncope
C. convulsive disorder
D. vasodepressor syncope

2. An 11-year-old boy referred from GP with grade III ejection systolic murmur mostly heard at left upper sternal border, it became fainter in upright position, not radiated to the back. The boy anthropometric measures are normal as well as his ECG.

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

A. aortic stenosis
B. adolescent ejection murmur
C. pulmonary valve stenosis
D. Still's murmur

3. You are evaluating a 14-year-old girl in cardiology clinic referred because of attacks of palpitation, you heard a mid-systolic click with late uniform, high-pitched systolic murmur, at apical area.

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

A. mitral valve prolapse
B. mitral valve stenosis
C. pulmonary hypertension
D. hypertrophic obstructive cardiomyopathy

4. In pediatrics; a cardiac cause is suspected if the history of chest pain is

A. increasing with inspiration
B. associated with panic attacks
C. increasing with palpation
D. associated with syncope

5. A house officer in duty is calling you, taking your opinion regarding a 5-year-old girl presented to ER with fever and query cardiac rhythm problem. He describes the pulse as (increasing with inspiration). The heart rate was 150 beats/min. Her ECG shows sinus rhythm.

Of the following, the **MOST** likely compatible rhythm abnormality is

A. sick sinus rhythm
B. sinus arrhythmia
C. atrial tachycardia
D. supraventricular tachycardia

6. You are treating a 4-month-old baby boy with narrow QRS complex tachycardia. His vitals are: HR 240 beats/min, B.P 75/50 mmHg, RR 50 breaths/min, temp 36.5^0 C.
Which of the following options would be the **MOST** effective management?

A. Ice face bag
B. Digoxin
C. Adenosine
D. Synchronized DC shock

7. A 7-month-old baby girl, who is a known case of recurrent SVT on amiodarone maintenance therapy.
Of the following, the **MOST** important follow-up investigation is

A. liver function test
B. renal function test
C. blood gas analysis
D. vitamin B_{12} level

8. You are evaluating a 6-year-old girl with dysrhythmia; her ECG shows complete dissociation between atrial and ventricular activity. Her heart rate is 70 bpm during rest, she had no sleep problems and had normal play and school activities.
Of the following, the **MOST** appropriate choice of management would be

A. watchful waiting
B. referring for temporary pacemaker
C. referring for permanent pacemaker
D. starting inotropic medications

9. You are evaluating a 3-month-old baby boy with ventricular septal defect. You heard a pansystolic murmur, at the lower left sternal border with mid-diastolic murmur at the apex and splitting of S_2.
You are **MOST** likely dealing with

A. small size VSD
B. large size VSD
C. VSD with pulmonary stenosis
D. VSD with mitral regurgitation

10. The **MOST** common type of ASD is

A. primum
B. secundum
C. sinus venosus
D. atrioventricular

11. A palpable impulse at the left lower sternal border is a feature of

A. atrial septal defect
B. ventricular septal defect
C. patent ductus arteriosus
D. aortic stenosis

12. You are counseling parents with a baby having a small ostium primum type of ASD. They are asking about the best way to deal with the defect.
What is the **MOST** appropriate answer?
A. Small primum defects usually closes spontaneously
B. We will start afterload reducing agents right now and see
C. It can be closed with closure device by catheterization
D. It needs surgery ultimately

13. To classify truncus arteriosus as a congenital heart disease, it would be classified as
A. stenotic defect
B. right-left defect
C. left-right defect
D. mixing defect

14. What is the **MOST** common electrolyte disturbance of furosemide?
A. Hypokalemia
B. Hypocalcemia
C. Hyponatremia
D. Hypouricemia

15. You are evaluating a 6-month-old baby girl with a continuous murmur at left infraclavicular area which is radiated to back. CXR shows increase pulmonary vascularity, and the ECG shows left ventricular hypertrophy. She is not gaining weight adequately and she had 2 episodes of chest infections.
What is the **MOST** appropriate measure?
A. Reassurance and watchful waiting
B. Referring her to cardiologist
C. Starting diuretic
D. Starting afterload reducing agents

16. Which of the following murmurs is radiated to the neck?
A. Pulmonary artery branch stenosis
B. Hypertrophic obstructive cardiomyopathy
C. Patent ductus arteriosus
D. Coarctation of aorta

17. Which of the following is more prone to develop early pulmonary vascular disease?
A. Large patent ductus arteriosus
B. Large ventricular septal defect
C. Complete atrioventricular defect
D. Large atrial septal defect

18. You are counseling a young parent with their 3-month-old baby girl who had diagnosed as pulmonary valve stenosis. The peak pressure gradient mentioned in echocardiography report was 30 mmHg. They are asking about the prognosis of such lesion.
What is the **MOST** appropriate answer?

A. Follow up of such lesion will determine its severity
B. Such a mild lesion usually does not progress
C. Urgent referral to pediatric cardiologist
D. Balloon dilatation will be very useful in future

19. You are counseling a parent of a 4-year-boy with severe aortic stenosis. You are explaining the possible risks and squeals of balloon valvuloplasty.
Of the following, the **HIGHEST** risk is

A. postoperative heart failure
B. valvular restenosis
C. valvular insufficiency
D. procedure failure

20. The ECG finding consistent with the endocardial cushion defect is

A. right axis deviation
B. superior axis
C. right ventricular hypertrophy
D. left ventricular hypertrophy

21. A 7-year-old boy referred to cardiac clinic for evaluation of murmur. He has referred from pediatric psychology clinic with report the child is highly verbal relative to his intelligence, and he is very sociable, hyper focusing on the eyes of others in social engagements, but he had many learning difficulties. He has broad forehead, a short nose, full cheeks, and a wide mouth with full lips. His teeth are small, widely spaced, and some are missing.
Of the following, the **MOST** likely cause of the murmur is

A. aortic stenosis
B. peripheral pulmonary artery stenosis
C. hypertrophic cardiomyopathy
D. coarctation of aorta

22. The genetic defect behind the patient described in the above scenario is

A. duplication of chromosome No. 17
B. translocation of chromosome No. 13
C. ring chromosome
D. microdeletion of chromosome No. 7

23. A 7-year-old girl referred to cardiology clinic for evaluation of an apical murmur, they had family history of an older sib who died because of thrombotic event. The girl appear tall for her age, has a long limb, pectus excavatum and high arched palate. She is not doing well in school and having visual problems.
Of the following, the **MOST** likely etiology is

A. Marfan syndrome

B. Ehler-Danlos syndrome
C. homocystinurea
D. Stickler syndrome

24. What is the **MOST** common congenital cyanotic heart disease in neonatal period?
A. Tetralogy of Fallot
B. Transposition of the great arteries
C. Tricuspid atresia
D. Truncus arteriosus

25. A 6-month-old baby boy who is a known case of tetralogy of Fallot, presented with severe cyanosis and tachypnea, the new echocardiography evaluation is showing markedly increase flow gradient across pulmonary valve (86mmHg), it also stated that the flow is critically reduced to main pulmonary artery. His anthropometric measures are good for age. You already managed the acute issue.
The **MOST** important next step in the management is to arrange for
A. Blalock-Taussing shunt
B. balloon dilatation through trans-femoral catheter
C. total correction
D. diagnostic catheterization

26. Which of the following has no role in management of hypoxic spells in tetralogy of Fallot?
A. Morphine sulfate
B. Sodium bicarbonate
C. Ketamine
D. Phenylephrine

27. The classical cardiac chest x-ray shadow of D-transposition of great arteries is
A. egg on a string
B. snowman appearance
C. boot shape
D. box shape

28. A 3-day-old baby boy presented with cyanosis. On examination; there was quiet tachypnea and single S_2. The pulses were weak and there was no murmur. The chest x-ray reveals increased pulmonary vascularity.
Of the following, the **MOST** likely diagnosis is
A. tetralogy of Fallot
B. transposition of the great arteries
C. tricuspid atresia
D. pulmonary atresia

29. A 1-month-old baby had tricuspid atresia and increasing evidence of cyanosis. He had a small restrictive VSD and hypoplastic right ventricle.
Of the following, the procedure of choice is
A. bidirectional Glenn

B. subclavian artery-to-pulmonary shunt
C. Fontan Procedure
D. Norwood procedure

30. A 2-week-old baby boy presented with history of increasing work of breathing for 4 days and respiratory distress for the last 24 hours. On examination his RR 70 breaths/min, HR 162 beats/min, B.P. 60/23mmHg, and O2 saturation is 83%. His peripheral pulses are bounding, and there is systolic murmur at the left sternal border with single S_2. Chest x-ray reveals cardiomegaly, increased pulmonary plethora and displaced pulmonary arteries.
Of the following, the **MOST** likely diagnosis is

A. patent ductus arteriosus
B. truncus arteriosus
C. total anomalous pulmonary venous return
D. hypoplastic left heart

31. An 18-month-old infant presented to ER with respiratory distress. He was well till 2 weeks ago when he started to have a flu like illness and fever, followed by increasing respiratory symptoms and persistent cough. His vitals are RR 62 breaths/min, HR 170 beats/min, B.P. 55/40 mmHg, and O_2 saturation 92%. You find a weak pulse, rales, S_3 and hepatomegaly.
Chest x-ray reveals cardiomegaly with increased lung plethora.
Of the following, the **MOST** beneficial pharmacological therapy is

A. diuretics
B. digoxin
C. ACE inhibitors
D. dopamine

SECTION 19

THE CARDIOVASCULAR SYSTEM

AHMED TAWFEQ

ANSWERS

1.**(D)** Typical syncopal events usually occur in the upright position or are related to changing position. Syncope may be associated with anxiety, pain, blood drawing or the sight of blood, fasting, a hot environment, or crowded places. The patient often appears pale. A prodrome, consisting of dizziness, light headedness, nausea, diaphoresis, visual changes (blacking out), or possibly palpitations, warns the patient and often prevents injury. Unconsciousness lasts for less than 1 minute. A return to normal consciousness occurs relatively quickly. Most of these syncopal episodes are vasovagal or neurocardiogenic in origin. The physical examination is normal. In cardiac syncope the examination usually is abnormal, there is exercise prodrome with variable symptoms of shortness of breath, fatigue and residual cyanosis as post syncopal events. In carotid sinus syncope usually, there is history of tight collar cloth. Convulsion remains always as challenging differential diagnosis.

2.**(B)** A type of innocent murmur in children ranging between 8-14 years, it's never exceeded grade III, soft with normal growth pattern and ECG as with all other types of innocent murmurs.

3.**(A)** Mitral valve prolapse (MVP) is the most common valvular abnormality, affecting approximately 2-3% of the population in the United States. MVP usually has a benign course, but it occasionally leads to serious complications, including clinically significant mitral regurgitation (MR), infective endocarditis, sudden cardiac death, and cerebrovascular ischemic events. The described murmur is typical for MVP.

4.**(D)** A history of chest pain associated with exertion, syncope, or palpitations or acute onset associated with fever suggests a cardiac etiology.

5.**(B)** Sinus arrhythmia is a common finding in children and represents a normal variation in the heart rate associated with breathing. The heart rate increases with inspiration and decreases with expiration, producing a recurring pattern on the electrocardiogram (ECG) tracing. Sinus arrhythmia is normal and does not require further evaluation or treatment.

6.**(C)** Increasing vagal tone maneuvers should be tried in vitally stable patient but it's still effective in few cases only, digoxin in selected cases, and synchronized cardioversion in hemodynamically compromised patients. Adenosine is the most effective treatment for this scenario as he is hemodynamically stable.

7.**(A)** Amiodarone is better to be avoided in maintenance therapy if alternative is available. It's potentially toxic medication and can be pro-arrhythmic. It can also cause liver toxicity and thyroid dysfunction.

8.**(A)** In cases of complete heart block, the pacemaker is indicated if the child is symptomatic, and the HR is below 60 bpm.

9.**(B)** Typical murmur in moderate-large VSD.

10.**(B)**

11.**(A)** In ASDs most patients are asymptomatic, findings can be manifested as prominent right ventricular impulse at the left lower sternal border (LLSB) often can be palpated. A soft (grade I or II) systolic ejection murmur in the region of the right ventricular outflow tract and a fixed split S2(due to overload of the right ventricle with prolonged ejection into the pulmonary circuit) are often audible. In pulmonary stenosis a similar impulse can be felt.

12.**(D)** Many secundum ASDs closes spontaneously, or can be closed with an ASD closure device in the catheterization laboratory. Primum and sinus venosus defects require surgical closure.

13.**(D)**

14.**(A)** Furosemide induces various electrolyte imbalances including hypokalemia, hypomagnesemia, hypocalcemia, hyponatremia, and hyperuricemia.

15.**(C)** Moderate-large PDAs may be managed initially with diuretics, but they eventually require closure. Elective closure of small, hemodynamically insignificant PDAs is controversial. Most PDAs can be closed in the catheterization laboratory by either coil embolization or a PDA closure device.

16.**(B)** Systolic murmurs maximal at the upper sternal borders are more likely to be ejection in type due to heart outflow abnormality or increased flow - aortic valve, subvalve or supravalvular stenosis and HOCM being maximal on the right radiating to the neck whilst pulmonary valve, subvalve or supravalvular stenosis or atrial septal defect murmurs are louder on the left and radiate to the back.

17.**(C)**. All above lesions can develop pulmonary hypertension, but the (C) defect is earliest to develop especially in Down syndrome followed B and A and the least is ASD.

18.**(B)** Valvular pulmonary stenosis usually does not progress, especially if it is mild.

19.**(C)** Balloon valvuloplasty (valvotomy), is usually the first interventional procedure for significant stenosis. It is not as successful as pulmonary balloon valvuloplasty and has a higher risk of significant valvular insufficiency. Surgical management is necessary when balloon valvuloplasty is unsuccessful or significant valve insufficiency develops.

20.**(B)** The ECG usually reveals left axis deviation and combined ventricular hypertrophy and may show combined atrial enlargement. Superior axis is a unique findings of EEG changes can be seen only in cushion defect and in tricuspid atresia where cardiac electricity is directed to between 270-360 degree.

21.**(A)** Aortic stenosis of supravalvular type is the most common associated cardiac problem in William syndrome followed by peripheral pulmonary artery stenosis.

22.**(D)** The genetic defect of William syndrome is (D) which is considered as structural chromosomal defect. The most common associated biochemical finding is hypercalcemia.

23.**(C)** Those patients may benefit from high doses of Vit B6 supplementation.

24.**(B)** The most common cyanotic heart disease is A representing about 10% of all congenital heart defects, while the commonest in neonatal period and represents only about 5% of congenital heart defects is B.

25.**(C)** A is still good option if no expert surgeon is available or to avoid long course of anesthesia in underweight baby, but the weight of the baby in scenario seems to be OK. Balloon dilatation has no role in subvalvular stenosis which always requiring surgery. Oral propranolol is of help for limited time, you can buy time by it meanwhile preparing for surgery. The occurrence of a cyanotic spell is an indication to proceed with surgical repair.

26.**(B)** Treatment of hypoxic spells consists of oxygen administration (although of minimal value given that worsening cyanosis is due to lack of pulmonary blood flow) and placing the child in the knee-chest position (to increase venous return). Traditionally, morphine sulfate was given (to relax the pulmonary infundibulum and for sedation). The use of ketamine is often beneficial (sedation, increases systemic vascular resistance [SVR], and can be given intramuscularly). If necessary, the SVR can be increased acutely through the administration of an α-adrenergic agonist (phenylephrine). Sodium bicarbonate is not a standard therapeutic option because of its potential side effects and can be used sparingly.

27.**(A)** The chest x-ray reveals increased pulmonary vascularity, and the cardiac shadow is classically an egg on a string created by the narrow superior mediastinum.

28.**(B)**. This is a scenario for duct dependent cyanotic heart disease. In A, C and D there will be no respiratory distress and CXR usually shows decrease lung vascularity (oligemic lung).

29.**(B)**. Management initially depends on the presence of a VSD and the amount of antegrade blood flow to the lungs. If there is no VSD, or it is small, prostaglandin E1 maintains pulmonary blood flow until surgery. Surgery is staged with an initial subclavian artery-to-pulmonary shunt (Blalock-Taussig procedure) typically followed by a two-stage procedure: bidirectional cavopulmonary shunt (bidirectional Glenn) and Fontan procedure. The need to B-T shunting is determined by the size of VSD; If VSD is small and restrictive, it may cause severe cyanosis (option B is needed initially then A and C) while if its large the baby may develops heart failure (so it's wise to go to option B and C together).

30.**(B)** A is non-cyanotic congenital heart disease, in C the presentation is similar but there will be no bounding pulses or wide pulse pressure, and there is usually wide split S2. In D the pulses are weak or absent. In all there is variable degree of hepatomegaly (markedly increased in HLHS). TAPVR may simulate respiratory distress syndrome of neonates.

31.**(D)** This scenario is compatible with heart failure secondary to viral myocarditis(with the resultant dilated cardiomyopathy). When symptoms of heart failure appear during rest. That means heart failure is in late stage, so in cases of severe cardiac decompensation, inotropes (as dopamine; dobutamine and milrinone) is indicated. In mild-moderate cases, supportive therapy along with ACE inhibitors, diuretics, and nitroglycerin/nitroprusside are indicated. However, in some instances, some of these drugs cannot be implemented initially because of hemodynamic instability.

SECTION 20

HAEMATOLOGY

HASANEIN GHALI

QUESIONS

1. In respect to history taking in a child suspected of having hematological disease, which of the following is considered a crucial information in approaching a five-year-old boy with long history of pallor, and splenomegaly?

A. The boy has history of severe neonatal jaundice with twice exchange transfusion within the first three days of life
B. The boy has history of prolonged neonatal jaundice that has required follow up till third month of life
C. The boy's aunt has history of frequent transfusions years ago, terminated by splenectomy
D. The maternal grandfather has done cholecystectomy at mid-thirties due to recurrent cholecystitis

2. Which of the following historical data strengthen the possibility of diagnosis of Glucose-6-Phosphate Dehydrogenase deficiency in a two-year-old boy presented with sudden onset of pallor and dark-colored urine?

A. History of prolonged neonatal jaundice
B. Positive family history of favism from maternal side
C. History of cholecystectomy from the paternal side
D. Consanguineous marriage of parents

3. Which of the following physical examination findings may point to the diagnosis of thalassemia major in a three-year-old boy presented with pallor and frequent transfusions since the age of six months?

A. Growth retardation
B. Splenomegaly
C. Jaundice
D. Large head

4.Considering the importance of physical examination in hematological disease, which of the following pushes the diagnosis of immune thrombocytopenia to the bottom of the list?

A. Bleeding mouth ulcers
B. Hemorrhagic chicken pox lesions
C. Left large cervical lymph node
D. Bronchial breathing in the upper right lung

5. Which of the following physical findings is least in contribution for a six-year-old boy with possible diagnosis of acute leukemia?

A. Hepatosplenomegaly
B. Lymphadenopathy
C. Growth impairment
D. Joint swelling

6. Which of the following signify extramedullary hematopoiesis in a five-year-old girl with chronic hemolytic process?

A. Short stature
B. Hepatosplenomegaly
C. Decayed teeth
D. Sleep apnea

7. Which of the following tests may give the most valuable information about the nature of the condition in a 10-year-old girl who presented with skin bleeding and was discovered to have dicytopenia by complete blood count?

A. Bone marrow aspiration
B. Pyruvate kinase enzyme assay
C. Coombs test
D. Blood culture

8. A mother of a 3-hour-old girl is enquiring about the cause of high hemoglobin level (18.0 gm/dl) for her newborn daughter. What would be the **BEST** response?

