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MCQs in Pediatrics

REVIEW OF
NELSON TEXTBOOK OF
PEDIATRICS

ZUHAIR M. ALMUSAWI

20
EDITION

MCQs in Pediatrics

**Review of Nelson TEXTBOOK of PEDIATRICS
(20th Edition)**

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Dedication

**We dedicate this book to our parents, wives, daughters, sons, and those whom without them we could not be able to write our book, those who are fighting to create peace and life (The Iraqi Military Forces and the Popular Mobilization)
May ALLAH bless them all.**

Special thanks are extended to our colleague, ALI ALMUSAWI for his help in designing the cover of this book.

Number of Deposition of This Book at Iraqi House of Books and Documents (Iraqi National Library) /2016

Preface

Perhaps the most effective way to use this book is to allow you one minute to answer each question in a given section in order to approximate the time limits imposed by the Iraqi and Arabic Board Pediatrics Examinations. As you proceed, indicate your answer to each question.

When you have finished answering the questions in a section, you should then spend as much time as you need verifying your answers and reading the explanations. Although you should pay special attention to the explanations for the questions you answered incorrectly, you should read every explanation. The authors had designed the explanations to reinforce and supplement the information tested by the questions. If, after reading the explanations, you want more information, you should study the text.

The information contained within this book was obtained by the authors from Nelson TEXTBOOK of PEDIATRICS (20th Edition).However , while every effort has been made to ensure its accuracy, **no responsibility for loss, damage or injury occasioned by any person acting or refraining from action as a result of information contained herein can be accepted by the authors.**

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PART I

The Field of Pediatrics QUESTIONS

AHMED TAWFIQ

1. The **MOST** common cause of under-5 mortality in developing world is
 - A. diarrheal disease
 - B. pneumonia
 - C. malaria
 - D. measles
 - E. neonatal disease
2. The **LEAST** common cause of death < 1 yr in USA is
 - A. congenital malformations
 - B. short gestation and low birth weight
 - C. sudden infant death syndrome
 - D. unintentional injuries
 - E. bacterial sepsis of newborn
3. In mental retardation, all the following are true **EXCEPT**
 - A. constitute 1-3% of children
 - B. 80% are mild form
 - C. in severe form socio-economic status is a considerable factor
 - D. higher percentage in low birthweight babies
 - E. fragile X chromosome is a recognized cause
4. An 8-month-old infant with confirmed diagnosis of spinal muscular atrophy type 1 referred back from tertiary care center with "DNAR" status. This means during acute illness or an emergency the child should
 - A. receive standard care and adrenaline injection
 - B. receive supportive care
 - C. receive active resuscitation but not mechanical ventilation
 - D. O₂ is not part of treatment options
 - E. none of the above
5. A full term neonate with Down syndrome and esophageal atresia admitted in Pediatric intensive care unit (PICU). The parents want "no interference" approach. Of the following, the **BEST** response is to
 - A. act according to parents well
 - B. arrange the operation with the surgeon
 - C. work for the patient best interest

- D. give supportive care only (DANR)
- E. wait for a clear legislation

6. You are resident in Pediatrics, called at midnight to attend delivery of expected baby with multiple malformations and oligohydromnias as had been reported in antenatal scan, mother in stage I labor so far.

Of the following, your **BEST** plan of action is to

- A. counsel the parents that you will go for (DNAR) status
- B. start your usual resuscitation waiting for your senior
- C. explain that the result of antenatal scan is final
- D. call your consultant or senior resident
- E. apply palliative care for the coming neonate

7. A 7-year-old boy presented to the out-patient department with fever and cough, you have diagnosed him as a case of pneumonia and decided to admit and give parental therapy, the child refused.

Of the following, the **BEST** response is to

- A. explain to father about dangers of omitting treatment
- B. try to explain the risk to the child
- C. give the chance for oral antibiotics and accept the child decision
- D. inform your consultant
- E. inform hospital ethical committee

8. You are treating acutely ill 6-year-old child with ascending paralysis, he eventually needs mechanical ventilation. Now you are in a situation of using the last bed in respiratory care unit (RCU).

Of the following, the **BEST** approach is to

- A. try to convince the parents that, there is no use of ventilation
- B. keep the bed for more treatable condition
- C. try bedside ventilation
- D. consider this case as potential treatable condition and use the last bed
- E. contact a nearby region RCU and transfer the child

9. You are managing a 10-year-old child with spastic quadriplasia, who is globally retarded, had recurrent seizures, and severe recurrent chest infections which requires frequent admissions to respiratory care unit (RCU) and ventilation. You are trying to convince the mother about non-benefit of future RCU admissions.

All of the following statements are true **EXCEPT**

- A. use an obligations of no bed availability
- B. consider mother emotional status and religious believes
- C. be empathetic
- D. explain disease chronicity and non-response status

E. explain the suffering of child in each ventilation

10. The well-child care (anticipatory guidance) intends to promote the physical and emotional well-being of children. The tasks of each well-child visit (which usually takes 18 min time) include all the following **EXCEPT**

- A. disease detection
- B. disease prevention
- C. treatment plans
- D. health promotion
- E. accident prevention

11. After the first year of life, the **MOST** common cause of death in children is mainly

- A. none-accidental injuries
- B. burn
- C. drowning
- D. accidents
- E. war

12. The **MOST** successful injury prevention strategy is

- A. car seats
- B. water safety
- C. changing in product design
- D. poison prevention
- E. fall prevention

13. Around 44,000-98,000 patients die in U.S. hospitals each year because of preventable medical errors, these figures can be reduced if adapting an emerging new science of Quality Improvement (QI).

The **TRUE** figure of children receive recommended health care in USA is about

- A. 16%
- B. 26%
- C. 36%
- D. 46%
- E. 80%

14. Patient culture can change the doctor treatment options and even the way of behavior with the patient.

All the following culture values are expected if he is dealing with Muslim patients **EXCEPT**

- A. fasting during Ramadan month
- B. modesty and female body cover
- C. forbiddance of touch and shake hands of opposite sex

- D. routine postmortem exam
- E. patriarchal way of decision making

- 1.(E). Causes of under-5 mortality differ markedly between developed and developing nations. In developing countries, 66% of all deaths resulted from infectious and parasitic diseases. Among the 42 countries having 90% of childhood deaths, diarrheal disease accounted for 22% of deaths, pneumonia 21%, malaria 9%, AIDS 3%, and measles 1%. Neonatal causes contributed to 33% of the total death but 24% of deaths are caused by severe infections and 7% by tetanus.
- 2.(E). Bacterial sepsis constitutes only 2%. Other distracters are arranged from common to least common.
- 3.(C). In severe form of mental retardation the socio-economic factor is not effective parameter while in mild forms it plays a role.
- 4.(B). DNAR status is a statement issued by medical ethical group (which is a group of consultants) regarding certain untreatable medical illnesses, it means do not attempt active resuscitation, so all supportive care, medications, and O2 are allowed, adrenaline is a medication used in active resuscitation.
- 5.(C). Pediatrician is a child advocate; he must take decision always to the "best interest" of the child that what is called beneficent paternalism in pediatrics which is the obligation to act in a younger child's best interest that takes moral precedence over the wishes of the child's parent(s), so in such a scenario arrangement with the surgeon to do the operation and calling the local hospital committee to protect the child is the best choice but this action is still need legislation in the third world. In a such situation the Pediatrician must explain the benefit of treatment to the parents (baby best interest) trying best efforts to save the baby keeping in mind Down syndrome babies are not DNAR status.
- 6.(D). In such situation when you have time (mother still in stage I labor), the best response is to call your direct senior resident or consultant then explain the result of antenatal scan trying to put the parents in the expected situation. Starting the usual resuscitation is the best choice when there is no time and the baby arrived. The decision of (DNAR) should be made by the ethical hospital committee or supervising consultant and should be powered by national legislations.
- 7.(C). In older children and in noncritical situation the principle of the "child assent" which means the child voice must be heard should be applied. In this scenario, the situation is mild and the child non acceptance of injectable medication should be respected. It is still in critical situation or more acute illness the child best interest should be sought and it may be against his well.
- 8.(D). It is clear in this scenario that the child suffers potentially treatable condition. In this situation use the best resources and the best care. The choices of (A&B) are

definitely non ethical. Other choices are optional when there is no bed is available in RCU taking the best interest for the patient in consideration.

9.(A). In such difficult situation (chronic disabling illnesses) and the child in vegetative state; use the approach from B to E steps and allow the final decision to the parents especially if you are not covered with clear legislations.

10.(C). This is a well child visit; treatment plans are not included here.

11.(D). The most common cause of death in children beyond the first few months of life is injuries which includes road traffic accidents and other none-intentional injuries, with up to 50-60% of total causes of death.

12.(C). The most successful injury-prevention strategies generally are those involving changes in product design. Such as making more secure cars, more suitable car seats ...etc.

13.(D).

14.(D). All the above values are expected from Muslim culture, except submitting the dead body for routine postmortem exam. The value (E) means the way of decision making is usually left to older person in the family.

PART II**Growth, Development, and Behavior
QUESTIONS****AHMED TAWFIQ**

1. In embryonic period, all the following are true **EXCEPT**
 - A. formation of blastocyst by 8 days
 - B. formation of endoderm and ectoderm by 2 weeks
 - C. formation of mesoderm by 6 weeks
 - D. formation of human like embryo by 8 weeks
 - E. the crown-rump length is about 3 cm

2. In fetal period, all the following are true **EXCEPT**
 - A. by 10 weeks the midgut returns to abdomen
 - B. by 12 weeks external genitalia becomes clearly distinguishable
 - C. by 24 weeks surfactant production begun
 - D. by 26 weeks recognizable human face formed
 - E. during third trimester the weight triples

3. All the following are recognizable teratogens **EXCEPT**
 - A. ethanol
 - B. antiepileptic medications
 - C. toxoplasmosis
 - D. hypothermia
 - E. mercury

4. Prenatal exposure to cigarette smoke is associated with
 - A. lower birthweight
 - B. shorter length
 - C. changes in neonatal neurodevelopmental status
 - D. neonatal diabetes
 - E. learning problems

5. A 16-year-old G₁P₁ mother complaining of reduced milk production and breasts engorgement. She stated that she feels unhappy and scared most of the times. Her husband is a soldier and she is living with his family.
Of the following, the **MOST** appropriate action is to
 - A. refer her to obstetrician
 - B. explain the appropriate way of milk expression
 - C. advice milk stimulant medications
 - D. apply Edinburgh postnatal scale

- E. stop breast feeding
6. A mother of a 2-week-fullterm baby noted doll's-eye movement of her baby eyes. Of the following, The **MOST** appropriate next action is to
- A. reassures her by informing her that, this is a normal reaction
 - B. refer the baby to an ophthalmologist
 - C. takes a detailed history of perinatal period
 - D. order brain ultrasound
 - E. order brain MRI
7. The age at which the infant achieve early head control with bobbing motion when pulled to sit is
- A. 2 mo
 - B. 3 mo
 - C. 4 mo
 - D. 5 mo
 - E. 6 mo
8. The age at which the infant can reach an object, grasp it and bring it to mouth and seems excited when see the food is
- A. 4 mo
 - B. 5 mo
 - C. 6 mo
 - D. 7 mo
 - E. 8 mo
9. By the age of 7 months the infant is able to do all the following **EXCEPT**
- A. transfer object from hand to hand
 - B. bounces actively
 - C. cruises
 - D. grasp uses radial palm
 - E. roll over
10. You are specialist explaining the developmental implication of a 6-month-old baby boy who "transfers object hand to hand" to college students. Of the following, the **BEST** statement describing that is a/an
- A. visuomotor coordination
 - B. voluntary release
 - C. comparison of objects
 - D. ability to explore small objects
 - E. increasing autonomy

11. Building a tower of 6 cubes by a 22-month-old child requires
- A. visuomotor coordination
 - B. ability uses objects in combination
 - C. visual, gross, and fine motor coordination
 - D. able to link actions to solve problems
 - E. symbolic thought
12. A 10-month-old child can do all the following **EXCEPT**
- A. follows one-step command without gesture
 - B. says "mama" or "dada"
 - C. points to objects
 - D. speaks first real word
 - E. inhibition to "no"
13. You are observing a 15-month-old toddler, he was able to do all the following **EXCEPT**
- A. walks alone
 - B. makes tower of 3 cubes
 - C. inserts raisin in a bottle
 - D. responds to his/her name
 - E. identifies 1 or more parts of body
14. The age by which the child can makes tower of 9 cubes and imitates circular stroke is
- A. 24 mo
 - B. 30 mo
 - C. 36 mo
 - D. 42 mo
 - E. 48 mo
15. The child who continue to search for a hidden subject, has achieved the developmental age of
- A. 7 mo
 - B. 9 mo
 - C. 11 mo
 - D. 13 mo
 - E. 15 mo
16. The child who's able to stands momentarily on 1 foot, makes tower of 10 cubes; imitates construction of "bridge" of 3 cubes; copies circle and imitates cross is
- A. 24 mo old
 - B. 30 mo old

- C. 36 mo old
- D. 42 mo old
- E. 48 mo old

17. A mother to a 4-year-old child who has pauses and repetitions of initial sounds visited outpatients department.

Of the following, the **MOST** appropriate advice is

- A. this is a normal phenomenon affecting about 5% of preschool children
- B. there is no need for action as 80% of affected children recover by their own
- C. tries to reduce pressures associated with speaking
- D. I'll refer him to ENT specialist for further evaluation
- E. I'll refer him to speech therapist

18. These facts are true regarding the developmental stage of preschool children **EXCEPT**

- A. handedness is achieved by 3 years of age
- B. boys are usually later than girls in achieving bladder control
- C. knowing gender by 4 years
- D. egocentric thinking
- E. masturbation

19. A worried mother of a 4-year-old boy describing attacks of inconsolable crying episodes of her child, taking long time, she stated also that he prefers to play alone.

Of the following, the **MOST** appropriate action is to

- A. reassures her that this is a normal phenomenon of temper tantrum
- B. seek more history regarding other skills and developmental domains
- C. refer her to pediatric psychiatry
- D. investigate social issues of the family
- E. investigate for the child abuse

20. A young couples of a 2-month-old baby girl with excessive crying seek your medical advice. Mother said that the baby is crying about 3 hr in a day, 2-3 days per week. She is intermittently spitting but she is gaining weight adequately. She is bottle fed baby.

Of the following, the **LEAST** important advice is to

- A. master the situation in relaxed manner
- B. adhere to precry cues
- C. change milk formula
- D. avoid sensory overstimulation
- E. reassure about benign nature of illness

21. Of those babies who have prolonged crying episodes in the first 2 mo of life, the percentage that will remain having similar episodes is about

- A. 1%
- B. 5%
- C. 10%
- D. 30%
- E. 50%

22. A medical student asked you during the morning round about his observation of infants cry in response to the cry of another infant.

Of the following, the **MOST** appropriate answer is that it represent

- A. an early sign of empathy development
- B. a sign of good hearing
- C. a startle reflex to a loud sound
- D. an early sign of fear development
- E. an early sign of autistic behavior

23. Between 2-6 months of life, the infant start to achieve a regular sleep–wake cycles.

All the following are true about infant sleep during this period **EXCEPT**

- A. total sleep hours are about 14-16 hr/24 hr
- B. sleeps about 9-10 hr concentrated at night
- C. sleeps 2 naps/day
- D. sleep electroencephalogram shows the mature pattern
- E. the sleep cycle time is similar to that of adults

24. A highly careful mother of a 10-month-old baby boy complaining that her baby has inadequate weight gain. She explained to you how she is so strict and so careful to feed her baby by spoon, but he was always refusing.

Of the following, the **LEAST** helpful advice is to

- A. respect infant independence
- B. offer softer diet
- C. use 2-spoons (1 for the child and 1 for the parent)
- D. use finger foods
- E. use high chair with tray table

25. The child who helps to undress; puts 3 words together (subject, verb, object); and handles spoon well, his/her **MOST** appropriate age is around

- A. 15 mo
- B. 18 mo
- C. 24 mo
- D. 30 mo
- E. 48 mo

26. The child who plays simple games (in “parallel” with other children); helps in dressing and washes hands, his/her **MOST** appropriate developmental age is around
- A. 15 mo
 - B. 18 mo
 - C. 24 mo
 - D. 30 mo
 - E. 48 mo
27. The child who is able to imitates construction of “gate” of 5 cubes; draws a man with 2-4 parts besides head and identifies longer of 2 lines, his/her **MOST** appropriate developmental age is around
- A. 24 mo
 - B. 30 mo
 - C. 48 mo
 - D. 54 mo
 - E. 60 mo
28. The birthweight usually quadruples by the age of
- A. 1.5 yr
 - B. 2 yr
 - C. 2.5 yr
 - D. 3 yr
 - E. 3.5 yr
29. Regarding the physical growth of preschool children (3-5 yr); all the following are true **EXCEPT**
- A. 4-5 kg weight increment/yr
 - B. 7-8 cm height increment/yr
 - C. head will grow only an additional 5-6 cm up to 18 yr
 - D. all 20 primary teeth have erupted by 3 yr
 - E. average wt 20 kg and height 40 inch by 4 yr
30. Regarding the physical growth of middle childhood (6-11 yr); all the following are true **EXCEPT**
- A. 3-3.5 kg wt increment/yr
 - B. 6-7 cm height increment/yr
 - C. brain myelinization stops by 8 yr
 - D. first deciduous tooth falls by 6 yr
 - E. risk for future obesity
31. The first permanent tooth to erupt is
- A. central incisor at 6 yr

- B. molar at 6 yr
- C. premolar at 6-7 yr
- D. lower canine at 6-7 yr
- E. upper canine at 6-7 yr

32. The following statements about sleep are true **EXCEPT**

- A. melatonin which is secreted in dark-light cycles is secreted from hypothalamus
- B. slow-wave sleep is the first cycle of sleep
- C. rapid eye movement (REM) sleep is responsible for dreams
- D. both cycles are needed for sufficient sleep
- E. REM sleep protect brain from injury

33. The **MOST** common cause of sleeping difficulty in the first 2 months of life is

- A. gastro-esophageal reflux
- B. formula intolerance
- C. colic
- D. developmentally sleeping behavior
- E. recurrent self resolving intussusceptions

34. A mother of 4-month-old baby boy complaining that the baby never goes to sleep unless he is rocked for quite long time. She needs to repeat the same issue whenever he is awake at night. She was surprised as no one of her previous babies had the similar behavior. She feels tired because of insufficient sleep.

Of the following, the **MOST** common cause of this baby problem is

- A. an early signs of ADHD
- B. behavioral insomnia of childhood, sleep-onset association
- C. primary sleep disorder
- D. restless legs syndrome
- E. sleep terrors

35. Parents of a 4-year-old boy complaining that he is refusing to go to bed, he remains active and playing.

Of the following, the **MOST** likely cause of this child problem is

- A. thyrotoxicosis
- B. ADHD
- C. primary sleep disorder
- D. limit setting; behavioral insomnia of childhood
- E. behavioral insomnia of childhood, sleep-onset association

36. All the following are included in basic principles of healthy sleep **EXCEPT**

- A. set bedtime routine
- B. avoid stimulating activities as playing computer games

- C. makes sure your child spends time outside every day
- D. gives a heavy meal within an hour or 2 of bedtime
- E. keeps the television set out of your child's bedroom

37. All the following are compatible with the definition of obstructive sleep apnea **EXCEPT**

- A. repeated episodes of prolonged upper airway obstruction
- B. apnea
- C. $\geq 30\%$ reduction in airflow
- D. $\geq 30\%$ O₂ desaturation
- E. disrupted sleep

38. Of the following, the **MOST** common cause of obstructive sleep apnea in children is

- A. adenotonsillar hypertrophy
- B. obesity
- C. allergies
- D. craniofacial abnormalities
- E. pharyngeal reactive edema due to gastroesophageal reflux

39. Persons with Down syndrome are at particularly high risk for obstructive sleep apnea with up to 70% prevalence.

All the following are considered as risk factors **EXCEPT**

- A. peculiar facial anatomy
- B. hypotonia
- C. developmental delay
- D. central adiposity
- E. hypothyroidism

40. All the following are parasomniac disorders **EXCEPT**

- A. narcolepsy
- B. sleepwalking
- C. sleep terror
- D. confusional arousal
- E. nightmare

PART II**Growth, Development, and Behavior
ANSWERS****AHMED TAWFIQ**

- 1.(C). Formation of mesoderm by 4 weeks.
- 2.(D). Usually by 10 weeks.
- 3.(D). Teratogens associated with gross physical and mental abnormalities include various infectious agents (toxoplasmosis, rubella, syphilis); chemical agents (mercury, thalidomide, antiepileptic medications, and ethanol), high temperature, and radiation.
- 4.(D). Prenatal exposure to cigarette smoke is associated with lower birthweight, shorter length, and smaller head circumference, as well as changes in neonatal neurodevelopmental assessments. Later, these children are at increased risk for learning problems, externalizing behavior disorders, and long-term health effects.
- 5.(D). Pediatricians must be alert to detect postpartum depression. Edinburgh Postnatal Depression Scale is the standard to apply; if the score achieved, the mother should be referred to psychiatrist.
- 6.(A). This is normal finding in neonatal period.
- 7.(B). This is a developmental skill of 3 months of age which include also; lifts head and chest with arms extended on prone; head above plane of body on ventral suspension; reaches toward and misses objects; waves at toy; listens to music and says “aah, ngah”.
- 8.(A). By this age he also able to lifts head and chest, with head in approximately vertical axis; achieve no head lag when pulled to sitting position, enjoys sitting with full truncal support when held erect, pushes with feet when sees raisin, but makes no move to reach for it, laughs out loud; and may show displeasure if social contact is broken.
- 9.(C). Cruises or walks’ holding to furniture is by 10 months of age.
- 10.(C). The developmental implication reflects cognitive development.
- 11.(C).
- 12.(D). This is a 12 months developmental skill.
- 13.(E). This is an 18 months developmental skill.
- 14.(B). A 30 months old child also able to goes up stairs alternating feet, makes vertical and horizontal strokes, refers to self by pronoun “I”; knows full name, helps put things away and pretends in play.
- 15.(B). A major milestone is the achievement by 9 mo of object permanence (constancy), the understanding that objects continue to exist, even when not seen. At 4-7 month of age, infants look down for ball that has been dropped but quickly give up if it is not seen. With object constancy, infants persist in searching. They will find objects hidden under a cloth or behind the examiner’s back.
- 16.(C).

17.(C). Developmental dysfluency or stuttering it includes a pauses and repetitions of initial sounds. Stress or excitement exacerbates these difficulties, which generally resolve on their own. Although 5% of preschool children will stutter, it will resolve in 80% of those children by age 8 yr. Children with stuttering should only be referred for evaluation if it is severe, persistent, or associated with anxiety, or if parental concern is elicited. The best response is to give guidance to parents to reduce pressures associated with speaking. Other statements can be used for reassurance and for explaining the natural history of the problem.

18.(C). Bowel and bladder control emerge during this period, with “readiness” for toileting having large individual and cultural variation. Bed-wetting is normal up to age 4 yr in girls and age 5 yr in boys. Thinking in this period characterized by magical thinking (the child might believe that people cause it to rain by carrying umbrellas, that the sun goes down because it is tired), and egocentrism which refers to a child’s inability to take another’s point of view. Gender identification usually achieved by 3 years of age. Curiosity about genitals and adult sexual organs is normal, as is masturbation. But excessive masturbation interfering with normal activity, acting out sexual intercourse, extreme modesty, or mimicry of adult seductive behavior all suggests the possibility of sexual abuse or inappropriate exposure.

19.(B). Classical temper tantrums of children usually result from fear, overtiredness, inconsistent expectations, or physical discomfort. Tantrums normally appear toward the end of the 1st yr of life and peak in prevalence between 2 and 4 years of age. Tantrums lasting more than 15 min or regularly occurring more than 3 times/day may reflect underlying medical, emotional, or social problems. In the scenario the episodes takes long time to resolve and the mother points to problem in social development. Here we need to investigate the developmental domains and to do systemic examination to checkout for any developmental/physical problem before referring the child to pediatric psychiatrist which the next most appropriate choice.

20.(C). Prolong crying episodes is present in about 20% of infants younger than 2 mo of age. Crying (longer than 3 hr/day, longer than 3 days/wk lasting longer than 1 wk) and more often crying/fussiness persisting longer than 3-5 mo might be associated with; child abuse; behavioral problems when an older child (anxiety, aggression, hyperactivity); decreased duration of breastfeeding and postnatal depression (uncertain which is the cause or effect). Most infants with crying/fussiness do not have gastroesophageal reflux, lactose intolerance, or cow-milk protein allergy.

21.(B).

22.(A).

23.(E). The sleep cycle remains shorter than in adults (50-60 min vs approximately 90 min). As a result, infants who arouses frequently during the night, considered as behavioral sleep problem.

24.(B). Poor weight gain at this age often reflects a struggle between an infant’s emerging independence and parent’s control of the feeding situation.

25.(C).

- 26.(D).
- 27.(C).
- 28.(C).
- 29.(A). 2kg/yr.
- 30.(C). Brain myelination continues into adolescence, with peak gray matter at 12-14 yr.
- 31.(B). The first permanent molars usually erupt between ages 6 and 7 years. For that reason, they often are called the “six-year molars.” They are among the “extra” permanent teeth in that they don’t replace an existing primary tooth. These important teeth sometimes are mistaken for primary teeth. The six-year molars also help determine the shape of the lower face and affect the position and health of other permanent teeth. The first deciduous teeth to fall are central incisors.
- 32.(A). Melatonin is secreted from pineal body. Two sleep cycles; the slow-wave sleep (SWS) (i.e., δ , or deep sleep) appears to be the most restorative form of sleep; it is entered into relatively quickly after sleep onset, it is preserved in the face of reduced total sleep time, and it increases (rebounds) after a night of restricted sleep. Rapid eye movement (REM) sleep (stage α or “dream” sleep) appears to be involved in (1) completing vital cognitive functions, such as the consolidation of memory; (2) promoting the plasticity of the central nervous system (CNS); and (3) protecting the brain from injury.
- 33.(D). Most sleep issues that are perceived as problematic at this stage represent a discrepancy between parental expectations and developmentally appropriate sleep behaviors. Newborns that are noted by parents to be extremely fussy and persistently difficult to console are more likely to have one of the mentioned medical issues.
- 34.(B). In this situation, the child learns to fall asleep only under certain conditions or associations, which typically require parental presence, such as being rocked or fed, and does not develop the ability to self-soothe. In babies, gradually weaning the child from dependence, and in older children introduction of more appropriate sleep associations that will be readily available to the child during the night (transitional objects, such as a blanket or toy), in addition to positive reinforcement (i.e., stickers for remaining in bed), is often beneficial.
- 35.(D). Is often the result of parental difficulties in setting limits and managing behaviour in general, and the inability or unwillingness to set consistent bedtime rules and enforce a regular bedtime in particular.
- 36.(D). Other advices also include; don’t send your child to bed hungry. A light snack (such as milk and cookies) before bed is a good idea. Heavy meals within an hour or 2 of bedtime, however, may interfere with sleep. Avoid products containing caffeine for at least several hours before bedtime. These include caffeinated sodas, coffee, tea, and chocolate. Keep your child’s bedroom quiet and dark. A low-level night light is acceptable for children who find completely dark rooms frightening. Keep your child’s bedroom at a comfortable temperature during the night ($<24^{\circ}\text{C}$). Don’t use your child’s

bedroom for time-out or punishment. Keep the television set out of your child's bedroom.

37.(D). $\geq 3\%$ O₂ desaturation.

38.(A).

39.(C). Macroglosia.

40.(A). Narcolepsy is a recurrent episode of excessive daytime sleepiness, while parasomnia is an episodic nocturnal behavior that often involves cognitive disorientation and autonomic and skeletal muscle disturbance.

PART III

Behavior and Psychiatric Disorders QUESTIONS

AHMED TAWFIQ

1. The percentage of mental illnesses that is encountered at least once in any stage during childhood is about

- A. 10%
- B. 20%
- C. 30%
- D. 40%
- E. 50%

2. Clinicians tools needed to recognize early symptoms of mental disorders are called "Mental Health Action Signs".

All the following are true **EXCEPT**

- A. involvement in many fights, wanting to badly hurt others
- B. not eating, throwing up, or using laxatives to make yourself lose weight
- C. feeling very sad or withdrawn for more than 2 mo
- D. sudden overwhelming fear for no reason
- E. worries or fears that get in the way of your daily activities

3. All the following are true associations between psychiatric illness in childhood and their treatment **EXCEPT**

- A. attention deficit/hyperactivity (ADHD) and atomoxetine
- B. anxiety and antidepressant
- C. aggression and atypical antipsychotic
- D. psychosis and typical antipsychotic
- E. depression and antidepressant

4. The following medications are truly matched to their major pharmacological groups **EXCEPT**

- A. methylphenidate stimulant
- B. atomoxetine serotonin-norepinephrine reuptake inhibitor
- C. escitalopram selective serotonin reuptake inhibitors
- D. fluoxetine..... tricyclic antidepressants
- E. risperidone.....atypical antipsychotics

5. Which one of the following statements is **FALSE** regarding rumination?

- A. seen only in infants and those with intellectual disability
- B. runs episodic course

- C. results from neglect
- D. in infancy can resolve spontaneously
- E. behavioral treatment is the treatment of choice

6. In pica disorder, the **MOST** appropriate statement is

- A. it is only related to eating clay and earth
- B. usually in children below 2 years
- C. not present with other eating disorders
- D. it need a period of more than a month to be stated
- E. it resulted in high fatalities

7. A 7-year-old boy presented with recurrent eye blinking behavior and recurrent extension of extremities, mother describe the movement as sudden, rapid, and repetitive movements, it was present in the last 9 months.

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

- A. Tourette's disorder
- B. persistent motor tic disorder
- C. provisional tic disorder
- D. post-viral encephalitis
- E. Sydenham chorea

8. All the following are recognized stereotypic movements **EXCEPT**

- A. hand shaking
- B. eye blinking
- C. body rocking
- D. head banging
- E. self-biting

9. A mother to a well 2-year-old girl with thumb sucking behavior, she is worried that the behavior may continue or may cause dental problem.

Of the following, the **BEST** response is to

- A. reassurance to mother
- B. leave the behavior as the complications usually started after 5 years
- C. ignore thumb sucking and encouraging a substituted behavior
- D. use of bitter ointments will resolve the problem early
- E. asses the social status of the family

10. A 7-year-old child brought by his father to your clinic, the child often express fear of being injured by a car accident during transport to school. He expresses this fear to his teachers and parents.

Of the following, the **TRUE** description of his reaction is

- A. non pathological anxiety

- B. school phobia
- C. separation anxiety
- D. generalized anxiety disorder
- E. panic disorder

11. All the following medical conditions can cause anxiety in a child **EXCEPT**

- A. antihistamine medications
- B. hypoparathyroidism
- C. prolonged school absences
- D. carbonated beverages
- E. lead poisoning

12. Regarding school refusal, which statement is **FALSE**?

- A. it is a complex disorder
- B. selective mutism is overlapping
- C. younger children usually have separation anxiety disorder
- D. older children usually suffer from obsessive disorders
- E. somatic symptoms are common

13. A school nurse called you to see a 6-year-old boy with school refusal, he is always crying after parental leaving, refuses to stay in the class room, the school nurse informed you that she tried her best during the last 4 weeks; and she met with the parents frequently.

Of the following, the **BEST** action is to

- A. refer the child to a pediatric psychologist
- B. start selective serotonin reuptake inhibitor treatment
- C. assess the home environment
- D. arrange for parent management training
- E. give the child special attention from the teacher

14. All the following are characteristic features of separation anxiety disorder **EXCEPT**

- A. not manifested below 3 years of age
- B. common up to 5% of children
- C. girls are more affected than boys
- D. child thinking often include fear of harm affecting parents
- E. not reported in children above 8 years of age

15. You are asked to explain breath holding spells concept to a staff nurse during morning round.

Of the following, the **BEST** statement; it is a/an

- A. manifestation of iron deficiency anemia
- B. type of seizure

- C. age typical expressions of frustration or anger
- D. psychic problem manifest when the child becomes older
- E. sort of tic disorder which resolve by age

16. A concerned parents of an 18-month-child which expressed a brief tonic movement after being upset and cried.

All the following are true advices **EXCEPT**

- A. investigate possibility of iron deficiency anemia
- B. avoid over concern behavior
- C. interfere early in the event by calming the child
- D. try to behave calmly, observe the child without being seen
- E. arrange for home rectal diazepam

17. You are evaluating a 5-year-old child with breath holding spells, the history given includes pallor with abnormal limb movement lasted for 5 minutes followed by sleep.

All the following are true responses/advices **EXCEPT**

- A. reassurance, behavioral instruction to parents and follow up
- B. order MRI brain
- C. order an ECG
- D. order an EEG
- E. neurological consultation

18. A concerned young parents asking about frequent lying behavior experienced by their 3-year-old girl.

The following advices are true **EXCEPT**

- A. it is a method of playing with the language
- B. it is a part of their magical thinking
- C. it indicates a potential for future lying behavior
- D. it is an approach to avoid unwanted confrontation with adults
- E. it is a way to describe things as they wish

19. All the following are true about truancy **EXCEPT**

- A. it is normal behavior in young children
- B. it represent disorganization within the home
- C. it may reflect underlying child abuse
- D. depression may be an association
- E. adolescent may be at risk of substance abuse

20. All the following are diagnostic criteria for major depressive episode **EXCEPT**

- A. hypomanic episode
- B. depressed mood
- C. loss of interest or pleasure

- D. significant weight loss
- E. hypersomnia

21. Approximately 90% of youths who complete suicide have a preexisting psychiatric illness.

Of the following, the **MOST** commonly encountered illness is

- A. major depression
- B. schizophrenia spectrum disorders
- C. conduct disorder
- D. chronic anxiety
- E. panic disorder

22. A 13-year-old adolescent female attained out-patient department (OPD), with a complaint of feeling fat especially over the stomach and thighs; she is also feeling cold, tired, weak, and lacking energy. Examination reveals, heart rate 46 beats/min, blood pressure 70/40 mm Hg, weight below 3rd centile, dry skin, and lanugo-type hair growth on face; lab investigations shows hypokalemia and hypophosphatemia.

Of the following, the **MOST** appropriate next action is

- A. admission to the hospital
- B. referral to psychiatrist
- C. referral to dietician
- D. make an scheduled OP visit to you and to psychiatrist
- E. offer partial hospital program

23. All the following are characteristic features of autistic spectrum disorder (ASD)

EXCEPT

- A. defective social communication
- B. highly restricted fixated interests
- C. scarceof gesture use
- D. stereotyped motor movements
- E. absence of routines

24. Diagnosis of autistic spectrum disorder (ASD) depends partly but importantly on assessment of language. All the following may raise your concern regarding language development and may indicate ASD **EXCEPT**

- A. absent babbling by 6 months
- B. absent gestures by 12 month
- C. absent single words by 16 month
- D. absent 2-word purposeful phrases by 24 month
- E. loss of language or social skills at any time

25. All the following are recognized in autistic spectrum disorder (ASD) **EXCEPT**

- A. 3 grades of severity
- B. superior intellectual functioning in select areas
- C. motor deficits
- D. epilepsy
- E. truancy

26. The **MOST** consistent statement of structural MRI brain finding of autistic spectrum disorders (ASD) is

- A. diffuse brain atrophy
- B. increase brain size
- C. focal fibrosis
- D. white matter degenerative changes
- E. gray matter degenerative changes

27. All the following should raise the suspicion of autistic spectrum disorders (ASD) **EXCEPT**

- A. sibling with ASD
- B. playmate concern
- C. parental concern
- D. caregiver concern
- E. pediatrician concern

28. Ideally all children should be subjected to routine screening for autistic spectrum disorders (in USA) at age of

- A. 06 and 12 mo
- B. 12 and 18 mo
- C. 18 and 24 mo
- D. 24 and 30 mo
- E. 30 and 18 mo

29. An 18-month-old male toddler was found to be symptomatic for autistic spectrum disorders (ASD) by routine screening testing. The recommended evaluation include all the following **EXCEPT**

- A. physical examination for dysmorphic features
- B. hearing tests
- C. brain CT
- D. wood's lamp
- E. chromosomal microarray

30. A 24-month-old child who is referred by health institute after parental concern of lonely play and delayed speech, he is pica eater. You have diagnosed him as autistic spectrum disorder (ASD).

Of the following, the **MOST** important next action is

- A. physical examination for dysmorphic features
- B. hearing tests
- C. lead level
- D. Wood's lamp
- E. chromosomal microarray

31. You are following a 6-year-old boy with autism, he is under structured psychosocial behavioral training program, there is frequent complains of aggression and self-injurious behavior.

Of the following, the **BEST** medication to control his behavior is

- A. methylphenidate
- B. risperidone
- C. escitalopram
- D. atomoxetine
- E. amantadine

32. Childhood psychosis may include all the following **EXCEPT**

- A. delusions
- B. loss of reality testing
- C. disorganized speech
- D. catatonic behavior
- E. acute phobic hallucination

33. A parents came to your clinic complaining that their 5-year-old boy had attacks of frightening with imagination of a snake crawling over him and he is acting as trying to remove it. You assessed the child and find no acute physical illness and he is cooperative and quite intelligent.

Of the following, the **MOST** appropriate explanation for the child behavior is

- A. acute phobic hallucination
- B. early sign of schizophrenia
- C. delusional infestation
- D. night terror
- E. bipolar disorder

1. **(B).**
2. **(C).** Feeling very sad or withdrawn for more than 2 mo.
3. **(D).** Atypical antipsychotic.
4. **(D).** These are the common medications used in Pediatric psychiatric disorders which pediatrician should be familiar with. Floxetine is common antidepressant and it is another selective serotonin reuptake inhibitors
5. **(A).** Rumination disorder is the repeated regurgitation of food, where the regurgitated food may be rechewed, reswallowed, or spit out, for a period of at least 1 mo following a period of normal function. Rumination disorder has also been in healthy individuals across the life span in addition to infants and those with intellectual disability.
6. **(D).** Pica involves the persistent eating of non-nutritive, non-food substances (e.g., paper, soap, plaster, charcoal, clay, wool, ashes, paint, earth) over a period of at least 1 mo. The eating behavior is inappropriate to the developmental level (e.g., the normal mouthing and tasting of objects in infants and toddlers) and, therefore, a minimum age of 2 yr is suggested. Children with pica are at increased risk for lead poisoning; iron-deficiency anemia; mechanical bowel problems, intestinal obstruction, intestinal perforations, dental injury, and parasitic infections. It can be fatal but usually based on substances ingested.
7. **(C).** A tic is a sudden, rapid, recurrent (in the same manner), non-rhythmic motor, movement or vocalization e.g. eye blinking, neck jerking, shoulder shrugging, extension of the extremities. Onset of tics is typically between ages 4 and 6 yr, in Tourette's disorder both multiple motor and one or more vocal tics have been present at some time during the illness, may be concurrently for > 1 year. In chronic or persistent tic disorder either one or more motor or vocal tics are present during the course of the disease while in provisional tic disorders the duration is < 1 yr. Chorea is involuntary, random, quick, jerking movements, most often of the proximal extremities, that flow from joint to joint (not in same manner), movements are abrupt, non-repetitive, and arrhythmic and have variable frequency and intensity.
8. **(B).** Eye blinking considered as simple motor tic, others are stereotypic movements. Unlike tic disorders, the stereotypic movements typically begin within the first 3 yr of life. These movements resolve over time. Among individuals with intellectual disability, the stereotyped behaviors may persist for years. Stereotypic movements are a common manifestation of a variety of neurogenetic disorders, such as Lesch-Nyhan syndrome, Rett syndrome, fragile X syndrome, Cornelia de Lange syndrome, and Smith-Magenis syndrome.

9.(C). Thumb sucking is common in infancy and in as many as 25% of children age 2 yr and 15% of children age 5 yr. Thumb sucking beyond 5 yr may be associated with sequelae (paronychia, anterior open bite). Like other rhythmic patterns of behavior, thumb sucking is self soothing. Basic behavioral management, including encouraging parents to ignore thumb sucking and instead focus on praising the child for substitute behaviors, is often effective treatment. Simple reminders and reinforcers, such as giving the child a sticker (or other rewards) for each block of time that he or she does not suck the thumb can also be considered. Use of noxious agents (bitter salves) this approach should rarely be necessary. Going deep in social history seems to be non-appropriate in a well child. Response A is true but incomplete.

10.(A). Generally anxiety classified to a pathological and non-pathological anxiety. In pathological form it is usually resulted in loss of function (i.e. become disabling, interfering with social interactions, development, and achievement of goals or quality of life, and can lead to low self-esteem, social withdrawal, and academic underachievement) also can be associated with somatic symptoms such as weight loss, pallor, tachycardia, tremors, muscle cramps, paresthesias, hyperhidrosis, flushing, hyperreflexia, and abdominal tenderness. In this scenario the child never refuses to go to school or to ride the car, he had no somatic symptoms, good explanation and reassurance can end the problem.

11.(B). Systemic disorders, hormonal disturbances, and drugs along with carbonated beverage can cause anxiety in children with it is symptoms and signs such as tachycardia e.g. hyperthyroidism, hyperparathyroidism, hypoglycemia, central nervous system disorders (brain tumors), migraine, asthma, lead poisoning, cardiac arrhythmias, and, rarely, pulmonary embolism, systemic lupus erythematosus, anaphylaxis, porphyria or pheochromocytoma. Side effect of anti-asthmatic agents, steroids, sympathomimetics, SSRIs (initiation), anticholinergic agents, and antipsychotics. Other like diet pills, antihistamines, drug withdrawal, and cold medicines can cause anxiety.

12.(D). School refusal or phobia it is a common problem, it occurs in approximately 1-2% of children, associated with anxiety in 40-50% of cases, depression in 50-60% of cases, and oppositional behavior in 50% of cases. Younger anxious children who refuse to attend school are more likely to have separation anxiety disorder whereas older anxious children usually refuse to attend school because of social phobia.

13.(A). Management of school refusal typically requires parent management training and family therapy, with school personnel is always indicated; anxious children often require special attention from teachers, counselors, or school nurses. Parents who are coached to calmly sending child to school and to reward the child for each completed day of school are usually successful; in this scenario this option had been used. In cases of ongoing school refusal, referral to a child psychiatrist or psychologist is indicated. SSRI treatment may be helpful. Young children with affective symptoms have a good prognosis.

14.(E). Separation anxiety disorder (SAD) is one of the most common childhood anxiety disorders with a prevalence of 3.5-5.4%. Separation anxiety behavior is developmentally normal when it begins about 10 mo of age and tapers off by 18 mo. By 3 yr of age, most children can accept the temporary absence of their mother or primary caregiver. SAD is more common in prepubertal children, with an average age of onset of 7.5 yr. Girls are more commonly affected than boys. SAD is characterized by unrealistic and persistent worries about separation from the home or parents. Numerous somatic symptoms and complaints of subjective distress. Symptoms vary depending on the child's age; children younger than 8 yr often have associated school refusal and excessive fear that harm will come to a parent; children 9-12 yr have excessive distress when separated from a parent; and those 13-16 yr often have school refusal and physical complaints.

15.(C). Breath holding spells are common during the first years of life and are age-typical expressions of frustration or anger. Subtypes of breath holding spells include cyanotic, pallid, or mixed episodes. Cyanotic are the dominant type and may include a brief loss of consciousness and a very brief tonic-clonic seizure. Pallid spells may be similar to vasovagal related syncopal events in older children and initiated from similar stimuli. Iron deficiency with or without anemia may be present and some children with breath-holding spell respond to iron therapy.

16.(E). All of the above in a sequence of BDCA represent an approach to deal with classical breath holding spells.

17.(A). In non-classical type of breath holding spells; medical conditions to consider should include seizures, Chiari crisis, dysautonomia, cardiac arrhythmias (as prolonged Q-T interval), and central nervous system lesions. So all the above optional investigations are justifiable.

18.(C). Lying is a normal behavior between 2-4 years and represents all the above except the option C.

19.(A). Truancy (running from school) is a never developmentally appropriate. Truancy may represent disorganization within the home, care taking needs of younger siblings, developing conduct problems, or emotional problems including depression or anxiety. Whereas younger children may threaten to run away out of frustration or a desire to get back at parents, older children who run away are almost always expressing serious underlying problem within themselves or their family, including violence, abuse, and neglect. Adolescent runaways are at high risk for substance abuse, unsafe sexual activity, and other risk-taking behaviors.

20.(A). Hypomanic or manic episodes are not features of major depressive episode. A period of at least 2 wks is required while in persistent depressive disorder require a period of 2 years for diagnosis.

21.(A). All mentioned distracters have a role in suicide but major depression is the most common one.

22.(A). Heart rate <50 beats/min and blood pressure <80/50 mmHg with hypokalemia and hypophosphatemia are indications for admission in anorexia nervosa; other

indications may include hypoglycemia, dehydration, temperature $<36.1^{\circ}\text{C}$, $<80\%$ healthy body weight, hepatic, cardiac, or renal compromise, suicidal intent and plan.

23.(E). Autism is typically recognized during the 2nd yr of life but can be seen earlier than 12 mo if developmental delays are severe. Initial symptoms most frequently involve delayed language accompanied by lack of social interest or odd play patterns. During the 2nd yr, odd and repetitive behaviors and the absence of typical play become more apparent. It is typical for parents to report that there was no period of normal development or that there was a history of unusual behaviors. Less commonly (in 20-40% of cases), a period of apparently normal development is reported before a loss of skills appears.

24.(A). Absence of babbling by 12 months is a red flag for language development and should raise a suspicion of ASD (autistic spectrum disorder).

25.(E). All these mentioned are recognized associations with ASD, it has 3 grades of severity, early diagnosis usually improves outcome, but those in the severe form continue to adulthood with problems of independent living, employment, social relationships, and mental health. Small group (mild form with communication abilities) can grow up to live self-sufficient lives in the community with employment. Truancy is a separate behavioral disorder.

26.(B). Increase in brain size and increased head circumference had been documented in autistic children. Other important finding in structural MRI is diffusion tensor imaging studies suggest aberrations in white matter tract development.

27.(B). Playmate concern is not considered.

28.(C).

29.(C). The recommended evaluation should rule out dysmorphic syndromes; macrocephaly; hearing problems; tuberous sclerosis and chromosomal abnormalities and genetic causes like fragile X syndrome and Rett syndrome.

30.(C). Lead level should be ordered in situation of that, the child is a pica eater or he is living in an area with high lead level in soil, air or water. Depending on clinical presentation, metabolic screen, thyroid stimulating hormone, EEG and specific genetic studies should be done.

31.(B). The drug of choice is risperidone, while methylphenidate is the drug of choice for ADHD in normal IQ children, atomoxetine is also a new medication for ADHD, and escitalopram is used for depression.

32.(E).

33.(A). In children with non-psychotic hallucinations, the symptoms of psychosis are absent; normal mental status examination is the key for its diagnosis. Non-psychotic hallucinations commonly occur in the context of severe traumatic stress, developmental difficulties, social and emotional deprivation. Hallucinations related to fantasy, culture beliefs, grief, hypnagogic hallucinations, night terrors, fever, and acute phobic hallucinations all are considered normal. In this scenario, the case typically has an acute phobic hallucination which is benign, common and occurs in previously healthy preschool children. The hallucinations are often visual or tactile, last

10-60 minutes, and occur at any time but most often at night. The child is quite frightened and might complain that bugs or snakes are crawling over him or her with attempt to remove them. The cause is unknown. The differential diagnosis includes drug overdose or poisoning, high fever, encephalitis, and psychosis. The child's fear is not alleviated by reassurance by the parents or physician, and the child is not amenable to reason. Findings on physical and mental status examinations are otherwise normal. Symptoms can persist for 1-3 days, slowly abating over 1-2 wk.

PART IV

Learning Disorders QUESTIONS

AHMED TAWFIQ

1. The definition of intellectual function is the capacity to think in the abstract, reason, problem solve, and comprehend. The score at which significant weaknesses in adaptive skills are evident is equal to or below
 - A. 55
 - B. 60
 - C. 65
 - D. 70
 - E. 75
2. The neurodevelopmental function (cognition) is dependent mainly on the development of the following functions **EXCEPT**
 - A. sensory and motor
 - B. language
 - C. visual-spatial
 - D. intellectual
 - E. cerebeller
3. Which statement is **FALSE** regarding specific learning disorders (SLD)?
 - A. it is a type of neurodevelopmental dysfunctions
 - B. the overall estimates of the prevalence of SLD range from 3-10%
 - C. it focuses on academic skill development
 - D. includes students did well in academic testing but not in intelligence testing
 - E. terms as dysgraphia or dyscalculia had been revolutionized
4. The major chemical modulator of attention is
 - A. dopamine
 - B. serotonin
 - C. nor-epinephrine
 - D. troponin
 - E. epinephrine
5. In between specific learning disorders (SLD), the **MOST** refractory to treat is
 - A. reading
 - B. mathematics
 - C. working memory
 - D. social cognition

- E. writing
6. All the following are risk factors for specific learning disorders (SLD) **EXCEPT**
- A. extreme prematurity
 - B. low birth weight
 - C. history of sibs or parents with SLD
 - D. history of admission to neonatal care unit
 - E. chronic medical problem
7. All the following are red flags in the history of children with specific learning disorders (SLD) **EXCEPT**
- A. parents concern about academic performance
 - B. inconsistency in marks report from grade to grade
 - C. struggling with home work activities
 - D. positive vision or hearing test
 - E. positive standard screening test
8. All the following are established modalities of treatment in a child with specific learning disorders (SLD) **EXCEPT**
- A. remediation of skills
 - B. demystification
 - C. bypass strategies
 - D. dietary interventions
 - E. strengthening of strength
9. The **MOST** common specific learning disorder (SLD) of childhood is
- A. attention-deficit/hyperactivity disorder (ADHD)
 - B. reading disorder (dyslexia)
 - C. spelling disorder
 - D. arithmetical skills disorders
 - E. mixed disorder of Scholastic Skills
10. The behavioral changes in attention-deficit/hyperactivity disorder (ADHD) should met all the following criteria **EXCEPT**
- A. developmentally inappropriately far comparing with other children of the same age
 - B. must begin before age 6 yr
 - C. must be present for at least for 6 mo
 - D. must be present in 2 or more settings
 - E. must not be secondary to another disorder

11. You are assessing an eight-year-old male child with attention-deficit/hyperactivity disorder (ADHD).

Of the following, the **LEAST** useful test/investigation is

- A. thyroid function test
- B. lead level
- C. EEG
- D. Blood film
- E. polysomnography

12. Of the following, the **MOST** common presenting behavior in girls with attention-deficit/hyperactivity disorder (ADHD) is

- A. inattentive
- B. hyperactive
- C. impulsive
- D. disruptive
- E. combined

13. Of the following, the **TRUE** statement of reading disorder is

- A. 25% of affected children have affected parents
- B. diagnosis is usually clinical
- C. IQ will remain same with time
- D. functional MRI is diagnostic
- E. boys are more affected than girls

- 1.(D). Neurodevelopmental dysfunctions reflect disruptions of neuroanatomic structure or psychophysiologic function and place a child at-risk for developmental, cognitive, emotional, behavioral, psychosocial, and adaptive challenges.
- 2.(E). Neurodevelopmental function is a basic brain process needed for learning and productivity. It needs all that process that mentioned above plus memory, social cognition and frontal lobe functions. The process of attention and executive functioning is heavily dependent on frontal lobe function.
- 3.(D). Specific developmental disorders of scholastic Skills (specific learning disorders) includes reading disorder, spelling disorder, disorder of arithmetical skills, and mixed disorder of Scholastic Skills. Dyslexia (reading disorder) is included in ICD-10 in a separate category of symbolic dysfunction. The terms, Dyscalculia (mathematics disorder), and Dysgraphia (written language disorder) are also used by investigators and clinicians, but their inclusion in diagnostic classification systems has been inconsistent and a source of some disagreement among experts. A student exhibiting a significant discrepancy between scores on tests of intelligence and tests of academic achievement could be classified as a student with an SLD, and would subsequently be eligible for Special Education Services i.e. those student may did very well in IQ testing but not in academic achievement or tests.
- 4.(A). Most brain processes are heavily dependent on functional arousal, alertness, and attention. Any malfunction within or across these systems will likely cause some degree of breakdown in other cognitive processes. Functional attention subsumes intact neuroanatomic and neurochemical brain systems. Structurally, brain regions involved include subcortical, cortical, and association areas throughout the brain. Primary structures involved include brainstem regions (e.g., basal ganglia), the limbic system (e.g., amygdala and hippocampus), and the frontal lobes (e.g., prefrontal cortex). The neurotransmitter dopamine, along with its neuronal pathways, has been identified as a major chemical modulator of attention. It is through the cognitive mechanisms of attention and executive functions that the child's brain acquires, organizes, and processes information. These mechanisms also allow the child to regulate, plan, and monitor their behaviors and thoughts.
- 5.(B). Delay in mathematical ability, known as mathematics disorder or dyscalculia, can be especially refractory to correction, partly because math involves the assimilation of both procedural knowledge (e.g., calculations) and higher-order cognitive processes (e.g., working memory).
- 6.(D). History of admission to neonatal care unit is not a specific marker for serious problem. It could be for minor cause or for observation only.

7.(B). Inconsistency in report from grade to grade may sometimes be caused by a difference in teaching styles or classroom demands.

8.(D).

9.(B). Reading disorder affect about 80% of children identified as learning disabled.

10.(B). It is important to systematically gather and evaluate information from a variety of sources, including the child, parents, teachers, physicians, and, when appropriate, other caretakers, over the course of both diagnosis and subsequent management. These behaviors should be evident before age of 12 yr.

11.(D). At first, hypertension, ataxia, and hyperthyroidism should be excluded. Other differential diagnosis should be considered as heavy metal poisoning (including lead), sensory deficits (hearing and vision), auditory and visual processing disorders, and neurodegenerative disorder, especially leukodystrophies. Fragile X syndrome, absence and sleeping disorders are also important differential diagnoses.

12.(A). While the hyperactive-impulsive and combined types are common in boys.

13.(B). The cardinal signs of dyslexia observed in school-age children and adults are a labored, effortful approach to reading involving decoding, word recognition, and text reading. 50% of affected children have affected parents. Boys and girls are equal.

PART V

Children with Special Needs QUESTIONS

AHMED TAWFIQ

1. In USA, the newly arrived international adoptees are subjected to many lab tests.

All the following tests need to be repeated after 3-6 months after arrival **EXCEPT**

- A. hepatitis B virus serology
- B. hepatitis C virus serology
- C. human immunodeficiency serology
- D. tuberculin skin test
- E. treponemal test

2. You are a newly employee Pediatrician to foster care children (children removed from their original families and put in suitable institutes for protection).

Those children have many significant problems, but the **MOST** common one is

- A. chronic medical problems
- B. abuse and neglect
- C. mental health concerns
- D. family relationship problems
- E. developmental problems

3. Witnessing violence, community violence, and media violence all are detrimental to children as it increases the impact of violence.

All the following adverse effects can be increased by increasing the impact of violence

EXCEPT

- A. poor school performance
- B. symptoms of anxiety and depression
- C. lower self-esteem
- D. cyber bullying
- E. post traumatic stress disorder

4. A 9-year-old male with good school performance and normal behavior. In the last 2 months he had changed his classroom. His teacher observed that he became introverted and his school performance reduced dramatically. He was always described by some of his new school mates as a "weak".

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

- A. school phobia
- B. anxiety disorder
- C. bullying
- D. hypothyroidism

- E. depression
5. The following factors show psychosocial impact of war on children **EXCEPT**
- A. loss of family members
 - B. separation from community
 - C. lack of education
 - D. acute stress reaction
 - E. displacement
6. The **MOST** reported type of child abuse in USA is
- A. neglect
 - B. physical abuse
 - C. sexual abuse
 - D. psychological maltreatment
 - E. trafficking
7. Regarding the physical abuse, the physical signs in abused children (as bite marks) may be found in up to
- A. 5%
 - B. 15%
 - C. 25%
 - D. 35%
 - E. 45%
8. The **MOST** common clue of physical abuse in children is
- A. history of inflicted trauma
 - B. burn marks
 - C. bruises
 - D. fractures
 - E. retinal hemorrhages
9. In children with physical abuse, when the inflicted trauma is burn injury. Of the following, the **MOST** suggestive one is
- A. burn in napkins with involvement of flexures
 - B. presence of splash mark
 - C. unclear borders of burn
 - D. glove and stock distribution
 - E. folk remedies
10. Skeletal injuries with high specificity in child abuse may include all the following **EXCEPT**
- A. tibial metaphyseal corner lesions

- B. posterior rib fracture
- C. scapular fracture
- D. first rib fracture
- E. clavicle fracture

11. A 3-month-old baby girl admitted to pediatric intensive care unit with severe head injury (evident by CT scan), the history given by parents was trivial and not informative. You suspected child abuse.

Of the following, the **MOST** helpful study to support your suspicion is

- A. infantogram (one shot for entire body)
- B. lateral and AP view X ray of the spine
- C. lateral and AP view X ray of skull
- D. radionuclide bone scan
- E. MRI brain

12. A 3-month-precious baby of a keen and highly educated family presented with history of URTI followed by deterioration of level of consciousness and seizure. CT brain shows intracranial bleeding with no evidence of skull fracture. Fundoscopy was normal. Mother gives history of difficult labour but with normal development, mild hypotonia, and macrocephaly which was reassured initially by general Pediatrician.

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

- A. residual birth trauma
- B. arteriovenous malformation
- C. acute viral encephalitis
- D. glutaric aciduria type 1
- E. abusive head trauma (AHT)

13. Retinal hemorrhages are an important marker of abusive head trauma (AHT). Whenever AHT is being considered, a dilated indirect eye examination by a pediatric ophthalmologist should be performed.

All the following are characteristic of retinal hemorrhage caused by AHT **EXCEPT**

- A. multiple
- B. involve more than one layer of the retina
- C. central
- D. traumatic retinoschisis
- E. various sizes

14. A 3-year-old female child with repeated admissions as bleeding tendency in a form of spontaneous ecchymosis and bleeding through the nose and urine. She had another daughter who died because of similar undiagnosed illness. The mother is a staff nurse and she is highly concerned about her child illness. Lab investigations usually show either PT and/or PTT prolongation with normal platelets, which usually normalized

after empiric vitamin K administration. The father is a petrol engineer and never seen accompanying the family.

Of the following, the **MOST** helpful investigation is

- A. factor VII assay
- B. factor II assay
- C. drug level
- D. von Willebrand factor assay
- E. platelets function test

15. In a child less than 3 years, the diagnosis of failure to thrive (FTT) is considered if

- A. weight is below the 3rd percentile
- B. weight drops down more than 2 major percentile lines
- C. weight for height is less than the 1st percentile
- D. BMI less than the 25th percentile
- E. midarm circumference < 15 cm

16. In patients with failure to thrive; chromosomal abnormality, intrauterine infection, and metabolic disorders need to be ruled out.

All the following factors are suggestive of metabolic problems **EXCEPT**

- A. insidious history
- B. recurrent vomiting
- C. neurologic symptoms
- D. cardiomyopathy
- E. renal symptoms

17. Approaching to a child with failure to thrive based on signs and symptoms.

Of the following, the **MOST** common cause behind a child has spitting, vomiting, and food refusal is

- A. gastroesophageal reflux
- B. chronic tonsillitis
- C. food allergies
- D. eosinophilic esophagitis
- E. inflammatory bowel disease

18. A 15-month-old male child with failure to thrive, diarrhea, and fatty stool.

Of the following, the **MOST** common possible cause of his illness is

- A. malabsorption
- B. intestinal parasites
- C. milk protein intolerance
- D. pancreatic insufficiency
- E. immunodeficiency

19. A 2-year-old child with failure to thrive, recurrent wheezing, and pulmonary infections.

Of the following, the **LEAST** common cause of his illness is

- A. asthma
- B. aspiration
- C. food allergy
- D. cystic fibrosis
- E. immunodeficiency

20. In a child with failure to thrive, the indications for hospitalization include severe malnutrition or failure of outpatient management. The period after which the child needs hospitalization, if he has not been responded to outpatient management, is about

- A. 1-2 wk
- B. 4 wk
- C. 2-3 mo
- D. 4 mo
- E. 5-6 mo

21. You are treating a 10-month-old child with failure to thrive.

Of the following, the minimal accepted catch-up weight gain is about

- A. 10 gm/kg/day
- B. 20-30 gm/kg/day
- C. 40-50 gm/kg/day
- D. 60 gm/kg/day
- E. 80 gm/kg/day

- 1.(E).
- 2.(D). All children in foster care have family relationship problems, while 40-60% for choice A, >70% for choice B, 80% for choice C, and around 60% for choice E.
- 3.(D). All these (except D) represent the impact of violence on children, while cyber bullying is a newly emerging form of bullying that takes place using electronic technology.
- 4.(C). Showing the power of some children on a weaker one can lead serious psychological and psychiatric effects. That child was a bullying-victim and should be protected.
- 5.(E). It represents physical impact.
- 6.(A). Physical abuse includes beating, shaking, burning, and biting. Sexual abuse has been defined as the involvement of dependent, developmentally immature children and adolescents in sexual activities. Neglect refers to omissions in care, resulting in actual or potential harm which include inadequate healthcare, education, and supervision, protection from hazards in the environment, and unmet physical needs (e.g., clothing, food) and emotional support. Psychological abuse includes verbal abuse and humiliation and acts that scare or terrorize a child. Trafficking in children (selling them abroad), for purposes of cheap labor and/or sexual exploitation, expose children to all of the forms of abuse just mentioned. Child neglect constitutes 78% of the reports.
- 7.(C). Sentinel injuries may be noted in approximately in 25% of abused infants and may precede the diagnosis by weeks or even months from the sentinel event.
- 8.(C). Features suggestive of inflicted bruises include (a) bruising in a pre ambulatory infant (occurring in just 2% of infants), (b) bruising of padded and less-exposed areas (buttocks, cheeks, under the chin, genitalia), (c) patterned bruising or burns conforming to shape of an object or ligatures around the wrists, and (d) multiple bruises, especially if clearly of different ages. History is the least dependable because it is either hidden or skewed.
- 9.(D). Immersion burn injury patterns; napkin area that spares flexor creases, immersion stocking burn, immersion glove burn, immersion buttocks burn. It usually shows a clear delineation between the burned and healthy skin, and uniform depth. The splash marks are usually absent.
- 10.(E). It is of low-specificity finding.
- 11.(D). Of the given options, option D is the best because it detect acute skeletal injuries (as fractures of posterior rib fracture that indicate shaking injury) before it became evident on X ray. If there is suspension in the first scan, another scan can be

performed after 2 days from the first one. Infantogram is not preferable as a one or 2 shot for entire body. Options B and C are only a part from skeletal survey, which is indicated at time of arrival and after 2 weeks. MRI brain is best taken 5-7 days after acute injury, MRIs are helpful in differentiating extra axial fluid, determining timing of injuries, assessing parenchymal injury, and identifying vascular anomalies.

12.(D). Patients with birth trauma may have asymptomatic subdural hematoma which may resolve by the end of first month of life. Arteriovenous malformation usually has subarachnoid hemorrhage. Encephalitis is possibility but unusually present with intracranial bleed. AHT is a possibility but presence of hypotonia and macrocephaly and absence of retinal hemorrhage make the diagnosis of Glutaric aciduria type 1 is the most likely one.

13.(C). It is often extending to the periphery (outside the posterior pole). D is highly characteristic.

14.(C). The presentation of medical child abuse (MCA) formerly called Munchausen syndrome by proxy; may vary in nature and severity. Consideration of MCA should be triggered when the reported symptoms are repeatedly noted by only one parent, appropriate testing fails to confirm a diagnosis, and seemingly appropriate treatment is ineffective. Drug level for warfarin may solve the problem in this case, the mother is staff nurse and she is familial with medical issues, the social history given is suggestive of disturb relationship between parents.

15.(B). Weight is below the 5th percentile, weight for height is less than the 5th percentile, BMI less than the 5th percentile, also defines FTT.

16.(A). The history of metabolic disorder is usually acute, severe, and potentially life-threatening, recurrent vomiting, liver dysfunction, neurologic symptoms, cardiomyopathy and myopathy, impairment of special senses, renal symptoms, or distinct dysmorphic features and/or organomegaly all are may be a part of metabolic illness.

17.(A). The first 4 causes are true causes but the commonest is (A). The choice (E) causes FTT with diarrhea and fatty stools.

18.(A).

19.(E).

20.(C). Inpatient care may include further diagnostic and laboratory evaluation, an assessment and implementation of adequate nutrition, and evaluation of the parent-child feeding interaction.

21.(B). Minimal catch-up growth should generally be 2-3 times the average weight gain for corrected age.

1. Regarding nutritional requirement during childhood, all the following are true **EXCEPT**

- A. nutrition and growth during 3-6 years of life predict adult stature and some health outcomes
- B. major risk period for growth stunting is between 4 and 24 months of age
- C. estimated average requirement (EAR) is the average daily nutrient intake level estimated to meet the requirements for 50% of the population
- D. recommended dietary allowance (RDA) is an estimate of the daily average nutrient intake to meet the nutritional needs of >97% of the individuals in a population
- E. the 3 components of energy expenditure are the basal metabolic rate, thermal effect of food, and energy for physical activity

2. The adequate intake (AI) for the total digestible carbohydrates in a nine-month-old boy is

- A. 60 gm/day
- B. 95 gm/day
- C. 120 gm/day
- D. 145 gm/day
- E. 170 gm/day

3. The adequate intake (AI) for the total fat in a nine-month-old boy is

- A. 10 gm/day
- B. 20 gm/day
- C. 30 gm/day
- D. 40 gm/day
- E. 50 gm/day

4. The adequate intake (AI) for the total protein in a nine-month-old boy is

- A. 9 gm/day
- B. 11 gm/day
- C. 13 gm/day
- D. 15 gm/day
- E. 17 gm/day

5. All the following amino acids are indispensable in human diet **EXCEPT**

- A. leucine
 - B. methionine
 - C. threonine
 - D. valine
 - E. alanine
6. Essential fatty acids deficiency is associated with all the following features **EXCEPT**
- A. desquamating skin rashes
 - B. alopecia
 - C. thrombocytosis
 - D. impaired immunity
 - E. growth deficits
7. All the following statements concerning iron requirement in children are true **EXCEPT**
- A. breast milk provides optimal intake of iron for the first 4 months
 - B. iron present in animal protein is more bioavailable than that found in vegetables
 - C. iron deficiency is the most common micronutrient deficiency
 - D. cow's milk is a good source of bioavailable iron
 - E. iron supplements can interfere with zinc absorption
8. Zinc deficiency is associated with increased risk for all the following **EXCEPT**
- A. stunting
 - B. impaired immune function
 - C. increased risk for respiratory diseases
 - D. increased risk for diarrheal diseases
 - E. increased risk for skin infections
9. The American academy of pediatrics change the vitamin D intake recommendation in 2010 to
- A. 200 IU/day
 - B. 400 IU/day
 - C. 600 IU/day
 - D. 800 IU/day
 - E. 1000 IU/day
10. Breast feeding has been suggested to have a possible protective effect against all the following **EXCEPT**
- A. otitis media
 - B. urinary tract infections
 - C. septicemia
 - D. childhood cancer

E. skin infections

11. An absolute contraindication to breastfeeding is

- A. hepatitis c infection of the mother
- B. alcohol intake
- C. herpes simplex lesion of the lips
- D. active pulmonary tuberculosis
- E. maternal high grade fever

12. Regarding breast engorgement; All the following are true **EXCEPT**

- A. usually happens in the first stage of lactogenesis
- B. poor breast feeding technique can cause engorgement
- C. breastfeeding immediately at signs of infant hunger will eventually prevent this
- D. to reduce engorgement, breasts should be softened prior to infant feeding with a combination of hot compresses and expression of milk
- E. between feedings, cold compresses applied, and oral nonsteroidal anti-inflammatory medications administered

13. All the following organisms can cause mastitis **EXCEPT**

- A. Staphylococcus aureus
- B. Escherichia coli
- C. Haemophilus influenza
- D. Klebsiella pneumoniae
- E. group B streptococcus

14. The **MOST** common cause of using infant formula is

- A. parental preference
- B. inborn error of metabolism
- C. maternal tuberculosis
- D. mastitis
- E. inadequate weight gain

15. Concerns to be considered among vegetarians are all the following **EXCEPT**

- A. higher bioavailability of iron
- B. lower B₁₂ levels
- C. risk of having lower level of fatty acids
- D. lower levels of calcium and vitamin D₃
- E. lower bioavailability of zinc

16. All the following are dimensions of food security **EXCEPT**

- A. access

- B. utilization
 - C. preparation
 - D. stability
 - E. availability
17. The **TRUE** hemoglobin cutoff to define anemia is
- A. 120 g/L for children 6-59 mo
 - B. 115 g/L for children 5-11 yr
 - C. 110 g/L for children 12-14 yr
 - D. 110 g/L for non-pregnant women
 - E. 120 g/L for men
18. The **MOST** commonly used index for nutritional status is
- A. height-for-age
 - B. weight-for-height
 - C. body mass index
 - D. mid-upper arm circumference
 - E. weight-for-age
19. For children <5 yr, the highest global prevalence of micronutrient and trace elements deficiencies is that of
- A. vitamin A deficiency
 - B. zinc deficiency
 - C. iron deficiency
 - D. iodine deficiency
 - E. copper deficiency
20. The **MOST** profound consequence of undernutrition is
- A. premature death
 - B. repeated infections
 - C. stunting
 - D. cell damage
 - E. developmental delay
21. "Reductive adaptation" process of evolution to a state of full malnutrition, all the following events will be ensued **EXCEPT**
- A. the liver makes glucose less readily
 - B. less heat production
 - C. gut produces less gastric acid and gut enzymes
 - D. evident responses to infection
 - E. reduced red cell mass

22. The outward sign of cell damage in malnutrition is
- A. repeated infections
 - B. wasting
 - C. edema
 - D. hepatomegaly
 - E. facial appearance
23. The recommended transition time from stabilization to rehabilitation phase in the treatment of malnutrition is
- A. 1 day
 - B. 3 days
 - C. 5 days
 - D. 7 days
 - E. 10 days
24. The WHO recommendation for antibiotic cover in the stabilization phase of treating malnourished boy free of initial complications is to use
- A. oral amoxicillin
 - B. parenteral penicillin
 - C. oral second generation cephalosporin
 - D. parenteral gentamicin
 - E. parenteral third generation cephalosporin
25. During treatment of malnutrition, the signal of entry to the rehabilitation phase is
- A. reduced edema
 - B. resolution of infection
 - C. disappearance of signs of micronutrient deficiency
 - D. constant blood sugar level
 - E. constant body temperature
26. In consideration of preventing infections in stabilization phase of malnutrition in unimmunized 10-month-old boy.
- Of the following, the vaccine that is recommended to be given is
- A. oral polio vaccine
 - B. acellular pertussis vaccine
 - C. measles vaccine
 - D. pneumococcal vaccine
 - E. H. influenza vaccine
27. The following vitamins and trace elements are recommended to be given during the stabilization phase of malnutrition **EXCEPT**
- A. iron

- B. vitamin A
 - C. folic acid
 - D. zinc
 - E. multivitamins
28. Emergency treatment in severe malnutrition includes all the following **EXCEPT**
- A. shock
 - B. hypoglycemia
 - C. dehydration
 - D. severe anemia
 - E. infections
29. The aim of the rehabilitation phase in the treatment of malnutrition is to
- A. repair cellular function
 - B. correct fluid and electrolyte imbalance
 - C. restore homeostasis
 - D. catch-up growth
 - E. prevent death from infection
30. The hallmark of refeeding syndrome is the development of severe
- A. hypophosphatemia
 - B. hypokalemia
 - C. hypomagnesemia
 - D. hypernatremia
 - E. hyperglycemia
31. Encephalopathy in refeeding syndrome is mainly a result of
- A. hypophosphatemia
 - B. hypokalemia
 - C. hypomagnesemia
 - D. thiamine deficiency
 - E. hyperglycemia
32. Thrombocytopenia in refeeding syndrome is mainly a result of
- A. hypophosphatemia
 - B. hypokalemia
 - C. hypomagnesemia
 - D. thiamine deficiency
 - E. hyperglycemia
33. Across all racial groups, the factor that confers protection against childhood obesity is

- A. gestational weight gain
- B. high birth weight
- C. maternal smoking
- D. breast feeding
- E. maternal education

34. Adeposity rebound, the period when the body fat is typically at the lowest level, is best represented at the age of

- A. 1 year
- B. 3 years
- C. 6 years
- D. 9 years
- E. 12 years

35. The gastrointestinal hormone/response that stimulates appetite in children is

- A. cholecystokinin
- B. glucagon-like peptide-1
- C. peptide yy
- D. vagal neuronal feedback
- E. ghrelin

36. All the following are childhood obesity-associated comorbidities later in life **EXCEPT**

- A. hypertension
- B. gallbladder disease
- C. tibia vara
- D. asthma
- E. type 1 diabetes mellitus

37. Genetic disorders associated with obesity can have all the following features **EXCEPT**

- A. dysmorphic features
- B. cognitive impairment
- C. vision abnormalities
- D. hearing abnormalities
- E. tall stature

38. The following are well recognized causes of secondary obesity **EXCEPT**

- A. myelodysplasia
- B. muscular dystrophy
- C. anti-psychotic medications
- D. achondroplasia
- E. anti-migraine medications

39. The following physical features might be discovered during physical examination of an obese 10-year-old boy, and each reflect the associated morbidity **EXCEPT**
- A. developmental delay suggests genetic disorder
 - B. day time sleepiness suggests sleep apnea
 - C. polyuria and nocturia suggest type 1 diabetes
 - D. hip pain suggests blount disease
 - E. abdominal pain suggests non alcoholic fatty liver disease
40. The investigations to be done as part of initial evaluation of a newly identified obesity in a 12-year-old girl are the following **EXCEPT**
- A. fasting plasma glucose
 - B. triglyceride level
 - C. lipoprotein level
 - D. liver function tests
 - E. renal function tests
41. The only Food and Drug Administration (FDA)-approved medication for obesity in children <16 yr old is
- A. orlistat
 - B. phentermine
 - C. topiramate
 - D. amylin
 - E. lorcaserin
42. The **MOST** characteristic lesion of vitamin A deficiency is
- A. xerophthalmia
 - B. corneal ulcers
 - C. Bitot spots
 - D. keratomalacia
 - E. keratoconus
43. All the following are helpful for diagnosis of marginal vitamin A deficiency **EXCEPT**
- A. dark adaptation test
 - B. conjunctival impression cytology
 - C. relative dose response
 - D. plasma retinol level
 - E. diet history
44. Chronic hypervitaminosis A is associated with all the following features **EXCEPT**
- A. seborrhea
 - B. alopecia
 - C. hepatosplenomegaly

- D. bulging fontanel
 - E. echymosis
45. The following are characteristic features of acute hypervitaminosis A **EXCEPT**
- A. nausea and vomiting
 - B. drowsiness
 - C. diplopia
 - D. cranial nerve palsies
 - E. seizures
46. The following conditions are risk factors for carotenemia **EXCEPT**
- A. liver disease
 - B. food faddist
 - C. diabetes mellitus
 - D. hypothyroidism
 - E. malabsorption
47. The classic clinical triad of Wernicke encephalopathy of thiamine deficiency is
- A. mental status changes, ocular signs, and ataxia
 - B. cardiac involvement, peripheral neuritis, and aphonia
 - C. depression, drowsiness, and poor mental concentration
 - D. increased intracranial pressure, meningismus, and coma
 - E. deterioration of school performance, seizures, and headache
48. Death from thiamine deficiency is usually due to
- A. renal involvement
 - B. repeated infections
 - C. lactic acidosis
 - D. cardiac involvement
 - E. increased intracranial pressure
49. Regarding thiamine deficiency
- A. features develop within 2-3 weeks of deficient intake
 - B. poor mental concentration and depression are late features of the disease
 - C. hoarseness or aphonia is a characteristic sign
 - D. the term (dry) beriberi reflects the features of raised intracranial pressure
 - E. the term (wet) beriberi reflects the features of renal dysfunction
50. Thiamine-responsive megaloblastic anemia syndrome is a rare disorder, it is characterized by the following **EXCEPT**
- A. diabetes mellitus
 - B. peripheral neuritis

- C. megaloblastic anemia
- D. sensorineural hearing loss
- E. autosomal recessive inheritance

51. Brown-Vialetto-Van Laere syndrome (BVVLS), a neurologic disorder characterized by progressive neurologic deterioration, sensorineural hearing loss, and pontobulbar palsy usually responds to treatment with high doses of

- A. niacin
- B. biotin
- C. riboflavin
- D. pyridoxine
- E. thiamin

52. The **MOST** convenient way to confirm a diagnosis of pellagra in children is

- A. skin biopsy
- B. urinary 2-pyridone
- C. response to niacin treatment
- D. urinary n1-methyl-nicotinamide
- E. physical signs of glossitis and dermatitis

53. The **MOST** characteristic manifestation of pellagra is

- A. dermatitis
- B. dementia
- C. diarrhea
- D. anorexia
- E. lassitude

54. The dietary reference intake of pyridoxine for a healthy eight-month-old infant is

- A. 0.1 mg/day
- B. 0.3 mg/day
- C. 0.5 mg/day
- D. 1.0 mg/day
- E. 3 mg/day

55. All the following manifestations of pyridoxine deficiency are seen in children

EXCEPT

- A. seizures
- B. cheilosis
- C. listlessness
- D. peripheral neuritis
- E. seborrheic dermatitis

56. Pyridoxine is not affected in patients receiving
- A. isoniazid
 - B. valproate
 - C. phenytoin
 - D. penicillamine
 - E. corticosteroids
57. Folate is important during embryogenesis for the development of
- A. lungs
 - B. genital organs
 - C. gastrointestinal system
 - D. central nervous system
 - E. cardiovascular system
58. All the following can cause folate deficiency **EXCEPT**
- A. celiac disease
 - B. sickle cell anemia
 - C. 6-mercaptopurine therapy
 - D. inflammatory bowel disease
 - E. methylene tetrahydrofolate reductase deficiency
59. Clinical features of cerebral folate deficiency include the following **EXCEPT**
- A. lethargy
 - B. microcephaly
 - C. ataxia
 - D. ballismus
 - E. blindness
60. One of the recognized non-hematological manifestations of folic acid deficiency in children is
- A. repeated respiratory abscesses
 - B. recurrent abdominal pain
 - C. recurrent seizures
 - D. growth retardation
 - E. teeth decay
61. All the following are features of hereditary folate malabsorption **EXCEPT**
- A. anal ulcers
 - B. chronic diarrhea
 - C. failure to thrive
 - D. megaloblastic anemia
 - E. opportunistic infections

62. The best indicator of chronic folate deficiency is
- A. serum folate level
 - B. RBC folate level
 - C. urinary folate level
 - D. bone marrow aspiration
 - E. CSF 5-methyltetrahydrofolate level
63. Effective supplementation of folate for prevention of neural tube defect is
- A. started at least 1 mo before conception, and continued through the first 2-3 mo of pregnancy
 - B. started at least 3 mo before conception, to be stopped after positive pregnancy test.
 - C. started after positive pregnancy test and continued through the first 2-3 mo of pregnancy.
 - D. started at least 1 mo before conception, to be stopped after positive pregnancy test.
 - E. started at least 3 mo before conception, and continued through the first 2-3 mo of pregnancy
64. The dietary reference intake (DRI) of folate for a healthy eight-month-old infant is
- A. 80 microgm/day
 - B. 150 microgm/day
 - C. 200 microgm/day
 - D. 300 microgm/day
 - E. 400 microgm/day
65. Massive doses of folate given by injection have the potential to cause
- A. ototoxicity
 - B. neurotoxicity
 - C. nephrotoxicity
 - D. severe abdominal pain
 - E. respiratory difficulty
66. Important mechanism for maintaining vitamin B12 nutriture include the following **EXCEPT**
- A. direct absorption
 - B. acidic pH in the ileum
 - C. enterohepatic circulation
 - D. binding with intrinsic factor
 - E. intestinal bacterial synthesis
67. Vitamin B₁₂ is important for all of the following **EXCEPT**

- A. hematopoiesis
- B. mental development
- C. skeletal development
- D. psychomotor development
- E. central nervous system myelination

68. All the following are real risk factors for vitamin B₁₂ deficiency **EXCEPT**

- A. celiac disease
- B. ileal resection
- C. Imerslund-gräsbeck disease
- D. helicobacter pylori infection
- E. use of proton pump inhibitors

69. Common observations with vitamin B₁₂ deficiency in children include all the following **EXCEPT**

- A. hypotonia
- B. peripheral neuritis
- C. developmental regression
- D. involuntary movements
- E. hyperpigmentation of the knuckles

70. The dietary reference intake of cobalamin for a healthy eight-month-old infant is

- A. 0.5 microgm/day
- B. 1.0 microgm/day
- C. 1.5 microgm/day
- D. 2.0 microgm/day
- E. 2.5 microgm/day

71. Radiographic features of scurvy are similar to that of

- A. copper deficiency
- B. magnesium deficiency
- C. vitamin A excess
- D. vitamin D excess
- E. mercury exposure

72. The best indicator of the body stores of vitamin C is

- A. plasma ascorbate concentration
- B. leukocyte ascorbate concentration
- C. urinary ascorbate level
- D. urinary amino acid level
- E. plasma amino acid level

73. The **MOST** specific but late radiographic feature of scurvy is
- A. ground-glass appearance of the shafts of the long bones
 - B. pencil outlining of the epiphysis and diaphysis
 - C. the white line of fränkel at the metaphysis
 - D. Trümmerfeld zone at the metaphysis
 - E. Pelkan spur at cortical ends
74. Ingestion of large doses of vitamin C can cause
- A. musculoskeletal pain
 - B. osmotic diarrhea
 - C. severe headache
 - D. respiratory symptoms
 - E. hematemesis
75. The following are recognized causes of craniotabes **EXCEPT**
- A. osteogenesis imperfect
 - B. rickets
 - C. syphilis
 - D. Sotos syndrome
 - E. normal newborn
76. All the following are recognized causes of rickets **EXCEPT**
- A. Mccune-Albright syndrome
 - B. epidermal nevus syndrome
 - C. tuberous sclerosis
 - D. neurofibromatosis
 - E. aluminum-containing antacids
77. Dominant features in X-linked hypophosphatemic rickets are
- A. head; craniotabes, frontal bossing, and caries
 - B. back; scoliosis, lordosis, and kyphosis
 - C. chest; rachitic rosary and Harrison groove
 - D. extremities; coxa vara and windswept deformity
 - E. general; listlessness and failure to thrive
78. Inorganic phosphorus in rickets is raised in
- A. vitamin D deficiency
 - B. dietary calcium deficiency
 - C. Fanconi syndrome
 - D. chronic renal failure
 - E. tumor-induced rickets

79. Parathyroid hormone level is reduced in
- A. vitamin D dependent rickets
 - B. X- linked hypophosphatemic rickets
 - C. hypophosphatasia
 - D. dietary calcium deficiency
 - E. vitamin D deficiency
80. 25-hydroxy vitamin D level is reduced in
- A. vitamin D deficiency
 - B. autosomal recessive hypophosphatemic rickets
 - C. autosomal dominant hypophosphatemic rickets
 - D. Fanconi syndrome
 - E. tumor-induced rickets
81. Urinary phosphorus is reduced in
- A. chronic renal failure
 - B. autosomal dominant hypophosphatemic rickets
 - C. autosomal recessive hypophosphatemic rickets
 - D. dietary calcium deficiency
 - E. Fanconi syndrome
82. The following hereditary forms of rickets may help to diagnose an apparently healthy mother with the same disease
- A. X- linked hypophosphatemic rickets
 - B. autosomal dominant hypophosphatemic rickets
 - C. autosomal recessive hypophosphatemic rickets
 - D. Fanconi syndrome
 - E. hereditary hypophosphatemic rickets with hypercalciuria
83. In evaluating the cause of rickets, which test is convenient if malabsorption is a consideration?
- A. PT
 - B. PTT
 - C. stool pH
 - D. barium study
 - E. hydrogen breath test
84. The standard method for determining a patient's vitamin D status is
- A. 25-hydroxylase level
 - B. 25-hydroxy vitamin D level
 - C. 1, 25-dihydroxy vitamin D level
 - D. D-binding protein

E. PTH level

85. Regarding vitamin E deficiency in premature infants, one of the following is **TRUE**
- A. it does not cross the placenta
 - B. hemolysis develops in the first week of life
 - C. premature formula that contains poly unsaturated fatty acids may carry a protective effect against hemolysis
 - D. excessive iron supplementation for premature infants may augment the oxidative stress
 - E. causes thrombocytopenia
86. The following conditions/agents are incorporated in a similar presentation to vitamin K deficiency bleeding **EXCEPT**
- A. liver disease
 - B. hereditary factor II deficiency
 - C. high dose salicylate
 - D. heparin
 - E. rodent poison
87. The first factor to be affected by deficiency of vitamin K is
- A. Factor I
 - B. Factor II
 - C. Factor VII
 - D. Factor IX
 - E. Factor X
88. The baby that is least vulnerable to develop vitamin K bleeding tendency is
- A. a three-day old neonate of a healthy mother that started immediate breastfeeding after birth
 - B. a three-year old child spent 21 days in intensive care unit receiving total parenteral nutrition
 - C. a nine-month old infant with prolonged history of diarrhea and multiple antibiotics therapy
 - D. a six-month old infant with cystic fibrosis missed to be diagnosed during early neonatal period
 - E. a six-hour neonate of an epileptic mother that received phenobarbital during pregnancy
89. One of the following matching is **TRUE** regarding the trace elements deficiencies and their consequences
- A. zinc and hypogonadism
 - B. selenium and dental caries

- C. iron and hypothyroidism
- D. fluoride and cardiomyopathy
- E. manganese and decreased alertness

90. One of the following matching is **TRUE** regarding the trace element excess and its consequence

- A. copper and congenital hypothyroidism
- B. manganese and hyperuricemia
- C. iron and abdominal pain
- D. zinc and cholestatic jaundice
- E. fluoride and nail changes

1. **(A).** The first 3 years.
2. **(B).** The AI is 60 gm/day for 0-6 months and 95 gm/day for 7-12 months.
3. **(C).** The AI is 31 gm/day for 0-6 months and 30 gm/day for 7-12 months.
4. **(B).** The AI is 9 gm/day for 0-6 months and 11 gm/day for 7-12 months.
5. **(E).** Alanine, aspartic acid, asparagine, glutamic acid, and serine are dispensable amino acids.
6. **(C).** Thrombocytopenia.
7. **(D).** Cow's milk is a poor source of bioavailable iron.
8. **(E).** Zinc is Essential for proper growth and development, and an important catalyst for about 100 specific enzymes.
9. **(C).** 600 IU/day.
10. **(E).** Diarrhea, otitis media, urinary tract infections, necrotizing enterocolitis, septicemia, infant botulism, celiac disease, childhood cancer, allergy, hospitalizations, and infant mortality all are conditions in which breastfeeding has been suggested to have a possible protective effect.
11. **(D).** Active pulmonary tuberculosis is a real contraindication to breastfeeding. Herpes simplex infection of the breast is another contraindication.
12. **(A).** Breast engorgement happens in the second stage of lactogenesis.
13. **(E).** Group A streptococcus, not group B.
14. **(A).**
15. **(A).** The level is similar in both vegetarian and non-vegetarian diets but the bioavailability is lower in vegetarians.
16. **(C).**
17. **(B).** Hemoglobin cutoffs to define anemia are 110 g/L for children 6-59 mo, 115 g/L for children 5-11 yr, and 120 g/L for children 12-14 yr. Cutoffs to define anemia for non-pregnant women are 120 g/L, 110 g/L for pregnant women, and 130 g/L for men.
18. **(E).** Weight-for-age is the most commonly used index of nutritional status, although a low value has limited clinical significance as it does not differentiate between wasting and stunting. Weight-for-age has the advantage of being somewhat easier to measure than indices that require height measurements.
19. **(A).** For children <5 yr, the global prevalence is estimated to be 33% for vitamin A deficiency, 29% for iodine deficiency, 17% for zinc deficiency, and 18% for iron-deficiency anemia. Prevalence of micronutrient deficiencies tends to be highest in Africa.
20. **(A).**

- 21.(D). Immune function is impaired, especially cell-mediated immunity. The usual responses to infection may be absent, even in severe illness, increasing the risk of undiagnosed infection.
- 22.(C). Edema and hair/skin changes are outward signs of cell damage.
- 23.(B). A controlled transition over 3 days is recommended to prevent the “refeeding syndrome.”
- 24.(A). If no complications; oral Amoxicillin 25 mg/kg twice daily for 5 days. If there are any complications (shock, hypoglycemia, hypothermia, skin lesions, urinary tract or respiratory tract infections, or lethargy); Gentamicin 7.5 mg/kg IV or IM once daily for 7 days and Ampicillin 50 mg/kg IV or IM every 6 hours for 2 days then oral Amoxicillin 25-40 mg/kg every 8 hours for 5 days.
- 25.(A). The signals for entry to this phase are reduced or minimal edema and return of appetite.
- 26.(C). Measles vaccine is given to unimmunized children age >6 mo.
- 27.(A). Iron should be avoided in this phase.
- 28.(E). Treatment of infections is one of the therapeutic directives for stabilization phase.
- 29.(D). The aim of the rehabilitation phase is to restore wasted tissues (i.e., catch-up growth).
- 30.(A). The hallmark of refeeding syndrome is the development of severe hypophosphatemia after the cellular uptake of phosphate during the 1st wk of starting to refeed. Serum phosphate levels of ≤ 0.5 mmol/L can produce weakness, rhabdomyolysis, neutrophil dysfunction, cardio-respiratory failure, arrhythmias, seizures, altered level of consciousness, or sudden death.
- 31.(D). Thiamine deficiency causes encephalopathy, lactic acidosis, and death.
- 32.(A). Hypophosphatemia causes leucocyte dysfunction, hemolysis, and thrombocytopenia.
- 33.(E). Prenatal factors including high preconceptual weight, gestational weight gain, high birth weight, and maternal smoking are associated with increased risk for later obesity. Paradoxically, intrauterine growth restriction with early infant catch-up growth is associated with the development of central adiposity and adult-onset cardiovascular risk. Breastfeeding is only modestly protective for obesity.
- 34.(C). Body fat levels decrease for approximately 5.5 yr until the period called adiposity rebound, when body fat is typically at the lowest level. Adiposity then increases until early adulthood.
- 35.(E). Gastrointestinal hormones, including cholecystikinin, glucagon-like peptide-1, peptide YY, and vagal neuronal feedback promote satiety. Ghrelin stimulates appetite.
- 36.(E). Type 2 diabetes mellitus is a childhood obesity-associated comorbidity.
- 37.(E). Short stature is a common finding in children with obesity due to genetic disorders.
- 38.(E). Antimigraine medications do not influence obesity in children. All other distracters are true causes of secondary obesity.

- 39.(C). Type 2 diabetes mellitus is influenced in obesity, not type 1.
- 40.(E). Glucose, Insulin, Triglyceride level, LDL and HDL, total cholesterol, and liver function tests are the initial markers to be done during evaluation of an obese 12-year girl.
- 41.(A). Orlistat decreases absorption of fat, resulting in modest weight loss. Complications include flatulence, oily stools, and spotting.
- 42.(A). In early vitamin A deficiency, the cornea keratinizes, becomes opaque, is susceptible to infection, and forms dry, scaly layers of cells (xerophthalmia).
- 43.(D). The plasma retinol level is not an accurate indicator of vitamin A status unless the deficiency is severe and liver stores are depleted, in which case low plasma retinol is likely to be evident.
- 44.(E). Manifestations of vitamin C deficiency.
- 45.(E). Nausea, vomiting, and drowsiness; less-common symptoms include diplopia, papilledema, cranial nerve palsies, and other symptoms suggesting pseudotumor cerebri are the hall mark of acute Hypervitaminosis A. Seizures are unlikely.
- 46.(E). Malabsorption is not associated with any risk of carotenemia.
- 47.(A). This triad is rarely reported in infants and young children with severe deficiency secondary to malignancies or feeding of defective formula.
- 48.(D). Death from thiamine deficiency usually is secondary to cardiac involvement. The initial signs are cyanosis and dyspnea, but tachycardia, enlargement of the liver, loss of consciousness, and convulsions can develop rapidly. The heart, especially the right side is enlarged.
- 49.(C). It can develop within 2-3 months of deficient intake. Poor mental status is an early feature. Hoarseness or aphonia caused by paralysis of the laryngeal nerve is a characteristic sign. Dry beriberi reflects neuronal involvement while wet beriberi reflects cardiac involvement.
- 50.(B). This is an autosomal recessive disorder, characterized by Megaloblastic anemia, diabetes mellitus, and sensorineural hearing loss.
- 51.(C). Mutations in genes coding for riboflavin transporter proteins have been identified in children with BVVLS.
- 52.(C). Rapid clinical response to niacin treatment is an important confirmatory test.
- 53.(A). Dermatitis, the most characteristic manifestation of pellagra, can develop suddenly or insidiously and may be initiated by irritants, including intense sunlight.
- 54.(B). 0-6 months: 0.1 mg/ day; 7-12 months: 0.3 mg/ day; 1-3 yr: 0.5 mg/day; 4-8 yr: 0.6 mg/day; 9-13 yr: 1.0 mg/day 14-18 yr: 1.2 - 1.3 mg/day.
- 55.(D). Peripheral neuritis is a feature of deficiency in adults but is not usually seen in children.
- 56.(B). Other distracters cause pyridoxine deficiency.
- 57.(D).
- 58.(C). Methotrexate, not 6-mercaptopurine.
- 59.(A). It manifests usually at 4-6 months of age. Other features are irritability (not lethargy), developmental delay, pyramidal tract signs, choreoathetosis and seizures.

- 60.(D). Non hematologic manifestations include glossitis, listlessness, and growth retardation not related to anemia.
- 61.(A). Hereditary folate malabsorption manifests at 1-3 months of age with recurrent or chronic diarrhea, failure to thrive, oral ulcerations, neurologic deterioration, megaloblastic anemia, and opportunistic infections.
- 62.(B). The normal RBC folate level is 150-600 ng/ mL of packed cells.
- 63.(A). All women desirous of becoming pregnant should consume 400-800 µg folic acid daily; the dose is 4 mg/ day in those having delivered a child with neural tube defect.
64. (A). The DRIs for folate are 65 µg for infants 0-6 months of age and 80µg between 6-12 months of age. For older children, the DRIs are 150 µg for ages 1-3 years; 200 µg for ages 4-8 years; 300 µg for ages 9-13 years; and 400 µg for ages 14-18 years.
- 65.(B).
- 66.(B). Alkaline pH not acidic.
- 67.(C).
- 68.(E). Use of proton pump inhibitors and/or histamine 2 receptor antagonists may increase the risk of deficiency but is not a real cause of deficiency.
- 69.(B). Irritability, developmental delay, developmental regression, and involuntary movements are the common neurologic symptoms in infants and children, whereas sensory deficits, paresthesias, and peripheral neuritis are seen in adults. Hyperpigmentation of the knuckles and palms is another common observation with B12 deficiency in children.
- 70.(A). 0-6 months: 0.4 µg/day; 7-12 months: 0.5 µg/day; 1-3 yr: 0.9 µg/day 4-8 years: 1.2 µg/day 9-13 years: 1.8 µg/day 14-18 years: 2.4 µg/day.
- 71.(A).
- 72.(B).
- 73.(D). The more specific but late radiologic feature of scurvy is a zone of rarefaction under the white line at the metaphysis . is zone of rarefaction (Trümmerfeld zone), a linear break in the bone that is proximal and parallel to the white line, represents area of debris of broken-down bone trabeculae and connective tissue.
- 74.(B). Larger doses of vitamin C can cause gastrointestinal problems, such as abdominal pain and osmotic diarrhea.
- 75.(D). Soto's syndrome causes large head.
- 76.(C). Aluminum-containing antacids cause phosphorus deficiency. McCune-Albright syndrome, epidermal nevus syndrome, and Neurofibromatosis enhance renal losses.
- 77.(D).
- 78.(D). All other distracters are associated with decreased level of inorganic phosphorus.
- 79.(C).
- 80.(A).
- 81.(A). Reduced in chronic kidney disease and dietary phosphorus deficiency only while elevated in all other forms of rickets.

- 82.(A). Undiagnosed disease in the mother is not unusual in X-linked hypophosphatemia.
- 83.(A). Direct measurement of other fat-soluble vitamins (A, E, and K) or indirect assessment of deficiency (prothrombin time for vitamin K deficiency) is appropriate if malabsorption is a consideration.
- 84.(B). The level of 1, 25-D is only low when there is severe vitamin D deficiency.
- 85.(D). It crosses the placenta significantly during the third trimester, which brings premature infants at risk. Hemolysis in premature infants typically started in the second month of life. Formulas containing poly unsaturated fatty acids may lead to exacerbation of deficiency as they make the red blood cells susceptible to oxidative stresses. In premature infants, deficiency causes hemolytic anemia and thrombocytosis, not thrombocytopenia.
- 86.(D). Anticoagulant warfarin and rodent poisons contain Coumarin derivatives that inhibit the action of vitamin K by preventing its recycling to an active form.
- 87.(C). Factor VII has the shortest half life of the coagulation factors.
- 88.(A). Delayed breast feeding is a risk factor to vitamin K bleeding tendency.
- 89.(A). Selenium deficiency causes cardiomyopathy and myopathy; Iron deficiency causes anemia and learning impairment; Manganese deficiency causes hypercholesterolemia and weight loss; Fluoride deficiency causes dental caries.
- 90.(C). Copper excess causes acute (Nausea, vomiting and hepatic coma) and chronic toxicity (liver and brain injury); Manganese excess causes neurological manifestations and cholestatic jaundice; Zinc excess causes abdominal pain, diarrhea and vomiting; Fluoride excess causes dental fluorosis.

PART VII

Fluid and Electrolyte Disorders QUESTIONS

QAHTAN ALOBAIDY

1. The ratio of the intracellular fluid volume to the extracellular fluid volume approaches adult levels at the age of
 - A. 1 yr
 - B. 2 yr
 - C. 3 yr
 - D. 4 yr
 - E. 5 yr
2. Expansion of the intravascular volume and increased intravascular pressure are the main causes of edema in
 - A. lymphatic obstruction
 - B. heart failure
 - C. protein-losing enteropathy
 - D. nephrotic syndrome
 - E. sepsis
3. The critical site for the renal regulation of sodium balance is the
 - A. nephron
 - B. proximal tubule
 - C. loop of Henle
 - D. distal tubule
 - E. collecting duct
4. More than 40% of total body sodium is in the
 - A. intravascular spaces
 - B. interstitial spaces
 - C. intracellular fluid
 - D. bone
 - E. gastrointestinal fluid
5. The **MOST** devastating consequence of untreated hypernatremia is
 - A. brain hemorrhage
 - B. seizures
 - C. central pontinemyelinolysis
 - D. extra pontinemyelinolysis
 - E. brain edema

6. In a child with hypernatremic dehydration, the first priority is restoration of intravascular volume by
- A. 3% saline
 - B. normal saline
 - C. lactated Ringer solution
 - D. 1/3 glucose saline
 - E. 1/2 glucose saline
7. Pseudohyponatremia is present in all the following **EXCEPT**
- A. multiple myeloma
 - B. immunoglobulin infusion
 - C. protein losing enteropathy
 - D. hypertriglyceridemia
 - E. hypercholesterolemia
8. Hypervolemic hyponatremia is caused by
- A. cirrhosis
 - B. cerebral salt wasting
 - C. pseudohypoaldosteronism type I
 - D. obstructive uropathy
 - E. bowel obstruction
9. Asymptomatic hyponatremia is seen in
- A. cirrhosis
 - B. tap water enema
 - C. child abuse
 - D. hyperglycemia
 - E. tubule-interstitial nephritis
10. Syndrome of inappropriate antidiuretic hormone secretion (SIADH) is characterized by
- A. extravascular volume expansion
 - B. high serum uric acid
 - C. high blood urea nitrogen
 - D. euvoletic hyponatremia
 - E. urine sodium <30 mEq/L
11. Spurious hyperkalemia or pseudohyperkalemia in children is usually exaggerated by
- A. blood transfusions
 - B. digitalis intoxication
 - C. thrombocytosis
 - D. renal failure

- E. malignant hyperthermia
12. The **MOST** common cause of hypokalemia in children is
- A. alkalemia
 - B. distal renal tubular acidosis
 - C. gastroenteritis
 - D. diabetic ketoacidosis
 - E. loop diuretic
13. Spurious hypokalemia occurs in patients with
- A. laxative abuse
 - B. high white blood cell count
 - C. hypomagnesaemia
 - D. posthypercapnia
 - E. Cushing syndrome
14. The mechanism of hypokalemia in emesis is mainly due to
- A. gastric loss of potassium
 - B. gastric loss of hydrochloric acid
 - C. low aldosterone level
 - D. dehydration
 - E. hypomagnesaemia
15. Renal manifestations of hypokalemia include all the following **EXCEPT**
- A. polyuria
 - B. urinary retention
 - C. decrease ammonia production
 - D. interstitial nephritis
 - E. renal cysts
16. Medications that cause renal losses of magnesium include all the following **EXCEPT**
- A. amphotericin
 - B. cisplatin
 - C. lithium
 - D. proton pump inhibitors
 - E. mannitol
17. ECG changes in hypomagnesaemia is characterized by
- A. peaking of the T waves and ST-segment depression
 - B. flattened T wave and depressed ST segment
 - C. flattening of the T wave and lengthening of the ST segment
 - D. prolonged PR, QRS, and QT intervals

- E. appearance of a U wave
18. Clinical manifestations of hypermagnesemia include all the following **EXCEPT**
- A. hypotonia
 - B. hyporeflexia
 - C. sleepiness
 - D. hypertension
 - E. vomiting
19. Treatment of hypermagnesemia include all the following **EXCEPT**
- A. intravenous hydration
 - B. loop diuretics
 - C. Kayexalate
 - D. exchange transfusion
 - E. intravenous calcium gluconate
20. The **MOST** common cause of hyperphosphatemia is
- A. acute hemolysis
 - B. vitamin D intoxication
 - C. renal insufficiency
 - D. cow's milk intake
 - E. hypoparathyroidism
21. Systemic calcification occurs when plasma calcium $\times$ plasma phosphorus, both measured in mg/dL is
- A. >90
 - B. >80
 - C. >70
 - D. >60
 - E. >50
22. All the following phosphorus binders are used in chronic renal insufficiency **EXCEPT**
- A. calcium carbonate
 - B. calcium acetate
 - C. sevelamer
 - D. lanthanum
 - E. aluminum
23. Normal anion gap metabolic acidosis can occur in
- A. renal failure
 - B. liver failure
 - C. severe anemia

- D. malignancy
- E. starvation ketoacidosis

24. The time for the kidneys to complete appropriate metabolic compensation for a primary respiratory process is

- A. 12-24 hr
- B. 24-36 hr
- C. 36-48 hr
- D. 48-72 hr
- E. 72-96 hr

25. Metabolic acidosis with increased anion gap occurs in

- A. ammonium chloride intake
- B. posthypocapnia
- C. urinary tract diversions
- D. hyporeninemic hypoaldosteronism
- E. severe anemia

26. Metabolic alkalosis with urinary chloride >20 meq/l occurs in

- A. loop diuretics
- B. cystic fibrosis
- C. Cushing syndrome
- D. emesis
- E. post-hypercapnia

27. In metabolic alkalosis secondary to gastric loss, all the following mechanisms prevent renal bicarbonate loss **EXCEPT**

- A. mineralocorticoid escape phenomenon
- B. reduce GFR
- C. sodium and bicarbonate resorption
- D. increase aldosterone level
- E. hypokalemia

28. Contraction alkalosis occurs in

- A. metformin
- B. diuretic use
- C. propofol
- D. methanol
- E. toluene

29. Measuring serum concentrations of renin and aldosterone differentiates children with metabolic alkalosis; both renin and aldosterone are elevated in

- A. renovascular disease
 - B. glucocorticoid-remediable aldosteronism
 - C. Liddle syndrome
 - D. Cushing syndrome
 - E. 11 β -hydroxylase deficiency
30. Increased production of CO₂ occurs in all the following **EXCEPT**
- A. fever
 - B. emesis
 - C. excess caloric intake
 - D. high levels of physical activity
 - E. increased respiratory muscle work
31. CNS manifestations of respiratory alkalosis include
- A. psychosis
 - B. anxiety
 - C. asterixis
 - D. paresthesia
 - E. hallucinations
32. Respiratory acidosis occurs in
- A. severe anemia
 - B. carbon monoxide poisoning
 - C. hypophosphatemia
 - D. hypotension
 - E. Liddle syndrome
33. The **MOST** important indication of mechanical ventilation in respiratory acidosis is
- A. Pco₂>75 mm Hg
 - B. concomitant metabolic acidosis
 - C. slowly responsive underlying disease
 - D. hypoxia that responds poorly to oxygen
 - E. tiring patient
34. Very-low birth weight infants can have insensible losses of
- A. 10-20 mL/kg/24 hr.
 - B. 30-40 mL/kg/24 hr.
 - C. 50-60 mL/kg/24 hr.
 - D. 70-80 mL/kg/24 hr.
 - E. 100-200 mL/kg/24 hr.

35. For each 1°C increase in temperature above 38°C leads to increase in maintenance water needs by

- A. 5-10%
- B. 10-15%
- C. 15-20%
- D. 20-25%
- E. 25-30%

36. The best fluid bolus giving to a child with isolated vomiting and severe dehydration is

- A. normal saline
- B. ringer lactate
- C. half-normal saline
- D. hypertonic (3%) saline
- E. 5% dextrose + half-normal saline

37. The **MOST** common manifestation of cerebral edema from an overly rapid decrease of serum sodium concentration during correction of hypernatremic dehydration is

- A. irritability
- B. hyperreflexia
- C. spasticity
- D. seizure
- E. coma

PART VII

Fluid and Electrolyte Disorders

ANSWERS

QAHTAN ALOBAIDY

1.(A).

2.(B). There is an increase in venous hydrostatic pressure from expansion of the intravascular volume, which is caused by impaired pumping by the heart, and the increase in venous pressure causes fluid to move from the intravascular space to the interstitial space.

3.(E). Even though the amount of sodium resorbed in this segment is less than in any other segment.

4.(D).

5.(A). As the extracellular osmolality increases, water moves out of brain cells, leading to a decrease in brain volume, this decrease can result in tearing of intra cerebral veins and bridging blood vessels as the brain moves away from the skull and the meninges, patients may have subarachnoid, subdural, and parenchymal hemorrhages. Seizures and brain edema are more common during correction of hypernatremia.

6.(B). Repeated boluses of normal saline (10-20 mL/kg) may be required to treat hypotension, tachycardia, and signs of poor perfusion (poor peripheral pulses and capillary refill time).

7.(C). Hypoalbuminemia caused by gastrointestinal disease (protein losing enteropathy) cause hypervolemichyponatremia, while pseudohyponatremia is a laboratory artifact that is present when the plasma contains very high concentrations of protein (multiple myeloma, intravenous immunoglobulin infusion) or lipid.

8.(A). Other distracters are causes of hypovolemic hyponatremia.

9.(D). Because the manifestations of hyponatremia are a result of the low plasma osmolality, patients with hyponatremia resulting from hyperosmolality do not have symptoms of hyponatremia.

10.(D). Because SIADH is a state of intravascular volume expansion, low serum uric acid and BUN levels are supportive of the diagnosis, the kidney increases sodium excretion in an effort to decrease intravascular volume to normal; thus, the patient has a mild decrease in body sodium(urine sodium >30 mEq/L).

11.(C). This phenomenon is exaggerated with thrombocytosis because of potassium release from platelets, for every 100,000/m³ increase in the platelet count; the serum potassium level rises by approximately 0.15 mEq/L.

12.(C).

13.(B). Spurious hypokalemia occurs in patients with leukemia and very elevated white blood cell counts if sample for analysis is left at room temperature, permitting the white blood cells to take up potassium from the plasma.

- 14.(B). The gastric loss of hydrochloric acid (HCl), leading to a metabolic alkalosis and a state of volume depletion. The kidney compensates for the metabolic alkalosis by excreting bicarbonate in the urine, but there is obligate loss of potassium and sodium with the bicarbonate. The volume depletion raises aldosterone levels, further increasing urinary potassium losses and preventing correction of the metabolic alkalosis and hypokalemia until the volume depletion is corrected.
- 15.(C). Hypokalemia stimulates renal ammonia production, an effect that is clinically significant if hepatic failure is present, because the liver cannot metabolize the ammonia. Hypokalemia impairs bladder function, potentially leading to urinary retention.
- 16.(C). Mild hypermagnesemia may occur in lithium ingestion.
- 17.(C). ECG changes in hyperkalemia include peaking of the T waves, this is followed, as the potassium level increases, by ST-segment depression, an increased PR interval, flattening of the P wave, and widening of the QRS complex. In hypokalemia a flattened T wave, a depressed ST segment, and the appearance of a U wave. In hypermagnesemia ECG changes include prolonged PR, QRS, and QT intervals.
- 18.(D). Elevated magnesium values are associated with hypotension because of vascular dilatation, which also causes flushing. Hypotension can be profound at higher concentrations from a direct effect on cardiac function.
- 19.(C). Kayexalate used for treatment of hypokalemia.
- 20.(C).
- 21.(C).
- 22.(E). Because of the risk of toxicity.
- 23.(A). In renal failure, there is retention of unmeasured anions, including phosphate, urate, and sulfate. The increase in unmeasured anions in renal failure is usually less than the decrease in the bicarbonate concentration. Renal failure is thus a mix of an increased gap and a normal gap metabolic acidosis. The normal gap metabolic acidosis is especially prominent in children with renal failure as a result of tubular damage, as occurs with renal dysplasia or obstructive uropathy, because these patients have a concurrent RTA. Other distractors are causes of high anion gap metabolic acidosis.
- 24.(E). While the respiratory compensation for a metabolic process happens quickly and is complete within 12-24 hr.
- 25.(E). Because of tissue hypoxia.
- 26.(C). Other distractors are causes of metabolic alkalosis and urinary chloride less than 15 meq/L.
- 27.(A).
- 28.(B). Diuretic use causes fluid loss without bicarbonate; thus, the remaining body bicarbonate is contained in a smaller total body fluid compartment.
- 29.(A). Aldosterone is high and renin is low in patients with adrenal adenomas or hyperplasia and glucocorticoid-remediable aldosteronism. Renin and aldosterone are low in children with Cushing syndrome, Liddle syndrome, licorice ingestion, 17 α -

hydroxylase deficiency, 11 β -hydroxylase deficiency, and 11 β -hydroxysteroid dehydrogenase deficiency.

30.(B).

31.(D). The paresthesia, tetany, and seizures may be partially related to the reduction in ionized calcium that occurs because alkalemia causes more calcium to bind to albumin.

32.(C). Hypophosphatemia cause respiratory muscle weakness.

33.(A).

34.(E).

35.(B).

36.(A). Ringer lactate should not be used because the lactate would worsen the alkalosis.

37.(D).

Pediatric Drug Therapy QUESTIONS

KHALID ALAARJI

1. Fentanyl anesthesia (used as analgesia to provide pain relief during operative procedures in neonates) reduces the incidence of all the following postoperative conditions **EXCEPT**

- A. acidosis
- B. tachycardia
- C. hypotension
- D. coagulation abnormalities
- E. intraventricular hemorrhage

2. Postoperative nausea and vomiting (PONV) may be related to the stress, trauma of surgery, and pain.

Of the following, the **MOST** likely drug that cause PONV is

- A. ketorolac
- B. droperidol
- C. acetaminophen
- D. opioid analgesics
- E. local anesthetic agents

3. Postoperative nausea and vomiting (PONV) occurs in the immediate postoperative period, within the 1st 1-2 hr, or several hours after surgery and anesthesia.

All the following may decrease the incidence of PONV **EXCEPT**

- A. hydration
- B. use of ketorolac
- C. preoperative fasting
- D. glucose supplementation
- E. prophylactic use of ondansetron

4. Malignant hyperthermia is an acute hypermetabolic syndrome that is triggered by inhalational anesthetic agents and succinylcholine.

Of the following, the **MOST** recognized clue to the risk of malignant hyperthermia is

- A. hypercarbia
- B. rapid onset of fever
- C. ventricular fibrillation
- D. acute hemoglobinuria
- E. rigid clenching of the masseter muscles

5. Postanesthetic apnea is most common within the 1st 12 hr after surgery and has been reported in premature infants up to 48 hr later.

General anesthesia should be avoided (except for emergency surgery) in full-term infants younger than postconceptual age of

- A. 44 wk
- B. 46 wk
- C. 48 wk
- D. 50 wk
- E. 52 wk

6. Upper respiratory illness (URI) can increase airway reactivity for up to 6 wk in both normal children and children with a history of reactive airway disease.

All the following should be taken in concern when preparing a child for operation with history of URI **EXCEPT**

- A. fever
- B. injected sclerae
- C. clear rhinorrhea
- D. productive cough
- E. increased mucous secretions

7. After Upper respiratory illness (URI), it is generally recommended to avoid general anesthesia in elective procedures for

- A. 1-2Wk
- B. 2-4Wk
- C. 4-6Wk
- D. 6-8Wk
- E. 8-10 wk

8. The child with asthma should be free of wheezing for at least several days before surgery, even if this necessitates an increase in β -agonist dosage and the addition of steroids.

Of the following, the **MOST** appropriate preoperative steroids regime in children with asthma (who are receiving asthma therapy or who have received steroid within the last year) is

- A. prednisone, 1 mg/kg given 24 and 12 hr before surgery
- B. prednisone, 2 mg/kg given 24 and 12 hr before surgery
- C. dexamethason, 0.6 mg/kg given 24 and 12 hr before surgery
- D. hydrocortisone IV, 5 mg/kg given 24 and 12 hr before surgery
- E. hydrocortisone IV, 10 mg/kg given 24 and 12 hr before surgery

9. Children with anterior mediastinal masses are at serious risk for airway compromise, cardiac tamponade, and vascular obstruction. The indication to prohibit general anesthesia is CT scan showing compression of the airway at the carina

- A. $\geq 30\%$
- B. $\geq 40\%$
- C. $\geq 50\%$
- D. $\geq 60\%$
- E. $\geq 70\%$

10. During general anesthesia in children with Down syndrome, the anesthesiologists should be aware of all the following **EXCEPT**

- A. macroglossia
- B. cardiac anomalies
- C. atlantoaxial instability
- D. upper airway obstruction
- E. temporo-mandibular subluxation

11. Of the following, the **MOST** acceptable hematocrit value for routine elective anesthesia is

- A. $\geq 28\%$
- B. $\geq 30\%$
- C. $\geq 32\%$
- D. $\geq 34\%$
- E. $\geq 36\%$

12. Preoperative psychologic preparation programs decrease the incidence of postoperative behavioral changes which last for up to 1 mo.

Of the following, the **MOST** appropriate advice is to use oral

- A. diazepam
- B. midazolam
- C. imipramine
- D. pentobarbital
- E. chloral hydrate

13. Aspiration of gastric contents is a perioperative disaster and, if superimposed on lung disease, may be rapidly fatal.

All the following are true about the guidelines for preoperative fasting **EXCEPT**

- A. clear, sweet liquids --- 2hr
- B. breast milk ----- 4hr
- C. infant formula ----- 6hr
- D. solid food ----- 8hr
- E. gelatin -----10hr

14. Nystagmus is a sign of poisoning with all the following **EXCEPT**

- A. cocaine
- B. alcohols
- C. ketamine
- D. anticonvulsant
- E. dextromethorphan

15. According to the National Poison Data Systems of the American Association of Poison Control Centers, approximately 50% of poisoning occurs in children

- A. ≤ 2 yr old
- B. ≤ 4 yr old
- C. ≤ 6 yr old
- D. ≤ 8 yr old
- E. ≤ 10 yr old

16. In poisoning, obtaining an accurate problem-oriented history is of paramount importance.

All the following are historical features that suggest a possible diagnosis of poisoning in patients without a witnessed exposure **EXCEPT**

- A. low levels of household stress
- B. multiple system organ dysfunction
- C. sudden alteration of mental status
- D. age of the child (toddler or adolescent)
- E. acute onset of symptoms without prodrome

17. Acetone odor is a sign of poisoning with all the following **EXCEPT**

- A. cyanide
- B. methanol
- C. salicylates
- D. paraldehyde
- E. isopropyl alcohol

18. Lacrimation is a sign of poisoning with

- A. atropin
- B. salicylate
- C. imipramin
- D. acetaminophine
- E. organophosphates

19. Mydriasis is a sign of poisoning with

- A. opioids
- B. clonidine

- C. hypnotics
 - D. organophosphates
 - E. tricyclic antidepressant
20. Constipation can be seen in poisoning with
- A. iron
 - B. lead
 - C. colchicine
 - D. cholinergics
 - E. antimicrobials
21. Hematemesis is a recognized finding in the following poisoning **EXCEPT**
- A. lead
 - B. iron
 - C. NSAIDs
 - D. caustics
 - E. salicylates
22. Which of the following poisoning may cause hyperglycemia?
- A. ethanol
 - B. quinine
 - C. caffeine
 - D. Beta blockers
 - E. salicylates (late)
23. All the following poisonings may cause peripheral neuropathy **EXCEPT**
- A. lead
 - B. arsenic
 - C. lithium
 - D. mercury
 - E. organophosphates
24. All the following poisonings may have a radiopaque shadow on KUB **EXCEPT**
- A. lead
 - B. arsenic
 - C. salicylates
 - D. calcium carbonate
 - E. enteric-coated pills
25. Prolonged PR interval is a sign of poisoning with
- A. lithium
 - B. fluconazole

- C. erythromycin
- D. carbamazepine
- E. diphenhydramine

26. Prolonged QTc is a sign of poisoning with

- A. amiodarone
- B. carbamazepine
- C. diphenhydramine
- D. cardiac glycosides
- E. tricyclic antidepressants

27. Antidotes are available for relatively few toxins, but early and appropriate use of an antidote is a key element in managing the poisoned patient.

The following matching are true **EXCEPT**

- A. β Blockers ----- glucagon
- B. opioids ----- naloxone
- C. iron ----- deferoxamine
- D. lead ----- BAL (dimercaprol)
- E. isoniazid ----- thiamine

28. The following matching are true regarding poisoning and antidotes **EXCEPT**

- A. acetaminophen ----- N-Acetylcysteine
- B. anticholinergics ----- physostigmine
- C. benzodiazepines ----- nalorphene
- D. carbon monoxide ----- 100% oxygen
- E. organophosphates ----- pralidoxime

29. The majority of poisonings in children are from ingestion, although exposures can also occur via inhalational, dermal, and ocular route.

All the following are true about decontamination **EXCEPT**

- A. the goal is to minimize absorption of the toxic substance
- B. decontamination should be routinely employed for every poisoned patient
- C. water should not be used for decontamination after exposure to phosphorus
- D. the efficacy of the intervention decreases with increasing time since exposure
- E. the specific method, depends on the properties of the toxin and the route of exposure

30. All the following poisonings may cause QTc prolongation **EXCEPT**

- A. lithium
- B. cisapride
- C. amiodarone
- D. antipsychotics

E. clarithromycin

31. Dermal and ocular decontamination begin with removal of any contaminated clothing and particulate matter, followed by flushing of the affected area with tepid water or normal saline.

Of the following, water should be used for dermal decontamination after exposure to

- A. phosphorus
- B. calcium oxide
- C. organophosphate
- D. elemental sodium
- E. titanium tetrachloride

32. Gastrointestinal (GI) decontamination is a controversial topic among medical toxicologists and is most likely to be effective in the 1st hour after an acute ingestion.

GI decontamination can be done even 1 hr after ingestion of the following **EXCEPT**

- A. antipsychotics
- B. anticholinergics
- C. massive pill ingestions
- D. enteric-coated salicylates
- E. sustained-release preparations

33. The majority of poisonings in children are from ingestion, although exposures can also occur via inhalational, dermal, and ocular routes.

All the following are described methods of GI decontamination **EXCEPT**

- A. gastric lavage
- B. whole-bowel irrigation
- C. induced emesis with ipecac
- D. single-dose activated charcoal
- E. multiple-dose activated charcoal

34. After a review of the evidence and assessment of the risks and benefits of ipecac use, the American Academy of Clinical Toxicology have published statements that advise to

- A. use ipecac with caution
- B. use ipecac in all poisoning
- C. abandon the use of ipecac
- D. use ipecac in some poisoning
- E. not use ipecac in persistently vomiting patient

35. Although gastric lavage was used routinely for many years, in most clinical scenarios of poisoning, the use of gastric lavage is no longer recommended due to all the following **EXCEPT**

- A. painful
 - B. time-consuming
 - C. can induce tachycardia
 - D. only removes a fraction of gastric contents
 - E. can delay administration of activated charcoal
36. All the following poisonings may cause QRS prolongation **EXCEPT**
- A. sotalol
 - B. quinidine
 - C. chloroquine
 - D. propranolol
 - E. procainamide
37. All the following substances are poorly adsorbed by activated charcoal **EXCEPT**
- A. iron
 - B. caustics
 - C. alcohols
 - D. hydrocarbons
 - E. benzodiazepines
38. Hypocalcemia is a sign of poisoning with
- A. ethanol
 - B. fluoride
 - C. Beta Blockers
 - D. salicylates (late)
 - E. calcium channel blockers
39. Alkalinization of urine is **MOST** useful in managing toxicity with
- A. iron
 - B. lead
 - C. methotrexate
 - D. phenothiazine
 - E. cyclophosphamide
40. All the following drugs toxicity can be removed by hemodialysis **EXCEPT**
- A. iron
 - B. methanol
 - C. salicylates
 - D. theophylline
 - E. valproic acid

41. Intralipid emulsions therapy (IET) act by sequestering fat-soluble drugs and decreasing their impact at target organs thus enhance the elimination in case of drug toxicity.

All the following can be eliminated by IET **EXCEPT**

- A. diltiazem
- B. verapamil
- C. amiodrone
- D. imipramine
- E. amitriptyline

42. All the following methods can be used to enhance the elimination of toxins **EXCEPT**

- A. hemodialysis
- B. urinary alkalization
- C. whole bowel irrigation
- D. intralipid emulsions therapy
- E. multiple-dose activated charcoal

43. Of the following, the **SINGLE** acute toxic dose of acetaminophen is

- A. ≥ 100 mg/kg
- B. ≥ 150 mg/kg
- C. ≥ 200 mg/kg
- D. ≥ 250 mg/kg
- E. ≥ 300 mg/kg

44. In toxic ingestion of acetaminophen, the serum level should be measured after

- A. 2 hr
- B. 4 hr
- C. 6 hr
- D. 8hr
- E. 10 hr

45. All the following poisonings may cause ataxia **EXCEPT**

- A. alcohols
- B. imipramin
- C. anticonvulsant
- D. carbon monoxide
- E. dextromethorphan

46. When considering the treatment of a patient with acetaminophen poisoning, and after assessment of the ABCs, it is helpful to place the patient into one of 4 stages.

Of the following, stage 1 is characterize by

- A. nausea and vomiting

- B. elevated alanine aminotransferase
- C. high international normalized ratio
- D. right upper quadrant abdominal pain
- E. elevated aspartate aminotransferase

47. The King's College criteria for consideration of liver transplantation in acetaminophen poisoning with acute liver failure (stage 3) include all the following **EXCEPT**

- A. coagulopathy (INR >6)
- B. acidemia (serum pH <7.3)
- C. serum lactic acid >3 mmol/L
- D. grade III or IV hepatic encephalopathy
- E. renal dysfunction (creatinine >3.4 mg/dL)

48. In acetaminophen poisoning, when the initiation of N-Acetylcysteine therapy is delayed, there is the greater the risk of acute liver failure.

N-Acetylcysteine should be instituted no later than

- A. 2 hr after ingestion
- B. 4 hr after ingestion
- C. 6 hr after ingestion
- D. 8 hr after ingestion
- E. 10 hr after ingestion

49. Wintergreen odor is a sign of poisoning with

- A. arsenic
- B. thallium
- C. selenium
- D. methyl salicylate
- E. organophosphates

50. The acute toxic dose of salicylates is >150 mg/kg, more significant toxicity is seen after ingestions of >300 mg/kg and severe, potentially fatal, toxicity is described after ingestions of

- A. ≥350 mg/kg
- B. ≥400 mg/kg
- C. ≥450 mg/kg
- D. ≥500 mg/kg
- E. ≥550 mg/kg

51. In early course of acute salicylism, the **MOST** dominate lab values are

- A. metabolic acidosis and hypoglycemia
- B. metabolic acidosis and hyperglycemia

- C. respiratory alkalosis and hypoglycemia
- D. metabolic acidosis and normoglycemia
- E. respiratory alkalosis and hyperglycemia

52. Salicylate ingestions are classified as acute or chronic, and acute toxicity is far more common in pediatric patients.

Which of the following is not a recognized feature of salicylate poisoning?

- A. tinnitus
- B. tachypnea
- C. diaphoresis
- D. bradycardia
- E. hyperthermia

53. A 2.5 year-old- child brought to emergency room immediately upon finding the boy with open bottle of some drugs (used by his grandfather). The child has vomited once, fussy, and lethargic. Physical evaluation reveals a 13-kg child who has tachypnea and tachycardia. Laboratory results include a blood sugar=225mg/dl, pH=7.5, PCO₂=20 mEq/L, and HCO₃=20 mEq/L.

Of the following, the **MOST** likely offending drug is

- A. iron
- B. atropine
- C. salicylate
- D. methylxanthine
- E. tricyclic antidepressant

54. Bradycardia is a sign of poisoning with

- A. antipsychotics
- B. anticholinergics
- C. antidepressants
- D. methylxanthines
- E. calcium channel blockers

55. In salicylate poisoning, hyperglycemia (early) and hypoglycemia (late) have been described.

Of the following, the **MOST** appropriate clue to administer IV glucose is

- A. vomiting
- B. convulsion
- C. palpitation
- D. diaphoresis
- E. altered mental status

56. Chronic salicylism have an insidious presentation, and patients can show marked toxicity at significantly lower salicylate levels than in acute toxicity.

Of the following, the **MOST** appropriate primary mode of therapy in chronic salicylism is

- A. IV glucose
- B. hemodialysis
- C. urinary alkalinization
- D. volume resuscitation
- E. gastric decontamination

57. Ibuprofen is well tolerated even in overdose. The acute dose of ibuprofen that result in more serious effects (altered mental status and metabolic acidosis) is

- A. >100 mg/kg
- B. >200 mg/kg
- C. >300 mg/kg
- D. >400 mg/kg
- E. >500 mg/kg

58. In acute toxic ingestion of ibuprofen, symptoms usually develop within 4-6 hr of ingestion and resolve within 24 hr.

All the following can result from acute ingestion of variable dose of ibuprofen **EXCEPT**

- A. gastric ulcers
- B. CNS depression
- C. abdominal pain
- D. renal insufficiency
- E. nausea and vomiting

59. Of the following, the **MOST** important manifestation of hydrocarbons toxicity is

- A. dysrhythmias
- B. hepatic toxicity
- C. intestinal necrosis
- D. bacterial pneumonitis
- E. aspiration pneumonitis

60. Salivation is a sign of poisoning with all the following **EXCEPT**

- A. mercury
- B. ketamine
- C. salicylates
- D. corrosives
- E. organophosphates

61. The most important manifestation of hydrocarbon toxicity is aspiration pneumonitis via inactivation of the type II pneumocytes and resulting surfactant deficiency.

Of the following, the propensity of a hydrocarbon to cause aspiration pneumonitis is

- A. inversely proportional to its viscosity
- B. inversely proportional to its viscosity and volatility
- C. directly proportional to its viscosity and volatility
- D. inversely proportional to its viscosity, and directly proportional to its volatility
- E. directly proportional to its viscosity, and inversely proportional to its volatility

62. Hydrocarbons with low viscosity and high volatility (kerosene, gasoline) spread rapidly across surfaces and cover large areas of the lungs when aspirated.

The quantity of aspirated hydrocarbon chemicals that can produce significant injury is less than

- A. 1 mL
- B. 3 mL
- C. 5 mL
- D. 7 mL
- E. 10 mL

63. Certain hydrocarbons have unique toxicities and can cause symptoms after ingestion, inhalation, or dermal exposures.

Of the following, the **MOST** likely cause of acute myelogenous leukemia is long-term exposure to

- A. gasoline
- B. benzene
- C. kerosene
- D. nitrobenzene
- E. methyl alcohol

64. Methemoglobinemia may result from exposure to

- A. gasoline
- B. benzene
- C. kerosene
- D. nitrobenzene
- E. carbon monoxide

65. All the following are clinical manifestations of hydrocarbons poisoning **EXCEPT**

- A. transient mild CNS depression
- B. cough is the first clinical finding
- C. chest radiographs may be initially normal
- D. pneumatoceles can appear 2-3 wk after exposure

- E. fever and leukocytosis indicate bacterial superinfection
66. Dysrhythmias may result from exposure to
- A. benzene
 - B. kerosene
 - C. nitrobenzene
 - D. carbon tetrachloride
 - E. halogenated hydrocarbons
67. Carbon monoxide (CO) is a colorless, odorless gas produced during the combustion of any carbon-containing fuel (the less efficient the combustion, the greater the amount of CO produced).
- All the following are true about CO poisoning **EXCEPT**
- A. CO displaces nitric oxide (NO) from proteins
 - B. NO is responsible for headache, syncope, and hypotension
 - C. HbCO levels are well correlated with clinical signs of toxicity
 - D. CO bind to cytochrome oxidase, disrupt cellular respiration
 - E. CO binds to hemoglobin with an affinity >200 times that of oxygen
68. Early symptoms of carbon monoxide (CO) poisoning are nonspecific and include headache, malaise, nausea, vomiting, and the skin may appear
- A. blue
 - B. pale
 - C. normal
 - D. grayish
 - E. cherry-red
69. In ambient air, the average half-life of HbCO is 4-6 hr, while after administration of 100% oxygen the half-life is
- A. 0.5-1 hr
 - B. 1-1.5 hr
 - C. 1.5-2 hr
 - D. 2-3 hr
 - E. 3-4 hr
70. All the following poisoning may cause QTc prolongation **EXCEPT**
- A. sotalol
 - B. propranolol
 - C. ketoconazole
 - D. erythromycin
 - E. phenothiazines

71. Caustics include acids and alkalis as well as a few common oxidizing agents, strong acids and alkalis can produce severe injury even in small-volume ingestions.

All the following are true about a caustic poisoning **EXCEPT**

- A. pH of <5 produce significant injury
- B. pH of >12 produce significant injury
- C. acids produce a coagulative necrosis
- D. alkalis produce a liquefaction necrosis
- E. prolong contact produce significant injury

72. Clinical manifestations of organophosphate toxicity relate to the accumulation of acetylcholine at peripheral nicotinic and muscarinic synapses and in the CNS.

All the following are symptoms of cholinergic excess at muscarinic receptors **EXCEPT**

- A. miosis
- B. emesis
- C. salivation
- D. bradycardia
- E. urine retention

73. Nicotinic signs and symptoms of organophosphates poisoning include all the following **EXCEPT**

- A. tachycardia
- B. hypotension
- C. fasciculation
- D. hypoventilation
- E. muscle weakness

74. All the following are true about treatment of organophosphate poisoning **EXCEPT**

- A. activated charcoal is unlikely to be of benefit
- B. without treatment, symptoms can persist for weeks
- C. atropine is primarily targeted to correct bradycardia
- D. washing exposed skin and removing exposed clothing
- E. pralidoxime breaks the bond between the enzyme and the organophosphate

75. All the following are done in the initial assessment of digoxin toxicity **EXCEPT**

- A. ECG
- B. serum calcium
- C. serum potassium
- D. renal function test
- E. serum digoxin level

76. All the following poisoning may cause hypoglycemia **EXCEPT**

- A. quinine

- B. ethanol
- C. sulfonyleureas
- D. beta Blockers
- E. calcium channel blockers

77. All the following matching are true **EXCEPT**

- A. salicylates ----- sodium bicarbonate
- B. organophosphates ----- atropine
- C. methemoglobinemia ----- methylene blue
- D. calcium channel blockers ----- insulin
- E. tricyclic antidepressants ----- flumazenil

78. Digoxin has a very narrow therapeutic index, therapeutic plasma digoxin concentrations are 0.5-2.0 ng/mL; a level >2 ng/mL is considered toxic and a level >6 ng/mL is considered potentially fatal.

All the following medications increase serum digoxin concentration **EXCEPT**

- A. tetracyclin
- B. amiodarone
- C. itraconazole
- D. clarithromycin
- E. spironolactone

79. In digoxin toxicity, Fab fragments bind free digoxin in both the intravascular and the interstitial space to form a pharmacologically inactive complex that is subsequently renally eliminated.

All the following are indications of Fab fragments **EXCEPT**

- A. renal failure
- B. hypertension
- C. ingestion >4 mg
- D. K^+ value >5-5.5 mEq/L
- E. life threatening dysrhythmias

80. Iron-containing products remain widely available, with the most potentially toxic being adult iron preparations.

Of the following, the severity of iron toxicity is **MOSTLY** related to the

- A. age of child
- B. time of presentation
- C. type of salt preparation
- D. number of tablets ingested
- E. amount of elemental iron ingested

81. The amount of elemental iron ingested that should be referred to medical care for evaluation is more than

- A. 10 mg/kg
- B. 20 mg/kg
- C. 30 mg/kg
- D. 40 mg/kg
- E. 50 mg/kg

82. A 2.5 year-old-boy presented with tachycardia, pallor, and fatigue. The history reveals excessive iron ingestion with unknown duration.

Of the following, the **MOST** likely time of ingestion is

- A. 30 min-6hr ago
- B. 6hr-24 hr ago
- C. 12hr-36 hr ago
- D. 36hr-48hr ago
- E. 48hr-72hr ago

83. Cyanosis (unresponsive to oxygen) is a sign of poisoning with

- A. atropine
- B. amiodarone
- C. vancomycin
- D. carbon monoxide
- E. elemental mercury

84. Iron is directly corrosive to the GI mucosa, leading to hematemesis, melena, ulceration, infarction, and potential perforation.

Of the following, the **MOST** appropriate method of gastrointestinal decontamination in iron toxicity is

- A. induced emesis with ipecac
- B. gastric lavage
- C. activated charcoal
- D. whole-bowel irrigation
- E. all of the above

85. Cardiovascular (CV) and CNS symptoms dominate the clinical presentation of tricyclic antidepressant (TCA) toxicity, symptoms typically develop within 1-2 hr of ingestion, and serious toxicity usually manifests within 6 hr of ingestion.

Of the following, the **MOST** common CV manifestation is

- A. hypotension
- B. sinus tachycardia
- C. widened QRS interval
- D. ventricular arrhythmias

E. prominent R wave in lead aVR

86. The **MOST** common cause of death in tricyclic antidepressant (TCA) overdose is

- A. seizures
- B. renal injury
- C. respiratory failure
- D. refractory hypotension
- E. ventricular arrhythmias

87. Asymptomatic child with tricyclic antidepressant toxicity, should be observed with continuous cardiac monitoring and serial ECGs for at least

- A. 4hr
- B. 6hr
- C. 8hr
- D. 10hr
- E. 12hr

88. Probiotics are oral supplement or a food product that contains a sufficient number of viable microorganisms (live microorganisms that are most often referred to as "friendly" or "good" bacteria).

Probiotics are indicated in all the following **EXCEPT**

- A. constipation
- B. irritable bowel syndrome
- C. protein losing enteropathy
- D. antibiotic-associated diarrhea
- E. inflammatory bowel disorders

89. Dietary supplements (as herbs) are commonly used and may be helpful adjunctive treatments for common childhood problems.

All the following matching are true **EXCEPT**

- A. Aloe vera ----- lice
- B. Ginger ----- Nausea
- C. Chamomile ----- dyspepsia
- D. Tea tree oil ----- acne remedies
- E. Peppermint -- Irritable bowel syndrome

- 1.(B). Reduce the incidence of other postoperative conditions as bradycardia, hypoglycemia, and death.
- 2.(D). The etiology may be also related to the stress and trauma of surgery combined with the emetic effects of anesthetic agents. Pain is an important cause of nausea and vomiting.
- 3.(C). Preoperative fasting does not decrease the incidence of nausea and vomiting.
- 4.(E). Acute myoglobinuria associated with a malignant hyperthermia triggering agent is another clue.
- 5 (A).
- 6.(C). Purulent green nasal discharge.
- 7.(C). URIs can increase the risk of laryngospasm and bronchospasm, reduce mucociliary clearance, and raise the risk of intraoperative atelectasis and hypoxemia. It is generally recommended to avoid general anesthesia for elective procedures for 4-6 wk after a URI.
- 8.(A).
- 9.(C).
- 10.(E). In children with Down syndrome, it is wise to exercise caution in stabilizing the cervical spine and also to avoid cervical flexion and extension.
- 11.(B). If there are reasons to expect significant blood loss or prolonged convalescence, anemia should be corrected preoperatively. In the emergency setting, transfusion may be required.
- 12.(B). Oral midazolam (0.5 mg/ kg) may decrease negative behavioral changes after surgery. Midazolam has the benefit of providing not only rapid-onset anxiolysis in 10-20 min but also very effective and rapid (10 min) amnesia.
- 13.(E). Guidelines for preoperative fasting ("2-4-6-8 Rule"), fruit juices, gelatin ----- 6hr.
- 14.(A). Cocaine causes mydriasis.
- 15.(C).
- 16.(A). High levels of household stress should suggest a possible diagnosis of poisoning.
- 17.(A). Bitter almonds odor is a sign of poisoning with cyanide.
- 18.(E).
- 19.(E). The other distracters may cause miosis.
- 20.(B). Antimicrobials, iron, cholinergics, and colchicines cause diarrhea.
- 21.(A). Lead may cause constipation.
- 22.(C). Hypoglycemia is a sign of other distracters.
- 23.(C). Lithium may cause delirium/psychosis and ataxia.
- 24.(C).

- 25.(A). Erythromycin and fluconazole may cause QTc prolongation while diphenhydramine and carbamazepine may cause QRS prolongation.
- 26.(A). The other distracters may cause wide QRS.
- 27.(E). Pyridoxine is antidote for isoniazid.
- 28.(C). Flumazenil is antidote for benzodiazepines.
- 29.(B). Decontamination should not be routinely employed for every poisoned patient. Instead, careful decisions regarding the utility of decontamination should be made for each patient and should include consideration of the toxicity and pharmacologic properties of the exposure, the route of the exposure, the time since the exposure, and the risks vs the benefits of the decontamination method.
- 30.(A). Lithium may cause PR interval prolongation.
- 31.(C). Dermal decontamination, especially after exposure to adherent or lipophilic (e.g., organophosphate) agents, should include thorough cleansing with soap and water. Water should not be used for decontamination after exposure to highly reactive agents, such as elemental sodium, phosphorus, calcium oxide, and titanium tetrachloride.
- 32.(A). Gastrointestinal decontamination strategies are most likely to be effective in the 1st hour after an acute ingestion. GI absorption may be delayed after ingestion of agents that slow GI motility (anticholinergic medications, opioids), massive pill ingestions, sustained-release preparations, and ingestions of agents that can form pharmacologic bezoars (e.g., enteric-coated salicylates).
- 33.(E). Whereas single-dose activated charcoal is used as a method of decontamination, multiple doses of activated charcoal (MDACs) can help to enhance the elimination of some toxins.
- 34.(C).
- 35.(C). Gastric lavage can induce bradycardia via a vagal response to tube placement.
- 36.(A). Sotalol may cause QTc prolongation.
- 37.(E). Charcoal is “activated” via heating to extreme temperatures, creating an extensive network of pores that provides a very large adsorptive surface area. Many, but not all, toxins are adsorbed onto its surface, thus preventing absorption from the GI tract.
- 38.(B).
- 39.(C). Urinary alkalinization (UA) enhances the elimination of some drugs that are weak acids by forming charged particles that are “trapped” within the renal tubules and thus excreted. Urinary alkalinization is accomplished via a continuous infusion of sodium bicarbonate-containing intravenous fluids, with a goal urine pH of 7.5-8. Alkalinization of the urine is most useful in managing salicylate and methotrexate toxicity.
- 40.(A). Toxins that are amenable to dialysis have the following properties: low volume of distribution (<1 L/kg), low molecular weight, low degree of protein binding, and high degree of water solubility.

- 41.(C). Lipophilic drugs are potentially bound by Intralipid emulsions, including calcium channel blockers (verapamil and diltiazem) and tricyclic antidepressants.
- 42.(C). Enhancing elimination is a potentially lifesaving intervention that results in improved clearance of a poison that has already been absorbed. Whole bowel irrigation used for gastrointestinal decontamination.
- 43.(C). In acetaminophen overdose, glutathione stores are overwhelmed, and free N-acetyl-p-benzoquinone imine is able to combine with hepatic macromolecules to produce hepatocellular necrosis.
- 44.(B). Acetaminophen levels obtained <4 hr after ingestion, unless “nondetectable,” are difficult to interpret and cannot be used to estimate the potential for toxicity.
- 45.(B). CNS toxicity of antidepressant can include lethargy, coma, myoclonic jerks, and seizures.
- 46.(A). Other findings present in stage 2.
- 47.(C). A serum lactic acid >3 mmol/L (after IV fluids) adds to both the sensitivity and specificity of the criteria to predict death without liver transplant. The degree of transaminase elevation is not a factor in this decision making process.
- 48.(D). The importance of instituting therapy with either IV or oral N-Acetylcysteine (NAC) no later than 8 hr from the time of ingestion cannot be overemphasized. No patient, no matter the size of the ingestion, who receives NAC within 8 hr of overdose, should die from liver failure. The further out from the 8 hr mark the initiation of therapy is delayed, the greater the risk of acute liver failure.
- 49.(D). Garlic odor is a sign of poisoning with other distracters.
- 50.(D).
- 51.(E). Classically, lab values from a patient poisoned with salicylates reveal a primary respiratory alkalosis and a primary, elevated anion gap, metabolic acidosis. Early in the course of acute salicylism, respiratory alkalosis dominates. As the respiratory stimulation diminishes, the patient will move toward the metabolic acidosis.
- 52.(D). The tachycardia results in large part from marked insensible losses from vomiting, tachypnea, diaphoresis, and uncoupling of oxidative phosphorylation.
- 53.(C). Early in the course of acute salicylism, respiratory alkalosis dominates. As the respiratory stimulation diminishes, the patient will move toward the metabolic acidosis. Hyperglycemia (early) and hypoglycemia (late) have been described.
- 54.(E). Tachycardia is a sign of poisoning with other distracters.
- 55.(E). Parenteral glucose should be provided in altered mental status as they may have CNS hypoglycemia not noted in a peripheral serum glucose test.
- 56.(C). Urinary alkalization enhances the elimination of salicylates by converting salicylate to its ionized form, “trapping” it in the renal tubules, and thus enhancing elimination.
- 57.(D). In children, acute doses of <200 mg/kg rarely cause toxicity, but ingestions of >400 mg/kg can produce more serious effects, including altered mental status and metabolic acidosis.

- 58.(A). GI bleeding and ulcers have been described with chronic use; they are rare in the setting of acute ingestion.
- 59.(E). The most important manifestation of hydrocarbon toxicity is aspiration pneumonitis via inactivation of the type II pneumocytes and resulting surfactant deficiency.
- 60.(A). Mercury, lead, arsenic, and bismuth cause gum lines.
- 61.(D). With low viscosity and high volatility, such as mineral spirits, naphtha, kerosene, gasoline, and lamp oil, spread rapidly across surfaces and cover large areas of the lungs when aspirated.
- 62.(A). Only small quantities (<1 mL) of such chemicals need be aspirated to produce significant injury.
- 63.(B). Benzene is known to cause cancer, most commonly acute myelogenous leukemia, after long-term exposure.
- 64.(D). Nitrobenzene, aniline, and related compounds can produce methemoglobinemia.
- 65.(E). Fever and leukocytosis are common accompanying signs in patients with pneumonitis and don't necessarily imply bacterial superinfection.
- 66.(E). Halogenated hydrocarbons (which contain a chlorine, bromine, or fluorine), can sensitize the myocardium to the effects of endogenous catecholamines. This can result in dysrhythmias and "sudden sniffing death".
- 67.(C). CO displaces oxygen and creates a conformational change in hemoglobin that impairs the delivery of oxygen to the tissues, leading to tissue hypoxia. HbCO levels are not well correlated with clinical signs of toxicity, likely because CO interacts with multiple proteins in addition to hemoglobin.
- 68.(E). At higher exposure levels, patients can develop mental status changes, confusion, ataxia, syncope, tachycardia, and tachypnea. Severe poisoning is manifested by coma, seizures, myocardial ischemia, acidosis, cardiovascular collapse, and potentially death. On exam, patients might have cherry-red skin.
- 69.(B). In ambient air, the average half-life of HbCO is 4-6 hr. This is dramatically reduced to 60-90 min by providing 100% oxygen at normal atmospheric pressures via a non-rebreather facemask. Severely poisoned patients might benefit from hyperbaric oxygen (HBO), which decreases the half-life of HbCO to 20-30 minutes.
- 70.(B). Propranolol may cause QRS prolongation.
- 71.(A). The severity of the corrosive injury depends on the pH and concentration of the product as well as the length of contact time with the product. Agents with a pH of <2 or >12 are most likely to produce significant injury.
- 72.(E). A commonly used mnemonic for the symptoms of cholinergic excess at muscarinic receptors is DUMBBELS, which stands for diarrhea/defecation, urination, miosis, bronchorrhea/bronchospasm, bradycardia, emesis, lacrimation, and salivation.
- 73.(B). Hypertension.
- 74.(C). Atropine dosing is primarily targeted to drying the respiratory secretions.

- 75.(B). The serum digoxin level should be assessed at least 6 hr after ingestion and carefully interpreted in the setting of clinical symptoms because the digoxin level alone does not entirely reflect the severity of intoxication. In acute ingestions, serum potassium is an independent marker of morbidity and mortality, with levels >5.5 mEq/L predicting poor outcomes. In chronic toxicity, serum potassium is less useful as a prognostic marker and may be altered due to concomitant use of diuretics.
- 76.(E). Calcium channel blockers may cause hyperglycemia.
- 77.(E). Sodium bicarbonate is antidote for tricyclic antidepressants.
- 78.(A). Numerous drug interactions affect plasma digoxin concentrations. Medications known to increase serum digoxin concentrations include the macrolides, erythromycin and clarithromycin, spironolactone, verapamil, amiodarone, and itraconazole.
- 79.(B). Clinically significant hypotension or other cardiovascular instability.
- 80.(E). The severity of an exposure is related to the amount of elemental iron ingested. Ferrous sulfate contains 20% elemental iron, ferrous gluconate 12%, and ferrous fumarate 33%. Multivitamin preparations and children's vitamins rarely contain enough elemental iron to cause significant toxicity.
- 81.(D). Pediatric patients who ingest >40 mg/kg of elemental iron should be referred to medical care for evaluation, although moderate to severe toxicity is typically seen with ingestions of >60 mg/kg.
- 82.(B). The 2nd stage, 6-24 hr after ingestion, is often referred to as the "quiescent phase," as GI symptoms typically have resolved. However, careful clinical exam can reveal subtle signs of hypoperfusion, including tachycardia, pallor, and fatigue.
- 83.(B). The other distracters may cause erythema.
- 84.(D).
- 85.(B). Sinus tachycardia is the most common cardiovascular manifestation of toxicity; however, patients can develop widening of the QRS complex, premature ventricular contractions, and ventricular arrhythmias. Refractory hypotension is a poor prognostic indicator and is the most common cause of death in TCA overdose.
- 86.(D). Refractory hypotension is a poor prognostic indicator and is the most common cause of death in TCA overdose.
- 87.(B). Children who remain completely asymptomatic with normal serial ECGs may be candidates for discharge after 6 hr of close observation.
- 88.(C). Probiotics also indicated in *Clostridium difficile*-associated diarrhea.
- 89.(A). Tea tree oil --- Anti-bacterial (acne remedies), pediculicide (lice), and Aloe vera -
-- Mild burns.

1. The **MOST** common reason for a sick child visit is
 - A. fever
 - B. altered mental status
 - C. vomiting
 - D. respiratory distress
 - E. abdominal pain

2. In pediatrics medical emergencies the survival rate can increase with good neurological outcome if rapid, effective, cardiopulmonary resuscitation (CPR) is done. The survival rate can reach up to
 - A. 10%
 - B. 30%
 - C. 50%
 - D. 70%
 - E. 90%

3. The first response to unwitnessed unresponsive infant is to
 - A. activate emergency response system
 - B. check pulse
 - C. start rescue breathing
 - D. start chest compression
 - E. do endotracheal intubation

4. The features of high quality CPR include all the following **EXCEPT**
 - A. rate at least 100/min
 - B. compression depth of 2/3 anterior-posterior diameter of the chest
 - C. allow complete chest recoil after each compression
 - D. minimize interruptions in chest compressions
 - E. avoid excessive ventilation

5. After activation of emergency medical system, the next action to unresponsive child is to
 - A. assess the air ways
 - B. check pulse
 - C. give rescue breathings
 - D. start chest compression

- E. attach automated external defibrillator (AED) to find shakable rhythm
6. A 12-month-old infant found unresponsive in kindergarten, you were there as health visitor, you asked for activation of EMS and to bring a nearby automated external defibrillator (AED) machine, you check the pulse; it was 60/min. Your next response is to
- A. open the air ways
 - B. give one breath every 3 seconds
 - C. begin cycles of 30 compressions with 2 breaths
 - D. begin a cycles of 15 compressions with 2 breaths
 - E. attach and use AED
7. The correct statement regarding the lower limit of systolic blood pressure for a neonate is
- A. $\geq 40 \text{ mm Hg} + (2 \times \text{age})$
 - B. $\geq 50 \text{ mm Hg} + (2 \times \text{age})$
 - C. $\geq 60 \text{ mm Hg} + (2 \times \text{age})$
 - D. $\geq 70 \text{ mm Hg} + (2 \times \text{age})$
 - E. $\geq 80 \text{ mm Hg} + (2 \times \text{age})$
8. The **MOST** common precipitating event for cardiac instability in infants and children is
- A. electrolyte disturbances
 - B. trauma
 - C. respiratory insufficiency
 - D. poisoning
 - E. myocarditis
9. Regarding Glasgow Coma scale in pediatrics, all the following are true **EXCEPT**
- A. in modified type it uses 15 score points
 - B. it has 3 components
 - C. valid as a prognostic scoring system
 - D. score ≤ 8 require aggressive management
 - E. verbal response component has 5 possible points
10. In pediatrics advanced life support (PALS) curriculum, regarding the format of ABCDE, all the following are true **EXCEPT**
- A. A refers to assessment of airways
 - B. B refers to assessment of breathing
 - C. C refers to assessment of circulation
 - D. D refers to assessment of dehydration
 - E. E refers to assessment of unidentified injuries

11. In secondary assessment of critically ill child; all the following are true **EXCEPT**
- A. interviewing a witness by medical personnel that involved in resuscitation
 - B. taking focused history
 - C. performing a focused medical examination
 - D. enquire about allergies and medications
 - E. enquire about timing of the last meal

12. All the following are part of tertiary assessment of critically ill child **EXCEPT**
- A. coagulation profile
 - B. renal function
 - C. echocardiography
 - D. arterial and venous catheters
 - E. lumbar puncture

13. A 2-year-old toddler was playing with candies and sweets in playroom, he developed an attack of wheezing and shortness of breath, the parents brought the child to emergency room, RR was 68/min with subcostal retractions, and O₂ saturation was 92%.

Of the following, the **MOST** proper next action is to

- A. arrange for urgent bronchoscopy
- B. arrange for bedside CXR
- C. arrange for CT chest
- D. encourage coughing
- E. reassure the parents

14. You received a call from your neighbor, he had a 12-month-old baby boy who suddenly chocked with a candy. When you arrive you find the baby unconscious.

Of the following, the **NEXT** proper action is to

- A. open airway and give rescue breathing
- B. give 5 back blows
- C. give 5 chest thrusts
- D. a series of 5 back blows and chest thrusts
- E. perform endotracheal intubation

15. All the following are absolute indications for endotracheal intubation **EXCEPT**
- A. inability to protect the airway against aspiration
 - B. failing to maintain adequate oxygenation
 - C. complete airway obstruction
 - D. failing to control blood carbon dioxide levels
 - E. paralysis is required for a procedure

16. All the following factors are attributing in definition of clinically significant bradycardia **EXCEPT**

- A. heart rate
- B. temperature
- C. perfusion
- D. mental status
- E. blood pressure

17. The **MOST** common pre-arrest rhythms in young children is

- A. bradyarrhythmia
- B. atrial flutter
- C. ventricular fibrillation
- D. supraventricular arrhythmia with WPW syndrome
- E. atrioventricular block

18. You are evaluating a 3-year-old conscious child with bradycardia and poor perfusion in the emergency department.

Of the following, the **MOST** important first step in treatment is

- A. maintaining adequate perfusion by chest compression
- B. maintaining the airways and assisted breathing
- C. treating precipitating factors as hypoglycemia
- D. epinephrine therapy
- E. atropine therapy

19. Atropine is used in the treatment of bradyarrhythmia that is associated with

- A. raised intracranial pressure
- B. hypoxia
- C. hypothermia
- D. tension pneumothorax
- E. hyperkalemia

20. Factors in favour of SVT versus sinus tachycardia in pediatrics include the following **EXCEPT**

- A. abrupt history
- B. polymorphic P wave
- C. absence of P wave
- D. poor perfusion
- E. constant RR

21. You were engaged in CPR for 3-year-old child found unresponsive in the ward. You were performing cycles of 15 chest compressions and 2 breathings alternatively with

another medical personal. After 2 min of resuscitation you evaluate the pulse which shows PEA (pulseless electrical activity) status. The AED already is attached.

Of the following, the correct **NEXT** action is to

- A. give a shock of 0.5 joule/kg
- B. give a shock of 1 joule/kg
- C. give a shock of 2 joules/kg
- D. give adrenaline 0.01mg/kg iv
- E. give adrenaline 0.1mg/kg iv

22. You were engaged in CPR for 3-year-old child found unresponsive in the ward. You were performing cycles of 15 chest compressions and 2 breathings alternatively with another medical personal. After 2 min of resuscitation the assistant told you that the pulse status is shockable. This means that the pulse is in status of

- A. pulseless electrical activity PEA
- B. asystole
- C. VT
- D. AF
- E. SVT

23. The perfusion pressure of brain (cerebral perfusion pressure) equals to

- A. intracranial pressure
- B. systolic blood pressure minus intracranial pressure
- C. mean blood pressure minus Intracranial pressure
- D. mean blood pressure
- E. mean blood pressure plus CSF pressure

24. All the following are recognized brain herniation due to increase intracranial pressure **EXCEPT**

- A. cingulate
- B. cerebral tonsillar
- C. transcalvrial
- D. uncal
- E. upward cerebellar

25. A 6-year-old male child brought to emergency department after been hit by a car while riding his bike. GCS was 6 and brain CT was normal. However, immediate MRI demonstrates multiple areas of punctate hemorrhages consistent with diffuse axonal injury. Regarding the classification of traumatic brain injury (TBI), the child in this scenario has

- A. mild TBI
- B. moderate TBI
- C. severe TBI

- D. pure axonal brain injury
- E. parenchymal brain hemorrhage

26. All the following are signs of increased intracranial pressure (ICP) and impending brain herniation **EXCEPT**

- A. pupillary dilation
- B. 4th cranial nerve palsy
- C. systemic hypertension
- D. bradycardia
- E. extensor posturing

27. All the following are regarded as first tier therapy of increased intracranial pressure **EXCEPT**

- A. intubation
- B. controlled mechanical ventilation
- C. head of bed elevation
- D. sedation and analgesia
- E. barbiturate infusion

28. All the following are important to diagnose brain death status in a child **EXCEPT**

- A. deep coma of unknown cause
- B. apnea
- C. absence of brainstem reflexes
- D. absence of motor response
- E. absence of confounding factors

29. All the following are red flags in evaluating a patient with syncope **EXCEPT**

- A. syncope with exercise
- B. family history of syncope
- C. presyncopal feeling of light headedness
- D. history of Kawasaki disease
- E. injury with syncope

30. An 11-year-old adolescent boy came to your clinic with a complaint of been collapsed while bathing with hot water after he felt dizzy. Father confirms that the boy was extremely pale when founded. He added that he took few minutes to recover, also he had past history of similar condition one month ago when he was urinating.

Of the following, the **MOST** likely explanation for this condition is

- A. long QT syndrome
- B. hypertrophic cardiomyopathy
- C. neurocardiogenic syncope
- D. drug abuse

E. seizure disorder

31. Of the following, the mandatory test/study for all patients presenting for the first time with syncope is

- A. ECG
- B. EEG
- C. echocardiography
- D. holter monitoring
- E. complete blood count

32. Ductus-dependent congenital heart lesions is a type of

- A. distributive shock
- B. obstructive shock
- C. cardiogenic shock
- D. hypovolemic shock
- E. septic shock

33. You are treating an 8-month-old baby in intensive care unit with cold shock. In the first 15 min he received adequate fluid therapy and a colleague had started dopamine for him. The baby condition is still unresponsive.

Of the following, the **BEST** option now is to start

- A. epinephrine
- B. hydrocortisone
- C. norepinephrine
- D. terlipressin
- E. angiotensin

34. Septic shock usually shows a combination of hypovolemic and distributive shock in addition to

- A. obstructive shock
- B. cardiogenic shock
- C. acute respiratory distress syndrome
- D. disseminated intravascular coagulation
- E. decreased steroid synthesis

35. The percentage of Fio_2 that can be delivered via the nasal cannula is up to

- A. 30%
- B. 40%
- C. 50%
- D. 60%
- E. 80%

36. The percentage of Fio_2 that can be delivered via a mask is up to
- A. 35%
 - B. 45%
 - C. 65%
 - D. 75%
 - E. 85%
37. Rapid and deep breathing without other signs of respiratory distress may be caused by the following **EXCEPT**
- A. diabetic ketoacidosis
 - B. renal tubular acidosis
 - C. encephalitis
 - D. CNS stimulants
 - E. heart failure
38. All the following are controlled mode of mechanical ventilation **EXCEPT**
- A. pressure support ventilation (PSV)
 - B. conventional mechanical ventilation (CMV)
 - C. intermittent mechanical ventilation (IMV)
 - D. synchronized Intermittent mechanical ventilation (SIMV)
 - E. synchronized Intermittent positive pressure ventilation (SIPPV)
39. A 2-hour-old fullterm baby delivered to a mother with gestational diabetes by CS, he developed an increasing respiratory distress and grunting with blood gases values indicative of respiratory failure.
- Of the following, the **BEST** mode to be used in mechanical ventilation is
- A. intermittent mechanical ventilation (IMV)
 - B. synchronized Intermittent mechanical ventilation (SIMV)
 - C. synchronized Intermittent positive pressure ventilation (SIPPV)
 - D. pressure-regulated volume control (PRVC)
 - E. pressure support ventilation (PSV)
40. In mechanical ventilation giving adequate PEEP can mainly reduces
- A. volutrauma
 - B. barotrauma
 - C. oxytrauma
 - D. pneumonia
 - E. post-extubation airway obstruction
41. All the following are recognized complications of mechanical ventilation which can result from use of high tidal volume and/or inspiratory pressure **EXCEPT**
- A. volutrauma

- B. barotrauma
- C. oxytrauma
- D. decrease surfactant production
- E. atelectotrauma

42. You are ventilating a fullterm baby with meconium aspiration syndrome, he was fine on the machine, suddenly the monitor shows severe drop in O₂ saturation. You checked the ventilator; it was working well with its connections.

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

- A. O₂ source supply
- B. endotracheal tube blockade
- C. self extubation
- D. low inspiratory pressure value
- E. low PEEP value

43. A 32-week-preterm baby on mechanical ventilation, he is recovering from RDS, his blood gas is satisfactory, and you reduced the ventilatory settings in process for weaning.

Of the following, the **MOST** valuable indicator that the baby is ready for extubation is

- A. frequency of respiration 35
- B. fraction of inspired O₂ 0.55
- C. peak expiratory end pressure 6
- D. inspiratory pressure 8
- E. inspiratory time 0.4 sec

44. Prior to extubation and in order to reduce airway narrowing after extubation it is advised to use

- A. dexamethasone IV
- B. betamethasone IV
- C. inhaled budesonide
- D. nebulized budesonide
- E. hydrocortisone IV

45. The **MOST** effective strategy to reduce ventilator-associated pneumonia (VAP) is

- A. reducing the period of ventilation
- B. elevation of the head of the bed to 30 degrees
- C. use of a protocol for oral decontamination
- D. liberal use of effective antibiotics
- E. frequent endotracheal tube suction

46. In surviving drowning patients; expecting brain edema can occur within

- A. 5 min

- B. 15 min
- C. 30 min
- D. 60 min
- E. several hours

47. Of the following, the **MOST** common factor that increases the likelihood of drowning in adolescent (USA) is

- A. epilepsy
- B. long QT syndrome
- C. alcohol
- D. myocarditis
- E. external trauma

48. All the following mechanisms are contributing in pathophysiology of drowning **EXCEPT**

- A. laryngospasm
- B. surfactant washout
- C. anoxic injury
- D. pulmonary aspiration
- E. circulatory overload

49. All the following are component of cold water shock resulted from immersion in cold water **EXCEPT**

- A. hypoventilation
- B. decrease breath holding ability
- C. hypertension
- D. ectopics
- E. SVT

50. You are treating a 6-year-old child who survived drowning after prolonged CPR, the mother is asking about the possibility of full neurological recovery of her child.

Of the following, the **BEST** response is that it is possible if he regained full consciousness within

- A. few hours
- B. 12 hours
- C. 24 hours
- D. 48 hours
- E. 72 hours

1.(A). Fever is the most common reason for a sick child visit. Most fevers are the result of self-limited viral infections. However, pediatricians need to be aware of the age-dependent potential for serious bacterial infections (e.g., urinary tract infections, sepsis, meningitis, pneumonia, dysentery, osteoarticular infection).

2.(B).

3.(A). A child found unresponsive from an unwitnessed collapse should be approached with a gentle touch and the verbal question, "Are you OK?" If there is no response, the caregiver should immediately shout for help and send someone to both activate the emergency response system (EMS) and locate an automated external defibrillator (AED).

4.(B). The Depth of chest compression should be at least 1/3 anterior-posterior diameter of chest, about 1 1/2 inches (4 cm) in infants and 2 inches (5 cm) in children.

5.(B). After activation of EMS the next step is to check for pulse and find definite pulse within 10 seconds.

6.(B). In summary, the plan of action for an unwitnessed infant found unresponsive as follows

- * Activate EMS and ask for AED machine.

- * Check for definite pulse, if found give one breath every 3 seconds.

- * Recheck pulse if it is < 60/min add chest compressions.

- * In case of no pulse found start directly chest compressions, for one rescuer give 30 compressions with 2 breaths and for 2 rescuers give 15 compressions and 2 breaths, reassess every 2 minutes.

- * Check for the rhythm if it is shockable or not, if it is shockable give one shock and continue CPR.

- * Continue CPR until health care provider comes or the victim moves.

7.(C). For a neonate ≥ 60 mm Hg, for an infant it is ≥ 70 mm Hg and for an older child it is ≥ 90 mm Hg, definitely the adult's value is inapplicable in children.

8.(C).

9.(C). Although the GCS has not been validated as a prognostic scoring system for infants and young children as it has been in adults, it is commonly used in the assessment of pediatric patients with an altered level of consciousness. The GCS is the most widely used method of evaluating a child's neurologic function and has 3 components. Individual scores for eye opening, verbal response, and motor response are added together, with a maximum of 15 points. Patients with a GCS score ≤ 8 require aggressive management, including stabilization of the airway and breathing with

endotracheal intubation and mechanical ventilation, respectively, and, if indicated, placement of an intracranial pressure monitoring device.

10.**(D).** The D means Disability which refers to assessment of the child's neurologic function in terms of the level of consciousness and cortical function. It can be assessed quickly by checking the pupillary light response and using Glasgow Coma scale, so the response D is only partially true. The causes of decreased level of consciousness in children are numerous and include conditions as diverse as respiratory failure with hypoxia or hypercarbia, hypoglycemia, poisonings or drug overdose, trauma, seizures, infection, and shock. All that system is represent the primary assessment of critically ill child.

11.**(A).** The components of a secondary assessment include a focused history and focused physical exam. The history should be targeted to information that could explain cardiorespiratory or neurologic dysfunction and should take the form of a SAMPLE history (Signs/symptoms, Allergies, Medications, past medical history, timing of Last meal, and Events leading to this situation). Medical personnel not engaged in resuscitative efforts can be dispatched to elicit history from witnesses or relatives. The physical exam during the secondary assessment is a thorough head-to-toe exam, although the severity of the child's illness or injury could necessitate curtailing portions of the exam or postponing nonessential elements until a later time.

12.**(E).** The tertiary assessment occurs in a hospital setting, where ancillary laboratory and radiographic assessments contribute to a thorough understanding of the child's condition. A basic blood chemistry profile, complete blood count, liver function tests, coagulation studies, and arterial blood gas analyses give fairly broad (but somewhat nonspecific) estimates of renal function, acid-base balance, cardiorespiratory function, and presence or absence of shock. Chest radiographs can be useful to evaluate both the heart and lungs, although more detailed estimates of heart function and cardiac output can be made. Choice E is not part of assessment.

13.**(A).** A history consistent with foreign-body aspiration is considered diagnostic. Any child in the proper setting with the sudden onset of choking, stridor, or wheezing has foreign body aspiration until proven otherwise. The choice A is the best answer as the child start to show signs of respiratory distress with borderline O2 sat. The choice B is the next appropriate action while encourage coughing may be appropriate at home when no respiratory distress is there.