A. The affinity of fetal RBC to oxygen is much greater than maternal RBC to provide the fetal tissue with enough amount of oxygen
B. Low partial pressure of oxygen in utero stimulates production of erythropoietin from the liver
C. The fetal bone marrow is more active than adult bone marrow, hence producing more RBC
D. The life span of fetal RBC is longer than 150 days

9. Observation and expectation in medicine is essential in the assessment of patients with possible hematological disease. A 6-year-old boy with suspected diagnosis of congenital neutropenia is expected to have which of the following?

A. Pallor
B. Heart failure
C. Cyanosis
D. Oral ulcers

10. You received a mother of a 2-month-old boy at the outpatient clinic, she is concerned that the routine checkup has shown a hemoglobin of 10 gm/dl, while it was 17 gm/dl at birth. On examination, the boy is active and thriving well, no worrying features could be identified during examination.
What is the **MOST** likely diagnosis?

A. Hereditary hemolytic anemia
B. Physiological anemia
C. Myelodysplasia
D. Megaloblastic anemia

11. Adolescent males maintain hemoglobin at normal values 1.5-2 gm/dl higher than females, the cause behind this is the

A. heart contractility is higher in males
B. androgen production in males

C. losses of blood through menses in females
D. nature of dietary content among males

12. A 6-year-old boy from an inner-city area, referred to OP clinic with pallor, the history was non-conclusive, and the examination was normal apart from pallor. His complete blood count showed hypochromic microcytic anemia.
Of the following, the **MOST** probable diagnosis from the above limited data is
A. Marrow infiltration
B. Hypothyroidism
C. Autoimmune hemolytic anemia
D. Copper deficiency

13. In regard to nutritional history, which of the following matches fit together?
A. Cow's milk diet and folate deficiency
B. Strict vegetarian and vitamin D deficiency
C. Pica and vitamin B12 deficiency
D. Malabsorption and vitamin E deficiency

14. Which of the following infections in a 10-year-old boy may suggest a hemolytic process as an underlying cause of his anemia?
A. Mycoplasma
B. Giardia Lamblia
C. Parvovirus
D. Hepatitis

15. A 6-year-old child presented to the outpatient clinic with history of pallor, you are explaining the nature of anemia to the colleague. An appropriate response of bone marrow to anemia implying hemolysis of blood loss includes
A. raised platelet count
B. elevated absolute reticulocyte number
C. reduction of monocyte count
D. sudden drop of hemoglobin level

16. A 5-year-old-boy presented with pallor for the last two days, he has normal physical examination. A complete blood count has shown anemia and reticulocytopenia.
The significance of this finding enlightens
A. acute onset of anemia
B. marrow-antibody mediated issue
C. intrinsic marrow disease is on his way
D. peripheral antibody mediated issue

17. An adolescent boy presented to the hematology clinic with suspected hemolysis.
Which of the following biochemical results signifies hemolysis?
A. Increased haptoglobin
B. Increased lactate dehydrogenase
C. Reduced aspartate aminotransferase
D. Reduced calcium level

18. Why a 10-month-old infant who is breast-fed is less likely to have Iron deficiency anemia than the same age infant who is on fresh cow milk?

A. Breast milk iron is higher in quantity
B. Bottle milk iron is irritant to the gastric mucosa
C. Breast milk iron is more palatable
D. Breast milk iron is more efficiently absorbed

19. In otherwise healthy child, the best diagnostic study for iron deficiency anemia is

A. serum iron
B. serum ferritin
C. therapeutic trial of oral iron
D. hepcidin level

20. Ordinal / sequential improvement of iron deficiency anemia includes

A. repletion of iron stores in 1-4 weeks
B. increase hemoglobin level in 2-3 days
C. reticulocytosis in 1-3 months
D. improved neurological function in 1-2 days

21. A 2-year-old boy had started receiving iron treatment for possible iron deficiency anemia. On examination, he was irritable, apathetic and pale, his growth parameters were below standards for his peers. Failure of iron therapy was considered after two weeks of treatment, noticed by failure of rise of hemoglobin.
In this forementioned case, the **MOST** likely cause is

A. ongoing blood loss
B. malabsorption
C. poor compliance
D. development of infection

22. Ophthalmological findings in anemia. Features and matches, which one is **TRUE**?

A. Retinopathy and dyskeratosis congenita
B. Optic atrophy and Fanconi anemia
C. Blocked lacrimal gland and Wilson disease
D. Blue sclera and iron deficiency anemia

23. Anemia and ataxia may suggest

A. iron deficiency
B. B_1 deficiency
C. B_{12} deficiency
D. vitamin E deficiency

24. A 3-year-old child with hypochromic microcytic anemia not responding to a trial of iron therapy has the following labs: Hemoglobin 9.3 gm/dl, mean cell volume of 59, mean cell hemoglobin concentration of 29, red blood cell count of 5.6, red cell distribution width of 12 is likely to have

A. blood loss
B. lead poisoning

C. βeta thalassemia minor
D. sideroblastic anemia

25. A 6-year-old boy living in an old grandpa home, presented with pallor, chronic constipation, pica, and abdominal pain. His blood film showed microcytic hypochromic anemia with basophilic stippling.
Of the following, the **MOST** likely diagnosis is
A. iron deficiency anemia
B. lead poisoning
C. beta thalassemia syndrome
D. hemoglobin Barts disease

26. A challenging clinical story of anemia in a 10-year-old boy with chronic inflammatory bowel disease and observed blood loss. The diagnosis was doubtful between anemia of inflammation and iron deficiency anemia. The main differentiation would be counted upon
A. bone marrow aspiration and iron staining
B. Mentzer index
C. red cell distribution width
D. serum ferritin

27. A 3-year-old boy presented to the hematology clinic with pallor since early months in life. The mother stated he received six times blood transfusion, the first one was at the age of 3 months, and the last one was three months ago. Past medical and family history were negative. On examination, he was an alert boy with prominent pallor. He does not have enlarged lymph nodes, or hepatosplenomegaly. He has short stature. His medical records have shown normochromic normocytic anemia among all blood smears done at different times.
of the following, the **MOST** likely diagnosis is
A. hereditary spherocytosis
B. Diamond-Blackfan syndrome
C. chronic myeloid leukemia
D. βeta thalassemia syndrome

28. A 3-year-boy presented with pallor for the last weeks. On examination, a well-appearing alert boy, pale, no lymph node enlargement, no organomegaly, no skin bleeding, and no joint swelling. His Hb 7.5 gm/dl, WBC 5,600/cmm, Platelets 177,000/cmm, N 20%, L 77%. Bone marrow aspiration showed mild erythroid hypoplasia. Other precursors are normal.
What is the **MOST** likely diagnosis?
A. Transient eryhtroblastopenia of childhood
B. Diamond-Blackfan syndrome
C. Fanconi anemia
D. Dyskeratosis congenita

29. Which of the following suggests bone marrow failure and mandate an examination of the bone marrow?
A. High reticulocyte count
B. Abnormal forms of leukocytes

C. Large platelets
D. Reduced mean cell volume

30. An 8-year-old boy presented to the casualty clinic with skin ecchymoses for the last few months. These lesions were noted spontaneously without trauma. Physical examination shows small head, hyperpigmented lesions in the trunk, and short stature. Abdominal sonogram showed abnormally positioned kidney. No lymph node enlargement or organomegaly was noted. He has a cousin with the same physical feature who passed away with intracranial bleeding at the age of 16 years. Complete blood count shows pancytopenia, with increased mean cell volume. Hemoglobin F was shown to be high.
Of the following, the **MOST** likely diagnosis is

A. Diamond-Blackfan syndrome
B. acquired aplastic anemia
C. Fanconi anemia
D. acute leukemia

31. A 7-year-old girl is referred to the hematology clinic for evaluation of her newly-developed skin ecchymoses. She is a known case of thalassemia since early life under regular transfusion program, yet got hepatitis C infections as a consequence. The mother stated an increment in the requirement of transfusions in the last few months, to be every-other-week instead of her past-usual monthly program. On examination, she has pallor, jaundice, dysmorphology of extramedullary hematopoiesis, and abdominal distension. Abdominal examination revealed hugely enlarged spleen reaching to right iliac fossa. Complete blood picture showed pancytopenia with platelets count of 44,000/cmm.
What is the **MOST** likely cause of low platelets in this patient?

A. Hepatitis C infection
B. Hypersplenism
C. Iron overload
D. Myelodysplasia

32. Out of the thalassemia syndromes, which disorder causes microcytic anemia with mild hemolysis?

A. Cooley anemia
B. Thalassemia intermediate
C. Silent carrier α thalassemia
D. Hemoglobin H

33. Which of the following sickle cell disorders is often asymptomatic?

A. Sickle cell anemia
B. Sickle cell – β thalassemia
C. Hb SC disease
D. Sickle-hereditary persistence of Hb F

34. A 5-year-old sickler presented to the emergency department with shortness of breath for few hours. This was preceded by initial chest pain then coughing. Physical examination reveals areas of decreased breath sounds and dullness on chest percussion.
Of the following, the **MOST** likely diagnosis is

A. pneumonia
B. pleural effusion
C. acute chest syndrome
D. pleurodynia

35. Which of the following manifestations are more frequently encountered in a 6-year-old male with sickle cell anemia?
A. Aplastic crises
B. Splenic sequestration crises
C. Painful crises
D. Priapism

36. Which of the following manifestations appears early in a 3-month-old infant with sickle cell anemia?
A. Anemia
B. Cerebrovascular accidents
C. Vaso-occlusive crises
D. Hematuria

37. A 2-year-old boy presented to the emergency department with sudden onset of pallor, associated with vomiting and abdominal pain. This presentation was preceded few-days by serious suppurative tonsillitis. The mother declared a healthy past history with no similar condition. His 2 older sisters are healthy. She denied any medicine intake except for amoxil and ibuprofen for his recent bacterial tonsillitis. His maternal uncle has four similar events during childhood managed by blood transfusions for each episode. On examination, the boy was very pale, icteric, apprehensive, with tachycardia and tachypnea, respiratory examination was negative, as is his abdominal note. He is passing red-colored urine. Complete blood count showed Hb 4.0 gm/dl, WBC 15,000/cmm, Platelets 455,000/cmm. his blood smear shows fragmented RBC with blisters. His reticulocyte count was 3%.
Of the following, the **MOST** likely diagnosis is
A. hereditary spherocytosis
B. G6PD deficiency
C. pure red cell aplasia
D. autoimmune hemolytic anemia

38. A negative osmotic fragility test is often seen in
A. hereditary spherocytosis
B. autoimmune hemolytic anemia
C. pyruvate kinase deficiency
D. hereditary pyropoikilocytosis

39. Which of the following conditions of thrombotic microangiopathy encounters depletion of clotting factors in a prominent manner?
A. Disseminated intravascular coagulation
B. Malignant hypertension
C. Thrombotic thrombocytopenic purpura
D. Hemolytic uremic syndrome

40. What is the **MOST** common hereditary cause of predisposition to thrombosis?
    A. Plasminogen deficiency
    B. Antithrombin deficiency
    C. Protein C deficiency
    D. Factor 5 Leiden

41. Initial management of thrombotic phenomena requires the use of
    A. Fibrinolytic agents
    B. Warfarin
    C. Heparin
    D. Plasma exchange

42. Coagulopathy and comparable laboratory test, which suit together?
    A. Disseminated intravascular coagulation and raised fibrinogen
    B. Liver failure and reduced D-Dimer
    C. Vitamin K deficiency and prolonged partial thromboplastin time
    D. Sepsis and reduced prothrombin time

43. Which of the following infections is well-known to cause disseminated intravascular coagulation?
    A. Toxoplasmosis
    B. Malaria
    C. Brucellosis
    D. Kala-Azar

44. A 12-year-old girl presented to the emergency department with menorrhagia and severe pallor. She has a background of infrequent epistaxis and cutaneous bruises. Upon reviewing her medical records, it was noted that she had frequent remarks during routine check stating the presence of trivial skin ecchymoses, attributed to trauma each time, though no serious bleeding. On physical examination, she has got multiple skin ecchymoses. No disconcerting features of lymphadenopathy or hepatosplenomegaly. Her initial complete blood count is normal. Bleeding time 15 minutes, partial thromboplastin time 50 seconds.
At this point the **MOST** likely diagnosis is
    A. Bernard Soulier syndrome
    B. Glanzmann thrombasthenia
    C. Von-Willebrand disease
    D. Hemophilia B

45. A 10-year-old boy who is a known case of hemophilia A was recently referred for possible continuous bleeding episodes despite usual dosing of factor VIII. Further evaluation discovered very high titers of antibodies.
At this moment, the **BEST** treatment option for his bleeding episodes is
    A. high dose / continuous factor VIII infusion
    B. cryoprecipitate infusion
    C. recombinant factor VII infusion
    D. plasma infusion

46. The dose of factor for replacement in hemophilia A and B can be calculated using the following fact:

A. One unit per kilogram of factor VIII increases plasma level 2%
B. Two units per kilogram of factor VIII increase plasma level 1%
C. One and half unit per kilogram of factor IX increases plasma level 2%
D. Two units per kilogram of factor IX increases plasma level 1.5%

47. A 6-year-old girl (of healthy consanguineous parents) with long history of presumptive diagnosis of immune thrombocytopenia since early months in life is likely to have

A. secondary immune thrombocytopenia
B. amegakaryocytic thrombocytopenia
C. Bernard Soulier syndrome
D. Glanzmann thrombasthenia

48. A previously healthy 5-year-old boy presented with sudden gum bleeding for the last day. The family denies previous similar episodes and confirms successful circumcision without any sequalae. Upon physical examination, he was found to have diffuse skin ecchymoses and petechiae, with negative note for pallor, lymph node enlargement or organ enlargement. His initial hematology test shows normal hemoglobin, white blood cells and differentials, and low platelets (4,000/cmm). Of the following, the **BEST** treatment option is

A. steroids
B. intravenous immunoglobulin
C. watchful waiting for more serious episodes
D. platelets transfusion

49. Which of the following scenarios is associated with raised thrombin time?

A. A 6-year-old boy with hemophilia A
B. A 10-year-old girl with Glanzmann thrombasthenia
C. A 10-month-old male with complicated sepsis
D. A 5-year-old girl with Type 2 von-Willebrand disease

50. A 5-year-old boy with known case of aplastic anemia, underwent transfusion of packed red blood cells for his anemia. One week later, he was brought to the casualty clinic for being yellowish and passing red-colored urine, with intermittent fever. Upon physical examination, he was found to have jaundiced, pale with low grade fever. His initial labs confirm the anemia and hyperbilirubinemia. His liver enzymes were normal in levels.
Of the following, the **MOST** likely diagnosis is

A. acute hemolytic transfusion reaction
B. delayed hemolytic transfusion reaction
C. febrile non-hemolytic transfusion reaction
D. allergic transfusion reaction

51. Steroids during treatment of transfusion reaction is of crucial rule in

A. acute hemolytic transfusion reaction
B. delayed hemolytic transfusion reaction
C. febrile non-hemolytic transfusion reaction
D. allergic transfusion reaction

HEMATOLOGY

HASANEIN GHALI

ANSWERS

1.**(C)** A detailed history regarding onset, severity, progression and associated symptoms along with family pedigree is crucial in identifying the approach to reach a diagnosis in hematological diseases.

2.**(B)** Positive history of favism from maternal side makes the possibility of the diagnosis of G6PD more convenient, as the disease runs sex-linked pattern of inheritance.

3.**(B)** Physical examination can help in reaching the diagnosis of blood diseases. Observation and detailed examination are invaluable steps. Pallor, jaundice, bleeding, growth parameters, lymph node enlargement and hepatosplenomegaly are essential in each examination.

4.**(C)** The five disconcerting features in immune thrombocytopenia that are essential-to-look-for are pallor, fever, lymphadenopathy, hepatomegaly, and splenomegaly.

5.**(C)** Childhood leukaemia usually presents with a rather acute form, making growth parameters of limited role in raising the diagnosis, among the forementioned items.

6.**(B)** Hemolysis may lead to marrow repopulation of cavities where hematopoiesis previously had ceased or may delay the shift of hematopoiesis.

7.**(A)** A bone marrow examination provides valuable information about processes that lead to underproduction of circulating cells.

8.**(B)** The normally high hemoglobin level of the fetus is a result of fetal erythropoietin production in the liver in response to low partial pressure of oxygen (Po2) in utero.

9.**(D)** A and B in Iron deficiency anemia, C in polycythemia.

10.**(B)** During the first few months of postnatal life, rapid growth, shortened RBC survival, and cessation of erythropoiesis cause a gradual decline in hemoglobin levels, with a nadir at 8–10 weeks of life. This so-called physiologic nadir is accentuated in premature infants with earlier timing and a lower nadir.

11.**(B)** The production of androgens at the onset of puberty in males causes them to maintain a normal hemoglobin value approximately 1.5–2g/dL higher than females.

12.**(D)** A and C has normochromic normocytic features, while B has macrocytic features.

13.**(D)** Cow's milk diet: iron deficiency, Strict vegetarian: vitamin B12 deficiency goat's milk diet: folate deficiency, Pica: plumbism, iron deficiency, Cholestasis, malabsorption: vitamin E deficiency.

14.**(A)** Mycoplasma and Malaria suggest hemolytic cause of anemia; EPV, Parvovirus and CMV suggest bone marrow suppression; blind loop and fish tapeworm suggest B12 deficiency; hepatitis suggest aplastic anemia.

15.**(B)** An appropriate bone marrow response to anemia includes an elevated absolute reticulocyte number, suggesting increased RBC production.

16.**(A)** Reticulocytopenia signifies either an acute onset of anemia such that the marrow has not had adequate time to respond, that reticulocytes are being destroyed in the marrow (antibody mediated), or that intrinsic bone marrow disease is present.

17.**(B)** Biochemical evidence of hemolysis includes raised levels of bilirubin and lactate dehydrogenase and a decrease in haptoglobin.

18.**(D)** Breast-fed infants are less likely to have iron deficiency than bottle-fed infants because, although there is less iron in breast milk, this iron is more efficiently absorbed. However, infants who continue to be exclusively breast-fed in the second half of the first year of life are at risk for iron deficiency.

19.**(C)** In an otherwise healthy child, a therapeutic trial of oral iron is the best diagnostic study for iron deficiency as long as the child is re-examined and a response is documented.

20.**(D)** The response to oral iron includes rapid subjective improvement, especially in neurologic function (within 24–48 hours) and reticulocytosis (48–72 hours); increase in hemoglobin levels (4–30 days); and repletion of iron stores (1–3 months).

21.**(B)** If the hemoglobin level fails to increase within 2 weeks after institution of iron treatment, careful re-evaluation for ongoing blood loss, development of infection, poor compliance, malabsorption, or other causes of microcytic anemia is required.

22.**(D)** Retinopathy matches hemoglobin SC disease, optic atrophy matches osteopetrosis, and blocked lacrimal gland matches dyskeratosis congenita.

23.**(C)** Ataxia and posterior column signs are recognized in vitamin B12 deficiency.

24.**(C)** If the mean corpuscular volume (MCV) divided by the RBC count is less than 12.5 (Mentzer index), the diagnosis is suggestive of thalassemia trait.

25.**(B)** The history of living in an older home (built before 1980) with chipped paint or lead dust should raise suspicion of lead poisoning, especially in a child with pica.

26.**(A)** Inflammation causes an increase in the production of hepcidin, interrupting the process of iron release by macrophages and the absorption of iron from the intestines

leading to anemia. In these circumstances, only a bone marrow aspiration with staining of the sample for iron can differentiate the two entities clearly.

27.**(B)** Diamond-Blackfan syndrome usually presents in the first few months of life or at birth with severe anemia and mild macrocytosis or normocytic anemia.

28.**(A)** Transient eryhtroblastopenia of childhood (TEC), a normocytic anemia caused by suppression of RBC synthesis, usually appears between 6 months and 5 years of age in an otherwise well- appearing child.

29.**(B)** Features that suggest bone marrow failure and mandate an examination of the bone marrow include a low reticulocyte count, teardrop forms of RBCs (implying marrow replacement, not just failure), presence of abnormal forms of leukocytes or myeloid elements more immature than band forms, small platelets, and an elevated MCV in the face of a low reticulocyte count.

30.**(C)**

31.**(B)** Hypersplenism may be the result of anatomic causes (portal hypertension or splenic hypertrophy from thalassemia), infections (including malaria or Epstein-Barr virus [EBV]), storage diseases (Gaucher disease), or malignancy (lymphomas, histiocytosis). Splenectomy is indicated only when the pancytopenia is of clinical significance.

32.**(D)** Hemoglobin H causes mild hemolytic and non-transfusion dependent, unless combined with the constant spring variant.

33.**(D)**

34.**(C)** The acute chest syndrome is a vasoocclusive event within the lungs with evidence of a new infiltrate on chest radiograph. Treatment involves early recognition and prevention of arterial hypoxemia. Oxygen, fluids, judicious use of analgesic medications, antibiotics, bronchodilators, and RBC transfusion (rarely exchange transfusion) are indicated as therapy for acute chest syndrome.

35.**(C)** Painful crises are the most common type of vasoocclusive crises.

36.**(A)** Anemia is chronic, onset 3–4 months of age; hemoglobin usually 6–10 g/dl.

37.**(B)** Early on, the hemolysis usually exceeds the ability of the bone marrow to compensate, so the reticulocyte count may be low for 3–4 days.

38.**(C)** The osmotic fragility test result is abnormal in any hemolytic disease in which spherocytes are present.

39.**(A)** Consumption of clotting factors are more prominent in DIC than in the other forms of thrombotic microangiopathy.

40.**(D)** Factor 5 Leiden is the most common hereditary cause of a predisposition to thrombosis, appearing in 3-5% of whites.

41.**(C)** Major vessel thrombosis, life-threatening thrombosis, or arterial thrombo- sis may necessitate treatment with fibrinolytic agents. In newborns, inherited deficiency syndromes may present as emergencies and necessitate replacement with plasma, antithrombin concentrates, or protein C concentrates.

42.**(C)** DIC and low fibrinogen; liver disease and raised D-Dimer, sepsis and prolonged prothrombin time.

43.**(B)** Meningococcemia (purpura fulminans), Other gram-negative bacteria (Haemophilus, Salmonella, and Escherichia coli), Rickettsia (Rocky Mountain spotted fever), Virus (cytomegalovirus, herpes, hemorrhagic fevers), Malaria, and Fungus.

44.**(C)** Partial thromboplastin time could be normal or prolonged in Von-Willebrand disease.

45.**(C)** Low titres are best treated with option A.

46.**(A)** The dose can be calculated using the knowledge that 1 U/kg body weight of factor VIII increases the plasma level 2%, whereas 1.5 U/kg of recombinant factor IX increases the plasma level 1%.

47.**(C)** The onset of thrombocytopenia early in life suggest inherited disorder, Bernard Soulier is associated with low levels of platelets.

48.**(B)** The diagnosis is immune thrombocytopenia.

49.**(C)** Prolonged thrombin time is noted in fibrin split products, DIC, hypofibrinogenemia, heparin, and uraemia.

50.**(B)** Onset 7–14 days after transfusion: pain, fever, jaundice, hemoglobinuria; fall in hemoglobin; and reticulocytopenia.

51.**(D)** Diphenhydramine with hydrocortisone for acute management of allergic transfusion reaction.

SECTION 21

ONCOLOGY

HASANEIN GHALI

QUESTIONS

1. What is the **BEST** screening radiographic study for a 5-year-old boy with suspected cervical lymphadenopathy, fever and weight loss?

A. Chest X-ray
B. CT neck and chest
C. MRI neck and chest
D. PET scan

2. Prolonged diarrhea as a presentation in a 4-year-old boy with suspected cancer is probably related to

A. Wilms' tumor
B. neuroblastoma
C. acute leukemia
D. lymphoma

3. A 2-year-old girl with suspected neuroblastoma can present with a variety of manifestations.
Of the following the **LEAST** in consideration is

A. miosis and ptosis
B. ataxia
C. abdominal mass
D. superior vena cava syndrome

4. Which of the following malignancies can present as pyrexia of unknown origin?

A. Wilms' tumor
B. Germ cell tumor
C. Hodgkin lymphoma
D. Retinoblastoma

5. A newly diagnosed 7-year-old girl with acute lymphoblastic leukemia can present with certain oncological emergencies.
Which of the following is **MOST** possible?

A. Tumor lysis syndrome
B. Spinal cord compression
C. Disseminated intravascular coagulation
D. Tracheal compression

6. A 4-year-old boy with features of superior vena cava syndrome is more likely to have

A. Neuroblastoma
B. Brain tumor
C. Lymphoma
D. Wilms' tumor

7. After induction therapy (Prednisolone, Asparaginase, Daunorubicin and Vincristine) to a 3-year-old girl with acute lymphoblastic leukemia, you noticed that she has a clumsy gait. Neurological examination revealed bilateral feet drop.
Which of the forementioned agents is responsible for this presentation?

A. Prednisolone
B. Asparaginase
C. Daunorubicin
D. Vincristine

8. A 7-year-old boy whose family is concerned about cognitive regression noted by his teachers at the school is approaching you for counselling. The boy had brain tumor at the age of 2 years and treated successfully with combination of chemotherapy, radiation therapy, and surgical resection. You explained that this is a possible sequalae of

A. Cranial radiation
B. Alkylating agents
C. Topoisomerase II inhibitors
D. Genetic predisposition

9. During a discussion with parents of a 6-year-old girl recently diagnosed with Wilms' tumor about the long-term sequalae of cancer therapy, you explained that she is going to receive Daunorubicin, Actinomycin and Vincristine as baseline treatment. The concern of this therapy should be raised about future development of

A. Infertility
B. Sepsis
C. Pulmonary fibrosis
D. Cardiomyopathy

10. A 3-year-old boy with 6-week history of pallor, fever, and skin ecchymoses. On examination, he was ill, pale, febrile, non-comfortable, with skin ecchymoses and bilateral cervical Lymph nodes, abdomen was soft with enlarged liver and spleen.
Of the following, the **MOST** likely diagnosis is

A. Acute leukemia
B. Infectious mononucleosis
C. Lymphoma
D. Neuroblastoma

11. A 5-year-old boy is diagnosed recently as acute lymphoblastic leukemia. You are conducting a first educational meeting with parents, talking about the nature of their son's disease after receiving the results of cytogenetic and molecular studies. His leukemia is stratified as lower risk due to

A. A white blood cell count of 63,000/cmm
B. CNS positive disease at diagnosis
C. t(12;21) was detected in his blood sample
D. Immune markers detect T cell subtype

12. A non-reported site of relapse in acute lymphoblastic leukemia is

A. Renal infiltration
B. Bone marrow involvement

C. Testicular involvement
D. CNS infiltrations

13. Trimethoprim-sulfamethoxazole prophylaxis is given during treatment of childhood acute lymphoblastic leukemia because those children are at higher risk of infections with
A. Escherichia coli
B. Pneumococcal pneumonia
C. Pneumocystis jirovecii
D. Pseudomonas Aeruginosa

14. Induction mortality in acute lymphoblastic leukemia in developing world is mainly due to
A. thrombosis
B. bleeding
C. tumor lysis
D. infection

15. Arsenic trioxide has been implicated in the treatment of which type of leukemia?
A. Acute promyelocytic leukemia (APL)
B. B-cell acute lymphoblastic leukemia
C. T-cell acute lymphoblastic leukemia
D. Non-APL myeloid leukemia

16. You received a newborn baby with stigmata suggestive of Down syndrome, with enlarged liver and spleen. A checkup CBC at 24-hours showed white cells of 125,000/cmm, Hemoglobin of 10 gm/dl, and platelets of 33,000. Blood film shows peripheral blast cells.
At this point, the **MOST** helpful step of management is
A. induction with chemotherapy
B. keep on supportive treatment
C. refer for radiotherapy to CNS
D. refer for bone marrow transplantation

17. You received a 3-year-old girl diagnosed recently with acute lymphoblastic leukemia, after long battery of tests investigating her pyrexia of unknown origin and non-specific bone pain. Upon reviewing her medical notes, you found that she was treated with steroids and analgesics three months prior to this hospitalization on the bases of other diagnosis. You are reviewing the mimickers of acute leukemia in children, the likely provided history points to
A. brucellosis
B. histiocytosis
C. juvenile idiopathic arthritis
D. immune thrombocytopenia

18. A common scenario of presentation for the sporadic form of Burkitt's lymphoma is
A. cervical swelling
B. abdominal mass
C. pyrexia of unknown origin
D. mediastinal mass

19. The **LEAST** informative evaluation step in assessing the staging of newly diagnosed 10-year-old girl with non-Hodgkin Lymphoma is

A. PET scan
B. bone marrow aspiration
C. cerebrospinal fluid cytology
D. whole body MRI scan

20. A comparative approach to a 6-year-old boy with non-Hodgkin lymphoma versus a same-age boy with Hodgkin lymphoma, a feature favors the first case include

A. Presence of B-symptom is of role in assigning risk adaptive therapy
B. PET scan is an excellent determinant of therapy response
C. Distant, noncontiguous metastases are common
D. Surgery and radiation therapy is more often encountered

21. The leading cause of childhood cancer death in developed regions is

A. leukemia
B. brain tumors
C. sarcomas
D. lymphomas

22. Which of the following syndromes is associated with increased risk for developing CNS tumors?

A. Sturge Weber syndrome
B. Russell Silver syndrome
C. Turcot syndrome
D. Turner syndrome

23. Which of the following is the imaging study of choice in brain tumors?

A. Skull X-ray
B. CT scan
C. MRI
D. PET scan

24. A 10-year-old boy who just recently has done surgical resection of a posterior fossa tumor, two days ago. During the last day, the mother has noted the boy is hesitant to speak, as if unable to produce words, with being apathetic in most of the times. His gait has become ataxic, which was not that case before surgery. The mother is concerned about the new symptoms.
According to the above features, the **MOST** likely explanation is

A. somnolence syndrome
B. micro bleeds resulting from surgical intervention
C. disease progression
D. cerebellar mutism

25. An educational meeting is being established with a family of an infant with brain tumor about the long-term consequences of delivered therapy.
Of the following, the **MOST** significant sequalae that carries a big concern to the family is

A. endocrine sequelae

B. renal insufficiency
C. decreased bone growth
D. neurocognitive deficits

26. A girl with big astrocytoma is recently seen in the outpatient clinic, suffering from features of raised intracranial pressure. You are meeting the family and discussing the importance of offering attention to the symptoms related to this entity.
Which of the following symptoms may point to raised intracranial pressure?

A. Ataxia
B. Failure of abduction of one eye
C. Seizures
D. Diminished coordination

27. A 9-month-old male presented with irritability for the last few weeks. He has no fever, or other complaints, but the mother was concerned about poor feeding noticed last days. Upon examination, he was found to be fussy and pale, with many skin nodules in both trunk and limbs. There was no skin bleeding, lymph node enlargement, or organomegaly. Initial evaluation shows anemia with normal white cells and platelets. Chest X-ray was normal, Abdominal sonogram revealed suspected left renal-related mass. CT scan confirmed the small left suprarenal mass. Bone marrow aspiration shows involvement by small round cell tumor. According to the forementioned data, what is the **MOST** likely diagnosis?

A. Wilms tumor
B. Hepatoblastoma
C. Neuroblastoma MS
D. Non-Hodgkin Lymphoma

28. A 2-year-old girl with raccoon eyes, periorbital ecchymoses associated with thrombocytopenia, is expected to have

A. rhabdomyosarcoma
B. neuroblastoma
C. acute leukemia
D. Hodgkin lymphoma

29. A 2-year-old boy with suspected neuroblastoma is **BEST** diagnosed with

A. urinary catecholamines
B. tissue biopsy
C. ^{123}I-MIBG
D. PET scan

30. Which of the following is the **MOST** common presentation in a 5-year-old boy with Wilms tumor?

A. Fever
B. Abdominal mass
C. Hypertension
D. Hematuria

31. While receiving a 3-year-old boy with newly diagnosed nephroblastoma, you want to look for metastatic sites.

Which of the following tests **BEST** consider the metastatic behavior of tumor?

A. Bone marrow aspiration
B. MRI abdomen and pelvis
C. CT chest
D. ^{123}I-MIBG

32. An infant with bilateral Wilms tumor is at special risk of certain consequences after finishing therapy for the disease.
Of the following, the **MOST** significant risk is

A. renal insufficiency
B. neurocognitive regression
C. liver failure
D. infertility

33. A girl who had received treatment with alkylating agents and/or radiation therapy is at risk of development of second malignancy, namely

A. retinoblastoma
B. rhabdomyosarcoma
C. osteosarcoma
D. Wilms tumor

34. Constitutional features such as fever, fatigue and weight loss may be the presenting features in which of the following tumors?

A. Osteosarcoma
B. Rhabdomyosarcoma
C. Retinoblastoma
D. Ewing sarcoma

SECTION 21

ONCOLOGY

HASANEIN GHALI

ANSWERS

1.**(A)** Mediastinal masses and pleural effusions can be detected on CXR.

2.**(B)** Vasoactive intestinal peptides.

3.**(D)** Also, periorbital ecchymoses, pallor, fever, thrombocytopenia, bone pain, arthralgia, hypertension, and soft tissue mass.

4.**(C)** Lymphoma, leukemia and neuroblastoma.

5.**(A)** Tumor lysis syndrome, febrile neutropenia, hyperleukocytosis, and bleeding.

6.**(C)** Lymphoma can present with tumor lysis syndrome and superior vena cava syndrome.

7.**(D)** Vincristine causes peripheral neuropathy, constipation, jaw pain, inappropriate ADH secretion and minimal myelosuppression.

8.**(A)** Cranial radiation is responsible for leukoencephalopathy, cognitive regression and pituitary dysfunction.

9.**(D)** Due to Anthracyclines.

10.**(A)** All are possible differential diagnoses; leukemia is the nearest target one.

11.**(C)** Favorable features include age 1-9.99 years, <50,000/cmm white cells at diagnosis, absent CNS disease at diagnosis, B lineage, hyperdiploidy, t(12;21), and negative minimal residual disease after induction therapy.

12.**(A)** Renal disease should point to further evaluation.

13.**(C)** Cell-mediated immunosuppression increases the risk of Pneumocystis jirovecii pneumonia.

14.**(D)** Infection in resource limited areas is the major concern of mortality during induction of acute lymphoblastic leukemia.

15.**(A)** Together with all-trans-retinoic acid.

16.**(B)** Newborns with trisomy 21 may have a condition known as transient myeloproliferative disorder, which produces elevated WBC counts with peripheral blasts, anemia, and thrombocytopenia. It usually resolves with supportive care only, but these

children have a significantly increased risk (30%) of developing acute leukemia (ALL or AML) within the next few months to years of life.

17.**(C)** A usual story of misdiagnosis of acute leukemia.

18.**(B)** The sporadic (North American) form of BL more commonly has an abdominal presentation (typically with pain), whereas the endemic (African) form frequently presents with tumors of the head and neck. The anterior mediastinum and cervical nodes are the usual primary sites for T-cell lymphomas.

19.**(D)** Whole body MRI.

20.**(C)** A, B and C are features of Hodgkin Lymphoma.

21.**(B)** Although leukemia is the most common malignancy among children, however, due its improved outcome, brain tumors is the leading cause of cancer death.

22.**(C)** Neurofibromatosis (types 1 and 2), Li-Fraumeni syndrome, tuberous sclerosis, Turcot syndrome, and von Hippel-Lindau syndrome, have an increased risk for developing a CNS tumor. However, most CNS tumors arise in children with no known underlying disorder or risk factor.

23.**(C)** The examination of choice.

24.**(D)** Posterior fossa syndrome (also known as “cerebellar mutism”) occurs in up to 25% of patients after resection of a posterior fossa tumor and is characterized by an acute decrease in speech, behavioural changes (e.g., irritability, apathy, or both), diffuse cerebellar dysfunction, and other neurologic abnormalities. It may begin within hours to days of surgery and is usually self-resolving within weeks to months, although some neurologic complications may persist.

25.**(D)** The neurocognitive deficits can be significant, particularly in infants and young children, and are the primary reason for the continued search for the lowest efficacious radiation therapy dose and the most conformal delivery methods.

26.**(B)** Symptoms of increased intracranial pressure are lethargy, headache, and vomiting (particularly in the morning on awakening). Inability to abduct the eye as the result of sixth cranial nerve palsy is a common sign of increased intracranial pressure.

27.**(C)** A unique category of neuroblastoma, stage MS, is defined in infants (<18 months old) with metastases limited to skin, liver, or bone marrow.

28.**(B)** Sign of orbital bone metastasis.

29.**(B)** Tissue biopsy is the best diagnostic tool. Catecholamines are produced by more than 90% of Neuroblastoma cases.

30.**(B)** The most common compliant is abdominal mass, with or without pain. Fever, hypertension and hematuria can be associated symptoms.

31.**(C)** Pulmonary involvement is the most common metastatic site, followed by regional lymph nodes.
32.**(A)** Patients with bilateral Wilms tumor are sometimes left with renal insufficiency or failure, requiring dialysis or a kidney transplant.

33.**(C)** Prior treatment for childhood cancer increases the risk for osteosarcoma as a second malignancy.

34.**(D)** The femur and pelvis are the most common sites. In addition to local pain and swelling, clinical manifestations may include constitutional symptoms such as fever, fatigue, and weight loss.