14.**(A).** In unconscious child, the child should be gently placed on the ground, supine, and then the provider should open the airway with the head-tilt/chin-lift maneuver and attempt mouth-to-mouth ventilation. If ventilation is unsuccessful, the airway is repositioned, and ventilation attempted again. If there is still no chest rise, attempts to remove a foreign body are indicated. In an infant <1 yr old, a combination of 5 back blows and 5 chest thrusts is administered, After each cycle of back blows and chest thrusts, the child's mouth should be visually inspected for the presence of the foreign body. If identified within finger's reach, it should be removed with a gentle finger sweep. If no foreign body is visual, ventilation is again attempted. If this is unsuccessful,

the head is repositioned, and ventilation attempted again. If there is no chest rise, the series of back blows and chest thrusts is repeated. For a conscious child >1 yr old, providers should give a series of 5 abdominal thrusts (Heimlich maneuver) with the child standing or sitting; this should occur with the child lying down if unconscious.

15.(C). In settings of known complete airway obstruction, endotracheal intubation should be avoided, and emergency cricothyroidotomy performed instead.

16.(B). A clinically significant bradycardia occurs when the heart rate is slow and there are signs of systemic hypoperfusion (i.e., pallor, altered mental status, hypotension, and acidosis). Symptomatic bradycardia occurs most often in the setting of hypoxia but can also be caused by hypoglycemia, hypocalcemia, other electrolyte abnormalities, and intracranial hypertension.

17.(A).

18.(B). Alleviating respiratory compromise and maintaining adequate breathing and oxygenation may correct clinically significant bradycardia. Start monitoring and obtain IV access. If bradycardia persists it is the time for chest compressions and if continues you need to start drug therapy. During resuscitation correct possible underlying causes referred collectively as the **6 Hs** (hypoxia, hypovolemia, hydrogen ions [acidosis], hypokalemia or hyperkalemia, hypoglycemia, hypothermia), and **5 Ts** (toxins, tamponade, tension pneumothorax, thrombosis [in either the pulmonary or cardiac circulations], and trauma [causing hypovolemia, intracranial hypertension, cardiac compromise or tamponade])

19.(A). Atropine is indicated in the case of increased vagal tone (e.g., in the setting of head injury with raised intracranial pressure) or primary atrioventricular block.

20.(D). In sinus tachycardia, the history and onset are consistent with a known cause of tachycardia, such as fever or dehydration and P waves are consistently present, are of normal morphology, and occur at a rate that varies somewhat. In SVT, onset is often abrupt without prodrome and P waves are absent or polymorphic, and when present, their rate is often fairly steady at or above 220 beats/min. Poor perfusion can present in either causes as in dehydration with tachycardia and in late stage of SVT.

21.(D). Pulseless electrical activity (PEA) is a clinical condition characterized by unresponsiveness and lack of palpable pulse in the presence of organized cardiac electrical activity. In this situation (and in asystole) adrenaline is indicated in cycles of 3-5 min, considering an advanced airways and reevaluation for the pulse status; if it became shockable give shock of 2 joules/kg. You need to look also for correctable causes (the 6H and 5T). If it became normal that goes for (ROSC) post-cardiac arrest care.

22.(C). Shockable rhythms mean either VF or VT.

23.(C). The intracranial pressure (ICP) is derived from the volume of its components and the bony compliance. The perfusion pressure of the brain (cerebral perfusion pressure [CPP]) is equal to the pressure of blood entering the cranium (mean arterial pressure) minus the ICP.

24.(B). In fact there are 6 types of brain herniation in addition to what have been mentioned in the question plus cerebellar tonsils and cerebral herniation through foramen magnum.

25.(C). The key for severity of TBI is GCS score, patient with GCS of 3-8 regarded as severe type while those between 9-12 regarded as moderate.

26.(B). The development of increased ICP with impending herniation may be heralded by new onset or worsening headache, depressed level of consciousness, vital sign changes (hypertension, bradycardia, irregular respirations), and signs of 6th (lateral rectus palsy) or 3rd (anisocoria [dilated pupil], ptosis, down-and-out position of globe as a result of rectus muscle palsies) cranial nerve compression.

27.(E). The first tier therapy also include neuromuscular blockade, CSF drainage, and the use of osmolar agents as hypertonic saline (3%) and mannitol. 2nd tier therapy include barbiturate infusion, decompressive craniectomy, mild hypothermia hyperventilation and lumbar CSF drainage.

28.(A).

29.(C). Typical prodromal symptoms prior to syncope include light headedness, dizziness, and nausea, sweating, and feeling hot or cold. Patients may report visual field changes and rushing in their ears.

30.(C). Typically, the patient with syncope will have been standing for a period of time, often on a hot day, or has gotten up suddenly from sleep or resting in a supine position. Occurrence in the shower is common, presumably caused by standing and vasodilation caused by hot water. For boys, the occurrence while urinating while standing is sometimes reported. The occurrence of syncope in girls while sitting or standing and having their hair brushed is common.

31.(A). All patients presenting with a first episode of syncope should have an electrocardiogram obtained, looking primarily for QT interval prolongation, preexcitation, ventricular hypertrophy, T-wave abnormalities, and conduction abnormalities. Other tests may be needed depending on the results of the initial evaluation.

32.(B). It represent a decreased in cardiac output secondary to direct impediment to right- or left-heart outflow or restriction of all cardiac chambers.

33.(A). After 15 min of treating shock, inotropes like dopamin need to be started, if the shock still unresponsive epinephrine for cold and norepinephrine for warm shock should be titrated. Hydrocortisone is considered after 60 min (Catecholamine resistant shock). Other options are used in warm shock and usually after other options are exhausted.

34.(B). Options C and D may be part of pathophysiology of septic shock, it is not a type.

35.(B). The typical FiO₂ value using this method is between 23% and 40%, and can be measured with following formula

FiO₂ %O₂ delivered = 21 %+(nasal cannula flow (L/min) X3), and the flow rate should be <5 L/min.

36.(C). Oxygen flow rates vary from 5-10 L/min, yielding typical Fio_2 values between 0.30 and 0.65.

37.(E). Causes rapid shallow breathing.

38.(A). Except the option A all other forms of mechanical ventilation variables are controlled either fully by the machine as in options B and C which need to paralyze the patient to get the synchrony or partially as in options D and E where there is patient trigger by an effort to respiration followed by the machine active work to do the preset ventilation values; ventilator–patient asynchrony can occur in this mode also because tidal volume, inflation pressure, and inspiratory time are determined by the ventilator alone. PSV allows the patient to control as much of the rate, tidal volume, and inspiratory time as much as possible. It is considered a gentler form of mechanical ventilation and is designed for patients with relatively minor lung disease or for those with neuromuscular weakness.

39.(D). IMV is not the choice because you need to paralyze the patient; options B and C both are good choice as they provide synchrony with the patient and can achieve the goal of ventilation by either controlling the pressure or volume values but in presence the option D in your ventilator, making it the best because it will reduces both baro and volutrauma.

40.(C). It can also reduce atelectotrauma.

41.(E). It can result from low PEEP.

42.(B). All the given options are true causes for sudden drop of O_2 . The possible causes of desaturating baby on ventilator are; Oxygen supply, machine and it is connections, settings, tube block, self extubation and the patient himself. In the given scenario, the most likely cause is B because of meconium aspiration. The best action to such scenario is to disconnect the baby from the machine and use Ambu bagging gently to check the air entry and rise of O_2 saturation. If no rise you need to change the tube. In case of unequal air entry, pneumothorax is another possibility.

43.(D). In absence of tachypnea, increased work of breathing, hypoxemia, hypercapnia, acidosis, diaphoresis, tachycardia, and hypotension and when the ventilator rate is <5 breaths/min or (between 5-10 cm H_2O in pressure controlled devices) indicate that the contribution of the ventilation to minute ventilation is minimal and the baby is ready to be extubated.

44.(A). Administration of intravenous dexamethasone 0.5 mg/kg every 6 hr for 4 doses prior to extubation has been shown to minimize the incidence of postextubation airway obstruction. In patients in whom postextubation airway obstruction develops, the need for re-intubation may be obviated by administration of nebulized racemic epinephrine and heliox.

45.(A). The options B and C are a true technique, but the most effective strategy to minimize VAP is regular assessment of extubation readiness and liberation from mechanical ventilation as soon as clinically possible.

46.(E). Several hours after cardiopulmonary arrest, cerebral edema may occur, although the mechanism is not entirely clear.

47.(C). The use of alcohol and drugs greatly increases the risk of drowning in teenagers and adults who die, 30-40% have positive blood alcohol level.

48.(E).

49.(A). Immersion in cold water has immediate respiratory and cardiovascular effects. Victims experience cold water shock, a dynamic series of cardiorespiratory physiologic responses that can cause drowning. In adults, immersion in icy water results in intense involuntary reflex hyperventilation and to a decrease in breath-holding ability to <10 sec, which leads to fluid aspiration. Severe bradycardia, the diving reflex, occurs in adults but is transient and rapidly followed by supraventricular and ectopic tachycardia and hypertension. There is no evidence that the diving reflex has any protective effect.

50.(E). Neurologic examination and progression during the 1st 24-72 hr are the best prognosticators of long-term CNS outcome. Children, who regain consciousness within 48-72 hr, even after prolonged resuscitation, are unlikely to have serious neurologic sequelae.

1. When an individual is at-risk genotype and not clinically express the condition; this phenomena is called
 - A. genetic imprinting
 - B. genetic anticipation
 - C. uniparental diasomy
 - D. non expressiveness
 - E. non penetrance

2. The genetic testing in an asymptomatic child with positive family history of Huntington chorea is called
 - A. predispositional testing
 - B. predictive testing
 - C. diagnostic testing
 - D. pharmacogenetic testing
 - E. linkage testing

3. A 5-year-old asymptomatic male child with strong positive family history of Huntington chorea, mother is highly concern about her child possible genetic affection, she asked for predictive genetic testing.
Of the following, the **MOST** appropriate explanation is that
 - A. whatever the result it will not affect the disease process
 - B. this is an age dependent type of genetic disorder and may not manifest ever
 - C. leave the decision to the child when he is grown up
 - D. it may lead to discrimination
 - E. we should weigh the benefits VS risks of testing

4. Predispositional genetic testing is more useful in
 - A. numerical chromosomal disorders
 - B. multifactorial disorders
 - C. single gene disorders
 - D. structural chromosomal disorders
 - E. pre arrangement for gene therapy

5. A 4-year-old female child, a known case of ALL, she is developing an increasing toxicity to methotrexate therapy.
Of the following, the **MOST** useful test is

- A. homocysteine level
 - B. prothrombin test
 - C. factor V leiden
 - D. MTHFR DNA testing
 - E. folate level
6. The following are indications for genetic counseling **EXCEPT**
- A. paternal age > 50 years
 - B. maternal age > 45 years
 - C. family history of intellectual disability
 - D. consanguinity
 - E. infertility
7. The following statements are true about genetic counseling **EXCEPT**
- A. constructing family pedigree up to the third degree
 - B. collecting health files about affected family members
 - C. performing appropriate genetic testing
 - D. refer to support groups
 - E. talking about rewarding gene therapy
8. Enzyme replacement therapy are available for the following disorders **EXCEPT**
- A. Wolman disease
 - B. Gaucher disease
 - C. Fabry disease
 - D. Mucopolysaccharidosis type VI
 - E. Pompe disease
9. DiGeorge syndrome is an example of
- A. contiguous gene disorders
 - B. single gene disorders
 - C. mitochondrial inheritance disorders
 - D. numeric chromosomal disorders
 - E. imprinting disorders
10. The phenomena of changing phenotypes (clinical presentations) associated with the same single-gene disorder is called
- A. variable expressiveness
 - B. variable penetrance
 - C. false positive
 - D. false negative
 - E. validity

11. The **MOST** important screening tool for genetic disorders is
- A. genetic study
 - B. family history
 - C. karyotyping
 - D. prenatal diagnosis
 - E. preimplantation genetic screen
12. One of the following is explained by digenetic inheritance
- A. vitamin D dependent rickets
 - B. retinitis pigmentosa
 - C. biotinidase deficiency
 - A. congenital lactic acidosis
 - B. glycogen storage disease type VI
13. Parent-to-child transmission (vertical transmission) is a character of autosomal dominant inheritance, for many patients with an autosomal dominant disorder there is no history of an affected family member.
- All the following are true explanations **EXCEPT**
- A. variable expressiveness
 - B. new mutation
 - C. somatic mutations (mosaicism)
 - D. digenetic inheritance
 - E. incomplete penetrance
14. The following are characteristic feature of autosomal dominant inheritance **EXCEPT**
- A. vertical transmission
 - B. male to male transmission
 - C. males and females are equally affected
 - D. skipped generation
 - E. both parents carried the affected gene
15. All the following are characteristic features of autosomal recessive inheritance **EXCEPT**
- A. vertical transmission
 - B. no affected family members in other generations
 - C. males and females are equally affected
 - D. consanguinity plays important role
 - E. 25% recurrence risk
16. The risk of a genetic disorder for the offspring of a first-cousin marriage (consanguinity marriage) is
- A. 3-5%

- B. 6-8%
 - C. 9-11%
 - D. 12-14%
 - E. 15-17%
17. Pseudodominant inheritance happens when
- A. an x-linked disorder appears as autosomal dominant
 - B. an autosomal dominant appears as recessive trait
 - C. an autosomal recessive disorder appears as autosomal dominant
 - D. skipped generations appears
 - E. an autosomal dominant appears as x-linked disorder
18. In genetic disorders, male to male transmission occurs in Y- linked inheritance and in
- A. X- linked inheritance
 - B. pseudodominant inheritance
 - C. digenic inheritance
 - D. autosomal dominant disorder
 - E. pseudogenetic inheritance
19. Females sometimes manifest signs of X- linked recessive disorders. All the following are possible explanations **EXCEPT**
- A. homozygosity for an X-linked trait
 - B. 45,XO female
 - C. 46,XY female
 - D. uniparental disomy
 - E. nonrandom X-inactivation
20. All the following are features of mitochondrial inheritance **EXCEPT**
- A. non-traditional inheritance
 - B. maternal inheritance
 - C. male to offspring transmission
 - D. both sexes are affected
 - E. mitochondrial DNA point mutations
21. The organ that can be spared in mitochondrial disorders is
- A. brain
 - B. eye
 - C. small intestine
 - D. liver
 - E. muscle

22. The following represent a non-traditional inheritance **EXCEPT**
- A. mitochondrial disorders
 - B. triplet repeat expansion diseases
 - C. imprinting defects
 - D. familial clustering
 - E. uniparental disomy
23. The following are a triplet repeat expansion disorders **EXCEPT**
- A. fragile X syndrome
 - B. myotonic dystrophy
 - C. Huntington disease
 - D. spinocerebellar ataxias
 - E. Kearns-Sayre syndrome
24. The observation of increasing severity of the disease and early age of onset in subsequent generation is called
- A. genetic imprinting
 - B. genetic anticipation
 - C. uniparental diasomy
 - D. non expressiveness
 - E. non penetrance
25. The observation of significantly increasing number of imprinting disorders as Beckwith-Wiedemann and Angelman syndrome are **MOST** likely due to
- A. new methods of prenatal diagnosis (amniocentesis)
 - B. new technology of gene therapy
 - C. assisted reproductive technology
 - D. increase radiation exposure
 - E. high pressure electricity chains in large cities
26. The following genetic scenarios are possible to explain Angelman syndrome **EXCEPT**
- A. inheriting both chromosomes 15 from the father
 - B. deletion of the maternal 15q12
 - C. mutation in an imprinted gene
 - D. mutation in the imprinting center
 - E. genetic anticipation
27. The **MOST** common genetic cause of Prader-Willi syndrome is
- A. inheriting both chromosomes 15 from the mother
 - B. deletion of the maternal 15q12
 - C. deletion of the paternal 15q12
 - D. mutation in an imprinted gene

- E. mutation in the imprinting center

28. The recurrence risk of multifactorial inheritance are characterized by all the following **EXCEPT**

- A. related to the incidence of the disease in community
- B. similar among all 1st-degree relatives
- C. no sex predilection
- D. increased when multiple family members are affected
- E. greater when the disorder is more severe

29. Chromosomal analysis is indicated in all the following **EXCEPT**

- A. recurrent premature deliveries
- B. postnatal problems in growth and development
- C. unexplained intellectual disability
- D. primary amenorrhea
- E. recurrent miscarriages (≥ 3)

30. Chromosomal abnormalities in live births constitute around

- A. 1-2%
- B. 3-4%
- C. 5-6%
- D. 7-8%
- E. 9-10%

31. A 17-year-old adolescent female with trisomy 21 became pregnant, the chance of affection of her offspring with the same problem is

- A. Not possible
- B. 25%
- C. 50%
- D. 75%
- E. 100%

32. All the following are recognized gastrointestinal features of Down syndrome in neonatal period **EXCEPT**

- A. annular pancreas
- B. tracheoesophageal fistula
- C. diaphragmatic hernia
- D. imperforate anus
- E. neonatal cholestasis

33. The **MOST** important reason for chromosomal analysis in persons suspected of having Down syndrome is to

- A. confirm clinical diagnosis
- B. detect mosaicism
- C. reassure the family
- D. detect translocations
- E. detect rare cases of ring chromosome

34. A chromosomal study of 22-year-old mother of a baby with Down syndrome t (14; 21) shows that; she is the carrier for the translocation. You explained that the recurrence rate will be approximately

- A. 2-4%
- B. 5-7%
- C. 8-10%
- D. 11-13%
- E. 33%

35. A highly educated young couple has their first baby with Down syndrome; they are asking you about his development.

Of the following, the **MOST** appropriate answer is

- A. programs of stimulation will achieve eventual normal development
- B. motor development is the mostly affected domain with fair cognitive delay
- C. developmental delay will be in the initial first few years, but it will normalize later on
- D. developmental delay is universal, all domains of development almost equally affected
- E. developmental delay is universal, social development is relatively spared but he will have problems with expressive language

36. You are explaining the motor development to a mother with a baby with Down syndrome, you told her that there is a wide range of time to achieve his milestone and as example the baby can walk between

- A. 10-18 mo
- B. 12-24 mo
- C. 12-45 mo
- D. 15-50 mo
- E. 18-60 mo

37. A 5-year-old child with Down syndrome complaining of intermittent symptoms of torticollis, weakness of the lower limbs and gait disturbances.

Of the following, the **NEXT** important step is to

- A. reassure the family
- B. send for radiological assessment of the neck
- C. send for thyroid antibodies

- D. send for radiological assessment of the airway
- E. send for thyroid function test

38. Quad screen is the four maternal serum tests that should be done for all pregnant women in second trimester as a screening for Down syndrome, which includes the following tests **EXCEPT**

- A. free β -human chorionic gonadotropin
- B. unconjugated estriol
- C. androsterone
- D. inhibin
- E. α -fetoprotein

39. All the following are features of Edwards syndrome (Trisomy 18) **EXCEPT**

- A. low birth weight
- B. closed fists
- C. cardiac malformations
- D. short sternum
- E. sloping forehead

40. All the following are features of Patau syndrome (Trisomy 13) **EXCEPT**

- A. scalp defects
- B. corneal abnormalities
- C. holoprosencephaly
- D. capillary hemangiomas
- E. prominent occiput

41. Robertsonian translocations involve 2 acrocentric chromosomes that fuse near the centromeric region with a subsequent loss of the short arms. It can occur in the following chromosomes **EXCEPT**

- A. 13
- B. 15
- C. 17
- D. 21
- E. 22

42. One of the common chromosomal deletions which can lead to retinoblastoma is

- A. 4p-
- B. 5p-
- C. 9p-
- D. 13q-
- E. 18p-

43. All the following are examples of microdeletion syndromes **EXCEPT**

- A. Williams
- B. Prader-Willi
- C. Angelman
- D. DiGeorge
- E. Wolf-Hirschhorn

44. An 18-month-old female child had been diagnosed as Turner syndrome (45, XO), FISH analysis shows Y chromosome mosaicism.

Of the following, the **MOST** appropriate next step is to

- A. refer to endocrinologist
- B. reassure as the course will be milder
- C. advise for feminizing hormones
- D. arrange for laparoscopic removal of gonads
- E. rear the child as male gender

45. All the following are recognized features of Turner syndrome **EXCEPT**

- A. female phenotype
- B. male phenotype
- C. hypothyroidism
- D. type I diabetes mellitus
- E. gonadal dysgenesis

46. All the following are characteristic features of fragile X chromosome of a 3-year-old male child **EXCEPT**

- A. intellectual disability
- B. autistic behavior
- C. macroorchidism
- D. hyperextensible finger joints
- E. characteristic facial features

1.(E).

2.(B).

3.(C). In fact, all the mentioned explanations about the disease are true but it is generally agreed that predictive genetic tests should be performed for children if the results of the test will benefit the medical management of the child. Otherwise, the test should be deferred until the child has an understanding of the risks and benefits of testing and can provide informed consent.

4.(B). In a disease cause by many genes and affected by environmental factors, controlling of environmental factors may reduce disease progression as in cigarette smoking and α 1-antitrypsin deficiency, familial breast cancer and mammography.

5.(D). Knowledge of individual genotypes (pharmacogenetic genetic testing) will guide pharmacologic therapy, allowing customization of choice of drug and dosage to avoid toxicity and provide a therapeutic response. An example of this is testing for polymorphisms within the methylenetetrahydrofolate reductase (MTHFR) gene for susceptibility of potentially increased toxicity to methotrexate antimetabolite therapy for treatment of acute lymphoblastic leukemia.

6.(B). Maternal age $\geq$ 35 is an indication for genetic counseling. Genetic counseling is a communication process in which the genetic contribution to health is explained, along with specific risks of transmission of a trait and options to manage the condition and its inheritance. The counselor is expected to present information in a neutral, nondirective manner and to provide support to the individual and family to cope with decisions that are made.

7.(E). Gene therapy still experimental and rewarding therapy is still a hope.

8.(A).

9.(A).

10.(A).

11.(B). The family history remains the most important screening tool for pediatricians in identifying a patient's risk for developing a wide range of diseases, from multifactorial conditions, such as diabetes and attention-deficit disorder, to single-gene disorders such as sickle cell anemia and cystic fibrosis.

12.(B). Digenic inheritance explains the occurrence of retinitis pigmentosa (RP) in children of parents who each carry a mutation in a different RP-associated gene. Both parents have normal vision, as would be expected, but their offspring who are double heterozygotes—having inherited both mutations—develop RP. Digenic pedigrees can exhibit characteristics of both autosomal dominant (vertical transmission) and autosomal recessive inheritance (1 in 4 recurrence risk).

13.(D). Double heterozygotes

14.(E). Autosomal dominant inheritance is determined by the presence of 1 abnormal gene on 1 of the autosomes (chromosomes 1-22). Autosomal genes exist in pairs, with each parent contributing 1 copy. In an autosomal dominant trait, a change in 1 of the paired genes has an effect on the phenotype.

15.(A). Horizontal transmission is the characteristic feature of autosomal recessive inheritance, the observation of multiple affected members of kindred in the same generation, but no affected family members in other generations.

16.(B). The risk of a genetic disorder for the offspring of a first-cousin marriage (6-8%) is about double the risk in the general population (3-4%).

17.(C). Pseudodominant inheritance refers to the observation of apparent dominant (parent to child) transmission of a known autosomal recessive disorder. This occurs when a homozygous affected individual has a partner who is a heterozygous carrier, and it is most likely to occur for relatively common traits, such as sickle cell anemia.

18.(D).

19.(D).

20.(C). Mitochondrial inheritance represents a non-traditional inheritance (not follow the usual Mendelian inheritance), both sexes can be affected from the affected mitochondrial DNA genome in the ovum (sperm contain few mitochondria which finishes during fertilization), so no male to offspring transmission.

21.(C). The mitochondria are the cell's suppliers of energy, so the organs that are most affected by the presence of abnormal mitochondria are those that have the greatest energy requirements, such as the brain, muscle, heart, and liver. Common manifestations include developmental delay, seizures, cardiac dysfunction, decreased muscle strength and tone, and hearing and vision problems. Examples of mitochondrial disorders include MELAS (*myopathy, encephalopathy, lactic acidosis, and stroke-like episodes*), MERRF (*myoclonic epilepsy associated with ragged red fibers*), and Kearns-Sayre syndrome (*ophthalmoplegia, pigmentary retinopathy, and cardiomyopathy*).

22.(D). The first three represent the non-traditional inheritance; familial clustering represents a form of pseudogenetic transmission, while uniparental disomy is a type of imprinting defects.

23.(E). Represent mitochondrial disorder.

24.(B).

25.(C). Several studies suggest that there may be a small but significantly increased incidence of imprinting disorders, specifically Beckwith-Wiedemann and Angelman syndrome, associated with assisted reproductive technologies such as in vitro fertilization and intracytoplasmic sperm injection. However, the overall incidence of these disorders in children conceived using assisted reproductive technologies is likely to be <1%.

26.(E). The first statement is called uniparental disomy when both copies of a chromosome is inherited from one parent, the second call genetic imprinting when one

gene copy is silenced in one side; either in mother or father resulting in each situation in different syndromes, third and fourth distracters is possible but are uncommon causes.

27.(C). In about 70% of cases missing paternal 15q12 lead to what is called genomic imprinting. The same clinical effect if the child inherited the same chromosome (15) from the mother (25-29%). In Angelman syndrome, paternal UPD of chromosome 15 is rarer and is observed in approximately 5% of the cases (missing the maternal chromosome 15).

28.(C). Some multifactorial inherited disorders have a sex predilection, as indicated by an unequal male: female incidence. Pyloric stenosis, for example, is more common in males, whereas congenital dislocation of the hips is more common in females. Where there is an altered sex ratio, the risk is higher for the relatives of an index case whose gender is less commonly affected than relatives of an index case of the more commonly affected gender. For example, the risk to the son of an affected female with infantile pyloric stenosis is 18%, compared with the 5% risk for the son of an affected male. An affected female presumably has a greater genetic susceptibility, which she can then pass on to her offspring.

29.(A). The specific indications for studies include advanced maternal age (>35 yr) or multiple abnormalities on fetal ultrasound (prenatal testing), multiple congenital anomalies, unexplained growth restriction in the fetus or postnatal problems in growth and development, ambiguous genitalia, unexplained intellectual disability with or without associated anatomic abnormalities, primary amenorrhea or infertility, recurrent miscarriages (≥ 3) or prior history of stillbirths and neonatal deaths, a 1st-degree relative with a known or suspected structural chromosome abnormality, clinical findings consistent with a known anomaly, some malignancies, and chromosome breakage syndromes (e.g., Bloom syndrome, Fanconi anemia).

30.(A). Clinical cytogenetics is the study of chromosomes: their structure, function, inheritance, and abnormalities. Chromosome abnormalities are very common and occur in approximately 1-2% of live births, 5% of stillbirths, and 50% of early fetal losses in the 1st trimester of pregnancy.

31.(C). Most males with Down syndrome are sterile, but some females have been able to reproduce, with a 50% chance of having trisomy 21 pregnancies.

32.(C). It also includes duodenal atresia and Hirschsprung disease.

33.(D). Chromosome analysis is indicated in every person suspected of having Down syndrome. If a translocation is identified, parental chromosome studies must be performed to determine whether one of the parents is a translocation carrier, which carries a high recurrence risk for having another affected child.

34.(B). Translocation (21; 21) carriers have a 100% recurrence risk for a chromosomally abnormal child, and other Robertsonian translocations, such as t(14;21), have a 5-7% recurrence risk when transmitted by females.

35.(E). Developmental delay is universal. Cognitive impairment does not uniformly affect all areas of development. Social development is relatively spared, but children

with Down syndrome have considerable difficulty using expressive language. Understanding these individual developmental strengths will maximize the educational process for children with Down syndrome. Persons with Down syndrome often benefit from programs aimed at stimulation, development, and education. These programs are most effective in addressing social skills that often appear advanced for the intellectual delay.

36.(C).

37.(B). The incidence of atlantoaxial subluxation or instability in Down syndrome is about 10-30%, affected patient should not allowed to participate in high risk sports as include diving starts in swimming, butterfly stroke, diving, pentathlon, high jump, equestrian sports, gymnastics, football, soccer, alpine skinning and warm up exercises placing stress on the head and neck

38.(C). Androsterone is generally considered to be an inactive metabolite of testosterone, and it is not related to quad test.

39.(E). Slopping forehead is a feature of Trisomy 13.

40.(E). Prominent occiput is a feature of Edwards's syndrome.

41.(C). It includes also chromosome number 14.

42.(D).

43.(E). Microdeletions involve loss of small chromosome regions so that the affected individuals can have a distinctive phenotype depending on the number of genes involved. All the mentioned syndromes are common microdeletions and the board candidate should be familial with their phenotypic descriptions and clinical manifestations. Wolf-Hirschhorn syndrome is chromosomal deletion syndrome.

44.(D). Phenotypic females with 45, X/46, XY mosaicism have a 15-30% risk of developing gonadoblastoma. The American Academy of Pediatrics has recommended the use of FISH analysis to look for Y-chromosome mosaicism in all 45, X patients. If Y chromosome material is identified, laparoscopic gonadectomy is recommended.

45.(D). Male phenotype can occur with mosaic type of Turner syndrome, and type II insulin resistance is the true answer.

46.(C). The macroorchidism may not be evident until puberty. The facial features, which include a long face, large ears, and a prominent square jaw, become more obvious with age.

Metabolic Disorders

QUESTIONS

KHALID ALAARJI

1. Inborn errors of metabolism (IEM) are hereditary biochemical disorders caused by single-gene mutations that result in alteration of

- A. chromosomal number or structure
- B. mitochondrial synthesis or function
- C. primary fat structure or the amount of fat synthesized
- D. primary protein structure or the amount of protein synthesized
- E. primary carbohydrate structure or the amount of carbohydrate synthesized

2. Inborn errors of metabolism (IEM) are hereditary biochemical disorders caused by single-gene mutations that encode specific proteins.

The following are the common characteristics of IEM **EXCEPT**

- A. the majority of conditions are inherited as autosomal recessive
- B. the later the appearance of clinical symptoms, the more severe is the disease
- C. early diagnosis is of paramount importance before irreversible organ damage
- D. the affected infant is normal at birth and becomes symptomatic later on in life
- E. there is a variation in the severity of the phenotype based on the gene mutation

3. All the following are widely used modalities for treatment of inborn errors of metabolism (IEM) **EXCEPT**

- A. special diets
- B. gene therapy
- C. peritoneal dialysis
- D. liver transplantation
- E. administration of the deficient metabolite

4. A 28-day-old girl presented with lethargy, poor feeding, and repeated vomiting for last 5 days; IV fluid and empirical antibiotics were started; later she develops repeated seizures not responding to IV calcium, glucose, B6, and anticonvulsant drugs. Septic screen is negative; serum ammonia is elevated with normal anion gap and normal pH. Family history reveals 2 siblings died with same scenario.

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

- A. galactosemia
- B. hyperglycinemia
- C. organic acidemias
- D. phenylketonurea

E. urea cycle defects

5. You suspect a metabolic problem in a 30-day-old girl presented with poor feeding, vomiting, lethargy, and convulsion. Previous sibling died with the same condition. Serum ammonia, pH, HCO_3 , and anion gap are normal.

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

- A. porphyria
- B. galactosemia
- C. organic acidemia
- D. urea cycle defect
- E. aminoacidopathy

6. A 3-year-old boy presented with history of hyperactivity and mild mental retardation. Past history revealed repeated projectile vomiting with normal abdominal sonography. On examination, there are eczematoid rash with lighter skin, microcephaly, and mild spasticity with exaggerated tendon reflexes.

Of the following, the **MOST** likely urine odor of this baby is

- A. musty
- B. rotting fish
- C. sweaty feet
- D. boiled cabbage
- E. swimming pool

7. A 6-year-old boy with eczematoid rash, abnormal face, and prominent maxilla complains from deterioration in school performance, hyperactivity, and seizures for the last 2 years.

Of the following, the **MOST** likely enzymatic deficiency is

- A. acid β -glucosidase
- B. β -hexosaminidases
- C. acid sphingomyelinase
- D. phenylalanine hydroxylase
- E. fumarylacetoacetate hydrolase

8. A 5-year-old mentally retarded girl presented with repeated convulsions and abnormal EEG finding. On examination, she had a light skin, small head, and exaggerated reflexes.

Of the following, the **MOST** appropriate method to confirm the diagnosis is

- A. Guthrie test
- B. flow cytometry
- C. tandem mass spectrometry
- D. measurement of phenylketones in the urine
- E. measurement of plasma phenylalanine concentration

9. In the classic phenylketonurea, the affected infant is normal at birth, intellectual disability develops gradually if the infant remains untreated, and cognitive delay may not be evident for the first few months.

All the following are true about neonatal screening program of this condition **EXCEPT**

- A. Guthrie test was the 1st method used
- B. optimal time for Guthrie test at 7-14 of life
- C. blood phenylalanine may rise to a diagnostic level as early as 4 hr after birth
- D. fluorometric and tandem mass spectrometry give a low false-positive rate
- E. diagnosis must be confirmed by measurement of plasma phenylalanine concentration

10. The mainstay of treatment of phenylketonurea (PKU) is especial diet.

All the following statements are true **EXCEPT**

- A. phenylalanine not totally restricted
- B. all patients kept on a phenylalanine-restricted diet lifelong
- C. especial diet should be used before and during pregnancy in mother with PKU
- D. especial diet should be started when blood phenylalanine levels above 10 mg/dL
- E. especial diet is not advocated in mild hyperphenylalaninemia whose levels are ≥ 6 mg/Dl

11. A 4-month-old boy presented with failure to thrive, rapid breathing, and repeated vomiting. On examination, there is hepatomegaly and abdominal sonography revealed renal calcification.

Of the following, the **MOST** likely urine odor of this infant is

- A. mousey
- B. rotting fish
- C. sweaty feet
- D. boiled cabbage
- E. swimming pool

12. A 10-month-old boy presented with failure to thrive, fever, jaundice, hepatomegaly, and severe rickets. Investigations revealed hypoglycemia and normal anion gap metabolic acidosis.

Of the following, the **MOST** likely enzyme deficiency is

- A. acid β -glucosidase
- B. β -hexosaminidases
- C. acid sphingomyelinase
- D. phenylalanine hydroxylase
- E. fumarylacetoacetate hydrolase

13. A 15-month-old boy had recurrent hospital admissions because of fever, jaundice, subcutaneous bleeding, and hypoglycemic fits; on examination, there is hepatomegaly. In the last attack, the child developed severe legs pain associated with retraction of the neck and trunk.

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

- A. galactosemia
- B. organic acidemia
- C. urea cycle defect
- D. tyrosinemia type 1
- E. maple syrup urine disease

14. The **MOST** appropriate method to confirm the diagnosis of tyrosinemia type 1 is by elevated level of

- A. α -fetoprotein
- B. plasma tyrosine
- C. serum methionine
- D. serum succinylacetone
- E. urinary 5-aminolevulinic acid

15. A 10-day-old boy presented with lethargy, poor feeding, repeated vomiting, and recurrent hypoglycemic fits not responding to IV glucose. On examination, he was hypertonic with severe opisthotonos alternating with flaccidity.

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

- A. tyrocinemia
- B. galactosemia
- C. phenyletonuria
- D. homocystinurea
- E. maple syrup urine disease

16. A 27-day-old girl presented with abnormal bicycling movement, lethargy, poor feeding, and repeated vomiting for the last 7 days, and then she developed rapid breathing, opisthotonos, and hypertonicity with bad odor urine.

Of the following, the **MOST** effective mode of therapy is

- A. good hydration
- B. peritoneal dialysis
- C. liver transplantation
- D. sufficient calories and nutrients
- E. diet low in branched-chain amino acids

17. The following are ocular manifestations of albinism **EXCEPT**

- A. red reflex
- B. strabismus

- C. refractive errors
- D. foveal hyperplasia
- E. lack of binocular vision

18. A 6-year-old tall and thin boy who had failed preparatory exam for school entry found to have myopia and subluxation of the ocular lens.

Of the following, the **MOST** important challenge is to know whether he is responsive to

- A. folic acid
- B. vitamin C
- C. vitamin E
- D. vitamin B6
- E. vitamin B12

19. A 6-year-old girl that looks tall and thin with light skin. On examination she has a peculiar malar flush, subluxation of the ocular lens, developmental delay, and severe hypertension.

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

- A. atherosclerosis
- B. hyperthyroidism
- C. thromboembolism
- D. coarctation of aorta
- E. renal artery stenosis

20. The **MOST** appropriate treatment of homocystinuria is

- A. betaine
- B. folic acid
- C. vitamin C
- D. vitamin B6
- E. methionine restriction

21. The diagnosis of homocystinuria is usually made after 3 yr of age when the ophthalmologist found

- A. cataracts
- B. glaucoma
- C. astigmatism
- D. ectopia lentis
- E. retinal detachment

22. An 11-month-old boy presented with repeated convulsions, poor eye contact, exaggerated startle response to noise, and large head. Ophthalmic exam revealed cherry-red spot.

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

- A. Tyrosinemia
- B. Fabry disease
- C. Gaucher disease
- D. Tay-Sachs disease
- E. Niemann-pick disease

23. One of the following is a characteristic feature for both Infantile Sandhoff disease and Tay-Sachs disease

- A. splenomegaly
- B. hepatomegaly
- C. cardiac involvement
- D. retinal cherry-red spots
- E. mild bony abnormalities

24. A 12-year-old boy presented with epistaxis, mild bruising, intermittent bone pain, and massive splenomegaly. Complete blood count shows thrombocytopenia and mild anemia. Radiological study revealed lytic lesions and Erlenmeyer flask deformity of the distal femur.

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

- A. Gaucher disease
- B. Sandhoff disease
- C. Tay-Sachs disease
- D. von Gierke disease
- E. Niemann-pick disease

25. A 9-year-old boy admitted to the hospital because of fracture of left femur due to a minor trauma. Past history revealed recurrent attacks of epistaxis and intermittent generalized bone pain. On examination, there is massive splenomegaly.

Of the following, the **MOST** appropriate practical treatment of this disease is

- A. gene therapy
- B. liver transplantation
- C. enzyme replacement
- D. substrate reduction agents
- E. bone marrow transplantation

26. A 2-year-old boy presented with failure to thrive, regression of developmental milestone, and difficult to contact with surroundings. On examination, there are hepatosplenomegaly and moderate lymphadenopathy.

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

- A. Gaucher disease
- B. Sandhoff disease
- C. Tay-Sachs disease

- D. Von Gierke disease
 - E. Niemann-pick disease
27. All the following are recognized feature of Niemann-pick disease type B **EXCEPT**
- A. normal IQ
 - B. progressive lung disease
 - C. prolonged neonatal jaundice
 - D. subtle or no neurologic involvement
 - E. cherry-red maculae in some patients
28. All the following lipidoses are inherited as an autosomal recessive traits **EXCEPT**
- A. Fabry disease
 - B. Krabbe disease
 - C. Gaucher disease
 - D. Tay-sachs disease
 - E. Niemann-pick disease
29. Niemann-pick disease type A is characterized by all the following **EXCEPT**
- A. failure to thrive
 - B. hepatosplenomegaly
 - C. progressive lung disease
 - D. psychomotor retardation
 - E. rapidly progressive neurodegenerative
30. A 7-month-old boy presented with recurrent attacks of hypoglycemic seizures especially during an acute illness. On examination, there are thin extremities, protuberant abdomen, and hepatomegaly.
- All the following are biochemical hallmarks of this disease **EXCEPT**
- A. lactic acidosis
 - B. hypoglycemia
 - C. hyperuricemia
 - D. hyperlipidemia
 - E. markedly elevated transaminase levels
31. A 2-year-old girl presented with rapid deep breathing, hypoglycemia, recurrent epistaxis, easy bruising, failure to thrive, fat cheek, and hepatomegaly.
- Of the following, the **MOST** likely cause of bleeding tendency is
- A. vascular disorders
 - B. clotting factors deficiency
 - C. Impaired hepatic function
 - D. depressed platelets count
 - E. impaired platelet aggregation and adhesion

32. A 2-year-old boy presented with growth retardation, hepatomegaly, and recurrent attacks of hypoglycemia; biochemical study reveals elevated blood lactate, triglyceride, and uric acid levels.

All the following are complications of this disease **EXCEPT**

- A. pancreatitis
- B. renal failure
- C. cardiomyopathy
- D. hepatic adenoma
- E. pulmonary hypertension

33. A 3-year-old girl presented with growth retardation, recurrent attacks of otitis media, diarrhea, and hypoglycemic seizures. On examination, there is hepatomegaly and her absolute neutrophil count (ANC) is 600/ μ L.

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

- A. Kostmann disease
- B. cyclic neutropenia
- C. drug-induced neutropenia
- D. glycogen storage disease Ib
- E. Shwachman-Diamond syndrome

34. The definitive diagnosis of type I glycogen storage disease GSD is by

- A. EMG
- B. liver biopsy
- C. muscle biopsy
- D. analysis of gene-based mutation
- E. enzyme assay of cultured skin fibroblasts

35. In von Gierke disease; treatment is designed to maintain normal blood glucose levels which is achieved by all the following **EXCEPT**

- A. restriction of fructose
- B. restriction of galactose
- C. administration of table sugar
- D. continuous nasogastric infusion of glucose
- E. oral administration of uncooked corn starch

36. A 9-month-old boy presented with persistent head lag, rapid breathing, difficult feeding, and hepatomegaly. Chest X- ray showed cardiomegaly.

Of the following, the **MOST** appropriate confirmatory test is

- A. liver biopsy
- B. muscle biopsy
- C. electrocardiography
- D. serum creatine phosphokinase

E. enzyme assay in dried blood spots

37. A 7-month-old girl presented with recurrent chest infection, choking during feeding, large protruded tongue, persistent head lag, and hepatomegaly. Chest X-ray showed cardiomegaly.

All the following are beneficial in the treatment of this patient **EXCEPT**

- A. exercise therapy
- B. high-protein diet
- C. cardiac transplantation
- D. nocturnal ventilatory support
- E. specific enzyme replacement therapy

38. A 28-day-old boy presented with prolonged neonatal jaundice, lethargy, poor feeding, and failure to regain birth weight. On examination there are hepatomegaly and cataract.

Of the following, the **MOST** likely injured organs in this disease are

- A. eyes, liver, and brain
- B. kidney, liver, and eyes
- C. kidney, liver, and brain
- D. kidney, heart, and brain
- E. pancreas, liver, and brain

39. Irreversible complication of untreated galactosemia is

- A. sepsis
- B. cataract
- C. renal injury
- D. myocardial damage
- E. intellectual disability

40. A 20-day-old girl presented with prolonged neonatal jaundice, lethargy, and poor feeding treated as sepsis by empirical antibiotics and nothing by mouth. The patient got partial response, when oral feeding resumed, the condition deteriorate again.

In this condition, the blood culture **MOST** likely reveals growth of

- A. *Escherichia coli*
- B. *Staphylococcal aureus*
- C. group B streptococcus
- D. *Listeria monocytogens*
- E. *Pseudomonas aeruginosa*

41. A 9-month-old boy with exclusive breast feeding presented with jaundice, vomiting, lethargy, hepatomegaly, and hypoglycemic fits when sweetened cereal added.

Laboratory findings showed prolonged clotting time, hypoalbuminemia, elevation of bilirubin and transaminase levels, and proximal tubular dysfunction.

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

- A. tyrosinemia
- B. galactosemia
- C. organic acidemias
- D. maple syrup urine disease
- E. hereditary fructose intolerance

42. All the following are common features between hereditary fructose intolerance and galactosemia **EXCEPT**

- A. failure to thrive
- B. impaired renal function
- C. impaired hepatic function
- D. reducing substance in urine
- E. impaired intellectual development

43. All the following should be restricted in hereditary fructose intolerance **EXCEPT**

- A. milk
- B. sorbitol
- C. fruit juice
- D. table sugar
- E. sweetened cereal

44. The earliest radiographic sign in Hurler syndrome is

- A. thick ribs
- B. thickened calvarium
- C. enlarged J-shaped sella
- D. abnormal spacing of teeth
- E. premature closure of lambdoid suture

45. Enzyme replacement is the main therapy of Hurler disease, it improve all the following **EXCEPT**

- A. growth rate
- B. joint mobility
- C. organomegaly
- D. cognition functions
- E. episodes of sleep apnea

46. A 9-year-old boy presented with tiredness on exertion; cardiac auscultation reveals a murmur in aortic area; echo study showed aortic valve disease. On examination he

had mild coarse facial features, corneal clouding, joint stiffness, and mild dysostosis multiplex but normal intelligence and stature.

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

- A. Hurler disease
- B. Scheie disease
- C. Hunter disease
- D. Morquio disease
- E. Sanfilippo disease

47. Hunter disease is characterized by all the following features **EXCEPT**

- A. short stature
- B. corneal clouding
- C. dysostosis multiplex
- D. intellectual disability
- E. coarse facial features

48. All the following mucopolysaccharidoses are inherited as autosomal recessive **EXCEPT**

- A. Hurler disease
- B. Scheie disease
- C. Hunter disease
- D. Morquio disease
- E. Sanfilippo disease

49. Which of the following mucopolysaccharidoses have normal intelligence?

- A. Sly disease
- B. Hurler disease
- C. Hunter disease
- D. Morquio disease
- E. Sanfilippo disease

50. A 5-year-old boy presented with chronic diarrhea. On examination he had hepatosplenomegaly, coarse facial features, joint stiffness, clear cornea, and short stature.

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

- A. Hurler disease
- B. Hunter disease
- C. Scheie disease
- D. Morquio disease
- E. Sanfilippo disease

51. In mucopolysaccharidosis, disproportionate severe CNS involvement with mild physical features is unique to
- A. Scheie disease
 - B. Hurler disease
 - C. Hunter disease
 - D. Morquio disease
 - E. Sanfilippo disease
52. All the following are skeletal manifestations of Morquio disease **EXCEPT**
- A. kyphosis
 - B. genua valga
 - C. waddling gait
 - D. short trunk and neck
 - E. bullet-shaped phalanges
53. Which of the following mucopolysaccharidoses is susceptible to life-threatening atlantoaxial instability and dislocation?
- A. Hurler disease
 - B. Scheie disease
 - C. Hunter disease
 - D. Morquio disease
 - E. Sanfilippo disease
54. The characteristic radiographic finding of Hurler disease is
- A. kyphosis
 - B. genua valgus
 - C. V-shaped configuration of the digits
 - D. hook-shaped appearance of L₁ vertebra
 - E. ovoid configuration of the vertebral bodies
55. A definitive diagnosis of mucopolysaccharidoses disorder is established by
- A. liver biopsy
 - B. enzyme assay
 - C. radiographic features
 - D. bone marrow examination
 - E. urinary glycosaminoglycan (GAG) test
56. Acute intermittent porphyria (AIP) could be exacerbated by all the following **EXCEPT**
- A. surgery
 - B. puberty
 - C. pregnancy

- D. premenstrual period
 - E. decreased carbohydrate intake
57. All the following drugs are unsafe in acute intermittent porphyria **EXCEPT**
- A. nifedipine
 - B. diclofenac
 - C. cimetidine
 - D. valproic acid
 - E. carbamazepine
58. All the following are clinical features of acute intermittent porphyria **EXCEPT**
- A. neuropathy
 - B. tachycardia
 - C. abdominal pain
 - D. bladder dysfunction
 - E. cutaneous photosensitivity
59. The least common presentation of acute intermittent porphyria is
- A. sensory loss
 - B. mental symptoms
 - C. muscle weakness
 - D. abdominal tenderness
 - E. pain in the extremities
60. The **MOST** common electrolyte abnormality in acute intermittent porphyria is
- A. hyperkalemia
 - B. hyponatremia
 - C. hypercalcemia
 - D. hypomagnesemia
 - E. hypophosphatemia
61. All the following are features of congenital erythropoietic porphyria (CEP) **EXCEPT**
- A. neuropathy
 - B. reddish urine
 - C. erythrodontia
 - D. photosensitivity
 - E. neonatal hyperbilirubinemia
62. The **MOST** important screening test of acute intermittent porphyria is
- A. plasma porphyrins
 - B. fibroblast porphyrins
 - C. fecal porphobilinogen

- D. erythrocyte porphyrins
- E. urinary porphobilinogen

63. In older infants and children, hypoglycemia is defined as whole blood glucose concentration less than

- A. 45 mg/dL
- B. 50 mg/dL
- C. 55 mg/dL
- D. 60 mg/dL
- E. 65 mg/dL

64. The least well-defined long-term sequelae of severe prolonged hypoglycemia is

- A. cerebral palsy
- B. personality affection
- C. cognitive impairment
- D. recurrent seizure activity
- E. autonomic dysregulation

65. The following are counter regulatory hormones which are increased as blood glucose falls **EXCEPT**

- A. cortisol
- B. glucagon
- C. epinephrine
- D. thyroid hormone
- E. growth hormone

66. A 3-year-old boy presented to emergency unit at 9:00 AM with history of difficult arousal from sleep, flue like illness, and low grade fever; the mother stated that he had slept without having his supper. Fasting blood sugar was 42 mg/dL.

The following metabolic changes are seen in this condition **EXCEPT**

- A. ketonuria
- B. low insulin level
- C. high alanine level
- D. high cortisol level
- E. high growth hormone level

67. At 10:00 AM Lylaa`s mother brought her 3-year-old daughter to emergency department with difficult arousal from sleep, low grade fever, and cough for the last 2 days. Fasting blood sugar was 36 mg/dL with ketonuria.

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

- A. galactosemia
- B. propionic acidemia

- C. ketotic hypoglycemia
- D. hereditary fructose intolerance
- E. medium-chain acyl-CoA dehydrogenase (MCAD) deficiency

68. A 5-year-old child with recurrent attacks of ketotic hypoglycemia.

Of the following, the **BEST** advice to the parents during any intercurrent illness is to

- A. administer a high-protein diet
- B. test the child's urine for ketones
- C. administer a high-carbohydrate diet
- D. do frequent monitoring of blood sugar
- E. administer intravenous glucose administration

69. The **MOST** specific and important laboratory finding in X-linked adrenoleukodystrophy (ALD) is elevated level of

- A. cortisol
- B. aldosterone
- C. epinephrine
- D. very long chain fatty acid
- E. adrenocorticotrophic hormone

70. In X-linked adrenoleukodystrophy (ALD), MRI shows characteristic cerebral white matter lesions mainly in

- A. frontal lobes
- B. temporal lobes
- C. frontoparietal lobes
- D. parietoccipital lobes
- E. temporoparietal lobes

71. All the following are secondary causes of hypercholesterolemia **EXCEPT**

- A. obesity
- B. cholestasis
- C. hypothyroidism
- D. anorexia nervosa
- E. nephrotic syndrome

72. All the following drugs are secondary causes of hypertriglyceridemia **EXCEPT**

- A. tegretol
- B. estrogen
- C. thiazides
- D. β -blockers
- E. anabolic steroids

73. Smith Lemli-Opitz Syndrome (SLOS) is characterized by all the following **EXCEPT**
- A. epispadias
 - B. cleft palate
 - C. microcephaly
 - D. retromicrognathia
 - E. mental retardation
74. All the following are recognized laboratory findings of medium-chain acyl-CoA dehydrogenase (MCAD) deficiency **EXCEPT**
- A. hypoglycemia
 - B. hypoketonuria
 - C. hypoketonemia
 - D. metabolic acidosis
 - E. elevated liver enzyme
75. A 3-year-old boy presented to emergency unit at 10:00 AM with history of difficult arousal from sleep, lethargy, vomiting, and seizures; the mother stated that he had slept without having his supper. Fasting blood sugar was 38mg/dL and urinary ketone was low.
- Of the following, the **MOST** likely diagnosis is
- A. galactosemia
 - B. propionic acidemia
 - C. ketotic hypoglycemia
 - D. hereditary fructose intolerance
 - E. medium-chain acyl-CoA dehydrogenase (MCAD) deficiency
76. The **MOST** common presentation of primary carnitine deficiency is
- A. liver disease
 - B. cardiomyopathy
 - C. skeletal myopathy
 - D. endocardial fibroelastosis
 - E. hypoketotic hypoglycemia
77. All the following are clinical abnormalities of Zellweger syndrome **EXCEPT**
- A. cataract
 - B. micrognathia
 - C. high forehead
 - D. small fontanells
 - E. redundant skin fold of neck
78. All the following are features of neonatal adrenoleukodystrophy (ALD) **EXCEPT**
- A. hepatomegaly

- B. impaired liver function
- C. severely impaired hearing
- D. chondrodysplasia punctata
- E. pigmentary degeneration of the retina

79. All the following are characteristic features of infantile Refsum disease **EXCEPT**

- A. ataxic gait
- B. hypertonia
- C. hearing loss
- D. impaired cognition
- E. retinal degeneration

80. The diagnostic assay of Zellweger syndrome is by elevated level of

- A. bile acid
- B. plasmalogen
- C. phytanic acid
- D. pristanic acid
- E. very long chain fatty acid

81. The normally functioning organ in Hutchinson-Gilford progeria syndrome (progeria) is

- A. eye
- B. ear
- C. skin
- D. liver
- E. heart

82. The **MOST** common initial manifestation of X-linked adrenoleukodystrophy (ALD) is

- A. seizure
- B. hyperactivity
- C. visual disturbance
- D. increased intracranial pressure
- E. impaired auditory discrimination

83. The **MOST** common presentation of mitochondrial β -oxidation of fatty acids disorders is

- A. cardiomyopathy
- B. muscle weakness
- C. renal tubulopathy
- D. hypoglycemic coma
- E. acute rhabdomyolysis

84. The specific clue to the diagnosis of fatty acid oxidation disorders may be the finding of hypoglycemia and

- A. hypoketonuria
- B. metabolic acidosis
- C. elevated liver enzyme
- D. elevated blood ammonia
- E. prolonged prothrombin time

85. Xanthomas are a feature of all the following **EXCEPT**

- A. familial hypercholesterolemia
- B. familial dysbetalipoproteinemia
- C. familial combined hyperlipidemia
- D. familial chylomicronemia (Frederickson type I)
- E. familial hypertriglyceridemia (Frederickson type V)

Metabolic Disorders

ANSWERS

KHALID ALAARJI

1.(D). Many childhood conditions are caused by single-gene mutations that encode specific proteins. These mutations can result in the alteration of primary protein structure or the amount of protein synthesized. The function of a protein, whether it is an enzyme, receptor, transport vehicle, membrane component, or structural element, may be compromised or abolished.

2.(B). The earlier the appearance of clinical symptoms, the more severe is the disease.

3.(B). Replacement of the mutant gene with a normal one (gene therapy) is still in the experimental phase.

4.(E). Elevation of blood ammonia is usually caused by defects of urea cycle enzymes. Infants with elevated blood ammonia levels from urea cycle defects commonly have normal serum pH and bicarbonate values; without measurement of blood ammonia, they may remain undiagnosed and succumb to their disease.

5.(E).

6.(A). Phenylketonurea is the most likely diagnosis; these children have an unpleasant odor of phenylacetic acid, which has been described as musty or mousey.

- Hawkinsinuria -----Swimming pool
- Isovaleric acidemias ----- Sweaty feet
- Trimethylaminuria ----- Rotting fish
- Tyrosinemia --- Boiled cabbage, rancid butte

7.(D). Phenylketonurea is the most likely diagnosis; deficiency of the enzyme phenylalanine hydroxylase (PAH) or of its cofactor tetrahydrobiopterin (BH4) causes accumulation of phenylalanine in body fluids and in the brain.

8.(E). In infants with positive screening results, diagnosis should be confirmed by quantitative measurement of plasma phenylalanine concentration.

9.(B). It is recommended that the blood for screening be obtained in the first 24-48 hr of life after feeding protein to reduce the possibility of false-negative results, especially in the milder forms of the condition.

10.(E). The mainstay of treatment of PKU is a low-phenylalanine diet. Most physicians advocate phenylalanine-restricted diet in patients with mild hyperphenylalaninemia whose levels are persistently above 6 mg/ dL (360 μ mole/L). It is generally accepted that infants with persistent (more than a few days) plasma levels of phenylalanine $\geq$ 6 mg/dL (360 μ mole/L) should be treated with a phenylalanine-restricted diet similar to that for classic PKU.

11.(D). Tyrosinemia is the most likely diagnosis; these children have an unpleasant odor which has been described as boiled cabbage, rancid butter.

12.(E). Tyrocinemia type 1 is a severe disease of the liver, kidney, and peripheral nerves are caused by a deficiency of the enzyme fumarylacetoacetate hydrolase.

- acid β -glucosidase ----- Gaucher disease
- phenylalanine hydroxylase ----- Phenylketonurea
- acid sphingomyelinase ---- Niemann-pick disease
- fumarylacetoacetate hydrolase -----Tyrosinemia
- β -hexosaminidases ----- Tay-Sachs disease

13.(D). Tyrocinemia type 1 is a severe disease of the liver, kidney, and peripheral nerves are caused by a deficiency of the enzyme fumarylacetoacetate hydrolase. An acute hepatic crisis commonly heralds the onset of the disease and is usually precipitated by an intercurrent illness that produces a catabolic state. Episodes of acute peripheral neuropathy occur in approximately 40% of affected children. Renal involvement is manifested as a Fanconi-like syndrome.

14.(D). The presence of elevated levels of succinylacetone in serum and urine is diagnostic for tyrosinemia type I. Increased levels of α -fetoprotein are present in the cord blood of affected infants, indicating intrauterine liver damage. Plasma tyrosine levels are usually elevated at diagnosis but this is a nonspecific finding and is dependent on dietary intake. Plasma levels of other amino acids, particularly methionine, may also be elevated in patients with liver damage. The urinary level of 5-aminolevulinic acid is elevated because of inhibition of 5-aminolevulinic hydratase by succinylacetone.

15.(E). In maple syrup urine disease, affected infants who are normal at birth develop poor feeding and vomiting in the 1st wk of life; lethargy and coma may ensue within a few days. Physical examination reveals hypertonicity and muscular rigidity with severe opisthotonos. Periods of hypertonicity may alternate with bouts of flaccidity manifested as repetitive movements of the extremities (boxing and bicycling). Cerebral edema may be present; convulsions occur in most infants, and hypoglycemia is common.

16.(B). Treatment of the acute state of maple syrup urine disease is aimed at hydration and rapid removal of the branched-chain amino acids and their metabolites from the tissues and body fluids. Because renal clearance of these compounds is poor, hydration alone may not produce a rapid improvement. Peritoneal dialysis or, preferably, hemodialysis is the most effective mode of therapy in critically ill infants.

17.(D). Ocular manifestations of albinism include hypopigmentation of iris and retina, foveal hypoplasia, reduced visual acuity, refractive errors, nystagmus, alternating strabismus, and a red reflex.

18.(D). Homocystinuria; this is the most common inborn error of methionine metabolism. Approximately 40% of affected patients respond to high doses of vitamin B6 and usually have milder clinical manifestations than those who are unresponsive to vitamin B6 therapy. These patients possess some residual enzyme activity.

19.(C). In Homocystinuria, thromboembolic episodes involving both large and small vessels, especially those of the brain, are common and may occur at any age. Optic

atrophy, paralysis, cor pulmonale, and severe hypertension (from renal infarcts) are among the serious consequences of thromboembolism, which is caused by changes in the vascular walls and increased platelet adhesiveness secondary to elevated homocystine levels.

20.(D). Treatment with high doses of vitamin B6 (200-1,000 mg/24 hr) causes dramatic improvement in most patients who are responsive to this therapy.

21.(D). Infants with homocystinuria are normal at birth. Clinical manifestations during infancy are nonspecific and may include failure to thrive and developmental delay. The diagnosis is usually made after 3 yr of age, when subluxation of the ocular lens (ectopia lentis) occurs.

22.(D). Tay-Sachs disease results from the deficiency of β -hexosaminidase activity. Clinical manifestations in infancy including loss of motor skills, increased startle reaction, and macular pallor and retinal cherry-red spots.

23.(D). Both of Sandhoff disease and Tay-Sachs disease in infant characterize by a neurologic features and retinal cherry-red spots. Infants with Sandhoff disease have hepatosplenomegaly, cardiac involvement, and mild bony abnormalities.

24.(A). Gaucher disease is a multisystemic lipidosis characterized by hematologic abnormalities, organomegaly, and skeletal involvement, the latter usually manifesting as bone pain and pathologic fractures. It is one of the most common lysosomal storage diseases, results from the deficient activity of the lysosomal hydrolase, acid β -glucosidase, which is encoded by a gene located on chromosome 1q21-q31. The pathologic hallmark of Gaucher disease is the Gaucher cell in the reticuloendothelial system, particularly in the bone marrow.

25.(C). Treatment of patients with Gaucher disease type 1 includes enzyme replacement therapy. The efficacy of enzyme replacement therapy with mannose-terminated recombinant human acid β -glucosidase has definitively been demonstrated. Most symptoms (organomegaly, hematologic indices, bone pain) are reversed by enzyme replacement therapy (60 IU/kg) administered by intravenous infusion every other week and the bone involvement can be stabilized or improved.

26.(E). Niemann-pick disease (NPD), inherited as autosomal recessive traits, the clinical manifestations and course of type A NPD is uniform and is characterized by a normal appearance at birth. Hepatosplenomegaly, moderate lymphadenopathy, and psychomotor retardation are evident by 6 mo of age, followed by neurodevelopmental regression and death by 3 yr. With advancing age, the loss of motor function and the deterioration of intellectual capabilities are progressively debilitating; and in later stages, spasticity and rigidity are evident. Affected infants lose contact with their environment. Type B disease is a non-neuronopathic form observed in children and adults NPD types A and B result from the deficient activity of acid sphingomyelinase. Type C disease is a neuronopathic form that results from defective cholesterol transport.

27.(C). Niemann-pick disease type B patients do not have neurologic involvement and have a normal IQ. Some patients with type B disease have cherry-red maculae or haloes

and subtle neurologic symptoms (peripheral neuropathy). In some type B patients, decreased pulmonary diffusion caused by alveolar infiltration becomes evident in late childhood or early adulthood and progresses with age. Prolonged neonatal jaundice occurs in type C.

28.(A). Fabry disease, inherited as X-linked recessive.

29.(C). The progressive deposition of sphingomyelin in the central nervous system results in the neurodegenerative course seen in type A, and in nonneural tissue in the systemic disease manifestations of type B, including progressive lung disease in some patients.

30.(E). In type I GSD (von Gierke disease), despite marked hepatomegaly, the liver transaminase levels are usually normal or only slightly elevated.

31.(E). In type I GSD (von Gierke disease), easy bruising and epistaxis are common and are associated with a prolonged bleeding time as a result of impaired platelet aggregation and adhesion.

32.(C). Type I GSD (von Gierke disease) complicated by pancreatitis, renal failure, hepatic adenoma, and pulmonary hypertension, while cardiomyopathy occurs in type II GSD (pompe disease).

33.(D). In patients with GSD Ib, the loss of mucosal barrier function as a result of inflammation, which is likely related to the disturbed neutrophil function, seems to be the main cause of diarrhea.

34.(B). The diagnosis of type I GSD is suspected on the basis of clinical presentation and the laboratory findings of hypoglycemia, lactic acidosis, hyperuricemia, and hyperlipidemia. Neutropenia is noted in GSD Ib patients, typically before 1 yr of age. Administration of glucagon or epinephrine results in little or no rise in blood glucose level, but the lactate level rises significantly. Before the glucose-6-phosphatase and glucose-6-phosphate translocase genes were cloned, a definitive diagnosis required a liver biopsy. Gene-based mutation analysis provides a noninvasive way to diagnose most patients with types Ia and Ib disease.

35.(C). Sucrose (table sugar, cane sugar, other ingredients), fructose (fruit, juice, high fructose corn syrup), lactose (dairy foods), and sorbitol should be avoided or limited.

36.(E). The confirmatory step for a diagnosis of Pompe disease is enzyme assay demonstrating deficient acid α -glucosidase. The enzyme assay is usually done in dried blood spots, leukocytes, blood mononuclear cells, muscle, and cultured skin fibroblasts.

37.(C). Specific enzyme replacement therapy (ERT) with recombinant human acid α -glucosidase is available for treatment of Pompe disease and is capable of preventing deterioration or reversing abnormal cardiac and skeletal muscle functions. A high-protein diet and exercise therapy may also be beneficial. Nocturnal ventilatory support, when indicated, should be used. It has been shown to improve the quality of life and is particularly beneficial during a period of respiratory decompensation.

38.(C). In galactosemia, without the transferase enzyme, the infant is unable to metabolize galactose-1-phosphate, the accumulation of which results in injury to kidney, liver, and brain.

- 39.(E). In galactosemia when the diagnosis is not made at birth, damage to the liver (cirrhosis) and brain (intellectual disability) becomes increasingly severe and irreversible.
- 40.(A). Patients with galactosemia are at increased risk for *Escherichia coli* neonatal sepsis; the onset of sepsis often precedes the diagnosis of galactosemia.
- 41.(E). Patients with hereditary fructose intolerance HFI are asymptomatic until fructose or sucrose (table sugar) is ingested (usually from fruit, fruit juice, or sweetened cereal). Symptoms may occur early in life, soon after birth if foods or formulas containing these sugars are introduced into the diet.
- 42.(E). In hereditary fructose intolerance, intellectual development is usually unimpaired.
- 43.(A). In hereditary fructose intolerance, treatment consists of the complete elimination of all sources of sucrose, fructose, and sorbitol from the diet. It may be difficult because these sugars are widely used additives, found even in most medicinal preparations.
- 44.(A). Hurler syndrome; Radiographs show a characteristic skeletal dysplasia known as dysostosis multiplex. The earliest radiographic signs are thick ribs and ovoid vertebral bodies. Skeletal abnormalities include enlarged, coarsely trabeculated diaphyses of the long bones with irregular metaphyses and epiphyses. With progression of the disease macrocephaly develops with thickened calvarium, premature closure of lambdoid and sagittal sutures, shallow orbits, enlarged J-shaped sella, and abnormal spacing of teeth with dentigerous cyst.
- 45.(D). Enzyme replacement using recombinant α -L-iduronidase has been approved for patients with MPS-I. It reduces organomegaly and ameliorates rate of growth, joint mobility, reduces the number of episodes of sleep apnea and urinary GAG excretion. The enzyme does not cross the blood–brain barrier and does not prevent deterioration of cognition and other neurologic functions.
- 46.(B). Scheie is a comparatively mild disorder characterized by joint stiffness, aortic valve disease, corneal clouding, and mild dysostosis multiplex. Patients with Scheie disease have normal intelligence and stature but have significant joint and ocular involvement.
- 47.(B). Patients with severe MPS-II have features similar to those of Hurler disease except for the lack of corneal clouding and the somewhat slower progression of somatic and central nervous system (CNS) deterioration.
- 48.(C). Hunter disease inherited as X- linked recessive.
- 49.(D). Morquio disease is characterized by short-trunk dwarfism, fine corneal deposits, a skeletal dysplasia that is distinct from other mucopolysaccharidoses, and preservation of intelligence.
- 50.(B).
- 51.(E). Patients with Sanfilippo disease are characterized by slowly progressive, severe CNS involvement with mild somatic disease. Such disproportionate involvement of the CNS is unique to MPS-III.

52.(E). Bullet-shaped phalanges in addition to metacarpals and phalanges are abnormally short, wide and deformed with proximal pointing of the metacarpals are feature of Hurler disease.

53.(D). In Morquio disease, instability of the odontoid process and ligamentous laxity is regularly present and can result in life-threatening atlantoaxial instability and dislocation.

54.(D). Hurler disease, anterosuperior hypoplasia of L-1 resulting in hook-shaped appearance. Sanfilippo disease, immature, ovoid configuration of the vertebral bodies. Morquio disease, genua valgus and kyphosis. Scheie disease, the carpal bones are small leading to V-shaped configuration of the digits.

55.(B). Any individual who is suspected of a mucopolysaccharidoses disorder based on clinical features, radiographic results, or urinary GAG screening tests should have a definitive diagnosis established by enzyme assay. Serum, leukocytes, or cultured fibroblasts are used as the tissue source for measuring lysosomal enzymes.

56.(C). Surprisingly, pregnancy is usually well tolerated, suggesting that beneficial metabolic changes may ameliorate the effects of high levels of progesterone.

57.(C).

58.(E). Cutaneous photosensitivity is a feature of Porphyria cutanea tarda.

59.(D). The common manifestations include mental symptoms; pain in the extremities, head, neck, or chest; muscle weakness; and sensory loss. Because all these manifestations are neurologic rather than inflammatory, there is little or no abdominal tenderness, fever, or leukocytosis.

60.(B). Hyponatremia is common during acute attacks. Inappropriate antidiuretic hormone secretion is often the most likely mechanism, but salt depletion from excess renal sodium loss, gastrointestinal loss, and poor intake have been suggested as causes of hyponatremia in some patients. Other electrolyte abnormalities may include hypomagnesemia and hypercalcemia.

61.(A). Neuropathic symptoms are absent, and there is no sensitivity to drugs, hormones, and carbohydrate restriction.

62.(E).

- Porphyria cutanea tarda ----- Plasma (or urine) porphyrins.
- Erythropoietic protoporphyria ---- Erythrocyte (or plasma) porphyrins.

63.(C). In older infants and children, a whole blood glucose concentration of <55 mg/dL (10-15% higher for serum or plasma) represents hypoglycemia, because counter regulatory mechanisms are activated at these glucose concentrations.

64.(B). Subtle effects on personality are also possible but have not been clearly defined.

65.(D). Thyroid hormone is not a counter regulatory hormone in response to hypoglycemia.

66.(C). Children with ketotic hypoglycemia have plasma alanine concentrations that are markedly reduced in the basal state after an overnight fast and decline even further with prolonged fasting. Alanine, produced in muscle, is a major gluconeogenic precursor.

67.(C). During acute episodes of medium-chain acyl-CoA dehydrogenase (MCAD) deficiency, hypoglycemia is usually present; plasma and urinary ketone concentrations are inappropriately low (hypoketotic hypoglycemia).

68.(B). During intercurrent illnesses, parents should be taught to test the child's urine for the presence of ketones, the appearance of which precedes hypoglycemia by several hours. In the presence of ketonuria, liquids of high carbohydrate content should be offered to the child.

69.(D). The most specific and important laboratory finding is the demonstration of abnormally high levels of VLCFA in plasma, red blood cells, or cultured skin fibroblasts. The test should be performed in a laboratory that has experience with this specialized procedure. Positive results are obtained in all male patients with ALD and in approximately 85% of female carriers of ALD.

70.(D). In 80% of patients, the lesions are symmetric and involve the periventricular white matter in the posterior parietal and occipital lobes. Approximately 50% show a garland of accumulated contrast material adjacent and anterior to the posterior hypodense lesions. In 12% of patients, the initial lesions are frontal.

71.(A). Obesity is secondary cause of hypertriglyceridemia.

72.(A). Tegretol is secondary causes of hypercholesterolemia.

73.(A). Genital anomalies; hypospadias, cryptorchidism, and sexual ambiguity (<50%).

74.(D). During acute episodes of MCAD deficiency, hypoglycemia is usually present. Plasma and urinary ketone concentrations are inappropriately low (hypoketotic hypoglycemia). Because of the hypoketonemia, there is little or no metabolic acidosis, which is expected to be present in many children with hypoglycemia.

75.(E). MCAD ; Previously undiagnosed affected patients usually present in the first 3 mo-5 yr of life with episodes of acute illness triggered by prolonged fasting (longer than 12-16 hr). During acute episodes of MCAD deficiency, hypoglycemia is usually present. Plasma and urinary ketone concentrations are inappropriately low (hypoketotic hypoglycemia).

76.(B). The most common presentation of primary carnitine deficiency is progressive cardiomyopathy with or without skeletal muscle weakness beginning at 1-4 yr of age.

77.(D). Craniofacial dysmorphisms are; high forehead, flat occiput, large fontanelle(s), wide sutures, shallow orbital ridges, low/broad nasal bridge, epicanthus, high arched palate, external ear deformity, and micrognathia.

78.(D). Chondrodysplasia punctata and renal cysts are absent.

79.(B). Early hypotonia and hepatomegaly with impaired function are common.

80.(E).

- Neonatal adrenoleukodystrophy ---- Phytanic acid (increased).
- Infantile Refsum disease ---- Pristanic acid (increased), Bile acid (Increased), and Plasmalogen level (decreased).

81.(D). In progeria; normally functioning systems are; Liver, kidney, thyroid, immune, gastrointestinal, and neurologic systems (other than stroke-related) remain intact. Intellect is normal for age.

82.(B). In the childhood cerebral form of ALD, symptoms are first noted most commonly between the ages of 4 and 8 yr. The most common initial manifestations are hyperactivity, which is often mistaken for an attention deficit disorder, and worsening school performance in a child who had previously been a good student.

83.(D). The most common presentation is an acute episode of life-threatening coma and hypoglycemia induced by a period of fasting because of defective hepatic ketogenesis.

84.(A). Fatty acid oxidation disorders are easily overlooked because the only specific clue to the diagnosis may be the finding of inappropriately low concentrations of urinary ketones in an infant who has hypoglycemia.

85.(C). Familial combined hyperlipidemia; an autosomal dominant condition characterized by moderate elevation in plasma LDL cholesterol and triglycerides, and reduced plasma HDL cholesterol.

1. Major cause of neonatal mortality in full-term newborn is
 - A. respiratory distress syndrome
 - B. necrotizing enterocolitis
 - C. bronchopulmonary dysplasia (BPD)
 - D. congenital anomalies
 - E. intraventricular hemorrhage

2. Generalized edema of the newborn may occur in the following conditions **EXCEPT**
 - A. prematurity
 - B. nonimmune hydrops
 - C. Turner syndrome
 - D. congenital nephrosis
 - E. Hurler syndrome

3. Soft areas in the occipital region suggest the irregular calcification and wormian bone formation usually associated with the following conditions **EXCEPT**
 - A. osteogenesis imperfecta
 - B. craniosynostosis
 - C. cleidocranial dysostosis
 - D. cretinism
 - E. Down syndrome

4. The following disorders are associated with a large anterior fontanel **EXCEPT**
 - A. congenital rubella syndrome
 - B. hypophosphatasia
 - C. Apert syndrome
 - D. Russell-Silver syndrome
 - E. vitamin A deficiency

5. Leukokoria (white pupillary reflex) in newborn infant suggests the following disorders **EXCEPT**
 - A. cataracts
 - B. tumor
 - C. congenital glaucoma
 - D. chorioretinitis
 - E. retinopathy of prematurity

6. A single umbilical artery in a newborn infant increases the risk for
- meningomyelocele
 - occult renal anomaly
 - omphalocele
 - gastroschisis
 - omphalitis
7. The following drugs can be given with caution to breast-feeding mother
- psychotropic drugs
 - amphetamines
 - bromocriptine
 - chloramphenicol
 - methimazole
8. Of the following, the condition which is associated with polyhydramnios is
- renal agenesis (Potter syndrome)
 - Prune-belly syndrome
 - pulmonary hypoplasia
 - intestinal pseudo-obstruction
 - diaphragmatic hernia
9. Low maternal serum α -fetoprotein (MSAFP) is associated with
- open neural tube defects
 - trisomy 21
 - gastroschisis
 - omphalocele
 - congenital nephrosis
10. A delay in fetal pulmonary maturation may be associated with
- hydrops fetalis
 - severe premature separation of the placenta
 - premature rupture of the fetal membranes
 - narcotic addiction
 - maternal hypertensive and renal vascular disease
11. True umbilical cord knots are seen in approximately 1% of births and are associated with the following conditions **EXCEPT**
- short cord
 - small fetal size
 - polyhydramnios
 - monoamniotic twinning
 - fetal demise

12. Regarding multiple gestation pregnancies, the following are true **EXCEPT**
- A. the reported incidence of spontaneous twinning is lowest in the Asian races
 - B. triplets are estimated to occur in 1 in 86² pregnancies in the USA
 - C. the incidence of monozygotic twins is unaffected by racial or familial factors
 - D. the overall incidence of multifetal gestation is unchanged
 - E. polyovular pregnancies are more frequent beyond the 2nd pregnancy
13. One of the following is a common neonatal respiratory problem associated with premature infants
- A. bronchopulmonary dysplasia
 - B. apnea
 - C. congenital pneumonia
 - D. pneumothorax
 - E. pneumomediastinum
14. One of the following drugs may cause pyloric stenosis if administered to a premature infant
- A. intravenous vitamin E
 - B. indomethacin
 - C. enteric gentamicin
 - D. prostaglandins
 - E. dexamethasone
15. The following factors pose a risk for poor academic performance of premature infants **EXCEPT**
- A. birthweight below 750 g
 - B. periventricular leukomalacia
 - C. antenatal exposure to magnesium sulfate
 - D. bronchopulmonary dysplasia
 - E. posthemorrhagic hydrocephalus
16. The incidence of all the following are increased in large for gestational age newborn **EXCEPT**
- A. cephalohematoma
 - B. hypocalcemia
 - C. hypoglycemia
 - D. congenital heart disease
 - E. developmental retardation
17. Sudden onset of hypotension in a very low birthweight (VLBW) infant suggests
- A. pneumothorax
 - B. necrotizing enterocolitis

- C. bacterial sepsis
 - D. myocarditis
 - E. hypoglycemia
18. Seizures beginning in the delivery room or shortly thereafter may be due to
- A. hypoxic-ischemic encephalopathy
 - B. unintentional injection of maternal local anesthetic into the fetus
 - C. intracranial hemorrhage
 - D. cerebral anomaly
 - E. hypoglycemia
19. After severe birth asphyxia, infants may have motor automatisms characterized by
- A. absence of oral-buccal-lingual movements
 - B. time-synchronized electroencephalographic discharges
 - C. significant cortical epileptic activity
 - D. good response to anticonvulsant therapy
 - E. a poor prognosis
20. Vomiting in the neonatal period is usually due to
- A. pyloric stenosis
 - B. milk allergy
 - C. overfeeding
 - D. stress ulcer
 - E. an inborn error of metabolism
21. Regarding cephalohematoma, all the following are true **EXCEPT**
- A. is a subperiosteal hemorrhage
 - B. it may extend across the midline and across suture lines
 - C. occur in 1-2% of live births
 - D. no discoloration of the overlying scalp
 - E. an underlying skull fracture may be associated with 10-25% of cases
22. Regarding intraventricular hemorrhage (IVH) of prematurity, the following are true **EXCEPT**
- A. it usually develops spontaneously
 - B. MRI is the preferred imaging technique for screening IVH
 - C. prophylactic administration of low-dose indomethacin reduces the incidence of severe IVH
 - D. majority of patients with IVH have no clinical symptoms
 - E. it may rarely manifest at birth

23. Early postnatal exposure to dexamethasone, within the 1st wk of life, in VLBW infants, is associated with the following **EXCEPT**
- A. metabolic derangements
 - B. poor growth
 - C. increased risk for sepsis
 - D. increased risk of spontaneous bowel perforation
 - E. cerebral palsy
24. The following are true regarding apnea **EXCEPT**
- A. apnea is a common problem in preterm infants
 - B. in term infants, apnea is always worrisome
 - C. obstructive apnea is characterized by absence of airflow but persistent chest wall motion
 - D. serious apnea is defined as cessation of breathing for longer than 20 sec
 - E. bradycardia follows the apnea by 1-2 sec in more than 95% of cases and is most often nodal
25. The risk for development of respiratory distress syndrome RDS increases with the following conditions **EXCEPT**
- A. prolonged rupture of membranes
 - B. maternal diabetes
 - C. multiple births
 - D. precipitous delivery
 - E. cold stress
26. The following are true regarding respiratory distress syndrome (RDS) **EXCEPT**
- A. signs of RDS usually appear within minutes of birth
 - B. in most cases, the symptoms and signs reach a peak within 7 days
 - C. apnea and irregular respirations are ominous signs
 - D. improvement is often heralded by spontaneous diuresis
 - E. mixed respiratory-metabolic acidosis may be seen
27. One of the following conditions can mimic RDS both clinically and radiographically
- A. persistent pulmonary hypertension
 - B. meconium aspiration syndrome
 - C. total anomalous pulmonary venous return
 - D. pulmonary lymphangiectasia
 - E. lobar emphysema
28. Administration of antenatal corticosteroids to women between 24 and 34 wk of gestation significantly reduces the following **EXCEPT**
- A. incidence and mortality of RDS

- B. postnatal growth
 - C. the overall neonatal mortality
 - D. need for and duration of ventilatory support
 - E. incidence of severe intraventricular hemorrhage
29. The basic defect requiring treatment in RDS is
- A. metabolic acidosis
 - B. circulatory insufficiency
 - C. hypothermia
 - D. inadequate pulmonary exchange of oxygen and carbon dioxide
 - E. electrolytes disturbance
30. Regarding CPAP, the following are true **EXCEPT**
- A. its prevents collapse of surfactant-deficient alveoli
 - B. its indicated, if oxygen saturation cannot be kept > 95%
 - C. its improves functional residual capacity FRC
 - D. its improves ventilation-perfusion matching
 - E. its reduces ventilatory needs
31. Prophylactic and rescue administrations of synthetic and natural surfactants have the following advantages **EXCEPT**
- A. reduces adverse outcomes
 - B. reduces neonatal mortality
 - C. decreases the risk for pneumothorax
 - D. decreases the risk for pulmonary interstitial emphysema
 - E. reduces bronco-pulmonary dysplasia BPD rates
32. Vitamin A supplementation given largely to infants < 1,000 g resulted in all the following **EXCEPT**
- A. a decrease in death
 - B. a decrease in bronco-pulmonary dysplasia BPD at 36 wk
 - C. less nosocomial sepsis
 - D. less retinopathy of prematurity
 - E. decreases the need for extracorporeal membrane oxygenation (ECMO)
33. Alkali therapy for the treatment of metabolic acidosis in RDS may result in all the following **EXCEPT**
- A. skin slough from infiltration
 - B. increased serum osmolarity
 - C. hypernatremia
 - D. hypocalcemia
 - E. hyperkalemia

34. Contributory factors for the development of broncho-pulmonary dysplasia (BPD) include the following **EXCEPT**

- A. immaturity
- B. dehydration during the 1st days of life
- C. chorioamnionitis
- D. symptomatic PDA
- E. malnutrition

35. Methylxanthines for the treatment of broncho-pulmonary dysplasia (BPD) have the following effects **EXCEPT**

- A. increase respiratory drive
- B. decrease apnea
- C. improve diaphragmatic contractility
- D. increase pulmonary vascular resistance PVR
- E. diuretic effects

36. One of the following is **FALSE** in the chest radiograph of transient tachypnea of the newborn (TTN)

- A. prominent pulmonary vascular markings
- B. fluid in the intralobar fissures
- C. overaeration
- D. flat diaphragms
- E. diffuse reticulogranular pattern

37. The following are true regarding meconium aspiration syndrome (MAS) **EXCEPT**

- A. (MAS) develops in 5% of meconium-stained infants
- B. 30% require mechanical ventilation
- C. 3-5% die
- D. overdistention of the chest may be prominent
- E. usually occurs in preterm or near-term infants

38. The following are predisposing factors for persistent pulmonary hypertension of the newborn (PPHN) **EXCEPT**

- A. anemia
- B. meconium aspiration syndrome
- C. early-onset sepsis
- D. hypoglycemia
- E. birth asphyxia

39. Associated anomalies have been reported in up to 30% of congenital diaphragmatic hernia cases; these include the following **EXCEPT**

- A. pulmonary hypoplasia

- B. CNS lesions
- C. esophageal atresia
- D. omphalocele
- E. cardiovascular lesions

40. Congenital diaphragmatic hernia (CDH) can be diagnosed on prenatal ultrasonography (between 16 and 24 wk of gestation) in > 50% of cases.