SECTION 22

NEPHROLOGY AND UROLOGY

QAHTAN ALOBAIDY

QUESTIONS

1. Which of the following hormones produced by the kidneys is secreted in response to low oxygen delivery?
   A. Renin
   B. Vasopressin
   C. Erythropoietin
   D. Vitamin D metabolites

2. A 4-year-old boy presented with fever and fit, lab work up revealed protein in urine. Which of the following is the **MOST** likely cause of this proteinuria?
   A. Transient proteinuria
   B. Orthostatic proteinuria
   C. Glomerular proteinuria
   D. Tubular proteinuria

3. Which of the following types of nephrotic syndrome can progress to end-stage kidney disease?
   A. Idiopathic membranous nephropathy
   B. Minimal change nephrotic syndrome
   C. Focal segmental glomerulosclerosis
   D. Membranoproliferative glomerulonephritis

4. Which of the following is a routine testing in newly diagnosed 5-year-old boy with nephrotic syndrome?
   A. Serum C3 complement
   B. Red blood cell urine test
   C. Urinary neutrophil gelatinase-associated lipocalin (NGAL)
   D. Antinuclear antibody

5. An 8-year-old girl presented with frequent relapsing nephrotic syndrome, her parents asking you to decrease her edema, your proper answer is that the cornerstone of edema management is by
   A. loop diuretics
   B. fluid restriction
   C. dietary salt restriction
   D. parenteral administration of 25% albumin

6. What are **MOST** common side effects of steroids in steroid-dependent nephrotic syndrome?
   A. Short stature
   B. Increased appetite
   C. Liability to infection

D. Truncal obesity

7. Steroid-dependent nephrotic syndrome means, relapse occurs within
A. 1 week of steroid discontinuation
B. 2 weeks of steroid discontinuation
C. 3 weeks of steroid discontinuation
D. 4 weeks of steroid discontinuation

8. A 5-year-old boy presented with hematuria, proteinuria, hypertension, edema, oliguria and acute kidney injury. He had history of un upper respiratory tract infection 10 days ago.
Which of the following is the **MOST** appropriate management?
A. Dietary sodium restriction, diuretics, and antihypertensive agents
B. Dialysis
C. Watchful waiting
D. Antibiotic treatment

9. Which of the following causes of hematuria typically has an excellent renal prognosis?
A. Thin basement membrane disease
B. Postinfectious glomerulonephritis
C. Rapidly progressive glomerulonephritis
D. Membranoproliferative glomerulonephritis

10. A 2-year-old child presented to emergency department with bloody diarrhea, followed in 7–10 days by weakness, lethargy, and oliguria. Physical examination reveals irritability, pallor, and petechiae. laboratory findings: peripheral blood smear reveals evidence of microangiopathic hemolysis with schistocytes and thrombocytopenia. Coombs test is negative, blood urea nitrogen and creatinine are elevated.
Which of the following management during the diarrheal phase may lessen the severity of acute kidney injury?
A. Treatment of hypertension
B. Red blood cell transfusion
C. Platelet transfusion
D. Intravenous hydration with isotonic saline

11. During the evaluation of a child with acute kidney injury (AKI), the laboratory and clinical findings reveal: low urine output, normal urinalysis, urine Na+ (mEq/L) <15, fractional excretion of sodium (FENa) (%) <1, urine osmolality (mOsm/L) >500, and normal renal ultrasound.
Which of the following causes his/her AKI?
A. Prerenal
B. Intrinsic
C. Postrenal
D. Multifactorial

12. Which of the following measures are initiated to shift potassium into cells in treatment of hyperkalemia in acute kidney injury?
A. Dialysis

B. Diuretics
C. Insulin/dextrose
D. Sodium-potassium exchange resins

13.A 9-year-old female had glomerulonephritis with acute kidney injury, on examination her blood pressure 190/110 mmHg (above the 99 th percentile).
Which of the following antihypertensive agents should be avoided in such case?
A. Diuretics
B. Vasodilators
C. Calcium channel blockers
D. Angiotensin converting enzyme inhibitor

14. Most metabolic complications of chronic kidney disease (CKD) do not begin to manifest until at least
A. stage 2 CKD
B. stage 3 CKD
C. stage 4 CKD
D. stage 5 CKD

15. In discussion with medical students and reviewing the causes of hypertension (HTN) in pediatric age group, particularly obese children and adolescents.
Which of the following is the **MOST** likely cause?
A. Renal causes
B. Endocrine causes
C. Primary hypertension
D. Obstructive sleep apnea

16. A previously well 12-year-old girl presented to pediatric emergency department with nausea, vomiting and headache. Her mother noticed, she had decreased activity and increased sleepiness in the last five days. Blood pressure is 190/130 mmHg.
Which of the following is the **BEST** parenteral antihypertensive agent for such case?
A. Nicardipine
B. Thiazide diuretics
C. Loop diuretics
D. Mannitol

17. Renal biopsy is recommended in children with nephrotic syndrome NS when the age is more than
A. 9 years
B. 11 years
C. 13 years
D. 15 years

18. A 4-year-old boy with neurogenic bladder presented with recurrent febrile UTI.
How much is the percentage of vesicoureteral reflux seen in this patient?
A. 25%
B. 50%

C. 75%
D. 100%

19. A 10-year-old child presented with fever and loin pain. Lab findings: CBP shows leukocytosis with shift to left and toxic granulation, GUE: pus cells 30/ HPF, RBC 10/ HPF, and positive nitrate test.
Which of the following studies is indicated in this child as an initial management?

A. CT abdomen
B. Renal ultrasound
C. Voiding cystourethrogram
D. Radionuclide cystogram

20. Which of the following is an Indication for surgical intervention in a 20-month-old boy with vesicoureteral reflux (VUR)?

A. Recurrent febrile UTI
B. Grades 1 and 2 primary VUR
C. Associated bowel and bladder dysfunction
D. Unilateral grade 4 primary VUR

21. A 1-year-old boy has flank masses, hepatosplenomegaly, hypertension, proteinuria, and hematuria.
Which of the following is the **MOST** likely diagnosis?

A. Wilms tumor
B. Multicystic dysplastic kidney
C. Autosomal recessive polycystic kidney disease
D. Autosomal dominant polycystic kidney disease

22. A 10-year-old boy presented to ER as gross hematuria with severe sharp intermittent pain in the flank radiating to the groin and associated with nausea and vomiting.
Which of the following is the initial study ordered for diagnosis?

A. Renal ultrasound
B. Plain abdominal radiography
C. Contrast helical computed tomography
D. Noncontrast helical computed tomography

23. Prenatal ultrasound showed left sided hydronephrosis, post-natal abdominal ultrasound confirmed crossed fused ectopia, his worried parents asking you about the prognosis of their baby condition.
Of the following, your proper answer is

A. excellent
B. good
C. fair
D. poor

24. A child has kidney stones; lab investigations reveal hypercalciuria as a cause of renal stones.
Which of the following dietary interventions is the **BEST** for this patient?

A. Low sodium and oxalate intake with normal calcium intake
B. Low sodium and oxalate intake with high calcium intake
C. Low sodium and oxalate intake with low calcium intake
D. Low sodium and oxalate intake without calcium intake

25. A 7-year-old female with involuntary urination during sleep. All investigations are within normal range.
Which of the following management provide safe and effective resolution of her enuresis?
A. Imipramine
B. Anticholinergics
C. Oral desmopressin
D. Bed-wetting alarms

26. Parents asking you about the ideal time for correction of their baby hypospadias. You explained to them that **MOST** pediatric urologists repair hypospadias before the age of
A. 6 months
B. 12 months
C. 18 months
D. 24 months

27. You examined a neonate weighing 700 Gm at delivery room with no testis in his scrotum.
Which of the following is the **MOST** likely diagnosis?
A. Retractile testes
B. Undescended testes
C. Absent testes
D. Ectopic testes

28. A 5-year-old boy presented with scrotal pain. On examination, the testicle is swollen and tender, with absent cremasteric reflex.
Which of the following is the **MOST** likely diagnosis?
A. Epididymitis
B. Incarcerated hernia
C. Testicular torsion
D. Torsion of the testicular epididymal appendix

SECTION 22

NEPHROLOGY AND UROLOGY

QAHTAN ALOBAIDY

ANSWERS

1.**(C)** Erythropoietin is secreted by interstitial cells in the renal medulla in response to low oxygen delivery and helps regulate bone marrow red blood cell production. Renin is released in response to decreased renal perfusion pressure, decreased sodium delivery to the DT, and sympathetic nervous system activity and helps maintain intraglomerular pressure. The most potent vitamin D analog—1,25(OH)2-cholecalciferol (calcitriol)—is produced by PT cells in response to parathyroid hormone and intracellular calcium and phosphorus concentrations.

2.**(A)** Transient proteinuria can be seen in children with fever, dehydration, or following seizure or vigorous exercise; it resolves within a few days, does not indicate renal disease, and needs no further evaluation or treatment.

3.**(C)** Focal segmental glomerulosclerosis (FSGS) accounts for ~10–20% of children with NS. Children with FSGS may present like those with MCNS or with less severe proteinuria. Some experts believe there is a spectrum of disease from MCNS to FSGS. A circulating factor that increases glomerular permeability is found in some patients with FSGS. More than 35% of children with FSGS progress to end-stage kidney disease.

4.**(A)** Routine testing typically includes a serum C3 complement. In children with NS, a low serum C3 implies a lesion other than MCNS. Renal ultrasound is often useful to rule out structural abnormalities of the urinary tract. Renal biopsy is recommended in children with persistent proteinuria including NS when MCNS is not suspected (e.g., kidney dysfunction, age >13 years, presence of hematuria, and hypertension).

5.**(C)** The cornerstone to edema management is dietary salt restriction. Severe edema may require use of loop diuretics or fluid restriction. When these therapies are not successful, cautious parenteral administration of 25% albumin (0.5–1 g/kg intravenously over 2–4hours) followed by an intravenous loop diuretic may achieve diuresis.

6.**(B)** Side effects of steroids, such as increased appetite and irritability, are most common in steroid-dependent and frequently relapsing patients who have greatest exposure to steroids.

7.**(B)**

8.**(A)** Treatment of PIGN involves supportive care with dietary sodium restriction, diuretics, and antihypertensive agents. Although treating the streptococcal infection does not prevent PIGN, antibiotic treatment is warranted with active streptococcal infection.

9.**(A)** Thin basement membrane disease (benign familial hematuria) is caused by autosomal dominant collagen mutations that lead to isolated GBM thinning and persistent hematuria.

There is often a family history of benign hematuria in first-degree relatives. As opposed to Alport syndrome, thin basement membrane disease is typically not progressive and usually has an excellent prognosis.

10.**(D)** Therapy for STEC-HUS is supportive and includes volume repletion, treatment of hypertension, and managing complications of AKI, including dialysis when indicated. Early and aggressive intravenous hydration with isotonic saline during the diarrheal phase may lessen the severity of AKI. Red blood cell transfusions are provided as needed. Platelet transfusions should be reserved for active hemorrhage or prior to invasive procedures as they may fuel the thrombotic microangiopathy process.

11.**(A)** Prerenal AKI is most commonly due to volume depletion but may be secondary to other mechanisms of glomerular hypoperfusion.

12.**(C)** Potassium intake and medications that increase potassium should be restricted. If hyperkalemia occurs, intravenous calcium will lower the risk of arrhythmia while measures are initiated to shift potassium into cells (bicarbonate, β-agonists, insulin/dextrose) and hasten removal (diuretics, sodium-potassium exchange resins, dialysis).

13.**(D)** It is logical to suspend all antihypertensive drugs, including ACEI and ARB, during management of acute hypertension. Suspension of ACEI and ARB may cause a rise in GFR, as a result of altered glomerular hemodynamics; this can be of additional value if the GFR is already very low.

14.**(B)** Most metabolic complications of CKD do not begin to manifest until at least stage 3 CKD. In stage 4 CKD, complications become more numerous and severe. Children with stage 5 CKD (ESKD) require treatment with either dialysis or kidney transplantation.

15.**(C)** Pediatric HTN has many causes that are either primary (essential) or secondary. Essential HTN is the most common cause of HTN in children and adolescents, particularly in those who are obese. Secondary HTN should be suspected in younger and/or non-overweight children with more severely elevated BP. Kidney disease is the most common cause of secondary HTN in children.

16.**(A)** Hypertensive emergency requires prompt hospitalization and may require parenteral antihypertensive treatment with nicardipine, labetalol, esmolol, or sodium nitroprusside.

17.**(C)** Renal biopsy is recommended in children with persistent proteinuria including nephrotic syndrome NS when Minimal change nephrotic syndrome MCNS is not suspected (e.g., kidney dysfunction, presence of hematuria, and hypertension) and the age more than 13 years old.

18.**(B)**

19.**(B)** Renal ultrasound (RUS) is typically obtained first to evaluate the kidneys and urinary tract. While the findings of renal and urinary tract anomalies make VUR more likely, the absence of these anomalies does not rule out VUR. Therefore a voiding cystourethrogram

(VCUG) is required to confirm the diagnosis of VUR in a child with recurrent febrile UTI. An international grading system based on VCUG imaging is useful to describe the severity of reflux. For follow-up studies, either VCUG or radionuclide cystogram (RNC) may suffice. In children with a normal RUS, a VCUG is recommended after the second febrile UTI.

20.**(A)** Indications for surgical intervention include VUR that does not resolve over time or is complicated by recurrent febrile UTI despite medical therapy. Grades 1 and 2 primary VUR will likely self-resolve as the intravesical ureteral segment lengthens with growth. Many cases of grades 3 and 4 primary VUR resolve when VUR is diagnosed before 2 years of age and is unilateral. Severe (grade 5) primary VUR and secondary VUR are less likely to resolve spontaneously.

21.**(C)** Autosomal recessive PKD (ARPKD) is characterized by marked bilateral renal enlargement due to numerous microcysts. In some cases, kidney failure develops in utero resulting in oligohydramnios and pulmonary hypoplasia. Congenital hepatic fibrosis is universally present and may lead to bile duct ectasia, biliary dysgenesis, and portal hypertension. Common clinical features include flank masses, hepatosplenomegaly, hypertension, proteinuria, and/or hematuria. End-stage kidney disease often occurs by early childhood.

22.**(A)** Renal ultrasound is usually the initial test of choice for suspected stones in children. Ultrasound can identify hydronephrosis and most stones in the kidney but can miss very small stones and stones distal to the kidney. Noncontrast helical computed tomography (CT) can identify stones of all sizes throughout the entire urinary tract. Plain abdominal radiography is less sensitive for identifying stones than ultrasound and CT.

23.**(A)** Children with renal ectopy or fusion abnormalities have an increased risk of hydronephrosis, VUR, urinary tract infection (UTI), and kidney stones. However, both conditions are asymptomatic with typically excellent prognosis for the majority of patients.

24.**(A)** Specific metabolic conditions require specific treatments. For children with hypercalciuria, dietary interventions include low sodium and oxalate intake with normal calcium intake. Children with hypercalciuria and recurrent stones despite dietary management may benefit from treatment with thiazide diuretics or potassium citrate.

25.**(D)** For children with isolated primary nocturnal enuresis, bed-wetting alarms provide safe and effective resolution of enuresis for greater than 70% of affected children. Oral desmopressin may also be considered in selected children with anticholinergics and imipramine less commonly used.

26.**(C)** Males with hypospadias should not be circumcised, particularly if the meatus is proximal to the glans, as the foreskin may be necessary for later repair. Most pediatric urologists repair hypospadias before 18 months of age.

27.**(B)** In neonates, cryptorchidism is more common with shorter gestation (20% in 2,000- to 2,500-g infants and 100% in <900-g infants).

28.**(C)** Torsion accounts for 40% of cases of acute scrotal pain and swelling and is the major cause of the acute scrotum in boys less than 6 years of age. The differential diagnosis of testicular pain includes trauma, incarcerated hernia, and torsion of the testicular epididymal appendix. Torsion of the appendix testis is associated with point tenderness over the lesion and minimal swelling. In adolescents, the differential diagnosis of testicular torsion also must include epididymitis, the most common cause of acute scrotal pain and swelling in older adolescents.

SECTION 23

ENDOCRINOLOGY

ZUHAIR ALMUSAWI

QUESTIONS

1. A diagnosis of DM is made based on four glucose abnormalities that may need to be confirmed by repeat testing, first of them is fasting serum glucose concentration of

A. 120 mg/dL or above
B. 123 mg/dL or above
C. 126 mg/dL or above
D. 130 mg/dL or above

2. Sporadic hyperglycemia can occur in children, usually in the setting of an intercurrent illness. When the hyperglycemic episode is clearly related to an illness or other physiologic stress, the probability of incipient diabetes is

A. <5%
B. <10%
C. <15%
D. <20%

3. T1D results from the autoimmune destruction of insulin- producing β cells (islets) of the pancreas. Which of the following vitamins deficiency triggers the autoimmune process?

A. Vitamin A
B. Vitamin B_{12}
C. Vitamin C
D. Vitamin D

4. The parents of an 18-month-old boy brought their baby because of urine frequency for the last few days, his fasting blood sugar was 110 mg/dL. Antibodies to islet cell antigens including antibodies to insulinoma-associated protein 2(IA-2), glutamic acid decarboxylase (GAD), and Zn Transporter 8 (ZnT8) were positive.
How much is the risk of developing diabetes in this boy?

A. The risk is 10–15%
B. The risk is 20-50%
C. The risk is 55–90%
D. The risk is 100%

5. What is the cause of artificially low measured serum sodium concentration in diabetic ketoacidosis?

A. Hyperglycemia
B. Prerenal azotemia
C. Dehydration
D. Poor oral intake

6. What is the most serious complication of DKA and its treatment?

A. Hypovolemic shock

B. Cerebral edema
C. Paralytic ileus
D. Pre-renal failure

7. Hyperglycemia in diabetic ketoacidosis is treated by fast-acting soluble insulin administered as a continuous IV infusion (0.1 U/kg/hr). Serum glucose concentrations should decrease at a rate no faster than
A. 50 mg/dL/hr
B. 100 mg/dL/hr
C. 150 mg/dL/hr
D. 200 mg/dL/hr

8. Which of the following vital signs is indicative of possible cerebral edema in diabetic ketoacidosis?
A. Bradycardia
B. Hypotension
C. Tachypnea
D. Hypothermia

9. How much is the typical starting total daily doses of insulin for prepubertal patient with new-onset T1D?
A. 0.3–0.5 U/kg per 24 hours
B. 0.5–0.7 U/kg per 24 hours
C. 0.7–1 U/kg per 24 hours
D. 1–1.5 U/kg per 24 hours

10. How long the honeymoon period can last in newly diagnosed T1D?
A. 6 months
B. 12 months
C. 18 months
D. 24 months

11. Measurements of HbA1c are inaccurate in patients with
A. Hemoglobinopathies
B. Iron deficiency anemia
C. B12 deficiency anemia
D. Folate deficiency anemia

12. Which of the following diseases should be tested annually in patients with T1D?
A. Immunoglobulin A deficiency
B. Addison disease
C. Peptic ulcer disease
D. Chronic autoimmune lymphocytic thyroiditis

13. A 5-year-old boy, while playing with his 7-year-old brother in the garden of their home developed headache, visual changes, diaphoresis, confusion, followed by seizure. He is a

known case of T1D for the last year, on lispro given with meals in combination with glargine given at bedtime. The boy took his second dose of lispro without lunch meal.
Of the following, the **MOST** appropriate urgent action is to give

A. glucose gel
B. fruit juice
C. intranasal glucagon
D. iv glucose

14.The parents of a 10-year-old asymptomatic obese boy with family history of T2D are worried about their child and ask for investigations. His fasting glucose was 128 mg/dL and his random glucose was 205 mg/dL.
Of the following, the **BEST** initial management is

A. lifestyle modifications
B. watchful waiting
C. metformin
D. insulin

15. Which of the following features is associated with epinephrine release in response to hypoglycemia?

A. Perspiration
B. Headache
C. Mental confusion
D. Staring

16. A 2-year-old boy presented with perspiration, pallor, somnolence, dysarthria, inability to concentrate, followed by seizure. The baby looks thin and small with history of being small for gestational age. Before this presentation he was suffering from upper respiratory tract infection with low grade fever and poor appetite.
Of the following, the **MOST** likely diagnosis is

A. drug poisoning
B. glucose-6-phosphatase deficiency
C. idiopathic ketotic hypoglycemia
D. malnutrition

17. A 15-month-old boy presented with severe hypoglycemia, massive hepatomegaly without splenomegaly, growth retardation, and lactic acidosis.
Of the following, the **MOST** likely diagnosis is