Findings on ultrasonography may include the following **EXCEPT**

- A. chest mass
- B. mediastinal shift
- C. gastric bubble
- D. oligohydramnios
- E. a liver in the thoracic cavity

41. A poor prognostic sign of congenital diaphragmatic hernia (CDH) is

- A. grunting
- B. use of accessory muscles
- C. early respiratory distress, within 6 hr of life
- D. cyanosis
- E. scaphoid abdomen

42. Eventration of the diaphragm may be associated with the following **EXCEPT**

- A. pulmonary hypoplasia
- B. pulmonary sequestration
- C. congenital heart disease
- D. chromosomal trisomies
- E. recurrent infections

43. All the following are true, regarding pathology of neonatal necrotizing enterocolitis (NEC) **EXCEPT**

- A. in fatal cases, gangrene may extend from the stomach to the rectum
- B. the greatest risk factor for NEC is prematurity
- C. in most situations, a pathogen is identified
- D. NEC is much less common in infants fed human milk
- E. NEC in term infants is often a "secondary" disease

44. Absolute indication for surgery in neonatal necrotizing enterocolitis (NEC) include

- A. positive result of abdominal paracentesis
- B. failure of medical management
- C. a single fixed bowel loop on radiographs
- D. abdominal wall erythema
- E. a palpable mass

45. The following factors are associated with decreased risk of significant jaundice **EXCEPT**
- A. gestational age ≥ 41 wk
 - B. male gender
 - C. exclusive bottle-feeding
 - D. black race
 - E. discharge from hospital after 72 hr
46. Jaundice, consisting of either indirect or direct bilirubin that is present at birth or appears within the 1st 24 hr of life may be due to all the following **EXCEPT**
- A. erythroblastosis fetalis
 - B. Crigler-Najjar syndrome
 - C. concealed hemorrhage
 - D. sepsis
 - E. congenital infections
47. A search to determine the cause of jaundice should be made in all the following conditions **EXCEPT**
- A. if it appears in the first 24-36 hr of life
 - B. if serum bilirubin is rising at a rate faster than 5 mg/dL/24 hr
 - C. if serum bilirubin is >12 mg/dL in a full-term infant
 - D. if jaundice persists after 10-14 days of life
 - E. if direct bilirubin fraction is >1 mg/dL at any time
48. Regarding breast milk jaundice, the following are true **EXCEPT**
- A. it develops in an estimated 2% of breast-fed term infants
 - B. maximal unconjugated bilirubin concentrations as high as 10-30 mg/dL reached during the 2nd-3rd week
 - C. jaundice may persist for 3-10 wk
 - D. phototherapy may be of benefit
 - E. kernicterus never occur
49. The therapeutic effect of phototherapy depends on the following factors **EXCEPT**
- A. distance between the lights and the infant
 - B. skin color
 - C. surface area of exposed skin
 - D. rate of hemolysis
 - E. in vivo metabolism and excretion of bilirubin
50. Early ultrasonographic signs of hydrops include
- A. double-bowel wall sign (bowel edema)
 - B. polyhydramnios

- C. ascites
- D. pleural effusions
- E. scalp edema

51. The blood smear of infant with hemolytic disease of the newborn (erythroblastosis fetalis) typically shows the following **EXCEPT**

- A. polychromasia
- B. increased reticulocyte count
- C. a marked increase in nucleated RBCs
- D. thrombocytopenia
- E. low white blood cell count

52. One of the following is **FALSE** in hemolytic disease of the newborn caused by blood group A and B incompatibility

- A. ABO incompatibility occurs in 20-25% of pregnancies
- B. A₁ is more antigenic than A₂
- C. natural antibodies against A and B factors are usually IgM antibodies that do not cross the placenta
- D. isoimmune hemolytic disease may be found in first-born infants irrespective of infant blood group
- E. most cases are mild

53. One of the following is **FALSE** in hemolytic disease of the newborn caused by Rh incompatibility

- A. infant is Rh Positive (D, sometimes C)
- B. never occur in first-born infant
- C. maternal antibody titers can help predict the severity of fetal disease
- D. severe anemia is frequent
- E. stillbirth/hydrops is frequent

54. One of the following is **FALSE** in plethora in the newborn infant (polycythemia)

- A. polycythemia is defined as a central Hct of 65% or higher
- B. peripheral (heelstick) Hct and Coulter counter results values are higher than central values
- C. the incidence of neonatal polycythemia is 8% in small for gestational age (SGA) infants
- D. many affected infants are asymptomatic
- E. neonatal RBCs have decreased deformability and filterability, which predispose to stasis in the microcirculation

55. Regarding hemorrhagic disease of the newborn, all the following are true **EXCEPT**

- A. age of onset is usually in 2-7 days old infants

- B. more frequent in breast-fed than in formula-fed infants
 - C. bleeding time is normal
 - D. oral vitamin K is as effective as intramuscular route in the prevention and treatment of the disease
 - E. prothrombin time (PT), and partial thromboplastin time (PTT) are prolonged
56. Apt test is useful for diagnosis of
- A. disseminated intravascular coagulopathy
 - B. swallowed blood syndrome
 - C. hemorrhagic disease of the newborn
 - D. neonatal thrombocytopenic purpura
 - E. congenital deficiency of factor VII
57. The normal cord at term is 55 cm long. Abnormally short cords are associated with the following **EXCEPT**
- A. fetal hypotonia
 - B. wrapping around fetal parts
 - C. uterine constraint
 - D. oligohydramnios
 - E. increased risk for complications of labor and delivery for both mother and infant
58. Surgical repair of umbilical hernia is advised in the following conditions **EXCEPT**
- A. if hernia persists to the age of 4-5 yr
 - B. causes symptoms
 - C. hernias that appear before the age of 6 mo
 - D. becomes strangulated
 - E. becomes progressively larger after the age of 1-2 yr
59. Generalized edema may be seen in the neonatal period with the following conditions **EXCEPT**
- A. congenital nephrosis
 - B. Turner syndrome
 - C. Hurler syndrome
 - D. hydrops fetalis
 - E. offspring of diabetic mothers
60. Diabetic mothers have a high incidence of all the following **EXCEPT**
- A. oligohydramnios
 - B. preeclampsia
 - C. pyelonephritis
 - D. preterm labor

E. chronic hypertension

61. In Infants of diabetic mothers, the following are true **EXCEPT**

- A. hypoglycemia develops in about 25-50% of infants of diabetic mothers
- B. increased weight of the placenta and all infant organs
- C. heart failure occurs in 5-10% of infants of diabetic mothers
- D. the incidence of congenital anomalies is increased threefold in infants of diabetic mothers
- E. lumbosacral agenesis is one of most common congenital anomalies

62. The following definitions are true **EXCEPT**

- A. a syndrome is defined as a pattern of multiple abnormalities that are related by pathophysiology and result from a common defined etiology
- B. dysplasia sequence is a poor organization of cells into tissues or organs
- C. an association refers to a nonrandom collection of malformations in which there is an unclear relationship among the malformations
- D. disruption sequence is a mechanical (uterine) forces that alter structure of intrinsically normal tissue
- E. malformation sequence is a single, local tissue morphogenesis abnormality that produces a chain of subsequent defects

63. Chromosomal causes of congenital malformations include

- A. Prader-Willi syndrome
- B. X-linked hydrocephalus
- C. Achondroplasia
- D. Treacher Collins syndrome
- E. Apert disease

64. One of the following definitions is **FALSE**

- A. brachydactyly=A condition of having short digits
- B. postaxial polydactyly=Extra finger or toe present on the medial side of the hand or foot
- C. clinodactyly=A medial or lateral curving of the fingers
- D. camptodactyly=Permanent flexion of one or more fingers
- E. syndactyly=Incomplete separation of the fingers

65. Intrauterine transplacental infections of significance to the fetus and/or newborn include the following **EXCEPT**

- A. hepatitis B virus (HBV)
- B. rubella
- C. toxoplasmosis
- D. Parvovirus B19

- E. varicella
66. Agents that commonly cause nosocomial/neonatal infection are
- A. group B streptococci GBS
 - B. coagulase-negative staphylococci
 - C. proteus
 - D. gonococci
 - E. chlamydiae
67. One of the following is **FALSE** regarding intrapartum antibiotics
- A. reduce vertical transmission of GBS
 - B. lessen neonatal morbidity after preterm rupture of membranes
 - C. prevent perinatal transmission of GBS
 - D. reduce the rates of late-onset GBS disease
 - E. has no effect on the rates of infection with non-GBS pathogens
68. In a cohort of 6,215 VLBW infants in the NICHD Neonatal Research Network, gram-positive agents were associated with which percent of cases of late-onset sepsis
- A. 90%
 - B. 70%
 - C. 50%
 - D. 30%
 - E. 10%
69. Osteitis may be a feature of one of the following transplacental infections
- A. cytomegalovirus
 - B. herpes simplex virus
 - C. varicella-zoster virus
 - D. rubella
 - E. T. gondii
70. Intracranial calcification may be a feature of one of the following transplacental infections
- A. cytomegalovirus
 - B. herpes simplex virus
 - C. varicella-zoster virus
 - D. syphilis
 - E. rubella
71. Limb hypoplasia may be a feature of the following transplacental infections
- A. cytomegalovirus
 - B. herpes simplex virus

- C. varicella-zoster virus
- D. HIV
- E. rubella

72. Common respiratory tract manifestations of neonatal bacterial infections is

- A. ethmoiditis
- B. otitis media
- C. mastoiditis
- D. retropharyngeal cellulitis
- E. empyema

73. Common skin and soft tissue manifestations of neonatal bacterial infections are

- A. facial cellulitis
- B. scalp abscess
- C. fasciitis
- D. breast abscess
- E. omphalitis

74. One of the following is not included in the definition of the systemic inflammatory response (SIRS) in neonates and pediatric patients

- A. temperature instability
- B. abnormal white blood cell (WBC) count
- C. respiratory dysfunction
- D. cardiac dysfunction
- E. perfusion abnormalities

75. Neonatal sepsis case fatality rate is highest for

- A. Staphylococcus—coagulase negative
- B. Group B streptococcus
- C. Escherichia coli
- D. Pseudomonas
- E. Candida albicans

76. Persistence of the umbilical cord beyond which time should prompt consideration of an underlying abnormality

- A. 10 days
- B. 20 days
- C. 30 days
- D. 40 days
- E. 50 days

77. The **MOST** important risk factor for necrotizing enterocolitis (NEC) in preterm infants is

- A. delivery by cesarean section
- B. breast feeding
- C. apgar score
- D. exposure to glucocorticoids during the first week of life
- E. gestational age and birth weight

78. Pneumatosis intestinalis is pathognomonic for

- A. Hirschsprung's disease
- B. necrotizing enterocolitis (NEC)
- C. pseudomembranous enterocolitis
- D. neonatal ulcerative colitis
- E. meconium ileus

79. The following factors suggest hemolytic disease as a cause of jaundice in the newborn **EXCEPT**

- A. bilirubin rise of >0.5 mg/dL/h
- B. reticulocytosis $>5\%$ at birth
- C. onset of jaundice before 24 hours of age
- D. significant decrease in hemoglobin
- E. failure of phototherapy to lower serum bilirubin levels

80. Which common sugar does the clinitest screen not detect?

- A. glucose
- B. fructose
- C. galactose
- D. sucrose
- E. lactose

81. The following are manifestations of hypocalcemia in the neonate **EXCEPT**

- A. jitteriness
- B. seizures
- C. carpopedal spasm
- D. high-pitched cry
- E. laryngospasm

82. When screening for intraventricular hemorrhage IVH, the best time to perform an ultrasound is

- A. first day of life
- B. second day of life
- C. third day of life

- D. fourth day of life
- E. fifth day of life

83. The antenatal administration of magnesium sulfate before preterm delivery has been suggested to

- A. reduce rates of necrotizing enterocolitis
- B. decrease the risk of cerebral palsy in surviving infants
- C. improve overall survival
- D. decrease respiratory distress syndrome in premature infants
- E. reduce rates of intraventricular hemorrhage

- 1.(D). Severe immaturity, respiratory distress syndrome, intraventricular hemorrhage, infection, necrotizing enterocolitis, and bronchopulmonary dysplasia (BPD) are major causes of mortality in preterms.
- 2.(C). Localized edema suggests a congenital malformation of the lymphatic system; when confined to one or more extremities of a female infant; it may be the initial sign of Turner syndrome.
- 3.(B). Persistently small fontanels suggest microcephaly, craniosynostosis, and congenital hyperthyroidism.
- 4.(E). Vitamin D deficiency rickets is associated with a large anterior fontanel.
- 5.(C). A cornea >1cm in diameter in a term infant (with photophobia and tearing) suggests congenital glaucoma and requires prompt ophthalmologic consultation.
- 6.(B). A single umbilical artery increases the risk for an occult renal anomaly.
- 7.(A). Amphetamines, bromocriptine, chloramphenicol, and methimazole are contraindicated in breast-feeding mother.
- 8.(E). Renal agenesis (Potter syndrome), Prune-belly syndrome, pulmonaryhypoplasia, and intestinal pseudo-obstruction are associated with oligohydramnios.
- 9.(B). Second-trimester screening (15-18 wk) of maternal serum α -fetoprotein (MSAFP) values is used to screen for open neural tube defects. About 90% of affected pregnancies can be detected by an elevated MSAFP value. Gastroschisis, omphalocele, congenital nephrosis, twins, and other abnormal conditions can also be identified. Low MSAFP is associated with incorrect gestational age estimates, trisomy 18 or 21, and intrauterine growth restriction.
- 10.(A). Earlier lung maturation may occur in the presence of severe premature separation of the placenta, premature rupture of the fetal membranes, narcotic addiction, or maternal hypertensive and renal vascular disease. A delay in pulmonary maturation may be associated with hydrops fetalis or maternal diabetes without vascular disease.
- 11.(A). Short cords and noncoiled cords occur with chromosome abnormalities and omphalocele.
- 12.(D). Although the incidence of spontaneous multifetal gestation has been stable over the years, the overall incidence of multifetal gestation is increasing as a result of treatment of infertility with ovarian stimulants (clomiphene, gonadotropins) and in vitro fertilization. Twins account for about 2.5% of births but about 20% of very low birthweight (VLBW) infants.
- 13.(B). Respiratory distress syndrome (hyaline membrane disease) and apnea are common neonatal respiratory problem associated with premature infants.

14.(D). Prostaglandins and erythromycin may cause pyloric stenosis if administered to a premature infant.

15.(C). Antenatal exposure to magnesium sulfate may have neuroprotective effects and may reduce the incidence of cerebral palsy in high-risk neonates.

16.(B). Large for gestational age LGA infants, regardless of their gestational age, have a higher incidence of birth injuries, such as cervical and brachial plexus injuries, phrenic nerve damage with paralysis of the diaphragm, fractured clavicles, cephalohematomas, subdural hematomas, and ecchymoses of the head and face. LGA infants are also at increased risk for hypoglycemia and polycythemia. The incidence of congenital anomalies, particularly congenital heart disease, is also higher in LGA infants than in term infants of normal weight. Intellectual and developmental retardation is statistically more common in high birthweight term and preterm infants than in babies of appropriate weight for gestational age

17.(A). Sudden onset of hypotension in a very low birthweight (VLBW) infant suggests pneumothorax, intraventricular hemorrhage, or subcapsular hepatic hematoma.

18.(B). Seizures beginning in the delivery room or shortly thereafter may be due to the unintentional injection of maternal local anesthetic into the fetus. Convulsions may also result from hyponatremia and water intoxication in the infant after the administration of large amounts of hypotonic fluid to the mother shortly before and during delivery.

19.(E). After severe birth asphyxia, infants may have motor automatisms characterized by oral-buccal-lingual movements, rotary limb activities (rowing, pedaling, and swimming), tonic posturing, or myoclonus. These motor activities are not usually accompanied by time-synchronized electroencephalographic discharges, may not signify cortical epileptic activity, respond poorly to anticonvulsant therapy, and are associated with a poor prognosis. Such automatisms may represent cortical depression that produces a brainstem release phenomenon or subcortical seizures.

20.(C). Vomiting during the 1st day of life suggests obstruction in the upper digestive tract or increased intracranial pressure. Roentgenographic studies are indicated when obstruction is suspected. Vomiting may also be a nonspecific symptom of an illness such as septicemia. It is a common manifestation of overfeeding, inexperienced feeding technique, or normal reflux and is rarely due to pyloric stenosis, milk allergy, duodenal ulcer, stress ulcer, an inborn error of metabolism (hyperammonemia, metabolic acidosis), or adrenal insufficiency.

21.(B). Cephalohematoma is a subperiosteal hemorrhage, hence always limited to the surface of one cranial bone, while caput succedaneum is a diffuse, sometimes ecchymotic, edematous swelling of the soft tissues of the scalp involving the area presenting during vertex delivery. It may extend across the midline and across suture lines.

22.(B). Ultrasonography is the preferred imaging technique for screening because it is noninvasive, portable, reproducible, and sensitive and specific for detection of IVH.

- 23.(E). Infants exposed to postnatal steroids after the 1st wk of life have an increased risk of cerebral palsy and developmental delay.
- 24.(E). Bradycardia follows the apnea by 1-2 sec in more than 95% of cases and is most often sinus, but on occasion it can be nodal. Vagal responses and, rarely, heart block are causes of bradycardia without apnea.
- 25.(A). The risk of RDS is reduced in pregnancies with chronic or pregnancy-associated hypertension, maternal heroin use, prolonged rupture of membranes, and antenatal corticosteroid prophylaxis.
- 26.(B). In most cases, the symptoms and signs reach a peak within 3 days, after which improvement is gradual.
- 27.(C). In the differential diagnosis, early-onset sepsis may be indistinguishable from RDS. In pneumonia manifested at birth, the chest roentgenogram may be identical to that for RDS. Maternal group B streptococcal colonization, identification of organisms on gram staining of gastric or tracheal aspirates or a buffy coat smear, and/or the presence of marked neutropenia may suggest the diagnosis of early-onset sepsis. Cyanotic heart disease (total anomalous pulmonary venous return) can also mimic RDS both clinically and radiographically. Echocardiography with color-flow imaging should be performed in infants who show no response to surfactant replacement, to rule out cyanotic congenital heart disease as well as ascertain patency of the ductus arteriosus and assess pulmonary vascular resistance (PVR).
- 28.(B). Postnatal growth is not adversely affected. Antenatal steroids do not increase the risk of maternal death, chorioamnionitis, or puerperal sepsis.
- 29.(D). The basic defect requiring treatment in RDS is inadequate pulmonary exchange of oxygen and carbon dioxide; metabolic acidosis and circulatory insufficiency are secondary manifestations.
- 30.(B). Warm humidified oxygen should be provided at a concentration initially sufficient to keep arterial oxygen pressure between 50 and 70 mm Hg (91-95% saturation) in order to maintain normal tissue oxygenation while minimizing the risk of oxygen toxicity. If oxygen saturation cannot be kept >90% at inspired oxygen concentrations of 40-70% or greater, applying CPAP at a pressure of 5-10 cm H₂O via nasal prongs is indicated and usually produces a rapid improvement in oxygenation. CPAP reduces collapse of surfactant-deficient alveoli and improves both FRC and ventilation-perfusion matching.
- 31.(E). The lack of reduction in BPD rates following surfactant replacement is probably, in part, due to the survival of infants with severe RDS who would have died without surfactant administration.
- 32.(E). Inhaled nitric oxide (iNO) decreases the need for extracorporeal membrane oxygenation (ECMO) in term and near-term infants with hypoxic respiratory failure or persistent pulmonary hypertension of the neonate.
- 33.(E). Alkali therapy may result in skin slough from infiltration, increased serum osmolarity, hyponatremia, hypocalcemia, hypokalemia, and liver injury when

concentrated solutions are administered rapidly through an umbilical vein catheter wedged in the liver.

34.(B). Overhydration during the 1st days of life may also contribute to the development of BPD. Vitamin A supplementation (5,000 IU intramuscularly 3 times/wk for 4 wk) in VLBW infants reduces the risk of BPD.

35.(D). Methylxanthines may also decrease PVR and increase lung compliance in infants with BPD, probably through direct smooth muscle relaxation.

36.(E). The distinctive features of transient tachypnea are rapid recovery of the infant and the absence of radiographic findings for RDS (hypoeration, diffuse reticulogranular pattern, air bronchograms) and other lung disorders.

37.(E). Meconium-stained amniotic fluid is found in 10-15% of births and usually occurs in term or post-term infants.

38.(A). Persistent pulmonary hypertension of the newborn (PPHN) occurs in term and post-term infants. Predisposing factors include birth asphyxia, MAS, early-onset sepsis, RDS, hypoglycemia, polycythemia, maternal use of nonsteroidal anti-inflammatory drugs with in utero constriction of the ductus arteriosus, maternal late trimester use of selective serotonin reuptake inhibitors, and pulmonary hypoplasia due to diaphragmatic hernia, amniotic fluid leak, oligohydramnios, or pleural effusions. PPHN is often idiopathic.

39.(A). Pulmonary hypoplasia and malrotation of the intestine are part of the lesion, not associated anomalies.

40.(D). Polyhydramnios

41.(C). Early respiratory distress, within 6 hr of life, is thought to be a poor prognostic sign.

42.(A). Congenital eventration may affect lung development, but it has not been associated with pulmonary hypoplasia.

43.(C). Clustering of cases suggests a primary role for an infectious agent. Various bacterial and viral agents, including *Escherichia coli*, *Klebsiella*, *Clostridium perfringens*, *Staphylococcus epidermidis*, astrovirus, norovirus, and rotavirus, have been recovered from cultures. Nonetheless, in most situations, no pathogen is identified.

44.(A). A surgeon should be consulted early in the course of treatment. Indications for surgery include evidence of perforation on abdominal roentgenograms (pneumoperitoneum) or positive result of abdominal paracentesis (stool or organism on Gram stain preparation from peritoneal fluid). Failure of medical management, a single fixed bowel loop on radiographs, abdominal wall erythema, and a palpable mass are relative indications for exploratory laparotomy.

45.(B). Male gender is a minor risk factor.

46.(B). Jaundice that first appears on the 2nd or 3rd day is usually physiologic but may represent a more severe form. Familial nonhemolytic icterus (Crigler-Najjar syndrome) and early-onset breast-feeding jaundice are seen initially on the 2nd or 3rd day.

47.(E). Direct bilirubin fraction is >2 mg/dL at any time.

- 48.(E). Although uncommon, kernicterus can occur in patients with breast milk jaundice.
- 49.(B). Dark skin does not reduce the efficacy of phototherapy.
- 50.(A). Early ultrasonographic signs of hydrops include organomegaly (liver, spleen, heart), the double-bowel wall sign (bowel edema), and placental thickening. Progression to polyhydramnios, ascites, pleural or pericardial effusions, and skin or scalp edema may then follow.
- 51.(E). The white blood cell count is usually normal but may be elevated.
- 52.(D). Although antibodies against A and B factors occur without previous immunization ("natural" antibodies), they are usually IgM antibodies that do not cross the placenta. However, IgG antibodies to A antigen may be present and these do cross the placenta, so A-O isoimmune hemolytic disease may be found in first-born infants.
- 53.(B). 5% of first-born infants may be affected.
- 54.(B). Peripheral (heelstick) Hct values are higher than central values, whereas Coulter counter results are lower than Hct values determined by microcentrifugation.
- 55.(D). Although oral vitamin K (birth, discharge, 3-4 wk: 1-2 mg) has been suggested as an alternative, oral vitamin K is less effective in preventing the late onset of bleeding due to vitamin K deficiency and thus cannot be recommended for routine therapy. The intramuscular route remains the method of choice.
- 56.(B). Apt devised the following test for differentiation of fetal from maternal blood: (1) Rinse a blood-stained diaper or some grossly bloody (red) stool with a suitable amount of water to obtain a distinctly pink supernatant hemoglobin solution; (2) centrifuge the mixture and decant the supernatant solution; (3) add 1 part of 0.25 N (1%) sodium hydroxide to 5 parts of the supernatant fluid. Within 1-2 min, a color reaction takes place: A yellow-brown color indicates that the blood is maternal in origin; a persistent pink indicates that it is from the infant. A control test with known adult or infant blood, or both, is advisable.
- 57.(B). Long cords (>70 cm) increase risk for true knots, wrapping around fetal parts (neck, arm), and/or prolapse. Straight untwisted cords are associated with fetal distress, anomalies, and intrauterine fetal demise.
- 58.(C). Most umbilical hernias that appear before the age of 6 mo disappear spontaneously by 1 yr of age. Even large hernias (5-6 cm in all dimensions) have been known to disappear spontaneously by 5-6 yr of age.
- 59.(B). Persistent edema of 1 or more extremities may represent congenital lymphedema (Milroy disease) or, in females, Turner syndrome.
- 60.(A). Diabetic mothers have a high incidence of polyhydramnios, preeclampsia, pyelonephritis, preterm labor, and chronic hypertension; their fetal mortality rate is greater than that of nondiabetic mothers, especially after 32 wk of gestation. Fetal loss throughout pregnancy is associated with poorly controlled maternal diabetes (especially ketoacidosis) and congenital anomalies.
- 61.(B). The probable pathogenic sequence is that maternal hyperglycemia causes fetal hyperglycemia, and the fetal pancreatic response leads to fetal hyperinsulinemia; fetal

hyperinsulinemia and hyperglycemia then cause increased hepatic glucose uptake and glycogen synthesis, accelerated lipogenesis, and augmented protein synthesis. Related pathologic findings are hypertrophy and hyperplasia of the pancreatic islet β cells, increased weight of the placenta and infant organs except for the brain, myocardial hypertrophy, increased amount of cytoplasm in liver cells, and extramedullary hematopoiesis. The incidence of congenital anomalies increased 3-fold in infants of diabetic mothers; cardiac malformations (ventricular or atrial septal defect, transposition of the great vessels, truncus arteriosus, double-outlet right ventricle, tricuspid atresia, coarctation of the aorta) and lumbosacral agenesis are most common.

62.(D). Disruption sequence is in utero tissue destruction after a period of normal morphogenesis while deformation sequence is a mechanical (uterine) force that alters structure of intrinsically normal tissue.

63.(A). X-linked hydrocephalus, Achondroplasia, Treacher Collins syndrome, and Apert disease are monogenic causes of congenital malformations.

64.(B). Postaxial polydactyly= Extra finger or toe present on the lateral side of the hand or foot. Preaxial polydactyly=Extra finger or toe present on the medial side of the hand or foot.

65.(A). Although HSV, HIV, hepatitis B virus (HBV), hepatitis C virus, and tuberculosis (TB) can each result in transplacental infection, the most common mode of transmission for these agents is intrapartum, during labor and delivery with passage through an infected birth canal (HIV, HSV, HBV), or postpartum, from contact with an infected mother or caretaker (TB) or with infected breast milk (HIV).

66.(B). Agents that commonly cause nosocomial infection are coagulase-negative staphylococci, gram-negative bacilli (*E. coli*, *Klebsiella pneumoniae*, *Salmonella*, *Enterobacter*, *Citrobacter*, *Pseudomonas aeruginosa*, *Serratia*), enterococci, *S. aureus*, and *Candida*. Viruses contributing to nosocomial neonatal infection include enteroviruses, CMV, hepatitis A, adenoviruses, influenza, respiratory syncytial virus (RSV), rhinovirus, parainfluenza, HSV, and rotavirus. Community-acquired pathogens such as *Streptococcus pneumoniae* may also cause infection in newborn infants after discharge from the hospital.

67.(D). Intrapartum chemoprophylaxis does not reduce the rates of late-onset GBS disease and has no effect on the rates of infection with non-GBS pathogens. Of concern is a possible increase in gram-negative infections (especially *E. coli*) in VLBW and possibly term infants in spite of a reduction in early GBS sepsis by intrapartum antibiotics.

68.(B). In a cohort of 6,215 VLBW infants in the NICHD Neonatal Research Network, gram-positive agents were associated with 70%, gram-negative with 18%, and fungi with 12% of cases of late-onset sepsis.

69.(D). Osteitis may be a feature of rubella and *T. pallidum*.

70.(A). Intracranial calcification may be a feature of CMV, HIV, toxoplasmosis, and *T. cruzi*.

71.(C).

72.(E). Pneumonia and empyema are common respiratory tract manifestations of neonatal bacterial infections.

73.(D). Breast abscess and impetigo are common skin and soft tissue manifestations of neonatal bacterial infections.

74.(B). Abnormal white blood cell (WBC) count or an increase in immature forms is included in the definition of the systemic inflammatory response SIRS in adults.

75.(D). Staphylococcus—coagulase negative (9.1%), Group B streptococcus (21.9%), Escherichia coli (34.0%), Pseudomonas (74.4%), and Candida albicans (43.9%).

76.(C). The umbilical cord generally dries up and sloughs by 2 weeks of life. Delayed separation can be normal up to 45 days. However, because neutrophilic and/or monocytic infiltration appears to play a major role in autodigestion, persistence of the cord beyond 30 days should prompt consideration of an underlying functional abnormality of neutrophils (leukocyte adhesion deficiency) or neutropenia.

77.(E). In an analysis of 15,072 neonates born at 98 centers over a 2-year period, the most important variables associated with NEC were gestational age and birth weight.

78.(E). Pneumatosis intestinalis can be seen in various other conditions, including Hirschsprung's disease, pseudomembranous enterocolitis, neonatal ulcerative colitis, and ischemic bowel disease. However, it is a characteristic finding in 85% of patients with NEC. Dark, concentric rings within the bowel wall represent hydrogen as a byproduct of bacterial metabolism

79.(B). Reticulocytosis (>8% at birth, >5% during first 2-3 days, >2% after first week).

80.(D). Reducing sugars (e.g., glucose, fructose, galactose, pentoses, and lactose) are detected, but sucrose is not a reducing sugar.

81.(C). Chvostek's sign (facial muscle twitching on tapping), and Trousseau's sign (carpopedal spasm) may be present, but more commonly these are absent during the neonatal period.

82.(D). In a series of infants studied by ultrasonography, approximately 50% had the onset of hemorrhage on the first day of life, 25% on the second day, and 15% on the third day. Thus, a single scan on the fourth day of life would be expected to detect >90% of IVHs.

83.(B). The antenatal administration of magnesium sulfate before preterm delivery has been suggested to decrease the risk of cerebral palsy in surviving infants but not improve overall survival. Prospective multicenter randomized trials are currently examining this potential benefit.

1. Adrenal production of androgen with development of underarm odor and faint genital hair (adrenarche) may occur as early as

- A. 4 yr
- B. 5 yr
- C. 6 yr
- D. 7 yr
- E. 8 yr

2. In males, the first visible sign of puberty is testicular enlargement, beginning as early as

- A. 8.5 yr
- B. 9.5 yr
- C. 10.5 yr
- D. 11.5 yr
- E. 12.5 yr

3. Penile growth occur during SMR

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

4. Peak growth occurs during SMR 4 when testis volumes reach approximately

- A. 3-4 cm³
- B. 5-6 cm³
- C. 7-8 cm³
- D. 9-10 cm³
- E. 11-12 cm³

5. Sperm may be found in the urine and nocturnal emissions may be noted at SMR

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

6. The first visible sign of puberty in females is the appearance of breast buds (thelarche) between

- A. 5 and 9 yr
- B. 6 and 10 yr
- C. 7 and 11 yr
- D. 8 and 12 yr
- E. 9 and 13 yr

7. Menses typically begins 2.5 yr after the onset of puberty at an average age of

- A. 9.5 yr
- B. 10.5 yr
- C. 11.5 yr
- D. 12.5 yr
- E. 13.5 yr

8. The belief that major structural brain development is completed in childhood is outdated. It is now clear that neuromaturation continues into the

- A. 2nd decade
- B. 3rd decade
- C. 4th decade
- D. 5th decade
- E. 6th decade

9. Urethritis is a sexually transmitted infections syndrome characterized by inflammation of the urethra, usually caused by an infectious etiology.

All of the following are true **EXCEPT**

- A. urethritis may present with meatal pruritus
- B. approximately 30-50% of males are asymptomatic
- C. on examination, the classic finding is erythema of the urethral meatus
- D. N. gonorrhoeae is one of the most commonly identified pathogen
- E. noninfectious causes of urethritis include urethral trauma or foreign body

10. In assessing the seriousness of adolescent drug abuse score, which of the following take +2 in the score?

- A. female sex
- B. age >15 years
- C. positive family history of drug abuse
- D. use before driving
- E. use of hallucinogens

11. Which of the following scores regarded as serious according to the assessment of seriousness of adolescent drug abuse score?

- A. 0-3
- B. 3-8
- C. 8-13
- D. 13-18
- E. 18-23

12. A 13-years-old male presented with delirium and mumbling speech, tachycardia, dry flushed skin, dilated pupils, myoclonus, slightly elevated temperature, urinary retention, and decreased bowel sounds, later on he developed seizure and dysrhythmia.

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

- A. antidepressant agents
- B. amphetamine
- C. barbiturates
- D. benzodiazepines
- E. edrophonium

13. Alcohol acts primarily as a central nervous system depressant. It produces all the following **EXCEPT**

- A. euphoria
- B. impaired short-term memory
- C. increased pain threshold
- D. hyperthermia
- E. respiratory depression

14. Smoking during pregnancy is associated with an average decrease in fetal weight by

- A. 100 g
- B. 200 g
- C. 300 g
- D. 400 g
- E. 500 g

15. Exposure to smokeless tobacco increases the users risk for

- A. lipoid pneumonia
- B. chronic cough
- C. cancers of the esophagus
- D. irritability
- E. decreased concentration

16. Lysergic acid diethylamide (LSD) is a very potent hallucinogen that is made from lysergic acid found in ergot, a fungus that grows on rye and other grains.

All the following are common somatic symptoms **EXCEPT**

- A. dizziness
- B. miosis
- C. nausea
- D. flushing
- E. hyperthermia

17. A 15-year-old female presented with delusions, paranoia, tachycardia, hypertension, hyperpyrexia, diaphoresis, piloerection, mydriasis, and hyperreflexia, later on she developed seizure, hypotension, and dysrhythmia.

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

- A. antidepressant agents
- B. amphetamine
- C. barbiturates
- D. benzodiazepines
- E. edrophonium

18. Which of the following is **TRUE** regarding pubertal gynecomastia?

- A. occurring in up to 20% of normal adolescent males
- B. onset typically is between 7 and 9 yr
- C. usually regresses within 6 mo
- D. surgery may be indicated in severe or persistent cases
- E. medical therapies have been approved for use in adolescents

19. Evaluation for pubertal delay in female should be done if she lacks any pubertal signs by the age of

- A. 12 yr
- B. 13 yr
- C. 14 yr
- D. 15 yr
- E. 16 yr

20. The age at which an evaluation for primary amenorrhea should be undertaken is

- A. 12 yr
- B. 13 yr
- C. 14 yr
- D. 15 yr
- E. 16 yr

21. Primary amenorrhea generally requires evaluation, if menstruation does not occur within

- A. 1 yr from the onset of puberty
- B. 2 yr from the onset of puberty

- C. 3 yr from the onset of puberty
- D. 4 yr from the onset of puberty
- E. 5 yr from the onset of puberty

22. The **MOST** commonly used method for contraception is

- A. condom
- B. withdrawal
- C. oral contraceptive pill
- D. spermicides
- E. intrauterine device

23. The contraceptive method with the highest failure rate even with perfect use is

- A. progestin releasing intra uterine device
- B. progestin-only injection
- C. combined oral contraceptives
- D. male condom
- E. spermicides

24. Depo-provera: an injectable progestin, medroxyprogesterone acetate available as a deep intramuscular injection (150 mg) or as a subcutaneous injection (104 mg) with typical use, the failure rate is 6%.

Potential adverse effects include

- A. heavy menstrual bleeding
- B. dysmenorrhea
- C. acne
- D. weight gain
- E. osteoporosis

- 1.(C). Adrenal production of androgen (chiefly dehydroepiandrosterone sulfate [DHEAS]) may occur as early as 6 yr of age, with development of underarm odor and faint genital hair (adrenarche).
- 2.(B), 3.(C), 4.(D), 5.(C). In males, the first visible sign of puberty and the hallmark of SMR 2 is testicular enlargement, beginning as early as 9.5 yr, followed by the development of pubic hair. This is followed by penile growth during SMR 3. Peak growth occurs when testis volumes reach approximately 9-10 cm³ during SMR 4. Under the influence of luteinizing hormone and testosterone, the seminiferous tubules, epididymis, seminal vesicles, and prostate enlarge. Sperm may be found in the urine by SMR 3; nocturnal emissions may be noted at this time as well.
- 6.(D), 7.(D). In females, typically the first visible sign of puberty and the hallmark of SMR 2 is the appearance of breast buds (thelarche), between 8 and 12 yr of age. A significant minority of females develops pubic hair (pubarche) prior to thelarche. Less visible changes include enlargement of the ovaries, uterus, labia, and clitoris, and thickening of the endometrium and vaginal mucosa. A clear vaginal discharge may be present prior to menarche (physiologic leukorrhea). Menses typically begins 2.5 yr after the onset of puberty, during SMR 3-4 (average age: 12.5 yr; normal range: 9-15 yr).
- 8.(B). This maturation is characterized by decreases in gray matter, increases in white matter, and an apparent increase in the efficiency of communication and connectivity between different brain regions.
- 9.(C). On examination, the classic finding is mucoid or purulent discharge from the urethral meatus. If no discharge is evident on exam, providers may attempt to express discharge by applying gentle pressure to the urethra from the base distally to the meatus 3-4 times.
- 10.(D). The score consist of 0, +1 and +2.
+2 include : use of drugs alone not in group, sad before drug use, recently poor school performance, use drugs before driving, history of accident, use drugs before and during school day and the use of Whiskey, opiates, cocaine, barbiturates.
- 11.(B). 0-3 less worrisome, 3-8 serious and 8-18 very serious.
- 12.(A). Patient in the question presented with anticholinergic syndrome and this syndrome can be caused by antihistamines, antiparkinsonian medication, atropine, scopolamine, amantadine, antipsychotic agents, antidepressant agents, antispasmodic agents, mydriatic agents, skeletal muscle relaxants, and many plants (notably jimson weed and Amanita muscaria).
- 13.(D). Hypothermia.

14.(B). Smoking during pregnancy is associated with an average decrease in fetal weight of 200 g; this decrease, added to the already smaller size of infants born to teenagers, increases perinatal morbidity and mortality.

15.(C). The 2 forms of smokeless tobacco (SLT) are “chew,” a leafy tobacco product sold in pouches, and “snuff,” a finely ground tobacco product sold in tins or packets. Exposure to SLT increases the users risk for oral cancers of the mouth, pharynx, larynx, and esophagus, as well as gum disease and nicotine addiction.

16.(B). Mydriasis.

17.(B). Patient in the question presented with sympathomimetic syndrome and this syndrome can be caused by cocaine, amphetamine, methamphetamine, and OTC decongestants (phenylpropanolamine, ephedrine, and pseudoephedrine). In caffeine and theophylline overdoses, similar findings, except for the organic psychiatric signs, result from catecholamine release.

18.(D). Occurring in up to 60% of normal adolescent males, onset typically is between 10 and 13 yr, usually regresses within 18-24 mo, no medical therapies for gynecomastia have been approved for use in adolescents.

19.(B).

20.(D), 21.(D). Age 15 yr is commonly considered the age at which an evaluation for primary amenorrhea should be undertaken. Evaluation should begin sooner if 4 yr have elapsed since the onset of puberty (breast development in most females).

22.(A). The most commonly used method is the condom, followed by withdrawal and then the pill.

23.(E). Failure rate are as follow: Progestin releasing intra uterine device 0.2%, progestin-only injection 0.2%, combined oral contraceptives 0.3%, male condom 2%, spermicides 18%.

24.(E). After 1 yr of use, 50% of users develop amenorrhea, which may be an added advantage for teens with heavy menstrual bleeding, dysmenorrhea, anemias, or blood dyscrasias. Although concern has been directed toward the potential for loss in bone mineral density in adolescents, thereby potentially increasing their risk for osteoporosis later in life, subsequent studies have found that bone density is recovered after discontinuation of the method and is considered safe for use in this population.

1. A 9-month-old boy presented with recurrent attacks of diarrhea, thrush, and failure to thrive shortly after birth. You suspect immune deficiency.

Of the following, the **MOST** common cause of this condition is

- A. hyper-IgE syndrome
- B. defect in phagocytic cells
- C. deficiencies in T-cell function
- D. defect in antibody production
- E. defect in complement proteins

2. A 17-month-old girl presented with history of recurrent attacks of pneumonia since the age of 8 months. You suspect a predominant B-cell defect.

Of the following, the **BEST** simple initial screening test is

- A. IgA measurement
- B. IgG measurement
- C. IgM measurement
- D. IgE measurement
- E. isohemagglutinins titer

3. A very high serum concentrations of one or more immunoglobulin classes, suggest all the following **EXCEPT**

- A. HIV infection
- B. chronic inflammation
- C. intestinal lymphangiectasia
- D. chronic granulomatous disease
- E. autoimmune lymphoproliferative syndrome

4. A 3-year-old boy presented with recurrent attacks of pneumonia and otitis media since the 1st birthday. Your diagnosis is X-linked agammaglobulinemia.

Of the following, the **MOST** likely offending organism is

- A. CMV
- B. Mycoplasma
- C. Pneumocystis jiroveci
- D. Staphylococcus aureus
- E. Streptococcus pneumonia

5. A 4.5-year-old boy presented with history of recurrent attack of secretory otitis media and purulent nasal discharge, Haemophilus influenza revealed by culture of ear discharge, on physical examination there is no tonsillar tissue and no palpable lymph nodes.

Of the following, the **MOST** appropriate test to confirm the diagnosis is measurement of

- A. flow cytometry
- B. IgA concentration
- C. isohemagglutinins titer
- D. IgG and IgM concentration
- E. antibodies to antigens of routine immunizations

6. A 3-year-old girl presented with recurrent attacks of chest infection and frequent episodes of chronic diarrhea, stool examination revealed Giardia lamblia and serum IgA level was very low.

Regarding this condition, all the following are true **EXCEPT**

- A. autosomal dominant inheritance
- B. phenotypically normal blood B cells
- C. administration of IVIG is not indicated
- D. environmental factors may trigger the disease
- E. it is associated with a celiac-like syndrome with dramatic response to gluten-free diet

7. A 2-year-old boy presented with failure to thrive, neonatal hypocalcemic seizure, and three attacks of sepsis. Examination reveals cleft palate and holo-systolic murmur.

Of the following, the genetic predisposition of this condition is

- A. microdeletions of chromosome 22q11.2
- B. interstitial deletion of the long arm of chromosome 15 (15q11-13)
- C. presence of two copies of paternally derived chromosome 11p15.5
- D. presence of mutated gene on long arm of chromosome 11 (11q22-23)
- E. presence of abnormal gene, on the proximal arm of the X chromosome at Xp11.22-11.23

8. Of the following, the findings that are **MOST** suggestive of DiGeorge syndrome are

- A. cataract and a congenital heart defect
- B. hypotonia and a congenital heart defect
- C. cutis aplasia and a congenital heart defect
- D. hypocalcemia and a congenital heart defect
- E. mongoloid slant to the eyes and a congenital heart defect

9. The **MOST** appropriate method to correct the immune deficiency in complete DiGeorge syndrome is

- A. vaccination
 - B. administration of IVIG
 - C. judicious use of antibiotics
 - D. transplantation of thymic tissue
 - E. transplantation of hematopoietic stem cells
10. In addition to infection and bleeding, the **MOST** common cause of death in Wiskott-Aldrich syndrome is
- A. renal failure
 - B. graft versus host disease
 - C. protracted bloody diarrhea
 - D. EBV-associated malignancies
 - E. development of autoimmune disease
11. The treatment of choice for a 3-year-old boy with Wiskott-Aldrich syndrome is
- A. splenectomy
 - B. use of killed vaccines
 - C. aggressive antibiotics
 - D. administration of IVIG
 - E. bone marrow or cord blood transplantation
12. All the following are features of ataxia-telangiectasia **EXCEPT**
- A. hepatic manifestation
 - B. high incidence of malignancy
 - C. chronic sinopulmonary disease
 - D. oculocutaneous telangiectasias
 - E. variable humoral and cellular immunodeficiency
13. The earliest clinical feature of ataxia-telangiectasia is
- A. ataxia
 - B. jaundice
 - C. inability to walk
 - D. ocular telangiectasias
 - E. cutaneous telangiectasias
14. Severe eosinophilia, defined as an absolute eosinophil count (AEC) more than
- A. 750 cells/ μ L
 - B. 1,000 cells/ μ L
 - C. 1,500 cells/ μ L
 - D. 5,000 cells/ μ L
 - E. 7,500 cells/ μ L

15. Which of the following statements is **TRUE** regarding diurnal variation of the absolute eosinophil count (AEC)?
- A. higher in the evening
 - B. lower in the afternoon
 - C. higher in the afternoon
 - D. lower in the early morning
 - E. higher in the early morning
16. Eosinophilia can be seen in all the following **EXCEPT**
- A. scabies
 - B. urticaria
 - C. corticosteroid therapy
 - D. Hodgkin disease
 - E. hypersensitivity drug reactions
17. The following conditions can cause eosinophilia **EXCEPT**
- A. malaria
 - B. filariasis
 - C. giardiasis
 - D. ascariasis
 - E. amebiasis
18. The highest white blood cells count is seen at
- A. birth
 - B. 12 hour of life
 - C. 1 week of life
 - D. 1 year of life
 - E. adolescence
19. Severe neutropenia is defined as absolute neutrophil count (ANC) less than
- A. 500/ μ L
 - B. 750/ μ L
 - C. 1000/ μ L
 - D. 1250/ μ L
 - E. 1500/ μ L
20. Chronic neutropenia by definition lasts longer than
- A. 1 mo
 - B. 3 mo
 - C. 6 mo
 - D. 9 mo
 - E. 12 mo

21. Chronic neutropenia may be induced by
- A. adenovirus
 - B. enteroviruses
 - C. cytomegalovirus
 - D. influenzas A and B
 - E. respiratory syncytial virus
22. A 4-year-old boy presented with high fever, decreased appetite, aphthous stomatitis, and cellulitis of the right calf muscle. His absolute neutrophil count (ANC) was 600/ μ L.
- The **MOST** likely organisms causing this condition are
- A. Staphylococcus aureus and Histoplasmosis
 - B. Staphylococcus aureus and rickettsial pox
 - C. Staphylococcus aureus and Cytomegalovirus
 - D. Staphylococcus aureus and Pneumocystis carinii
 - E. Staphylococcus aureus and gram-negative bacteria
23. Recurrent infections with neutropenia are a distinctive feature of
- A. Pompe disease
 - B. Hurler disease
 - C. Gaucher disease
 - D. Glycogen storage disease Ia (GSDIa)
 - E. Glycogen storage disease Ib (GSDIb)
24. All the following drugs can induce neutropenia **EXCEPT**
- A. penicillin
 - B. tetracycline
 - C. phenobarbital
 - D. phenothiazine
 - E. chloramphenicol
25. A 15-year-old boy with rheumatoid arthritis, on quinine for 6 months, presented with fever, aphthous stomatitis, and fatigue for the last 5 days. His absolute neutrophil count (ANC) was 700/ μ L.
- The **MOST** next appropriate therapeutic measure to be done when quinine withdrawal fails to improve neutropenia is
- A. administration of IVIG
 - B. aggressive use of antibiotics
 - C. transplantation of hematopoietic stem cells
 - D. administration of pulse methylprednisolone
 - E. administration of recombinant human granulocyte colony-stimulating factor

26. A 9-year-old boy with acute lymphoblastic leukemia on chemotherapy, presented with fever, aphthous stomatitis, and fatigue for the last 5 days. His absolute neutrophil count (ANC) was $450/\mu\text{L}$ and hemoglobin level was 10gm/dl .

The **NEXT** appropriate therapeutic measure is

- A. administration of IVIG
- B. transplantation of hematopoietic stem cells
- C. administration of pulse methylprednisolone
- D. aggressive use of broad-spectrum antibiotics
- E. administration of recombinant human granulocyte colony-stimulating factor

27. A 13-month-old girl presented with failure to thrive, repeated chest infection, and fatty offensive diarrhea. Hemoglobin level 8.8gm/dl , platelet count $150/\text{mm}^3$, white blood cell count $3.500/\text{mm}^3$, with absolute neutrophil count (ANC) $500/\mu\text{L}$.

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

- A. Kostmann disease
- B. cyclic neutropenia
- C. drug-induced neutropenia
- D. acute lymphoblastic leukemia
- E. Shwachman-Diamond syndrome

28. Leukemoid reactions (WBC count $> 50,000/\mu\text{L}$) are characterized by abundant

- A. blasts
- B. band form
- C. myelocytes
- D. promyelocytes
- E. metamyelocytes

29. Leukemoid reactions (WBC count $>50,000/\mu\text{L}$) are caused by all the following **EXCEPT**

- A. shigellosis
- B. septicemia
- C. salmonellosis
- D. chronic granulomatous disease
- E. leukocyte adhesion defect with infection

30. Shifting to the left (proportion of immature neutrophil cells $>5\%$) may occasionally be caused by all the following **EXCEPT**

- A. burns
- B. trauma
- C. hemorrhage
- D. acute hemolysis
- E. megaloblastic anemia

31. A 15-year-old boy presented with high fever, lymphadenopathy, hepatosplenomegaly, recurrent skin infections, silvery hair, and photophobia. Last year, he developed ataxia.

The **MOST** characteristic finding in this condition is

- A. neutropenia
- B. thrombocytopenia
- C. prolonged bleeding time
- D. prolonged prothrombine time
- E. prolonged partial prothrombine time

32. In patient with Chédiak-Higashi syndrome, the only curative therapy is hematopoietic stem cell transplantation which correct all the following **EXCEPT**

- A. neuropathy
- B. immunologic function
- C. hematopoietic function
- D. natural killer cell deficiency
- E. conversion to the accelerated phase

33. A 3-year-old boy presented with recurrent subcutaneous abscesses, lymphadenitis, osteomyelitis, and family history of recurrent infections.

The hallmark of this condition is

- A. hypocalcemia
- B. atopic dermatitis
- C. thrombocytopenia
- D. granuloma formation
- E. prolonged bleeding time

34. The **MOST** common pathogen in chronic granulomatous disease (CGD) is

- A. *S. aureus*
- B. *Aspergillus*
- C. *Salmonella*
- D. *Mycobacterium*
- E. *Candida albicans*

35. The diagnosis of chronic granulomatous disease CGD is **MOST** often made by

- A. DNA analysis
- B. flow cytometry
- C. neutrophil G6PD assay
- D. erythrocyte G6PD assay
- E. nitroblue tetrazolium dye test

36. To reduce the number of bacterial infection in patients with chronic granulomatous disease (CGD), they should be given daily

- A. penicillin
- B. cloxacillin
- C. interferon- γ
- D. itraconazole
- E. trimethoprim-sulfamethoxazole

37. In chronic granulomatous disease (CGD), the best method to detect and follow up deep-seated infection is

- A. ESR
- B. blood culture
- C. WBC count
- D. C-reactive protein
- E. immature to mature neutrophil ratio

38. All the following are characteristic in properdin deficiency **EXCEPT**

- A. dermal vasculitis
- B. male predominance
- C. systemic lupus erythematosus
- D. susceptibility to *N. meningitidis* meningitis
- E. normal serum hemolytic complement activity

39. All the following matching about genetic deficiencies of plasma complement components and associated clinical findings are true **EXCEPT**

- A. C4 ----- discoid lupus erythematosus
- B. C1q----- systemic lupus erythematosus
- C. C1 INH ----- hereditary angioedema
- D. factor H---- atypical hemolytic-uremic syndrome
- E. properdin ----- *N. meningitidis* meningitis

40. The complement component of both the classical and alternative pathways is

- A. C2
- B. C3
- C. C4
- D. C5
- E. C6

41. All the following conditions can induce lymphopenia **EXCEPT**

- A. sepsis
- B. typhoid
- C. brucellosis

- D. corticosteroid use
- E. cartilage-hair hypoplasia

- 1.(C). Children with defects in antibody production, phagocytic cells, or complement proteins have recurrent infections with encapsulated bacteria and may grow and develop normally despite their recurring infections, unless they develop bronchiectasis from repeated lower respiratory tract bacterial infections or persistent enteroviral infections of the central nervous system. By contrast, patients with deficiencies in T-cell function usually develop opportunistic infections or serious illnesses from common viral agents early in life, and they fail to thrive.
- 2.(A). If the IgA level is normal, selective IgA deficiency, which is the most common B-cell defect, is excluded, as are most of the permanent types of hypogammaglobulinemia, as IgA is usually very low or absent in those conditions. If IgA is low, IgG and IgM should also be measured.
- 3.(C). There is a possible loss of immunoglobulins through the urinary or gastrointestinal tracts (nephrotic syndrome, protein-losing enteropathies, intestinal lymphangiectasia).
- 4.(E). They acquire infections with extracellular pyogenic organisms, such as *Streptococcus pneumoniae* and *Haemophilus influenzae*, unless they are given prophylactic antibiotics or immunoglobulin therapy.
- 5.(A). The diagnosis of X-linked agammaglobulinemia (XLA) should be suspected if lymphoid hypoplasia is found on physical examination (minimal or no tonsillar tissue and no palpable lymph nodes). Flow cytometry is an important test to demonstrate the absence of circulating B cells, which will distinguish this disorder from common variable immunodeficiency, the hyper-IgM syndrome, and transient hypogammaglobulinemia of infancy.
- 6.(E). An isolated absence or near absence (<10 mg/dL) of serum and secretory IgA is the most common well-defined immunodeficiency disorder. IgA deficiency is associated with a celiac-like syndrome, which may or may not respond to a gluten-free diet.
- 7.(A). A. DiGeorge syndrome, B. Prader-Willi syndrome, C. Beckwith-Wiedemann syndrome, D. Wiskott-Aldrich syndrome, and E. ataxia-telangiectasia.
- 8.(D). Digeroge syndrome; associated with congenital heart disease (conotruncal, atrial, and ventricular septal defects). The diagnosis is often first suggested by hypocalcemic seizures during the neonatal period.
- 9.(D).
- 10.(D). Wiskott-Aldrich syndrome, an X-linked recessive syndrome, is characterized by atopic dermatitis, thrombocytopenic purpura with normal-appearing megakaryocytes but small defective platelets, and undue susceptibility to infection. Survival beyond the

teens is rare; infections, bleeding, and EBV-associated malignancies are major causes of death.

11.(E). Treatment; Good supportive care includes appropriate nutrition, routine IVIG, use of killed vaccines, aggressive management of eczema and associated cutaneous infections, platelet transfusion for serious bleeding episodes, splenectomy if a transplant is not going to be done, and high-dose IVIG with systemic steroids for autoimmune complications. Bone marrow or cord blood transplantation is the treatment of choice and is usually curative.

12.(A). The most prominent clinical features are progressive cerebellar ataxia, oculocutaneous telangiectasias, chronic sinopulmonary disease, a high incidence of malignancy, and variable humoral and cellular immunodeficiency.

13.(A). Ataxia typically becomes evident soon after these children begin to walk and progresses until they are confined to a wheelchair, usually by the age of 10-12 yr. The telangiectasias begin to develop at 3-6 yr of age.

14.(D). Moderate (AEC 1,500-5,000 cells/ μ L) and severe (AEC >5,000 cells/ μ L).

15.(E). The absolute eosinophil count (AEC) is calculated as the white blood cell count/ μ L $\times$ percent of eosinophils, it is usually <450 cells/ μ L and varies diurnally, with eosinophil numbers higher in the early morning and diminishing as endogenous glucocorticoid levels rise.

16.(C). Exogenous and endogenous glucocorticoid suppresses absolute eosinophil count.

17.(C). Eosinophilia are caused by tissue-invasive as helminth infections; ascariasis, filariasis, amebiasis, malaria, and scabies whereas *enterobius vermicularis* and *Giardia lamblia* are solely intraluminal in gastrointestinal tract.

18.(A). For newborns, the mean WBC count at birth is high, followed by a rapid fall beginning at 12 hr through the 1st wk of life. Thereafter, values are stable until 1 yr of age, after which a slow steady decline in the WBC count continues throughout childhood until adult values are reached during adolescence.

19.(A). Mild neutropenia, is an absolute neutrophil count (ANC) of 1,000-1,500/ μ L. Moderate neutropenia, is an absolute neutrophil count (ANC) of 500- 1,000/ μ L. Severe neutropenia, is an absolute neutrophil count (ANC) <500/ μ L.

20.(B). Chronic neutropenia by definition lasts longer than 3 months and arises from reduced production, increased destruction or excessive splenic sequestration of neutrophils.

21.(C). Chronic neutropenia often accompanies infection with Epstein-Barr virus, cytomegalovirus, or HIV while the other distracters cause acute neutropenia.

22.(E). Infections commonly associated with neutropenia include cellulitis, furunculosis, perirectal inflammation, colitis, sinusitis, and otitis media, as well as more serious infections such as pneumonia, deep tissue abscess, and sepsis.

23.(E). Recurrent infections with neutropenia are a distinctive feature of glycogen storage disease (GSD) type Ib. As in classic von Gierke disease (GSD Ia), glycogen storage in GSD Ib causes massive hepatomegaly and severe growth retardation.

24.(B). The process of neutropenia likely arises from effects of drugs, such as propylthiouracil or penicillin, that act as haptens to stimulate antibody formation, or drugs, such as quinine, that induce immune complex formation. Late-onset neutropenia can occur after rituximab therapy. Idiosyncratic reactions, for example to chloramphenicol, are unpredictable with regard to dose or duration of use. Hypersensitivity reactions are rare and may involve arene oxide metabolites of aromatic anticonvulsants (phenytoin or phenobarbital).

25.(E).

26.(D). A decline in the WBC count typically occurs 7-10 days after administration of the anticancer drug and may persist for 1-2 wk. Patients with chemotherapy/ radiation-related neutropenia and fever must be treated aggressively with broad-spectrum antibiotics.

27.(E). Shwachman-Diamond syndrome (SDS) is an autosomal recessive disorder classically characterized by neutropenia, pancreatic insufficiency, and short stature with skeletal abnormalities. The initial symptoms are usually steatorrhea and failure to thrive because of malabsorption, which usually develops by 4 mo of age, although the gastrointestinal symptoms may be subtle in some patients and go unrecognized. Virtually all patients with SDS have neutropenia, with the ANC periodically $<1000/\mu\text{L}$.

28.(B). Leukemoid reactions are usually neutrophilic, and unlike true leukemia, show only small proportions of immature myeloid cells, consisting primarily of band forms, occasional metamyelocytes, and progressively rarer myelocytes, promyelocytes, and blasts.

29.(D).

30.(E). Higher degrees of left shift with more immature neutrophil precursors are indicative of serious bacterial infections and may be a direct sign of depletion of the bone marrow reserve pool of neutrophils. Marked left shift may occasionally be encountered with trauma, burns, surgery, acute hemolysis, or hemorrhage.

31.(C). Patients with Chédiak-Higashi syndrome (CHS) have prolonged bleeding times with normal platelet counts, resulting from impaired platelet aggregation associated with a deficiency of the dense granules containing adenosine diphosphate and serotonin.

32.(A).

33.(D). Granuloma formation and inflammatory processes are a hallmark of chronic granulomatous disease CGD and may be the presenting symptoms that prompt testing for CGD if they cause pyloric outlet obstruction, bladder outlet or ureter obstruction, or rectal fistulas and granulomatous colitis simulating Crohn disease.

34.(A). The most common pathogen is *S. aureus*, although any catalase-positive microorganism may be involved.

35.(B). The diagnosis is most often made by performing flow cytometry using dihydrorhodamine 123 (DHR) to measure oxidant production through its increased fluorescence when oxidized by H_2O_2 .

36.(E). A placebo-controlled study found that interferon- γ significantly reduces the number of hospitalizations and serious infections. Itraconazole administered prophylactically reduces the frequency of fungal infections.

37.(A). The erythrocyte sedimentation rate (ESR) can be quite helpful. If the child does not have a deep-seated infection, the ESR will be normal or will normalize within several days with standard management. However, if it does not, a search for deep tissues is warranted, as is consideration of empiric antibiotics.

38.(C). Typical discoid lupus erythematosus is common.

39.(A). Systemic lupus erythematosus (SLE) is very common in C4 deficiency.

40. (B).

41.(C). Chronic bacterial infections such as tuberculosis and brucellosis may lead to a sustained lymphocytosis.

PART XV

Allergic Disorders QUESTIONS

HASANEIN GHALI

1. The recognition of allergic rhinitis as a major chronic respiratory disease of children rests largely on all the following features **EXCEPT**
 - A. high prevalence
 - B. effect on quality of life
 - C. school performance
 - D. other comorbidities
 - E. presentation in infancy
2. The diagnosis of allergic rhinitis is established by the time the child reaches age of
 - A. 2 yr
 - B. 4 yr
 - C. 6 yr
 - D. 8 yr
 - E. 10 yr
3. A significant risk factor in the development of allergic rhinitis in children is
 - A. positive family history
 - B. serum IgA higher than 100 iu/ml
 - C. alcoholic mother
 - D. diabetic mother
 - E. father with renal disease
4. The critical period for the development of allergic rhinitis exists when the genetically susceptible individual at greatest risk of sensitization in
 - A. early adolescence
 - B. late adolescence
 - C. school age
 - D. toddlerhood
 - E. early infancy
5. The occurrence of 3 or more episodes of rhinorrhea in the first year of life is associated with allergic rhinitis (AR) at age of
 - A. 11 yr
 - B. 9 yr
 - C. 7 yr
 - D. 5 yr

E. 3 yr

6. Early introduction of all the following decrease the risk of asthma and allergic rhinitis

EXCEPT

- A. rye
- B. fish
- C. egg
- D. meat
- E. wheat

7. The exposure to which of the following early in childhood protects against the development of atopy

- A. smoking
- B. alcohol
- C. rodents
- D. dogs
- E. indoor allergen

8. The main differentiating item between intermittent versus persistent allergic rhinitis is

- A. school performance
- B. troublesome symptoms
- C. daily activities
- D. number of attacks per week
- E. sleep pattern

9. Allergic shiners represent

- A. an upward rubbing of the nose with an open palm
- B. continuous open- mouth breathing
- C. dark circles under the eyes
- D. transverse crease of the nose
- E. conjunctival edema and itching

10. Allergic rhinitis may be complicated by secondary bacterial infection as sinusitis.

Of the following, the **MOST** likely clue to infection is

- A. hyperemia
- B. swollen turbinates
- C. thick nasal secretions
- D. bluish mucus membranes
- E. edematous mucus membranes

11. Causes of hormonal rhinitis include all the following **EXCEPT**

- A. hypothyroidism
 - B. exercise
 - C. cancer
 - D. vaculitides
 - E. systemic lupus erythematosus
12. The reported rate of remission of allergic rhinitis among children is about
- A. 1-10%
 - B. 11-20%
 - C. 21-30%
 - D. 31-40%
 - E. 41-50%
13. What percentage of allergic rhinitis patients has allergic conjunctivitis?
- A. >30%
 - B. >40%
 - C. >50%
 - D. >60%
 - E. >70%
14. The percentage of allergic rhinitis in asthmatic patients is 78%, while the percentage of asthma in patients with allergic rhinitis is
- A. 18%
 - B. 28%
 - C. 38%
 - D. 48%
 - E. 58%
15. To avoid false-negative skin test results in atopic host, most sedating antihistamine should be withheld for 3-4 days, and nonsedating antihistamines for 5-7 days, while montelukast should be withheld for
- A. 1 day
 - B. 3 day
 - C. 5 day
 - D. 7 day
 - E. 9 day
16. The best method for detection of allergen-specific IgE is
- A. skin tests
 - B. serum immune assay for specific IgE
 - C. nasal smear study
 - D. eosinophilia in blood film

- E. total serum IgE concentration
17. Second generation antihistamines are preferable to first generation antihistamines because they have
- A. longer action
 - B. less sedation
 - C. more potency
 - D. more palatable effect
 - E. more shelf life
18. The **MOST** effective therapy for persistent allergic rhinitis is
- A. oral desloratadine
 - B. oral montelukast
 - C. subcutaneous omalizumab
 - D. nasal budesonide spray
 - E. nasal ipratropium bromide spray
19. Approximately 80% of all asthmatic patients report disease onset prior to the age of
- A. 2 yr
 - B. 4 yr
 - C. 6 yr
 - D. 8 yr
 - E. 10 yr
20. Of the following, the major risk factor for persistent asthma is
- A. eczema
 - B. allergic rhinitis
 - C. wheezing apart from colds
 - D. food allergen sensitization
 - E. $\geq 4\%$ peripheral blood eosinophils
21. Of the following, the strongest identifiable factor for the persistence of childhood asthma is
- A. allergy
 - B. male gender
 - C. low birth weight
 - D. parental asthma
 - E. lower respiratory tract infection
22. The following actions may help reduce the likelihood of asthma development **EXCEPT**
- A. healthy diet

- B. immunization
- C. active lifestyle
- D. avoidance of tobacco smoking
- E. prolonged breastfeeding > 4 months

23. Recurrent coughing and wheezing occurs in 35% of preschool-age children.

Those who continue to have persistent asthma into later childhood are approximately

- A. one-fifth
- B. one-fourth
- C. one-third
- D. one-half
- E. two-thirds

24. Indications of severe exacerbation of asthma include the following **EXCEPT**

- A. breathlessness
- B. accessory muscles use
- C. labored breath
- D. PEF or FEV1 value <70% of personal best values
- E. mental status changes

25. Spirometry is a helpful objective measure of airflow limitation; it depends on patient's ability to properly perform a full, forceful, and prolonged expiratory maneuver.

Spirometry usually feasible in children

- A. ≥4 yr of age
- B. ≥6 yr of age
- C. ≥8 yr of age
- D. ≥10 yr of age
- E. ≥12 yr of age

26. The **MOST** vital initial treatment in the management of severe asthma exacerbations is

- A. supplemental oxygen
- B. inhaled β -agonist
- C. intramuscular injection of epinephrine
- D. inhaled ipratropium
- E. intramuscular injection of β -agonist

27. A patient with exacerbation of asthma being handled and managed in the ER may be discharged home if all the following parameters are met **EXCEPT**

- A. normal physical activity
- B. PEF >70% of predicted or personal best

- C. exercise tolerance
- D. oxygen saturation >92% while the patient is breathing room air for 4 hr
- E. no more use of accessory muscles

28. All the following are considered as risk factors for asthma morbidity and mortality **EXCEPT**

- A. poverty
- B. air pollution exposure
- C. female gender
- D. poor response to systemic steroid therapy
- E. sudden asphyxia episodes

29. The best “rescue” medication in the treatment of acute asthma symptoms is

- A. oral SABA
- B. inhaled SABA
- C. oral corticosteroid
- D. inhaled ipratropium
- E. inhaled corticosteroid

30. The best treatment option for step -6- severe persistent asthma in a 6-year-old boy is

- A. medium dose inhaled corticosteroids with long acting B-agonists
- B. high dose inhaled corticosteroids with leukotriene receptor antagonist
- C. low dose inhaled corticosteroids with leukotriene receptor antagonist
- D. high dose inhaled corticosteroids with long acting B-agonist and oral corticosteroids
- E. high dose inhaled corticosteroids with long acting B-agonist and oral corticosteroids along with omalizumab therapy

31. A common finding of chest radiograph in a child with asthma is

- A. peribronchial thickening
- B. atelectasis
- C. pneumothorax
- D. bronchiectasis
- E. pneumomediastinum

32. Bronchodilator response to an inhaled β -agonist (e.g., albuterol) is greater in asthmatic patients than nonasthmatic persons, the rate of improvement in FEV1 consistent with asthma is

- A. $\geq 10\%$
- B. $\geq 12\%$
- C. $\geq 14\%$

- D. $\geq 16\%$
- E. $\geq 18\%$

33. The diurnal variation in Peak Expiratory Flow (PEF) that is consistent with asthma is more than

- A. 10%
- B. 20%
- C. 30%
- D. 40%
- E. 50%

34. In emergency department, the patient may be discharged to home if there is symptomatic improvement, normal physical findings, PEF $>70\%$ of predicted or personal best, and oxygen saturation $>92\%$ in room air for 4hr.

Of the following, the **MOST** likely discharge medication used is

- A. inhaled β -agonist only
- B. oral corticosteroid only
- C. inhaled corticosteroid only
- D. inhaled β -agonist plus oral corticosteroid
- E. oral β -agonist plus inhaled corticosteroid

35. All the following are a recognized features of a “well-controlled” asthma in a 7-year-old boy **EXCEPT**

- A. FEV1:FVC ratio $>80\%$
- B. FEV1of $>80\%$ of predicted
- C. daytime symptoms ≤ 2 days/wk
- D. ≥ 4 exacerbations in the past year
- E. need a rescue bronchodilator ≤ 2 days/wk

36. Use of a stoplight zone system (green, yellow, red) tailored to each child’s “personal best” PEF values can optimize effectiveness and interest.

Of the following, the PEF value in yellow zone is

- A. $<10\%$
- B. 10-30%
- C. 30-50%
- D. 50-80%
- E. 80-100%

37. All levels of persistent asthma should be treated with daily medications to improve long-term control.

All the following are long-term controller medications **EXCEPT**

- A. albuterol

- B. omalizumab
- C. montelukast
- D. inhaled corticosteroid
- E. sustained-release theophylline

38. Adverse effects of frequently administered β -agonist therapy in asthma include all the following **EXCEPT**

- A. tremor
- B. irritability
- C. tachycardia
- D. hypokalemia
- E. mouth dryness

39. Atopic dermatitis affects many children worldwide, the exact percentage approximating

- A. 5-10%
- B. 10-30%
- C. 30-50%
- D. 50-70%
- E. 70-90%

40. The hallmark of atopic dermatitis is

- A. intense pruritus
- B. cutaneous reactivity
- C. lichenification
- D. fibrotic papules
- E. dry skin

41. Atopic dermatitis typically begins in

- A. infancy
- B. toddlerhood
- C. preschooler age
- D. school age
- E. adolescence

42. The cardinal feature of atopic dermatitis is

- A. skin rash
- B. lichenification
- C. fibrotic papules
- D. intense pruritus
- E. dry skin

43. All the following are triggers for pruritus in atopic dermatitis **EXCEPT**

- A. grass
 - B. tree nuts
 - C. high humidity
 - D. herpes simplex
 - E. excessive sweating
44. In infancy, atopic dermatitis is usually acute and spares the
- A. extensor surfaces of the extremities
 - B. forehead
 - C. scalp
 - D. diaper area
 - E. cheeks
45. Older children with chronic atopic dermatitis have lichenification and a tendency to be localized to
- A. flexural folds of the extremities
 - B. forehead
 - C. scalp
 - D. diaper area
 - E. cheeks
46. The following are the major clinical features of atopic dermatitis (AD) **EXCEPT**
- A. pruritus
 - B. family history
 - C. relapsing dermatitis
 - D. facial eczema in infants
 - E. extensor eczema in adolescents
47. A high index of suspicion of which of the following conditions is to be undertaken in a patient with atopic dermatitis and failure to thrive
- A. Wiscott-Aldrich syndrome
 - B. severe combined immune deficiency
 - C. Histiocytosis
 - D. hyper IgE syndrome
 - E. chronic granulomatous disease
48. One of the following metabolic conditions is not deemed in the differential diagnosis of atopic dermatitis
- A. zinc deficiency
 - B. pyridoxine deficiency
 - C. cobalamin deficiency
 - D. niacin deficiency

E. phenylketonuria

49. The first line therapy of atopic dermatitis (AD) is

- A. moisturizers
- B. cyclosporine
- C. antihistamine
- D. tar preparations
- E. topical corticosteroids

50. Prevention of atopic dermatitis in infancy includes the following measures **EXCEPT**

- A. breast feeding
- B. feeding with a hypoallergenic hydrolyzed formula
- C. use of probiotics
- D. elimination of implicated food allergen from the mother diet
- E. use of special type napkins

51. Which of the following vitamin deficiencies often accompanies severe atopic dermatitis?

- A. E
- B. C
- C. A
- D. D
- E. B12

52. Predictive factors of a poor prognosis for atopic dermatitis includes all the following **EXCEPT**

- A. widespread atopic dermatitis in childhood
- B. filaggrin gene null mutations
- C. concomitant allergic rhinitis and asthma
- D. family history of atopic dermatitis in parents or siblings
- E. late age at onset of atopic dermatitis

53. The best choice of antibiotic in treating localized impetiginous lesions in patients with atopic dermatitis is

- A. oral erythromycin
- B. oral cephalexin
- C. topical mupirocin
- D. topical ampicillin
- E. topical gentamycin

54. IgE anti-bodies against *Malassezia furfur* have been found in patients with atopic dermatitis of

- A. head and neck
- B. hands and arms
- C. legs and feet
- D. upper chest
- E. lower abdomen

55. One of the following statements is **TRUE** regarding skin tests in identifying food allergies in patients with atopic dermatitis

- A. negative skin and blood test results for allergen-specific ige have a low predictive value for excluding suspected allergens
- B. positive results of skin or blood tests using foods often correlate with clinical symptoms and no need to be confirmed with controlled food challenges
- C. extensive elimination diets are commonly required
- D. even with multiple majority of patients react to more than 3 foods
- E. potential allergens can be identified by a careful history and performing selective skin prick tests or in vitro blood testing for allergen- specific IgE

56. One of the following medications used in treatment of atopic dermatitis should be discontinued after failure to achieve good results within 4-6 weeks

- A. methotrexate
- B. azathioprine
- C. cyclosporine
- D. mycophenolate mofetil
- E. omalizumab

57. Systemic corticosteroids are rarely indicated in the treatment of atopic dermatitis because

- A. toxic side effects after long term use
- B. rebound flare after therapy discontinuation
- C. cannot do more than what topical can
- D. tapering is required even after short term use
- E. antimetabolites can do better in modifying disease course

58. The least potent topical steroid used in treatment of atopic dermatitis is

- A. fluticasone
- B. desonide
- C. betamethasone dipropionate
- D. hydrocortisone
- E. clobetasol propionate

59. Which topical steroid is preferable to use for treatment of atopic dermatitis in the face?

- A. fluticasone 0.005%
- B. desonide 05%
- C. betamethasone dipropionate 0.05%
- D. hydrocortisone 1%
- E. clobetasol propionate 0.05%

60. In atopic dermatitis, the presence of punched out erosions, vesicles, and infected skin lesions that fail to respond to oral antibiotics suggests infection with

- A. Herpes zoster
- B. Herpes simplex
- C. cutaneous warts
- D. Trichophyton rubrum
- E. Molluscum contagiosum

61. Exfoliative dermatitis may develop in patients with atopic dermatitis and extensive skin involvement, usually caused by inappropriate therapy or superinfection with

- A. E. coli
- B. Herpes zoster
- C. Herpes simplex
- D. Streptococcal pyogen
- E. Trichophyton rubrum

62. Reactions to stinging and biting insects may cause

- A. a limited lesion confined to the primary site
- B. a pronounced localized reaction
- C. a pronounced systemic reaction due to immediate hypersensitivity
- D. a pronounced systemic reaction due to delayed hypersensitivity
- E. no reaction

63. Systemic allergic responses to insects are attributed to IgE antibody response caused primarily by

- A. ticks
- B. spiders
- C. hymenoptera
- D. scorpions
- E. triatoma (kissing bug)

64. The **MOST** notorious stinging insect is

- A. honeybee
- B. harvester ants
- C. hornet
- D. bumblebee

- E. yellow jackets
65. Delayed/Late reaction to stinging venom may precipitate all the following **EXCEPT**
- A. vasculitis
 - B. serum sickness
 - C. encephalopathy
 - D. hemolytic anemia
 - E. nephrotic syndrome
66. The **MOST** reliable diagnostic modality for detection of venom-specific IgE is
- A. in vitro serum assay for venom-specific IgE
 - B. skin test
 - C. serum tryptase level
 - D. plasma histamine
 - E. radioallergosorbent assay
67. In the presence of convincing history of a severe systemic reaction, the next diagnostic step for those with initially negative skin test is
- A. in vitro serum assay for venom-specific IgE
 - B. repeat skin test after 4-6 weeks
 - C. serum tryptase level
 - D. plasma histamine
 - E. basophil histamine release test
68. Sting sites rarely become infected possibly owing to
- A. cleansing the area immediately after insect
 - B. the over use of antibacterial creams for the area
 - C. venom constituents have antibacterial action
 - D. vasospasm after stinging impedes bacterial invasion
 - E. venom immunotherapy has antibacterial properties
69. Anaphylactic reactions after a Hymenoptera sting are treated exactly like anaphylaxis from any cause.
- Of the following, the drug of choice is
- A. oxygen
 - B. epinephrine
 - C. antihistamines
 - D. corticosteroids
 - E. intravenous fluids
70. A **TRUE** indication of venom immunotherapy in a six-year-old boy is
- A. large local reaction with positive skin test and positive in vitro test

- B. generalized cutaneous reaction with positive skin test and negative in vitro test
- C. generalized cutaneous reaction with positive in vitro test and negative skin test
- D. systemic reaction with positive skin test and negative in vitro test
- E. systemic reaction with negative skin test and negative in vitro test

71. Skeeter syndrome is a large local reaction to stinging and biting insects.

Of the following, the **TRUE** statement is

- A. Ig E mediated response
- B. usually occurs in older children
- C. usually followed by anaphylaxis
- D. mosquito is the responsible agent
- E. misdiagnosed as erythema nodosum

72. The eye is a common target of allergic disorders because of its marked vascularity and direct contact with allergens in the environment.

Of the following, the **MOST** immunologically active tissue of the external eye is

- A. iris
- B. sclera
- C. eyelids
- D. eyelashes
- E. conjunctiva

73. The **MOST** common hypersensitivity response of the eye is

- A. allergic conjunctivitis
- B. vernal keratoconjunctivitis
- C. atopic keratoconjunctivitis
- D. giant papillary conjunctivitis
- E. contact allergy

74. The **MOST** sight-threatening type of ocular allergy is

- A. contact allergy
- B. vernal keratoconjunctivitis
- C. giant papillary conjunctivitis
- D. seasonal allergic conjunctivitis
- E. perennial allergic conjunctivitis

75. The **MOST** common presenting complain of allergic conjunctivitis is

- A. pain
- B. itching
- C. redness
- D. blurred vision
- E. purulent discharge

76. Topical ophthalmic medications usually cause
- A. contact allergy
 - B. allergic conjunctivitis
 - C. vernal keratoconjunctivitis
 - D. atopic keratoconjunctivitis
 - E. giant papillary conjunctivitis
77. Contact lenses are associated with
- A. contact allergy
 - B. allergic conjunctivitis
 - C. vernal keratoconjunctivitis
 - D. atopic keratoconjunctivitis
 - E. giant papillary conjunctivitis
78. Conjunctivitis medicamentosa is a consequence of chronic use of
- A. antihistamines
 - B. steroids
 - C. decongestants
 - D. anti-inflammatory
 - E. mast cell stabilizers
79. Non IgE mediated urticaria can be caused by
- A. milk
 - B. hymenoptera
 - C. Epstein-barr virus
 - D. latex
 - E. blood
80. Chronic urticaria may be caused by
- A. latex
 - B. peanut
 - C. IV immunoglobulin
 - D. streptococcal pharyngitis
 - E. systemic lupus erythematosus
81. The drug of choice for cold-induced urticaria is
- A. loratadine
 - B. epinephrine IM
 - C. cyproheptadine
 - D. diphenhydramine
 - E. oral corticosteroid

82. Acute urticaria is a self-limited illness requiring little treatment. All the following can be used **EXCEPT**
- A. loratadine
 - B. hydroxyzine
 - C. epinephrine IM
 - D. H2 antihistamines
 - E. oral corticosteroid
83. Skin biopsy for diagnosis of possible urticarial vasculitis is recommended for
- A. urticarial lesions that persist at different locations for >24 hr
 - B. those with non-pigmented or non-purpuric components
 - C. those that burn more than itch
 - D. those with associated collagen vascular diseases
 - E. those appeared after peanuts ingestion
84. The differential diagnosis of chronic urticaria includes the following **EXCEPT**
- A. cutaneous mastocytosis
 - B. systemic mastocytosis
 - C. complement-mediated mast cell degranulation in malignancies
 - D. cutaneous blistering disorders
 - E. dermatographism
85. The **MOST** common childhood symptoms of hereditary angioedema is
- A. cutaneous non-pitting but pruritic edema associated with urticaria
 - B. cutaneous pitting but non-pruritic edema not associated with urticaria
 - C. cutaneous pitting and pruritic edema associated with urticaria
 - D. cutaneous non-pitting and non-pruritic edema associated with urticaria
 - E. cutaneous non-pitting and non-pruritic edema not associated with urticaria
86. All the following factors can amplify anaphylaxis **EXCEPT**
- A. exercise
 - B. vaccination
 - C. upper respiratory tract infection
 - D. fever
 - E. emotional stress
87. One of the principal pathologic features in fatal anaphylaxis is
- A. acute bronchial obstruction
 - B. hypotension
 - C. behavioral change
 - D. abdominal pain
 - E. vomiting

88. Sudden collapse in the presence of cutaneous symptoms in a previously healthy child should raise suspicion of
- A. vasovagal collapse
 - B. anaphylaxis
 - C. aspiration
 - D. pulmonary embolism
 - E. seizure disorder
89. Cutaneous symptoms may be absent in anaphylaxis in
- A. 5%
 - B. 10%
 - C. 20%
 - D. 30%
 - E. 40%
90. The best diagnostic test for anaphylaxis in a child exposed to an allergen 3 hours before arrival to hospital is
- A. plasma histamine
 - B. skin test
 - C. radioallergosorbent assay
 - D. plasma tryptase
 - E. immunoCAP IgE test
91. After institution of treatment of anaphylaxis, the patient should be monitored in the emergency room for at least
- A. 1 hour
 - B. 4 hours
 - C. 6 hours
 - D. 12 hours
 - E. 24 hours
92. In cases of food-associated exercise-induced anaphylaxis, the parents are advised that children must not take exercise after ingestion of the triggering food for
- A. 2-3 hours
 - B. 4-6 hours
 - C. 8-12 hours
 - D. 13-18 hours
 - E. 19-24 hours
93. All the following medications are encountered to cause serum sickness **EXCEPT**
- A. ciprofloxacin
 - B. meropenem

- C. rituximab
- D. trimethoprim sulfate
- E. carbamazepine

94. Once the offending agent causing serum sickness is discontinued, the symptoms resolve spontaneously within

- A. 1-4 days
- B. 1-4 weeks
- C. 5-8 weeks
- D. 9-12 weeks
- E. 13-18 weeks

95. Which of the following results of investigations is unlikely in serum sickness?

- A. elevated erythrocyte sedimentation rate
- B. thrombocytosis
- C. reduced C_3 activity
- D. negative microbial cultures
- E. microscopic hematuria

96. Skin biopsy is not usually necessary for confirming the diagnosis of serum sickness because

- A. the complement C_3 and C_4 yield better results
- B. the clinical examination is enough for the diagnosis
- C. skin test is superior to biopsy
- D. the histopathological findings are not specific
- E. it may be complicated by local infection

97. All the following are complications of serum sickness **EXCEPT**

- A. carditis
- B. glomerulonephritis
- C. Guillain-barre syndrome
- D. peripheral neuritis
- E. colitis

98. All the following are symptoms of acute IgE mediated food allergy **EXCEPT**

- A. urticaria
- B. pruritis
- C. abdominal pain
- D. diarrhea
- E. wheezing

99. One of the following presentations is a non-IgE mediated food allergy

- A. Heiner syndrome
- B. gastrointestinal anaphylaxis
- C. rhino conjunctivitis
- D. oral allergy syndrome
- E. angioedema

100. The onset of presentation of food allergy is late in

- A. hen's egg white
- B. peanuts
- C. shellfish
- D. soybean
- E. cow's milk

101. Food protein-induced enterocolitis syndrome is characterized by all the following **EXCEPT**

- A. manifests in the first months of life
- B. vomiting occurs 1-3 hours after feeding
- C. hypotension occurs in approximately 15% of cases
- D. commonly improved with ingestion of soy protein based formula
- E. usually resolve by the age of three years of life

102. The **MOST** common cause of food protein induced enteropathy is

- A. peanuts
- B. fish
- C. cow's milk
- D. eggs
- E. tree nuts

103. The **MOST** severe form of food protein-induced enteropathy is

- A. celiac disease
- B. eosinophilic esophagitis
- C. oral allergy syndrome
- D. acute gastrointestinal allergy
- E. food protein proctocolitis

104. Wheezing occurs in approximately which percent of IgE-mediated food allergic reactions?

- A. 15%
- B. 25%
- C. 35%
- D. 45%
- E. 55%

105. Administration of the following vaccine is contraindicated in children with egg allergy
- A. MMR
 - B. Influenza
 - C. DPT
 - D. yellow fever
 - E. rabies
106. The only way to establish the diagnosis of cell-mediated food reactions is
- A. breath hydrogen test
 - B. endoscopy
 - C. elimination and challenge test
 - D. IgE testing
 - E. skin test
107. Unpredictable drug reactions include
- A. dose dependent
 - B. drug toxicity
 - C. drug interactions
 - D. adverse effects
 - E. allergic reaction
108. Which of the following drugs can cause Stevens - Johnson syndrome?
- A. dapsone
 - B. hydralazine
 - C. sulfonamide
 - D. procainamide
 - E. bleomycin
109. Which one of the following agents is implicated in causing cutaneous lupus?
- A. corticosteroids
 - B. calcium-channel blockers
 - C. allopurinol
 - D. B-lactam antibiotics
 - E. sulfonamide
110. Fixed drug eruption can be caused by
- A. allopurinol
 - B. cephalosporin
 - C. nonsteroidal anti-inflammatory drug
 - D. nitrofurantoin
 - E. infliximab

111. One of the following drug-induced allergic reactions is not included as an indictment of allopurinol

- A. exanthem
- B. Stevens-Johnson syndrome
- C. toxic epidermal necrolysis
- D. pulmonary fibrosis
- E. interstitial nephritis

112. Drug Rash with Eosinophilia and Systemic Symptoms (DRESS) can be caused by all the following **EXCEPT**

- A. anticonvulsants
- B. sulfonamides
- C. B-lactam antibiotics
- D. minocycline
- E. allopurinol

113. Systemic lupus erythematosus can be caused by

- A. isoniazid
- B. sulfonamide
- C. hydrochlorothiazide
- D. allopurinol
- E. nonsteroidal anti-inflammatory drug

114. Vasculitis can be caused by

- A. isoniazid
- B. sulfonamide
- C. penicillamine
- D. calcium channel blockers
- E. tetracycline

115. All the following are risk factors for adverse drug reactions **EXCEPT**

- A. prior exposure
- B. route of administration
- C. dosing schedule
- D. genetic predisposition
- E. atopy

116. Of the following, the agent that does not share an identical side chain with penicillin is

- A. cephalixin
- B. cefotaxime
- C. cefixime

- D. ceftazidime
- E. ceftriaxone

- 1.(E). Recognition of allergic rhinitis as a major chronic respiratory disease of children rests largely on its high prevalence, detrimental effects on quality of life and school performance, and comorbidities. The prevalence peaks late in childhood.
- 2.(C). The symptoms may appear in infancy; with the diagnosis generally established by the time the child reaches age 6 yr. The prevalence peaks late in childhood.
- 3.(A). Positive family history is the major risk factor. Other risk factors are raised serum IgE more than 100 IU/ml, smoking mother, and those with heavy exposure to indoor allergens.
- 4.(E).
- 5.(C).
- 6.(D). There is a decreased risk of asthma, allergic rhinitis (AR), and atopic sensitization with early introduction to wheat, rye, oats, barley, fish and eggs.
- 7.(D). The exposure to dogs, cats and endotoxin early in childhood protects against the development of atopy.
- 8.(D). The symptoms of intermittent allergic rhinitis occur on <4 days per week or for <4 consecutive weeks. In persistent AR symptoms occur on >4 days per week and/or for >4 consecutive weeks.
- 9.(C). Dark circles under the eyes. (A) represent allergic salute, while (B) represent allergic gape.
- 10.(C). Thick, purulent nasal secretions indicate the presence of infection.
- 11.(E).
- 12.(B).
- 13.(D). Allergic conjunctivitis, characterized by itching, redness and swelling of the conjunctivae, has been reported in at least 20% of the population and in more the 70% of patients with allergic rhinitis, most frequently in older children and young adults.
- 14.(C).
- 15.(A).
- 16.(A). Epicutaneous skin tests provide the best method for detection of allergen-specific IgE (positive predictive value of 48.7% for the epidemiologic diagnosis of AR). They are inexpensive and sensitive, and the risks and discomfort are minimal.
- 17.(B).
- 18.(D). Patients with more persistent, severe symptoms require intranasal corticosteroids, the most effective therapy for allergic rhinitis, a treatment that may be beneficial also for concomitant allergic conjunctivitis.
- 19.(C).

20.(A). Early childhood risk factors for persistent asthma have been identified and have been described as major (parent asthma, eczema, inhalant allergen sensitization) and minor (allergic rhinitis, wheezing apart from colds, $\geq 4\%$ peripheral blood eosinophils, food allergen sensitization) risk factors.

21.(A).

22.(B).

23.(C). Of these, approximately one-third who continue to have persistent asthma into later childhood, and approximately two-thirds improve on their own through their teen years.

24.(D). Severe airflow limitation (PEF or FEV1 value $< 50\%$ of personal best or predicted values).

25.(B).

26.(A). Supplemental oxygen is the best to start with.

27.(C).

28.(C). Male gender.

29.(B). Inhaled SABA, up to 3 treatments in 1 hr. If the child has an incomplete response to initial treatment with rescue medication (persistent symptoms and/or a PEF value $< 80\%$ of personal best), a short course of oral corticosteroid therapy (prednisone 1-2 mg/kg/day [not to exceed 60 mg/day] for 4 days), in addition to inhaled β -agonist therapy, should be instituted.

30.(D).

31.(A). Peribronchial thickening and flattening of diaphragm.

32.(B). In asthmatic patients, an improvement in $FEV1 \geq 12\%$ or > 200 mL in response to bronchodilator is consistent with asthma, while FEV1 typically decreases during or after exercise by $> 15\%$.

33.(B).

34.(D). Discharge medications include administration of an inhaled β -agonist up to every 3-4 hr plus a 3-7 day course of an oral corticosteroid.

35.(D). Children with well-controlled asthma have daytime symptoms ≤ 2 days/wk and need a rescue bronchodilator ≤ 2 days/wk; an $FEV1$ of $> 80\%$ of predicted (and an $FEV1:FVC$ ratio $> 80\%$ for children 5-11 yr of age); no interference with normal activity; and < 2 exacerbations in the past year.

36.(D). The green zone (80-100% of personal best) indicates good control; the yellow zone (50-80%) indicates less-than-optimal control and necessitates increased awareness and treatment; the red zone ($< 50\%$) indicates poor control and greater likelihood of an exacerbation, requiring immediate intervention.

37.(A). Long-term controller medications are inhaled corticosteroid, LABAs, leukotriene modifiers, sustained-release theophylline, and nonsteroidal antiinflammatory agents. An anti-IgE preparation, omalizumab (Xolair), is approved by the FDA for use as an add-on therapy in children ≥ 12 yr that have moderate to severe allergic asthma that is difficult to control.

38.(E).

39.(B).

40.(E). The hallmark of atopic dermatitis is dry skin, is results from compromise of the epidermal barrier, which leads to excess transepidermal water loss, allergen penetration, and microbial colonization.

41.(A). Approximately 50% of patients experience symptoms in the 1st yr of life, and an additional 30% are diagnosed between 1 and 5 yr of age.

42.(D). Intense pruritus, especially at night, and cutaneous reactivity are the cardinal features of atopic dermatitis.

43.(C). Foods (cow milk, egg, peanut, tree nuts, soy, wheat, and shellfish), aeroallergens (pollen, grass, animal dander, and dust mites), infection (staphylococcus, herpes simplex, and molluscum), reduced humidity, excessive sweating, and irritants (wool, acrylic, soaps, toiletries, fragrances, and detergents) can exacerbate (trigger) pruritus and scratching.

44.(D).

45.(A).

46.(E). The major clinical features in atopic dermatitis are (pruritus, facial and extensor eczema in infants and children, flexural eczema in adolescents, relapsing dermatitis and personal or family history).

47.(C). Histiocytosis, other mentioned disorders are associated with infections.

48.(C).

49.(A). Because patients with AD have impaired skin barrier function from reduced lipid levels, they present with diffuse, abnormally dry skin, or xerosis. Moisturizers are first-line therapy.

50.(E).

51.(D). Vitamin D deficiency often accompanies severe AD. Vitamin D enhances skin barrier function, reduces corticosteroid requirements to control inflammation and augments skin antimicrobial function.

52.(E).

53.(C). Erythromycin and azithromycin are usually beneficial for patients who are not colonized with a resistant *S. aureus* strain; a first-generation cephalosporin (cephalexin) is recommended for macrolide-resistant *S. aureus*. Topical mupirocin is useful in the treatment of localized impetiginous lesions, with systemic antibiotics for widespread infections.

54.(A). There has been particular interest in the role of *Malassezia furfur* in atopic dermatitis because it is lipophilic yeast commonly present in the seborrheic areas of the skin.

55.(E). For the other stems; A- high predictive value, B- Positive skin test often do not correlate with clinical symptoms, C- Rarely required, and D- The majority react to less than 3 foods.

56.(D).

- 57.(B). The dramatic clinical improvement that may occur with systemic corticosteroids is frequently associated with a severe rebound flare of atopic dermatitis after therapy discontinuation.
- 58.(D). Hydrocortisone is labeled as group 7 topical steroids which is the least potent one.
- 59.(D). Use the least potent steroid in the face and intertriginous areas.
- 60.(B). Herpes simplex virus (HSV) can provoke recurrent dermatitis and may be misdiagnosed as *S. aureus* infection, which can be diagnosed by a Giemsa-stained Tzanck smear of cells scraped from the vesicle base or by viral polymerase chain reaction or culture.
- 61.(C). Exfoliative dermatitis associated with generalized redness, scaling, weeping, crusting, systemic toxicity, lymphadenopathy, and fever, and is usually caused by superinfection (e.g., with toxin-producing *S. aureus* or HSV infection) or inappropriate therapy. In some cases, the withdrawal of systemic glucocorticoids used to control severe AD precipitates exfoliative erythroderma.
- 62.(A).
- 63.(C).
- 64.(E). Yellow jackets are aggressive and ground dwelling, and they linger near activities involving food.
- 65.(D).
- 66.(B).
- 67.(A). In vitro serum assay for venom-specific IgE is recommended if skin test results are negative in the presence of a convincing history of a severe systemic reaction.
- 68.(C).
- 69.(B). Adjunctive treatment includes antihistamines, corticosteroids, intravenous fluids, oxygen, and transport to the emergency room.
- 70.(D). Systemic reaction at any age with positive skin test / in vitro test is an indication for venom immunotherapy. Large local reaction and generalized cutaneous reactions in children are not indications for both diagnostic tests (skin and in vitro tests) and venom immunotherapy.
- 71.(D). Skeeter syndrome is a large local reaction to mosquito bite, it happens usually in young children and misdiagnosed as cellulitis. This is not an Ig E mediated reaction but because of the vasoactive and irritant material from the mosquito, it does not cause anaphylaxis.
- 72.(E).
- 73.(A). Allergic conjunctivitis is the most common hypersensitivity response of the eye, affecting approximately 25% of the general population and 30% of children with atopy.
- 74.(B). Vernal keratoconjunctivitis and atopic keratoconjunctivitis are sight-threatening entities and causes visual morbidity and suspected cases need to be referred to an ophthalmologist.
- 75.(B). Patients with allergic conjunctivitis complain of variable ocular itching, rather than pain, with increased tearing.

- 76.(A). Contact allergy typically involves the eyelids but can also involve the conjunctivae. It is being recognized more frequently in association with increased exposure to topical medications, contact lens solutions, and preservatives.
- 77.(E). Giant papillary conjunctivitis has been linked to chronic exposure to foreign bodies, such as contact lenses, both hard and soft, ocular prostheses, and sutures.
- 78.(C). Topical decongestants act as vasoconstrictors; adverse effects of topical include burning and rebound hyperemia or conjunctivitis medicamentosa with chronic use.
- 79.(C). Acute urticaria can also result from non-IgE-mediated stimulation of mast cells, caused by radiocontrast agents, viral agents (including hepatitis B and Epstein-Barr virus), opiates, and nonsteroidal anti-inflammatory agents.
- 80.(E). Autoimmune disease, like SLE, Juvenile rheumatoid arthritis, thyroid disease, inflammatory bowel diseases and celiac disease, are well established causes of chronic urticaria.
- 81.(C).
- 82.(D). H2 antihistamines used in chronic urticaria.
- 83.(D). Skin biopsy is recommended for urticarial lesions that persist at the same location for >24 hr, those with pigmented or purpuric components, and those that burn more than itch. Collagen vascular diseases such as systemic lupus may manifest urticarial vasculitis as a presenting feature.
- 84.(E).
- 85.(E).
- 86.(B). Vaccination is a trigger, not a cofactor in anaphylaxis, others are cofactors.
- 87.(A). Principal pathologic features in fatal anaphylaxis include acute bronchial obstruction with pulmonary hyperinflation, pulmonary edema, intraalveolar hemorrhaging, visceral congestion, laryngeal edema, and urticaria and angioedema. Acute hypotension is attributed to vasomotor dilation and/or cardiac dysrhythmias.
- 88.(B). Other choices are to be considered in cases where there are no cutaneous manifestations.
- 89.(C).
- 90.(D). Plasma histamine is elevated for a brief period but is unstable and difficult to measure in a clinical setting. Plasma tryptase is more stable and remains elevated for several hours but often is not elevated, especially in food-induced anaphylactic reactions.
- 91.(B). Patients may experience biphasic anaphylaxis, which occurs when anaphylactic symptoms recur after apparent resolution. More than 90% of biphasic responses occur within 4 hours, so patients should be observed for at least 4 hr before being discharged from the emergency department.
- 92.(A).
- 93.(A).
- 94.(B). Symptoms lasting longer suggest another diagnosis.
- 95.(B). Thrombocytopenia is often present.
- 96.(D). Not specific and not diagnostic.

- 97.(E).
- 98.(B).
- 99.(A). Heiner syndrome (food induced pulmonary hemosiderosis) is a non-IgE mediated food allergy.
- 100.(C). The onset is usually during adulthood. Others are earlier in presentation.
- 101.(D). Commonly provoked by ingestion of milk and soy based protein formula.
- 102.(C).
- 103.(A).
- 104.(B).
- 105.(D).
- 106.(C). There are no laboratory studies to help identify foods responsible for cell-mediated reactions. Consequently, elimination diets followed by food challenges are the only way to establish the diagnosis.
- 107.(E).
- 108.(C). Antibacterial sulfonamides, anticonvulsants, oxycam NSAIDs, and allopurinol have been implicated in causing Stevens - Johnson syndrome and toxic epidermal necrolysis.
- 109.(B). Hydrochlorothiazide and calcium channel blockers can cause cutaneous lupus.
- 110.(C). Tetracycline, NSAIDs, and carbamazepine are implicated in causing fixed drug eruption.
- 111.(D). Nitrofurantoin, bleomycin and methotrexate are implicated in causing pulmonary fibrosis.
- 112.(C). They can cause anaphylaxis.
- 113.(A). Along with Hydralazine and procainamide.
- 114.(C). Along with Hydralazine and propylthiouracil.
- 115.(E). Atopy does not appear to predispose patients to allergic reactions to low-molecular-weight compounds, but atopic patients in whom an allergic reaction develops have a significantly increased risk of serious reaction.
- 116.(C).

1. Nonsteroidal anti-inflammatory drugs (NSAIDs) are prescribed to decrease acute and chronic inflammation associated with various rheumatic diseases; however, many adverse effects have been encountered with long term use.

Of the following, the NSAID that has the highest toxicity is

- A. celecoxib
- B. meloxicam
- C. indomethacin
- D. naproxen
- E. ibuprofen

2. A 7-year-old boy developed small hypopigmented depressed scars after fingernail scratches; he has been treated with nonsteroidal antiinflammatory drugs (NSAIDs) for a rheumatic disease 6 weeks ago.

Of the following, the NSAID that is **MOST** likely to cause such a unique skin reaction is

- A. celecoxib
- B. meloxicam
- C. indomethacin
- D. naproxen
- E. ibuprofen

3. Hydroxychloroquine sulfate is an antimalarial drug important in the treatment of SLE and dermatomyositis, particularly cutaneous manifestations of disease and to reduce lupus flares.

Of the following, the **MOST** important procedure that should be done routinely during the course of administration is

- A. gastric endoscopy
- B. bone marrow examination
- C. muscle biopsy
- D. glucose-6-phosphate dehydrogenase enzyme level assay
- E. ophthalmological examination

4. Sulfasalazine is an effective drug in many rheumatic diseases; however, it is associated with severe systemic hypersensitivity reaction.

It is approved in all the following rheumatic disease **EXCEPT**

- A. polyarticular juvenile idiopathic arthritis (JIA)
- B. oligoarticular JIA

- C. systemic JIA
- D. peripheral arthritis associated with juvenile ankylosing spondylitis
- E. enthesitis associated with juvenile ankylosing spondylitis

5. You are meeting parents of a 14-year-old girl who has been treated with a monthly intravenous cyclophosphamide for SLE-associated renal failure for the last 6 months; the mother is asking about the long-term complications of this drug.

All the following are long-term complications **EXCEPT**

- A. bone marrow suppression
- B. bladder cancer
- C. leukemia
- D. lymphoma
- E. infertility

6. Juvenile idiopathic arthritis has many subtypes. Arthritis must be present to make a diagnosis of any subtype; involved joints often have the following signs **EXCEPT**

- A. swelling
- B. warm
- C. erythema
- D. limitation of movement
- E. pain on movement

7. Oligoarthritis, the most common subtype of juvenile idiopathic arthritis, is defined as involving ≤ 4 joints within the 1st 6 mo of disease onset.

Of the following, the **MOST** commonly affected joint is

- A. hip
- B. knee
- C. elbow
- D. wrist
- E. metatarsal

8. Oligoarthritis predominantly affects the joints of the lower extremities rather than upper extremity joints. Which of the following joints is never a presenting sign of oligoarthritis?

- A. hip
- B. knee
- C. ankle
- D. metatarsal
- E. interphalangeal

9. A 4-year-old girl recently diagnosed with persistent oligoarticular juvenile idiopathic rheumatoid arthritis (JIA); she has 3 involved joints including the right knee, right ankle, and left elbow; antinuclear antigen (ANA) is significantly positive.

Of the following, the **MOST** important step in the management of this girl is

- A. regular examination of locomotor system
- B. periodic slit-lamp examination
- C. periodic ANA monitoring
- D. periodic erythrocyte sedimentation rate (ESR) monitoring
- E. frequent C-reactive protein (CRP) monitoring

10. Antinuclear antigen (ANA) measurement test is useful in some rheumatologic diseases especially with persistent oligoarticular juvenile idiopathic rheumatoid arthritis (JIA).

All the following are more likely to be correlated with ANA positivity **EXCEPT**

- A. anterior uveitis
- B. younger age at disease onset
- C. female sex
- D. symmetrical arthritis
- E. lower number of involved joints over time

11. Rheumatoid factor (RF)–positive polyarthritis is characterized by aggressive symmetric inflammation of joints of both upper and lower extremities.

Of the following, the extra articular manifestation that is almost exclusively occur in RF-positive individuals is

- A. fever
- B. evanescent rash
- C. uveitis
- D. extensor surfaces nodules
- E. pericarditis

12. A 5-year-old boy develops acute onset of high spiking fevers, lymphadenopathy, hepatosplenomegaly, and purpura; he has been diagnosed with systemic Juvenile idiopathic arthritis (JIA) since early childhood treated with antiinflammatory drugs. You suspect macrophage activating syndrome (MAS).

Of the following, the **BEST** test that distinguishes MAS from a flare of the primary disease is

- A. leucopenia
- B. falling ESR
- C. hypofibrinogenemia
- D. hypertriglyceridemia
- E. evidence of hemophagocytosis in the bone marrow

13. Rheumatoid factor (RF)–positive polyarthritis usually accounts for < 10% of all juvenile idiopathic arthritis (JIA) cases. The articular manifestation pattern is characterized by involvement of ≥ 5 joints in both upper and lower extremities.

Of the following, the **LEAST** effective drug to induce remission for this subtype of JIA is

- A. NSAIDs
- B. methotrexate
- C. TNF- α antagonists
- D. IL-1 inhibitors
- E. IL-6 inhibitors

14. A 5-year-old boy has systemic juvenile idiopathic arthritis (sJIA) with systemic manifestations including fever, hepatosplenomegaly, lymphadenopathy, and pericarditis.

Of the following, the **BEST** initial treatment for this boy is

- A. NSAIDs
- B. systemic glucocorticoids
- C. TNF- α inhibitors
- D. IL-1 antagonists
- E. IL-6 antagonists

15. You are discussing with the medical students the role of systemic steroids for the management of rheumatologic diseases.

Your discussion should include all the following statements **EXCEPT**

- A. they are recommended for management of severe systemic illness
- B. they are used for bridge therapy during the wait for therapeutic response to a disease modified anti rheumatic drugs (DMARD)
- C. they are effective for control of uveitis
- D. they prevent joint destruction
- E. they impose risks of severe toxicities

16. Chronic uveitis is one of the extraarticular manifestations of various subtypes of juvenile idiopathic arthritis.

All the following are risk factors for the development of uveitis **EXCEPT**

- A. oligoarthritis subtype
- B. female gender
- C. ANA-positivity
- D. onset of arthritis earlier than 6 yr of age
- E. severity of arthritis

17. The child with polyarticular JIA often has a more prolonged course of active joint inflammation and requires early and aggressive therapy.

Of the following, the predictor that carries the **WORST** prognosis is

- A. old age at onset
- B. rheumatoid factor (RF) seronegativity
- C. absence of rheumatoid nodules
- D. small numbers of affected joints
- E. hip joint involvement

18. Spondyloarthritis may be overlapped clinically with other forms of juvenile idiopathic arthritis (JIA). Clinical manifestations that help to distinguish spondyloarthritis from other forms of juvenile arthritis include all the following **EXCEPT**

- A. arthritis of the sacroiliac joints
- B. arthritis of the hips
- C. enthesitis
- D. asymptomatic uveitis
- E. gastrointestinal inflammation

19. Enthesitis-Related Arthritis (ERA) is a subtype of rheumatic disease that falls in the juvenile idiopathic arthritis (JIA) category.

Of the following, the clinical feature that is highly suggestive of ERA is

- A. symmetrical arthritis early in the disease course
- B. early involvement of more than 5 joints
- C. involvement of upper limb joints
- D. Inflammation of the small joints of the foot
- E. asymmetrical enthesitis

20. Spondyloarthritis are complex diseases in which susceptibility is largely genetically determined. Which of the following characteristics is never associated with such diseases?

- A. enthesitis
- B. peripheral arthritis
- C. axial arthritis
- D. HLA-B27 positivity
- E. rheumatoid factor positivity

21. You are discussing with medical students Juvenile Ankylosing Spondylitis (JAS) subtype of spondyloarthritis. One of the students asks you about the features that distinguish this disease from the adult-onset ankylosing spondylitis (AOAS).

All the following are true regarding JAS in comparison with AOAS **EXCEPT**

- A. it is present in patients <16 yr old
- B. axial disease occurs more frequently early in the disease course
- C. inflammatory back pain is less frequent at disease onset
- D. enthesitis occurs more commonly
- E. peripheral arthritis is more common

22. Reactive arthritis is defined as joint inflammation caused by a sterile inflammatory reaction following a recent infection.

Of the following, the **LEAST** likely micro-organism that may cause reactive arthritis is

- A. *Clostridium difficile*
- B. *Salmonella paratyphi*
- C. *Shigella flexneri*
- D. *Yersinia enterocolitica*
- E. *Campylobacter jejuni*

23. The distinction between postinfectious arthritis and reactive arthritis is not always clear. All the following are common features of postinfectious arthritis, rather than reactive arthritis **EXCEPT**

- A. there is a transient joint swelling or pain
- B. it lasts less than 6 wk
- C. chronic spondyloarthritis is a common sequelae
- D. it may be encountered after group A streptococcus infection
- E. rubella infection is a common causative agent

24. You are meeting parents of 2-year-old girl who is going to receive rubella vaccine; the mother is concerned regarding arthritis that may develop after immunization.

Of the following, the statement that should be included during the discussion is

- A. it is usually frequently encountered after immunization
- B. it typically affects the large joints of the upper extremity
- C. it is more common in girls
- D. it is uncommon in children
- E. arthralgia usually begins immediately after administration of immunization

25. Postinfectious arthritis describes arthritis that occurs after infectious illnesses, mainly viruses.

Of the following, the **LEAST** likely causative viral infection is

- A. rubella
- B. varicella-zoster
- C. cytomegalovirus
- D. Epstein-Barr virus
- E. herpes-simplex virus

26. A 4-year-old boy has an acute severe left groin pain that refers to thigh. Examination reveals limitation of movement of the left hip joint. Lab investigations reveal normal blood count and indices, and normal ESR; radiologic imaging shows widening of the hip joint space with effusion. You suspect transient synovitis.

Of the following, the **BEST** therapeutic approach for this boy is

- A. systemic steroids

- B. methotrexate
- C. aspiration of joint fluid
- D. intravenous antibiotics
- E. observation

27. Reactive arthritis has the potential for evolving to chronic arthritis, especially after bacterial enteric infection or genitourinary tract infection (UTI).

Of the following, the microorganism that is **MOST** likely causing chronic arthritis after UTI is

- A. *Escherichia coli*
- B. *Chlamydia trachomatis*
- C. *Proteus mirabilis*
- D. *Pseudomonas aeruginosa*
- E. *Staphylococcus saprophyticus*

28. Systemic lupus erythematosus (SLE) is a chronic autoimmune disease characterized by multisystem inflammation and the presence of circulating autoantibodies directed against self-antigens. It occurs in both children and adults.

All the following are features of childhood SLE **EXCEPT**

- A. it has a more severe course
- B. there is more widespread organ involvement
- C. it usually presents before 5 years of age
- D. fever, fatigue, hematologic abnormalities, and arthritis are common clinical manifestations
- E. although renal disease is asymptomatic, it is often present as nephrotic syndrome in adolescent age group

29. Skin is a commonly involved organ by SLE. There are different cutaneous manifestations.

Of the following, the skin manifestation that is **MOST** suggestive of SLE in children is

- A. malar rash
- B. discoid rash
- C. photosensitive rash
- D. cutaneous vasculitis
- E. livedo reticularis

30. Drug-induced lupus refers to the presence of SLE manifestations triggered by exposure to specific medications, including antibiotics.

Of the following, the drug that is **MOST** likely associated with drug-induced lupus is

- A. isoniazid
- B. rifampin
- C. nitrofurantoin

- D. penicillin
- E. tetracycline

31. Systemic lupus erythematosus (SLE) is often characterized by periods of flare and disease quiescence or may follow a more smoldering disease course.

All the following lab tests correlate with active disease **EXCEPT**

- A. positive anti-nuclear antibody titer
- B. positive anti-double-stranded DNA level
- C. low serum complement level
- D. high erythrocyte sedimentation rate
- E. elevated C-reactive protein (CRP) value

32. A 12-year-old female adolescent has a recent diagnosis of SLE; she has a faint malar rash and mild arthritis including both elbow and knee joints as well as the joints of small fingers.

Of the following, the **MOST** appropriate initial therapy for this patient is

- A. systemic steroids
- B. methotrexate
- C. leflunomide
- D. cyclophosphamide
- E. hydroxychloroquine

33. A 3-week-old male baby develops malar rash involving the face and periorbital area after exposure to sun light; you suspect neonatal lupus.

Of the following, the **NEXT** step of management is

- A. measurement of anti-Ro and anti-La antibodies of the mother
- B. measurement of anti-Ro and anti-La antibodies of the baby
- C. cardiac evaluation of the mother
- D. cardiac evaluation of the baby
- E. hematological evaluation of the baby

34. You are meeting a pregnant mother who has had a previous baby with congenital heart block due to neonatal lupus; she has a positive anti-SSA and anti-SSB antibodies. All the following are therapeutic modalities to prevent occurrence of congenital fetal cardiac complications **EXCEPT**

- A. fluorinated corticosteroids
- B. intravenous immunoglobulin
- C. plasmapheresis
- D. hydroxychloroquine
- E. methotrexate

35. Juvenile dermatomyositis (JDM) is the most common inflammatory myositis in children characterized by skin rash and proximal muscle weakness.

All the following are common cutaneous manifestations of JDM **EXCEPT**

- A. heliotrope rash of the eyelids
- B. photosensitivity to ultraviolet light
- C. facial erythema sparing the nasolabial folds
- D. Gottron papules
- E. periungual telangiectasias

36. Treatment of juvenile dermatomyositis (JDM) includes pharmacological and non-pharmacological therapy. The mainstay of pharmacological therapy is corticosteroids that should be integrated with non-pharmacological therapy.

All the following are recommended as part of non-pharmacological treatment program in JDM **EXCEPT**

- A. physical therapy
- B. occupational therapy
- C. avoidance of sun exposure
- D. bed rest
- E. social and psychological support

37. Most complications from Juvenile dermatomyositis (JDM) are related to prolonged and severe weakness; secondary complications from medical treatments are also common.

Of the following, the **LEAST** likely recognized complications of JDM is

- A. aspiration pneumonia
- B. gastrointestinal bleeding
- C. cardiac arrhythmias
- D. muscle atrophy
- E. skin calcifications

38. Juvenile localized scleroderma (JLS) is generally insidious with skin manifestations that vary according to disease subtype. Up to 25% of children with LS have extracutaneous manifestations, including arthritis and neurological symptoms.

Of the following, the subtype that is **MOST** commonly associated with neurological manifestations is

- A. plaque morphea
- B. eosinophilic fasciitis
- C. generalized morphea
- D. en coup de sabre
- E. morphea profunda

39. Juvenile systemic scleroderma (JSS) has a prolonged course with periods of remission and exacerbation. It is characterized by multisystem organ involvement including skin, locomotor system, CNS, and viscera.

Of the following, the **MOST** common early cutaneous manifestation is

- A. edema of the dorsum of the hands and fingers
- B. induration and fibrosis of the skin
- C. flexion contractures at the elbows, hips, and knees
- D. skin ulceration over pressure points
- E. subcutaneous calcifications

40. You are discussing Raynaud phenomenon (RP) and Raynaud disease (RD) with medical students. You state that RP is usually associated with rheumatic diseases, while RD is independent of an underlying rheumatic disease.

Of the following, the feature that is **MOST** likely consistent with RD, rather than RP is

- A. presence in early childhood
- B. absence of tissue necrosis and gangrene
- C. asymmetric occurrence
- D. presence of periungual telangiectasia
- E. associated pain and paresthesia

41. Behçet disease (BD) is a primary variable vessel vasculitis, characterized by exacerbations and remissions. The hallmark of the disease is oral ulceration.

All the following are characteristic features of the oral ulcers **EXCEPT**

- A. very painful
- B. recurrent
- C. involve any location in the oral cavity
- D. heal with scarring
- E. last 3-10 days

42. You are evaluating a 16-year-old female adolescent who has been diagnosed with Behçet disease since the age of 12 year; she recently develops oral genital ulcers; examination reveals multiple oral ulcers involving the hard palate, multiple genital ulcers involving the labia majora, and erythema nodosum. There is no major organ involvement and uveitis.

Of the following, the **BEST** treatment for this patient is

- A. colchicine
- B. azathioprine
- C. cyclophosphamide
- D. systemic steroids
- E. interferon alpha

43. Primary Sjögren syndrome, although rare, may occur in children between 9-10 year.

Of the following, the **MOST** common manifestation in children is

- A. recurrent parotitis
- B. sicca symptoms
- C. polyarthritis
- D. vulvovaginitis
- E. hepatitis

44. Familial Mediterranean fever (FMF) is a recessively inherited autoinflammatory disease usually characterized by recurrent self-limited episodes of fever, serositis, arthritis, and skin rash.

Of the following, the hallmark cutaneous finding of this disease is

- A. erysipelas-like erythema overlying the dorsum of the foot
- B. morbilliform rash
- C. migratory rash overlying area of myalgia
- D. cold-induced urticaria like lesions
- E. generalized pustulosis

45. Amyloidosis is the most serious complication of familial Mediterranean fever (FMF), and in its absence FMF patients may live a normal life span.

Of the following, the organ that is not affected by secondary amyloidosis of FMF is

- A. kidney
- B. lung
- C. nerve
- D. heart
- E. teste

46. Amyloidosis is a disease caused by protein misfolding; the type referred as amyloid A (AA), amyloidosis usually develops in patients with chronic inflammatory states, especially juvenile idiopathic arthritis.

In which of the following subtypes of JIA there is a highest prevalence of AA amyloidosis?

- A. systemic JIA
- B. oligoarticular JIA
- C. polyarticular JIA
- D. psoriatic arthritis
- E. enthesitis-related arthritis

47. Although there is no established therapy to AA amyloidosis, colchicine may be an effective drug to prevent the development of amyloidosis.

Of the following, the disease that is respond to colchicine in preventing AA amyloidosis is

- A. juvenile idiopathic arthritis

- B. ankylosing spondylitis
- C. familial Mediterranean fever
- D. hyper IgD syndrome
- E. cryopyrin-associated periodic syndrome

48. Kawasaki disease (KD) is an acute febrile illness of childhood characterized by vasculitis with a predilection for the coronary arteries.

Predictors of poor outcome across several studies include all the following **EXCEPT**

- A. old age
- B. male gender
- C. persistent fever
- D. poor response to IVIG
- E. elevated C-reactive protein levels

49. Kawasaki disease (KD) has unique clinical manifestations; however, less consistent clinical presentation may occur.

All the following are common classic clinical features of KD **EXCEPT**

- A. bilateral non exudative bulbar conjunctivitis
- B. cracked lips
- C. edema and erythema of the hands and feet
- D. vesicular rash
- E. unilateral non suppurative cervical lymphadenopathy

50. You are evaluating a 1-year-old boy with an established diagnosis of Kawasaki disease (KD) since 4 days; the caring nurse asks you about cardiac involvement during this stage.

All the following are possible cardiac complications at this stage **EXCEPT**

- A. myocarditis
- B. pericarditis
- C. coronary artery aneurysm
- D. mitral regurgitation
- E. cardiogenic shock

51. A follow up two-dimensional echocardiography is performed to a 1.5-year-old boy with Kawasaki disease (KD) 3 weeks after the diagnosis; it shows a small solitary aneurysm of the left anterior descending coronary artery without thrombosis.

Of the following, the **MOST** appropriate therapy for this child is

- A. aspirin for 6 months
- B. life-long aspirin
- C. IVIG and aspirin for 14 days
- D. aspirin and clopidogrel antiplatelet for 8 weeks
- E. aspirin and warfarin for 6 months

52. Henoch-Schönlein purpura (HSP) is the most common vasculitis of childhood affecting small vessels in the skin, joints, gastrointestinal tract, and kidney. Gastrointestinal involvements occur in up to 80% of children with HSP.

Of the following, the **LEAST** gastrointestinal manifestation that may occur in children with HSP is

- A. abdominal pain
- B. vomiting and diarrhea
- C. paralytic ileus
- D. melena
- E. intussusception

53. A 5-year-old boy has acute onset of abdominal pain and melena; he has been diagnosed with Henoch-Schönlein purpura (HSP) one week before. Examination reveals symmetrical palpable purpura overlying the lower extremities and buttock; other examinations are unremarkable.

Of the following, the **BEST** treatment for this boy is

- A. supportive measures
- B. steroids
- C. azathioprine
- D. cyclophosphamide
- E. mycophenolate mofetil

54. Renal disease is the major long-term complication, occurring in 1-2% of children with Henoch-Schönlein purpura (HSP). Chronic HSP renal disease is managed with a variety of immunosuppressants.

Of the following, the **LEAST** effective drug for the treatment of renal disease in HSP is

- A. prednisolone
- B. azathioprine
- C. cyclophosphamide
- D. cyclosporine
- E. mycophenolate mofetil

55. Childhood vasculitis encompasses a broad spectrum of diseases that share in common inflammation of the blood vessels. Vascular injury includes small, medium, and large vessels.

Of the following, the disease that affects predominantly large blood vessels is

- A. Henoch-Schönlein purpura
- B. granulomatosis with polyangiitis (wegener granulomatosis)
- C. childhood polyarteritis nodosa
- D. Kawasaki disease
- E. Takayasu arteritis

- 1.(C). Certain agents (indomethacin) have a higher risk of toxicity than others (ibuprofen); naproxen has an intermediate risk. Selective COX-2 inhibitors (such as celecoxib and meloxicam) inhibit receptors responsible for promoting inflammation with potential for fewer gastrointestinal adverse effects.
- 2.(D). Naproxen is more likely than other NSAIDs to cause pseudoporphyria. Pseudoporphyria is more likely to occur in fair-skinned individuals and on sun-exposed areas. If pseudoporphyria develops; the inciting NSAID should be discontinued because scars can persist for years or be permanent.
- 3.(E). The most significant potential adverse effect is retinal toxicity, which occurs rarely but results in irreversible color blindness or loss of central vision. Complete ophthalmologic examinations, including assessment of peripheral vision and color fields, are conducted at baseline and every 6-12 mo to screen for retinal toxicity.
- 4.(C). Sulfasalazine is generally considered contraindicated in children with active systemic JIA because of increased hypersensitivity reactions.
- 5.(A). Nausea, vomiting, anorexia, alopecia, mucositis, hemorrhagic cystitis, and bone marrow suppression are potential short-term adverse effects.
- 6.(C). Involved joints are often swollen, warm to touch, and painful on movement or palpation with reduced range of motion, but usually are not erythematous.
- 7.(B). In oligoarthritis, it predominantly affects the large joints of the lower extremities, such as the knees and ankles. Isolated involvement of upper extremity large joints is less common.
- 8.(A). Isolated involvement of the hip is almost never a presenting sign and suggests extended oligoarticular juvenile idiopathic arthritis.
- 9.(B). The presence of a positive ANA confers increased risk for asymptomatic anterior uveitis, requiring periodic slit-lamp examination.
- 10.(D). ANA positivity is more likely to be correlated with asymmetric arthritis.
- 11.(D). Rheumatoid nodules on the extensor surfaces of the elbows, spine, and over the Achilles tendons, although unusual, are associated with a more severe course and almost exclusively occur in RF-positive individuals.
- 12.(E). The most important indicators of MAS include a falling platelet count, extreme hyperferritinemia, increased liver enzymes, falling leukocyte count, persistent, continuous fever $\geq 38^{\circ}\text{C}$, falling ESR, hypofibrinogenemia, and hypertriglyceridemia. The diagnosis is confirmed by bone marrow biopsy demonstrating hemophagocytosis.
- 13.(A). NSAIDs alone rarely induces remission in children with polyarthritis or systemic JIA. Methotrexate is the oldest and least toxic effective drug. Biologic medications that

inhibit proinflammatory cytokines, such as TNF- α , IL-1, and IL-6, demonstrated excellent disease control.

14.(B). TNF inhibition is not as effective for the systemic symptoms found in SJIA. When systemic symptoms dominate systemic steroids are started followed by the initiation of IL-1 or IL-6 antagonist therapy, which often induces a dramatic and rapid response.

15.(D). Systemic steroids do not prevent joint destruction.

16.(E). There is no association between the activity or severity of arthritis and uveitis.

17.(E). Disease involving the hip and hand and wrist is associated with a poorer prognosis and may lead to significant functional impairment. All other mentioned factors carry a good outcome.

18.(D). Symptomatic, rather than asymptomatic, eye inflammation (acute anterior uveitis) is a clinical feature suggestive of spondyloarthritis.

19.(D). In ERA, the arthritis is typically asymmetric and involves 4 or fewer joints during the 1st 6mo of the disease. The most frequently affected joints are the knees, ankles, and hips. Enthesitis is typically symmetric and most commonly affects the lower limbs. Inflammation of the small joints of the foot, or tarsitis, is highly suggestive of ERA.

20.(E). Positive rheumatoid factor is never associated with all subtypes of spondyloarthritis.

21.(B). In comparison to adult-onset AS, axial disease and inflammatory back pain are less frequent at disease onset, while enthesitis and peripheral arthritis is more common.

22.(A). Reactive arthritis typically follows enteric infection with *Salmonella* sp., *Shigella flexneri*, *Yersinia enterocolitica*, *Campylobacter jejuni*, orogenitourinary tract infection with *Chlamydia trachomatis*. *Escherichiacoli* and *Clostridium difficile* are also causative enteric agents, although less common.

23.(C). Postinfectious arthritis does not necessarily share the typical spondyloarthritis pattern of joint involvement.

24.(D). Rubella-associated arthropathy may follow natural rubella infection and, infrequently, rubella immunization. It typically occurs in young women, with an increased frequency with advancing age, and is uncommon in preadolescent children and in males. Arthralgia of the knees and hands usually begins within 7 days of onset of the rash or 10-28 days after immunization.

25.(D).

26.(C). In transient synovitis (toxic synovitis), aspiration of joint fluid is often necessary to exclude septic arthritis and typically results in dramatic clinical improvement.

27.(B).

28.(C). Childhood SLE is rare before 5 yr of age.

29.(B).

30.(A).

31.(A). ANA titers are not reflective of disease activity; therefore, repeating ANA titers is not helpful in disease management.

32.(E). Hydroxychloroquine is recommended for all individuals with SLE if tolerated. In addition to treating mild SLE manifestations such as rash and mild arthritis, hydroxychloroquine prevents SLE flares, improves lipid profiles, and may have a beneficial impact on mortality and renal outcomes. Corticosteroids are a mainstay for treatment of significant manifestations of SLE. Methotrexate, leflunomide, and azathioprine are often used to treat persistent moderate disease, including arthritis, significant cutaneous or hematologic involvement, and pleural disease. Cyclophosphamide is reserved for the most severe, potentially life threatening SLE manifestations, such as renal, neurologic, and cardiopulmonary disease.

33.(D). The most serious complication due to neonatal lupus is congenital heart block.

34.(E).

35.(C). In JDM, facial erythema is crossing the nasolabial folds, in contrast to the malar rash without nasolabial involvement typical of systemic lupus erythematosus.

36.(D). Bed rest is not indicated, because weight bearing improves bone density and prevents contractures.

37.(C). Cardiac involvement by JDM is rare.

38.(D). Children with en coup de sabre may have symptoms unique to central nervous system involvement, such as seizures, hemifacial atrophy, ipsilateral uveitis, and learning/behavioral changes.

39.(A).

40.(B). RD often begins in adolescence and is characterized by symmetric occurrence, the absence of tissue necrosis and gangrene, and the lack of manifestations of an underlying rheumatic disease. Children have normal nail-fold capillaries. Pain and paresthesia indicate ischemic changes, associated only with RP due to rheumatic diseases.

41.(D). The oral ulcers heal without scarring. In contrast; the genital ulcers heal with scars.

42.(A). In patients without major organ involvement, colchicine significantly improves oral and genital ulcers, skin features, and disease activity. In pediatric patients with vascular involvement with venous thrombosis, steroids and azathioprine have been used, whereas those with pulmonary arterial or cardiac involvement are initially treated with cyclophosphamide. Interferon alpha can be used for CNS vasculitis.

43.(A). Recurrent parotid gland enlargement and parotitis are the most common manifestations in children (>70%), whereas sicca syndrome (dry mouth, painful mucosa, sensitivity to spicy foods, halitosis, widespread dental caries) predominate in adults.

44.(A).

45.(C). Amyloidosis may develop most commonly in the kidneys, gastrointestinal tract, spleen, lungs, testes, thyroid, and adrenals. Rarely, cardiac amyloidosis may develop; macroglossia and amyloid neuropathy is generally not seen with the amyloidosis of FMF.

- 46.(A). JIA is a rheumatic disease that is associated with the development of AA amyloidosis with the highest prevalence in patients with systemic JIA followed by those with polyarticular disease.
- 47.(C). Unlike AA amyloidosis associated with FMF, AA amyloidosis associated with other autoinflammatory diseases (including TRAPS, cryopyrin-associated periodic syndrome, and HIDS) and chronic rheumatic diseases (JIA, RA, and ankylosing spondylitis) do not respond to colchicine.
- 48.(A). Young age is poor predictor. Other lab findings associated with poor outcome are neutrophilia, thrombocytopenia, transaminitis, hyponatremia, hypoalbuminemia, elevated levels of N-terminal-probrain natriuretic protein.
- 49.(D). Typical rash is maculopapular, erythema multiforme, or scarlatiniform. Bullous, pustular, or vesicular rashes are uncommon skin manifestations.
- 50.(C). Coronary artery aneurysm usually develops in the subacute phase in the 2nd or 3rd week of the illness.
- 51.(B). The patient with KD who has had a small solitary aneurysm should continue aspirin indefinitely. Patients with larger or numerous aneurysms may require the addition of other antiplatelet agents or anticoagulation.
- 52.(E). Intussusception, mesenteric ischemia, and intestinal perforation are uncommon in children with HSP.
- 53.(B). Steroid is most often used to treat significant gastrointestinal involvement or other life-threatening manifestations.
- 54.(A).
- 55.(E).

1. Approximately 80% of blood cultures that will be positive are identified at
 - A. 1st 24 hr from incubation
 - B. 25 – 48 hr from incubation
 - C. after 3 day from incubation
 - D. after 5 day from incubation
 - E. after 7 day from incubation

2. The optimal amount of blood to collect from a pediatric patient for blood culture depend on
 - A. age
 - B. weight
 - C. length
 - D. BMI
 - E. laboratory standards

3. The IgM response occurs earlier in the illness, generally peaking at 7-10 days after infection, and usually disappears within a few weeks, but for some infections it can persist for months such as
 - A. measles
 - B. mumps
 - C. rubella
 - D. hepatitis A
 - E. varicella

4. Vaccines are defined as whole or parts of microorganisms administered to prevent an infectious disease.
Which of the following is a live attenuated vaccine?
 - A. hepatitis A
 - B. hepatitis B
 - C. pneumococcal
 - D. varicella
 - E. diphtheria

5. Which of the following is a T-lymphocyte independent vaccine?
 - A. hepatitis A
 - B. hepatitis B

- C. pneumococcal
 - D. varicella
 - E. diphtheria
6. Rotavirus vaccine should not be initiated for infants older than
- A. 11 wk
 - B. 13 wk
 - C. 15 wk
 - D. 17 wk
 - E. 19 wk
7. The final dose of rotavirus vaccine must be administered no later than
- A. 6 mo of age
 - B. 8 mo of age
 - C. 10 mo of age
 - D. 12 mo of age
 - E. 18 mo of age
8. Immunization is one of the most beneficial and cost-effective disease prevention measures. As a result of effective and safe vaccines, which of the following diseases has been eradicated?
- A. smallpox
 - B. polio
 - C. measles
 - D. rubella
 - E. pertussis
9. Infants, children, and adolescents in Iraq are routinely immunized against
- A. 10 diseases
 - B. 11 diseases
 - C. 12 diseases
 - D. 13 diseases
 - E. 14 diseases
10. The **MOST** common adverse reaction to intramuscular immunoglobulin is
- A. pain at the injection site
 - B. flushing
 - C. headache
 - D. chills
 - E. nausea
11. All the following are major recommended indications for IVIG **EXCEPT**

- A. replacement therapy for primary immunodeficiency disorders
- B. Kawasaki disease
- C. hepatitis A prophylaxis
- D. immune-mediated thrombocytopenia
- E. prophylaxis of infection following bone marrow transplantation

12. Serious reactions to IVIG include all the following **EXCEPT**

- A. anaphylactoid events
- B. thromboembolic disorders
- C. aseptic meningitis
- D. carditis
- E. renal insufficiency

13. A toxoid is a modified bacterial toxin that is made non-toxic but still able to induce an active immune response against the toxin.

Which of the following vaccine is a toxoid?

- A. hepatitis A
- B. hepatitis B
- C. pneumococcal
- D. varicella
- E. diphtheria

14. Hepatitis A vaccine, licensed for administration to children 12 mo of age and older. The 2 doses in the series should be separated by at least

- A. 2 mo
- B. 4 mo
- C. 6 mo
- D. 1 yr
- E. 2 yr

15. The minimum interval between the 2 doses of MMR is

- A. 2 wk
- B. 4 wk
- C. 2 mo
- D. 6 mo
- E. 1 yr

16. The minimum age for the last dose of hepatitis B vaccine is

- A. 16 weeks
- B. 20 weeks
- C. 24 weeks
- D. 28 weeks

E. 32 weeks

17. Preterm infant, weight 1600 gm, should not receive the following vaccine at birth

- A. BCG
- B. hepatitis B, if born to a HBs Ag negative mother
- C. polio
- D. DPT
- E. MMR

18. Which of the following vaccines is contraindicated for a patient with X-linked agammaglobulinemia?

- A. BCG
- B. hepatitis B
- C. DPT
- D. MMR
- E. varicella

19. Which of the following vaccines is contraindicated for a patient with chronic renal disease?

- A. pneumococcal
- B. hepatitis B
- C. live attenuated influenza
- D. varicella
- E. hepatitis A

20. Waterless hand hygiene products are effective in killing most microbes but do not remove dirt or debris and are ineffective against

- A. Pseudomonas
- B. hepatitis A
- C. Salmonella
- D. C. difficile
- E. S. aureus

21. All the following are recognized skin infections or infestations in children in childcare **EXCEPT**

- A. impetigo
- B. pediculosis
- C. scabies
- D. erythrasma
- E. tinea corporis

22. A 3-year-old child diagnosed with shigella infection, he can return to his daycare

- A. 2 days after initiation of treatment
- B. after diarrhea resolves
- C. after diarrhea resolves and results of a single stool culture is negative for these organisms
- D. after diarrhea resolves and results of 2 stool cultures are negative for these organisms
- E. after diarrhea resolves and results of 3 stool cultures are negative for these organisms

23. Oral rehydration is the mainstay of treatment for pediatric traveler's diarrhea and the drug of choice is

- A. metronidazol
- B. azithromycin
- C. amoxicillin
- D. trimethoprim-sulfamethoxazol
- E. erythromycin

24. Fever is defined as a rectal temperature of

- A. $\geq 37.5^{\circ}\text{C}$
- B. $\geq 37.6^{\circ}\text{C}$
- C. $\geq 38^{\circ}\text{C}$
- D. $\geq 38.1^{\circ}\text{C}$
- E. $\geq 38.5^{\circ}\text{C}$

25. Three mechanisms can produce fever: pyrogens, heat production exceeding loss, and defective heat loss. Endogenous pyrogens include

- A. antigen-antibody complexes
- B. complement components
- C. lymphocyte products
- D. interferons β and γ
- E. androgenic steroid metabolites

26. Drugs that are known to cause fever include

- A. chlorpheniramine
- B. allopurinol
- C. diphenhydramine
- D. acetazolamide
- E. adenosine

27. Ingestion of dirt is an important clue to infection with

- A. amebiasis
- B. giardiasis

- C. malaria
- D. trichinosis
- E. toxoplasmosis

28. Bulbar conjunctivitis in a child with FUO suggests

- A. leptospirosis
- B. coxsackievirus infection
- C. tuberculosis
- D. infectious mononucleosis
- E. lymphogranuloma venereum

29. Multiple blood cultures may be required to detect bacteremia associated with

- A. malaria
- B. pyelonephritis
- C. brucellosis
- D. osteomyelitis
- E. pneumonia

30. Primary immunodeficiencies are compromised states that result from genetic defects affecting 1 or more arms of the immune system while secondary immunodeficiencies result from infection, malignancy, or as an adverse effect of immunomodulating or immunosuppressing medications.

Which of the following represent secondary immune deficiency?

- A. Shwachman-Diamond syndrome
- B. cystic fibrosis
- C. Chédiak-Higashi syndrome
- D. Omenn syndrome
- E. ataxia-telangiectasia

31. Which type of fever is persistent and varies by more than 0.5°C (0.9°F)/day?

- A. intermittent
- B. hectic
- C. sustained
- D. remittent
- E. relapsing

32. All the following are causes of very high temperatures (>41°C) **EXCEPT**

- A. malignant hyperthermia
- B. malignant neuroleptic syndrome
- C. infection
- D. drug fever
- E. heat stroke

33. Relative bradycardia (when the pulse rate remains low in the presence of fever) can accompany all the following conditions **EXCEPT**

- A. typhoid fever
- B. brucellosis
- C. leptospirosis
- D. visceral leishmaniasis
- E. drug fever

34. The **MOST** common serious bacterial infection in infant aged 1-3 mo is

- A. pyelonephritis
- B. meningitis
- C. pneumonia
- D. septic arthritis
- E. osteomyelitis

35. Neutropenia is defined as an absolute neutrophil count of less than

- A. 1,000 cells/mm³
- B. 1,500 cells/mm³
- C. 2,000 cells/mm³
- D. 2,500 cells/mm³
- E. 3,000 cells/mm³

36. Leukocyte adhesion defects are caused by defects in the β chain of integrin (CD18), which is required for the normal process of neutrophil aggregation and attachment to endothelial surfaces.

It is characterized by all the following **EXCEPT**

- A. delayed cord separation
- B. recurrent infections
- C. ecthyma gangrenosum
- D. neutropenia
- E. survival is usually <10 yr

37. Penicillins are the drugs of choice for pediatric infections caused by the following **EXCEPT**

- A. group A Streptococcus
- B. H .influenzae
- C. Treponema pallidum
- D. L. monocytogenes
- E. N. meningitides

38. Cefixime is a third-generation cephalosporin active against all the following **EXCEPT**

- A. Streptococci

- B. Staphylococci
- C. H. influenza
- D. Neisseria gonorrhoeae
- E. Proteus vulgaris

39. Cephalexin is a cephalosporin active against *S. aureus*, *Streptococcus*, *E. coli*, *Klebsiella*, and *Proteus*. To which generation of cephalosporins it belongs?

- A. 1st
- B. 2nd
- C. 3rd
- D. 4th
- E. 5th

40. Meropenem is a carbapenem antibiotic with broad-spectrum activity but has no activity against

- A. *P. aeruginosa*
- B. *S. maltophilia*
- C. *L. monocytogenes*
- D. *S. aureus*
- E. *N. meningitidis*

41. Metronidazole is highly effective in the treatment of infections caused by anaerobes. It can increase the level of which of the following drugs if given at the same time

- A. carbamazepine
- B. rifampin
- C. phenobarbital
- D. phenytoin
- E. valproic acid

42. Cephalosporins are widely used in pediatric practice, both in oral and parenteral formulations. Which of the following is a 2nd generation cephalosporins?

- A. cefazolin
- B. cephalexin
- C. cefuroxime
- D. ceftazidime
- E. ceftaroline

43. Which cephalosporin should not be mixed or reconstituted with a calcium-containing product, such as Ringer solution or parenteral nutrition containing calcium?

- A. cefazolin
- B. cefotaxime

- C. ceftriaxone
- D. ceftazidime
- E. cefepime

44. Toxic shock syndrome is an acute and potentially severe illness characterized by all the following **EXCEPT**

- A. desquamation on the hands and feet
- B. myalgias
- C. focal neurologic abnormalities
- D. conjunctival hyperemia
- E. strawberry tongue

45. Which of the following represent a major criterion for the diagnosis of staphylococcal toxic shock syndrome?

- A. rash
- B. conjunctival hyperemia
- C. myalgia
- D. thrombocytopenia
- E. vomiting

46. Kawasaki disease closely resembles toxic shock syndrome clinically. However, many of the clinical features of toxic shock syndrome are usually absent or rare in Kawasaki disease like

- A. fever unresponsive to antibiotics
- B. hyperemia of mucous membranes
- C. erythematous rash
- D. desquamation
- E. diffuse myalgia

47. *S. pneumoniae* is the most frequent cause of bacteremia, bacterial pneumonia, otitis media, and bacterial meningitis in children. Children at increased risk of pneumococcal infections include those with the following conditions **EXCEPT**

- A. megaloblastic anemia
- B. deficiencies in humoral immunity
- C. HIV infection
- D. cerebrospinal fluid leak
- E. cochlear implants

48. Group A streptococcus can be subdivided into more than 220 serotypes on the basis of the M protein antigen, M serotyping is valuable for epidemiologic studies; specific group A streptococcus diseases tend to be associated with certain M types. Of the following, the M type associated with glomerulonephritis is

- A. 1
- B. 6
- C. 12
- D. 18
- E. 29

49. Scarlet fever is an upper respiratory tract infection associated with a characteristic rash.

Of the following, which statement is **TRUE**?

- A. it is caused by an infection with pyrogenic endotoxin producing group A streptococcus
- B. the rash appears 96 hr after onset of symptoms
- C. the rash begins to fade after 1-2 weeks
- D. before desquamation, the reddened papillae are prominent, giving the tongue a strawberry appearance
- E. the milder form can be confused with Kawasaki disease

50. Impetigo (or pyoderma) has traditionally been classified into 2 clinical forms: bullous and nonbullous.

Of the following, which statement is **TRUE**?

- A. bullous impetigo is more common
- B. nonbullous lesions are most common on the trunk and perineum
- C. regional lymphadenitis is commonly associated with nonbullous lesions
- D. nonbullous impetigo is generally accompanied by fever
- E. bullous impetigo usually involve the face and extremities

51. In which of the following circumstances the diagnosis of acute rheumatic fever can be made without strict adherence to Jones criteria?

- A. when chorea occurs as the only major manifestation of acute rheumatic fever
- B. when indolent carditis is the only manifestation months after the apparent onset of acute rheumatic fever
- C. in a limited number of patients with recurrences of acute rheumatic fever in particularly high-risk populations
- D. all of the above
- E. none of the above

52. Because no clinical or laboratory finding is pathognomonic for acute rheumatic fever, 5 major and 4 minor criteria with evidence of recent group A streptococcus infection are required for the diagnosis. Migratory polyarthritits represent one of the major criteria, which of the following statements is **TRUE** regarding this major criterion?

- A. occurs in approximately 50% of patients

- B. typically involves small joints
 - C. the pain can precede and can appear to be disproportionate to the objective findings
 - D. rheumatic arthritis is almost deforming
 - E. there is often a proportional relationship between the severity of arthritis and the severity of cardiac involvement
53. Acute rheumatic carditis usually presents as cardiac murmurs and
- A. tachycardia
 - B. chest pain
 - C. dyspnea
 - D. syncopal attack
 - E. tachypnea
54. Patients with acute rheumatic carditis and more than minimal cardiomegaly should receive prednisone 2 mg/kg/day in 4 divided doses for
- A. 7-10 days
 - B. 2-3 weeks
 - C. 6-8 weeks
 - D. 2-3 months
 - E. 4-6 months
55. The drug of choice for Sydenham chorea is
- A. aspirin
 - B. prednisone
 - C. phenobarbital
 - D. haloperidol
 - E. chlorpromazine
56. To prevent first attack of acute rheumatic fever after acute group A streptococcus pharyngitis, appropriate antibiotic therapy should be instituted before
- A. 3rd day of illness
 - B. 5th day of illness
 - C. 7th day of illness
 - D. 9th day of illness
 - E. 11th day of illness
57. Prophylaxis for people who have had acute rheumatic fever with carditis but without residual heart disease persist for
- A. 5 yr or until 21 yr of age, whichever is longer
 - B. 10 yr or until 21 yr of age, whichever is longer
 - C. 5 yr or until 40 yr of age, whichever is longer

- D. 10 yr or until 40 yr of age, whichever is longer
- E. Lifelong

58. Group B streptococcus (GBS), or *Streptococcus agalactiae*, is a major cause of neonatal bacterial sepsis.

Of the following, the **MOST** common syndrome associated with childhood GBS disease beyond early infancy is

- A. bacteremia without a focus
- B. meningitis
- C. ventriculitis
- D. septic arthritis
- E. pneumonia

59. Recommended duration of therapy for manifestations of group B streptococcus are as follow

- A. bacteremia without a focus: 7 days
- B. meningitis: 3-6 weeks
- C. ventriculitis: at least 8 weeks
- D. septic arthritis: 3-4 weeks
- E. osteomyelitis: 6-8 weeks

60. Diphtheria is an acute toxic infection; toxic cardiomyopathy occurs in 10-25% of patients with respiratory diphtheria and is responsible for 50-60% of deaths.

Of the following, the mainstay of therapy is

- A. antitoxin
- B. penicillins
- C. erythromycin
- D. clindamycin
- E. rifampin

61. A painless, slow-growing, hard mass producing cutaneous fistulas, a condition commonly known as lumpy jaw is usually caused by

- A. *Staphylococcus*
- B. *Actinomyces*
- C. *Nocardia*
- D. *Yersinia*
- E. *Leptospira*

62. The **MOST** common secondary site involved in cases of pulmonary nocardiosis is

- A. brain
- B. skin
- C. kidney

- D. liver
- E. bone

63. Which of the following is a gram negative bacterium?

- A. *Staphylococcus aureus*
- B. *Streptococcus pneumoniae*
- C. *Actinomyces*
- D. *Haemophilus influenza*
- E. *Nocardia*

64. Regarding epidemiology of *Neisseria meningitidis*, all the following are true **EXCEPT**

- A. meningococci are transmitted during close contact via aerosol droplets or exposure to respiratory secretions
- B. meningococci survive for long periods in the environment
- C. smoking and respiratory viral infection are associated with increased rates of carriage and disease
- D. the highest rate of meningococcal disease occurs in infancy
- E. most cases of meningococcal disease are sporadic

65. The **MOST** common clinical manifestation of meningococcal infection is

- A. asymptomatic carriage
- B. meningococcal meningitis
- C. bacteremia without sepsis
- D. meningococcal septicemia
- E. pneumonia

66. In meningococcal purpura, necrotic skin lesions are less common among children treated with

- A. penicillin G
- B. ampicillin
- C. cefotaxime
- D. ceftriaxone
- E. meropenem

67. The **MOST** common complication of acute severe meningococcal septicemia is

- A. arthritis
- B. focal skin infarction
- C. endocarditis
- D. pneumonia
- E. peritonitis

68. A poor prognostic factor for invasive meningococcal disease on presentation is

- A. hypertension
- B. leukocytosis
- C. alkalosis
- D. meningitis
- E. normal erythrocyte sedimentation rate

69. Of the following, the **MOST** effective agent for prophylaxis of meningococcal disease is

- A. ceftriaxone
- B. rifampin
- C. ampicillin
- D. penicillin
- E. amoxicillin

70. Gonorrhea is manifested by a spectrum of clinical presentations from asymptomatic carriage, to the characteristic localized urogenital infections, to disseminated systemic infection.

Regarding disseminated gonococcal infection, the following statement is **TRUE**

- A. hematogenous dissemination occurs in 10-30% of all gonococcal infections
- B. men account for the majority of cases
- C. meningitis and osteomyelitis are the most common manifestations
- D. skin lesions found in 75% of patients
- E. acute endocarditis is an uncommon but often fatal manifestation

71. All patients who are presumed or proven to have gonorrhea should be evaluated for concurrent presence of all the following infections **EXCEPT**

- A. syphilis
- B. hepatitis B
- C. HIV
- D. C. trachomatis
- E. HSV2

72. Children who have bacteremia or arthritis caused by gonococcal infections should be treated with ceftriaxone (50 mg/kg/day; maximum: 1 g/day if weighs <45 kg) for a minimum of

- A. 3 days
- B. 5 days
- C. 7 days
- D. 14 days
- E. 21 days

73. The **MOST** common etiology of joint and bone infections in young children is

- A. Staphylococcus aureus
- B. Streptococcus pneumoniae
- C. Kingella kingae
- D. Haemophilus influenza
- E. Neisseria meningitidis

74. Diagnosis of chancroid in infants and children is a strong evidence of sexual abuse.

Chancroid is caused by

- A. Haemophilus ducreyi
- B. Syphilis
- C. C. trachomatis
- D. Kingella kingae
- E. Neisseria gonorrhea

75. The **MOST** important clinical manifestation of M.catarrhalis infection in children is

- A. pneumonia
- B. bronchitis
- C. otitis media
- D. pharyngitis
- E. cystitis

76. The **MOST** common reason for which children receive antibiotics is

- A. pneumonia
- B. bronchitis
- C. otitis media
- D. pharyngitis
- E. cystitis

77. Infants younger than 3 mo of age with suspected pertussis usually are admitted to hospital, as are many between 3 and 6 mo of age unless witnessed paroxysms are not severe, as well as are patients of any age if significant complications occur.

Typical paroxysms that are not life threatening have the following feature

- A. duration >45 sec
- B. blue color change
- C. bradycardia <60 beats/min in infants
- D. oxygen desaturation that spontaneously resolves at the end of the paroxysm
- E. post-tussive unresponsiveness

78. All the following regarding assessment and care of infants with pertussis are true **EXCEPT**

- A. infants with potentially fatal pertussis may appear well between episodes

- B. a paroxysm must be witnessed before a decision is made between hospital and home care
- C. suctioning of nose, oropharynx, or trachea should be performed on a preventive schedule
- D. feeding in the period following a paroxysm may be more successful than after napping
- E. family education, recruitment as part of the team, and continued support after discharge are essential

79. Hospital discharge of infants with pertussis is appropriate in all the following circumstances **EXCEPT**

- A. over a 24-hr period disease severity is unchanged or diminished
- B. intervention is not required during paroxysms
- C. nutrition is adequate
- D. no complication has occurred
- E. parents are adequately prepared for care at home

80. The preferable antibiotic in neonate with pertussis is

- A. azithromycin
- B. erythromycin
- C. clarithromycin
- D. trimethoprim-sulfamethoxazole
- E. amoxycilline

81. Echocardiography should be performed in critically ill infants with pertussis to detect presence of

- A. myocarditis
- B. heart failure
- C. congenital heart defect
- D. pulmonary hypertension
- E. right atrial dilatation

82. The least common clinical feature of typhoid fever in children is

- A. diarrhea
- B. abdominal pain
- C. pallor
- D. splenomegaly
- E. headache

83. All the following are clinical features of typhoid fever **EXCEPT**

- A. incubation period is usually 30-45 days
- B. macular or maculopapular rash (rose spots) may be visible around the 7th-10th

day of the illness

- C. if complications not occur, the symptoms and physical findings gradually resolve within 2-4 wk
- D. typhoid fever usually manifests as high-grade fever with a wide variety of associated features
- E. relative bradycardia, neurologic manifestations, and gastrointestinal bleeding, are rare in children

84. Regarding diagnosis of typhoid fever, one of the following is **TRUE**

- A. blood cultures are positive in 65-80% of the patients
- B. urine culture results become positive within the 1st wk
- C. thrombocytosis may be a marker of severe illness
- D. diagnosis by Widal test alone is prone to error
- E. leukocytosis is rare in young children

85. All the following are clinical features of shigellosis **EXCEPT**

- A. an incubation period of 12 hr to several days
- B. most children never progress to the stage of bloody diarrhea
- C. untreated diarrhea can last more than 4 weeks
- D. neurologic findings are the most common extraintestinal manifestations
- E. neonatal shigellosis is rare

86. Cholera is a dehydrating diarrheal disease caused by *Vibrio cholerae*, of more than 200 serogroups; the serogroups that have been associated with epidemics are

- A. O131
- B. O139
- C. O151
- D. O159
- E. O165

87. Persons with which blood group are at increased risk for developing severe disease of cholera?

- A. A
- B. B
- C. AB
- D. O
- E. Blood group not related to the severity of the disease

88. Seizure in cholera is commonly due to

- A. hypocalcemia
- B. hyponatremia
- C. hypernatremia

- D. hypoglycemia
- E. fever

89. Complications of campylobacter jejuni can include acute and late onset complications that may present after the acute infection has resolved.

Of the following, the **MOST** common late-onset complication is

- A. reactive arthritis
- B. immunoglobulin A nephropathy
- C. immune complex glomerulonephritis
- D. hemolytic anemia
- E. carditis

90. **MOST** Campylobacter isolates are susceptible to

- A. aminoglycosides
- B. cephalosporins
- C. rifampin
- D. penicillins
- E. trimethoprim

91. The genus Yersinia is a member of the family Enterobacteriaceae and comprises more than 14 named species, 3 of which are established as human pathogens.

Which of the following is **MOST** often associated with mesenteric lymphadenitis?

- A. Yersinia enterocolitica
- B. Yersinia pseudotuberculosis
- C. Yersinia pestis
- D. Yersinia mollaretii
- E. Yersinia rohdei

92. Patients with conditions leading to iron overload are at higher risk of developing infections with

- A. Aeromonas
- B. Pseudomonas aeruginosa
- C. Yersinia
- D. Francisella tularensis
- E. Campylobacter

93. The **MOST** common complication of Y. enterocolitica infection in younger children is

- A. reactive arthritis
- B. erythema multiforme
- C. hemolytic anemia
- D. thrombocytopenia

E. septicemia

94. Tularemia is a zoonotic infection caused by the gram-negative bacterium *Francisella tularensis*.

Of the following, the **MOST** common forms of tularemia diagnosed in children is

- A. ulceroglandular
- B. pneumonia
- C. oropharyngeal
- D. oculoglandular
- E. typhoidal

95. Human brucellosis is caused by organisms of the genus *Brucella* and continues to be a major public health problem worldwide.

All the following are true **EXCEPT**

- A. symptoms can be acute or insidious
- B. most patients present with fever, arthralgia/arthritis, and hepatosplenomegaly
- C. some present as a fever of unknown origin
- D. variable fever pattern
- E. invasion of the nervous system occurs in approximately 20% of cases

96. Which antibiotics should be avoided in patients with botulism?

- A. penicillin
- B. cephalosporin
- C. macrolide
- D. aminoglycoside
- E. trimethoprim-sulfamethoxazole

97. Tetanus is an acute spastic paralytic illness historically called lock-jaw that is caused by the neurotoxin produced by *Clostridium tetani*.

Of the following, the **TRUE** statement is

- A. tetanus is most often localized
- B. incubation period typically is 2-6 weeks
- C. patient remains conscious and there is no pain
- D. tetanic paralysis becomes more severe in the 4th wk after onset
- E. cephalic tetanus occurs in association with chronic otitis media

98. Worsening of acne in adolescent female on anti-tuberculous treatment is caused by

- A. isoniazid
- B. rifampin
- C. pyrazinamide
- D. ethambutol

E. streptomycin

99. Isoniazid is accompanied by significant drug–drug interactions, which of the following is **TRUE**?

- A. aluminum salts increase absorption of isoniazid
- B. isoniazid increase toxicity of carbamazepine
- C. rifampin decreased hepatotoxicity of isoniazid
- D. isoniazid decrease level of warfarin
- E. prednisolone decreased isoniazid metabolism

100. The ideal agent for treating fungal urinary tract infections is

- A. amphotericin B
- B. fluconazole
- C. voriconazole
- D. micafungin
- E. caspofungin

101. Candida is a common cause of oral mucous membrane infections (thrush) and perineal skin infections (Candida diaper dermatitis) in young infants.

All the following are true **EXCEPT**

- A. candida species are the third most common cause of bloodstream infection in premature infants
- B. up to 10% of full-term infants are colonized as the result of vertical transmission from the mother at birth
- C. histamine-2 blockers facilitate Candida colonization and overgrowth
- D. significant risk factors for neonatal invasive candidiasis include the presence of a central venous catheter
- E. the cumulative incidence is <0.3% among infants <750 g birthweight admitted to the NICU

102. NICUs with a high incidence of invasive candidiasis should consider prophylaxis with fluconazole in infants with a birthweight of

- A. <750 g
- B. <1,000 g
- C. <1,500 g
- D. <2,000 g
- E. <2,500 g

103. Antiviral chemotherapy typically requires a delicate balance between targeting critical steps in viral replication without interfering with host cellular function.

Which of the following antiviral blocks M2 protein ion channel?

- A. acyclovir

- B. amantadine
- C. ganciclovir
- D. foscarnet
- E. vidarabine

104. In neonates with HSV infection including CNS involvement, to improve neurodevelopmental outcome, suppressive therapy with oral acyclovir should be used for

- A. 3 mo
- B. 6 mo
- C. 9 mo
- D. 12 mo
- E. 18 mo

105. Acyclovir is a safe and effective therapy for herpes simplex virus (HSV) infections. All the following are true **EXCEPT**

- A. activity against CMV is less pronounced
- B. activity against Epstein-barr virus is modest, both in vitro and clinically
- C. acyclovir therapy in a nursing mother is not a contraindication to breastfeeding
- D. main route of elimination is hepatic
- E. high doses of acyclovir are associated with neurotoxicity

106. Ribavirin is a guanosine analog that has broad-spectrum activity against a variety of viruses, particularly RNA viruses.

All the following are true **EXCEPT**

- A. its precise mechanism of action is incompletely understood
- B. aerosolized ribavirin effective for parainfluenza, influenza, and measles infections
- C. ribavirin is generally nontoxic, particularly when administered by aerosol
- D. ribavirin and its metabolites concentrate in hepatocytes
- E. conjunctivitis and bronchospasm have been reported following exposure to aerosolized drug

107. The portal of entry of measles virus is through the respiratory tract or conjunctivae following contact with large droplets or small-droplet aerosols in which the virus is suspended.

Patients with measles are infectious

- A. 1 day before to up to 4-6 days after the onset of rash
- B. 2 days before to up to 4-6 days after the onset of rash
- C. 3 days before to up to 4-6 days after the onset of rash
- D. 4 days before to up to 4-6 days after the onset of rash
- E. 5 days before to up to 4-6 days after the onset of rash

108. Measles is a serious infection characterized by high fever, an enanthem, cough, coryza, conjunctivitis, and a prominent exanthem.

All the following are true **EXCEPT**

- A. incubation period is 8-12 days
- B. Koplik spots have been reported in 50-70% of measles cases
- C. the rash begins on the forehead, behind the ears, and on the upper neck
- D. the rash fades over about 3 days in the same progression as it evolved
- E. in more severe cases, generalized lymphadenopathy may be present

109. The **MOST** common complication of measles is

- A. aseptic meningitis
- B. pharyngitis
- C. otitis media
- D. pneumonia
- E. cystitis

110. Subacute sclerosing panencephalitis (SSPE) is a chronic complication of measles with a delayed onset and an outcome that is nearly always fatal.

SSPE is characterized by

- A. females are affected twice as often as males
- B. clinical manifestations begin insidiously 3-6 yr after primary measles infection
- C. the hallmark of the 1st stage is massive myoclonus
- D. the 3rd stage is characterized by loss of critical centers
- E. clinical trials using isoprinosine suggest significant benefit

111. The **MOST** common finding among infants with congenital rubella syndrome is

- A. psychomotor retardation
- B. cataracts
- C. deafness
- D. patent ductus arteriosus
- E. neonatal purpura

112. Mumps virus is in the family Paramyxoviridae and the genus Rubulavirus.

Mumps is characterized by

- A. incubation period ranges from 7-11 days
- B. unilateral parotitis rarely becomes bilateral
- C. pale opening of the Stensen duct
- D. parotid swelling peaks in approximately 7 days
- E. a morbilliform rash is rarely seen

113. The **MOST** common complication of mumps is

- A. meningitis

- B. conjunctivitis
- C. optic neuritis
- D. pneumonia
- E. thrombocytopenia

114. Intrauterine mumps infection have been associated with

- A. glaucoma
- B. limb anomalies
- C. pulmonary stenosis
- D. renal agenesis
- E. no fetal malformations

115. Mumps virus is neurotropic and is thought to enter the CNS via the choroid plexus and infect the choroidal epithelium and ependymal cells. CNS involvement is characterized by

- A. symptomatic CNS involvement occurs in 40-60% of infected individuals
- B. encephalitis most commonly manifests 10 days after the parotitis
- C. CNS symptoms usually resolve in 3 days
- D. CSF protein content is usually elevated
- E. facial palsy is a less-common CNS complication

116. Hematogenous dissemination of HSV to the central nervous system occur in

- A. neonates
- B. individuals with eczema
- C. severely malnourished children
- D. infants on steroids
- E. all the above

117. Herpes whitlow is HSV infection of the

- A. eye
- B. mouth
- C. fingers
- D. ear
- E. lip

118. The **MOST** common cause of recurrent aseptic meningitis is

- A. mumps
- B. EBV
- C. adenoviruses
- D. HSV
- E. influenza virus

119. In neonatal HSV encephalitis, skin vesicles occur in approximately

- A. 20%
- B. 40%
- C. 60%
- D. 80%
- E. 100%

120. In patients with neonatal HSV infection who receive IV treatment for 2-3 weeks then suppressive treatment for 6 months, they should be monitored by

- A. absolute neutrophil count ANC
- B. liver function tests
- C. renal function tests
- D. platelet count
- E. Hb%

121. Varicella-zoster virus (VZV) causes primary, latent, and recurrent infections.

All the following statements are true **EXCEPT**

- A. varicella is a serious disease in young infants
- B. within households, transmission of VZV occurs at a rate of 65-86%
- C. Herpes zoster is more common in winter
- D. the lifetime risk for herpes zoster for individuals with a history of varicella is 20-30%
- E. Herpes zoster is very rare in healthy children younger than 10 yr of age

122. Varicella is an acute febrile rash illness that is common in Iraq.

Of the following, the **TRUE** statement is

- A. it usually begins 4-6 days after exposure
- B. subclinical varicella is common
- C. mild abdominal pain may occur 24-48 hr before the rash appears
- D. temperature elevation usually as high as 41.1°C
- E. lesions often appear first on the extremities

123. All the following are true about varicella in unvaccinated individuals **EXCEPT**

- A. simultaneous presence of lesions in the same stages of evolution
- B. distribution of the rash is predominantly central
- C. many children have vesicular lesions on the eyelids
- D. exanthem may be much more extensive in children with skin disorders
- E. hypopigmentation or hyperpigmentation of lesion sites persists for days to weeks in some children

124. Encephalitis and acute cerebellar ataxia are well-described neurologic complications of varicella.

All the following are true **EXCEPT**

- A. morbidity from central nervous system complications is highest among patients younger than 5 yr and older than 20 yr
- B. nuchal rigidity, altered consciousness, and seizures characterize meningoencephalitis
- C. patients with cerebellar ataxia have a gradual onset of gait disturbance, nystagmus, and slurred speech
- D. neurologic symptoms usually begin 2-6 days after the onset of the rash but may occur during the incubation period or after resolution of the rash
- E. clinical recovery is typically occurring after 96 hr, and is usually gradual

125. Oral therapy with acyclovir (20 mg/kg/dose; maximum: 800 mg/dose) given as 4 doses/day for 5 days can be used to treat uncomplicated varicella in individuals at increased risk for moderate to severe varicella.

It includes all the following individuals **EXCEPT**

- A. nonpregnant individuals older than 12 yr of age
- B. individuals older than 12 mo of age with chronic cutaneous disorders
- C. individuals receiving short-term corticosteroid therapy
- D. individuals receiving long-term salicylate therapy
- E. individuals with chronic hematological disorders

126. Epstein-Barr virus (EBV) is shed in oral secretions after acute infection for

- A. 2 wk
- B. 4 wk
- C. 2 mo
- D. 4 mo
- E. 6 mo

127. What percent of the world's population infected by Epstein-Barr virus (EBV)?

- A. 5%
- B. 25%
- C. 50%
- D. 75%
- E. 95%

128. Infectious mononucleosis is the best-known clinical syndrome caused by Epstein-Barr virus (EBV). It is characterized by

- A. symptomatic hepatitis
- B. elevated liver enzymes
- C. massive splenic enlargement
- D. huge hepatomegaly
- E. occasional palatal petechiae

129. The **MOST** common long-term sequela associated with congenital CMV infection is
- A. renal failure
 - B. hearing loss
 - C. heart failure
 - D. vision loss
 - E. hepatic failure
130. Human herpesvirus 6 (HHV-6A and HHV-6B) and human herpesvirus 7 (HHV-7) cause infection in infancy and early childhood. HHV-6B is responsible for the majority of cases of roseola infantum (exanthema subitum or sixth disease).
- All the following are true **EXCEPT**
- A. 95% of children being infected with HHV-6 by 2 yr of age
 - B. peak age of primary HHV-6B infection is 6-9 mo of life
 - C. congenital infection with HHV-6 occurs in 1% of newborns
 - D. congenital infection with HHV-7 is well recognized
 - E. breast milk does not play a role in transmission of either HHV-6 or HHV-7
131. Amantadine and rimantadine are effective only against influenza A viruses and are not approved for use in children younger than
- A. 6 mo
 - B. 18 mo
 - C. 3 yr
 - D. 5 yr
 - E. 10 yr
132. Typically, the first sign of infection in infants with respiratory syncytial virus (RSV) is
- A. rhinorrhea
 - B. cough
 - C. low-grade fever
 - D. increased respiratory rate
 - E. subcostal retractions
133. For military services, vaccines are available for human adenoviruses types
- A. 1 and 4
 - B. 2 and 5
 - C. 3 and 6
 - D. 4 and 7
 - E. 5 and 8
134. Respiratory tract infections are common manifestations of human adenoviruses (HAdV) infections in children and adults.