A. pyruvate carboxylase deficiency
B. galactosemia
C. hereditary fructose intolerance
D. glucose-6-phosphatase deficiency

18. Which of the following is a cause of hypoglycemia without ketonuria?

A. Isolated growth hormone deficiency
B. Adrenocorticotropic hormone deficiency
C. Epinephrine deficiency
D. Hyperinsulinism

19. Which of the following suppresses GH secretion?

A. Stress
B. Hyperglycemia
C. Starvation
D. Exercise

20. What is the upper-to-lower segment ratio in a normal term infant?

A. 1:1
B. 1.4:1
C. 1.7:1
D. 2:1

21. Full term neonates with congenital GH deficiency usually have

A. normal birth length and weight
B. short birth length and normal weight
C. normal birth length and low birth weight
D. short birth length and low birth weight

22. A 15-month-old boy presented with hypotonia, small hands and feet, obesity with an insatiable appetite, developmental delay, hypogonadism, and almond-shaped eyes.
Of the following, the **MOST** likely diagnosis is

A. Laurence-Moon-Bardet-Biedl syndrome
B. Noonan syndrome
C. Prader-Willi syndrome
D. Russell-Silver syndrome

23. Which of the following causes of short stature has increased GH and decreased IGF-1)?

A. Laron dwarfism
B. Pygmies
C. Hypothyroidism
D. Glucocorticoid excess

24. A 14-year-old boy came with his father complaining from short stature in comparison with his school mate with no signs of puberty, his bone age and height age of 11 year, his old brother had same problem at same age, but he is now well regarding puberty and average adult height.
Of the following, the **EXPECTED** spontaneous puberty usually begins after

A. one year
B. two years
C. three years
D. four years

25. A 17-year-old boy with normal proportions and growth throughout childhood presented with delayed puberty and an upper-to-lower ratio of 0.8 and an arm span greater than his height.
Of the following, the **MOST** likely cause of his delayed puberty is

A. Klinefelter syndrome
B. XY gonadal dysgenesis
C. hypogonadotropic hypogonadism
D. constitutional delay

26. A 14-year-old female presented with normal feminization, absence of pubic hair, and primary amenorrhea.
Of the following, the **MOST** likely diagnosis is
A. imperforate hymen
B. androgen insensitivity
C. vaginal septum
D. hypothyroidism

27. Sexual precocity (precocious puberty) in females is classically defined as secondary sexual development occurring before the age of
A. 6 years
B. 7 years
C. 8 years
D. 9 years

28. A 2-year-old female baby brought by her mother due to isolated appearance of bilateral breast tissue without any other abnormal physical findings.
Of the following, the **MOST** appropriate next step is
A. full laboratory investigations
B. pelvic ultrasound study
C. breast ultrasound study
D. re-evaluation at intervals of 6–12 months

29. premature pubarche in males is defined as the isolated appearance of pubic hair before age of
A. 7 years
B. 8 years
C. 9 years
D. 10 years

30. A young mother brought her 10-day-old full term boy with the following result of neonatal screening test, low serum total T4 concentration, normal TSH.
Of the following, the **MOST** likely diagnosis is
A. primary hypothyroidism
B. hypothalamic (trh) tertiary hypothyroidism
C. pituitary (TSH) secondary hypothyroidism
D. congenital thyroxine-binding globulin deficiency

31. An11-year-old girl presented with pallor, hoarse voice, muscle weakness, constipation, and weight gain. On examination, she has dry scaly skin, sparse brittle hair, and a firm, non-tender diffuse goiter with a pebble-like surface.
Which of the following is the **BEST** confirmatory test?

A. Serum antithyroid peroxidase
B. Thyroid biopsy
C. Thyroid scan
D. Serum TSH and free T4

32. A 14-year-old girl presented with mood instability, tremor, anxiety, inability to concentrate, and insidious weight loss. On examination, BP 150/70 mmHg, PR 130 beats/minute, she has a firm homogeneous goiter with brisk DTR. Lab investigation reveals normal serum free T4, elevated serum T3, and suppressed TSH.
Of the following, the **FIRST-LINE** therapy is
A. propylthiouracil
B. methimazole
C. propranolol
D. iodine

33. A 25-day-old full term neonate weighing 2.8 kg (birth weight was 2.5 kg) presented with irritability and poor feeding. Heart rate 130 beats/min, HB 18 gm/dl.
The mother is known to be thyrotoxic with an elevated thyroid stimulating immunoglobulin treated surgically before pregnancy.
Of the following, the **BEST** treatment is
A. methimazole
B. β blocker
C. watchful waiting
D. radioiodine

34. How much is the incidence of carcinoma of the thyroid in children 5 -9 years old of all pediatric cancers?
A. 1%
B. 5%
C. 11%
D. 15%

35. Which of the following is the best reflection of vitamin D sufficiency in the body?
A. 1,25-hydroxyvitamin D
B. 25-hydroxyvitamin D
C. 1-dihydroxyvitamin D
D. Calcium

36. Which of the following conditions is characterized by↑ calcium, ↑ phosphate, ↓parathyroid hormone, and normal 25(OH)D?
A. Immobilization
B. Hyperparathyroidism
C. Familial hypophosphatemic rickets
D. Pseudohypoparathyroidism

37. What is the most common cause of female ambiguous genitalia?
A. Congenital virilizing adrenal hyperplasia

B. Gonadal dysgenesis
C. Dysgenetic testes
D. Tumor infiltration of clitoris

38. What is the classic approach to sex assignment in patient with ambiguous genitalia?
A. Parents desire
B. Feasibility of genital reconstruction
C. Karyotype
D. Gonadal histology

39. A 1st cousin parents brought their 1st 20-day-old breast fed male baby complaining from vomiting, lethargy, poor weight gain, and dehydration. Lab. Investigations reveals hyponatremia, hypoglycemia, and hyperkalemia. Septic screen and ultra sound abdomen are negative.
Of the following, the **MOST** likely diagnosis is
A. pyloric stenosis
B. late onset neonatal sepsis
C. inadequate milk intake
D. congenital adrenal hyperplasia

40. What are the signs of overtreatment of 21-hydroxylase deficiency?
A. Weight loss
B. Growth stunting
C. Skeletal advance
D. Early appearance puberty

41. A 9-year- old female presented with hyperpigmentation, salt craving, anorexia, weakness, and postural hypotension. Lab. findings reveals fasting hypoglycemia, hyponatremia, hyperkalemia, and elevated plasma renin activity.
Of the following, the **MOST** likely diagnosis is
A. Cushing syndrome
B. Addison disease
C. 11-hydroxylase deficiency
D. polycystic ovarian syndrome

42. A 10-year-old obese girl presented with marked failure of longitudinal growth, hirsutism, weakness, nuchal fat pad, acne, striae, and hypertension.
Of the following, the **MOST** frequent cause is
A. adrenal adenoma
B. nodular adrenal hyperplasia
C. Cushing disease
D. long-term administration of exogenous glucocorticoids

SECTION 23

ENDOCRINOLOGY

ZUHAIR ALMUSAWI

ANSWERS

1.**(C)** A diagnosis of DM is made based on four glucose abnormalities that may need to be confirmed by repeat testing: (1) fasting serum glucose concentration of 126 mg/dL or above, (2) a random venous plasma glucose of 200 mg/dL or above with symptoms of hyperglycemia, (3) an abnormal oral glucose tolerance test (OGTT) with a 2-hour postprandial serum glucose concentration of 200 mg/dL or above, and (4) a hemoglobin A1c (HbA1c) of 6.5% or above.

2.**(A)**

3.**(D)** T1D results from the autoimmune destruction of insulin- producing β cells (islets) of the pancreas. In addition to the presence of diabetes susceptibility genes, an unknown environmental insult presumably triggers the autoimmune process. A variety of studies have produced conflicting data regarding a host of environmental factors. These include cow's milk feeding at an early age, viral infectious agents (coxsackievirus, enterovirus, cytomegalovirus, mumps, rubella), vitamin D deficiency, and perinatal factors. T1D is thought to be primarily a T-cell–mediated disease.

4.**(C)** Antibodies to islet cell antigens may be seen months to years before the onset of β dysfunction. These include islet cell antibodies; insulin autoantibodies; and antibodies to insulinoma-associated protein 2(IA-2), glutamic acid decarboxylase (GAD), Zn Transporter 8 (ZnT8), among others. The risk for diabetes increases with the number of antibodies detected in the serum. In individuals with only one detectable antibody, the risk is only 10–15%, while in individuals with three or more antibodies the risk is 55–90%.

5.**(A)** Serum sodium concentrations may be elevated, normal, or low, depending on the balance of sodium and free water losses. The measured serum sodium concentration is artificially low, however, because of hyperglycemia. The corrected Na can be calculated using the equation: Corrected Na = measured Na + [1.6(glucose –100)/100]. Hyperlipidemia also contributes to the decrease in measured serum sodium.

6.**(B)**

7.**(B)** When serum glucose concentrations decrease to less than 250–300 mg/dL, glucose should be added to the IV fluids.

8.**(A)** Symptoms indicative of possible cerebral edema includes a decreased sensorium, sudden severe headache, vomiting, change in vital signs (bradycardia, hypertension, apnea), a dilated pupil, ophthalmoplegia, or seizure.

9.**(B)** A patient already diagnosed with T1D may be restarted on the prior doses if they were adequate. For a patient with new-onset T1D, typical starting total daily doses are

approximately 0.5–0.7 U/kg per 24 hours for prepubertal patients and approximately 0.7–1 U/kg per 24 hours for adolescents, using any number of the available insulin combinations.

10.**(D)** The honeymoon period, usually starts in the first weeks of therapy, often continues for 3–6 months, and can last 2 years.

11.**(A)** HbA1c should be measured four times a year, and the results should be used for counseling of patients. The American Diabetes Association has set HbA1c targets for children at less than 7.5%. Measurements of HbA1c are inaccurate in patients with hemoglobinopathies. Glycosylated albumin or fructosamine can be used in these cases.

12.**(D)** Chronic autoimmune lymphocytic thyroiditis is particularly common and can result in hypothyroidism. Because symptoms can be subtle, thyroid function tests should be performed annually. Other disorders that occur with increased frequency in children with T1D include celiac disease, immunoglobulin A deficiency, Addison disease, and peptic ulcer disease.

13.**(C)** Symptoms of hypoglycemia include those resulting from neuroglycopenia (headache, visual changes, confusion, irritability, or seizures) and those resulting from the catecholamine response (tremors, tachycardia, diaphoresis, or anxiety). Mild episodes can be treated with administration of rapidly absorbed oral glucose (glucose gel or tablets or fruit juices). More severe episodes that result in seizures or loss of consciousness at home should be treated with glucagon, which can be given as an injection or intranasal. IV glucose should be given in hospital settings.

14.**(A)** Asymptomatic patients with mildly elevated glucose values (slightly >126 mg/dL for fasting or slightly >200 mg/dL for random glucose) may be managed initially with lifestyle modifications, including nutrition therapy (dietary adjustments) and increased exercise, which has been shown to decrease insulin resistance.

15.**(A)** Features associated with epinephrine release are perspiration, palpitation (tachycardia), pallor, paresthesia, trembling, anxiety, weakness, nausea, and vomiting. While features associated with cerebral glucopenia are, headache, mental confusion, somnolence, dysarthria, personality changes, inability to concentrate, staring, hunger, convulsions, ataxia, coma, diplopia, and stroke.

16.**(C)** A common cause of new-onset hypoglycemia is idiopathic ketotic hypoglycemia, usually seen in children between 18 months and 8 years of age. Patients have symptoms of hypoglycemia after a period of prolonged fasting, often in the setting of an intercurrent illness with decreased feeding.

17.**(D)** Some glycogen storage diseases (GSDs) occur in a variety of subtypes that differ in severity. Among the subtypes that result in hypoglycemia, the most severe form is glucose-6-phosphatase deficiency, which is characterized by severe hypoglycemia, massive hepatomegaly, growth retardation, and lactic acidosis.

18.**(D)** Hypoglycemia without ketonuria suggests hyperinsulinism or a defect in fatty acid oxidation.

19.**(B)** GH secretion is enhanced by α-adrenergic stimulation, hypoglycemia, stress, starvation, exercise, early stages of sleep, GH-releasing peptides (GHRPs), sex steroids, thyroxine, glucocorticoids, ghrelin, and clonidine. It is suppressed by β-adrenergic stimulation and hyperglycemia.

20.**(C)** The upper-to-lower segment ratio is the ratio of the upper segment (determined by subtraction of the measurement from the symphysis pubis to the floor [known as the lower segment] from the total height) to the lower segment. A normal term infant has an upper-to-lower ratio of 1.7:1, a 1-year-old has a ratio of 1.4:1, and a 10-year-old has a ratio of 1:1.

21.**(A)** Infants with congenital GH deficiency achieve a normal or near-normal birth length and weight at term, but the growth rate slows after birth, most noticeably after age 2–3 years. These children become progressively shorter for age and tend to have an elevated weight: height ratio.

22.**(C)** The Prader-Willi syndrome (PWS) includes fetal and infantile hypotonia, small hands and feet, postnatal acquired obesity with an insatiable appetite, developmental delay, hypogonadism, and almond-shaped eyes. PWS is caused by a lack of expression of paternally inherited genes in the region of chromosome 15q11.2-q13. The majority of patients (70%) have a deletion of the paternal allele, whereas one-quarter have maternal uniparental disomy [UPD (15q) m].

23.**(A)** Patients with Laron dwarfism usually have a prominent forehead, hypoplastic nasal bridge, delayed dentition, sparse hair, blue sclerae, delayed bone maturation and osteoporosis, progressive adiposity, hypercholesterolemia, and low blood glucose. They have elevated serum GH concentrations, although serum IGF-1 and IGF-BP3 concentrations are low.

24.**(A)** Patients with constitutional delay have delayed onset of pubertal development and significant bone age delay (2 standard deviations below the mean, which is equal to a 1.5- to 2-year delay as a teenager). Usually, height gain is below, although fairly parallel to, the normal percentiles on the growth curve. The prepubertal nadir, or deceleration before their pubertal growth spurt, is prolonged or protracted. A family history of delayed puberty in a parent or sibling is reassuring. Spontaneous puberty usually begins in these patients by the time the bone age reaches 12 years in males and 11 years in females.

25.**(C)** As a cause of delayed or absent puberty, hypogonadotropic hypogonadism may be difficult to distinguish from constitutional delay. Hypogonadotropic hypogonadism precludes spontaneous entry into gonadarche; adrenarche usually occurs to some degree. Throughout childhood and in early puberty, patients with hypogonadotropic hypogonadism have normal proportions and growth. When these patients reach adulthood, they often have an upper-to-lower ratio below the lower limit of normal of 0.9 and an arm span greater than their height due to prolonged time growing.

26.**(B)** The complete syndrome of androgen insensitivity includes normal feminization, absence of pubic or axillary hair, and primary amenorrhea. In this syndrome, all müllerian structures, including ovaries, uterus, fallopian tubes, and upper third of the vagina, are lacking; the karyotype is 46, XY, and patients have intra-abdominal testes.

27.**(C)** Sexual precocity (precocious puberty) is classically defined as secondary sexual development occurring before the age of 9 years in males or 8 years in females.

28.**(D)** Benign premature thelarche is the isolated appearance of unilateral or bilateral breast tissue in females, usually at ages 6 months to 3 years. Laboratory investigations are not usually necessary, but a pelvic ultrasound study rarely may be indicated to exclude ovarian disease. Females with this condition should be re-evaluated at intervals of 6–12 months to ensure that apparent premature thelarche is not the beginning of progression into precocious puberty. The prognosis is excellent; if no progression occurs, no treatment other than reassurance is necessary.

29.**(C)** The isolated appearance of pubic hair before age 6–7 years in females or before age 9 years in males is termed premature pubarche, usually resulting from adrenarche. It is relatively common. If the pubic hair is associated with any other feature of virilization (clitoral or penile enlargement or advanced bone age) or other signs (acne, rapid growth, or voice change), a detailed investigation for a pathologic cause of virilization is indicated.

30.**(D)** Although not a hypothyroid condition, congenital thyroxine-binding globulin deficiency occurs in about 1 in 10,000 live births and is associated with low serum total T4 concentration, normal TSH, and normal serum free T4. This is a euthyroid condition and does not require treatment with thyroid hormone because it is merely a binding protein abnormality. It is commonly X-linked dominant.

31.**(A)** The diagnosis may be confirmed by serum antithyroid peroxidase (previously antimicrosomal) and antithyroglobulin antibodies. Neither biopsy nor thyroid scan is indicated in Hashimoto thyroiditis, although a thyroid scan with reduced uptake may differentiate Hashitoxicosis from Graves' disease.

32.**(B)** Medical therapy to block thyroid hormone synthesis consists of methimazole (0.4–0.6 mg/kg/day once or twice daily) or propylthiouracil (5–7 mg/kg/day divided every 8 hours). Both medications are equally effective; however, propylthiouracil is no longer a first-line therapy secondary to concerns of severe liver injury and acute liver failure. A β blocker, such as propranolol or atenolol, is started if symptoms are severe to control cardiac manifestations and is tapered as the methimazole takes effect. Iodine administration, which may suppress thyroid function but becomes ineffective in a few weeks, is sometimes used as a preparation for surgery but never for long-term therapy.

33.**(C)** Treatment for a severely affected neonate includes methimazole and, as needed, a β blocker to help decrease symptoms. Because the half-life of the immunoglobulin is several weeks, spontaneous resolution of neonatal thyrotoxicosis resulting from transplacental passage of TSIs usually occurs by 2–3 months of age. Observation without treatment is indicated in patients who are minimally affected.

34.**(A)** Carcinoma of the thyroid is rare in children (1% of all pediatric cancers in the 5- to 9-year-old age group and up to 7% of cancers in the 15- to 19-year-old age group).

35.**(B)** The serum concentration of 25-hydroxyvitamin D is a better reflection of vitamin D sufficiency than the measurement of 1,25-hydroxyvitamin D.

36.**(A)**

TABLE **176.1** Important Physiologic Changes in Bone and Mineral Diseases

CONDITION	CALCIUM	PHOSPHATE	PARATHYROID HORMONE	25(OH)D
Primary hypoparathyroidism	↓	↑	↓	Nl
Pseudohypoparathyroidism	↓	↑	↑	Nl
Vitamin D deficiency	Nl(↓)	↓	↑	↓
Familial hypophosphatemic rickets	Nl	↓	Nl (sl↑)	Nl
Hyperparathyroidism	↑	↓	↑	Nl
Immobilization	↑	↑	↓	Nl

37.**(A)** Congenital virilizing adrenal hyperplasia is the most common cause of female ambiguous genitalia; it is most commonly the result of an enzyme deficiency that impairs synthesis of glucocorticoids but does not affect androgen production. The impaired cortisol secretion leads to adrenocorticotropic hormone (ACTH) hypersecretion, which stimulates hyperplasia of the adrenal cortex and excessive adrenal production of androgens.

38.**(B)** The major goal is a rapid identification of any life-threatening disorders (salt loss and shock caused by the salt-losing form of CAH). Although the classic approach to sex assignment has been based on the feasibility of genital reconstruction and potential fertility rather than on karyotype or gonadal histology, the effects of prenatal androgen must be considered.

39.**(D)** The dominant clinical features of congenital adrenal mineralocorticoid deficiency are hyponatremia and hyperkalemia, usually developing by 5–7 days after birth but not immediately after birth. Vomiting, dehydration, and acidosis soon follow as does hypotensive shock from glucocorticoid deficiency. Death may occur if the disorder remains undiagnosed and untreated. In females, the ambiguity of the external genitalia is an obvious clue that salt-losing congenital adrenal hyperplasia (CAH) or simple virilizing CAH must be ruled out.