All the following are true **EXCEPT**

- A. HAdVs cause 5-10% of all childhood respiratory disease
- B. primary infections in infants may manifest as bronchiolitis or pneumonia
- C. HAdV pneumonia may manifest as features more typical of bacterial disease
- D. pharyngitis typically includes symptoms of coryza, sore throat, and fever
- E. HAdV can be identified in <5% of children with isolated pharyngitis

135. Rotaviruses are in the Reoviridae family and cause disease in virtually all mammals and birds.

All the following statements are true **EXCEPT**

- A. infection typically begins after an incubation period of <48 hr (range: 1-7 days)
- B. fever, vomiting and frequent watery stools are present in about 50-60% of cases
- C. vomiting and fever typically abate after the 4th day of illness
- D. dehydration may develop and progress rapidly particularly in infants
- E. most severe disease typically occurs among children 4-36 mo of age

136. Common adverse effects of nitazoxinide include

- A. diarrhea
- B. flatulence
- C. increased appetite
- D. fever
- E. pruritus

137. One of the following statements regarding tinidazole is **TRUE**

- A. it is FDA approved for treatment of trichomoniasis
- B. it can be in infancy
- C. it should be given in 2 divided doses
- D. it is excreted via urine only
- E. it carries a pregnancy category A

138. All the following can cause human amoebic meningoencephalitis **EXCEPT**

- A. Naegleria
- B. Acanthamoeba
- C. Balamuthia
- D. Entamoeba histolytica
- E. Sappinia

139. One of the following statements regarding Giardia lamblia is **TRUE**

- A. life cycle is composed of 3 stages
- B. each ingested cyst produces 2 trophozoites in the caecum
- C. trophozoites contain 4 oval nuclei anteriorly

- D. cyst viability is not affected by the usual concentrations of chlorine used to purify water for drinking
- E. Giardia genotypes difference lead to wide spectrum of clinical manifestations

140. All the following statements regarding giardiasis are true **EXCEPT**

- A. incubation period of Giardia infection usually is 1-2 wk
- B. most infections are asymptomatic
- C. symptomatic infections occur more frequently in adults than in children
- D. stools do not contain blood, mucus, or fecal leukocytes
- E. malabsorption of sugars, fats, and fat-soluble vitamins is well documented

141. Giardiasis should be considered in children who have the following presentations **EXCEPT**

- A. acute dysenteric diarrhea
- B. persistent diarrhea
- C. failure to thrive
- D. malabsorption
- E. chronic crampy abdominal pain

142. Pregnant mother seek your advice about the stool examination of her asymptomatic 15 -month-old child which contain giardia cyst, your advice should be

- A. reassurance, no treatment
- B. inidazole 50 mg/kg once
- C. nitazoxinide 200 mg bid for 3 days
- D. metronidazole 15 mg/kg/day in 3 divided doses for 5-7 days
- E. albendazole 400 mg once a day for 5 days

143. The largest protozoan that parasitizes human is

- A. Entamoeba histolytica
- B. Giardia lamblia
- C. Balantidium coli
- D. Isospora belli
- E. Cryptosporidium

144. The leading protozoal cause of diarrhea in children worldwide is

- A. Entamoeba histolytica
- B. Giardia lamblia
- C. Balantidium coli
- D. Isospora belli
- E. Cryptosporidium

145. The **MOST** common cause of death in kala azar is

- A. severe anemia
- B. bleeding
- C. secondary bacterial infection
- D. hepatic failure
- E. renal failure

146. Hand-foot-mouth disease is one of the more distinctive rash syndromes; it is **MOST** frequently caused by

- A. coxsackievirus A6
- B. coxsackievirus A16
- C. enterovirus 71
- D. coxsackievirus B2
- E. coxsackievirus B12

1.(A).

2.(B).

3.(D). Hepatitis A and West Nile Virus.

4.(D). Vaccines can consist of whole inactivated microorganisms (e.g., polio and hepatitis A), parts of the organism (e.g., acellular pertussis, HPV, and hepatitis B), polysaccharide capsules (e.g., pneumococcal and meningococcal polysaccharide vaccines), polysaccharide capsules conjugated to protein carriers (e.g., Hib, pneumococcal, and meningococcal conjugate vaccines), live attenuated microorganisms (measles, mumps, rubella, varicella, rotavirus, and live-attenuated influenza vaccines), and toxoids (tetanus and diphtheria).

5.(C). T-lymphocyte independent vaccines are associated with poor immune responses in children <2 yr of age, short-term immunity, and absence of an enhanced or booster response on repeat exposure to the antigen. With some polysaccharide vaccines, repeat doses actually are associated with reduced responses, as measured by antibody concentrations, compared to 1st doses (i.e., hyporesponsive). To overcome problems of plain polysaccharide vaccines, polysaccharides have been conjugated, or covalently linked, to protein carriers, converting the vaccine to a T-lymphocyte dependent vaccine. In contrast to plain polysaccharide vaccines, conjugate vaccines induce higher-avidity antibody, immunologic memory leading to booster responses on repeat exposure to the antigen, long-term immunity, and herd protection by decreasing carriage of the organism.

6.(C).

7.(B). Two rotavirus vaccines are available, RotaTeq (RV5) and Rotarix (RV1). With both vaccines, the 1st dose can be administered as early as 6 wk of age and must be administered by 14 wk 6 days. The final dose in the series must be administered no later than 8 mo of age. The RV5 vaccine is administered in 3 doses at least 4 wk apart. The RV1 vaccine is administered in 2 doses at least 4 wk apart. Immunization should not be initiated for infants 15 wk of age and older as stated in the immunization schedule.

8.(A). As a result of effective and safe vaccines, smallpox has been eradicated; polio is close to worldwide eradication.

9.(B). Infants, children, and adolescents in the Iraq routinely are immunized against 11 diseases: BCG, poliomyelitis, hepatitis B, diphtheria, tetanus, pertussis, H. influenzae type b (Hib) disease, rotavirus, measles, mumps and rubella.

10.(A). The most common adverse reaction to immunoglobulin is pain and discomfort at the injection site and, less commonly, flushing, headache, chills, and nausea.

11. **(C).** In hepatitis A prophylaxis, intramuscular IG is recommended.
12. **(D).**
13. **(E).** Toxoids are (tetanus and diphtheria).
14. **(C).**
15. **(B).**
16. **(C).** The minimum interval between 2nd and 3rd dose is 8 weeks and at least 16 weeks after 1st dose. Minimum age for the last dose is 24 weeks.
17. **(B).** Preterm infants generally can be vaccinated at the same chronologic age as full-term infants according to the recommended childhood immunization schedule. An exception is the birth dose of hepatitis B vaccine. Infants weighing ≥ 2 kg and who are stable may receive a birth dose. However, hepatitis B vaccination should be deferred in infants weighing < 2 kg at birth until 30 days of age, if born to a HBs Ag negative mother. All preterm, low birth weight infants born to HBs Ag positive mothers should receive hepatitis B immunoglobulin and hepatitis B vaccine within 12 hr of birth. However, such infants should receive an additional 3 doses of vaccine starting at 30 days of age.
18. **(A).** Oral polio, smallpox, live attenuated influenza, and BCG all these vaccines are contraindicated for this patient.
19. **(C).**
20. **(D).** Waterless hand hygiene products increase hand hygiene compliance and save time; these agents are the preferred agents for routine hand hygiene when hands are not visibly soiled. These products are effective in killing most microbes but do not remove dirt or debris. However, they are ineffective against *C. difficile* spores, requiring the use of other cleansing products during hospital *C. difficile* outbreaks.
21. **(D).**
22. **(D).**
23. **(B).** The drug of choice is azithromycin (10 mg/kg once daily for up to 3 days, with maximum daily dose of 500 mg). Ciprofloxacin (10 mg/kg per dose twice a day for up to 3 days, maximum dose of 500 mg twice a day) is an alternative for children > 1 yr of age. Amoxicillin, trimethoprim-sulfamethoxazole (cotrimoxazole), and erythromycin should not be prescribed for self-treatment of traveler's diarrhea, because of widespread resistance among diarrheal pathogens.
24. **(C).** Fever is defined as a rectal temperature $\geq 38^{\circ}\text{C}$ (100.4°F) and a value $> 40^{\circ}\text{C}$ (104°F) is called hyperpyrexia.
25. **(D).** Endogenous pyrogens include the cytokines interleukins 1 and 6, tumor necrosis factor α , and interferons β and γ . Some substances produced within the body are not pyrogens but are capable of stimulating endogenous pyrogens. Such substances include antigen-antibody complexes in the presence of complement, complement components, lymphocyte products, bile acids, and androgenic steroid metabolites.
26. **(B).** Drugs that are known to cause fever include vancomycin, amphotericin B, and allopurinol.

- 27.(E). Ingestion of dirt is a particularly important clue to infection with *Toxocara canis* (visceral larva migrans) or *Toxoplasma gondii* (toxoplasmosis).
- 28.(A). Palpebral conjunctivitis in a febrile patient may be a clue to measles, coxsackievirus infection, tuberculosis, infectious mononucleosis, lymphogranulomavenereum, or cat-scratch disease. In contrast, bulbar conjunctivitis in a child with FUO suggests Kawasaki disease or leptospirosis.
- 29.(D). Multiple or repeated blood cultures may be required to detect bacteremia associated with infective endocarditis, osteomyelitis, or deep-seated abscesses.
- 30.(B). All other distractors represent primary immune deficiency.
- 31.(D). Intermittent fever is an exaggerated circadian rhythm that includes a period of normal temperatures on most days; extremely wide fluctuations may be termed septic or hectic fever. Sustained fever is persistent and does not vary by more than 0.5°C (0.9°F)/day. Remittent fever is persistent and varies by more than 0.5°C (0.9°F)/day. Relapsing fever is characterized by febrile periods that are separated by intervals of normal temperature.
- 32.(C). Temperatures in excess of 41°C (105.8°F) are most often associated with a noninfectious cause.
- 33.(D).
- 34.(A). Pyelonephritis is the most common serious bacterial infection in this age group and is also more common in uncircumcised infant boys and infants with urinary tract anomalies. *E. coli* is the most common pathogen identified in bacteremic infants, the majority having pyelonephritis.
- 35.(A). Neutropenia is defined as an absolute neutrophil count of <1,000 cells/mm³ and can be associated with significant risk for developing severe bacterial and fungal disease.
- 36.(D). Because the defect involves leukocyte migration and adherence, the neutrophil count in the peripheral blood is usually extremely elevated but pus is not found at the site of infection.
- 37.(B). Although there has been ever-increasing emergence of resistance to penicillins but they remain the drugs of choice for pediatric infections caused by group A and group B *Streptococcus*, *Treponema pallidum* (syphilis), *L. monocytogenes*, and *N. meningitidis*.
- 38.(B). Active against streptococci, *H. influenzae*, *M. catarrhalis*, *Neisseria gonorrhoeae*, *Serratia marcescens*, and *Proteus vulgaris*. No antistaphylococcal or antipseudomonal activity. Dose: 8 mg/kg/24 hr divided q 12-24 hr PO.
- 39.(A). Dose: 25-100 mg/kg/24 hr divided q 6-8 hr PO.
- 40.(B). Carbapenem antibiotic with broad-spectrum activity against Gram-positive cocci and Gram-negative bacilli, including *P. aeruginosa* and anaerobes. No activity against *S. maltophilia*.
- 41.(D). It may increase levels of warfarin, phenytoin, and lithium.
- 42.(C). The first-generation cephalosporins (e.g., cefazolin, a parenteral formulation, and cephalexin, an oral equivalent) are commonly used for management of skin and

soft-tissue infections caused by susceptible strains of *S. aureus* and group A *Streptococcus*. The second-generation cephalosporins (e.g., cefuroxime, cefoxitin) have better activity against Gram-negative bacterial infections than do first-generation cephalosporins and are used to treat respiratory tract infections, urinary tract infections, and skin and soft-tissue infections. A variety of orally administered second-generation agents (cefaclor, cefprozil, loracarbef, cefpodoxime) are commonly used in the outpatient management of sinopulmonary infections and otitis media. The third-generation cephalosporins (cefotaxime, ceftriaxone, and ceftazidime) are typically used for serious pediatric infections, including meningitis and sepsis. A fourth generation cephalosporin, called cefepime, has activity against *P. aeruginosa* and retains good activity against methicillin-susceptible staphylococcal infections. A fifth generation cephalosporin, ceftaroline has been licensed.

43.(C). Ceftriaxone should not be mixed or reconstituted with a calcium-containing product, because particulate formation can result. Cases of fatal reactions with ceftriaxone–calcium precipitates in lungs and kidneys in neonates have been reported.

44.(C). Toxic shock syndrome (TSS) is an acute and potentially severe illness characterized by fever, hypotension, erythematous rash with subsequent desquamation on the hands and feet, and multisystem involvement, including vomiting, diarrhea, myalgias, nonfocal neurologic abnormalities, conjunctival hyperemia, and strawberry tongue.

45.(A).

MAJOR CRITERIA (ALL REQUIRED)

1. Acute fever; temperature $>38.8^{\circ}\text{C}$ (101.8°F)
2. Hypotension
3. Rash (erythroderma with convalescent desquamation)

MINOR CRITERIA (ANY 3 OR MORE)

1. Mucous membrane inflammation (vaginal, oropharyngeal or conjunctival hyperemia, strawberry tongue)
2. Vomiting, diarrhea
3. Liver abnormalities (bilirubin or transaminase greater than twice upper limit of normal)
4. Renal abnormalities (urea nitrogen or creatinine greater than twice upper limit of normal, or greater than 5 white blood cells per high-power field)
5. Muscle abnormalities (myalgia or creatinine phosphokinase greater than twice upper limit of normal)
6. Central nervous system abnormalities (alteration in consciousness without focal neurologic signs)
7. Thrombocytopenia ($100,000/\text{mm}^3$ or less)

46.(E). Many of the clinical features of TSS are usually absent or rare in Kawasaki disease, including diffuse myalgia, vomiting, abdominal pain, diarrhea, azotemia, hypotension, acute respiratory distress syndrome, and shock.

47.(A). Children at increased risk of pneumococcal infections include those with sickle cell disease, asplenia, deficiencies in humoral (B cell) and complement-mediated immunity, HIV infection, certain malignancies (e.g., leukemia, lymphoma), chronic heart, lung, or renal disease (particularly nephrotic syndrome), cerebrospinal fluid leak, and cochlear implants.

48.(C). A pharyngeal strains (e.g., M type 12) are associated with glomerulonephritis; skin strains (e.g., M types 49, 55, 57, and 60) are considered nephritogenic. Several pharyngeal serotypes (e.g., M types 1, 3, 5, 6, 18, 29), but no skin strains, are associated with acute rheumatic fever.

49.(E). It is caused by an infection with pyrogenic exotoxin (erythrogenic toxin)–producing GAS. The rash appears within 24-48 hr after onset of symptoms, although it may appear with the first signs of illness. After 3-4 days, the rash begins to fade and is followed by desquamation. After desquamation, the reddened papillae are prominent, giving the tongue a strawberry appearance. The milder form can be confused with viral exanthems, Kawasaki disease, and drug eruptions.

50.(C). Nonbullous impetigo is the more common form. The lesions may occur anywhere but are most common on the face and extremities. Regional lymphadenitis is common. Nonbullous impetigo is generally not accompanied by fever or other systemic signs or symptoms. Bullous impetigo is less common and occurs most often in neonates and young infants. The usual distribution involves the face, buttocks, trunk, and perineum.

51.(D).

52.(C). Arthritis occurs in approximately 75% of patients with acute rheumatic fever and typically involves larger joints, particularly the knees, ankles, wrists, and elbows. Involvement of the spine, small joints of the hands and feet, or hips is uncommon. Rheumatic arthritis is almost never deforming. There is often an inverse relationship between the severity of arthritis and the severity of cardiac involvement.

53.(A). Acute rheumatic carditis usually presents as tachycardia and cardiac murmurs, with or without evidence of myocardial or pericardial involvement.

54.(B). Patients with carditis and more than minimal cardiomegaly and/or congestive heart failure should receive corticosteroids. The usual dose of prednisone is 2 mg/kg/day in 4 divided doses for 2-3 wk followed by half the dose for 2-3 wk and then tapering of the dose by 5 mg/24 hr every 2-3 days. When prednisone is being tapered, aspirin should be started at 50 mg/kg/day in 4 divided doses for 6 wk to prevent rebound of inflammation.

55.(C). Phenobarbital (16-32 mg every 6-8 hr PO) is the drug of choice. If phenobarbital is ineffective, then haloperidol (0.01-0.03 mg/kg/24 hr divided bid PO) or chlorpromazine (0.5 mg/kg every 4-6 hr PO) should be initiated. Some patients may benefit from a few week courses of corticosteroids.

56.(D).

57.(B).

- Rheumatic fever without carditis : 5 yr or until 21 yr of age, whichever is longer

- Rheumatic fever with carditis but without residual heart disease : 10 yr or until 21 yr of age, whichever is longer
- Rheumatic fever with carditis and residual heart disease: 10 yr or until 40 yr of age, whichever is longer; sometimes lifelong prophylaxis

58.(A). Early-onset neonatal GBS disease presents as sepsis; pneumonia and meningitis. Late-onset neonatal GBS disease most commonly manifests as bacteremia (45-65%) and meningitis (25-35%). Invasive GBS disease in children beyond early infancy is uncommon. Bacteremia without a focus is the most common syndrome associated with childhood GBS disease beyond early infancy.

59.(D).

- bacteremia without a focus 10 days
- meningitis 2-3 wk
- ventriculitis at least 4 wk
- septic arthritis 3-4 wkase
- osteomyelitis 3-4 wkas

60.(A). Specific antitoxin is the mainstay of therapy and should be administered on the basis of clinical diagnosis. The role of antimicrobial therapy is to halt toxin production, treat localized infection, and prevent transmission of the organism to contacts.

61.(B). Three major forms of actinomycosis—cervicofacial, abdominal and pelvic, and pulmonary. In the patient with cervicofacial actinomycosis, there is often a history of oral trauma, oral surgery, dental procedures, or caries, facilitating entry of organisms into cervicofacial tissues. Cervicofacial actinomycosis usually manifests as a painless, slow-growing, hard mass and can produce cutaneous fistulas, a condition commonly known as lumpy jaw. Less frequently, cervicofacial actinomycosis manifests clinically as an acute pyogenic infection with a tender, fluctuant mass with trismus, firm swelling, and fistulas with drainage containing the characteristic sulfur granules.

62.(A). The brain is the most common secondary site and is involved in 15-40% of cases of pulmonary nocardiosis. Brain abscess is the most common presentation, and meningitis is the second most common presentation.

63.(D). Actinomyces, staphylococcus, streptococcus, diphtheria, enterococcus, listeria and nocardia are gram positive.

64.(B).

65.(A). The most common clinical manifestation of meningococcal infection is asymptomatic carriage of the organism in the nasopharynx.

66.(D).

67.(B). The most common complication of acute severe meningococcal septicemia is focal skin infarction, which most commonly affects the lower limbs and can lead to substantial scarring and require skin grafting.

68.(E). Poor prognostic factors on presentation include hypothermia or extreme hyperpyrexia, hypotension or shock, purpura fulminans, seizures, leukopenia, thrombocytopenia (including disseminated intravascular coagulation), acidosis, and high circulating levels of endotoxin and tumor necrosis factor- α . The presence of

petechiae for <12 hr before admission, absence of meningitis, and low or normal erythrocyte sedimentation rate indicate rapid, fulminant progression and poorer prognosis.

69.(A). Ceftriaxone and ciprofloxacin are the most effective agents for prophylaxis, the latter being the drug of choice in some countries. Rifampin is most widely used but fails to eradicate colonization in 15% of cases. Neither penicillin nor ampicillin treatment eradicates nasopharyngeal carriage and should not be routinely used for prophylaxis.

70.(E). Hematogenous dissemination occurs in 1-3% of all gonococcal infections, more frequently after asymptomatic primary infections than symptomatic infections. Women account for the majority of cases, with symptoms beginning 7-30 days after infection and within 7 days of menstruation. The most common manifestations are asymmetric arthralgia, petechial or pustular acral skin lesions, tenosynovitis, suppurative arthritis, and, rarely, carditis, meningitis, and osteomyelitis. Only 25% of patients complain of skin lesions.

71.(E). All patients who are presumed or proven to have gonorrhea should be evaluated for concurrent syphilis, hepatitis B, HIV, and *C. trachomatis* infection.

72.(C). Children who have bacteremia or arthritis should be treated with ceftriaxone (50 mg/kg/day; maximum: 1 g/day if weighs <45 kg) for a minimum of 7 days. Meningitis should be treated for 10-14 days, and endocarditis for a minimum of 28 days, with ceftriaxone (50 mg/kg/ dose q12h with maximum of 1-2 g IV q12h). Neonatal gonococcal ophthalmia is treated effectively with a single dose of ceftriaxone (50 mg/kg IM, not to exceed 125 mg); a single dose of cefotaxime (100 mg/kg IM) is an acceptable alternative. The conjunctivae should be irrigated frequently with physiologic saline solution.

73.(C). Kingellakingae is being increasingly recognized as the most common etiology of joint and bone infections in young children.

74.(A). Chancroid is caused by *Haemophilus ducreyi*, a fastidious Gram negative bacillus.

75.(C). The most important clinical manifestation of *M.catarrhalis* infection in children is otitis media. Colonization and infection with *M. catarrhalis* are increasing in countries in which pneumococcal conjugate vaccines are used widely.

76.(C). Otitis media is the most common reason for which children receive antibiotics. On the basis of culture of middle ear fluid obtained by tympanocentesis, the predominant causes of acute otitis media are *Streptococcus pneumoniae*, *H. influenzae*, and *M. catarrhalis*.

77.(D). Typical paroxysms that are not life threatening have the following features: duration <45 sec; red but not blue color change; tachycardia, bradycardia (not <60 beats/min in infants), or oxygen desaturation that spontaneously resolves at the end of the paroxysm; whooping or strength for brisk self-rescue at the end of the paroxysm; self-expectorated mucus plug; and post-tussive exhaustion but not unresponsiveness.

78.(C). Suctioning of nose, oropharynx, or trachea should not be performed on a "preventive" schedule.

79.(A). Over a 48-hr period disease severity is unchanged or diminished.

80.(A).

- Azithromycin: Recommended agent 10 mg/kg/day in a single dose for 5 days
- Erythromycin: Not preferred
- Clarithromycin: Not recommended
- trimethoprim-sulfamethoxazole: Contraindicated for infants <2 mo of age

81.(D). Progressive pulmonary hypertension in very young infants and secondary bacterial pneumonia are severe complications of pertussis and are the usual causes of death. Echocardiography should be performed in critically ill infants with pertussis to detect presence of pulmonary hypertension and to intervene expeditiously.

82.(E). Diarrhea 36%, abdominal pain 21%, pallor 20%, splenomegaly 17% and headache 4%.

83.(A). The incubation period of typhoid fever is usually 7-14 days but depends on the infecting dose and ranges between 3 and 30 days.

84.(D). Results of blood cultures are positive in 40-60% of the patients seen early in the course of the disease, and stool and urine culture results become positive after the 1st wk. Thrombocytopenia may be a marker of severe illness and may accompany disseminated intravascular coagulopathy. Blood leukocyte counts are frequently low in relation to the fever and toxicity, there is a wide range in counts; in younger children leukocytosis is common and may reach 20,000-25,000 cells/ μ L.

85.(C). Untreated diarrhea can last 7-10 days; only approximately 10% of patients have diarrhea persisting for longer than 10 days. Persistent diarrhea occurs in malnourished infants, children with AIDS, and occasionally previously normal children.

86.(B). Of the more than 200 serogroups, only serogroups O1 and O139 have been associated with epidemics, although some non-O1, non-O139 V.cholerae strains (e.g., O75 and O141) are pathogenic and can cause small outbreaks.

87.(D). Persons with blood group O, decreased gastric acidity, malnutrition, immunocompromised state, and absence of local intestinal immunity (prior exposure by infection or vaccination) are at increased risk for developing severe disease.

88.(D).

89.(A). The most common late-onset complications include reactive arthritis and Guillain-Barré syndrome.

90.(A). Most *Campylobacter* isolates are susceptible to macrolides, fluoroquinolones, aminoglycosides, chloramphenicol, tetracyclines, and clindamycin, and are resistant to cephalosporins, rifampin, penicillins, trimethoprim, and vancomycin.

91.(B). *Yersinia enterocolitica* is by far the most common *Yersinia* species causing human disease and it produces fever, abdominal pain that can mimic appendicitis, and diarrhea. *Yersinia pseudotuberculosis* is most often associated with mesenteric lymphadenitis. *Yersinia pestis* is the agent of plague and most commonly causes an acute febrile lymphadenitis (bubonic plague) and less commonly occurs as septicemic, pneumonic, pharyngeal, or meningeal plague. Other *Yersinia* organisms are uncommon causes of infections of humans.

- 92.(C). Patients with conditions leading to iron overload are at higher risk of developing Yersinia infections.
- 93.(E). Septicemia is more common in younger children, and reactive arthritis is more common in older patients.
- 94.(A). Ulceroglandular and glandular disease are the 2 most common forms of tularemia diagnosed in children. The most common glands involved are the cervical or posterior auricular nodes owing to a tick bite on the head or neck. If an ulcer is present, it is erythematous and painful and may last from 1-3 wk. The ulcer is located at the portal of entry. After the ulcer develops, regional lymphadenopathy ensues.
- 95.(E). Headache, mental inattention, and depression may be demonstrated in patients with brucellosis, invasion of the nervous system occurs in only approximately 1% of cases.
- 96.(D). Aminoglycoside antibiotics should be avoided because they may potentiate the blocking action of botulinum toxin at the neuromuscular junction.
- 97.(E). Tetanus is most often generalized but may also be localized. The incubation period typically is 2-14 days but may be as long as months after the injury. Because tetanus toxin does not affect sensory nerves or cortical function, the patient unfortunately remains conscious, in extreme pain, and in fearful anticipation of the next tetanic seizure. The tetanic paralysis usually becomes more severe in the 1st wk after onset, stabilizes in the 2nd wk, and ameliorates gradually over the ensuing 1-4 wk.
- 98.(A). Major adverse events include hepatotoxicity in 1% of children and approximately 3% of adults (increasing with age) and dose-related peripheral neuropathy. Minor adverse events include rash, worsening of acne, epigastric pain with occasional nausea and vomiting, decreased vitamin D levels, and dizziness.
- 99.(B). Aluminum salts decrease absorption of isoniazid, rifampin increase hepatotoxicity of isoniazid, isoniazid increase level of warfarin and prednisolone increased isoniazid metabolism.
- 100.(B). Concentrations of fluconazole are 10-20– fold higher in the urine than blood, making it an ideal agent for treating fungal urinary tract infections.
- 101.(E). The cumulative incidence is <0.3% among infants >2,500 g birthweight admitted to the NICU. The cumulative incidence increases to 8% for infants <750 g birthweight.
- 102.(B). Twice weekly fluconazole at 3 and 6 mg/kg/dose decreases rates of both colonization with Candidaspecies and invasive fungal infections.
- 103.(B). While the others inhibits viral DNA polymerase.
- 104.(B). The use of suppressive therapy with oral acyclovir for 6 mo has been demonstrated to improve neurodevelopmental outcome.
- 105.(D). The main route of elimination is renal, and dosage adjustments are necessary for renal insufficiency. Toxicity is observed typically only in exceptional circumstances: for example, if administered by rapid infusion to a dehydrated patient or a patient with underlying renal insufficiency, acyclovir can crystallize in renal tubules and produce a reversible obstructive uropathy.

106.(D). Ribavirin and its metabolites concentrate in red blood cells and can persist for several weeks and, in rare instances, may be associated with anemia.

107.(C).

108.(D). The rash fades over about 7 days in the same progression as it evolved, often leaving a fine desquamation of skin in its wake.

109.(C). Acute otitis media is the most common complication of measles and pneumonia is the most common cause of death in measles.

110.(E). Males are affected twice as often as females, and there appear to be more cases reported from rural than urban populations. Clinical manifestations of SSPE begin insidiously 7-13 yr after primary measles infection. The hallmark of the 2nd stage is massive myoclonus, which coincides with extension of the inflammatory process site to deeper structures in the brain, including the basal ganglia. The 4th stage is characterized by loss of critical centers that support breathing, heart rate, and blood pressure. Death soon ensues.

111.(C). Nerve deafness is the single most common finding among infants with congenital rubella syndrome.

112.(E). The incubation period for mumps ranges from 12-25 days but is usually 16-18 days. Parotitis may be unilateral initially but becomes bilateral in approximately 70% of cases. The opening of the Stensen duct may be red and edematous. The parotid swelling peaks in approximately 3 days and then gradually subsides over 7 days.

113.(A). The most common complications of mumps are meningitis, with or without encephalitis, and gonadal involvement. Uncommon complications include conjunctivitis, optic neuritis, pneumonia, nephritis, pancreatitis, and thrombocytopenia.

114.(E). No fetal malformations have been associated with intrauterine mumps infection.

115.(E). Symptomatic CNS involvement occurs in 10-30% of infected individuals, but CSF pleocytosis has been found in 40-60% of patients with mumps parotitis. The meningoencephalitis may occur before, along with, or following the parotitis. It most commonly manifests 5 days after the parotitis. In typical cases, symptoms resolve in 7-10 days. The CSF protein content is normal or mildly elevated. Less-common CNS complications of mumps include transverse myelitis, aqueductal stenosis, and facial palsy.

116.(A). Hematogenous dissemination of virus to the central nervous system appears to only occur in neonates.

117.(C). Herpes whitlow is a term generally applied to HSV infection of fingers or toes, although strictly speaking it refers to HSV infection of the paronychia.

118.(D). HSV is the most common cause of recurrent aseptic meningitis (Mollaret meningitis).

119.(C). Infants with encephalitis typically present at 8-17 days of life with clinical findings suggestive of bacterial meningitis, including irritability, lethargy, poor feeding,

poor tone, and seizures. Fever is relatively uncommon, and skin vesicles occur in only approximately 60% of cases.

120.(A). The absolute neutrophil count should be measured at weeks 2 and 4 after initiation treatment and then monthly.

121. (C). Herpes zoster is caused by the reactivation of latent VZV. It is not common in childhood and shows no seasonal variation in incidence.

122.(C). The illness usually begins 14-16 days after exposure, although the incubation period can range from 10-21 days. Subclinical varicella is rare. Temperature elevation is usually 37.8-38.9°C but may be as high as 41.1°C. Varicella lesions often appear first on the scalp, face, or trunk.

123.(A). The simultaneous presence of lesions in various stages of evolution is characteristic of varicella.

124.(E). Clinical recovery is typically rapid, occurring within 24-72 hr, and is usually complete.

125.(E).

126.(E). EBV is shed in oral secretions consistently for more than 6 mo after acute infection and then intermittently for life.

127.(E). EBV infects more than 95% of the world's population.

128.(B). The classic physical examination findings are generalized lymphadenopathy (90% of cases), splenomegaly (50% of cases), and hepatomegaly (10% of cases). Epitrochlear lymphadenopathy is particularly suggestive of infectious mononucleosis. Symptomatic hepatitis or jaundice is uncommon, but elevated liver enzymes are very common. Splenomegaly to 2-3 cm below the costal margin is typical (15-65% of cases) and is seen in most cases by ultrasonography; massive enlargement is uncommon. Palatal petechiae at the junction of the hard and soft palate are frequently seen.

129.(B). Hearing loss is the most common long-term sequela associated with congenital CMV infection, the failure of an infant to pass a newborn hearing screening exam should raise the possibility of congenital CMV infection. Hearing loss in the older infant and young child should also alert the clinician to the possibility of congenital CMV infection, as approximately 50% of infants with hearing loss associated with congenital CMV infection will pass an initial hearing screening exam but develop hearing loss in later infancy and early childhood.

130.(D). Congenital infection with HHV-7 has not been demonstrated.

131.(D).

132.(A).

133.(D). Vaccines are available for HAdV types 4 and 7, but are used only for military populations.

134.(E). The virus can be identified in 15-20% of children with isolated pharyngitis, mostly in preschool children and infants.

135.(C). Vomiting and fever typically abate during the 2nd day of illness, but diarrhea often continues for 5-7 days.

136.(A). Common adverse effects include abdominal pain, diarrhea, and nausea. Rare side effects include anorexia, flatulence, increased appetite, fever, pruritus, and dizziness.

137.(A). It is FDA approved for treatment of trichomoniasis and for giardiasis and amebiasis in children 3 yr of age and older. In the treatment of giardiasis, it has the advantages of very few side effects and only requiring a single dose. It is excreted via urine and feces. Tinidazole carries a pregnancy category C classification and can be detected in breast milk. Breastfeeding should be interrupted during treatment and for 3 days after treatment.

138.(D). Naegleria, Acanthamoeba, Balamuthia, and Sappinia are small, free living amebas that cause human amebic meningoencephalitis.

139.(D). The life cycle of *G. lamblia* (also known as *Giardia intestinalis* or *Giardia duodenalis*) is composed of 2 stages: trophozoites and cysts. Each ingested cyst produces 2 trophozoites in the duodenum. *Giardia* trophozoites contain 2 oval nuclei anteriorly, a large ventral disk, a curved median body posteriorly, and 4 pairs of flagella. Studies suggest that different *Giardia* genotypes may cause unique clinical manifestations, but these findings appear to vary according to the geographic region tested.

140.(C).

141.(A). Giardiasis should be considered in children who have acute nondysenteric diarrhea, persistent diarrhea, intermittent diarrhea and constipation, malabsorption, chronic crampy abdominal pain and bloating, failure to thrive, or weight loss.

142.(D). Asymptomatic excretors generally are not treated except in specific instances such as outbreak control, prevention of household transmission by toddlers to pregnant women and patients with hypogammaglobulinemia or cystic fibrosis, and situations requiring oral antibiotic treatment where *Giardia* may produce malabsorption of the antibiotic. Tinidazole not recommended below 3 years old as albendazole not recommended below age of 6 years. Nitazoxanide 1-3 yr: 100 mg bid for 3 days

143.(C).

144.(E). *Cryptosporidium* is recognized as a leading protozoal cause of diarrhea in children worldwide and is a common cause of outbreaks in childcare centers; it is also a significant pathogen in immunocompromised patients.

145.(C). The late stage of the illness is often complicated by secondary bacterial infections, which frequently are a cause of death.

146.(B). Hand-foot-and-mouth disease, one of the more distinctive rash syndromes, is most frequently caused by coxsackievirus A16, sometimes in large outbreaks, and can also be caused by enterovirus 71; coxsackie A viruses 5, 6, 7, 9, and 10; coxsackie B viruses 2 and 5; and some echoviruses.

1. All the following are causes oropharyngeal dysphagia **EXCEPT**
 - A. cerebral palsy
 - B. esophagitis
 - C. brain tumors
 - D. cerebrovascular accidents
 - E. hyperthyroidism

2. All the following are common causes of emesis in childhood **EXCEPT**
 - A. gastroenteritis
 - B. toxic ingestion
 - C. pertussis syndrome
 - D. peptic ulcer
 - E. otitis media

3. Secretory diarrhea can be caused by
 - A. neuroblastoma
 - B. laxative abuse
 - C. lactase deficiency
 - D. irritable bowel syndrome
 - E. thyrotoxicosis

4. All the following metabolic disorders can cause constipation **EXCEPT**
 - A. hypercalcemia
 - B. hyperkalemia
 - C. hypothyroidism
 - D. diabetes mellitus
 - E. diabetes insipidus

5. All the following suggest a potentially serious organic etiology of abdominal pain **EXCEPT**
 - A. age >5 yr
 - B. fever
 - C. flank pain
 - D. awakening from sleep
 - E. referred pain to shoulder

6. All the following are common causes of gastrointestinal bleeding in infancy **EXCEPT**
- A. bacterial enteritis
 - B. intussusception
 - C. Meckel diverticulum
 - D. anal fissure
 - E. lymphonodular hyperplasia
7. Delayed eruption of the primary teeth can be due to the following **EXCEPT**
- A. familial
 - B. hypopituitarism
 - C. hyperthyroidism
 - D. cleidocranial dysplasia
 - E. trisomy 21
8. All the following are features of Stickler syndrome **EXCEPT**
- A. prominent joints
 - B. autosomal recessive inheritance
 - C. hypotonia
 - D. mitral valve prolapse
 - E. retinal detachment
9. Periodontitis is often associated with the following conditions **EXCEPT**
- A. leukocyte adhesion defects
 - B. hypophosphatasia
 - C. leukemia
 - D. vitamin D resistant rickets
 - E. histiocytosis X
10. Aphthous-like lesions may be associated with the following conditions **EXCEPT**
- A. inflammatory bowel disease
 - B. Behcet disease
 - C. gluten-sensitive enteropathy
 - D. SWEET syndrome
 - E. herpetic gingivostomatitis
11. Eruption cyst over the erupting tooth is characterized by the following **EXCEPT**
- A. smooth
 - B. painful
 - C. blue or blue-black
 - D. no treatment is indicated
 - E. resolves after eruption of the tooth

12. All the following criteria must be met for the consensus definition of cyclical vomiting syndrome **EXCEPT**
- A. at least 5 attacks in any interval
 - B. recurrent episodes of intense vomiting and nausea lasting 1 hr to 10 days and occurring at least 1 wk apart
 - C. vomiting during episodes occurs ≥ 4 times/hr for ≥ 1 hr
 - D. return to baseline health between episodes
 - E. usually attributed to another disorder
13. One of the following cause osmotic diarrhea
- A. toxigenic Escherichiacoli
 - B. carcinoid
 - C. neuroblastoma
 - D. glucose-galactose malabsorption
 - E. congenital chloride diarrhea
14. Acute diarrhea in infancy is commonly caused by
- A. primary disaccharidase deficiency
 - B. overfeeding
 - C. Hirschsprung toxic colitis
 - D. adrenogenital syndrome
 - E. neonatal opiate withdrawal
15. Advanced dentition for age and sex is seen in
- A. nutritional disturbances
 - B. hypopituitarism
 - C. cleidocranial dysplasia
 - D. hyperthyroidism
 - E. trisomy 21
16. Mottled enamel (permanent teeth) may be seen in
- A. renal failure
 - B. immunosuppression
 - C. neutrophil chemotactic deficiency
 - D. maternal infections
 - E. uncontrolled juvenile diabetes
17. Pierre Robin syndrome consists of the following **EXCEPT**
- A. micrognathia
 - B. high arched palate
 - C. cleft palate
 - D. small tongue

E. foreshortened floor of the mouth

18. The following are features of achalasia **EXCEPT**

- A. dysphagia for solids and liquids
- B. may be accompanied by undernutrition
- C. may be accompanied by chronic cough
- D. may be misdiagnosed as asthma
- E. usually diagnosed before school age

19. Contrast (usually barium) radiographic study of the esophagus and upper gastrointestinal tract has poor sensitivity and specificity in the diagnosis of

- A. achalasia
- B. esophageal strictures
- C. GERD
- D. hiatal hernia
- E. gastric outlet obstruction

20. Gastroesophageal reflux disease (GERD) is the most common esophageal disorder in children of all ages.

Of the following, the **MOST** common symptom in infants is

- A. excessive crying
- B. regurgitation
- C. failure to thrive
- D. abnormal posturing
- E. sleeping disturbances

21. The following procedures satisfactorily improve gastroesophageal reflux disease (GERD) symptoms in infants **EXCEPT**

- A. modified feeding volumes
- B. hydrolyzed infant formulas
- C. prone position, when the infant is awake and observed
- D. avoidance of smoke exposure
- E. seated position

22. Erosive esophagitis is found in approximately 12% of children with gastroesophageal reflux disease (GERD) symptoms and is more common in the following **EXCEPT**

- A. girls
- B. older children
- C. neurologically abnormal children
- D. children with severe chronic respiratory disease
- E. children with hiatal hernia

23. Hypertrophic pyloric stenosis has been associated with the following **EXCEPT**

- A. eosinophilic gastroenteritis
- B. Apert syndrome
- C. Zellweger syndrome
- D. trisomy 21
- E. Cornelia de Lange syndrome

24. Oral and intravenous atropine sulfate (pyloric muscle relaxant) has been described when surgical treatment is not available for hypertrophic pyloric stenosis with a success rate of

- A. 20%
- B. 40%
- C. 60%
- D. 80%
- E. 90%

25. Meckel diverticulum has been conveniently referred to by the “rule of 2s,” which explains the classic presentation of this congenital anomaly. One of the following is **FALSE** in this rule

- A. found in approximately 2% of the general population
- B. usually located 2 feet proximal to the ileocecal valve
- C. approximately 2 inches in length
- D. contain 2 types of ectopic tissue (pancreatic or gastric)
- E. found twice as commonly in males

26. Confirmation of a Meckel diverticulum can be difficult.

Of the following, the **MOST** sensitive study is

- A. plain abdominal radiographs
- B. barium studies
- C. radionuclide technetium-99m scans
- D. abdominal ultrasound
- E. abdominal CT scan

27. Hirschsprung disease has been seen in association with the following **EXCEPT**

- A. microcephaly
- B. mental retardation
- C. autism
- D. cleft lip
- E. hydrocephalus

28. In healthy full-term infants, meconium is passed within 48 hr of birth in

- A. 99%

- B. 90%
- C. 80%
- D. 70%
- E. 60%

29. Ileus is the failure of intestinal peristalsis caused by loss of coordinated gut motility without evidence of mechanical obstruction. In children, ileus accompanies the following metabolic abnormalities **EXCEPT**

- A. uremia
- B. hypokalemia
- C. hypocalcemia
- D. hypermagnesemia
- E. acidosis

30. The classic triad of intussusception, (pain, a palpable sausage-shaped abdominal mass, and bloody or currant jelly stool) is seen in

- A. $\leq 20\%$ of patients
- B. $\leq 30\%$ of patients
- C. $\leq 40\%$ of patients
- D. $\leq 50\%$ of patients
- E. $\leq 60\%$ of patients

31. Regarding foreign bodies in the stomach and intestine, all the following are true **EXCEPT**

- A. coins are the most commonly ingested foreign body
- B. perforation is estimated to be $<1\%$ of all objects ingested.
- C. most objects pass through the intestine in 4-6 days
- D. cathartics should be avoided
- E. ingestion of batteries usually leads to problems

32. A bezoar is an accumulation of exogenous matter in the stomach or intestine. Regarding bezoar, all the following are true **EXCEPT**

- A. trichobezoars are composed of the patient's own hair
- B. lactobezoars can be attributed to the high casein or calcium content of some premature formulas.
- C. phytobezoars are composed of a combination of plant and animal material
- D. lactobezoars usually resolve when feedings are withheld for 24-48 hr
- E. sunflower seed bezoars are reported to cause small bowel obstruction

33. The usual presenting symptoms of peptic ulcer disease in infants and younger children are the following **EXCEPT**

- A. feeding difficulty

- B. vomiting
- C. epigastric pain
- D. crying episodes
- E. hematemesis

34. *H. pylori* are among the most common bacterial infections in humans.

All the following are true **EXCEPT**

- A. *H. pylori* is a Gram-negative, S-shaped rod
- B. *H. pylori* infection can manifest with abdominal pain or vomiting
- C. *H. pylori* is classified by the World Health Organization as a group I carcinogen
- D. If *H. pylori* is identified in a child with no symptoms, no therapy should be offered
- E. *H. pylori* can be associated, though rarely, with chronic autoimmune thrombocytopenia

35. One of the following features is **MORE** common in ulcerative colitis than in Crohn disease

- A. abdominal pain
- B. growth failure
- C. rectal bleeding
- D. mouth ulceration
- E. strictures

36. A neonate is presented with watery and voluminous stool which is mistaken for urine.

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

- A. congenital glucose-galactose malabsorption
- B. microvillus inclusion disease
- C. congenital bile acid malabsorption
- D. congenital enterokinase deficiency
- E. congenital trypsinogen deficiency

37. The **MOST** common extraintestinal manifestation of celiac disease is

- A. osteoporosis
- B. short stature
- C. arthritis
- D. iron-deficiency anemia
- E. aphthous stomatitis

38. The following neurologic manifestations are seen in celiac disease **EXCEPT**

- A. myopathy
- B. epilepsy

- C. irritability
- D. cerebral calcifications
- E. cerebellar ataxia

39. The following conditions can be associated with celiac disease **EXCEPT**

- A. Williams syndrome
- B. Down syndrome
- C. Turner syndrome
- D. Klinefilter syndrome
- E. Sjogren syndrome

40. Abetalipoproteinemia is a rare autosomal recessive disorder of lipoprotein metabolism (Bassen-Kornzweig syndrome). It is characterized by the following **EXCEPT**

- A. presence of acanthocytes in the peripheral blood smear
- B. extremely low plasma levels of cholesterol
- C. very low triglycerides level
- D. absent deep tendon reflexes
- E. normal intellectual development

41. The following infections may cause malabsorption in immunocompromised children **EXCEPT**

- A. Shigella
- B. Salmonella
- C. enteropathogenic E. coli
- D. Giardia
- E. rotavirus

42. In infants and very young toddlers chronic diarrhea can appear following infectious enteritis. The pathogenesis of the diarrhea is not always clear and may be related to

- A. food protein allergy
- B. bacterial overgrowth
- C. giardiasis
- D. Strongyloides stercoralis
- E. eosinophilic gastroenteropathy

43. Vitamin B12 and bile salts are only absorbed in

- A. distal ileum
- B. duodenum
- C. proximal jejunum
- D. colon
- E. proximal ileum

44. Management of the diarrhea in chronically malnourished children is based on
- A. intravenous therapy
 - B. standard osmolarity oral rehydration solutions
 - C. reduced osmolarity oral rehydration solutions
 - D. slow resumption of feeds
 - E. nothing by mouth for 24 hour
45. The **MOST** common congenital disorder associated with exocrine pancreatic insufficiency is
- A. Shwachman- Diamond syndrome
 - B. Johanson-Blizzard syndrome
 - C. Pearson bone marrow syndrome
 - D. isolated pancreatic enzyme deficiency
 - E. cystic fibrosis
46. In the blue diaper syndrome, symptoms can include all the following **EXCEPT**
- A. vomiting
 - B. diarrhea
 - C. failure to thrive
 - D. nephrocalcinosis
 - E. ocular abnormalities
47. Features of Tangier disease include the following **EXCEPT**
- A. orange tonsils
 - B. hepatosplenomegaly
 - C. relapsing neuropathy
 - D. orange-brown spots on the colon and ileum
 - E. increased plasma cholesterol
48. Newborns with congenital chloride diarrhea present with severe life-threatening secretory diarrhea during the 1st few wk of life.
Of the following, the **MOST** common laboratory finding is
- A. metabolic alkalosis
 - B. hyperchloremia
 - C. hyperkalemia
 - D. hyponatremia
 - E. hypercalcemia
49. Acrodermatitis enteropathica requires long term treatment with elemental zinc
- A. 0.5 mg/kg/day
 - B. 1 mg/kg/day
 - C. 1.5mg/kg/day

- D. 2 mg/kg/day
 - E. 2.5 mg/kg/day
50. Direct person-to-person contact outbreaks of gastroenteritis are usually caused by
- A. Shigella
 - B. Salmonella
 - C. Rotavirus
 - D. Giardia
 - E. Clostridium difficile
51. Guillain-Barré syndrome can be a sequel in the following foodborne bacterial illness
- A. Bacillus anthracis
 - B. Bacillus cereus
 - C. Brucella abortus
 - D. Campylobacter jejuni
 - E. Clostridium botulinum
52. Antibiotics are not indicated and may be detrimental in the following foodborne bacterial illness
- A. Enterotoxigenic E. coli
 - B. Enterohemorrhagic Escherichia coli O157:H7
 - C. Campylobacter jejuni
 - D. Brucella abortus
 - E. Bacillus anthracis
53. The first-line treatment for children with Vibrio cholera is
- A. azithromycin
 - B. ciprofloxacin
 - C. doxycycline
 - D. ampicillin
 - E. TMP-SMX
54. Appendicitis-like symptoms (diarrhea, vomiting, fever, and abdominal pain) may be seen in the following foodborne bacterial illness
- A. Vibrio vulnificus
 - B. Yersinia enterocolitica
 - C. Shigella spp.
 - D. Staphylococcus aureus (preformed enterotoxin)
 - E. Listeria monocytogenes
55. Risks associated with severe dehydration that might necessitate intravenous resuscitation include

- A. age < 9 mo
- B. postmaturity
- C. fever >38°C if 3-36 mo of age
- D. secretory diarrhea
- E. depressed level of consciousness

56. The low-osmolality World Health Organization (WHO) oral rehydration solution (ORS) has the following per liter **EXCEPT**

- A. 75 mEq of sodium
- B. 50 mEq of chloride
- C. 20 mEq of potassium
- D. 75 mmol of glucose
- E. total osmolality of 245 mOsm

57. Immunoglobulin A (IgA) nephropathy is an extraintestinal manifestation of the following enteric infection

- A. Salmonella
- B. Shigella
- C. Yersinia
- D. Campylobacter
- E. Cryptosporidium

58. Continued enteral feeding in diarrhea aids in recovery from the episode, but the following should be avoided

- A. rice
- B. potatoes
- C. lean meats
- D. fruits
- E. juices

59. Zinc supplementation in children with diarrhea in developing countries leads to the following **EXCEPT**

- A. decreased use of ORS
- B. reduced duration of diarrhea
- C. reduced severity of diarrhea
- D. reduced hospital admission
- E. improving diarrhea recovery rates

60. Ondansetron is an effective and less-toxic antiemetic agent and can be given as a useful adjunct to the treatment of vomiting in ambulatory settings, it is usually given

- A. intravenously
- B. intramuscularly

- C. subcutaneously
- D. sublingually
- E. orally

61. Nitazoxanide is an anti-infective agent effective in the treatment of a wide variety of pathogens including the following **EXCEPT**

- A. *Giardia lamblia*
- B. *E. histolytica*
- C. *Shigella*
- D. *C. difficile*
- E. Rotavirus

62. Ciprofloxacin is the drug of choice for the treatment of

- A. *Salmonella*
- B. *Shigella*
- C. *Campylobacter jejuni*
- D. *Clostridium difficile*
- E. *Cyclospora* spp.

63. Increased fecal calprotectin concentration is seen in

- A. fat malabsorption
- B. carbohydrate malabsorption
- C. intestinal inflammation
- D. pancreatic function
- E. reduced intestinal surface

64. The following are alarm symptoms of functional abdominal pain **EXCEPT**

- A. persistent left lower quadrant pain
- B. unexplained fever
- C. genitourinary tract symptoms
- D. dysphagia
- E. nocturnal diarrhea

65. A variety of laboratory tests have been used in the evaluation of children with suspected appendicitis.

Of the following, the **MOST** sensitive and specific one is

- A. leukocyte count
- B. urinalysis
- C. electrolytes
- D. C-reactive protein
- E. serum amyloid A protein

66. Plain abdominal radiographs may be helpful in selected cases of abdominal pain/suspected appendicitis. Plain abdominal x-rays can demonstrate the following findings in acute appendicitis **EXCEPT**

- A. sentinel loops of bowel and localized ileus
- B. scoliosis from psoas muscle spasm
- C. a colonic air–fluid level above the right iliac fossa
- D. a RLQ soft-tissue mass
- E. a calcified appendicolith (50% of cases)

67. Findings that suggest advanced appendicitis on ultrasound include the following **EXCEPT**

- A. asymmetric wall thickening
- B. increased local tenderness to compression
- C. abscess formation
- D. associated free intraperitoneal fluid
- E. surrounding tissue edema

68. The clinical presentation of one of the following closely mimics appendicitis

- A. Meckel diverticulitis
- B. mesenteric adenitis
- C. sickle cell disease
- D. pancreatitis
- E. ovarian torsion

69. There are many anomalies associated with anorectal malformations.

Of the following, the **MOST** common are

- A. kidney anomalies
- B. cardiac anomalies
- C. esophageal atresia
- D. spina bifida
- E. tethered cord

70. The risk of colon cancer is 100% in the following condition

- A. Peutz-Jeghers syndrome
- B. Cowden syndrome
- C. Gardner syndrome
- D. Juvenile polyposis
- E. Bannayan-Riley- Ruvalcaba syndrome

71. Hamartomatous polyps, which represent the most common intestinal tumors of childhood have the following features **EXCEPT**

- A. occurring in 1-2% of children

- B. present in the 1st decade
- C. most often at ages 2-5 yr
- D. most commonly in the colon
- E. often multiple

72. Inguinal hernias are one of the most common conditions seen in pediatric practice and the most common surgical procedure performed in pediatric surgical practice. Regarding the incidence of congenital indirect inguinal hernia, all the following are true

EXCEPT

- A. approaches 30% in very-low birth weight infants
- B. more common in boys
- C. approximately 60% of inguinal hernias occur on the right side
- D. bilateral hernias is higher in boys
- E. an increased incidence in twins

73. The features of the Johanson-Blizzard syndrome include the following **EXCEPT**

- A. exocrine pancreatic deficiency
- B. hypoplasia of the alae nasi
- C. congenital deafness
- D. hyperthyroidism
- E. ectodermal scalp defects

74. The test of choice for acute pancreatitis is

- A. serum amylase
- B. serum lipase
- C. hyperglycemia
- D. hypocalcemia
- E. elevated γ -glutamyl transpeptidase

75. The first choice for diagnosis of pancreatic pseudocysts is

- A. transabdominal ultrasonography
- B. CT scanning
- C. magnetic resonance cholangiopancreatography
- D. endoscopic retrograde cholangiopancreatography
- E. endoscopic ultrasound

76. Stippled calcifications of the patellas and greater trochanter are found in the following intrahepatic cholestasis

- A. idiopathic neonatal hepatitis
- B. Aagaard's syndrome
- C. Zellweger (cerebrohepatorenal) syndrome
- D. neonatal iron storage disease

- E. Byler disease
77. One of the following rules out biliary atresia
- A. history of prematurity
 - B. normal size of liver
 - C. normal consistency of liver
 - D. consistently pigmented stools
 - E. no familial incidence
78. The success rate for establishing good bile flow after the Kasai operation is much higher (90%) if performed before
- A. 4 wk of life
 - B. 6 wk of life
 - C. 8 wk of life
 - D. 10 wk of life
 - E. 12 wk of life
79. For patients with advanced liver disease, hepatic transplantation has a success rate >85%. If the operation is technically feasible, it will prolong life and might correct the metabolic error in diseases such as
- A. tyrosinemia
 - B. galactosemia
 - C. hereditary fructose intolerance
 - D. Zellweger syndrome
 - E. Crigler-Najjar
80. Persistence of unconjugated hyperbilirubinemia at levels >20 mg/dL after the 1st wk of life in the absence of hemolysis should suggest
- A. galactosemia
 - B. tyrosinemia
 - C. α 1-Antitrypsin deficiency
 - D. Crigler-Najjar type I
 - E. Maple serum urine disease
81. Liver histology demonstrates normal architecture, but hepatocytes contain black pigment similar to melanin in
- A. Dubin-Johnson syndrome
 - B. Rotor syndrome
 - C. Gilbert syndrome
 - D. Crigler-Najjar syndrome
 - E. Zellweger syndrome

82. Kayser-Fleischer (K-F) ring is a brown discoloration at the outer margin of the cornea; the following are true **EXCEPT**

- A. might not be present in younger children
- B. requires a slit-lamp examination
- C. is permanent even after treatment
- D. is present in 95% of patients with neurologic symptoms
- E. is deposition of copper in Descemet's membrane

83. A major attempt in Wilson disease should be made to restrict dietary copper intake to <1 mg/day.

Of the following, the food that should be avoided is

- A. liver
- B. meat
- C. egg
- D. chicken
- E. fish

84. Neonatal iron storage disease (NISD), also known as neonatal hemochromatosis is a rapidly fatal, progressive illness characterized by the following **EXCEPT**

- A. hypoprothrombinemia
- B. hypoalbuminemia
- C. hyperferritinemia
- D. hyperbilirubinemia
- E. hyperglycemia

85. No parenteral transmission occur in the following hepatotropic viruses

- A. HAV
- B. HBV
- C. HCV
- D. HDV
- E. HEV

86. HBV is present in high concentrations in

- A. serous exudates
- B. saliva
- C. vaginal fluid
- D. semen
- E. urine

87. Risk factors for HBV infection in children and adolescents include acquisition by the following **EXCEPT**

- A. intravenous drug

- B. contaminated needles
 - C. sexual contact
 - D. intimate contact with carriers
 - E. sharing toys
88. Regarding epidemiology of HBV, the following are true **EXCEPT**
- A. intrauterine infection occurs in 2.5%
 - B. antigenemia appear 1-3 mo after birth
 - C. HBsAg is inconsistently recovered in human milk of infected mothers
 - D. breastfeeding of nonimmunized infants by infected mothers confer a greater risk of hepatitis
 - E. HBV has 8 genotypes (A-H)
89. Routine screening for HBV infection requires assay of multiple serologic markers, all the following are true **EXCEPT**
- A. HBsAg is the first serologic marker of infection to appear
 - B. Anti-HBc is a valuable serologic marker of acute HBV infection
 - C. anti-HBs and anti- HBc are detected in persons immunized with hepatitis B vaccine
 - D. HBeAg is a marker of infectivity
 - E. HBsAg levels fall before symptoms wane
90. HBV is usually spread by
- A. breastfeeding
 - B. kissing
 - C. hugging
 - D. sharing utensils
 - E. sharing needles
91. The **MOST** common hepatobiliary disease associated with inflammatory bowel disease (IBD) is
- A. hepatic abscess
 - B. portal vein thrombosis
 - C. sclerosing cholangitis
 - D. autoimmune hepatitis
 - E. biliary carcinoma
92. Total parenteral nutrition (TPN) can cause a variety of liver diseases. Of the following, the **MOST** severe complication is
- A. hepatic steatosis
 - B. gallbladder damage
 - C. bile duct damage

- D. cholestasis
- E. sclerosing cholangitis

93. Autoimmune hepatitis is a clinical diagnosis based on certain diagnostic criteria; no single test will make this diagnosis. Important positive features include the following **EXCEPT**

- A. female gender
- B. primary elevation in alkaline phosphatase
- C. elevated γ -globulin levels
- D. presence of autoantibodies
- E. characteristic histologic findings

94. The gallbladder is congenitally absent in approximately 0.1% of the population. Hypoplasia or absence of the gallbladder can be associated with

- A. biliary dyskinesia
- B. cystic fibrosis
- C. cirrhosis
- D. cholestasis
- E. Wilson disease

95. Biliary sludge or cholelithiasis can be detected in >40% of children who are treated with prolonged course of

- A. cefotaxime
- B. meropenem
- C. vancomycin
- D. erythromycin
- E. ceftriaxone

96. The **MOST** common presentation of portal hypertension is

- A. jaundice
- B. splenomegaly
- C. esophageal hemorrhage
- D. ascites
- E. dilated cutaneous collateral vessels in the periumbilical region

97. The **MOST** common indication for liver transplantation in children is

- A. Alagille syndrome
- B. congenital hepatic fibrosis
- C. hepatocellular carcinoma
- D. biliary atresia
- E. Wilson disease

98. Ascites is the pathologic accumulation of fluid within the peritoneal cavity.

Of the following, the **MOST** common cause in children is

- A. neoplasia
- B. hepatic
- C. cardiac
- D. trauma
- E. infection

99. Primary peritonitis usually refers to bacterial infection of the peritoneal cavity without a demonstrable intraabdominal source.

Of the following, the **MOST** common isolated bacteria are

- A. group A streptococci
- B. Staphylococci
- C. Escherichia coli
- D. Mycobacterium tuberculosis
- E. Pneumococci

100. Acute secondary peritonitis most often results from entry of enteric bacteria into the peritoneal cavity through a necrotic defect in the wall of the intestines or other viscus.

It **MOST** commonly follows

- A. perforation of the appendix
- B. rupture of a Meckel diverticulum
- C. intussusception
- D. peptic ulceration
- E. necrotizing enterocolitis

1. **(B).** Esophagitis is a cause of esophageal dysphagia.
2. **(D).** Peptic ulcer is a rare cause of emesis in childhood.
3. **(A).** Laxative abuse and lactase deficiency are causes of osmotic diarrhea while irritable bowel syndrome and thyrotoxicosis are caused by increased bowel motility.
4. **(B).** Hypokalemia causes constipation.
5. **(A).** Age less than 5 year may suggest serious organic etiology.
6. **(C).** Meckel diverticulum is a rare cause.
7. **(C).** Hypothyroidism is a cause of delayed eruption.
8. **(B).** Stickler syndrome, an autosomal dominant condition that includes other findings such as prominent joints, arthritis, hypotonia, hyperextensible joints, mitral valve prolapse, and ocular problems (retinal detachment, myopia, cataracts).
9. **(D).** Periodontitis in children before puberty is a rare disease that often begins between the time of eruption of the primary teeth and the age of 4 or 5 yr. The disease occurs in localized and generalized forms. There is rapid bone loss, often leading to premature loss of primary teeth. It is often associated with systemic problems, including neutropenia, leukocyte adhesion or migration defects, hypophosphatasia, Papillon-Lefvre syndrome, leukemia, and histiocytosis X.
10. **(E).** The aphthous ulcer (canker sore) is a distinct oral lesion, prone to recurrence. Aphthous ulcers are reported to develop in 20% of the population. Their etiology is unclear, but allergic or immunologic reactions, emotional stress, genetics, and injury to the soft tissues in the mouth have been implicated. Aphthous-like lesions may be associated with inflammatory bowel disease, Behcet disease, gluten-sensitive enteropathy, periodic fever-aphthae-pharyngitis-adenitis syndrome, Sweet syndrome, HIV infection (especially if ulcers are large and slow to heal), and cyclic neutropenia. Clinically, these ulcers are characterized by well-circumscribed, ulcerative lesions with a white necrotic base surrounded by a red halo.
11. **(B).** Eruption cyst is a smooth, painless swelling over the erupting tooth.
12. **(E).** Not attributed to another disorder.
13. **(D).** Lactase deficiency, glucose-galactose malabsorption, lactulose, and laxative abuse are causes of osmotic diarrhea.
14. **(B).** Rare causes of acute diarrhea in infancy include
 - Primary disaccharidase deficiency
 - Hirschsprung toxic colitis
 - Adrenogenital syndrome
 - Neonatal opiate withdrawal

- 15.(D). Delayed eruption of the 20 primary teeth can be familial or indicate systemic or nutritional disturbances such as hypopituitarism, hypothyroidism, cleidocranial dysplasia, trisomy 21, and multiple syndromes.
- 16.(A). Mottled enamel (permanent teeth) is seen in kidney failure and cystic fibrosis.
- 17.(D). The tongue is usually of normal size, but the floor of the mouth is foreshortened. The air passages can become obstructed, particularly on inspiration, usually requiring treatment to prevent suffocation.
- 18.(E). It is uncommon before school age.
- 19.(C). Contrast (usually barium) radiographic study of the esophagus and upper gastrointestinal tract is performed in children with vomiting and dysphagia to evaluate for achalasia, esophageal strictures and stenosis, hiatal hernia, and gastric outlet or intestinal obstruction. It has poor sensitivity and specificity in the diagnosis of GERD.
- 20.(B). Infantile reflux manifests more often with regurgitation (especially postprandially), signs of esophagitis (irritability, arching, choking, gagging, feeding aversion), and resulting failure to thrive; symptoms resolve spontaneously in the majority of infants by 12-24 mo.
- 21.(E). Seated position worsens infant reflux and should be avoided in infants with GERD.
- 22.(A). More common in boys.
- 23.(D). Trisomy 18.
- 24.(D).
- 25.(E). Meckel diverticulum is found twice as commonly in females.
- 26.(C). The most sensitive study is a Meckel radionuclide scan, which is performed after intravenous infusion of technetium-99m pertechnetate. The mucus-secreting cells of the ectopic gastric mucosa take up pertechnetate, permitting visualization of the Meckel diverticulum. The uptake can be enhanced with various agents, including cimetidine, ranitidine, glucagon, and pentagastrin. The sensitivity of the enhanced scan is approximately 85%, with a specificity of approximately 95%. A false-negative scan may be seen in anemic patients; although false-positive results are uncommon, they have been reported with intussusception, appendicitis, duplication cysts, arteriovenous malformations, and tumors.
- 27.(D). Cleft palate
- 28.(A). In 99% of healthy full-term infants, meconium is passed within 48 hr of birth.
- 29.(C). In children, it is most often associated with abdominal surgery or infection (gastroenteritis, pneumonia, peritonitis). Ileus also accompanies metabolic abnormalities (e.g., uremia, hypokalemia, hypercalcemia, hypermagnesemia, and acidosis) or administration of certain drugs, such as opiates, vincristine, and antitumor agents such as loperamide when used during gastroenteritis.
- 30.(B). The classic triad of pain, a palpable sausage-shaped abdominal mass, and bloody or currant jelly stool is seen in <30% of patients with intussusception.
- 31.(E). Ingestion of batteries rarely leads to problems, but symptoms can arise from leakage of alkali or heavy metal (mercury) from battery degradation in the

gastrointestinal tract. Batteries can also generate electrical current and thereby cause low-voltage electrical burns to the intestine.

32.(E). Sunflower seed bezoars are reported to cause rectal pain and constipation as a result of the seed shells being associated with fecal impaction. Endoscopic removal is indicated, as these bezoars are refractory to enema or lavage management.

33.(C). School-age children and adolescents more commonly present with epigastric pain and nausea, presentations generally seen in adults.

34.(D). If *H. pylori* is identified, even in a child with no symptoms, eradication therapy should be offered

35.(C). Rectal bleeding, diarrhea, mucus, pus, rectal involvement, toxic megacolon, sclerosing cholangitis, and crypt abscesses are more common in ulcerative colitis.

36.(B). In secretory diarrhea caused by disorders such as congenital chloride diarrhea and microvillus inclusion disease, the stool is watery and voluminous and can be mistaken for urine.

37.(D). The most common extraintestinal manifestation of celiac disease is iron-deficiency anemia, unresponsive to iron therapy. Osteoporosis may be present; in contrast to the situation in adults, it can be reversed by a gluten-free diet, with restoration of normal peak bone densitometric values. Other extraintestinal manifestations include short stature, arthritis and arthralgia, epilepsy with bilateral occipital calcifications, peripheral neuropathies, cardiomyopathy, isolated hypertransaminasemia, dental enamel hypoplasia, aphthous stomatitis, and alopecia.

38.(A). Peripheral neuropathy.

39.(D). Some diseases, many with an autoimmune pathogenesis, are found with a higher-than-normal incidence in celiac disease patients. Among these are type 1 diabetes, autoimmune thyroid disease, Addison disease, Sjögren syndrome, autoimmune cholangitis, autoimmune hepatitis, primary biliary cirrhosis. Such associations have been interpreted as a consequence of the sharing of identical HLA haplotypes. The relation between celiac disease and other autoimmune diseases is poorly defined; once those diseases are established, they are not influenced by a gluten-free diet. Other associated conditions include selective IgA deficiency and Down, Turner, and Williams syndromes.

40.(E). Intellectual development tends to be slow.

41.(C). Malabsorption is mainly seen after infection with *Campylobacter*, *Shigella*, *Salmonella*, *Giardia*, *cryptosporidium*, *coccidioidosis*, and *rotavirus*. These infectious causes of malabsorption are more common in immunocompromised children.

42.(A). The pathogenesis of the diarrhea is not always clear and may be related to secondary lactase deficiency, food protein allergy, antibiotic-associated colitis (including pseudomembranous colitis caused by *Clostridium difficile* toxin), or a combination of these.

43.(A). Vitamin B12 and bile salts are only absorbed in the distal ileum.

- 44.(C). Management of the diarrhea in chronically malnourished children is based on 3 principles: oral rehydration to correct dehydration, rapid resumption of feeds with avoidance of periods of nothing by mouth, and treating the etiology of the diarrhea.
- 45.(E). Cystic fibrosis is the most common congenital disorder associated with exocrine pancreatic insufficiency.
- 46.(B). Symptoms can include digestive disturbances such as vomiting, constipation, poor appetite, failure to thrive, hypercalcemia, nephrocalcinosis, fever, irritability, and ocular abnormalities.
- 47.(E). Features of Tangier disease include orange tonsils, hepatosplenomegaly, relapsing neuropathy, orange-brown spots on the colon and ileum, diarrhea in association with decreased plasma cholesterol levels (apolipoprotein A-I and A-II), and normal or elevated triglyceride levels. Specific therapy for Tangier disease has not yet been established.
- 48.(A). Laboratory findings are metabolic alkalosis, hypochloremia, hypokalemia, and hyponatremia (with high plasma renin and aldosterone activities).
- 49.(B). Acrodermatitis enteropathica requires long term treatment with elemental zinc 1 mg/kg/day.
- 50.(A). Direct person-to-person contact outbreaks of gastroenteritis are usually caused by norovirus and Shigella species. Unknown agents are seen in 30-40%; other pathogens include Salmonella, rotavirus, Giardia, Cryptosporidium, Clostridium difficile, and C. jejuni.
- 51.(D). Treatment of Campylobacter jejuni is supportive care. For severe cases, antibiotics, such as azithromycin and quinolones, may be indicated early in the diarrheal disease. Guillain-Barré syndrome can be a sequel.
- 52.(B). Treatment of Enterohemorrhagic Escherichia coli (EHEC) E. coli O157:H7 include supportive care, monitor renal function, hemoglobin, and platelets closely. E. coli O157:H7 infection is also associated with hemolytic uremic syndrome (HUS), which can cause lifelong complications. Studies indicate that antibiotics might promote the development of HUS. Antidiarrheal agents like Imodium may also increase the risk of developing HUS.
- 53.(A). Treatment of Vibrio cholerae is supportive care with aggressive oral and intravenous rehydration. Doxycycline is recommended as first-line treatment for adults, whereas azithromycin is recommended as first-line treatment for children and pregnant women. Ciprofloxacin and doxycycline recommended as second-line drugs for children.
- 54.(B). Signs and symptoms of Yersinia enterocolitica and Yersinia pseudotuberculosis are, appendicitis-like symptoms (diarrhea and vomiting, fever, abdominal pain) occur primarily in older children and young adults; might have a scarlatiniform rash or erythema nodosum with Y. Pseudotuberculosis.
- 55.(E). Risks associated with severe dehydration that might necessitate intravenous resuscitation include: age <6 mo; prematurity; chronic illness; fever >38°C (100.4°F) if

younger than 3 mo or $>39^{\circ}\text{C}$ (102.2°F) if 3-36 mo of age; bloody diarrhea; persistent emesis; poor urine output; sunken eyes; and a depressed level of consciousness.

56.(B). The low-osmolality World Health Organization (WHO) oral rehydration solution (ORS) containing 75 mEq of sodium, 64 mEq of chloride, 20 mEq of potassium, and 75 mmol of glucose per liter, with total osmolality of 245 mOsm/L, is now the global standard of care and more effective than home fluids, including decarbonated soda beverages, fruit juices, and tea. These are not suitable for rehydration or maintenance therapy because they have inappropriately high osmolalities and low sodium concentrations.

57.(D). Characterized by recurrent episodes of blood in the urine, this condition results from deposits of the protein IgA in the glomeruli. IgA nephropathy can progress for years with no noticeable symptoms. Men seem more likely to develop this disorder than women.

58.(E). Fatty foods or foods high in simple sugars (juices, carbonated sodas) should be avoided.

59.(A). Administration of zinc in community settings leads to increased use of ORS and reduction in the inappropriate use of antimicrobials.

60.(D). Because persistent vomiting can limit oral rehydration therapy, a single sublingual dose of an oral dissolvable tablet of ondansetron (4 mg 4-11 yr and 8 mg for children older than 11 yr [generally 0.2 mg/kg]) may be given.

61.(C). Nitazoxanide, an anti-infective agent, is effective in the treatment of a wide variety of pathogens, including *C. parvum*, *G. lamblia*, *E. histolytica*, *Blastocystis hominis*, *C. difficile*, and rotavirus.

62.(B). The drug of choice for shigella is Ciprofloxacin, ampicillin, ceftriaxone, azithromycin, or TMP-SMX.

63.(C). Abnormalities in the digestive-absorptive function tests suggest small bowel involvement, whereas intestinal inflammation, as demonstrated by increased fecal calprotectin or lactoferrin, supports colitis.

64.(A). Pain that wakes up the child from sleep, persistent right upper or right lower quadrant pain.

65.(E). Serum amyloid A protein is consistently elevated in patients with acute appendicitis with a sensitivity and specificity of 86% and 83%, respectively.

66.(E). A calcified appendicolith (5-10% of cases),

67.(B). Decreased local tenderness to compression.

68.(A). Meckel diverticulitis is an infrequent condition, but the clinical presentation closely mimics appendicitis and the diagnosis is usually made at surgery.

69.(A). There are many anomalies associated with anorectal malformations. The most common are anomalies of the kidneys and urinary tract in conjunction with abnormalities of the sacrum. This complex is often referred to as the caudal regression syndrome.

70.(C). 100% in Familial adenomatous polyposis (FAP) and Gardner syndrome.

- 71.(E). Polyps may be found anywhere in the gastrointestinal (GI) tract, most commonly in the colon or rectum; they are often solitary but may be multiple.
- 72.(D). The incidence of bilateral hernias is higher in girls and appears to be 20-40%.
- 73.(D). The features of the Johanson-Blizzard syndrome include exocrine pancreatic deficiency, aplasia or hypoplasia of the alae nasi, congenital deafness, hypothyroidism, developmental delay, short stature, ectodermal scalp defects, and absence of permanent teeth, urogenital malformations, and imperforate anus.
- 74.(B). Serum lipase is now considered the test of choice for acute pancreatitis as it is more specific than amylase for acute inflammatory pancreatic disease and should be determined when pancreatitis is suspected.
- 75.(A). Because of its ease, availability, and reliability, ultrasonography is the first choice.
- 76.(C). Affected infants have severe, generalized hypotonia and markedly impaired neurologic function with psychomotor retardation. Patients have an abnormal head shape and unusual facies, hepatomegaly, renal cortical cysts, stippled calcifications of the patellas and greater trochanter, and ocular abnormalities.
- 77.(D). Consistently pigmented stools rule against biliary atresia. The finding of bile-stained fluid on duodenal intubation also excludes biliary atresia.
- 78.(C). The success rate for establishing good bile flow after the Kasai operation is much higher (90%) if performed before 8 wk of life. Therefore, early referral and prompt evaluation of infants with suspected biliary atresia is important.
- 79.(A). It will prolong life and might correct the metabolic error in diseases such as α 1-antitrypsin deficiency, tyrosinemia, and Wilson disease.
- 80.(D). The diagnosis of CN type I is based on the early age of onset and the extreme level of bilirubin elevation in the absence of hemolysis. In the bile, bilirubin concentration is <10 mg/dL compared with normal concentrations of 50-100 mg/dL; there is no bilirubin glucuronide. Definitive diagnosis is established by measuring hepatic glucuronyl transferase activity in a liver specimen obtained by a closed biopsy.
- 81.(A). Liver histology demonstrates normal architecture, but hepatocytes contain black pigment similar to melanin. Liver function is normal and prognosis is excellent. The most commonly reported symptoms are abdominal pain and fatigue, jaundice, dark urine, and slight enlargement of the liver. Jaundice fluctuates in intensity and is aggravated by intercurrent disease.
- 82.(C). After adequate treatment, Kayser-Fleischer rings resolve.
- 83.(A). A major attempt should be made to restrict dietary copper intake to <1 mg/day. Foods such as liver, shellfish, nuts, and chocolate should be avoided.
- 84.(E). NISD is a rapidly fatal, progressive illness characterized by hepatomegaly, hypoglycemia, hypoprothrombinemia, hypoalbuminemia, hyperferritinemia, and hyperbilirubinemia. The coagulopathy is refractory to therapy with vitamin K. Liver pathology demonstrates severe liver injury with acute and chronic inflammation, fibrosis, and cirrhosis.
- 85.(E). Rare in HAV, common in HBV, HCV, and HDV, never in HEV.

- 86.(A). HBV is present in high concentrations in blood, serum, and serous exudates and in moderate concentrations in saliva, vaginal fluid, and semen.
- 87.(E). HBV is not thought to be transmitted via indirect exposure, such as sharing toys.
- 88.(D). Breastfeeding of nonimmunized infants by infected mothers does not confer a greater risk of hepatitis than does formula feeding.
- 89.(C). Only anti-HBs is present in persons immunized with hepatitis B vaccine, whereas both anti-HBs and anti-HBc are detected in persons with resolved infection.
- 90.(E). HBV is not spread by breastfeeding, kissing, hugging, or sharing water or utensils.
- 91.(C). Sclerosing cholangitis is the most common hepatobiliary disease associated with IBD, occurring in 2-8% of adult patients with ulcerative colitis and less often in Crohn disease.
- 92.(D). Cholestasis is the most severe complication and can lead to progressive fibrosis and cirrhosis. It is the major factor limiting effective long-term use of TPN in children and adults. Risk factors for TPN-associated cholestasis include prolonged duration of TPN, prematurity, low birthweight, sepsis, necrotizing enterocolitis, and short bowel syndrome.
- 93.(B). Primary elevation in transaminases and not alkaline phosphatase.
- 94.(B). Hypoplasia or absence of the gallbladder can be associated with extrahepatic biliary atresia or cystic fibrosis.
- 95.(E). Prolonged use of high-dose ceftriaxone, a third-generation cephalosporin, has been associated with the formation of calcium-ceftriaxone salt precipitates (biliary pseudolithiasis) in the gallbladder. Biliary sludge or cholelithiasis can be detected in >40% of children who are treated with ceftriaxone for at least 10 days. In rare cases, children become jaundiced and develop abdominal pain; precipitates usually resolve spontaneously within several months after discontinuation of the drug.
- 96.(C). Bleeding from esophageal varices is the most common presentation. Splenomegaly, sometimes with hypersplenism, is the next most common presenting feature in portal vein obstruction and may be discovered first on routine physical examination.
- 97.(D). Biliary atresia is the most common indication for liver transplantation in children, followed by metabolic and inborn disorders, autoimmune and familial cholestatic disorders, and acute hepatic necrosis.
- 98.(B). In children, hepatic and renal diseases are the most common causes.
- 99.(E). Pneumococci (most common), group A streptococci, enterococci, staphylococci, and Gram-negative enteric bacteria, especially *Escherichia coli* and *Klebsiella pneumoniae*, are most commonly found. *Mycobacterium tuberculosis*, *Neisseria meningitidis*, and *Mycobacterium bovis* are rare causes.
- 100.(A). It most commonly follows perforation of the appendix. Other causes include incarcerated hernias, rupture of a Meckel diverticulum, midgut volvulus, intussusception, hemolytic uremic syndrome, peptic ulceration, inflammatory bowel disease, necrotizing cholecystitis, necrotizing enterocolitis, typhlitis, and traumatic perforation.

1. Grunting is produced by expiration against a partially closed glottis and is an attempt to maintain positive airway pressure during expiration for as long as possible.

It is **MOST** commonly prominent in

- A. epiglottitis
- B. hyaline membrane disease
- C. asthma
- D. croup
- E. choanal atresia

2. The pulmonary vascular resistance and the pulmonary artery pressure to the systemic arterial resistance, from 3 mo after birth through childhood and adolescence is approximately

- A. 15% of the systemic values
- B. 25% of the systemic values
- C. 35% of the systemic values
- D. 45% of the systemic values
- E. 55% of the systemic values

3. Chest CT is of a particular importance in the evaluation of the following lesions **EXCEPT**

- A. mediastinal lesions
- B. cystic parenchymal lesions
- C. pulmonary embolisms
- D. vascular rings
- E. bronchiectasis

4. Rigid bronchoscopy is preferentially indicated for

- A. persistent pneumonia
- B. extracting foreign bodies
- C. atelectasis
- D. persistent wheeze
- E. interstitial disease

5. The following infections can cause sudden death in infants **EXCEPT**

- A. bronchiolitis (respiratory syncytial virus)
- B. infant botulism

- C. pertussis
- D. Staph. pneumonia
- E. sepsis

6. Declines of 50% or more in rates of sudden infant death syndrome (SIDS) in the USA and around the world have occurred in the past decade. The reductions in risk appear to be related primarily to

- A. decreases in placing infants prone for sleep
- B. exclusive breast-feeding
- C. adequate immunizations
- D. avoidance of soft sleeping surface
- E. avoidance of bed sharing with parent(s)

7. Foreign bodies are often placed in the nose by small children and developmentally delayed children.

Of the following, the **MOST** common presenting clinical symptom is

- A. mucopurulent nasal discharge
- B. foul nasal odor
- C. epistaxis
- D. nasal obstruction
- E. mouth breathing

8. Common causes of childhood epistaxis include the following **EXCEPT**

- A. epistaxis digitorum
- B. foreign bodies
- C. significant gastroesophageal reflux into the nose
- D. dry air
- E. allergic rhinitis

9. Of the following, the **MOST** common childhood cause of nasal polyposis is

- A. chronic sinusitis
- B. allergic rhinitis
- C. Samter triad
- D. Kartagener syndrome
- E. cystic fibrosis

10. The following may be effective as a common cold treatment **EXCEPT**

- A. vitamin C
- B. first generation antihistamine
- C. topical or oral adrenergic agents
- D. ipratropium bromide
- E. honey

11. Lemierre syndrome is characterized by septic thrombophlebitis of the internal jugular veins with septic pulmonary emboli, producing hypoxia and pulmonary infiltrates, it is a complication of pharyngitis caused by
- A. group A β -hemolytic streptococcus
 - B. *Arcanobacterium haemolyticum*
 - C. *Mycoplasma pneumoniae*
 - D. *Fusobacterium necrophorum*
 - E. *Corynebacterium diphtheriae*
12. Persistent cough may need to be sought beyond the lungs, because cough receptors also reside in the following regions **EXCEPT**
- A. pharynx
 - B. paranasal sinuses
 - C. stomach
 - D. nose
 - E. external auditory canal
13. Habit cough (“psychogenic cough” or “cough tic”) must be considered in any child with a cough that has lasted for weeks or months, that has been refractory to treatment and is
- A. abrupt
 - B. loud
 - C. barking quality
 - D. all day, never during sleep
 - E. paroxysmal
14. A “silent chest” with infiltrates should arouse suspicion of all the following **EXCEPT**
- A. alveolar proteinosis
 - B. bronchopulmonary dysplasia
 - C. *Pneumocystis jiroveci* infection
 - D. interstitial pneumonitis
 - E. tumors
15. The barking cough typical of croup is rare in
- A. laryngotracheobronchitis
 - B. acute epiglottitis
 - C. acute infectious laryngitis
 - D. spasmodic croup
 - E. measles croup
16. Children with croup should be hospitalized for any of the following **EXCEPT**
- A. progressive stridor

- B. severe stridor at rest
 - C. cyanosis
 - D. depressed mental status
 - E. congenital heart disease
17. Bacterial tracheitis is characterized by all the following **EXCEPT**
- A. Staphylococcus aureus is the most commonly isolated pathogen
 - B. incidence and severity do not differ by sex
 - C. mean age is between 5 and 7 yr
 - D. considered a primary bacterial illness
 - E. more common than epiglottitis in vaccinated populations
18. The **MOST** common congenital laryngeal anomaly that produces stridor is
- A. vocal cord paralysis
 - B. laryngomalacia
 - C. congenital subglottic stenosis
 - D. congenital laryngeal web
 - E. congenital subglottic hemangioma
19. Risk factors for persistent wheezing include all the following **EXCEPT**
- A. parental history of asthma
 - B. paternal smoking
 - C. persistent rhinitis (apart from acute upper respiratory tract infections)
 - D. eczema at <1 yr of age
 - E. frequent episodes of wheezing during infancy
20. Cardiovascular causes of wheezing include
- A. right atrial enlargement
 - B. left atrial enlargement
 - C. right ventricular enlargement
 - D. left ventricular enlargement
 - E. coarctation of aorta
21. Regarding congenital lobar emphysema (CLE), the following are true **EXCEPT**
- A. familial occurrence has been reported
 - B. usually no cause of CLE can be identified
 - C. usually become apparent in the neonatal period
 - D. many cases are diagnosed by antenatal ultrasonography
 - E. left upper lobe is the most affected site

22. Bronchiolitis obliterans (BO) is a rare chronic obstructive lung disease of the bronchioles and smaller airways. An insult to the lower respiratory tract occurs, resulting in fibrosis of the small airways.