40.**(B)** The treatment of 21-hydroxylase deficiency requires hydrocortisone and fludrocortisone in the case of the salt-losing form. Therapy must be adjusted throughout childhood at regular intervals. Overtreatment will cause growth stunting and weight gain (cushingoid features), whereas undertreatment will cause excessive height gain, skeletal advance, and early appearance puberty, ultimately jeopardizing adult height potential.

41.**(B)** Clinical manifestations of Addison disease are hyperpigmentation, salt craving, postural hypotension, fasting hypoglycemia, anorexia, weakness, and episodes of shock

during severe illness. Subnormal baseline and ACTH-stimulated cortisol values confirm the diagnosis. The presence of hyponatremia, hyperkalemia, and elevated plasma renin activity indicate mineralocorticoid deficiency.

42.**(D)** Classic clinical manifestations of Cushing syndrome in children include progressive central or generalized obesity, marked failure of longitudinal growth, hirsutism, weakness, a nuchal fat pad, acne, striae, hypertension, and often hyperpigmentation when ACTH is elevated. The most frequent cause is long-term administration of exogenous glucocorticoids due to chronic medical conditions.

SECTION 24

NEUROLOGY

HAYDER ALMUSAWI

QUESTIONS

1. Which of the following central nervous system reflexes of infancy appears at age 8-10 months?

A. Grasp
B. Rooting
C. Parachute
D. Babinski

2. During the assessment of olfactory nerve if we use ammonia instead of perfumes, we stimulate the

A. 3rd cranial nerve
B. 5th cranial nerve
C. 7th cranial nerve
D. 9th cranial nerve

3. A 12-year-old male presented with sudden onset of double vision for the last 2 days. On examination, the right eye deviates down and out with associated ptosis and a dilated pupil. There is no preceding febrile illness, trauma, recent vaccination or drug intake.
Of the following, the **MOST** likely is

A. 2nd cranial nerve palsy
B. 3rd cranial nerve palsy
C. 4th cranial nerve palsy
D. 6th cranial nerve palsy

4. The corneal reflex tests cranial nerves V ophthalmic division (sensory arc) and motor arc of

A. 7th cranial nerve
B. 9th cranial nerve
C. 10th cranial nerve
D. 12th cranial nerve

5. A 9-year-old boy presented with incomplete closure of the right eye and deviation of the mouth angle to the left side with absence of forehead wrinkles and no additional clinically relevant finding.
Of the following, the **MOST** likely affected site is

A. right peripheral facial nerve
B. left peripheral facial nerve
C. right side of upper motor neurons in the brain
D. left side of upper motor neurons in the brain

6. At which age an infant can turn his head and eyes to localize a sound?

A. 40 days
B. 60 days
C. 90 days
D. 120 days

7. During analysis of cerebrospinal fluid, the **MOST** likely cause of increased lymphocytes and decreased CSF glucose is
A. bacterial infection
B. brain abscess
C. parasitic infection
D. fungal infection

8. The electroencephalogram with diffuse slow wave activity most likely suggest
A. brain tumour
B. brain abscess
C. metabolic encephalopathy
D. stroke

9. A 12-year-old boy presented with history of recurrent attacks of severe frontal headache, pounding in nature which is aggravated by activity and last about 3 hours, associated with nausea, vomiting, pallor, and an intense desire to seek a quiet, dark room for rest.
Of the following, the **MOST** likely cause of these attacks is
A. tension headache
B. migraine headache
C. increased intracranial pressure
D. sinusitis

10. A 9-year-old boy presented with history of recurrent attacks of mild global headache, squeezing in nature, last about 5 hours, not associated with nausea nor vomiting, and do not disrupt daily activities.
Of the following, the **MOST** likely cause of these attacks is
A. tension headache
B. migraine headache
C. increased intracranial pressure
D. sinusitis

11. A 14-year-old boy presented with history of recurrent attacks of severe migraine headache, not responding to acetaminophen and NSAIDs.
Of the following, the second line medication to abort the attacks is
A. cyproheptadine
B. amitriptyline
C. rizatriptan
D. propranolol

12. A 5-year-old boy is noted by his mother to experience 10-15 seconds lapses in consciousness, eyelid fluttering and lip smacking. electroencephalographic (EEG) patterns consist of generalized 3-Hz spike-and-wave activity.

What is the **MOST** likely diagnosis?

A. Complex partial Seizure
B. Juvenile myoclnic Epilepsy
C. Absence epilepsy
D. Tics disorders

13. A previously healthy and developmentally normal 18-month-old boy presents with generalised seizure lasting for about 9 minutes. There is no past or family history of seizures. Subsequent physical examination reveals a playful child who has a temperature of 39.2^0C and nasal discharge.
Of the following, the **MOST** likely diagnosis is

A. bacterial meningitis
B. viral encephalitis
C. febrile seizure
D. epilepsy

14. An otherwise healthy 1-year-old girl presented with history of several episodes of cyanosis and loss of consciousness after crying. On examination; normal neurological, cardiac, and respiratory system, lab study reveals mild hypochromic microcytic anaemia.
What is the **MOST** likely diagnosis?

A. Epilepsy
B. Breath-holding spells
C. Cyanotic heart disease
D. Benign infantile myoclonus

15. A 6-year-old boy referred to you for evaluation of plaque like lesions. He has subcutaneous nodules, cutaneous schwannomas and posterior capsular cataracts, with no family history of same lesions.
Of the following, the **MOST** likely diagnosis is

A. Neurofibromatosis type II
B. Von Recklinghausen disease
C. Tuberous sclerosis
D. Incontinentia pigmenti

16. What is the **MOST** appropriate rate that phenytoin can be administered intravenously in a child with status epilepticus?

A. 1 mg/kg/min
B. 2 mg/kg/min
C. 3 mg/kg/min
D. 4 mg/kg/min

17. An 8-year-old boy presented with history of abnormal movements around the face and mouth associated with drooling and rhythmic guttural sound which occurs only during sleep or on awakening, the interictal EEG shows bilateral centrotemporal sharp waves.
What is the **MOST** likely diagnosis?

A. Juvenile myoclonic epilepsy
B. Benign Rolandic epilepsy

C. Lennox-Gastaut syndrome
D. Landau-Kleffner syndrome

18. Absence seizures typically begin in the early school years and usually resolve by late childhood or adolescence and if does not remit, nearly half will go on to develop

A. juvenile myoclonic epilepsy
B. simple partial epilepsy
C. centrotemporal epilepsy
D. complex partial epilepsy

19. What is the **BEST** medication to treat an adolescent girl with juvenile myoclonic epilepsy?

A. Valproic acid
B. Ethosuximide
C. Levetiracetam
D. Phenytoin

20. A 6-month-old male presented with history of brief attacks of neck and arm flexion followed by sustained muscle contraction lasting less than 2 seconds, this occur frequently when the child is awakening or going to sleep, each jerk is followed by a brief period of relaxation with repeated spasms in clusters of variable duration.
What is the **MOST** likely EEG findings for this child?

A. Chaotic high-voltage slow waves
B. Persistent focal slowing
C. Diffuse slow wave activity
D. Normal

21. A 7-year-old boy presented with history of loss of previously acquired language, his EEG is highly epileptiform in sleep, the peak area of abnormality in perisylvian region.
What is the **MOST** likely diagnosis?

A. Landau-Kleffner syndrome
B. Lennox-Gastaut syndrome
C. Rett syndrome
D. Cortical spinal syndrome

22. For most children antiseizure medications can be weaned off after 2 years without seizures.
Which of the following conditions require lifelong treatment?

A. Progressive myoclonic epilepsy
B. Absence seizures
C. Centrotemporal epilepsy
D. Infantile spasm

23. A 9-month-old girl brought by her parents for evaluation of delayed motor development. On examination the infant has severe hypotonia, generalized weakness, tongue fasciculation, and absent of deep tendon reflexes.
The diagnosis is best made by

A. creatine phosphokinase
B. electromyography
C. muscle biopsy
D. genetic study

24. A 9-year-old boy presented with history of numbness in hands and feet followed by weak feeling in legs and inability to walk, he had history of acute gastroenteritis 10 days ago. Physical examination reveals symmetrical lower limb weakness and areflexia.
What is the **MOST** likely finding in CSF examination of this patient?
A. Decreased protein levels with significant pleocytosis
B. Decreased protein levels without significant pleocytosis
C. Elevated protein levels with significant pleocytosis
D. Elevated protein levels without significant pleocytosis

25. An 8-year-old boy presented with history of weakness of the ankles and frequent tripping. Examination shows high-arched feet, bilateral weakness of foot dorsiflexors, with normal sensation. The patient had family history of a relative confined to wheelchair with difficulties in performing everyday tasks with his hands.
What is the **MOST** likely diagnosis?
A. Chronic inflammatory demyelinating polyneuropathy
B. Hereditary motor sensory neuropathy
C. Werdnig-Hoffmann disease
D. Kugelberg-Welander syndrome

26. A 9-year-old female presented with a chief complaint of double vision, worsening during the day. Examination shows bilateral ptosis, increasing in severity during the day and improved after rest.
What is the **MOST** likely diagnosis?
A. Third nerve palsy
B. Juvenile botulism
C. Juvenile myasthenia
D. Idiopathic ophthalmoplagia

27. A 7-month-old boy presented with history of constipation and poor feeding. On examination; the baby looks lethargic, hypotonic, with 3rd, 4th, and 6th cranial nerves palsy. The mother added honey to his food in the last month.
What is the **MOST** likely diagnosis?
A. Muscular dystrophy
B. Infantile botulism
C. Infantile myasthenia
D. Congenital myopathy

28. A 9-year-old boy with acute leukaemia on chemotherapy developed progressive headache, vomiting, and lethargy over 2 weeks. Examination shows papilledema, and his head CT scan is normal.
What is the **MOST** likely diagnosis?
A. Meningitis

B. Arterial ischemic stroke
C. Cerebral sinovenous thrombosis
D. Haemorrhagic stroke

29. The incidence of paediatric stroke is 2.5–10 per 100,000 children and is higher among neonates, iron-deficiency anaemia is a risk factor for
A. all types of strokes
B. arterial ischemic stroke
C. cerebral sinovenous thrombosis
D. haemorrhagic stroke

30. Which of the following causes appendicular ataxia?
A. Pilocytic astrocytoma
B. Medulloblastoma
C. Acute postinfectious cerebellar ataxia
D. Ethanol intoxication

31. What is the **MOST** common cause of acute ataxia in childhood?
A. Encephalitis
B. Labyrinthitis
C. Drug intoxications
D. Guillain-Barré syndrome

32. A 17-year-old boy presented with history of inability to make accurate, smooth, and coordinated movements. On examination he has diminished proprioception and vibration sense, absent deep tendon reflexes, and nystagmus.
Of the following, the **EXPECTED** echocardiograph finding is
A. moderate pulmonary valve stenosis
B. atrial septal defect
C. hypertrophic cardiomyopathy
D. severe mitral valve prolapse

33. A state of responsiveness to pain but not to other stimuli, defined as
A. lethargy
B. obtundation
C. stuporous
D. coma

34. A 6-month-old boy presented with developmental delay. On examination, he is unable to make eye contact with retinal cherry red spots on ophthalmoscopy. His liver and spleen are enlarged 4 cm below costal margin and he is unable to hold his head upright during arm traction.
Of the following, the **MOST** likely diagnosis is
A. Tay-Sachs disease
B. Metachromatic leukodystrophy
C. Krabbe disease
D. Niemann-Pick disease

35. A previously healthy 2-year-old boy develops progressive gait ataxia, spasticity, optic atrophy, and intellectual deterioration. Examination reveals absent deep tendon reflexes. Lab findings shows increased cerebral spinal fluid protein and slowing of motor nerve conduction velocity.
Of the following, the **MOST** likely diagnosis is

A. Tay-Sachs disease
B. Metachromatic leukodystrophy
C. Krabbe disease
D. Niemann-Pick disease

36. A 6-month-old boy previously well, develops extreme irritability, hyperpyrexia, vomiting, seizures, hypertonia, and blindness.
Of the following, the **MOST** likely diagnosis is

A. Tay-Sachs disease
B. Metachromatic leukodystrophy
C. Krabbe disease
D. Niemann-Pick disease

37. Which type of mucopolysaccharidoses presents at birth?

A. Type I
B. Type II
C. Type VI
D. Type VII

38. A 19-month-old girl presented with history of developmental regression, loss of purposeful hand movements, loss of verbal communication skills, gait apraxia, and stereotypic repetitive hand movements that resemble washing, wringing, or clapping of the hands.
Of the following, the **MOST** likely diagnosis is

A. Rett syndrome
B. subacute necrotizing encephalomyelopathy
C. MELAS syndrome
D. MERRF syndrome

39. The presence of six or more café-au-lait spots larger than 5 mm in a prepubescent child suggests the diagnosis of

A. Neurofibromatosis type II
B. Von Recklinghausen disease
C. Tuberous sclerosis
D. Incontinenitia pigmenti

SECTION 24

NEUROLOGY

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ANSWERS

1.**(C)** Grasp and rooting reflexes appear at birth and disappear at age 3-4 months of age while parachute reflex appears at age 8-10 months and never disappear.

2.**(B)** Smell can be assessed in verbal, cooperative children older than 2 years of age. Aromatic substances (perfumes, vanilla) should be used instead of volatile substances (ammonia), which irritate the nasal mucosa and stimulate the trigeminal nerve.

3.**(B)** Abnormalities of 3^{rd},4^{th} and 6^{th} cranial nerves may cause diplopia (double vision). With unilateral third cranial (oculomotor) nerve palsy, the involved eye deviates down and out (infraducted, abducted), with associated ptosis and a dilated (mydriatic) pupil.

4.**(A)** The corneal reflex tests cranial nerves V ophthalmic division (sensory arc) and VII (motor arc).

5.**(A)** Weakness of all unilateral muscles of the face, including the forehead, eye, and mouth, indicates a lesion of the ipsilateral peripheral facial nerve (e.g., Bell palsy). Since the upper third of the face receives bilateral cortical innervation, if the weakness affects only the lower face and mouth, a contralateral lesion of upper motor neurons in the brain (e.g., tumor, stroke, abscess) must be considered.

6.**(D)** Four-month-old infants can turn the head and eyes to localize a sound.

7.**(D)** In bacterial infection, brain or parameningeal abscess and parasitic infection increased polymorphonuclear cells and decreased CSF glucose.

8.**(C)** Persistent focal slowing suggests an underlying structural abnormality (e.g., brain tumor, abscess, stroke). Diffuse slow wave activity suggests bilateral cerebral involvement (e.g., increased ICP, metabolic encephalopathy). Spikes, polyspikes, and spike and- wave abnormalities, either in a localized region (focal) or bihemispherical (generalized), suggest an increased risk for seizures.

9.**(B)** Migraine headaches are common type of recurrent headaches and frequently begin in childhood. Headaches are stereotyped attacks of frontal, bitemporal, or unilateral, moderate to severe, pounding or throbbing pain that are aggravated by activity and last 1–72 hours. Associated symptoms include nausea, vomiting, pallor, photophobia, phonophobia, and an intense desire to seek a quiet, dark room for rest.

10.**(A)** Tension-type headaches are the most common type of recurrent primary headaches in children and adolescents. Because they are generally mild, without associated symptoms, they typically do not disrupt daily activities. The pain is global and squeezing or pressing in

character and can last for hours or days. There is no associated nausea, vomiting, phonophobia, or photophobia.

11.**(C)** If first-line medications are insufficient, triptan agents may be considered. Triptans, available in injectable, nasal spray, oral disintegrating, and tablet form, are serotonin receptor agonists that can alleviate migraine symptoms promptly.

12.**(C)** Seizures in which the primary clinical feature is staring can be either absence (generalized) or focal seizures. The clinical hallmark of absence seizures is a brief (<15 seconds) loss of awareness accompanied by eyelid fluttering or simple automatisms, such as fumbling with the fingers and lip smacking. Absence seizures usually begin between 4 and 6 years of age. Neurologic examination and brain imaging are normal. The characteristic electroencephalographic (EEG) patterns consist of generalized 3-Hz spike-and-wave activity. An absence seizure can often be provoked by hyperventilation.

13.**(C)** Febrile seizures represent the most common cause of seizures among children between 6 months and 5 years of age, occurring in about 4% of all children. By definition, a febrile seizure occurs in the presence of fever. Simple febrile seizures are generalized at onset, last less than 15 minutes, and occur only once in a 24-hour period. If the seizure has focal features, lasts longer than 15 minutes, or recurs within 24 hours, the seizure is referred to as a complex febrile seizure.

14.**(B)**

15.**(A)** Neurofibromatosis type 2 (NF2) is an autosomal dominant disorder with an incidence of 1 in 25,000. Half the cases have no family history. Posterior capsular or cortical cataracts are common, and skin lesions include plaque like lesions, subcutaneous nodules, and cutaneous schwannomas. Lisch nodules, café-au-lait spots, and axillary freckling (seen in NF1) are not features of NF2.

16.**(A)**

17.**(B)** Childhood epilepsy with centrotemporal spikes, also known as benign Rolandic epilepsy, is among the most common epilepsy syndromes and usually begins between ages 5 and 10 years. The seizures typically occur only during sleep or on awakening. Affected children usually have focal motor seizures involving the face and arm (abnormal movement or sensation around the face and mouth, drooling, rhythmic guttural sound, impaired speech and swallowing). Seizures sometimes evolve to bilateral convulsions. The interictal EEG classically demonstrates independent bilateral centrotemporal sharp waves but is otherwise normal. With a classic history and EEG and a normal neurologic examination, the diagnosis can be made with confidence and neuroimaging is not required. Seizures usually respond promptly to antiseizure medication. Intellectual outcome is typically normal and the epilepsy resolves after puberty.

18.**(A)** JME is the most common generalized epilepsy among adolescents and young adults. Onset is typically in early adolescence with myoclonic jerks (exacerbated in the morning,

often causing the patient to drop objects), generalized tonic-clonic seizures, and absence seizures. Seizures usually resolve promptly with antiseizure medication.

19.**(C)** JME is classically treated with valproic acid; however, there are concerns about side effects and teratogenicity. Levetiracetam has a much lower risk of teratogenicity and is generally well tolerated. As such, levetiracetam is now favoured, particularly for girls and women since treatment of JME is usually lifelong.

20.**(A)** Persistent focal slowing suggests an underlying structural abnormality (e.g., brain tumor, abscess, stroke). Diffuse slow wave activity suggests bilateral cerebral involvement (e.g., increased ICP, metabolic encephalopathy) while in infantile spasm chaotic high-voltage slow waves, spikes, and polyspikes.

21.**(A)** Acquired epileptic aphasia (Landau-Kleffner syndrome) is characterized by the abrupt loss of previously acquired language in young children. The language disability is an acquired cortical auditory deficit (auditory agnosia). The EEG is highly epileptiform in sleep, the peak area of abnormality often being in the dominant perisylvian region (language areas). This diagnosis should be considered for young patients with clear language regression, as it is a potentially treatable entity.

22.**(A)** Children with JME, progressive myoclonic epilepsy, atypical absence seizures, and Lennox-Gastaut syndrome usually require lifelong treatment with antiseizure medication.