Of the following, the **BEST** mean of establishing its diagnosis is

- A. chest radiographs
- B. pulmonary function tests
- C. ventilation-perfusion scans
- D. chest CT
- E. open lung biopsy

23. Pulmonary edema is traditionally separated into two categories according to cause (cardiogenic and non-cardiogenic). Non-cardiogenic pulmonary edema, in its most severe state, is also known as acute respiratory distress syndrome (ARDS).

The following radiographic features is usually present in non-cardiogenic pulmonary edema

- A. septal lines
- B. peribronchial cuffing
- C. pleural effusions
- D. air bronchograms
- E. even or central distribution of edema

24. The recurrent aspiration of small quantities of gastric, nasal, or oral contents can lead to several clinical presentations, including recurrent bronchitis or bronchiolitis; recurrent pneumonia; atelectasis; wheezing; cough; apnea; and/or laryngospasm.

The **MOST** common underlying problem associated with recurrent pneumonias in hospitalized children is

- A. oropharyngeal incoordination
- B. esophageal foreign body
- C. nasoenteric tube
- D. poor oral hygiene
- E. bronchopulmonary dysplasia

25. The **MOST** common etiology of pulmonary infiltrates with eosinophilia (PIE) is

- A. simple pulmonary eosinophilia
- B. acute eosinophilic pneumonia
- C. chronic eosinophilic pneumonia
- D. allergic bronchopulmonary aspergillosis
- E. parasitic infections

26. The **MOST** frequent pathogens of pneumonia in children 5 yr and older is

- A. *Streptococcus pneumoniae*
- B. *Mycoplasma pneumoniae*

- C. group A streptococci
 - D. H. influenzae (type b, nontypable)
 - E. adenovirus
27. The **MOST** frequent pathogen of pneumonia in children 4 mo-4 yr is
- A. Streptococcus pneumoniae
 - B. Mycoplasma pneumoniae
 - C. group A streptococci
 - D. H. influenzae (type b, nontypable)
 - E. Respiratory syncytial virus
28. Recurrent pneumonia is defined as
- A. 2 or more episodes in a single year, with radiographic clearing between occurrences
 - B. 2 or more episodes in a single year, without radiographic clearing between occurrences
 - C. 3 or more episodes in a single year, with radiographic clearing between occurrences
 - D. 3 or more episodes in a single year, without radiographic clearing between occurrences
 - E. 4 or more episodes in a single year
29. The **MOST** consistent clinical manifestation of pneumonia is
- A. subcostal retractions
 - B. grunting
 - C. tachypnea
 - D. fever
 - E. cough
30. The following are indications for admission to a hospital in children with pneumonia **EXCEPT**
- A. age <6 mo
 - B. sickle cell anemia
 - C. multiple lobe involvement
 - D. moderate to severe respiratory distress
 - E. vomiting
31. In the developed world, the **MOST** common cause of clinically significant bronchiectasis is
- A. primary ciliary dyskinesia
 - B. foreign body aspiration
 - C. aspiration of gastric contents

- D. immune deficiency syndromes
 - E. cystic fibrosis
32. The **MOST** common complaint in patients with bronchiectasis is
- A. cough and production of copious purulent sputum
 - B. hemoptysis
 - C. fever
 - D. anorexia
 - E. poor weight gain
33. Cystic fibrosis (CF) can be presented with all the following **EXCEPT**
- A. salt depletion
 - B. pansinusitis
 - C. rectal polyposis
 - D. pancreatitis
 - E. cholelithiasis
34. The earliest pathologic lesion in the lung of a patient with cystic fibrosis is
- A. bronchiolitis
 - B. bronchitis
 - C. bronchiolectasis
 - D. bronchiectasis
 - E. interstitial lung disease
35. Arterial blood gas study in cystic fibrosis shows
- A. hypernatraemic hypochloraemic metabolic alkalosis
 - B. hyponatraemic hypochloraemic metabolic acidosis
 - C. hyponatraemic hyperchloraemic metabolic alkalosis
 - D. hyponatraemic hypochloraemic metabolic alkalosis
 - E. hypernatraemic hyperchloraemic metabolic acidosis
36. The **MOST** common presenting feature of cystic fibrosis is
- A. failure to thrive
 - B. persistent respiratory symptoms
 - C. abnormal stools
 - D. meconium ileus
 - E. hepatobiliary disease
37. Regarding cystic fibrosis, all the following are true **EXCEPT**
- A. delayed sexual development
 - B. more than 95% of males are azoospermic
 - C. sexual function is generally impaired

- D. high incidence of inguinal hernia
- E. diminished female fertility rate

38. All the following conditions are associated with false-positive sweat test results **EXCEPT**

- A. eczema
- B. anorexia nervosa
- C. hypothyroidism
- D. Klinefelter syndrome
- E. malnutrition

39. The finding of the following pathogen on culture of the lower airways (sputum) strongly suggests a diagnosis of cystic fibrosis

- A. *B. cepacia*
- B. gram-negative rods
- C. nontuberculous mycobacterial species
- D. *Mycoplasma*
- E. *S. aureus*

40. All the following inhalation therapies in cystic fibrosis are true **EXCEPT**

- A. β -agonists may decrease PaO_2 acutely
- B. human recombinant DNase improves pulmonary function
- C. N-acetylcysteine is toxic to ciliated epithelium
- D. nebulized hypertonic saline improves mucociliary clearance
- E. aerosolized antibiotics are often used when the airways are colonized with *S. aureus*

41. The following are clinical manifestations of primary ciliary dyskinesia (PCD) **EXCEPT**

- A. recurrent pneumonia
- B. chronic otitis media
- C. chronic pansinusitis
- D. dextrocardia
- E. retinitis pigmentosa

42. The following are causes of diffuse alveolar hemorrhage (DAH) syndromes with pulmonary capillaritis **EXCEPT**

- A. Goodpasture syndrome
- B. Henoch-Schonlein purpura
- C. Wegener granulomatosis
- D. Heiner syndrome
- E. Behcet syndrome

43. Pathologic findings of pulmonary hemosiderosis in association with cow's milk hypersensitivity include all the following **EXCEPT**

- A. elevations of IgA
- B. peripheral eosinophilia
- C. alveolar deposits of IgG, IgA
- D. alveolar deposits of C3
- E. high titers to cow's milk protein

44. The classic laboratory findings of idiopathic pulmonary hemosiderosis (IPH) are the following **EXCEPT**

- A. microcytic hypochromic anemia
- B. elevated reticulocyte count
- C. normal plasma bilirubin
- D. reduced serum iron
- E. elevated iron-binding capacity

45. The treatment of choice of idiopathic pulmonary hemosiderosis (IPH) is

- A. transfusion of blood products
- B. systemic corticosteroids
- C. cyclophosphamide
- D. chloroquine
- E. lung transplantation

46. A commonly encountered risk factor for deep venous thrombosis (DVT) and pulmonary embolism (PE) in the pediatric population is

- A. central venous catheter
- B. hematologic malignancies
- C. sickle cell disease
- D. nephrotic syndrome
- E. antiphospholipid antibody syndrome

47. The **MOST** common form of pulmonary malignancy in children is

- A. bronchial carcinoid
- B. adenoid cystic carcinoma
- C. mucoepidermoid carcinoma
- D. metastatic lesions
- E. pulmonary blastoma

48. Pleurisy or inflammation of the pleura is often accompanied by an effusion. Of the following, the **MOST** common cause of pleural effusion in children is

- A. tuberculosis
- B. heart failure

- C. bacterial pneumonia
- D. rheumatoid arthritis
- E. metastatic intrathoracic malignancy

49. The **MOST** common cause of pneumomediastinum in older children and teenagers is

- A. dental extractions
- B. adenotonsillectomy
- C. tuberculosis
- D. mycoplasma pneumonia
- E. acute asthma

50. The following conditions are associated with increased intrathoracic pressure causing pneumothorax in children **EXCEPT**

- A. asthma
- B. pneumatocele
- C. bronchiolitis
- D. cystic fibrosis
- E. airway foreign body

51. One of the following is a rare cause of hydrothorax in children

- A. cardiac disease
- B. renal disease
- C. hepatic disease
- D. severe nutritional edema
- E. vascular obstruction by neoplasms

52. Of the following, the **LEAST** likely cause of hemothorax in children is

- A. chest trauma
- B. intrathoracic neoplasms
- C. costal exostoses
- D. blood dyscrasias
- E. rupture of an aneurysm

53. All the following are true in chylothorax **EXCEPT**

- A. fluid may be clear
- B. fluid triglyceride level is >110 mg/dL
- C. pleural fluid:serum triglyceride ratio is >1.0
- D. pleural fluid:serum cholesterol ratio is <1.0
- E. cells are primarily B lymphocytes

54. Bronchopulmonary dysplasia (BPD) is a syndrome characterized by signs and symptoms of chronic lung disease that originates in the neonatal period. An accepted definition includes a postnatal oxygen requirement for

- A. 7 days
- B. 14 days
- C. 21 days
- D. 28 days
- E. 35 days

55. The **MOST** common physical finding of the pulmonary exam in bronchopulmonary dysplasia (BPD) is

- A. tachypnea
- B. mouth breathing
- C. increased anteroposterior diameter of the chest
- D. intercostal retractions
- E. clear breath sounds

56. The etiology of wheezing in bronchopulmonary dysplasia (BPD) may be due to all the following **EXCEPT**

- A. lower airway inflammation
- B. bronchial smooth muscle irritation
- C. intraluminal exudate
- D. bronchial smooth muscle hypertrophy
- E. airway malacia

57. The following are features of pectus excavatum (funnel chest) **EXCEPT**

- A. occurs in 1 : 400 births
- B. usually associated with a connective tissue disorder
- C. 9 : 1 male preponderance
- D. accounts for >90% of congenital chest wall anomalies
- E. positive family history in one third of cases

58. Pectus carinatum is characterized by all the following **EXCEPT**

- A. accounting for 5-15% of congenital chest wall anomalies
- B. females are affected 4 times more often than males
- C. high familial occurrence
- D. common association of mild to moderate scoliosis
- E. association with mitral valve disease

59. Acute respiratory insufficiency is often the **MOST** prominent clinical manifestation of

- A. Duchenne muscular dystrophy

- B. spinal muscular atrophy
- C. congenital myotonic dystrophy
- D. myasthenia gravis
- E. Guillain-Barre syndrome

60. Asphyxiating thoracic dystrophy (Jeune syndrome) is associated with the following skeletal abnormalities **EXCEPT**

- A. narrowed thorax
- B. horizontal ribs
- C. short extremities
- D. bell-shaped chest cage
- E. low clavicles

61. Physical examination and an upright, posteroanterior radiograph with subsequent measurement of the angle of curvature (Cobb technique) remain the gold standard for assessment of scoliosis.

Scoliosis is defined when curves

- A. ≥ 5 degrees
- B. ≥ 10 degrees
- C. ≥ 15 degrees
- D. ≥ 20 degrees
- E. ≥ 25 degrees

- 1.(B). Grunting is most beneficial in alveolar diseases that produce widespread loss of FRC, such as in pulmonary edema, hyaline membrane disease, and pneumonia. Grunting is also effective in small airway obstruction (bronchiolitis) to maintain a higher positive pressure in the airway during expiration, decreasing the airway collapse.
- 2.(A). The PVR is ~50% of the systemic arterial resistance 3 days after birth. In the next several wk after birth as pulmonary arterial musculature in the tunica media involutes, there is a further decline in PVR and therefore in pulmonary artery pressure. Two to 3 mo after birth, the PVR and the pulmonary artery pressure are ~15% of the systemic values, a relationship that exists through childhood and adolescence.
- 3.(D). MRI is an excellent procedure to delineate hilar and vascular anatomy associated with vascular rings or slings.
- 4.(B). Rigid bronchoscopy is preferentially indicated for extracting foreign bodies, for removing tissue masses, and in patients with massive hemoptysis. In other cases, the flexible scope offers the advantages that it can be passed through endotracheal or tracheostomy tubes can be introduced into bronchi that come off the airway at acute angles, and can be safely and effectively inserted with topical anesthesia and conscious sedation.
- 5.(D). The following infections can cause sudden death in infants: sepsis, meningitis, encephalitis, brain abscess, hepatitis, pyelonephritis, bronchiolitis (respiratory syncytial virus), infant botulism, and pertussis.
- 6.(A). The reductions in risk appear to be related primarily to decreases in placing infants prone for sleep and increases in placing them supine. A number of other risk factors also have significant associations with SIDS; although many are nonmodifiable and most of the modifiable factors have not changed appreciably, self-reported maternal smoking prevalence during pregnancy has decreased by 25% in the past decade.
- 7.(A). Presenting clinical symptoms include history of insertion of foreign bodies (86%), mucopurulent nasal discharge (24%), foul nasal odor (9%), epistaxis (6%), nasal obstruction (3%), and mouth breathing (2%).
- 8.(C). Common causes of nosebleeds from the anterior septum include digital trauma, foreign bodies, dry air, and inflammation, including upper respiratory tract infections, sinusitis, and allergic rhinitis. Young infants with significant gastroesophageal reflux into the nose rarely present with epistaxis secondary to mucosal inflammation.
- 9.(E). Cystic fibrosis is the most common childhood cause of nasal polyposis and should be suspected in any child <12 yr old with nasal polyps, even in the absence of typical

respiratory and digestive symptoms; as many as 30% of children with cystic fibrosis acquire nasal polyps.

10.(A). Vitamin C, guaifenesin, and inhalation of warm, humidified air are no more effective than placebo for the treatment of cold symptoms.

11.(D). Lemierre syndrome is a serious complication of *F. necrophorum* pharyngitis and is characterized by septic thrombophlebitis of the internal jugular veins with septic pulmonary emboli, producing hypoxia and pulmonary infiltrates.

12.(D). Because cough receptors also reside in the pharynx, paranasal sinuses, stomach, and external auditory canal, the source of a persistent cough may need to be sought beyond the lungs.

13.(D). Habit cough (“psychogenic cough” or “cough tic”) must be considered in any child with a cough that has lasted for weeks or months, that has been refractory to treatment, and that disappears with sleep or with distraction.

14.(B). A “silent chest” with infiltrates should arouse suspicion of alveolar proteinosis, *Pneumocystis jiroveci* infection, genetic disorders of surfactant synthesis and secretion causing interstitial pneumonitis, or tumors.

15.(B). This dramatic, potentially lethal condition is characterized by an acute rapidly progressive and potentially fulminating course of high fever, sore throat, dyspnea, and rapidly progressing respiratory obstruction. The barking cough typical of croup is rare.

16.(E). Children with croup should be hospitalized for any of the following: progressive stridor, severe stridor at rest, respiratory distress, hypoxia, cyanosis, depressed mental status, poor oral intake, or the need for reliable observation.

17.(D). Bacterial tracheitis often follows a viral respiratory infection (especially laryngotracheitis), so it may be considered a bacterial complication of a viral disease, rather than a primary bacterial illness.

18.(B). Laryngomalacia is the most common congenital laryngeal anomaly and the most common cause of stridor in infants and children. Sixty percent of congenital laryngeal anomalies in children with stridor are due to laryngomalacia.

19.(B). Multiple studies have tried to predict which early wheezers will go on to have asthma in later life. Risk factors for persistent wheezing include parental history of asthma and allergies, maternal smoking, persistent rhinitis (apart from acute upper respiratory tract infections), eczema at <1 yr of age, and frequent episodes of wheezing during infancy.

20.(B). Cardiovascular causes of wheezing include dilated chambers of the heart including massive cardiomegaly, left atrial enlargement, and dilated pulmonary arteries.

21.(B). In 50% of cases, a cause of CLE can be identified. Congenital deficiency of the bronchial cartilage, external compression by aberrant vessels, bronchial stenosis, redundant bronchial mucosal flaps, and kinking of the bronchus caused by herniation into the mediastinum have been described as leading to bronchial obstruction and subsequent CLE and commonly affects the left upper lobe.

- 22.(E). Open lung biopsy or transbronchial biopsy remains the best means of establishing the diagnosis of BO.
- 23.(D). Air bronchograms is usually present in non cardiogenic edema while the other findings are usually found in cardiogenic pulmonary edema.
- 24.(A). Oropharyngeal incoordination is reportedly the most common underlying problem associated with recurrent pneumonias in hospitalized children. In one series of 238 children hospitalized with recurrent pneumonia, 48% were found to have dysphagia as the underlying problem.
- 25.(E). The most common etiology of PIE syndromes includes parasite infections and drug reactions.
- 26.(B). *Streptococcus pneumoniae* (pneumococcus) is the most common bacterial pathogen in children 3 wk to 4 yr of age, whereas *Mycoplasma pneumoniae* and *Chlamydia pneumoniae* are the most frequent pathogens in children 5 yr and older.
- 27.(E). The most frequent pathogens of pneumonia in children 4 mo-4 yr (in order of frequency) are Respiratory syncytial virus, other respiratory viruses (parainfluenza viruses, influenza viruses, and adenovirus), *S. pneumoniae*, *H. influenzae* (type b, nontypable), *Mycoplasma pneumoniae*, and group A streptococcus.
- 28.(A). Recurrent pneumonia is defined as 2 or more episodes in a single year or 3 or more episodes ever, with radiographic clearing between occurrences.
- 29.(C). Tachypnea is the most consistent clinical manifestation of pneumonia. Increased work of breathing accompanied by intercostal, subcostal, and suprasternal retractions, nasal flaring, and use of accessory muscles is common.
- 30.(B). Sick cell anemia with acute chest syndrome.
- 31.(E). In the developed world, cystic fibrosis is the most common cause of clinically significant bronchiectasis.
- 32.(A). The most common complaints in patients with bronchiectasis are cough and production of copious purulent sputum. Younger children may swallow the sputum. Hemoptysis is seen with some frequency. Fever can occur with infectious exacerbations. Anorexia and poor weight gain may occur as time passes.
- 33.(C). CF is responsible for most cases of exocrine pancreatic insufficiency in early life and is the major cause of severe chronic lung disease in children. It is also responsible for many cases of salt depletion, nasal polyposis, pansinusitis, rectal prolapse, pancreatitis, cholelithiasis, and insulin-dependent hyperglycemia. CF may manifest as failure to thrive and, occasionally, as cirrhosis or other forms of hepatic dysfunction.
- 34.(A). The earliest pathologic lesion in the lung is that of bronchiolitis (mucous plugging and an inflammatory response in the walls of the small airways); with time, mucus accumulation and inflammation extend to the larger airways (bronchitis).
- 35 (D).
- 36.(B). Failure to thrive 37.5%; persistent respiratory symptoms 45.6%; abnormal stools 28.8%; meconium ileus 19.9%; hepatobiliary disease 1.2%

- 37.(C). More than 95% of males are azoospermic because of failure of development of Wolffian duct structures, but sexual function is generally unimpaired.
- 38.(E). Dilution, malnutrition, edema, insufficient sweat quantity, hyponatremia, and cystic fibrosis transmembrane conductance regulator (CFTR) mutations with preserved sweat duct function are causes of false negative sweat test.
- 39.(E). The finding of *S. aureus* or *P. aeruginosa* on culture of the lower airways (sputum) strongly suggests a diagnosis of CF. In particular, mucoid forms of *P. aeruginosa* are often recovered from CF lungs. *B. cepacia* recovery also suggests CF. A wide range of other organisms are frequently recovered, particularly in advanced lung disease; they include a variety of gram-negative rods, fungi, and nontuberculous mycobacterial species.
- 40.(E). Aerosolized antibiotics are often used when the airways are colonized with *Pseudomonas* as part of daily therapy. Aerosolized tobramycin, TOBI, used as a suppressive therapy (on 1 month, off 1 month) may reduce symptoms, improve pulmonary function, and alleviate the need for hospitalization.
- 41.(D). Left-right laterality defects are found in PCD; 50% of patients have situs inversus totalis with transposition of the thoracic and abdominal organs.
- 42.(D). Heiner syndrome is a cause of diffuse alveolar hemorrhage (DAH) syndromes without pulmonary capillaritis.
- 43.(A). Pathologic findings have included elevations of IgE.
- 44.(C). The anemia of IPH can mimic a hemolytic anemia. Elevations of plasma bilirubin are caused by absorption and breakdown of hemoglobin in the alveoli.
- 45.(B). In IPH, early treatment with systemic corticosteroids is the treatment of choice.
- 46.(A). A commonly encountered risk factor for DVT and PE in the pediatric population is the presence of a central venous catheter. The presence of a catheter in a vessel lumen as well as instilled medications can induce endothelial damage and favor thrombus formation.
- 47.(D). Metastatic lesions are the most common forms of pulmonary malignancy in children; primary processes include Wilms tumor, osteogenic sarcoma, and hepatoblastoma.
- 48.(C). The most common cause of pleural effusion in children is bacterial pneumonia.
- 49.(E). Acute asthma is the most common cause of pneumomediastinum in older children and teenagers. Simultaneous pneumothorax is unusual in these patients.
- 50.(B). Pneumatocele, lung abscess, and bronchopleural fistula are infectious causes of pneumothorax in children.
- 51.(E). Hydrothorax is most often associated with cardiac, renal, or hepatic disease. It can also be a manifestation of severe nutritional edema and hypoalbuminemia. Rarely, it results from vascular obstruction by neoplasms, enlarged lymph nodes, pulmonary embolism, or adhesions. It may occur from a ventriculoperitoneal shunt or peritoneal dialysis and has been reported in congenital parvovirus B19 infection.
- 52.(E). Rupture of an aneurysm is unlikely during childhood.
- 53.(E). The cells are primarily T lymphocytes.

- 54.(D). An accepted definition includes an oxygen requirement for 28 days postnatally, and the disorder is graded as mild, moderate or severe on the basis of supplemental oxygen requirement and gestational age.
- 55.(A). Physical findings of the pulmonary exam vary with the severity of disease. Tachypnea is a common finding.
- 56.(C). The etiology of wheezing in BPD may be lower airway inflammation, bronchial smooth muscle irritation, bronchial smooth muscle hypertrophy, and airway malacia.
- 57.(B). It may be associated with a connective tissue disorder (Marfan or Ehlers-Danlos syndrome).
- 58.(B). Males are affected 4 times more often than females.
- 59.(E). Acute respiratory insufficiency is often the most prominent clinical manifestation of several acute neuromuscular disorders, such as high-level spinal cord injury, poliomyelitis, Guillain-Barre syndrome, and botulism. Although much more insidious in its clinical course, respiratory dysfunction constitutes the leading cause of morbidity and mortality in progressive neuromuscular disorders (e.g., Duchenne muscular dystrophy, spinal muscular atrophy, congenital myotonic dystrophy, myasthenia gravis, and Charcot-Marie-Tooth disease).
- 60.(E). Physical examination reveals a narrowed thorax that, at birth, is much smaller than the head circumference. The ribs are horizontal, and the child has short extremities. Chest radiographs demonstrate a bell-shaped chest cage with short, horizontal, flaring ribs and high clavicles.
- 61.(B). Curves >10 degrees define the presence of scoliosis.

1. CHARGE association is characterized by the following cardiac feature
 - A. endocardial cushion defect
 - B. supraaortic stenosis
 - C. tetralogy of Fallot
 - D. patent ductus arteriosus
 - E. anomalous pulmonary venous return
2. Hypertrophic cardiomyopathy is a recognized association with
 - A. infant of diabetic mother
 - B. Marfan syndrome
 - C. William syndrome
 - D. trisomy 21 (Down syndrome)
 - E. XO (Turner syndrome)
3. The following are characters of venous hum **EXCEPT**
 - A. continuous murmur
 - B. heard in infraclavicular region
 - C. grades I-III/VI
 - D. louder with patient in supine position
 - E. changes with compression of jugular vein
4. Chest pain in the pediatric patient often generates a significant amount of patient and parental concern.
Of the following, the **MOST** common cause is
 - A. myocarditis
 - B. pericarditis
 - C. pleurisy
 - D. pneumothorax
 - E. pneumonia
5. Congenital heart disease causing cyanosis without respiratory distress include the following **EXCEPT**
 - A. tricuspid atresia
 - B. aortic stenosis
 - C. Ebstein anomaly
 - D. pulmonary atresia

- E. tetralogy of Fallot
6. The **MOST** common cyanotic cardiac lesion to present in the newborn period is
- A. dextroposed transposition of the great arteries
 - B. hypoplastic left heart syndrome
 - C. tetralogy of Fallot
 - D. truncus arteriosus
 - E. pulmonary atresia
7. The ECG shows left ventricular hypertrophy and a superior QRS axis (between 0° and -90°) in
- A. tricuspid atresia
 - B. Ebstein anomaly
 - C. pulmonary atresia
 - D. pulmonary stenosis
 - E. tetralogy of Fallot
8. The **MOST** common cause of death from cardiac defects in the first month of life is
- A. d-Transposition without associated lesions
 - B. hypoplastic left heart syndrome
 - C. pulmonary atresia
 - D. truncus arteriosus
 - E. complex single ventricle
9. All the following are causes of heart failure in full-term neonate **EXCEPT**
- A. asphyxial cardiomyopathy
 - B. coarctation of aorta
 - C. hypoplastic left heart
 - D. transposition of great arteries
 - E. ventricular septal defect
10. One of the following is a sign of right-sided heart failure
- A. edema
 - B. tachypnea
 - C. orthopnea
 - D. wheezing
 - E. pulmonary edema
11. The **MOST** common cause of syncope in children is
- A. Wolff-Parkinson-White syndrome
 - B. prolonged QT syndrome
 - C. atrioventricular block

- D. neurocardiogenic syncope
- E. myocarditis

12. Of the following, the **MOST** common clinical sign of coarctation of the aorta in older children is

- A. cardiac enlargement
- B. notching of the inferior border of the ribs
- C. a systolic ejection click or thrill in the suprasternal notch
- D. differential blood pressure: arms > legs
- E. diminished or absent femoral or lower-extremity pulses

13. The chest x-ray may help to differentiate the types of congenital heart defects. Increased pulmonary markings (increased pulmonary blood flow) is seen in

- A. pulmonary atresia
- B. truncus arteriosus
- C. tetralogy of Fallot
- D. tricuspid atresia
- E. Ebstein anomaly

14. The following are causes of congestive heart failure and cardiomegaly during the newborn period but no murmur **EXCEPT**

- A. endocardial fibroelastosis
- B. asphyxia
- C. glycogen storage disease (Pompe disease)
- D. paroxysmal supraventricular tachycardia
- E. sepsis

15. SVT in children differ from physiologic sinus tachycardia by all the following **EXCEPT**

- A. sudden onset and termination
- B. persistent ventricular rate of >180 bpm
- C. fixed RR interval on ECG
- D. abnormal P-wave shape or axis or absent P waves
- E. evident change in heart rate with activity

16. The following cardiac lesions are at increased risk for bacterial endocarditis **EXCEPT**

- A. mitral insufficiency
- B. aortic stenosis
- C. atrial septal defect secundum
- D. coarctation of the aorta
- E. patent ductus arteriosus

17. The following factors are most strongly associated with the development of coronary artery disease in patients with Kawasaki disease **EXCEPT**
- A. duration of fever of >16 days
 - B. first-degree heart block
 - C. cardiomegaly
 - D. male gender
 - E. age of <1 year
18. The following are major side effects of PGE₁ **EXCEPT**
- A. apnea
 - B. fever
 - C. cutaneous flushing
 - D. seizures
 - E. hypertension
19. One of the following cardiac lesions have been identified more in male
- A. transposition of the great arteries
 - B. atrial septal defect
 - C. VSD
 - D. PDA
 - E. pulmonic stenosis
20. In **MOST** patients with an ASD, the characteristic physical finding is
- A. a right ventricular systolic lift
 - B. a fixed splitted 2nd heart sound
 - C. a systolic ejection murmur
 - D. a short, rumbling mid-diastolic murmur
 - E. a mild left precordial bulge
21. Indications for surgical closure of a VSD include the following **EXCEPT**
- A. patients in whom clinical symptoms and failure to thrive cannot be controlled medically
 - B. infants between 6 and 12 mo of age with large defects associated with pulmonary hypertension
 - C. patients older than 24 mo with a Qp : Qs ratio greater than 2 : 1.
 - D. patients with a supracristal VSD of any size
 - E. severe pulmonary vascular disease nonresponsive to pulmonary vasodilators
22. Regarding supracristal ventricular septal defect, the following are true **EXCEPT**
- A. a supracristal VSD is complicated by aortic insufficiency in 50-90% of patients
 - B. the incidence is higher in Asian children

- C. aortic insufficiency is most often not recognized until late in the 1st decade of life
 - D. the murmur of a supracristal VSD is usually heard at the mid to upper left sternal border
 - E. closure of supracristal ventricular VSDs at the time of diagnosis is not recommended in an asymptomatic child
23. All the following are signs of coarctation of the aorta **EXCEPT**
- A. femoral pulses are weak or absent
 - B. bounding pulses of the arms
 - C. femoral pulse occurs slightly before the radial pulse
 - D. blood pressure in the legs is lower than that in the arms
 - E. precordial impulse and heart sounds are usually normal
24. All the following are causes of chronic pulmonary venous hypertension **EXCEPT**
- A. congenital mitral stenosis
 - B. total anomalous pulmonary venous return with obstruction
 - C. peripheral pulmonary stenosis
 - D. left atrial myxomas
 - E. cor triatriatum
25. Regarding mitral valve prolapse; all the following are true **EXCEPT**
- A. it is predominantly congenital
 - B. it is usually sporadic
 - C. it is more common in girls
 - D. the dominant abnormal signs are auscultatory
 - E. antibiotic prophylaxis is recommended during surgery and dental procedures
26. Sinus bradycardia is due to slow discharge of impulses from the sinus node, the heart's natural pacemaker. Sinus bradycardia in neonates is considered if sinus rate is
- A. <100 beats/min
 - B. <90 beats/min
 - C. <80 beats/min
 - D. <70 beats/min
 - E. <60 beats/min
27. The following procedures may abort an attack of SVT **EXCEPT**
- A. placing of the face in ice water
 - B. straining
 - C. breath holding
 - D. standing on head
 - E. ocular pressure

28. The following drugs may cause long Q-T syndromes (LQTS) **EXCEPT**
- A. trimethoprim/sulfamethoxazole
 - B. erythromycin
 - C. imipramine
 - D. pentostam
 - E. risperidone
29. The **MOST** common cause of death in competitive athletes is
- A. aortic stenosis
 - B. hypertrophic cardiomyopathy
 - C. coronary artery abnormalities
 - D. myocarditis
 - E. long Q-T syndrome (LQTS)
30. The leading causative agents for endocarditis in pediatric patients are
- A. group D enterococci
 - B. viridans-type streptococci
 - C. *Pseudomonas aeruginosa*
 - D. fungal organisms
 - E. *Serratia marcescens*
31. The following is a major Duke criterion for the diagnosis of endocarditis
- A. new valve regurgitant flow by echocardiography
 - B. Osler nodes
 - C. single positive blood culture
 - D. serologic evidence of infection
 - E. high erythrocyte sedimentation rate
32. The lower limit of pulse rate in neonate at rest is
- A. 50/min
 - B. 60/min
 - C. 70/min
 - D. 80/min
 - E. 90/min
33. Careful evaluation of the character of the pulses is an important early step in the physical diagnosis of congenital heart disease. A wide pulse pressure with bounding pulses may suggest the following **EXCEPT**
- A. patent ductus arteriosus
 - B. aortic insufficiency
 - C. cardiomyopathy
 - D. anemia

- E. anxiety
34. Tall (>2.5 mm), narrow, and spiked P waves are seen in
- A. Ebstein anomaly
 - B. ventricular septal defect [VSD]
 - C. patent ductus arteriosus [PDA]
 - D. severe mitral stenosis
 - E. mitral regurgitation
35. In utero heart failure, often with fetal pleural and pericardial effusions and generalized ascites (non-immune hydrops fetalis) may occur in
- A. ventricular septal defect
 - B. coarctation of aorta
 - C. d-Transposition of great arteries
 - D. Ebstein anomaly
 - E. single ventricle
36. Of the following, the cardiac lesion resulting in increased volume load is
- A. valvular pulmonic stenosis
 - B. patent ductus arteriosus
 - C. valvular aortic stenosis
 - D. coarctation of the aorta
 - E. mitral stenosis
37. Of the following, the cyanotic cardiac lesion with decreased pulmonary blood flow is
- A. transposition of the great vessels
 - B. single ventricle
 - C. truncus arteriosus
 - D. tricuspid atresia
 - E. total anomalous pulmonary venous return without obstruction
38. The predominant manifestations of primary pulmonary hypertension include the following **EXCEPT**
- A. exercise intolerance
 - B. precordial chest pain
 - C. syncope
 - D. low arterial oxygen saturation
 - E. dizziness
39. The following are common side effects of sildenafil **EXCEPT**
- A. flushing

- B. elevated liver function tests
 - C. headache
 - D. diarrhea
 - E. myalgia
40. Postoperative (cardiac surgery) pulmonary hypertension can be managed with
- A. hyperventilation and inhaled nitric oxide
 - B. catecholamines
 - C. phosphodiesterase inhibitors
 - D. nitroprusside
 - E. diuretics
41. Patients who have undergone surgery entailing the use of cardiopulmonary bypass, should be watched carefully for
- A. epilepsy
 - B. learning disabilities
 - C. anemia
 - D. pulmonary hypertension
 - E. arrhythmias
42. The postpericardiotomy syndrome is characterized by all the following **EXCEPT**
- A. fever
 - B. decreased appetite
 - C. listlessness
 - D. vomiting
 - E. chest pain
43. For one of the following, total repair really achieved, with no requirement for long-term follow-up
- A. atrial septal defects
 - B. ventricular septal defects
 - C. valvar pulmonic stenosis
 - D. uncomplicated tetralogy of Fallot
 - E. isolated patent ductus arteriosus
44. Pregnancy is contraindicated in mothers with
- A. severe pulmonary hypertension
 - B. ventricular septal defects
 - C. prosthetic valves
 - D. atrial septal defects
 - E. unoperated cyanotic congenital heart disease

45. Surgical repair of VSD at the time of diagnosis should be considered in
- A. small supracristal VSD
 - B. nonrestrictive VSD
 - C. hemodynamically significant VSD
 - D. restrictive VSD
 - E. VSD with heart failure
46. The **MOST** common long-term complication for those patients who underwent early repair of complete atrioventricular canal before the development of pulmonary vascular disease is
- A. left AV valve regurgitation
 - B. subaortic stenosis
 - C. residual ventricular level shunts
 - D. complete heart block
 - E. endocarditis
47. Patients with a small PDA have the following criteria **EXCEPT**
- A. continuous murmur heard best at the left upper sternal border
 - B. normal peripheral pulses
 - C. normal pulmonary artery pressure by echocardiography
 - D. asymptomatic and live a normal life expectancy
 - E. risk for endocarditis is extremely low
48. Severe pulmonary valve stenosis defined as a peak gradient of
- A. >40 mm Hg
 - B. >45 mm Hg
 - C. >50 mm Hg
 - D. >55 mm Hg
 - E. >60 mm Hg
49. Symptoms in patients with aortic stenosis depend on the severity of the obstruction. Critical aortic stenosis associated with the following **EXCEPT**
- A. cardiomegaly
 - B. pulmonary edema
 - C. weak pulses in all extremities
 - D. pale skin
 - E. maximum intensity of the murmur
50. All the following are true regarding the diagnosis of coarctation of the aorta **EXCEPT**
- A. cardiac enlargement are noted with severe coarctation
 - B. notching of the ribs is common by late childhood
 - C. doppler is useful for demonstrating the specific site of the obstruction

- D. CT and MRI are valuable noninvasive tools for evaluation of coarctation when the echocardiogram is equivocal
 - E. diagnostic catheterization is mandatory before surgery
51. Shone complex consist of the following **EXCEPT**
- A. coarctation of the aorta
 - B. subvalvar aortic stenosis
 - C. valvar aortic stenosis
 - D. pulmonic stenosis
 - E. mitral stenosis
52. Paroxysmal hypercyanotic attacks (hypoxic, “blue,” or “tet” spells) are a particular problem during the 1st 2 yr of life. They are characterized by
- A. early evening occurrence
 - B. an increase in intensity of the systolic murmur
 - C. unpredictable onset
 - D. metabolic alkalosis
 - E. more frequent spells in patients with marked cyanosis at rest
53. Depending on the frequency and severity of hypercyanotic attacks in tetralogy of Fallot, all the following procedures are true **EXCEPT**
- A. placement of the infant in the knee-chest position
 - B. administration of oxygen
 - C. injection of morphine subcutaneously
 - D. rapid correction with intravenous sodium bicarbonate irrespective of the spell severity
 - E. intravenous administration of propranolol
54. The typical radiologic configuration in tetralogy of Fallot as seen in the anteroposterior view consists of all the following **EXCEPT**
- A. normal heart size
 - B. some elevation of the cardiac apex
 - C. convexity in the region of the main pulmonary artery
 - D. right-sided aortic arch
 - E. diminished pulmonary vascularity
55. The severity of symptoms of Ebstein anomaly and the degree of cyanosis are highly variable and depend on the extent of
- A. cardiac dysrhythmias
 - B. displacement of the tricuspid valve
 - C. atrial right-to-left shunt
 - D. pulmonary vascular resistance

E. polycythemia

56. D-transposition of the great vessels, a common cyanotic congenital anomaly, accounts for $\approx 5\%$ of all congenital heart disease. In this anomaly, all the following are true **EXCEPT**

- A. the systemic veins return to the right atrium
- B. the pulmonary veins return to the left atrium
- C. the connections between the atria and ventricles are normal
- D. the aorta is posterior and to the right of the pulmonary artery
- E. the aorta is anterior and to the right of the pulmonary artery

57. Obstructed total anomalous pulmonary venous return (TAPVR) is a pediatric cardiac surgical emergency because

- A. prostaglandin therapy is usually not effective
- B. severe pulmonary congestion develop
- C. left atrium may be small
- D. left ventricle may be small
- E. pulmonary hypertension develop

58. In neonates with total anomalous pulmonary venous return and marked pulmonary venous obstruction, the chest x-ray demonstrates

- A. small heart
- B. enlarged heart
- C. prominent pulmonary artery
- D. prominent right ventricle
- E. subaortic stenosis

59. In truncus arteriosus, a single arterial trunk arises from the heart and supplies the systemic, pulmonary, and coronary circulations. All the following are true **EXCEPT**

- A. VSD is always present
- B. the truncus overriding the defect
- C. both ventricles are at systemic pressure
- D. heart failure ensues immediately after birth
- E. clinical cyanosis is usually mild

60. Clinical manifestations of hypoplastic left-heart syndrome include

- A. cyanosis always obvious in the 1st 48 hr of life
- B. deep-blue color skin
- C. bounding peripheral pulses
- D. occasional association with Turner syndrome
- E. autosomal recessive inheritance is usual

61. Polysplenia (left isomerism) is characterized by
- A. right-sided stomach
 - B. decreased pulmonary blood flow
 - C. absent gallbladder
 - D. howell-jolly bodies
 - E. severe cyanosis
62. Pentalogy of Cantrell consists of the following **EXCEPT**
- A. ectopia cordis
 - B. midline supra umbilical abdominal defect
 - C. deficiency of the anterior diaphragm
 - D. defect of the upper sternum
 - E. intracardiac defect
63. The **MOST** common cause of pulmonary hypertension in pediatric patients is
- A. idiopathic pulmonary arterial hypertension
 - B. pulmonary venoocclusive disease
 - C. left-sided valvular heart disease
 - D. chronic obstructive pulmonary disease
 - E. interstitial lung disease
64. All patients with clinical evidence of a PDA are at increased risk for endocarditis. As result, PDA should be considered for closure **EXCEPT**
- A. PDA with severe irreversible pulmonary hypertension
 - B. small, hemodynamically insignificant PDA
 - C. small, hemodynamically significant PDA
 - D. moderate size PDA
 - E. large PDA
65. Phenytoin is used for treatment of
- A. supraventricular tachycardia
 - B. atrial fibrillation
 - C. atrial flutter
 - D. ventricular tachycardia
 - E. digitalis intoxication
66. Side effects of propranolol may include
- A. hypothyroidism
 - B. elevated triglycerides
 - C. school performance problems
 - D. hepatic toxicity
 - E. pulmonary fibrosis

67. The following criteria are indications for further investigation of premature ventricular contractions (PVCs) that could require suppressive therapy **EXCEPT**

- A. multiform PVCs
- B. disappear during exercise
- C. R-on-T phenomenon
- D. extreme frequency of beats
- E. presence of underlying heart disease

68. In urgent situations of supraventricular tachycardia (SVT) when symptoms of severe heart failure have already occurred.

Of the following, the initial management is

- A. adenosine by rapid intravenous push
- B. DC cardioversion (0.5-2 J/kg)
- C. verapamil
- D. digoxin
- E. amiodarone

69. Ventricular tachycardia (VT) is defined as at least 3 premature ventricular contractions PVCs at >120 beats/min. Hemodynamically unstable patients with VT should be immediately treated with

- A. catheter ablation
- B. ICD implantation
- C. DC cardioversion
- D. amiodarone
- E. procainamide

70. In 90% of cases of endocarditis, the causative agent is recovered from the first 2 blood cultures. Antimicrobial pretreatment of the patient reduces the yield of blood cultures to

- A. 10%
- B. 30%
- C. 40%
- D. 60%
- E. 80%

71. Despite the use of antibiotic agents, mortality remains high, in the range of 20-25%. Serious morbidity occurs in 50-60% of children with documented infective endocarditis.

Of the following, the **MOST** common morbidity is

- A. heart failure
- B. pulmonary emboli
- C. mycotic aneurysms
- D. acquired ventricular septal defect

E. heart block

72. Anthracycline cardiotoxicity (doxorubicin [Adriamycin]) on rare occasion causes acute inflammatory myocardial injury, but more classically results in dilated cardiomyopathy (DCM) and occurs in up to 30% of patients given a cumulative dose of doxorubicin exceeding

- A. 250 mg/m²
- B. 350 mg/m²
- C. 450 mg/m²
- D. 550 mg/m²
- E. 650 mg/m²

73. Restrictive cardiomyopathy (RCM) accounts for <5% of cardiomyopathy cases. RCM is characterized by all the following EXCEPT

- A. normal ventricular chamber dimensions
- B. normal myocardial wall thickness
- C. preserved systolic function
- D. normal atrial chamber dimensions
- E. high ventricular diastolic pressure

74. Diastolic dysfunction and normal systolic function are recognized features of

- A. dilated cardiomyopathy
- B. hypertrophic cardiomyopathy
- C. left ventricular noncompaction
- D. restrictive cardiomyopathy
- E. arrhythmogenic right ventricular cardiomyopathy

75. In Asia, the following hepatitis virus appears to be a significant cause of viral myocarditis

- A. hepatitis A virus
- B. hepatitis B virus
- C. hepatitis C virus
- D. hepatitis D virus
- E. hepatitis E virus

76. The vast majority of tumors originating from the heart are benign. Of the following, the **MOST** common pediatric cardiac tumors are

- A. fibromas
- B. rhabdomyomas
- C. myxomas
- D. hemangiomas
- E. papillomas

77. Afterload reducers are not indicated in children with heart failure secondary to
- A. cardiomyopathy
 - B. severe mitral insufficiency
 - C. aortic insufficiency
 - D. ventricular septal defect
 - E. aortic stenosis
78. Adverse reactions to angiotensin-converting enzyme inhibitors (ACEIs) include the following **EXCEPT**
- A. hypotension
 - B. hypokalemia
 - C. maculopapular pruritic rash
 - D. renal toxicity
 - E. chronic cough
79. The definition of hypertension in children is
- A. average systolic blood pressure (SBP) and/or diastolic BP that is ≥ 95 th percentile for age, sex, on ≥ 3 occasions
 - B. average systolic blood pressure (SBP) and/or diastolic BP that is ≥ 95 th percentile for age, sex, and height on ≥ 2 occasions
 - C. average systolic blood pressure (SBP) and/or diastolic BP that is ≥ 95 th percentile for age, sex, and height on ≥ 3 occasions
 - D. average systolic blood pressure (SBP) and/or diastolic BP that is ≥ 90 th percentile for age, sex, and height on ≥ 3 occasions
 - E. average systolic blood pressure (SBP) and/or diastolic BP that is ≥ 99 th percentile for age, sex, and height on ≥ 3 occasions
80. Renal childhood diseases responsible for chronic hypertension include
- A. hemolytic-uremic syndrome
 - B. acute tubular necrosis
 - C. congenital dysplastic kidney
 - D. pyelonephritis
 - E. renal trauma
81. Measuring serum potassium is essential in a child with confirmed hypertension because hyperkalemia may be seen in
- A. Gordon syndrome
 - B. Liddle syndrome
 - C. glucocorticoid remedial aldosteronism
 - D. apparent mineralcorticoid excess
 - E. pheochromocytoma

1. **(C).** CHARGE association (coloboma, heart, atresia choanae, retardation, genital and ear anomalies) is associated with TOF, aortic arch and conotruncal anomalies.
2. **(A).** Marfan syndrome is associated with dilated and dissecting aorta, aortic valve regurgitation, mitral valve prolapsed, William syndrome with supravalvular aortic stenosis, peripheral pulmonary stenosis, Trisomy 21 (Down syndrome) with endocardial cushion defect, VSD, ASD, PDA, while XO (Turner syndrome) associated with coarctation of aorta, aortic stenosis.
3. **(D).** Louder with patient in upright position.
4. **(E).** Common causes are costochondritis, Trauma or muscle overuse/strain, asthma (often exercise-induced), severe cough, pneumonia, reflux esophagitis, anxiety, hyperventilation, precordial catch syndrome (Texidor's twinge), sickle cell vasoocclusive crisis, and idiopathic.
5. **(B).**
6. **(A).** Although dextroposed transposition of the great arteries represents only about 5% of congenital heart defects, it is the most common cyanotic lesion to present in the newborn period.
7. **(A).**
8. **(B).** Hypoplastic left heart syndrome accounts for 1% of all congenital heart defects but is the most common cause of death from cardiac defects in the first month of life.
9. **(E).** Fluid overload, PDA, VSD, and Cor pulmonale (BPD) are causes of heart failure in premature neonate.
10. **(A).** If left-sided failure is predominant, tachypnea, orthopnea, wheezing, and pulmonary edema are seen. Hepatomegaly, edema, and distended neck veins are signs of right-sided failure.
11. **(D).** In otherwise healthy children, neurocardiogenic syncope is most common. This entity goes by a number of terms, including vasovagal syncope, neurally mediated syncope, and autonomic syncope.
12. **(D).** The most common clinical sign of coarctation of the aorta in older children is differential blood pressure: arms > legs (100%).
13. **(B).**

Decreased pulmonary markings (diminished pulmonary blood flow)

- Pulmonary atresia or severe stenosis
- Tetralogy of Fallot
- Tricuspid atresia
- Ebstein anomaly

Increased pulmonary markings (increased pulmonary blood flow)

- Transposition of great arteries
 - Total anomalous pulmonary venous return
 - Truncus arteriosus
- 14.(A). Endocardial fibroelastosis is a cause after the neonatal period.
- 15.(E). Little change in heart rate with activity, crying, or breath holding.
- 16.(C).
- 17.(B). Dysrhythmias (other than first-degree heart block) is an associated factor.
- 18.(E). Apnea, fever, cutaneous flushing, seizures, hypotension, and bradycardia/tachycardia.
- 19.(A). Gender differences in the occurrence of specific cardiac lesions have been identified. Transposition of the great arteries and left-sided obstructive lesions are slightly more common in boys (~65%), whereas atrial septal defect, VSD, PDA, and pulmonic stenosis are more common in girls.
- 20.(B). In most patients with an ASD, the characteristic finding is that the 2nd heart sound is widely split and fixed in its splitting during all phases of respiration.
- 21.(E).
- 22.(E). Closure of all supracristal ventricular VSDs at the time of diagnosis is commonly recommended to prevent the development of aortic regurgitation, even in an asymptomatic child.
- 23.(C). The radial and femoral pulses should always be palpated simultaneously for the presence of a radial-femoral delay. Normally, the femoral pulse occurs slightly before the radial pulse. A radial-femoral delay occurs when blood flow to the descending aorta is dependent on collaterals, in which case the femoral pulse is felt after the radial pulse.
- 24.(C). A variety of lesions may give rise to chronic pulmonary venous hypertension, which when extreme may result in pulmonary arterial hypertension and right-sided heart failure. These lesions include congenital mitral stenosis, mitral insufficiency, total anomalous pulmonary venous return with obstruction, left atrial myxomas, cor triatriatum (stenosis of a common pulmonary vein), individual pulmonary vein stenosis, and supravalvular mitral rings.
- 25.(E). This lesion is not progressive in childhood, and specific therapy is not indicated. Antibiotic prophylaxis is no longer recommended during surgery and dental procedures.
- 26.(B). Sinus rate <90 beats/min in neonates and <60 beats/min in older children is considered to be sinus bradycardia.
- 27.(E). Ocular pressure must never be performed, and carotid sinus massage is very rarely effective.
- 28.(D). The other antiprotozoal agent, pentamidine isethionate may cause long Q-T syndrome (LQTS).
- 29.(B). The most common cause of death in competitive athletes is hypertrophic cardiomyopathy, with or without obstruction to left ventricular outflow.
- 30.(B). Viridans-type streptococci (α -hemolytic streptococci) and *Staphylococcus aureus* remain the leading causative agents for endocarditis in pediatric patients.

- 31.(A). The **Duke criteria** help in the diagnosis of endocarditis. **Major criteria** include (1) positive blood cultures (2 separate cultures for a usual pathogen, 2 or more for less typical pathogens), and (2) evidence of endocarditis on echocardiography (intracardiac mass on a valve or other site, regurgitant flow near a prosthesis, abscess, partial dehiscence of prosthetic valves, or new valve regurgitant flow).
- 32.(C). Lower limit of normal is 70/min, average 125/min, and upper limit is 190/min.
- 33.(C). The presence of diminished pulses in all extremities is associated with pericardial tamponade, left ventricular outflow obstruction, or cardiomyopathy.
- 34.(A). Tall (>2.5 mm), narrow, and spiked P waves are indicative of **right atrial enlargement** and are seen in congenital pulmonary stenosis, Ebstein anomaly of the tricuspid valve, tricuspid atresia, and sometimes cor pulmonale. These abnormal waves are most obvious in leads II, V₃R, and V₁. Similar waves are sometimes seen in thyrotoxicosis. **Broad P waves**, commonly **bifid** and sometimes **biphasic**, are indicative of left atrial enlargement. They are seen in some patients with large left-to-right shunts (ventricular septal defect [VSD], patent ductus arteriosus [PDA]) and with severe mitral stenosis or regurgitation. Flat P waves may be encountered in hyperkalemia.
- 35.(D). One notable exception is the case of severe regurgitant lesions, most commonly of the tricuspid valve. In these lesions (e.g., Ebstein anomaly or severe right ventricular outflow obstruction, the parallel fetal circulation cannot compensate for the volume load imposed on the right side of the heart. In utero heart failure, often with fetal pleural and pericardial effusions and generalized ascites (nonimmune hydrops fetalis) may occur.
- 36.(B). The most common lesions in this group are those that cause left-to-right shunting: atrial septal defect, ventricular septal defect (VSD), AV septal defects (AV canal), and patent ductus arteriosus.
- 37.(D). This group of congenital heart lesions include (tetralogy of Fallot, pulmonary atresia with an intact septum, tricuspid atresia, total anomalous pulmonary venous return with obstruction).
- 38.(D). Arterial oxygen saturation is usually normal unless there is an associated intracardiac shunt.
- 39.(B). Flushing, headache, diarrhea, hypotension, fluid retention, exacerbation of heart failure, anemia, elevated liver function tests, and palpitations are common side effects of bosentan.
- 40.(A).
- 41.(B). Patients who have undergone surgery entailing the use of cardiopulmonary bypass, especially in the newborn period, should be watched carefully during their early school years for signs of mild to moderate learning disabilities, which are often amenable to early remedial intervention. The risk is higher in patients who have undergone repair using hypothermic total circulatory arrest than in those where systemic blood flow is maintained using cardiopulmonary bypass.
- 42.(E). The postpericardiotomy syndrome may occur toward the end of the 1st postoperative week or may sometimes be delayed until weeks or months after surgery.

This febrile illness is characterized by fever, decreased appetite, listlessness, nausea, and vomiting. Chest pain is not always present, so a high index of suspicion should be maintained in any recently postoperative patient.

43.(E). Many argue that only for isolated patent ductus arteriosus is total repair really achieved, with no requirement for long-term follow-up.

44.(A). Pregnancy is contraindicated in mothers with severe pulmonary hypertension, severe obstructive lesions, and Marfan syndrome, aortic root >40 mm.

45.(A). The exception to this rule is patients with small supracristal or perimembranous VSD with associated prolapse of the aortic cusp into the defect resulting in progressive aortic regurgitation. These patients should be considered for surgical repair at the time of diagnosis to prevent progressive aortic valve damage.

46.(A). Overall, for those patients who underwent early repair before the development of pulmonary vascular disease, the long-term prognosis is good. The most common long-term complication is left AV valve regurgitation, with approximately 5-10% of patients requiring surgical revision for left AV valve repair or replacement during follow-up.

47.(E). A silent PDA is a tiny defect that cannot be heard by auscultation and is only detected by other nonclinical means such as echocardiography. Life expectancy is always normal in this population and the risk for endocarditis is extremely low. Patients with a small PDA have an audible long-ejection or continuous murmur heard best at the left upper sternal border that radiates to the back. In addition, they have normal peripheral pulses. Because there is negligible left to right shunting these patients have normal left aorta (LA) and left ventricle (LV) size and normal pulmonary artery pressure by echocardiography and chest x-ray. These patients like those with silent PDAs are asymptomatic and live a normal life expectancy. They have a higher risk for endocarditis.

48.(C). For those patients with severe stenosis (defined as a peak gradient of >50 mm Hg), the majority ultimately require an intervention, either surgery or balloon valvuloplasty by age 25 yr.

49.(E). If cardiac output is significantly decreased, the intensity of the murmur at the right upper sternal border may be minimal.

50.(E). In cases that are well defined by echocardiography, CT, or MRI, diagnostic catheterization is not usually required before surgery.

51.(D). Congenital mitral stenosis is a rare anomaly that can be isolated or associated with other defects, the most common being subvalvar and valvar aortic stenosis and coarctation of the aorta (Shone complex).

52.(C). The spells occur most frequently in the morning on initially awakening or after episodes of vigorous crying. Temporary disappearance or a decrease in intensity of the systolic murmur is usual as flow across the right ventricular outflow tract diminishes. The onset is usually spontaneous and unpredictable. Spells are associated with reduction of an already compromised pulmonary blood flow, which, when prolonged, results in severe systemic hypoxia and metabolic acidosis. Infants who are only mildly

cyanotic at rest are often more prone to the development of hypoxic spells because they have not acquired the homeostatic mechanisms to tolerate rapid lowering of arterial oxygen saturation, such as polycythemia.

53.(D). Because metabolic acidosis develops when arterial Po₂ is <40 mm Hg, rapid correction (within several minutes) with intravenous administration of sodium bicarbonate is necessary if the spell is unusually severe and the child shows a lack of response to the foregoing therapy.

54.(C). Concavity in the region of the main pulmonary artery.

55.(B). The severity of symptoms and the degree of cyanosis are highly variable and depend on the extent of displacement of the tricuspid valve and the severity of right ventricular outflow tract obstruction.

56.(D). In normally related great vessels, the aorta is posterior and to the right of the pulmonary artery; in d-transposition of the great arteries (d-TGA), the aorta is anterior and to the right of the pulmonary artery (the *d* indicates a dextropositioned aorta, transposition indicates that it arises from the anterior right ventricle).

57.(A). Obstructed TAPVR is a pediatric cardiac surgical emergency because prostaglandin therapy is usually not effective.

58.(A). In neonates with marked pulmonary venous obstruction, the chest x-ray demonstrates a very dramatic perihilar pattern of pulmonary edema and a small heart.

59.(D). When pulmonary vascular resistance is relatively high immediately after birth, pulmonary blood flow may be normal; as pulmonary resistance drops in the 1st mo of life, blood flow to the lungs is greatly increased and heart failure ensues.

60.(D). Although cyanosis may not always be obvious in the 1st 48 hr of life, a grayish-blue color of the skin is soon apparent and denotes a mix of cyanosis and poor perfusion. All of the peripheral pulses may be weak or absent. This lesion may be isolated or associated in 5-15% of patients with known genetic syndromes, such as Turner syndrome, trisomy 13, 18, or 21, Jacobsen syndrome (11q deletion), Holt-Oram syndrome, and Rubinstein-Taybi syndrome. Occasionally it is familial and inherited as an autosomal recessive trait.

61.(C). All other options are features of asplenia (right isomerism).

62.(D). Defect of the lower sternum.

63.(A). Idiopathic or familial disease is the most common in pediatric patients (~55%), followed by pulmonary hypertension secondary to congenital heart disease (~35%) and chronic respiratory disorders (~15%).

64.(A). All PDAs except for small silent PDAs and those patients with severe irreversible pulmonary hypertension should be considered for closure.

65.(E). Other options are treated by procainamide.

66.(C). Other options are side effects of amiodarone.

67.(B). It is important to distinguish PVCs that are benign from those that are likely to lead to more severe arrhythmias. The former usually disappear during the tachycardia of exercise. If they persist or become more frequent during exercise, the arrhythmia may have greater significance.

68.(B). In urgent situations when symptoms of severe heart failure have already occurred, synchronized DC cardioversion (0.5-2 J/kg) is recommended as the initial management.

69.(C).

70.(D). Antimicrobial pretreatment of the patient reduces the yield of blood cultures to 50-60%. The microbiology laboratory should be notified if the patient has received antibiotics so that more sophisticated methods can be used to recover the offending agent.

71.(A). The most common is heart failure caused by vegetations involving the aortic or mitral valve. Myocardial abscesses and toxic myocarditis may also lead to heart failure without characteristic changes in auscultatory findings and, occasionally, to life-threatening arrhythmias.

72.(D). Exceeding 550 mg/m². The risk of toxicity appears to be exacerbated by concomitant radiation therapy.

73.(D). Dramatic atrial dilation can occur as a result of the abnormal myocardial compliance and high ventricular diastolic pressure.

74.(D).

75.(C). Coxsackievirus and other enteroviruses, adenovirus, parvovirus, Epstein-Barr virus, parechovirus, influenza virus, and cytomegalovirus are the most common causative agents in children, though most known viral agents have been reported. In Asia, hepatitis C virus appears to be significant as well.

76.(B). Rhabdomyomas are the most common pediatric cardiac tumors and are associated with tuberous sclerosis in 70-95% of cases.

77.(E). ACEIs and ARBs are not generally used in the presence of stenotic lesions of the left ventricular outflow tract because of concern over coronary perfusion.

78.(B). Adverse reactions to ACEIs include hypotension and its sequelae (weakness, dizziness, syncope) and hyperkalemia.

79.(C). The National High Blood Pressure Education Program Working Group on High Blood Pressure in Children and Adolescents published the Fourth Report on the Diagnosis, Evaluation, and Treatment of High Blood Pressure in Children and Adolescents (Fourth Report) in 2004.

The Fourth Report defined hypertension as average systolic blood pressure (SBP) and/or diastolic BP that is ≥ 95 th percentile for age, sex, and height on ≥ 3 occasions. Prehypertension was defined as average SBP or diastolic BP that are ≥ 90 th percentile but < 95 th percentile.

80.(C). Other options are responsible for acute/ intermittent hypertension.

81.(A). Measuring serum potassium is essential because hypokalemia may be present in Liddle syndrome, glucocorticoidremedial aldosteronism, and apparent mineralcorticoid excesssyndrome, while hyperkalemia may be seen in Gordon syndrome.

1. The hematologic features of congenital hypoplastic anemia (Diamond-Blackfan Anemia) include all the following **EXCEPT**
 - A. normocytic anemia
 - B. reticulocytopenia
 - C. deficiency or absence of red blood cell bone marrow precursors
 - D. elevated fetal hemoglobin (Hb F)
 - E. elevated serum iron levels
2. The **MOST** specific test that is helpful to differentiate Diamond-Blackfan anemia from transient erythroblastopenia of childhood is
 - A. hemoglobin electrophoresis
 - B. reticulocytes count
 - C. erythrocyte adenosine deaminase (ADA) enzyme level assay
 - D. bone marrow examination
 - E. mean corpuscular volume (MCV)
3. Anemia and reticulocytopenia that occur in the 2nd half of infancy period is **LEAST** likely due to
 - A. congenital hypoplastic anemia (Diamond-Blackfan anemia)
 - B. transient erythroblastopenia of childhood
 - C. a protracted, prolonged course of the anemia of hemolytic disease of the newborn
 - D. aplastic crises complicating various types of chronic hemolytic anemias
 - E. Fanconi anemia
4. The main stay of treatment for congenital hypoplastic anemia is
 - A. corticosteroids
 - B. androgen
 - C. antithymocyte globulin (ATG)
 - D. fully matched-related stem cell transplantation
 - E. fully matched-unrelated stem cell transplantation
5. All the following are true regarding Transient Erythroblastopenia of Childhood (TEC) **EXCEPT**
 - A. it is more common than congenital hypoplastic (Diamond-Blackfan) anemia
 - B. most of the affected children are older than 12 mo at onset

- C. Parvovirus B19 infections is a common causative agent
 - D. virtually all children recover within 1-2 months
 - E. corticosteroid therapy is of no value
6. The valuable diagnostic feature that differentiate anemia of chronic disorder from iron deficiency anemia is
- A. low serum iron
 - B. low or normal serum transferrin
 - C. normal bone marrow cellularity
 - D. normochromic normocytic RBC
 - E. leukocytosis
7. All the following are true regarding physiologic anemia of infancy **EXCEPT**
- A. it is exaggerated in premature infants
 - B. the hemoglobin concentration range is between 9-11 g/dL
 - C. the hemoglobin concentration rarely falls below 10 g/dL in healthy infants
 - D. vitamin E deficiency does not play a role
 - E. iron deficiency is a contributing factor
8. Folic acid deficiency may develop in the following children **EXCEPT**
- A. a term infant fed on goat's milk
 - B. an infant fed on unfortified powdered milk
 - C. an infant fed on pasteurized cow's milk
 - D. a child with chronic use of phenytoin anticonvulsant
 - E. a child with trimethoprim prophylaxis for recurrent urinary tract infection
9. A 1-year-old child has folic acid deficiency since the age of 4 months, the best indicator of this deficiency is
- A. significant increment of mean corpuscular volume MCV
 - B. significant fall of reticulocytes count
 - C. high level of lactate dehydrogenase LDH
 - D. decrease level of RBC folate
 - E. increase number of hypersegmented neutrophils
10. The first laboratory marker in progressive iron deficiency anemia is
- A. depletion of bone marrow hemosiderin
 - B. falling of serum ferritin
 - C. decrease of serum iron and increase of the iron-binding capacity
 - D. decrease hemoglobin synthesis
 - E. hypochromic microcytic anemia
11. An extremely high RBC distribution width (RDW) is **MOST** likely consistent with

- A. iron deficiency anemia
- B. megaloblastic anemia
- C. sideroblastic anemia
- D. thalassemia
- E. sickle cell anemia

12. The laboratory finding that is **MOST** likely consistent with the diagnosis of spherocytosis is

- A. high reticulocytes count of more than 10%
- B. normal mean corpuscular volume MCV
- C. low mean corpuscular hemoglobin MCH
- D. high mean corpuscular hemoglobin concentration MCHC
- E. indirect hyperbilirubinemia

13. In hereditary spherocytosis, all the following will resolve postsplenectomy **EXCEPT**

- A. osmotic fragility
- B. anemia
- C. reticulocytosis
- D. hyperbilirubinemia
- E. hypoplastic/aplastic crises

14. Splenectomy is recommended in all the following conditions **EXCEPT**

- A. a 6-year-old child with hereditary spherocytosis and significant hemolysis
- B. a 7-year-old child with hereditary elliptocytosis and a hemoglobin level of 7 g/dL and corrected reticulocytes count of more than 15%
- C. an 8-year-old child with hereditary stomatocytosis with hemolysis
- D. a 6-year-old child with thalassemia major with splenomegaly and frequent blood transfusion requirement
- E. a 12-year-old child with chronic immune thrombocytopenia not responding to all modalities of medical treatment

15. The best diagnostic test for paroxysmal nocturnal hemoglobinuria (PNH) is

- A. acidified serum hemolysis (HAM) test
- B. sucrose lysis test
- C. complement assay
- D. flow cytometry
- E. bone marrow study

16. All the following may occur as a renal manifestation in a child with sickle cell anemia **EXCEPT**

- A. polyuria
- B. hyposthenuria

- C. nephrotic syndrome
- D. hematuria
- E. acute renal injury

17. The least common infection that may occur in a 6-year-old child with sickle cell anemia is

- A. Pneumococcus sepsis
- B. Haemophilus influenzae meningitis
- C. Salmonella osteomyelitis
- D. E.coli urinary tract infection
- E. Parvovirus B19 aplastic episode

18. Blood transfusion therapy in a child with sickle cell anemia is indicated in all the following conditions **EXCEPT**

- A. acute chest syndrome
- B. stroke
- C. chronic intolerable pain syndrome
- D. avascular necrosis of the hip joint
- E. splenic sequestration

19. The best assessment of iron overload for patients with thalassemia major is achieved by

- A. liver MRI
- B. bone marrow biopsy
- C. serum iron
- D. serum ferritin
- E. total iron binding capacity

20. The criteria for successful bone marrow transplantation in thalassemic patients include all the following **EXCEPT**

- A. age younger than 15-year-old
- B. no hepatomegaly
- C. no bone deformities
- D. no iron overload
- E. full HLA-matched sibling

21. The best treatment for spinal cord compression at the vertebral canal with neurological symptoms caused by extramedullary hematopoiesis in thalassemic patients is

- A. blood transfusion
- B. splenectomy
- C. iron chelation therapy

- D. local radiotherapy
- E. vitamin D supplementation

22. You are evaluating a 4-year-old child with β -thalassemia major; he is on chronic transfusion therapy since the age of 1 year; you suspect transfusion-induced hemosiderosis.

Of the following, the organ that is **LEAST** likely to be affected by iron deposition at this time is

- A. liver
- B. pancreas
- C. heart
- D. pituitary gland
- E. thyroid gland

23. A healthy 5-mo-old boy appears pale. Examination is unremarkable. Lab findings include: Hb 8.1 g/dL; WBC 4,800/mm³; platelets 144,000/mm³; MCV, 111 fL; blood film showed hypersegmented neutrophils; serum B12, 65 pg/mL (low). The infant is vigorously breast fed.

Of the following, the **NEXT** step of the management is

- A. performing Schilling test
- B. check serum B12 of the mother
- C. parenteral administration of vitamin B12
- D. administration of both folic acid and vitamin B12
- E. transfuse packed RBCs

24. A 9-month-old child with a hemoglobin concentration of 10 gm/dL and marked microcytosis; serum iron and total iron binding capacity are within normal limits; serum ferritin and hemoglobin electrophoresis are also normal.

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

- A. iron deficiency anemia
- B. sideroblastic anemia
- C. β -thalassemia minor
- D. α -thalassemia trait
- E. anemia due to chronic disease

25. Hereditary intrinsic factor deficiency (HIFD), formerly called congenital pernicious anemia, differs from the typical adult pernicious anemia by all the following **EXCEPT**

- A. it is prominent at around 3 mo of age
- B. stomach secretes acid normally
- C. stomach is histologically normal
- D. there are no antibodies to parietal cells
- E. there are no associated endocrine disorders

26. Increased incubated osmotic fragility test that is not corrected by the addition of glucose is **MOST** likely suggestive of
- A. hereditary spherocytosis
 - B. hereditary elliptocytosis
 - C. hereditary stomatocytosis
 - D. glucose-6-phosphate dehydrogenase deficiency
 - E. pyruvate kinase deficiency
27. All the following may cause autoimmune hemolytic anemia **EXCEPT**
- A. systemic lupus erythematosus (SLE)
 - B. immunodeficiency
 - C. mycoplasma pneumoniae infection
 - D. lymphoproliferative disorder
 - E. penicillin drug administration
28. All the following are correct regarding treatment of cold agglutinin disease **EXCEPT**
- A. patient should avoid exposure to cold
 - B. patient should be treated for underlying disease
 - C. plasmapheresis is a modality of treatment
 - D. glucocorticoids treatment is a modality of treatment
 - E. patient can be treated by splenectomy when there is no response to other modalities of treatment
29. Fragmentation hemolysis by mechanical injury may be seen in all the following **EXCEPT**
- A. extensive burns
 - B. Kasabach-Merritt syndrome
 - C. after cardiac surgery for prosthetic heart valve replacement
 - D. thrombotic thrombocytopenic purpura (TTP)
 - E. hemolytic uremic syndrome (HUS)
30. All the following conditions may be associated with iron deficiency **EXCEPT**
- A. hookworm infestation
 - B. prolonged intravascular hemolysis
 - C. celiac disease
 - D. congenital heart disease with right to left shunt
 - E. prolonged use of isoniazid (INH)
31. Parvo virus B19 infection may cause transient pancytopenia in
- A. thalassemia major
 - B. sickle cell anemia
 - C. hereditary spherocytosis

- D. hereditary elliptocytosis
- E. pyruvate kinase deficiency

32. Fresh frozen plasma (FFP) transfusion is the best treatment for which of the following conditions

- A. hemophilia a
- B. hemophilia b
- C. factor vii deficiency
- D. immunoglobulin replacement therapy
- E. bleeding due to warfarin therapy

33. Transfusion of fresh frozen plasma (FFP) is efficacious for the treatment of deficiency of all the following coagulation factors **EXCEPT**

- A. factor V
- B. factor X
- C. factor XI
- D. factor XIII
- E. protein C

34. Fresh frozen plasma (FFP) transfusion/replacement is indicated in all the following conditions **EXCEPT**

- A. a 9-year-old child with chronic liver disease with prolonged clotting times who needs a liver biopsy
- B. a 6-year-old child with protein S deficiency and significant deep venous thrombosis
- C. an 11-year-old child with thrombotic thrombocytopenic purpura (TTP) post allogenic bone marrow transplantation
- D. a 2-day-old preterm neonate with clotting deficiencies to prevent intraventricular hemorrhage
- E. a 1-week-old term neonate with hemorrhage secondary to vitamin K deficiency

35. The main reason behind gamma irradiation of the blood products is to prevent

- A. transfusion-related bacterial infections
- B. transfusion-related viral infections
- C. graft-versus-host disease (GVHD)
- D. allergic reactions
- E. transfusion-related acute lung injury (TRALI)

36. CMV-transmitted infection is least likely occurring in transfusion of which of the following blood products?

- A. whole blood
- B. packed RBCs

- C. granulocytes
 - D. platelets
 - E. fresh frozen plasma
37. A prolonged thrombin time with normal reptilase time is consistent with
- A. dysfibrinogenemia
 - B. afibrinogenemia
 - C. factor XIII deficiency
 - D. disseminated intravascular coagulopathy (DIC) with high level of fibrin split products
 - E. heparin overdose
38. In hemophilia A, factor VIII level activity should be increased to 100% in
- A. epistaxis
 - B. iliopsoas bleeding
 - C. tooth extraction
 - D. hematuria
 - E. gum bleeding
39. A 9-year-old male child is about to do elective tonsillectomy; preoperative investigations are all normal except for a significant prolonged partial thromboplastin time PTT; there was no previous bleeding after circumcision and emergency appendectomy.
- Of the following, the **MOST** likely diagnosis is
- A. hemophilia A
 - B. hemophilia B
 - C. hemophilia C
 - D. prekallikrein deficiency
 - E. factor XIII deficiency
40. Spontaneous intracranial hemorrhage is more likely to occur in
- A. factor VII deficiency
 - B. factor VIII deficiency
 - C. factor IX deficiency
 - D. factor XI deficiency
 - E. factor XIII deficiency
41. Systemic amyloidosis may be associated with deficiency of
- A. factor VII
 - B. factor VIII
 - C. factor IX
 - D. factor X

E. factor XI

42. In which of the following conditions you should use a plasma that is frozen within 24 hours after collection?

- A. factor V deficiency
- B. factor VII deficiency
- C. factor IX deficiency
- D. factor X deficiency
- E. factor XI deficiency

43. All the following are true regarding factor XIII **EXCEPT**

- A. the half-life is 5-7 days
- B. PT, PTT, and thrombin time (TT) are prolonged when the factor is deficient
- C. clot solubility test with 5m urea is a screening test for factor XIII deficiency
- D. factor assay is a specific test to detect its level
- E. the hemostatic level is only 2-3%

44. In severe liver disease with prolongation of the bleeding time, the best effective treatment before doing liver biopsy is

- A. intravenous vitamin K
- B. intramuscular vitamin K
- C. oral vitamin K
- D. fresh frozen plasma
- E. intravenous DDAVP

45. Vitamin K-dependent clotting factors include all the following **EXCEPT**

- A. factor I
- B. factor II
- C. factor VII
- D. factor IX
- E. protein S

46. A 16-year-old female is found to have multiple bruising and hypochromic microcytic anemia; during her first pregnancy she noticed disappearance of the bruises; she has a past history of uncomplicated emergency appendectomy at the age of 10 year.

Of the following, the drug **MOST** likely beneficial for alleviation of her symptoms is

- A. ferrous sulphate
- B. desmopressin acetate (DDAVP)
- C. recombinant factor VIII
- D. recombinant factor IX
- E. aminocaproic acid

47. Low platelets count may be encountered in all the following **EXCEPT**
- A. Von Willebrand disease (VWD) type 2 B
 - B. VWD type 2 N
 - C. Platelet type pseudo VWD
 - D. Bernard Soulier syndrome
 - E. Wiskott Aldrich syndrome
48. Which of the following is **FALSE** about factor V Leiden mutation?
- A. it is the most common inherited risk factor for thrombosis
 - B. heterozygous individuals have less risk for thrombosis than homozygotes
 - C. heterozygotes have an increased risk of arterial thrombosis
 - D. there is an increased frequency of thrombosis while receiving oral contraceptive agents
 - E. there is an increased risk of recurrent abortions
49. Arterial thrombosis is the least likely cause of
- A. stroke
 - B. a cold and pulseless lower extremity
 - C. renal infarction
 - D. myocardial infarction
 - E. pulmonary embolism
50. Lupus anticoagulants usually affect the following laboratory test
- A. bleeding time (BT)
 - B. partial thromboplastin time (PTT)
 - C. prothrombin time (PT)
 - D. thrombin time (TT)
 - E. clot solubility test
51. Which of the following is **FALSE** regarding standard unfractionated heparin therapy?
- A. it acts by enhancing the antithrombin III activity
 - B. it can be used safely during pregnancy
 - C. it should not be given for a child with bacterial endocarditis
 - D. it can be given intramuscularly
 - E. it needs monitoring by PTT test
52. All the following are true regarding low molecular weight heparin (LMWH) in comparison with unfractionated heparin (UFH) **EXCEPT**
- A. it is more stable
 - B. it is ease to titrate
 - C. it is monitored by activated PTT

- D. it can be used subcutaneously
- E. it can be used as an outpatient

53. A healthy 5-day-old male neonate develops bruising and melena. The pregnancy, and delivery course were unremarkable. The mother received heparin therapy for recurrent abortions secondary to lupus anticoagulants. The infant is vigorously breastfeeding. Examination reveals only multiple bruises. Lab tests reveals: hemoglobin, 8.4 g/dL; WBC, 8600/mm³; platelets count, 200000/mm³; PTT, 74 seconds; PT, 28 seconds; serum fibrinogen is normal.

Of the following, the **MOST** likely cause of this bleeding is

- A. disseminated intravascular coagulopathy (DIC)
- B. hemophilia
- C. vitamin K deficiency
- D. heparin therapy
- E. lupus anticoagulants

54. A previously healthy 10-month-old boy develops pallor over 2 weeks. Examination is unremarkable except for pallor. Lab tests includes: hemoglobin, 4.9 g/dL; WBC, 6700/mm³; platelets count, 180000/mm³; MCV, 79 fL; reticulocyte count 0.8%; hemoglobin electrophoresis, normal.

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

- A. pure red cell aplasia
- B. transient erythroblastopenia of childhood
- C. thalassemia minor
- D. aplastic anemia
- E. parvovirus infection

55. You are evaluating a 5-year-old male child with multiple ecchymoses distributed all over the extremities and trunk; examination reveal a palpable spleen 3 cm below the left costal margin. Lab findings include: hemoglobin, 11.3 g/dL; white blood cell count, 8700/mm³; platelets count, 21000/mm³.

Of the following, the most proper **NEXT** step is

- A. bleeding time
- B. coagulation profile
- C. platelets aggregation test
- D. abdominal ultrasonography
- E. bone marrow study

56. Both quantitative and qualitative platelets defects are seen in

- A. generalized vasculitis
- B. Bernard-Soulier syndrome
- C. Glanzmann's Thrombasthenia

- D. type I von Willebrand disease
- E. uremia

57. Which of the following viruses is more likely to be associated with chronic immune thrombocytopenia?

- A. cytomegalovirus
- B. parvovirus
- C. influenza virus
- D. parainfluenza virus
- E. human immune deficiency virus

58. Which of the following is least likely to be associated with prolonged thrombocytopenia in children?

- A. immune thrombocytopenia
- B. systemic lupus erythematosus (SLE)
- C. HIV infection
- D. type 2B von Willebrand disease
- E. Wiskott-Aldrich syndrome (WAS)

59. Which of the following drugs is least likely to cause thrombocytopenia in children?

- A. valproic acid
- B. phenytoin
- C. sulfonamides
- D. trimethoprim-sulfamethoxazole
- E. heparin

60. Microthrombocytes are **MOST** likely to be seen in

- A. immune thrombocytopenia (ITP)
- B. Bernard-Soulier syndrome
- C. Wiskott-Aldrich syndrome (WAS)
- D. Glanzmann's Thrombasthenia
- E. type 2B von Willebrand disease

61. Thrombocytopenia that improves with age is **MOST** likely a feature of

- A. congenital amegakaryocytic thrombocytopenia
- B. Bernard-Soulier syndrome
- C. Wiskott-Aldrich syndrome
- D. thrombocytopenia absent radius syndrome (TAR)
- E. aplastic anemia

62. You are going to explain the role of anti-D therapy to the parents of a 4-year-old boy with acute immune thrombocytopenia (ITP) who has several ecchymotic lesions located over extremities and trunk. There is no mucosal bleeding.

An important statement that should be included during the discussion is

- A. anti-D therapy is more effective than IVIG therapy in acute ITP
- B. anti-D therapy is very effective to all children with ITP
- C. when anti-D therapy is given, monitoring of hemoglobin concentration is recommended
- D. anti-D therapy is used in acute ITP only
- E. anti-D therapy can be used as initial treatment for children with acute ITP

63. A 9-month-old boy appears pale. Examination reveals a palpable liver 3 cm below the right costal margin and palpable spleen 4 cm below the left costal margin. Lab findings include: hemoglobin concentration, 6.4 g/dl; mean corpuscular volume (MCV), 67 fL; and WBC, 15,000/mm³.

Of the following, The **MOST** valuable test to confirm the diagnosis is

- A. blood smear
- B. serum iron, total iron binding capacity, and serum ferritin
- C. hemoglobin electrophoresis
- D. bone marrow study
- E. abdominal ultrasonography

64. You are explaining the risk of sepsis after splenectomy to medical students. An important statement that should be included during this discussion is that risk is especially high in children

- A. older than 5 yr at the time of surgery
- B. with hereditary spherocytosis
- C. with chronic immune thrombocytopenia (ITP)
- D. after trauma
- E. with Hodgkin lymphoma

65. You are evaluating a 7-year-old male child with cervical lymphadenopathy; the **MOST** likely finding that raise your suspicion of malignancy is

- A. matted nodes
- B. fluctuant nodes
- C. fixed nodes
- D. presence of systemic symptoms
- E. no regression in the size of the nodes to its normal within 2 weeks

66. Of the following, the **MOST** accurate reflection of active erythropoiesis of the bone marrow is

- A. hemoglobin concentration

- B. packed cell volume (PCV)
- C. reticulocyte count
- D. mean corpuscular volume (MCV)
- E. reticulocyte index

67. Hematologic disease with upper limb orthopedic congenital abnormalities with presence of thumbs is **MOST** likely seen in

- A. Fanconi anemia
- B. thrombocytopenia with Absent Radii (TAR) Syndrome
- C. congenital Amegakaryocytic Thrombocytopenia
- D. Glanzmann Thrombasthenia
- E. Bernard-Soulier Syndrome

68. Which of the following is not a feature of Wiskott–Aldrich Syndrome (WAS)

- A. thrombocytopenia
- B. eczema
- C. recurrent otitis media and pneumonia
- D. propensity to develop autoimmune disorders
- E. giant platelets by blood smear

69. A 5-year-old child has multiple bruises on her extremities and oral mucosal bleeding of 3 days duration; she had a mild respiratory tract infection 2 weeks before; physical examination was normal apart from multiple ecchymoses and petechiae.

Of the following, the **NEXT** diagnostic step is

- A. complete blood count
- B. prothrombin time
- C. bleeding time
- D. partial thromboplastin time
- E. antinuclear antibody titer

70. Of the following, the **LEAST** likely feature of Bernard-Soulier syndrome is

- A. autosomal recessive inheritance
- B. severe thrombocytopenia
- C. giant Platelets
- D. abnormal ristocetin test
- E. platelet transfusion is the only reliable therapy

71. Each of the following may cause thrombocytopenia **EXCEPT**

- A. heparin therapy
- B. aspirin ingestion
- C. disseminated intravascular coagulation (DIC)
- D. systemic lupus erythematosus (SLE)

E. hemolytic-uremic syndrome (HUS)

72. Which of the following children with acute immune thrombocytopenia is **MOST** likely to have a benefit from splenectomy?

- A. an 8-year-old girl with multiple bruises and platelets count of 10,000/ml
- B. a 6-year-old boy with mild epistaxis and platelets count of 20,000/ml
- C. a 4-year-old boy with subdural hematoma and platelets count of 100,000/ml after receiving IVIG
- D. an 11-year-old boy with severe rectal bleeding and platelets count of 20000/ml not responding to medical treatment
- E. a 13-year-old female adolescent with moderate menstrual bleeding and platelets count of 50000/ml

73. Of the following, the **LEAST** likely feature of acute immune thrombocytopenia (ITP) is

- A. a preceding viral infection
- B. isolated thrombocytopenia
- C. mucocutaneous bleeding
- D. isolated splenomegaly
- E. increased number of megakaryocytes in the bone marrow

74. A 3-year-old male child develops hematoma and bruising of his right hand next day after falling on the ground; the mother stated that her child has a poor wound healing and a history of delayed umbilical separation during the neonatal period.