23.**(D)** The diagnosis of SMA is made by genetic testing. Creatine phosphokinase (CK) may be normal or mildly elevated. Although not currently used to diagnose SMA, muscle biopsy specimens show grouped atrophy, and electromyogram shows fasciculations, fibrillations, and denervation.

24.**(D)** The cerebrospinal fluid in Guillain-Barré syndrome is sometimes normal early in the illness but classically shows elevated protein levels without significant pleocytosis (**albuminocytologic dissociation**). MRI with gadolinium may reveal enhancement of the spinal nerve roots. Electromyography (EMG) and nerve conduction velocity are not always required but can provide corroborative diagnostic evidence and prognostic indicators.

25.**(B)** Hereditary motor sensory neuropathy (also called charcot-marie-tooth disease) is a group of inherited, progressive, distal peripheral nerve diseases. Motor components generally dominate the clinical picture, with sensation and autonomic functions affected later. There is variability of severity between types of HMSN and even within families affected by the same type. Some patients never have more than a mild foot deformity, loss of ankle reflexes, and electrophysiologic abnormalities. Others in the same family may be confined to a wheelchair and have difficulties performing everyday tasks with the hands.

26. **(C)** Rapid fatigue of muscles distinguishes myasthenia from other neuromuscular disorders, with progressive worsening over the day or with repetitive activity and improvement after rest. Although myasthenia can produce weakness in any muscle group, variable ptosis, diplopia, ophthalmoplegia, and facial weakness are typical presenting symptoms. There can be any degree of involvement. In some children, the disease never

advances beyond ophthalmoplegia and ptosis (ocular myasthenia). Others may have a progressive and potentially life-threatening illness that involves all musculature, including that of respiration and swallowing.

27.**(B)** Infant botulism results from intestinal infection by Clostridium botulinum, which produces a neurotoxin that blocks presynaptic cholinergic transmission. Infants may ingest dust, soil, or food (e.g., honey, poorly canned foods) contaminated with spores. Infants typically present with constipation and poor feeding. Hypotonia and weakness develop progressively, along with cranial nerve dysfunction manifested by decreased gag reflex, diminished eye movements, decreased pupillary contraction, and ptosis. Autonomic dysfunction can result in fluctuations in heart rate and blood pressure. Severely affected infants may develop respiratory failure.

28.**(C)**

29.**(C)** Symptoms of cerebral sinovenous thrombosis may progress more gradually and be more variable and nonspecific than arterial ischemic stroke. In cerebral sinovenous thrombosis, acute focal deficits may occur or the child may have progressive signs of elevated intracranial pressure, including headache, papilledema, diplopia (most often from cranial nerve VI palsy), seizures, lethargy, or confusion. Hemorrhagic stroke tends to present acutely, with a sudden “thunderclap” headache, hemorrhagic stroke may also present with loss of consciousness, nuchal rigidity, focal deficits, or seizures and can be rapidly fatal.

30.**(A)** Truncal ataxia reflects disturbances of the midline cerebellar vermis (e.g., medulloblastoma, acute postinfectious cerebellar ataxia, ethanol intoxication). Appendicular ataxia reflects disturbances of the ipsilateral cerebellar hemisphere (e.g., pilocytic astrocytoma).

31.**(C)** The most common causes of acute ataxia in childhood are postinfectious acute cerebellar ataxia and drug intoxications.

32.**(C)** Friedreich ataxia is a relentlessly progressive, autosomal recessive disorder. Children present in the late elementary school years with ataxia, dysmetria, dysarthria, diminished proprioception and vibration sense, absent deep tendon reflexes, and nystagmus. Many develop hypertrophic cardiomyopathy and skeletal abnormalities (e.g., high-arched feet, hammer toes, kyphoscoliosis).

33.**(C)** Lethargic patients have difficulty maintaining an aroused state. Patients who are obtunded have decreased arousal but are responsive to stimuli. Stupor is a state of responsiveness to pain but not to other stimuli. Coma is a state of unresponsive unconsciousness and is caused by dysfunction of the cerebral hemispheres bilaterally, the bilateral thalami, and/or the brainstem.

34.**(D)** Classic Niemann-Pick disease (type A) should be suspected in infants who exhibit the combination of hepatosplenomegaly, developmental delay, interstitial lung disease, and retinal cherry red spots. Cognitive regression, myoclonic seizures, hypotonia, and jaundice are also noted within the first year of life.

35.**(B)** Metachromatic leukodystrophy with the late-infantile form present between 1 and 2 years of age. After a period of normal development, progressive gait ataxia, spasticity, optic atrophy, intellectual deterioration, and absent reflexes occur. Diagnostic testing reveals increased cerebral spinal fluid protein (a sign of CNS demyelination) and slowing of motor nerve conduction velocity (a sign of peripheral demyelination).

36.**(C)** Krabbe disease, demyelination of the CNS and peripheral nervous system results in upper and lower motor neuron signs. Individuals with the infantile form (85– 90% of cases) appear to be normal during the first months of life. Symptoms begin by 6 months of age and include extreme irritability, hyperpyrexia, vomiting, seizures, hypertonia, and blindness. Bone marrow transplant during infancy may be a treatment option.

37.**(D)** Signs and symptoms are usually not present at birth with the exception of the more severe forms of MPS type VII.

38.**(A)** MELAS syndrome (mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke like episodes); MERRF syndrome (myoclonus, epilepsy, and ragged red fibers), which can also manifest as combinations of dementia, sensorineural hearing loss, optic nerve atrophy, peripheral neuropathy, and occasionally cardiomyopathy with Wolff-Parkinson-White syndrome, Subacute necrotizing encephalomyelopathy, or Leigh disease ,symptoms usually begin before 2 years of age and consist of hypotonia, feeding difficulties, respiratory irregularity, weakness of extraocular movements, and ataxia. Blood and CSF lactate and pyruvate levels are elevated.

39.**(B)** Neurofibromatosis type 1 (NF1), also known as von Recklinghausen disease, is an autosomal dominant disorder with an incidence of approximately 1 in 3,000. The cardinal features of neurofibromatosis are café-au-lait spots, axillary or inguinal freckling, cutaneous neurofibromas, and iris hamartomas (Lisch nodules).

SECTION 25

DERMATOLOGY

ZAHRAA Z ALMUSAWI

QUESTIONS

1. What is the **MOST** common diagnoses in children attending pediatric dermatology clinics?

A. Atopic dermatitis
B. Impetigo
C. Acne vulgaris
D. Molluscum contagiosum

2. A 16-year-old girl presented with typical acne lesions on face and chest six months ago, she used many topical and oral drugs with no improvement, on the contrary the lesion became worse with development of red cysts and nodule.
Of the following, the **MOST** appropriate agent to be used now is

A. hormonal agents
B. oral isotretinoin
C. topical retinoid
D. oral tetracycline

3. An 18-month-old boy with well controlled atopic dermatitis of the face presented with fever, and exacerbation of facial lesion by the new appearance of pustules, fissures, and crusted lesions. Of note, the mother has lip sores for the last seven days.
What is the **MOST** likely cause of this acute worsening of the atopic lesion?

A. Staphylococcus aureus
B. Herpes simplex virus
C. Fungal infection
D. Streptococcus pyogene

4. Corticosteroids are available in different vehicles for the treatment of atopic dermatitis, which of the following vehicles is the **BEST** to be used in children?

A. Lotions
B. Gels
C. Creams
D. Ointments

5. A 2-year-old asthmatic boy with well controlled atopic dermatitis of face using low potency topical corticosteroids, presented with pustules, erythema, crusting, with lack of response to adequate low potency topical anti-inflammatory therapy.
Of the following, the **MOST** appropriate action is to add

A. high potency topical steroid
B. topical mupirocin
C. oral antibiotic
D. topical acyclovir

6. A 6-month-old infant brought to dermatology clinic suffering from ill-defined, scaly, pink patches and plaques affecting the perianal region and the buttocks while sparing the groin folds and other occluded areas.
Of the following, the **MOST** likely diagnosis is

A. allergic contact dermatitis
B. candida dermatitis
C. seborrheic dermatitis
D. irritant diaper dermatitis

7. A 2-month-old infant presented with greasy, scaly, erythematous, nonpruritic patches and plaques on the face and posterior auricular folds with thick, greasy and waxy, yellow-white scaling and crusting of the scalp.
Of the following, the **MOST** likely diagnosis is

A. seborrheic dermatitis
B. atopic dermatitis
C. tinea capitis
D. Langerhans cell histiocytosis

8. A 15-year-old boy developed a generalized eruption on the trunk and proximal extremities, multiple 0.5- to 1-cm, oval, red macules with a fine, bran like scale. Two weeks before, a solitary 4-cm, pink, oval patch with central clearing was found on his proximal thigh.
Of the following, the **MOST** likely diagnosis is

A. atopic dermatitis
B. seborrheic dermatitis
C. pityriasis rosea
D. Langerhans cell histiocytosis

9. Which of the following is characterized by Auspitz sign?

A. Atopic dermatitis
B. Pityriasis rosea
C. Langerhans cell histiocytosis
D. Psoriasis

10. A 4-month- old infant presented with multiple poorly demarcated, gray-blue patches up to 10 cm in size in lumbosacral area and buttocks.
Of the following, the **MOST** likely diagnosis is

A. café-au-lait macules
B. epidermal nevi
C. salmon patches
D. dermal melanocytosis

11. Which of the following skin lesions is liable for risk of melanoma?

A. Port-wine stain
B. Giant congenital melanocytic nevus
C. Mastocytoma
D. Nevus depigmentosus

12. At what age, superficial hemangiomas reach their maximal size?

A. 3 months
B. 5 months
C. 1 year
D. 4 years

13. A 6-month-old female infant referred to emergency department with progressive hoarseness and stridor for the last two weeks, she has facial hemangioma on the chin and jaw.

Of the following, the **MOST** appropriate action is to refer for

A. laryngoscopy
B. inhaled budesonide
C. inhaled epinephrine
D. CPAP

14. A 9-month-old girl infant presented with large midline cutaneous hemangioma in the lumbosacral area.

Which of the following is the **MOST** appropriate action?

A. CT of the spine
B. MRI of the spine
C. US of the spine
D. Watchful waiting

15. A 3-year-old boy brought by his mother to dermatology clinic complaining from friable, pink-red papules which arise after a minor trauma to the neck, 2- weeks ago. The lesion is growing rapidly into 5-mm, bright red, vascular, pedunculated papule.

Of the following, the **MOST** definitive treatment option is

A. topical β blockers
B. surgical excision
C. pulsed dye laser
D. cryosurgery

16. An 8-month-old girl presented with a 10-cm pink patch on right check seen at birth, with time, it darkens to a port-wine color with slightly thickened surface.

Of the following, the **MOST** successful treatment modality is

A. watchful waiting
B. topical β blockers
C. pulsed dye laser
D. maxillofacial intervention

17. A 6-year-old boy presented with abrupt onset of round, deep red, well-demarcated macules and papules with a dusky gray center. The size of most lesions is approximately 1 cm in diameter which consists of concentric rings.

Of the following, the **MOST** appropriate treatment is

A. oral antihistamines
B. systemic corticosteroids

C. acyclovir
D. topical steroids

18. A 4-year-old boy developed fever, malaise, and upper respiratory symptoms for 5 days followed by red macules which appear suddenly and coalesce into large patches, with a predominant distribution over the face and trunk. These lesions evolve rapidly into bullae and areas of necrosis around 40% body surface area with involvement of upper and lower lips and oral mucosa, with bilateral conjunctival injection and erosions.
Of the following, the **MOST** likely diagnosis is
A. Stevens-Johnson syndrome
B. toxic epidermal necrolysis
C. reactive infectious mucocutaneous eruption
D. staphylococcal scalded skin syndrome

19. Which of the following is a recognized cause of reactive infectious mucocutaneous eruption?
A. Coxsackie virus
B. Herpes simplex
C. Staphylococcus aureus
D. Mycoplasma pneumonia

20. Which of the following differentiate Stevens-Johnson syndrome from patients with Kawasaki disease?
A. Conjunctival injection
B. Hyperemia of the mucous membranes
C. Necrosis of the mucosal surfaces
D. Intertriginous region involvement

21. A 9-month-old boy infant brought by his mother to dermatology outpatient clinic with diffuse erythema, scaling, and pinpoint papules, along with pustules, vesicles, and nodules on axillae, umbilicus, wrist, and face. The mother has similar lesions sparing the face with severe itching.
Which of the following is the **MOST** helpful confirmatory test?
A. Skin scrapings
B. Cytologic examination
C. Wood light examination
D. Skin biopsy

22. A 3-year-old boy presented to dermatology clinic with skin lesion in the trunk since birth. By examination, there is a white patch of 4 cm surrounded by redness and telangiectasia. By diascopy it became indistinguishable from surrounding skin.
Of the following, the **MOST** likely diagnosis is
A. nevus anemicus
B. pityriais alba
C. vitiligo
D. nevus depigmentosus

23. A 5-month-old infant presented with intense erythema and superficial erosions with satellite pustules in diaper area mostly in skin folds and genitalia
What is the **MOST** useful adjuvant test in the diagnosis of this case?

A. Tzanck test
B. KOH test
C. Skin scrapings
D. Woods light

24. Which of the following skin lesions can be altered by time and external factors?

A. Macule
B. Bullae
C. Wheal
D. Lichenification

25. What is the **MOST** common form of childhood psoriasis?

A. Pustular
B. Plaque
C. Inverse
D. Guttate

26. A 3-month-old infant presented with thick greasy, waxy scales and crusts in the scalp.
Of the following, the **LEAST** likely site to be involved by this dermatosis is

A. abdomen
B. axilla
C. ear
D. eyebrows

27. A school aged girl presented with itching, erythema and scaling in the scalp and the nape of the neck that persist despite using OTC lotions. On examination, there is brown and white nits.
Of the following, the treatment option that **MOST** likely to be associated with resistance is

A. benzyl alohol
B. spinosad
C. malathion
D. permethrin

SECTION 25

DERMATOLOGY

ZAHRAA Z ALMUSAWI

ANSWERS

1.**(A)** The most common cutaneous diseases encountered in community settings are dermatophytosis, acne vulgaris, seborrheic dermatitis, atopic dermatitis (eczema), verrucae (warts), tumors, psoriasis, vitiligo, and infections such as herpes simplex and impetigo. The most common diagnoses in children attending pediatric dermatology clinics include atopic dermatitis, seborrheic dermatitis, impetigo, tinea capitis, acne vulgaris, verrucae vulgaris, and molluscum contagiosum.

2.**(B)** For recalcitrant or severe nodulocystic acne, oral isotretinoin may be instituted. Isotretinoin, an oral analog of vitamin A, normalizes follicular keratinization, reduces sebum production, and decreases 5α-dihydrotestosterone formation and androgen receptor-binding capacity. A course of isotretinoin is the only medication that can permanently alter the course of acne and induce a durable remission.

3.**(B)** Signs of concomitant infection include acute worsening of disease in an otherwise well-controlled patient, resistance to standard therapy, fever, and presence of pustules, fissures, punched out erosions, or exudative or crusted lesions. Eczema herpeticum and eczema vaccinatum can be life threatening if not treated.

4.**(D)** Corticosteroids are available in different vehicles. In general, ointments are preferred because of their increased efficacy, occlusive nature, and tolerability. Creams may be slightly less effective for a given steroid ingredient but may be more cosmetically acceptable for older patients or in warmer climates. Lotions may cause more irritation and are generally less potent. Sprays, foams, solutions, and gels can be especially useful for hair-bearing areas. Creams, lotions, sprays, solutions, and gels can be particularly irritating when applied to atopic skin and should generally be avoided on areas of open skin.

5.**(B)** Secondary impetigo with S. aureus is the most common secondary skin infection found in atopic dermatitis, followed by group A streptococcus infection. Infection manifests with pustules, erythema, crusting, flare of disease, or lack of response to adequate anti-inflammatory therapy. Localized lesions can be treated with topical mupirocin.

6.**(D)** Diaper dermatitis is a common problem in infants and most commonly is a form of irritant contact dermatitis. The dermatitis is caused by irritation from urine and feces, typically affecting the perianal region and the buttocks while sparing the protected groin folds and other occluded areas.

7.**(A)** Seborrheic dermatitis in infants begins during the first month and persists during the first year of life. It is also called cradle cap because of the thick, greasy and waxy, yellow-white scaling and crusting of the scalp. Greasy, scaly, erythematous, nonpruritic patches and plaques may extend to the face and posterior auricular folds, sometimes involving the entire body.

8.**(C)** Pityriasis rosea is a benign, self-limited eruption that may occur at any age, with peak incidence during adolescence. A solitary 2- to 5-cm, pink, oval patch with central clearing, the so-called herald patch, is the first manifestation of the eruption. The herald patch typically is found on the trunk or proximal thigh and is often misdiagnosed as fungal or eczematous in origin. One to 2 weeks later, a generalized eruption occurs on the torso and proximal extremities. Multiple 0.5- to 2-cm, oval to oblong, red or tan macules with a fine, bran like scale are characteristically arranged parallel to skin tension lines (Christmas tree pattern). Rarely, the eruption may have an inverse distribution involving the axillae and groin or a popular or papulovesicular appearance.

9.**(D)** The most common variety is plaque-type psoriasis (psoriasis vulgaris), which can be localized or generalized. The lesions consist of round, well-demarcated, pink to red plaques measuring 1–7 cm with micaceous scale, which is distinctive in its thick, silvery appearance with pinpoint bleeding points revealed on removal of the scales (Auspitz sign).

10.**(D)** This is a congenital lesion caused by entrapment of melanocytes in the dermis during their migration from the crest into the epidermis. Although most of these lesions are found in the lumbosacral area (Mongolian spot), they also at other sites, such as the buttocks, flank, extremities, or rarely) the face. Single or multiple poorly demarcated, gray-blue patches up to 10 cm in size may be present. Most lesions gradually disappear during the first few years of life; aberrant lesions in unusual sites are more likely to persist.

11.**(B)** Malignant melanoma develops in approximately 2–10% of patients with giant congenital melanocytic nevi, in either the cutaneous lesion or the neural melanocytes.

12.**(B)** Superficial hemangiomas reach their maximal size by 5 months with the most rapid growth occurring before 8 weeks of life, but deep hemangiomas may grow for longer. Hemangiomas then undergo slow, spontaneous involution, which takes 3–10 years.

13.**(A)** Subglottic hemangiomas manifest as hoarseness and stridor; progression to respiratory failure may be rapid. Symptomatic airway hemangiomas develop in more than 50% of infants with extensive facial hemangiomas on the chin and jaw (beard distribution); any infant with a beard hemangioma should be urgently referred for laryngoscopy.

14.**(B)** MRI of the spine is indicated in all patients with large midline cutaneous hemangiomas in the lumbosacral area.

15.**(B)** A pyogenic granuloma is an acquired, benign vascular tumor commonly seen in children. Initially, the lesions appear as friable, granulation tissue-like, pink-red papules that often arise after minor trauma, growing rapidly over a period of weeks into a 2- to 10-mm, bright red, vascular, often pedunculated papule. They may occur anywhere on the body, but the head, neck, and upper extremities are most commonly affected. When traumatized, these lesions may bleed profusely, often requiring emergent medical attention. Surgical excision is the most definitive treatment option, although pulsed dye laser, cryosurgery, electrodesiccation, and topical β blockers can be useful.

16.**(C)** Port-wine stains (nevus flammeus, capillary malformation) are malformations of the superficial capillaries of the skin. These lesions are present at birth, permanent in nature, and grow with the child. The most successful treatment modality in use is the pulsed dye laser, which can result in 80–90% improvement in these lesions after a series of treatment sessions and can avoid future complications associated with vascular dilation. Treatment is more effective if undertaken in infancy. Overgrowth of underlying muscle and bone can occur and is frequently seen with facial lesions. Affected patients often need maxillofacial intervention from malalignment that develops.

17.**(A)** Most EM cases in children are precipitated by herpes simplex virus infection, although the infection may no longer be apparent by the time EM develops. Symptomatic treatment is usually sufficient. Oral antihistamines help suppress pruritus, stinging, and burning. The use of systemic corticosteroids is controversial but may be considered for severe mucosal disease. Antiviral medications targeting herpes simplex virus do not alter the course of the EM, although children with recurrent EM may be candidates for prophylactic antivirals. The prognosis is excellent, with most lesions lasting no more than 2 weeks. Healing occurs without scarring.

18.**(B)** SJS is defined as epidermal detachment of less than 10% of the body surface area, whereas SJS/TEN overlap has 10–30% and TEN has greater than 30% body surface area involvement.

19.**(D)** Reactive infectious mucocutaneous eruption is a newly recognized disorder characterized by a respiratory illness prodrome including fever and cough, severe mucositis (most often involving at least two mucosal surfaces), and a scarce to absent rash. When present, the rash associated with RIME is most often targetoid, but varying morphologies have been described. In contrast to SJS/TEN, which is caused by medications, RIME is caused by infection with Mycoplasma pneumoniae, Chlamydophila pneumoniae, or respiratory viruses such as rhinovirus, enterovirus, influenza, parainfluenza, and human metapneumovirus.

20.**(C)** Patients with Kawasaki disease may also have conjunctival injection and hyperemia of the mucous membranes; however, necrosis of the mucosal surfaces does not occur, and blistering, erosions, and severe crusting are not observed. The mucosal changes of staphylococcal scalded skin syndrome are minor, and the blistering of the skin is more superficial and favors intertriginous regions.

21.**(A)** The diagnosis of scabies can be confirmed by microscopic visualization of the mite, eggs, larvae, or feces in scrapings of papules or burrows examined under oil immersion. Skin biopsy is rarely necessary but may be useful if lesions have become nodular.

22.**(A)** Nevus anemicus is a white patch present since birth in variable sites caused by decreased blood flow through capillaries in the dermal papillae, by using diascopy this birthmark become indistinguishable from surrounding skin differentiate it from the other differential diagnosis mentioned.

23.**(B)** secondary candida infection can complicate irritant diaper dermatitis; the diagnosis of candidiasis is established by the presence of budding yeast and pseudohyphae on KOH examination.

24.**(D)** lichenifaication is an accentuated skin markings that result from epidermal thickening due to rubbing.

25.**(B)** Plaque type psoriasis is the most common variety of psoriasis.

26.**(A)** Seborrheic dermatitis is a common, chronic inflammatory disease that classically present in infants as cradle cap, it is theorized to represent an abnormal inflammatory response to commensal Malassezia species in sebum rich areas that include the scalp, eyebrows, eyelids, nasolabial folds, external auditory canals, posterior auricular folds and intertriginous areas.

27.**(D)** The OTC 1% permethrin cream rinses or lotions efficacy is poor due to resistance.

SECTION 26

ORTHOPEDICS

HAYDER ALMUSAWI

QUESTIONS

1. Which of the following orthopaedic terminologies is **TRUE**?
   A. Abduction: movement toward midline
   B. Adduction: movement away from midline
   C. Equinus: plantar flexion of the forefoot
   D. Pes cavus: flat foot

2. At what age the effect of in-utero-positioning resolve?
   A. 3–6 months
   B. 7–12 months
   C. 1-2 years
   D. 3–4 years

3. At what age toddlers walk independently?
   A. 12 months
   B. 15 months
   C. 18 months
   D. 24 months

4. A 5-year-old child presented with history of painful limping.
Of the following, the **MOST** likely cause is
   A. developmental dysplasia of the hip
   B. Legg-Calve-Perthes disease
   C. transient monoarticular synovitis
   D. lower extremity length inequality

5. Which of the following is **MOST** likely cause of painless limping in a 12-year-old boy?
   A. Myositis
   B. Rheumatologic disorder
   C. Juvenile idiopathic arthritis
   D. Developmental dysplasia of the hip

6. What is the **MOST** common fracture in paediatrics age group?
   A. Torus fracture
   B. Complete fracture
   C. Greenstick fracture
   D. Bowing fracture

7. An 18-month-old boy presented with history of crushing trauma to his right elbow, the patient developed progressive swelling of the affected area, and become agitated later on. On examination; the elbow was tense, markedly swollen, the pulse was not detected in

cubital region with severe pain that was not well controlled with prescribed medication and eventually have decreased sensation.
Of the following, the **MOST** likely cause is

A. compartment syndrome
B. deep vein thrombosis
C. cellulitis
D. rhabdomyolysis

8. An infant with congenital muscular torticollis, metatarsus adductus, and clubfoot should undergo a careful examination of the

A. upper extremities
B. spine
C. hips
D. lower extremities

9. Which of the following is a risk factor for developmental dysplasia of the hip?

A. Male infants
B. Face presentation
C. Firstborn child
D. Polyhydramnios

10. A 3-year-old female presented with history of abnormal gait, she is the 1st baby delivered by caesarean section due to breech presentation and oligohydramnios. On examination; the child has increased lumbar lordosis with waddling gait.
What is the **MOST** helpful test during physical examination?

A. Klisic test
B. Ortolani test
C. Barlow test
D. Galeazzi test

11. What is the imaging study of choice for initial evaluation of a 6-month-old infant with suspected developmental dysplasia of the hip?

A. Ultrasound
B. Plain radiographs
C. CT scan
D. MRI

12. What is the imaging study of choice for a 7-week-old infant with a positive Ortolani test?

A. Ultrasound
B. Plain radiograph
C. CT scan
D. MRI

13. A 5-year-old boy presented with limping and pain in the right hip joint and anterior thigh. Investigations reveal; WBC 10,000/cmm, ESR 6 mm/hr, anteroposterior and frog-leg radiographs of the hip are normal while ultrasonography reveals joint effusion.
What is the **MOST** likely diagnosis?

A. Toxic synovitis
B. Septic arthritis
C. Osteomyelitis
D. Legg-Calve-Perthes disease

14. What is the mainstay of treatment of transient synovitis?
A. Bed rest
B. NSAID
C. Antibiotics
D. Aspirin

15. A 7-year-old boy presented with painless limp associated with intermittent hip and anterior thigh pain. On examination; there is decreased internal rotation and abduction with thigh muscle spasm.
Of the following, the **MOST** likely diagnosis is
A. toxic synovitis
B. septic arthritis
C. slipped capital femoral epiphysis
D. Legg-Calve-Perthes disease

16. A 12-year-old obese boy presented with history of right hip pain; and inability to ambulate. On examination; the patient holds the affected extremity in external rotation, when the hip is flexed, it externally rotates with limitation of internal rotation.
Of the following, the **MOST** likely diagnosis is
A. toxic synovitis
B. septic arthritis
C. slipped capital femoral epiphysis
D. Legg-Calve-Perthes disease

17. A 5-year-old girl presented with history of W-sitting, kissing kneecaps and with running have an egg-beater gait.
What is the **MOST** likely cause?
A. Internal femoral torsion
B. Internal tibial torsion
C. External tibial torsion
D. Blount disease

18. Which of the following is a common cause of out-toeing?
A. External tibial torsion
B. Metatarsus adductus
C. Talipes equinovarus
D. Developmental dysplasia

19. During discussing the physiology of knock-knees with medical students, one of them asks at which age it usually resolved?
A. 2-4 years
B. 5-8 years

C. 9 - 12 years
D. >12 years

20. A 13-year-old male presented with history of pain during and after activity. On examination; tenderness and local swelling over the tibial tubercle.
What is the **MOST** likely diagnosis?
A. Discoid Lateral Meniscus
B. Osgood-Schlatter disease
C. Osteochondritis dissecans
D. Patellofemoral pain syndrome

21. An adolescent athletic female presented with history of anterior knee pain that worsens with activity, going up and down stairs, and soreness after sitting in one position for an extended time. On examination; there is no associated swelling. Palpating and compressing the patellofemoral joint with the knee extended elicits pain.
What is the **MOST** likely diagnosis?
A. Discoid Lateral Meniscus
B. Osgood-Schlatter disease
C. Osteochondritis dissecans
D. Patellofemoral pain syndrome

22. Which of the following is found in an infant with clubfoot?
A. Midfoot valgus
B. Forefoot abduction
C. Hindfoot equinus and valgus
D. Varying degrees of rigidity

23. What is the **MOST** common foot disorder in infants?
A. Clubfoot
B. Metatarsus adductus
C. Calcaneovalgus
D. Cavovarus

24. The mother of an 8-month-old infant with metatarsus adductus asks you about the age at which her child needs evaluation by pediatric orthopedist, if he does not improve.
The **BEST** answer will be
A. 9 months
B. 12 months
C. 18 months
D. 24 months

25. Hypermobile pes planus cannot be diagnosed until after the age of
A. 2 years
B. 4 years
C. 6 years
D. 8 years

26. A 9-year-old girl presented with heel pain increases with activity and decreases with rest. On examination; pain on palpation of the posterior calcaneus and tight heel cords.
What is the **MOST** likely diagnosis?

A. Calcaneal apophysitis
B. Cavovarus foot
C. Idiopathic avascular necrosis
D. Tarsal coalition

27. The shoulder actually comprises four joints, which one is commonly referred to as the shoulder joint?

A. Glenohumeral joint
B. Acromioclavicular joint
C. Sternoclavicular joint
D. Scapulothoracic joint

28. In which of the following condition computed tomography may be necessary to evaluate the great vessels?

A. An anteriorly displaced medial clavicle fracture
B. A posteriorly displaced medial clavicle fracture
C. An anteriorly displaced distal clavicle fracture
D. A posteriorly displaced distal clavicle fracture

29. An adolescent athlete complains of an insidious onset of low back pain persisting over 6 weeks. The pain tends to worsen with activity and extension of the back and improves with rest, the pain is radiated to the buttocks.
Of the following, the **MOST** likely cause is

A. diskitis
B. ankylosing spondylitis
C. vertebral osteomyelitis
D. spondylolisthesis

30. A 4-year-old child presented with back pain, abdominal pain, irritability, and refusal to walk or sit. The child holds his spine in a straight position with loss of lumbar lordosis. The white blood cell count is normal, but the ESR and CRP are high.
Of the following, the **MOST** likely cause is

A. diskitis
B. ankylosing spondylitis
C. spondylolisthesis
D. spondylolysis

31. A 10-year-old child presented with elbow pain, decreased range of motion, and tenderness to palpation over the capitellum. Radiographs reveal fragmentation of the capitellum.
Of the following, the **MOST** likely diagnosis is

A. medial humeral epicondyle apophysitis
B. Panner disease
C. osteochondritis dissecans of the capitellum
D. Little Leaguer's elbow

SECTION 26

ORTHOPEDICS

HAYDER ALMUSAWI

ANSWERS

1.**(C)** Abduction: movement away from midline, adduction: movement toward or across midline, equinus: plantar flexion of the forefoot, hindfoot, or entire foot, pes cavus: high medial arch of the foot, pes planus: flat foot.

2.**(D)** By the age of 3–4 years, the effects of in utero positioning have usually resolved.

3.**(C)** Toddlers will generally walk independently by 18 months of age.

4.**(C)**

5.**(D)** Causes of painful limping in children age 3-10 years include: septic arthritis, osteomyelitis, myositis, transient monoarticular synovitis, trauma, rheumatologic disorders, juvenile idiopathic arthritis, spondylodiscitis and malignancy.
Causes of painless limping in children age 11 years to maturity include: slipped capital femoral epiphysis (chronic; stable), developmental dysplasia of the hip, acetabular dysplasia, lower extremity length inequality and neuromuscular disorder.

6.**(B)** Complete fractures occur when both sides of bony cortex are fractured. This is the most common fracture and may be classified as comminuted, oblique, transverse, or spiral, depending on the direction of the fracture line.

7.**(A)** Compartment syndrome is an orthopedic emergency that results from hemorrhage and soft tissue swelling within the tight fascial compartments of an extremity. The most common sites are the lower leg (tibial fracture) and arm (supracondylar fracture). Swollen and tight compartments and pain with passive stretching are present. Affected patients have severe pain not well controlled with prescribed medication and may eventually have decreased sensation in the dermatomes supplied by the nerves located in the compartment or decreased perfusion of the extremity if not promptly identified and treated.

8.**(C)** Congenital muscular torticollis, metatarsus adductus, and clubfoot are associated with DDH. An infant with any of these three conditions should receive a careful examination of the hips.

9.**(C)** Physiologic risk factors for DDH include a generalized ligamentous laxity, perhaps from maternal hormones that are associated with pelvic ligament relaxation (estrogen and relaxin). Female infants are at higher risk (9:1); family history is positive in 20% of all patients with DDH. Other risk factors include breech presentation, firstborn child (60%), oligohydramnios, and intrauterine and postnatal infant positioning.

10.**(A)** The Klisic test is useful in identifying bilateral DDH; it is done by placing the third finger over the greater trochanter and the index finger over the anterior superior iliac spine,

then drawing an imaginary line between the two. The line should point to the umbilicus in a normal child. However, in a dislocated hip, the greater trochanter is elevated, which causes the line to project lower (between the umbilicus and pubis).

11.**(B)**

12.**(A)** Ultrasound is used for initial evaluation of infants with DDH and should be done for females with a positive family history or breech presentation in both sexes. Ultrasound should be obtained after 6 weeks of age in the absence of abnormal examination findings to avoid confusion with physiologic laxity. Because the femoral head doesn't begin to ossify until 4–6 months of age, plain radiographs can be misleading until patients are older. Plain radiographs are the imaging study of choice once the child reaches 6 months of age.

13.**(A)**

14.**(A)** Transient synovitis, also known as toxic synovitis, is a common cause of limping in children. The etiology of transient synovitis is uncertain; possible causesare viral illness and nonspecific inflammatory reactions. The mean age at onset is 6 years, with a range of 3–8 years. It is twice as common in male children. An acute onset of pain in the groin/hip, anterior thigh, or knee occurs. Patients with transient synovitis are often afebrile, walk with a painful limp, and have normal to minimally elevated white blood cell count (WBC), C-reactive protein (CRP), and erythrocyte sedimentation rate (ESR). Anteroposterior and frog-leg radiographs of the hip are usually normal. Ultrasonography may reveal a joint effusion. The mainstay of treatment is bed rest and minimal weight bearing until the pain resolves.

15.**(D)** Legg-Calve-Perthes disease (LCPD) is idiopathic avascular necrosis (osteonecrosis) of the capital epiphysis of the femoral head. LCPD commonly presents in patients 3–12 years of age, with a mean age of 7 years. It is four to five times more common in boys. Patients may not present for several weeks because of minimal discomfort; the classic presentation is a child with an atraumatic, painless limp. There may be mild or intermittent hip/groin, anterior thigh, or knee pain. Decreased internal rotation and abduction with some discomfort, thigh muscle spasm, and anterior thigh muscular atrophy may be present.

16.**(C)** Slipped capital femoral epiphysis (SCFE) is an adolescent hip disorder that is an orthopedic emergency. Approximately 20% of patients with SCFE will have bilateral involvement at presentation, and another 20–40% may progress to bilateral involvement. The average age is 10–16 years, with a mean of 12 years in males and 11 years in females. The presentation is variable, based on severity and type of slip. Patients will often report hip, thigh, or knee pain; limp or inability to ambulate; and decreased hip range of motion. There may or may not be a traumatic event. The patient usually holds the affected extremity in external rotation. As the hip is flexed, it will progressively externally rotate (obligate external rotation). There is usually a limitation of internal rotation, but there may also be a loss of flexion and abduction. If the patient can bear weight, it is typically an antalgic gait with the affected leg in external rotation. It is important to examine both hips given the high rate of bilateral involvement.

17.**(A)** Internal femoral torsion or femoral anteversion is the most common cause of in-toeing in children 2 years or older. The rotation is maximal between 4 and 6 years of age, and then decreases. It occurs twice as often in females. Many cases are associated with generalized ligamentous laxity. The etiology of femoral anteversion is likely congenital and is common in individuals with abnormal sitting habits such as W-sitting.

18.**(A)** Common causes of in-toeing include internal femoral torsion or anteversion, internal tibial torsion, metatarsus adductus, talipes equinovarus (clubfoot) and developmental dysplasia. Common causes of out-toeing include external femoral torsion or retroversion, external tibial torsion. calcaneovalgus feet, hypermobile pes planus (flatfoot) and slipped capital femoral epiphysis.

19.**(B)** Physiologic knock-knees are most common in 3–4-year-olds and usually resolve between 5 and 8 years of age.

20.**(B)** Osgood-Schlatter disease is a common cause of knee pain at the insertion of the patellar tendon on the tibial tubercle. It usually occurs after a growth spurt and is more common in boys. The age at onset is typically 11 years for females and 13–14 years for males. Patients will present with pain during and after activity and often have tenderness and local swelling over the tibial tubercle.

21.**(D)** Idiopathic anterior knee pain is a common complaint in adolescents. It is particularly prevalent in adolescent female athletes. Previously, this was referred to as chondromalacia of the patella, but this term is incorrect as the joint surfaces of the patella are often normal. It is now known as patellofemoral pain syndrome (PFPS). The patient will present with anterior knee pain that worsens with activity, going up and down stairs, and soreness after sitting in one position for an extended time. There is usually no associated swelling. The patient may complain of a grinding sensation under the kneecap. Palpating and compressing the patellofemoral joint with the knee extended elicits pain.

22.**(D)** The infant will have midfoot cavus, forefoot adduction, hindfoot equinus and varus, and varying degrees of rigidity.

23.**(B)**

24.**(D)** True metatarsus adductus resolves spontaneously over 90% of the time without treatment, so reassurance is all that is needed. Metatarsus adductus that does not improve within 2 years needs evaluation by a pediatric orthopedist.

25.**(C)** Hypermobile pes planus cannot be diagnosed until after 6 years of age; before that, it is developmental pes planus.

26.**(A)** Calcaneal apophysitis is a common cause of heel pain among active young people. The mean age of presentation for girls is about 9 years of age and for boys about 11–12 years. The common presentation is a young athlete who develops heel pain with activity that decreases with rest. Swelling is rare, but limping may be associated with severe disease. The child will have pain on palpation of the posterior calcaneus and often tight heel cords

27.**(A)**

28.**(B)**

29.**(D)**

30.**(A)** Diskitis is an intervertebral disk space infection that does not cause associated vertebral osteomyelitis. The most common organism is Staphylococcus aureus. The infection can occur at any age but is more common in patients under 6 years of age.

31.**(B)** Panner disease is an osteochondritis of the capitellum (lateral portion of distal humeral epiphysis) that occurs spontaneously in late childhood. Clinical features include elbow pain, decreased range of motion, and tenderness to palpation over the capitellum. Radiographs reveal fragmentation of the capitellum. Treatment is activity restriction and follow-up radiographs to demonstrate spontaneous reossification of the capitellum over several months. There is usually no need for further treatment or imaging studies. This is not to be confused with osteochondritis dissecans of the capitellum, which usually will occur in adolescents involved with throwing sports.