Of the following, the **MOST** valuable test for this case is

- A. bleeding time
- B. partial thromboplastin time
- C. prothrombin time
- D. thrombin time
- E. clot solubility test

75. A 5-year-old child, with hemophilia A of severe type, presents to the emergency unit with a groin pain after a minor trauma to his back; his blood pressure is 60/30 mm Hg; his pulse rate is 180/min; he holds his right hip in a flexion position with internal rotation.

Of the following, the **NEXT** step in the management of this child is

- A. factor VIII replacement therapy
- B. intravenous 1-deamino-8-d-arginine vasopressin (DDAVP)
- C. factor VIII assay
- D. abdominal ultrasonography
- E. abdominal computed tomography (CT) scan

76. In a child with a recent diagnosis of hemophilia A who developed moderate epistaxis, the aim of the treatment is to increase the level of factor VIII to
- A. 5-20%
 - B. 35-50%
 - C. 55-60%
 - D. 65-70%
 - E. 80-100%
77. Allogeneic bone marrow stem cell transplantation is least likely to be required in
- A. Fanconi anemia
 - B. thrombocytopenia with Absent Radii (TAR) Syndrome
 - C. congenital Amegakaryocytic Thrombocytopenia (CAT)
 - D. Wiskott –Aldrich Syndrome (WAS)
 - E. severe combined immune deficiency
78. Inherited thrombocytopenias include all the following **EXCEPT**
- A. congenital Amegakaryocytic Thrombocytopenia
 - B. thrombocytopenia with Absent Radii (TAR) Syndrome
 - C. Glanzmann’s Thrombasthenia
 - D. Bernard-Soulier Syndrome
 - E. Wiskott–Aldrich Syndrome (WAS)
79. Which of the following is the best treatment for a 5-year-old girl with acute immune thrombocytopenia (ITP) and platelets count of 30,000/ml and mild petechial rash all over her body?
- A. intravenous immunoglobulin (IVIG)
 - B. anti-D gamma globulin
 - C. corticosteroids
 - D. platelets transfusion
 - E. no treatment
80. The standard care for **MOST** children with severe hemophilia is
- A. prevention by F VIII replacement therapy to prevent spontaneous bleeding and early joint deformities
 - B. aggressive treatment by F VIII replacement therapy when significant bleeding occurs
 - C. avoid trauma
 - D. avoid aspirin and other NSAID
 - E. avoid violent contact sports

- 1.(A). The RBCs in Diamond-Blackfan anemia are usually macrocytic for age, but there is no hypersegmentation of neutrophils.
- 2.(C). Erythrocyte adenosine deaminase (ADA) activity is increased in most patients with congenital hypoplastic anemia, a finding that helps distinguish this disorder from acquired transient erythroblastopenia of childhood. Also, because elevated ADA activity is not a fetal RBC feature, measurement of this enzyme is helpful in diagnosing Diamond-Blackfan anemia in very young infants. Hemoglobin F may be increased in 20% of cases with transient erythroblastopenia of childhood at the time of diagnosis.
- 3.(C). The anemia of hemolytic disease of the newborn can have a protracted course, but this usually terminates spontaneously at 5-8 wk of age. Aplastic anemic crises, frequently caused by parvovirus B19 infections, may complicate various types of chronic hemolytic disease, but usually after the first several months of life.
- 4.(A). Corticosteroid therapy is beneficial in three fourths of patients; the mechanism of its effect is unknown. Stem cell transplantation from a related histocompatible donor has a role in children who do not respond to corticosteroids and who have demonstrated a several-year need for RBC transfusions; The survival results for matched-related donors have been very encouraging, but the responses have been much inferior with the use of partially mismatched siblings or matched unrelated donors.
- 5.(C). In rare instances, a prolonged case of apparent TEC may be caused by parvovirus-induced RBC aplasia, occurring in children with hemolytic anemia or congenital or acquired immunodeficiencies.
- 6.(B). The serum iron level in anemia of chronic disorder is low, but without the increase in total iron-binding capacity (serum transferrin) that occurs in iron deficiency. This pattern of low serum iron and low to normal iron-binding protein (serum transferrin) is a regular and valuable diagnostic feature.
- 7.(E). The iron previously stored from the degraded fetal RBCs can be used for hemoglobin synthesis. The supply of stored iron is sufficient for hemoglobin synthesis, even in the absence of dietary iron intake, until approximately 20 wk of age. Unless there has been significant blood loss, iron stores should be sufficient to maintain erythropoiesis early on.
- 8.(C). Goat's milk and powdered milk are deficient with folic acid. Human breast milk, pasteurized cow's milk and infant formulas provide adequate amounts of folic acid. Anticonvulsants may decrease folate absorption. Trimethoprim may affect folate metabolism.
- 9.(D).

10.(A). In progressive iron deficiency, a sequence of biochemical and hematologic events occurs. First, the tissue iron stores represented by bone marrow hemosiderin disappear.

11.(C). RDW is usually high in IDA, megaloblastic anemia, and sideroblastic anemia, but it is extremely high in the last.

12.(D). The MCHC is a measure of cellular hydration status. A high value (36-38 g/ dL) is characteristic of spherocytosis and a low value is commonly associated with iron deficiency.

13.(A). After splenectomy, osmotic fragility often improves because of diminished splenic conditioning and less RBC membrane loss, but not resolved.

14.(C). Hemolytic anemia may be associated with hereditary stomatocytosis, but splenectomy is not a recommended treatment. Persistent symptomatic thrombocytosis may follow splenectomy if the hemolysis is not eliminated or markedly decreased. Patients have developed a life-threatening tendency toward in situ thrombosis after splenectomy in association with the abnormal adherence of the stomatocytic RBC to vascular endothelium in conjunction with the thrombocytosis.

15.(D). Flow cytometry is the best diagnostic test for PNH. With the use of anti-CD59 for RBCs and anti-CD55 and anti-CD59 for granulocytes, flow cytometry is more sensitive than the classic RBC lysis tests in detecting the reduced glycolipid-bound membrane proteins.

16.(E). Renal function is progressively impaired by diffuse glomerular and tubular fibrosis leading to chronic, rather than acute, renal shutdown.

17.(D). There is an increased incidence of urinary tract infection with bacteremia in young women with sickle cell anemia.

18.(D). Blood transfusions usually do not reverse ischemic damage. Even while on chronic transfusion therapy children can still have transient cerebral ischemia as well as avascular necrosis of the hips and shoulders.

19.(A). Quantitative liver iron by approved MRI technology is the best indicator of total-body iron stores and should be obtained in chronically transfused patients after chronic transfusion therapy has initiated.

20.(C).

21.(D). Extramedullary hematopoiesis can occur in the vertebral canal, compressing the spinal cord and causing neurologic symptoms; the latter is a medical emergency requiring immediate local radiation therapy to halt erythropoiesis.

22.(C). Iron is initially deposited in the liver. Liver hemosiderosis develops after 1 yr of chronic transfusion therapy and is followed by iron deposition in the endocrine system. This leads to a high rate of hypothyroidism, hypogonadotropic gonadism, growth hormone deficiency, hypoparathyroidism, and diabetes mellitus. After 10 yr of transfusion, cardiac dysfunction secondary to hemosiderosis begins.

23.(B). Older children and adults have sufficient vitamin B12 stores to last 3-5 yr. However, in young infants born to mothers with low vitamin B12 stores (breast fed

infants of vegans mothers or having pernicious anemia), clinical signs of cobalamin deficiency can become apparent in the first 4-5 mo of life.

24.(D). The α -thalassemia traits present as a microcytic anemia that can be mistaken for iron-deficiency anemia. The hemoglobin electrophoresis is normal. The diagnosis is confirmed by DNA testing.

25.(A). In HIFD, symptoms become prominent at an early age (6-24 mo), consistent with exhaustion of vitamin B12 stores acquired in utero.

26.(E). Autohemolysis is moderately or markedly increased in pyruvate kinase deficiency, but the addition of glucose does not regularly correct the abnormality as it does in hereditary spherocytosis.

27.(E). Penicillin may cause hemolysis via the "hapten" mechanism (immune but not autoimmune) by binding tightly to the RBC membrane. Antibodies to the drug, either newly or previously formed, bind to the drug molecules on RBCs, mediating their destruction in the spleen.

28.(E). Splenectomy is not useful in cold agglutinin disease.

29.(A). Extensive burns may directly damage the RBCs and result in hemolysis that results in the formation of spherocytes. Blood loss and marrow suppression may contribute to anemia and require blood transfusion; others cause RBC destruction by mechanical injury.

30.(E). Secondary iron deficiency may complicate the intravascular hemolysis because of urinary hemoglobin and hemosiderin iron loss. Cardiovascular defects involving right-to-left shunts interfere with proper oxygenation leading to secondary polycythemia which may cause iron deficiency due to increased RBC demands production. INH therapy may cause secondary sideroblastic anemia with high serum iron and increased transferrin saturation.

31.(B). Parvovirus B19 is classically associated with isolated red cell aplasia, but in patients with sickle cell disease or immunodeficiencies it can result in transient pancytopenia.

32.(E). Transfusion of FFP is no longer recommended for treatment of patients with severe hemophilia A or B or for factor VII deficiency because safer factor VIII, IX, and VII concentrates are available; also it is not indicated for correction of hypovolemia or as immunoglobulin replacement therapy, because safer alternatives exist; An important use of FFP, albeit rare in children, is for rapid reversal of warfarin effects in patients who are actively bleeding or who require emergency surgery (i.e., in whom functional deficiencies of factors II, VII, IX, and X cannot be rapidly reversed by vitamin K).

33.(D). Transfusion of FFP is efficacious for the treatment of deficiencies of clotting factors II, V, X, and XI. Factor XIII and fibrinogen deficiencies are treated with cryoprecipitate. FFP also contains several anticoagulant proteins (antithrombin III, protein C, and protein S), whose deficiencies have been associated with thrombosis.

34.(D). In neonates, Clotting times are prolonged, owing to developmental deficiency of clotting proteins, and FFP should be transfused only after reference to normal values expected for the birthweight and age of the infant. The use of prophylactic FFP

transfusions to prevent intraventricular hemorrhage in premature infants is not recommended.

35.(C).

36.(E). CMV infections may be transmitted by leukocyte containing blood component transfusions; FFP does not transmit such infection because it is acellular component.

37.(E). Unlike the thrombin time, the reptilase time is not sensitive to heparin and is prolonged only by reduced or dysfunctional fibrinogen and fibrin split products.

38.(B).

39.(D). Deficiencies of the contact factors prolong the PTT but are not causes of clinical bleeding. These factors include factor XII, prekallikrein, and high molecular weight kininogen.

40.(A).

41.(D). Systemic amyloidosis may be associated with factor X deficiency owing to the adsorption of factor X on the amyloid protein. In the setting of amyloidosis, transfusion therapy is often not successful because of the rapid clearance of factor X.

42.(A). Factor V is lost rapidly from FFP. It is therefore important to use FFP that is less than 2 mo old. Because the plasma half-life of factor VII is 2-4 hr, therapy with FFP is difficult and often complicated by fluid overload.

43.(B). The usual screening tests for hemostasis are normal in patients with factor XIII deficiency, antiplasmin deficiency, and plasminogen activator inhibitor deficiency.

44.(E). DDAVP (0.3 µg/kg IV) has been found to be effective in shortening the bleeding time and has been used effectively to augment hemostasis prior to liver biopsy.

45.(A). Vitamin K-dependent clotting factors include factors II, VII, IX, X, protein C, and protein S.

46.(B). Von Willebrand disease (VWD) is the most likely cause for this case; menstruating female may present with iron deficiency due to menorrhagia. Because VWF is an acute-phase protein, stress will increase its level; thus, patients may not bleed during appendectomy and childbirth. Bruising symptoms may diminish during pregnancy, because the VWF levels may double or triple during pregnancy.

47.(B). Type 2N VWD is caused by the reduction of factor VIII binding by VWF. This disorder has also been termed autosomal hemophilia. With this variant, platelet interaction with VWF is normal but the 2N VWF binds weakly (or not at all) to factor VIII, resulting in rapid clearance of factor VIII that is weakly complexed to VWF.

48.(C). Individuals who are heterozygous have a 5-7-fold increase in risk of venous thrombosis

49.(E). Pulmonary embolism is a feature of venous thrombosis.

50.(B). The lupus anticoagulant causes a prolongation of the PTT that fails to correct on 1:1 mixing with normal plasma. The antibody is directed against the phospholipid used as a reagent in the PTT.

51.(D).

52.(C). Unlike UFH, which is monitored using the aPTT, LMWH is monitored via the anti-Xa activity.

53.(C). Heparin does not cross the placenta, and so it does not affect the baby. DIC causes prolonged PTT, PT, thrombocytopenia and low serum fibrinogen. Hemophilia causes prolonged PTT only. Lupus anticoagulants may cause thrombosis rather than bleeding and usually associated with prolonged PTT because of the presence of antiphospholipids that affect the phospholipid reagent.

54.(B). Normal hemoglobin electrophoresis usually excludes thalassemia and Diamond-Blackfan syndrome. Anemia due to parvovirus infection usually occurs in children with hemolytic anemia or immunodeficiency. In aplastic anemia, the anemia is usually macrocytic rather than normocytic.

55.(E). Although spleen may be palpable in 10% of children with ITP, bone marrow examination is mandatory when there is an unusual presentation.

56.(B). In Bernard-Soulier syndrome there is moderate thrombocytopenia and abnormal ristocetin test.

57.(E).

58.(A). In 70-80% of children who present with acute ITP, spontaneous resolution of their ITP will occur within 6 mo.

59.(E). Common drugs used in pediatrics that cause thrombocytopenia include valproic acid, phenytoin, sulfonamides, and trimethoprim-sulfamethoxazole. Heparin-induced thrombocytopenia is seldom seen in pediatrics.

60.(C). WAS is characterized by thrombocytopenia with tiny platelets, eczema, and recurrent infections due to immune deficiency.

61.(D). Thrombocytopenia of TAR syndrome frequently remits over the first few years of life.

62.(C). The role of IV anti-D in initial therapy of acute ITP is under investigation; the response appears to be somewhat slower than after IVIG; it is effective for Rh+ children; it can be used in acute or chronic ITP.

63.(C). β -thalassemia major is the most likely diagnosis; hemoglobin electrophoresis is quite helpful to confirm the diagnosis. Blood smear usually shows hypochromic microcytic RBCs but it is difficult to differentiate between iron deficiency anemia and thalassemia.

64.(E). This risk of sepsis is especially high in children younger than 5 yr at the time of surgery. The risk of sepsis is less after splenectomies performed for trauma, RBC membrane defects, and immune thrombocytopenia than when there is pre-existing immune deficiency (Wiskott-Aldrich syndrome, Hodgkin disease) or reticuloendothelial blockade (storage diseases, severe hemolytic anemias).

65.(C). A firm fixed node to the surrounding tissue should always raise the question of malignancy, regardless of the presence or absence of systemic symptoms or other abnormal physical findings. Fluctuance suggests abscess formation. Tuberculous nodes may be matted.

66.(E). The Reticulocyte production index (RPI, also called a corrected reticulocyte count) is a calculated value used in the diagnosis of anemia. This calculation is

necessary because the raw reticulocyte count is misleading in anemic patients. Reticulocyte index = reticulocyte count × (observed hematocrit/normal hematocrit).

67.(B). TAR is characterized by absent radii with presence of thumbs; this distinguishes from Fanconi anemia in which thumbs are absent.

68.(E).

69.(A).

70.(B). Bernard-Soulier Syndrome is characterized by moderate, rather than severe, thrombocytopenia.

71.(B). Aspirin may affect platelets function rather than the count.

72.(D). Severe life-threatening bleeding not responding to all modalities of medical treatment is an indication for emergent splenectomy.

73.(D). Splenomegaly is present in only 10% of cases of acute ITP.

74.(E). The most likely diagnosis is factor XIII deficiency.

75.(A). Iliopsoas bleeding is life threatening that needs immediate factor replacement therapy.

76.(B). When mild to moderate bleeding occurs, level of factor VIII or IX must be raised to a hemostatic level, in the range of 35-50%.

77.(B). In TAR, thrombocytopenia frequently remits but some cases may require allogenic bone marrow transplantation.

78.(C). Glanzmann's thrombasthenia is characterized by an abnormal platelets function and normal platelets count.

79.(E). Spontaneous recovery occurs in the majority of children with ITP; treatment is required when the platelets count is less than 20,000/ml.

80.(A).

1. Which of the following malignant tumors is least likely to occur in adults in comparison with children?

- A. acute Lymphoblastic Leukemia (ALL)
- B. osteosarcoma
- C. medulloblastoma
- D. retinoblastoma
- E. Hodgkin lymphoma

2. A 9-year-old boy develops acute myelogenous leukemia (AML) one year after completion of therapy for soft tissue sarcoma at his right thigh.

Which of the following chemotherapeutic agents is **MOST** likely the cause of secondary acute myelogenous leukemia AML in this boy?

- A. cyclophosphamide
- B. vincristine
- C. etoposide
- D. doxorubicin
- E. cisplatin

3. Of the following, the genetic syndrome **MOST** likely associated with increased risk of optic glioma is

- A. Down syndrome
- B. neurofibromatosis 1
- C. monosomy 7
- D. Bloom's syndrome
- E. ataxia-telangiectasia

4. Constellation of aniridia and hemihypertrophy is strongly associated with increased risk of which of the following tumors?

- A. rhabdomyosarcoma
- B. hepatoblastoma
- C. Wilms' tumor
- D. neuroblastoma
- E. medulloblastoma

5. During the routine exam of an infant, the parents state a strong family history of adenomatous polyposis.

The statement that should be included during the discussion is

- A. the infant is at increased risk for colonic adenocarcinoma
- B. the infant is at increased risk of acute lymphocytic leukemia
- C. the infant is at increased risk of intestinal Burkitt lymphoma
- D. the infant is at increased risk of hepatoblastoma
- E. the infant is at increased risk of germ cell tumor

6. Undescended testis is a risk factor for the development of which of the following tumors?

- A. rhabdomyosarcoma
- B. leukemia
- C. yolk sac tumor
- D. lymphoma
- E. neuroblastoma

7. Epstein-Barr virus (EBV) infection is more likely to be associated with all the following malignancies **EXCEPT**

- A. Burkitt lymphoma
- B. nasopharyngeal carcinoma
- C. T-cell lymphoma
- D. Hodgkin lymphoma
- E. hepatocellular carcinoma

8. Which of the following malignancies is least likely to occur in a 10-month-old infant?

- A. neuroblastoma
- B. nephroblastoma
- C. retinoblastoma
- D. hepatoblastoma
- E. Ewing sarcoma

9. Hepatitis C virus infection is a risk factor for which of the following malignancy?

- A. hepatoblastoma
- B. splenic lymphoma
- C. Hodgkin lymphoma
- D. nasopharyngeal carcinoma
- E. cervical carcinoma

10. Nearly all cervical carcinomas contain human papillomavirus (HPV). Which of the following types is never associated with such malignancy?

- A. type 6
- B. type 16
- C. type 18

- D. type 31
- E. type 33

11. Which of the following chromosomal abnormalities in acute lymphoblastic leukemia of childhood carry a favorable outcome?

- A. t(12;21)
- B. t(4;11)
- C. t(9;22)
- D. hypodiploidy
- E. 11q23

12. You are meeting with parents of 10-year-old child who recently develops acute lymphoblastic leukemia (ALL); the mother has a concern about risk of CNS relapse.

Which of the following is **LEAST** likely to increase the risk of CNS relapse in children with ALL?

- A. first traumatic lumbar puncture (LP)
- B. T-cell leukemia
- C. cranial nerve involvement at the time of diagnosis
- D. presence of lymphoblast in the CSF at any time during treatment
- E. age of more than 10 year at the time of the diagnosis

13. Children with ALL who carry poor outcome include all the following **EXCEPT**

- A. age younger than 1 year and older than 10 year
- B. T-cell immunophenotype
- C. hyperdiploidy chromosomal abnormality
- D. initial leukocyte count of > 50,000
- E. poor response to initial therapy

14. Which of the following chromosomal abnormalities of childhood ALL carries the highest risk of relapse despite intensive chemotherapy?

- A. t(9;22)
- B. t(4;11)
- C. hypodiploidy
- D. t(1;19)
- E. t(12;21)

15. You are meeting the parents of a child with ALL who want to go for a trip with their child. The **MOST** nontoxic phase of the treatment of ALL that allow the family to have a safe trip is

- A. before starting treatment
- B. remission induction
- C. consolidation

- D. delayed intensification
- E. interim maintenance

16. You are evaluating a 6-year-old child with ALL on interim maintenance phase who has frequent mucositis and myelosuppression that needs frequent discontinuation of his treatment.

Of the following, the **MOST** valuable test for this child is

- A. complete blood count
- B. bone marrow study
- C. pharmacogenetic testing of the thiopurine s-methyltransferase (tpmt) gene
- D. lumbar puncture
- E. renal function test

17. You are evaluating a 9-year-old boy child with ALL who recently develops testicular relapse; an important statement that should be mentioned to his parents is

- A. testicular relapse occurs in the majority of boys with ALL
- B. it usually occurs during the course of therapy
- C. such relapse occurs as painful swelling of one or both testes
- D. the diagnosis is confirmed by ultrasonography of the affected testis
- E. the majority of affected boys can be successfully retreated, and the survival rate is good

18. Granulocytic sarcoma masses are commonly seen in which type of the following malignancies?

- A. neuroblastoma
- B. rhabdomyosarcoma
- C. soft tissue sarcoma
- D. ALL
- E. AML

19. Which of the following types of translocation of childhood AML that typically associated with granulocytic sarcoma mass?

- A. t(15:17)
- B. t(8:21)
- C. inv(16)
- D. t(6:9)
- E. inv(3)

20. You are explaining the risk of leukemia in children with Down syndrome to medical students; your discussion will include all the following statements **EXCEPT**

- A. acute leukemia occurs more frequently in children with Down syndrome than in the general population

- B. AML is more common in children with Down syndrome in the 1st 3 yr of life than ALL as compared with the ratio of ALL/AML in general population
- C. in children with Down syndrome who have AML, the expected outcome of treatment is slightly inferior to that for other children
- D. patients with Down syndrome who develops acute leukemia demonstrate a remarkable sensitivity to antimetabolites
- E. neonates with Down syndrome may develop a transient leukemia or myeloproliferative disorder

21. A healthy 20-day-old male neonate with Down syndrome appears pale; examination reveals a palpable liver 6 cm below the right costal margin and palpable spleen 3 cm below the left costal margin; lab findings include: hemoglobin, 8.8 g/dl; platelets count, 55000/mm³; white blood cell count, 18700/mm³ with 10% blast cells; bone marrow examination is consistent with acute leukemia.

Of the following, the **BEST** approach for the management is

- A. intensive chemotherapy
- B. low dose chemotherapy
- C. pulses of chemotherapy
- D. bone marrow transplantation
- E. close follow up

22. Which of the following types of leukemia is **MOST** likely to develop in a 2-year-old child with Down syndrome who has been developed transient myeloproliferative disorder in the neonatal period?

- A. ALL
- B. CML
- C. AML M1
- D. AML M6
- E. AML M7

23. Which of the following chromosomal abnormalities is often characteristic of infantile ALL?

- A. t(4;11)
- B. t(12;21)
- C. t(9;22)
- D. t(10;14)
- E. t(1;19)

24. Hodgkin lymphoma in children usually has favorable outcome; however, the prognosis may be grim in some cases.

Poor prognostic factors in Hodgkin Lymphoma include all the following **EXCEPT**

- A. age of more than 15 year at the time of the diagnosis

- B. stage IV disease
- C. presence of "B" symptoms
- D. poor response to therapy manifested by positron emission tomography (PET) scan positivity
- E. presence of bulky mediastinal mass

25. A 12-year-old male adolescent, recently diagnosed with Hodgkin lymphoma; the PET-CT scan shows an involvement of the left cervical and supraclavicular lymph nodes; he has a history of drenching night sweating and high grade fever.

Based on Ann Arbor Staging Classification for Hodgkin Lymphoma, the patient is classified as

- A. stage IA
- B. stage IB
- C. stage IIA
- D. stage IIB
- E. stage IIIB

26. Which of the following manifestations is more specific to occur in anaplastic large cell lymphoma (ALCL) than other types of non-Hodgkin Lymphoma (NHL)?

- A. primary intestinal involvement
- B. bone marrow involvement
- C. CNS involvement
- D. primary cutaneous involvement
- E. primary mediastinal involvement

27. Tumor lysis syndrome (TLS) is a common complication during the treatment of pediatric malignancies, which of the following malignancies is more likely to manifest such complication?

- A. acute myelogenous leukemia AML
- B. nephroblastoma
- C. neuroblastoma
- D. Burkitt lymphoma
- E. Rhabdomyosarcoma

28. Metabolic derangement secondary to tumor lysis syndrome is commonly encountered after starting chemotherapy in children with malignancy; it include all the following **EXCEPT**

- A. hyperuricemia
- B. hypernatremia
- C. hyperkalemia
- D. hyperphosphatemia
- E. hypocalcemia

29. A 5-year-old child, recently diagnosed with ileocecal Burkitt lymphoma; lab investigations reveal: serum uric acid 12 mg/dl, serum sodium, 145 meq/dl; serum potassium, 4.5 meq/dl; serum phosphate 4.4 meq/dl; serum calcium, 8.9 mg/dl; blood urea, 22 mg/dl; serum creatinine, 0.8 mg/dl.

Of the following, the **MOST** effective treatment is

- A. excessive hydration
- B. sodium bicarbonate
- C. xanthine oxidase inhibitor
- D. recombinant urate oxidase
- E. no treatment

30. A 6-year-old male child presented with a cerebellar mass; after undergoing complete surgical resection of the mass, the histology of the mass reveals pilocytic astrocytoma.

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

- A. radiotherapy
- B. chemotherapy
- C. observation
- D. concomitant chemo-radiotherapy
- E. radiotherapy followed by chemotherapy

31. A 6-year-old child complains of bilateral thigh pain, motor weakness and some sensory deficits of both lower limbs; he has a history of bladder dysfunction over the past 2 weeks; MRI of the spinal cord shows a mass arising from the filum terminale and conus medullaris causing some pressure effect. A CNS tumor is suspected.

Which of the following tumor is **MOST** likely arising in such site?

- A. medulloblastoma
- B. anaplastic astrocytoma
- C. myxopapillary ependymoma
- D. choroid plexus carcinoma
- E. oligodendroglioma

32. You are discussing the risk of radiotherapy with the parents of a child with medulloblastoma; the mother has a concern about the late neurological complications post radiotherapy.

The statement that should be included in the discussion that late neurological sequelae post radiotherapy is more severe with

- A. focal radiotherapy rather than craniospinal irradiation
- B. children with an age of less than 3 year
- C. concomitant chemo-radiotherapy
- D. low grade tumors rather than high grade tumors
- E. infratentorial tumors rather than supratentorial tumors

33. The least common late neurologic sequelae that may be encountered post craniospinal irradiation for a 9-year-old child with medulloblastoma is

- A. microcephaly
- B. learning disabilities
- C. cognitive impairment
- D. neuroendocrine dysfunction
- E. second malignancy

34. Chemotherapy has a major role in many childhood CNS tumors. In which of the following tumors chemotherapy is not effective?

- A. medulloblastoma
- B. pilocytic astrocytoma
- C. craniopharyngioma
- D. pineoblastoma
- E. supratentorial primitive neuroectodermal tumors (SPNETs)

35. Childhood primary brain stem tumors are heterogenous group of tumors; the outcome usually depends on tumor location.

Which of the following brain stem tumors, depending on the site of tumor, carry the worst prognosis?

- A. focal
- B. dorsally exophytic
- C. cervicomedullary
- D. diffuse intrinsic
- E. none of the above

36. Extraneural metastasis from primary brain tumors is less likely to occur. Which of the following primary brain tumor is **MOST** commonly metastasizing extraneurally?

- A. medulloblastoma
- B. primitive neuroectodermal tumor (PNET)
- C. ependymoma
- D. malignant glioma
- E. choroid plexus carcinoma

37. Childhood primary brain tumors are less likely to metastasize extraneurally. Which of the following is **MOST** likely considered a risk factor for extraneural metastasis?

- A. supratentorial tumor
- B. age of the patient of less than 10 year
- C. female gender
- D. ventriculoperitoneal (VP) shunt insertion
- E. glial tumors

38. Small round blue cell tumors refer to tumors that have round blue cells histologically. They include all the following **EXCEPT**

- A. neuroblastoma
- B. non-Hodgkin lymphoma
- C. osteosarcoma
- D. Ewing sarcoma
- E. rhabdomyosarcoma

39. A 9-mo-old infant develops a left adrenal mass; histological examination reveals a significant degree of undifferentiated round blue cells consistent with neuroblastoma. Which of the following genetic characteristics of the tumor cells carry a better outcome?

- A. amplification of the MYCN (N-myc) proto-oncogene
- B. hyperdiploidy
- C. loss of heterozygosity of 1p chromosome
- D. loss of heterozygosity of 11q chromosome
- E. gain of 17q chromosome

40. A 1-year-old child develops right eye ptosis, myosis, and loss of sweating; you suspect neuroblastoma.

Of the following, the **MOST** valuable investigation to confirm the diagnosis is

- A. Computerized tomography (CT) scan of the neck and chest
- B. CT scan of the abdomen
- C. CT scan of the brain
- D. magnetic resonance imaging (MRI) of the brain
- E. X-ray the chest

41. Neuroblastoma is a highly malignant childhood neoplasm. Metastatic spread can occur via local invasion or distant hematogenous or lymphatic routes.

Of the following, the **LEAST** common site of metastases in neuroblastoma is

- A. long bones
- B. bone marrow
- C. lung
- D. skin
- E. liver

42. Neuroblastoma can be associated with paraneoplastic syndrome. All the following are paraneoplastic features of neuroblastoma **EXCEPT**

- A. uncontrollable jerking eye and body movements
- B. cerebellar ataxia and poor coordination
- C. increased sweating and hypertension
- D. unilateral ptosis, myosis, and anhidrosis

- E. profound secretory diarrhea

43. You are evaluating a 6-mo-old girl with a firm abdominal mass. Radiography reveals a calcified small right suprarenal mass. The tumor is completely resected and histologically is confirmed as neuroblastoma. Metastatic work-up shows massive liver involvement, 10% bone marrow involvement by tumor cells, and multiple subcutaneous nodules involvement. There is no bony involvement. The *N-myc* oncogene from the tumor is not amplified.

According to the international neuroblastoma staging system, the infant is stratified as

- A. stage I
- B. stage II A
- C. stage III
- D. stage IV
- E. stage IV S

44. A 10-mo-old boy has a left suprarenal mass. Surgery is accomplished with complete surgical removal of the mass as well as the non-adherent ipsilateral lymph nodes; multiple contralateral lymph nodes biopsies are taken during surgery. The histology reveals poorly differentiated neuroblastoma with microscopic involvement of the ipsilateral lymph nodes; the contralateral lymph nodes are negative for tumor cells.

Of the following, the **BEST** therapeutic approach for this infant is

- A. chemotherapy
- B. radiotherapy
- C. concomitant chemo-radiotherapy
- D. chemotherapy followed by radiotherapy
- E. close observation

45. Children with high-risk neuroblastoma have poor survival. Current treatment consists of all the following **EXCEPT**

- A. surgery
- B. intensive chemotherapy
- C. radiation
- D. allogenic bone marrow transplantation
- E. differentiating agents (13-cis-retinoic acid)

46. A 2-year-old male child has an asymptomatic right flank mass discovered incidentally by the mother while bathing. Suspicion of Wilms' tumor is raised.

Of the following, the **LEAST** likely investigation to be performed in this child is

- A. plain abdominal radiography
- B. computerized tomography (CT) scan of the abdomen
- C. CT scan of the chest
- D. Biopsy of the mass

- E. ultrasonography with Doppler imaging of the abdominal blood vessels

47. Biopsy for children with Wilms` tumor is not usually performed as it may upstage the disease.

All the following are indications of accomplishing renal biopsy for a child with suspicion of Wilms` tumor **EXCEPT**

- A. age of 2-3 year at presentation
- B. signs of infection and inflammation around the tumor
- C. significant abdominal lymph nodes enlargement radiologically
- D. absence of renal parenchyma radiologically
- E. radiological appearance of intratumoral calcification

48. A 2-year-old child is being evaluated for a right flank mass; radiological appearance is consistent with right renal Wilms` tumor. During radical nephrectomy there is intra-abdominal tumor rupture stated by the surgeon.

Of the following, the **BEST** therapeutic approach for this child is

- A. chemotherapy
- B. radiotherapy
- C. concomitant chemotherapy-radiotherapy
- D. another surgery
- E. observation

49. Although children with Wilms` tumor have a favorable prognosis, there are some adverse factors that affect outcome.

All the following are poor prognostic factors in children with Wilms` tumor **EXCEPT**

- A. young age at diagnosis
- B. advanced stage
- C. large tumors
- D. anaplastic histology
- E. loss of heterozygosity at chromosome 1p and 16q

50. Although it is not common, about 15% of children with Wilms` tumor suffer relapse, most relapses occur early (within 2 yr of diagnosis).

Factors associated with a favorable outcome after relapse include all the following **EXCEPT**

- A. low stage (I/II) at diagnosis
- B. no prior radiotherapy
- C. anaplastic histology
- D. relapse to lung only
- E. late relapse more than 12 mo from nephrectomy

51. A 2-mo-old male infant has a left flank mass discovered incidentally by the mother; radiological imaging reveals a left renal mass.

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

- A. nephroblastoma
- B. neuroblastoma
- C. mesoblastic nephroma
- D. clear cell sarcoma
- E. rhabdoid tumor of the kidney

52. A healthy 4-year-old child has a left flank mass. Computerized tomography (CT) scan of the abdomen shows a localized left renal mass. Radical nephrectomy is performed; the histology shows clear cell sarcoma of the kidney.

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

- A. CT scan of the chest
- B. CT scan of the brain
- C. bone scan
- D. magnetic resonance imaging (mri) of the brain
- E. skeletal survey

53. Renal cell carcinoma (RCC) is rare in children, accounting for <5% of all renal tumors of childhood.

All the following are true regarding RCC in children **EXCEPT**

- A. patients may present with frank hematuria, flank pain, and/or a palpable mass
- B. it can be asymptomatic and detected incidentally
- C. it has a propensity to metastasize to the lungs, bone, liver, and brain
- D. it can be associated with von hippel–lindau disease
- E. local lymph node involvement has an adverse outcome

54. Alveolar type of rhabdomyosarcoma (RMS) accounts for approximately 1/3 of all cases of pediatric RMS and carries the poorest prognosis.

Of the following, the **MOST** common site of involvement by alveolar type RMS is

- A. orbit
- B. middle ear
- C. extremities
- D. vagina
- E. bladder

55. In pediatric rhabdomyosarcoma, stages are dependent on primary site whether favorable or unfavorable.

Which of the following sites of involvement is considered unfavorable?

- A. vagina
- B. uterus

- C. testis
- D. orbit
- E. paranasal sinuses

56. Osteosarcoma is the most common primary malignant bone tumor in children and adolescents; it has multiple subtypes and requires different modalities of treatment including surgery and chemotherapy.

Of the following, the subtype of osteosarcoma which is treated by surgery alone is

- A. fibroblastic
- B. chondroblastic
- C. telangiectatic
- D. parosteal
- E. periosteal

57. Of the following, the **WORST** prognostic factor in pediatric osteosarcoma is

- A. primary pelvic bone tumor
- B. poor histologic response to treatment
- C. bony metastases at the time of diagnosis
- D. lung metastases at the time of diagnosis
- E. chondroblastic subtype

58. Radiotherapy is an effective modality of treatment in variable pediatric solid tumors.

Of the following, the **LEAST** responsive tumor to radiotherapy is

- A. rhabdomyosarcoma
- B. neuroblastoma
- C. nephroblastoma
- D. osteosarcoma
- E. Ewing sarcoma

59. A 12-year-old male adolescent has pain of the right upper thigh that gradually becomes more severe and most often at night; it usually relieved by taking salicylates medication. Examination reveals limping, atrophy, and weakness of the right lower extremity. Plain radiography shows a round lucent lesion at the diaphysis of the right upper femur, about 0.5 cm in diameter, surrounded by sclerotic cortical bone formation.

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

- A. Ewing sarcoma
- B. osteosarcoma
- C. osteoid osteoma
- D. osteoblastoma
- E. nonossifying fibroma

60. During a routine examination of a 10-mo-old male infant, you find a white pupillary reflex of right eye; the eye movements are normal. You suspect retinoblastoma.

Of the following, the **BEST** confirmatory diagnostic evaluation of this infant is

- A. indirect ophthalmoscopy with slit-lamp examination
- B. examination under general anesthesia by an experienced ophthalmologist
- C. retinal biopsy
- D. orbital ultrasonography
- E. brain MRI

61. A 9-mo-old infant is recently diagnosed with bilateral retinoblastoma; examination under anesthesia reveals bilateral multifocal involvement of the retina. An important step in the management is

- A. radiotherapy of both eyes
- B. retinal examination of 1st degree relatives
- C. orbital ultrasonography
- D. bilateral bone marrow examination
- E. bilateral enucleation

62. The serum α -fetoprotein (AFP) level is elevated with some malignant germ cell tumors (GCTs) especially endodermal sinus tumors; it can be used as a measure of response to treatment and during follow up after completion of chemotherapy. However, it is normally elevated during infancy.

At which age does AFP physiologically fall to normal adult level?

- A. three month
- B. eight month
- C. one year
- D. three year
- E. ten year

63. The β subunit of human chorionic gonadotropin (HCG) is a useful surface marker in some malignant germ cell tumors (GCTs); it is secreted by syncytiotrophoblasts.

In which of following GCTs this surface marker is characteristically elevated?

- A. teratoma
- B. endodermal sinus tumor
- C. germinoma
- D. gonadoblastoma
- E. embryonal carcinoma

64. You are discussing with medical students the role of chemotherapy in malignant germ cell tumors (GCTs); you state that GCTs are sensitive to some types of chemotherapy.

Of the following, the **MOST** effective chemotherapeutic agent in GCTs is

- A. vincristine
- B. cyclophosphamide
- C. cisplatin
- D. methotrexate
- E. cytosine Arabinoside

65. Hepatoblastoma occurs predominantly in children younger than 3 yr of age. Although etiology is unknown, there are many associated risk factors.

All the following are associated risk factors for development of hepatoblastoma **EXCEPT**

- A. familial adenomatous polyposis
- B. Beckwith-Wiedemann syndrome
- C. prematurity
- D. low birth weight
- E. hepatitis C infection

66. Hepatoblastoma is the most common neoplasm of the liver; it arises from the precursors of hepatocytes. It is of different histological classification which has a direct correlation with clinical outcome.

Of the following, the histological type that predict the **MOST** favorable outcome is

- A. epithelial type of pure fetal histology
- B. epithelial type of pure embryonal histology
- C. epithelial type of mixed histology (fetal and embryonal)
- D. mixed type of epithelial and mesenchymal elements
- E. undifferentiated type

67. Hemangiomas are the most common benign tumors of infancy, occurring more often in the head and neck region.

Of the following, the infant at higher risk of development of hemangioma is

- A. full term infant
- B. male gender infant
- C. infant of woman who had chorionic villus sampling
- D. infant delivered by cesarean section
- E. infant of diabetic mother

68. You are examining an infant with multiple cutaneous hemangiomas; you suspect an involvement of internal organs.

Of the following, the **MOST** common site of visceral involvement by hemangiomas is

- A. brain
- B. heart
- C. lung
- D. liver
- E. pancreas

69. Kasabach-Merritt syndrome is a benign vascular tumor which may be life-threatening; it is characterized by all the following **EXCEPT**

- A. rapidly enlarging lesion
- B. thrombocytopenia
- C. microangiopathic hemolytic anemia
- D. coagulopathy
- E. association with infantile hemangiomas

70. A 3-day-old neonate has a large soft, painless mass involving the right lower part of the head and neck that transilluminate; computerized tomography reveals a cystic mass infiltrating the neck and intrathoracic region mostly the mediastinum.

Of the following, the **BEST** modality of treatment for this neonate is

- A. aspiration of mass
- B. surgical resection
- C. Injection of sclerosing agent
- D. laser therapy
- E. systemic interferon

71. You are meeting with parents of 12-year-old girl who recently diagnosed with papillary thyroid carcinoma (PTC). The statement that should be included in your discussion is

- A. PTC has a grim overall prognosis
- B. chemotherapy is the primary therapy
- C. children with PTC do not require radioactive iodine therapy (^{131}I)
- D. supraphysiologic levothyroxine is required during the course of treatment
- E. long-term follow-up involves monitoring of calcitonin/carcinogenic embryonic antigen tumor markers

72. You are examining a 12-year-old female adolescent with a small nevus in the left thigh; her mother is concerned regarding development of melanoma in future.

All the following findings raise the suspicion of melanoma **EXCEPT**

- A. rapidly enlarging nevus
- B. nevus with changing colors
- C. nevus with irregular margins
- D. easily bleeds nevus
- E. Spitz nevus

73. Although melanoma is relatively rare in children, some risk factors may increase its incidence.

All the following are risk factors for development of melanoma **EXCEPT**

- A. positive family history of melanoma
- B. dark skinned child

- C. giant hairy nevus
- D. dysplastic nevus syndrome
- E. xeroderma pigmentosum

74. Nasopharyngeal carcinoma is one of the most common nasopharyngeal tumors in pediatric patients. Many factors may affect the prognosis.

Which of the following carries the worst outcome?

- A. elevated lactate dehydrogenase (LDH) level
- B. advanced local disease
- C. extensive cervical lymph nodes involvement
- D. Epstein-Barr virus (EBV) DNA levels
- E. evidence of distant metastases

75. You are evaluating a 6-year-old male child with Langerhans cell histiocytosis (LCH); you states to the parents that skeletal survey and bone scan are important investigations as bones are the most common sites of involvement.

Of the following, the **MOST** common site of bone involvement by LCH is

- A. skull
- B. mandible
- C. vertebra
- D. pelvis
- E. femur

76. Langerhans cell histiocytosis (LCH) has an extremely variable presentation. Single or multiple organs may be affected by the disease.

Of the following, the **LEAST** likely organ affected by LCH is

- A. skin
- B. bone
- C. middle ear
- D. lung
- E. lymph nodes

77. In Langerhans cell histiocytosis (LCH), systemic manifestations are more likely to occur in patients with multisystem disease who are at high risk of mortality (i.e., "risk organ"-positive patients). All the following are risk organs in LCH **EXCEPT**

- A. bone
- B. liver
- C. spleen
- D. lung
- E. hematopoietic system

- 1.(D). Retinoblastoma is an embryonal tumor, usually occurs in childhood.
- 2.(C). Epipodophyllotoxins like etoposide may cause secondary AML early within two years after administration; alkylating agents like cyclophosphamide may cause secondary AML as far as 5 years after therapy.
- 3.(B). Neurofibromatosis 1 is strongly associated with optic glioma. Down syndrome and Monosomy 7 are associated with increased risk of AML. Bloom's syndrome and ataxia-telangiectasia are associated with increased risk of ALL.
- 4.(C). Beckwith-Wiedemann syndrome, an overgrowth syndrome characterized by macrosomia, macroglossia, hemihypertrophy, omphalocele, and renal anomalies, is associated with an increased risk of Wilms` tumor, hepatoblastoma, rhabdomyosarcoma, neuroblastoma, and adrenal cortical carcinoma, while aniridia is associated with Wilms` tumor.
- 5.(D). Beckwith-Wiedemann syndrome, hemihypertrophy, Gardner syndrome, and family history of adenomatous polyposis are associated with increased risk of hepatoblastoma.
- 6.(C).
- 7.(E). Hepatocellular carcinoma is associated with hepatitis B and C infection.
- 8.(E). During the 1st yr of life, embryonal tumors such as neuroblastoma, nephroblastoma (Wilms tumor), retinoblastoma, rhabdomyosarcoma, hepatoblastoma, and medulloblastoma are most common. As children age, bone malignancies, Hodgkin disease, gonadal germ cell malignancies (testicular and ovarian carcinomas), and other carcinomas increase in incidence.
- 9.(B). Hepatitis C virus infection is a risk factor for hepatocellular carcinoma and is also associated with splenic lymphoma.
- 10.(A). High-risk HPVs include types 16 and 18 but also types 31, 33, 35, 45, and 56, which are also commonly found in women without lesions. The low-risk HPVs, including 6 and 11 that are commonly found in genital warts, are almost never associated with malignancies.
- 11.(A).
- 12.(E).
- 13.(C). More favorable characteristics include a rapid response to therapy, hyperdiploidy, trisomy of specific chromosomes (4, 10, and 17), and rearrangements of the ETV6-RUNX1 (formerly TEL-AML1) genes.
- 14.(B). Infants with ALL, along with patients who present with specific chromosomal abnormalities, such as t(4; 11), have an even higher risk of relapse despite intensive therapy. However, the poor outcome of Philadelphia chromosome positive ALL with

t(9;22) has dramatically changed by the addition of imatinib to an intensive chemotherapy backbone.

15.(E).

16.(C). Pharmacogenetic testing of the thiopurine S-methyltransferase (TPMT) gene, which encodes one of the metabolizing enzymes of mercaptopurine, can identify patients who are wild type (normal TPMT enzyme activity), heterozygous (slightly decreased TPMT enzyme activity), or homozygous (low or absent enzyme activity). Decreased TPMT enzyme activity results in an accumulation of a toxic metabolite of mercaptopurine and results in severe myelosuppression, requiring dose reductions of the chemotherapy.

17.(E). Testicular relapse occurs in less than 2% of boys with ALL, usually after completion of therapy. Such relapse occurs as painless swelling of one or both testes. The diagnosis is confirmed by biopsy of the affected testis. Treatment includes systemic chemotherapy and possibly local irradiation. A high proportion of boys with a testicular relapse can be successfully retreated, and the survival rate of these patients is good.

18.(E).

19.(B). Discrete masses, known as chloromas or granulocytic sarcomas, can occur in the absence of apparent bone marrow involvement and typically are associated with a t(8;21) translocation. Chloromas also may be seen in the orbit and epidural space.

20.(C). In AML, patients with Down syndrome have much better outcomes than non-Down syndrome children, with a >80% long-term survival rate.

21.(E). Approximately 10% of neonates with Down syndrome develop a transient leukemia or myeloproliferative disorder which usually resolves within the 1st 3 mo of life. Although these neonates can require temporary transfusion support, they do not require chemotherapy unless there is evidence of life-threatening complications. However, those patients require close follow-up, because 20-30% will develop typical leukemia by 3 yr of life.

22.(E). Twenty-thirty percent of patients who have Down syndrome and who develop transient leukemia or myeloproliferative disorder will develop typical leukemia (often acute megakaryocytic leukemia) by 3 yr of life (mean onset, 16 mo).

23.(A). More than 80% of the cases of infantile ALL demonstrate rearrangements of the MLL gene, found at the site of the 11q23 band translocation, the majority of which are the t(4;11). This subset of patients largely accounts for the very high relapse rate.

24.(A).

25.(D). Stage II includes involvement of 2 or more lymph node regions on the same side of the diaphragm (II) or localized involvement of an extralymphatic organ or site and 1 or more lymph node regions on the same side of the diaphragm (IIE). HL is subclassified into A or B categories: A is used to identify asymptomatic patients and B is for patients who exhibit any B symptoms.

26.(D). The primary site of tumor involvement and metastasis pattern varies by pathologic subtype. Lymphoblastic lymphoma (LBL) commonly manifests as an intrathoracic or mediastinal supradiaphragmatic mass and also has a predilection for

spreading to the bone marrow and CNS. Burkitt lymphoma (BL) commonly manifests as abdominal (sporadic type) or head and neck (endemic type) tumor and can metastasize to the bone marrow or CNS. Diffuse large B-cell lymphoma (DLBCL) commonly manifests as either an abdominal or mediastinal primary and, rarely, disseminates to the bone marrow or CNS. ALCL manifests either as a primary cutaneous manifestation (10%) or as systemic disease (90%) with dissemination to liver, spleen, lung, or mediastinum. Bone marrow or CNS disease is rare in ALCL.

27.(D). TLS can occur from rapid cell turnover, which is especially common in Burkitt lymphoma.

28.(B).

29.(D). Recombinant urate oxidase is preferred in patients with a high risk of tumor lysis. Frequently, only a single dose is needed; however, repeat doses can be given if a subsequent rise in uric acid is seen.

30.(C). With complete surgical resection the overall survival approaches 80-100%.

31.(C). Myxopapillary ependymoma (WHO grade I) is a slow-growing tumor arising from the filum terminale and conus medullaris; it carries good prognosis.

32.(B).

33.(A). Craniospinal radiation in children younger than 3 yr of age results in severe late neurologic sequelae, including microcephaly, learning disabilities, cognitive impairment, and neuroendocrine dysfunction. Similarly, in older children, late sequelae, such as learning disabilities, neuroendocrine dysfunction, and/or second malignancies, can occur.

34.(C). There is no role for chemotherapy in craniopharyngioma.

35.(D). Surgical resection is the primary treatment approach for focal and dorsally exophytic tumors and leads to a favorable outcome. Histologically, these 2 groups usually are low-grade gliomas. Cervicomedullary tumors, owing to their location, may not be amenable to surgical resection but are sensitive to radiation therapy. Diffuse intrinsic tumors, characterized by the diffuse infiltrating pontine glioma, are associated with a very poor outcome independent of histologic diagnosis.

36.(A).

37.(D). Ventriculoperitoneal shunts have been known to allow extraneural metastases, primarily within the peritoneal cavity but also systemically.

38.(C).

39.(B). Hyperdiploidy confers better prognosis if the child is younger than 1 yr of age at diagnosis. Amplification of MYCN is strongly associated with advanced tumor stage and poor outcomes. Other chromosomal abnormalities, including loss of heterozygosity of 1p, 11q, and 14q, and gain of 17q, are commonly found in neuroblastoma tumors and are also associated with worse outcomes.

40.(A). Horner syndrome, characterized by unilateral ptosis, myosis, and anhidrosis, is associated with a thoracic or cervical primary tumor (e.g., superior cervical ganglion). Symptoms do not resolve with tumor resection.

41. **(C).** The most common sites of metastasis are the regional or distant lymph nodes, long bones and skull, bone marrow, liver, and skin. Lung and brain metastases are rare, occurring in >3% of cases.
42. **(D).** Horner syndrome results from direct invasion of the neural foramina by tumor, causing nerve root compression. opsoclonus–myoclonus–ataxia syndrome is of paraneoplastic nature of autoimmune origin. Neuroblastoma can produce catecholamines that can cause increased sweating and hypertension; it may release vasoactive intestinal peptide, causing a profound secretory diarrhea.
43. **(E).** Infants younger than 1 yr of age also can present in unique fashion, termed stage 4S, with widespread subcutaneous tumor nodules, massive liver involvement, limited bone marrow disease, and a small primary tumor without bone involvement or other metastases.
44. **(E).** The infant has stage 2A and is stratified as low risk. The usual treatment for children with low-risk neuroblastoma is surgery for stages 1 and 2 and observation for stage 4S with cure rates generally >90% without further therapy.
45. **(D).** High-dose chemotherapy with autologous stem cell rescue, and not allogeneic bone marrow transplantation, is one of the treatment modality for high risk patients.
46. **(D).** Although biopsy is a reliable diagnostic tool, it is discouraged as it results in disease upstaging. A core needle biopsy obtained via a posterior approach should be performed in cases of unusual presentation (older age, signs of infection, inflammation) or unusual imaging findings (significant adenopathy, no renal parenchyma seen, intratumoral calcification).
47. **(A).** Older age, more than 5 years, is an indication to perform renal biopsy.
48. **(C).** The child is stratified as stage III; so he needs chemo-radiotherapy.
49. **(A).** Prognostic factors for risk-adapted therapy include age, stage, tumor weight, and loss of heterozygosity at chromosomes 1p and 16q. Histology plays a major role in risk stratification of WT. Absence of anaplasia is considered a favorable histologic finding.
50. **(C).** Factors associated with a favorable outcome after relapse include low stage (I/II) at diagnosis, treatment with vincristine and actinomycin D only, no prior radiotherapy, favorable histology, relapse to lung only, and interval from nephrectomy to relapse 12 mo or longer.
51. **(C).** Mesoblastic nephroma is the most common solid renal tumor identified in the neonatal period. Most of the patients are diagnosed before 3 mo of age, whereas Wilms' tumor is rarely diagnosed before 6 mo of age. Radical nephrectomy is the treatment of choice and may be sufficient by itself.
52. **(C).** Bone is the most common site of distant metastasis in clear cell sarcoma of the kidney followed by lung, abdomen, retroperitoneum, brain, and liver.
53. **(E).** Unlike the case for adult RCC, local lymph node involvement is not a poor prognostic indicator in pediatric RCC. Nephrectomy alone may be adequate for early-stage RCC.
54. **(C).** Alveolar tumors occur most often in the trunk and extremities.

- 55.(E). Favorable sites include female genital, paratesticular, and head and neck (nonparameningeal) regions; all other sites are considered unfavorable. Involvement of paranasal sinuses by rhabdomyosarcoma is considered as unfavorable site.
- 56.(D). Parosteal osteosarcoma is a low-grade, well-differentiated tumor that does not invade the medullary cavity and most commonly is found in the posterior aspect of the distal femur. Surgical resection alone often is curative in this lesion, which has a low propensity for metastatic spread.
- 57.(C). All the mentioned factors carry poor prognosis, but multiple bony metastases at the time of diagnosis is the worst.
- 58.(D). Osteosarcoma is a radio-resistant tumor.
- 59.(C). Osteoid osteomas are diagnosed between 5 and 20 yr of age. The clinical pattern is characteristic, consisting of unremitting and gradually increasing pain that often is worst at night and is relieved by aspirin. Boys are more affected than girls. Radiographs are distinctive, showing a round or oval metaphyseal or diaphyseal lucency (nidus) surrounded by sclerotic bone.
- 60.(B). Imaging studies are not diagnostic, and biopsies are contraindicated. Indirect ophthalmoscopy with slit-lamp evaluation can detect retinoblastoma tumors, but a complete evaluation requires an examination under general anesthesia by an experienced ophthalmologist to obtain complete visualization of both eyes, which also facilitates photographing and mapping of the tumors.
- 61.(B). Retinoblastoma can be either hereditary or sporadic. Hereditary cases usually are diagnosed at a younger age and are multifocal and bilateral, whereas sporadic cases are usually diagnosed in older children who tend to have unilateral, unifocal involvement. All 1st-degree relatives of children with known or suspected hereditary retinoblastoma should have retinal examinations to identify retinomas or retinal scars.
- 62.(B). Infants normally have higher levels of AFP, which fall to normal adult levels by about age 8 mo; consequently, high AFP levels must be interpreted with caution in this age group.
- 63.(C). Elevation of the β subunit of human chorionic gonadotropin, which is secreted by syncytiotrophoblasts, is seen with choriocarcinoma and germinomas.
- 64.(C). Cisplatin-based chemotherapy regimens usually are curative in GCTs that cannot be completely resected, even if metastases are present.
- 65.(E). Hepatitis B or C infection is often associated with hepatocellular carcinoma.
- 66.(A). The pure fetal histology subtype predicts a more favorable outcome and the small cell undifferentiated subtype is associated with normal AFP levels and predicts a worse outcome.
- 67.(C). The risk of hemangioma is 3-5 times higher in girls than boys. The risk is doubled in premature infants and 10 times higher in offspring of women who had chorionic villus sampling.
- 68.(D). An ultrasonographic scan or MRI of the liver should be performed if multiple cutaneous lesions are present.

69.(E). All the features of Kasabach-Merritt syndrome results from platelet and red blood cell trapping and activation of the clotting system within the vasculature of the hemangioma. This syndrome is associated with kaposiform hemangioendotheliomas or tufted angiomas but not with infantile hemangiomas.

70.(C). Localized cystic hygroma lesions may be surgically resected, but this can be difficult, owing to their infiltrative nature. Recurrence is common with incompletely resected lesions. Aspiration can provide temporary relief in an emergency and reaccumulation will occur. Treatment by injection of sclerosing agents (by streptococcal immunotherapeutic agent OK-432) is the treatment of choice; its use will prevent the need for surgery in most cases. Laser therapy is not used in such age. Although spontaneous regression has been reported but is not typical.

71.(D). PTC has an excellent overall prognosis even in the presence of metastatic disease. The primary therapy is a total thyroidectomy and lymph node dissection. 131I is used postoperatively to treat distant metastasis and unresectable residual neck disease. The TSH level is suppressed by giving supraphysiologic levothyroxine as TSH stimulates tumor growth. Long-term follow-up involves monitoring of thyroglobulin tumor markers.

72.(E). Spitz nevus is a benign harmless skin lesion. However, making the distinction from diagnosis of melanoma can be difficult.

73.(B). Patients with fair skin are at particularly high risk for development of melanoma.

74.(E). Most pediatric patients present with advanced locoregional disease. LDH is elevated but it is not specific. Epstein-Barr virus DNA levels correlate with disease stage, have prognostic value and can be used to monitor for recurrence. The outcome depends on the extent of disease; patients with distant metastases have a very poor prognosis.

75.(A). LCH has an extremely variable presentation. The skeleton is involved in 80% of patients and may be the only affected site, especially in children older than 5 yr of age. Bone lesions may be single or multiple and are seen most commonly in the skull.

76.(D). Bone is the most common site of involvement by LCH. About 50% of patients experience skin involvement at some time during the course of disease. Localized or disseminated lymphadenopathy is present in approximately 33% of patients. Otitis media is present in 30-40% of patients. In 10-15% of patients, pulmonary infiltrates are found on radiography. The lesions may range from diffuse fibrosis and disseminated nodular infiltrates to diffuse cystic changes.

77.(A).

1. The part of the glomerulus which serves as a supporting and probably has a role in the regulation of glomerular blood flow, filtration, and removal of macromolecules is the
 - A. glomerular basement membrane
 - B. mesangium
 - C. bowman's capsule
 - D. parietal epithelial cell
 - E. visceral epithelial
2. Collectively, the glomerular endothelial cell, basement membrane, and podocytes form the
 - A. glomerulus
 - B. filtration membrane
 - C. juxtamedullary nephron
 - D. nephron
 - E. renal corpuscle
3. The ultra-filtrate which is filtered through the glomerular capillary walls contains all of the following **EXCEPT**
 - A. electrolytes
 - B. low-molecular-weight proteins
 - C. creatinine
 - D. albumin
 - E. glucose
4. In contrast to the concentration of blood urea nitrogen, the serum creatinine level is primarily influenced by
 - A. state of hydration
 - B. nitrogen balance
 - C. muscle mass
 - D. hemorrhage
 - E. vomiting
5. Mesangial cell disease include all the following **EXCEPT**
 - A. IgA nephropathy
 - B. membranous nephropathy

- C. mesangioproliferative glomerulonephritis
- D. diabetic nephropathy
- E. class II lupus nephritis

6. Hematuria is defined as the presence of at least 5 red blood cells per microliter of urine, false-positive results may be seen in urinary dipstick reading in all the following **EXCEPT**

- A. an alkaline urine (pH > 8)
- B. contamination with hydrogen peroxide
- C. presence of formalin
- D. hemoglobinuria
- E. myoglobinuria

7. Rhabdomyolysis is always clinically significant as it may lead to acute renal injury, it can occur secondary to

- A. severe electrolyte abnormalities
- B. bacterial endocarditis
- C. transfusion reactions
- D. mycoplasma infection
- E. autoimmune hemolytic anemia

8. Glomerular hematuria is associated with

- A. urinary microscopic findings of RBC casts
- B. minimal proteinuria on dipstick (<100 mg/dL)
- C. gross hematuria that is bright red or pink
- D. the presence of leukocytes cast
- E. normal urinary RBC morphology

9. In hematuria, renal biopsy is indicated in all the following **EXCEPT**

- A. some children with persistent microscopic hematuria
- B. decreased renal function
- C. proteinuria
- D. hypertension
- E. persistent high serum complement C3

10. IgA nephropathy is characterized by

- A. hematuria occur after skin infection
- B. proteinuria is often >1000 mg/24 hr
- C. severe hypertension
- D. normal serum levels of C3
- E. serum IgA levels have a diagnostic value

11. A 5-year-old male presented with dark color urine 1 to 2 days after an upper respiratory tract infection, GUE: RBC +++, albumin +; blood urea: 25 mg/dl; serum creatine: 0.6 mg/dl; with normal serum complement. Poor prognostic indicators at follow-up include all of the following **EXCEPT**

- A. persistent hypertension
- B. diminished renal function
- C. significant proteinuria
- D. histologically diffuse mesangial proliferation
- E. recurrent macroscopic hematuria

12. The primary treatment of IgA nephropathy is appropriate blood pressure control and management of significant proteinuria. Regarding the treatment, which of the following is **TRUE**?

- A. ACEI are ineffective in reducing proteinuria
- B. corticosteroids not improve renal function
- C. tonsillectomy is currently recommended
- D. successful kidney transplantation
- E. cyclophosphamide is effective in improving renal function

13. Alport Syndrome AS, is a genetically heterogeneous disease caused by mutations in the genes coding for type IV collagen, it is characterized by

- A. asymptomatic microscopic hematuria
- B. proteinuria < 1 g/24 hr.
- C. congenital sensor neural hearing loss
- D. ocular abnormalities in 80% of X-linked type
- E. leiomyomatosis of the esophagus is common

14. Which of the following is pathognomonic in diagnosis of X-linked type of hereditary nephritis?

- A. gross hematuria
- B. discontinuous epidermal basement membrane staining
- C. platelet abnormalities
- D. hearing loss
- E. macular flecks

15. Thin basement membrane disease TBMD is defined by the presence of persistent microscopic hematuria and isolated thinning of the GBM on electron microscopy, all the following are true **EXCEPT**

- A. significant proteinuria is rare
- B. episodic gross hematuria can be present
- C. may be transmitted as an autosomal dominant trait
- D. heterozygous mutations in the COL4A4 genes

- E. renal insufficiency is common
16. The best single test to document cutaneous streptococcal infection is
- A. low serum C3 level
 - B. elevated antistreptolysin O titer
 - C. positive streptozyme screen
 - D. antideoxyribonuclease B level
 - E. depressed serum CH50
17. A 10-year-old male (had a boil in the left thigh and mild fever one month ago) presented with headache, malaise, lethargy, joint pain, decreased urine output, and puffiness of the face; GUE: RBC cast ++, albumin ++; low C3 level and normal C4 level. Which of the following is **TRUE** about his disease?
- A. acute nephritic syndrome 40%
 - B. nephrotic syndrome 50%
 - C. acute renal failure 90%
 - D. ↑ ASO titers 70%
 - E. hypertension 70%
18. Renal biopsy in acute post streptococcal glomerulonephritis should be considered in all the following **EXCEPT**
- A. acute renal failure
 - B. nephrotic syndrome
 - C. absence of evidence of streptococcal infection
 - D. low C3 level in the first 2 months
 - E. initially normal complement levels
19. Acute post- infectious glomerulonephritis is one of the most common glomerular causes of gross hematuria in children. Of the following, the **TRUE** statement is
- A. can follow fungal infection
 - B. early systemic antibiotic, eliminate the risk of glomerulonephritis
 - C. all family members of patient need antibiotic treatment
 - D. antibiotic affect the natural history of the disease
 - E. recurrences are common
20. The **MOST** common etiology of secondary membranous nephropathy (MN) in children is
- A. neuroblastoma
 - B. measles
 - C. systemic lupus erythematosus
 - D. chronic hepatitis C

- E. penicillin
21. The well-known complication of membranous nephropathy is
- A. renal vein thrombosis
 - B. hypertension 2%
 - C. hematuria acute renal failure
 - D. gross hematuria
 - E. nephritic syndrome
22. Type I membranoproliferative glomerulonephritis is
- A. less common than type II membranoproliferative glomerulonephritis
 - B. not to be mediated by immune complexes
 - C. associated with partial lipodystrophy
 - D. called dense deposit disease
 - E. characterized by low C3 complement level
- A. 23. Renal involvement in childhood systemic lupus erythematosus (SLE) is present in
- A. 100% of patients
 - B. 80% of patients
 - C. 60% of patients
 - D. 40% of patients
 - E. 20% of patients
24. WHO classification of lupus nephritis is based on a combination of features including light microscopy, immunofluorescence, and electron microscopy. Of the following, the **WORST** outcome is associated with
- A. class I nephritis
 - B. class II nephritis
 - C. class III nephritis
 - D. class IV nephritis
 - E. class V nephritis
25. In treatment of lupus nephritis, plasmapheresis is effective in
- A. all patients
 - B. classes III and IV
 - C. maintenance therapy
 - D. reducing proteinuria
 - E. accompanying thrombotic thrombocytopenic purpura
26. Henoch-Schönlein purpura is the most common small vessel vasculitis in childhood, aggressive therapy may be reasonable in those with

- A. isolated microscopic hematuria
- B. insignificant proteinuria
- C. severe systemic manifestations
- D. >50% crescents on renal biopsy
- E. IgA deposits intensity

27. "Rapidly progressive" RPGN describes the clinical course of several forms of glomerulonephritis whose unifying feature is the histopathologic finding of crescents in the majority of glomeruli. Plasmapheresis has been reported to be of benefit in patients with

- A. post infectious glomerulonephritis
- B. immunoglobulin A nephropathy
- C. Henoch-Schönlein purpura
- D. Good pasture disease
- E. endocarditis

28. All the following diseases can cause a pulmonary-renal syndrome **EXCEPT**

- B. systemic lupus erythematosus
- C. Henoch-Schönlein purpura
- D. microscopic polyangiitis
- E. granulomatosis with polyangiitis
- F. IgA nephropathy

29. Hemolytic-uremic syndrome is a common cause of community acquired acute kidney injury in young children; atypical type may be triggered by

- A. toxin-producing E-coli
- B. raw milk
- C. Streptococcus pneumoniae
- D. shiga toxin
- E. cyclosporine

30. In hemolytic-uremic syndrome, thrombotic microangiopathies are associated with all the following **EXCEPT**

- A. genetically determined factor H deficiency
- B. neuraminidase-producing Streptococcus pneumonia infection
- C. systemic lupus erythematosus
- D. malignancy
- E. hypotension

31. In hemolytic-uremic syndrome (HUS). Of the following, the **MOST** correct statement is

- A. majority of patients of diarrhea-associated enteropathictype develops HUS

- B. stool culture is often positive in patients who have diarrhea-associated HUS
- C. kidney biopsy is rarely indicated to diagnose HUS
- D. partial thromboplastin and prothrombin times are usually low
- E. Coombs test is usually negative in pneumococci-induced HUS

32. In the treatment of diarrhea-associated hemolytic-uremic syndrome. One of the following options has no beneficial role

- A. dialysis
- B. early intravenous volume expansion
- C. control of hypertension
- D. eculizumab
- E. red cell transfusions

33. All the following matching are true **EXCEPT**

- A. nephrotic syndrome---Penicillin
- B. nephrogenic diabetes insipidus ---cisplatin
- C. nephrolithiasis ---furosemide
- D. renal tubular acidosis---lithium
- E. interstitial nephritis---cimetidine

34. Renal cortical necrosis is a rare cause of acute renal failure occurring secondary to extensive ischemic damage of the renal cortex.

Of the following, the **LEAST** common cause is

- A. perinatal asphyxia
- B. severe congenital heart disease
- C. severe hemolytic-uremic syndrome
- D. amniotic fluid embolism
- E. infectious endocarditis

35. All the following causes hematuria associated with rapid development of microangiopathic hemolytic anemia or enlargement of the kidney(s) **EXCEPT**

- A. nephrotic syndrome
- B. renal vein thrombosis
- C. polycystic kidney disease
- D. Wilms tumor
- E. hemolytic-uremic syndrome

36. Hypercalciuria can be seen in

- A. hypoparathyroidism
- B. corticosteroid therapy
- C. vitamin D deficiency
- D. oral thiazide diuretics therapy

E. Addison`s disease

37. Clinical manifestations of sickle cell nephropathy SSN include all the following **EXCEPT**

- A. hematuria
- B. polyuria
- C. hemolytic-uremic syndrome
- D. renal tubular acidosis
- E. nephrotic-range proteinuria

38. Autosomal dominant polycystic kidney disease is the most common hereditary human kidney disease, all the following are true **EXCEPT**

- A. gross or microscopic hematuria
- B. may be seen in neonates
- C. abnormal renal sonography in the absence of symptoms
- D. is a multiorgan disorder
- E. right sided valvular heart disease

39. Hemorrhagic cystitis can occur in response to all the following **EXCEPT**

- A. cyclophosphamide
- B. adenovirus infection
- C. cyclosporine
- D. polyoma BK virus infection
- E. amyloidosis

40. All the following are treatment options of hemorrhagic cystitis **EXCEPT**

- A. antibiotic
- B. intensive intravenous hydration
- C. forced diuresis
- D. analgesia
- E. spasmolytic drugs

41. In exercise hematuria; one of the following is **TRUE**

- A. A blood clots rarely seen in urine
- B. no dysuria
- C. abnormal findings on cystoscopy
- D. resolution after 7 days
- E. less common in males

42. False-negative results in urine dipstick measurement of protein can occur in

- A. very high urine pH (>7.0)
- B. large volume of urine output

- C. contamination of the urine with blood
- D. highly concentrated urine specimen
- E. contamination of the urine with antiseptic

43. Which of the following methods offer more precise information regarding urine protein excretion?

- A. frothy urine
- B. dipstick testing
- C. micro albuminuria
- D. spot urine for protein/creatinine ratio
- E. 24 hr. urine for protein and creatinine excretion

44. Transient proteinuria can be seen in all the following **EXCEPT**

- A. temperature $>38.3^{\circ}\text{C}$
- B. exercise
- C. over hydration
- D. cold exposure
- E. seizures

45. Orthostatic proteinuria is increased amounts of protein in the upright position.

Of the following, the **MOST** appropriate answer is

- A. usually symptomatic
- B. hematuria may be present
- C. renal dysfunction are absent
- D. the cause is well-known
- E. edema usually positive

46. All the following are secondary causes of nephrotic syndrome **EXCEPT**

- A. measles
- B. malaria
- C. syphilis
- D. toxoplasmosis
- E. schistosomiasis

47. Children with nephrotic syndrome are especially susceptible to infections such as cellulitis, spontaneous bacterial peritonitis, and bacteremia, all the following are contributory factors **EXCEPT**

- A. urinary losses of immunoglobulin G
- B. urinary loss of complement factors
- C. impaired opsonization of microorganisms
- D. immunosuppressive therapy
- E. urinary losses of immunoglobulin A

48. Nephrotic syndrome is a hypercoagulable state resulting from
- A. hemodilution
 - B. decreased platelet number
 - C. changes in coagulation factor levels
 - D. decrease in hepatic production of fibrinogen
 - E. high level of antithrombin III
49. When you investigate for first attack of minimal change nephrotic syndrome (MCNS), one of the following is **TRUE**
- A. microscopic hematuria is present in 20% of children
 - B. spot urine protein : creatinine ratio should be <2.0
 - C. serum complement levels are low
 - D. renal biopsy is routinely performed
 - E. 2+ proteinuria
50. In treatment of minimal change nephrotic syndrome, all the following are true **EXCEPT**
- A. tuberculosis must be ruled out prior to starting immunosuppressive therapy
 - B. steroid therapy may be initiated without a diagnostic renal biopsy
 - C. prednisone should be administered as a single daily dose of 60 mg/m²/day
 - D. remission consists of a urine <1+ protein on urine dipstick for 3 consecutive days
 - E. pleural effusion, is a potential complication of parenteral albumin therapy
51. Steroid resistance nephrotic syndrome is defined as failure to achieve remission after
- A. 4wk of corticosteroid therapy
 - B. 6 wk. of corticosteroid therapy
 - C. 8 wk. of corticosteroid therapy
 - D. 10 wk. of corticosteroid therapy
 - E. 12 wk. of corticosteroid therapy
52. To reduce the risk of serious infections in children with nephrotic syndrome, all the following immunizations can be used **EXCEPT**
- A. pneumococcal 13-valent conjugant vaccine
 - B. influenza vaccine annually to the child
 - C. pneumococcal 23-valent polysaccharide vaccine
 - D. varicella-zoster vaccine
 - E. influenza vaccination annually to their household contacts
53. After achievement of remission in children with nephrotic syndrome, they should
- A. restrict their activities
 - B. restrict sodium intake

- C. be considered chronically ill
- D. maintain an unrestricted diet
- E. decrease water/fluid intake

54. Congenital nephrotic syndrome is defined as nephrotic syndrome manifesting at birth or within the 1st 3 mo of life,

The etiologies include all the following **EXCEPT**

- A. syphilis
- B. herpes infection
- C. mercury exposure
- D. mutations in the NPHS1
- E. infantile systemic lupus erythematosus

55. The definitive treatment of primary congenital nephrotic syndrome is

- A. angiotensin-converting enzyme inhibitors
- B. prostaglandin synthesis inhibitors
- C. aggressive nutritional support
- D. bilateral nephrectomies
- E. renal transplantation

56. All the following matching are true **EXCEPT**

- A. proximal RTA----trimethoprim
- B. distal RTA----amphotericin B
- C. hyperkalemic RTA----cyclosporine
- D. distal RTA----lithium
- E. proximal RTA----gentamicin

57. Cystinosis is a systemic disease caused by a defect in the metabolism of cysteine that results in accumulation of cystine crystals in most of the major organs of the body.

All the following are recognized features **EXCEPT**

- A. renal insufficiency
- B. rickets
- C. hypothyroidism
- D. glaucoma
- E. fever

58. Treatment of cystinosis is directed at correcting the metabolic abnormalities associated with Fanconi syndrome or chronic renal failure, and the use of

- A. cystine eye drops
- B. oral cysteine
- C. prostaglandin synthesis inhibitor
- D. growth hormone

- E. angiotensin-converting enzyme inhibitors
59. Urinalysis in patients with Fanconi syndrome may show all the following **EXCEPT**
- A. urine pH is alkaline
 - B. glycosuria
 - C. uricosuria
 - D. phosphaturia
 - E. elevated urinary sodium
60. Distal RTA can be presented with
- A. hypocalciuria
 - B. hypercitraturia
 - C. phosphaturia
 - D. conductive deafness
 - E. growth failure
61. Medullary sponge kidney is a relatively rare sporadic disorder in children. It is characterized by
- A. cystic dilation of the proximal tubule
 - B. nephrolithiasis
 - C. proximal renal tubular acidosis
 - D. concentrated urine
 - E. cortical nephrocalcinosis
62. Pathogenesis of hyperkalemic (Type IV) renal tubular acidosis is due to
- A. hyperaldosteronism
 - B. inhibiting ammonia genesis
 - C. pseudohyperaldosteronism
 - D. massive bicarbonate wasting
 - E. all of the above
63. Urinary indices in patients with type IV RTA include all the following **EXCEPT**
- A. may be alkaline or acidic
 - B. elevated sodium level
 - C. inappropriately low potassium level
 - D. foul-smell
 - E. aminoaciduria
64. Nephrogenic diabetes insipidus is a disorder of water metabolism characterized by an inability to concentrate urine, even in the presence of antidiuretic hormone. Of the following, the **MOST** correct statement is
- A. most common inheritance is autosomal dominant

- B. can be caused by hypokalemia
- C. mutations in the AQP2 gene in X-linked form
- D. irritability and crying are uncommon features
- E. secondary form present with hyponatremia

65. All the following are options in the treatment of nephrogenic diabetes insipidus **EXCEPT**

- A. free access to water
- B. furosemide
- C. hydrochlorothiazide
- D. amiloride
- E. indomethacin

66. Bartter syndrome is a group of disorders characterized by hypokalemic metabolic alkalosis with hypercalciuria and salt wasting.

It can be presented with all of the following **EXCEPT**

- A. maternal polyhydramnios
- B. severe episodes of recurrent dehydration
- C. failure to thrive in classic Bartter syndrome
- D. deafness is associated with antenatal Bartter syndrome
- E. hypertension due to hyperaldosteronism

67. The diagnosis of Bartter syndrome typically include

- A. markedly low serum rennin
- B. high urinary calcium levels
- C. nephrocalcinosis
- D. hypomagnesaemia
- E. renal biopsy

68. All the following are options in the treatment of Bartter syndrome **EXCEPT**

- A. potassium supplementation
- B. aldosterone antagonist
- C. high-sodium diet
- D. Indomethacin
- E. kidney transplantation

69. Gitelman syndrome is a rare cause of autosomal recessive hypokalemic metabolic alkalosis, the distinctive feature is

- A. Hypercalciuria
- B. hypermagnesuria
- C. hyponatremia
- D. low aldosterone levels

- E. elevated prostaglandin E secretion

70. The juvenile nephronophthisis JN is a group of inherited genetically determined cystic renal diseases, it can be presented with all the following **EXCEPT**

- A. oliguria
- B. unexplained anemia
- C. growth failure
- D. chronic renal failure
- E. retinal degeneration

71. Acute kidney injury AKI is a clinical syndrome in which a sudden deterioration in renal function results in the inability of the kidneys to maintain fluid and electrolyte homeostasis.

Of the following, the **MOST** correct matching is

- A. low urine sodium ($UNa < 20$ mEq/L) : intrinsic AKI
- B. fractional excretion of sodium $> 2\%$: prerenal AKI
- C. elevated urine osmolality ($UOsm > 500$ mOsm/kg) : post renal AKI
- D. specific gravity of < 1.010 : intrinsic AKI
- E. red blood cell casts : prerenal AKI

72. Medical management of acute kidney injury include all the following **EXCEPT**

- A. bladder catheter should be placed immediately
- B. intravenous administration of boluses isotonic saline
- C. furosemide may be administered as a single IV dose
- D. fluid restriction is essential if there is no response to a diuretic challenge
- E. mannitol is not effective in prevention of pigment induced renal failure

73. Hyperkalemia can lead to cardiac arrhythmia, cardiac arrest, and death. Serum potassium can be lowered by all the following **EXCEPT**

- A. regular insulin with glucose
- B. calcium gluconate
- C. sodium bicarbonate
- D. Kayexalate
- E. Dialysis

74. Treatment of hypocalcemia in acute kidney injury include all the following **EXCEPT**

- A. low-phosphorus diet
- B. intra venous phosphate binders
- C. calcium should not be given intravenously
- D. sevelamer
- E. avoid aluminum-based binders

75. Indications for dialysis in acute kidney injury include all the following **EXCEPT**
- A. anuria/oliguria
 - B. persistent hypercalcemia
 - C. severe metabolic acidosis
 - D. volume overload
 - E. blood urea nitrogen >100-150 mg/dl
76. Many patients with acute kidney injury require dialysis support for 1-3 wk, which of the following is **TRUE** statement
- A. Intermittent hemodialysis is useful in patients with unstable hemodynamic status
 - B. continuous renal replacement therapy is useful in patients with stable hemodynamic status
 - C. in peritoneal dialysis ,hypo-osmolar dialysate is infused into the peritoneal cavity
 - D. anticoagulation is not necessary in peritoneal dialysis
 - E. urea and creatinine clearance more with peritoneal dialysis than intermittent hemodialysis
77. Chronic kidney disease in children <5-yr-old is **MOST** commonly a result of
- A. lupus nephritis
 - B. familial juvenile nephronophthisis
 - C. Alport syndrome
 - D. focal segmental glomerulosclerosis
 - E. autosomal dominant polycystic kidney disease
78. Pathogenesis of chronic kidney disease (CKD) include all the following **EXCEPT**
- A. hyperfiltration injury
 - B. hyperkalemia
 - C. proteinuria
 - D. hypertension
 - E. hyperphosphatemia
79. End-stage renal disease (ESRD) represents the state in which a patient's renal dysfunction has progressed to the point at which homeostasis and survival can no longer be sustained with native kidney function and maximal medical management. Of the following, the **TRUE** statement is
- A. the ultimate goal for children is dialysis
 - B. renal replacement therapy be initiated at stage 5 CKD
 - C. the time to actually initiate dialysis include impaired school performance
 - D. peritoneal dialysis is performed 3 times weekly
 - E. peritoneal dialysis use cyclical-driven therapy increases the risk of peritonitis

80. Kidney transplantation is the optimal therapy for children with end-stage renal disease, all the following are true **EXCEPT**

- A. relative contraindication is preexisting metastatic malignancy
- B. successful transplantation leads to improvement in their linear growth
- C. preemptive transplantation mean transplantation without prior dialysis
- D. optimal outcomes result when the child weighs >15 kg
- E. urologic problems should be addressed before surgery

81. Recurrent disease in the renal graft occur in all cases of

- A. focal segmental glomerulosclerosis
- B. primary oxalosis
- C. membranoproliferative glomerulonephritis type II
- D. IgA nephropathy
- E. congenital nephrotic syndrome

82. Indications for bilateral native nephrectomies before renal transplantation include

- A. Denys-Drash syndrome
- B. IgA nephropathy
- C. chronic glomerulonephritis
- D. Prune belly syndrome
- E. medullary cystic disease

83. Before renal transplantation, hemoglobin levels should be maintain at the level between

- A. 7 and 8 g/dL
- B. 9 and 10 g/dL
- C. 11 and 12 g/dL
- D. 13 and 14 g/dL
- E. 15 and 16 g/dL

84. Risk factor for renal graft thrombosis include

- A. prior history of peritoneal dialysis
- B. arterial hypertension
- C. donor age (>2 yr.)
- D. recipient age (>5 yr.)
- E. cold ischemia time <24 hr.

85. Infections need to be identified and treated before renal transplantation. Infectious disease screening includes all the following **EXCEPT**

- A. tuberculosis skin test
- B. cytomegalovirus IgG
- C. measles antibody

- D. hepatitis A serology
 - E. varicella titer
86. Antithymocyte globulin and thymoglobulin act against
- A. interleukin-2 Receptor Antibodies
 - B. T-lymphocyte antigens
 - C. B cells
 - D. CD20 epitope
 - E. mammalian target of rapamycin
87. Of the following, the **BEST** choice for maintenance immunosuppression in renal transplantation is
- A. cyclosporin
 - B. basiliximab
 - C. daclizumab
 - D. rituximab
 - E. belatacept
88. The side-effect profile of cyclosporine in children include all the following **EXCEPT**
- A. hypertrichosis
 - B. gingival hyperplasia
 - C. neutropenia
 - D. coarsening facial features
 - E. hyperlipidemia
89. Corticosteroids remain an integral part of many immunosuppressive protocols despite their multifaceted toxicities. In children, the **MOST** noteworthy side effect is
- A. obesity
 - B. hypertension
 - C. aseptic necrosis of bone
 - D. retarded skeletal growth
 - E. cushingoid faces
90. Polyomavirus nephropathy is an important cause of allograft dysfunction; the increased incidence is thought to be the result of
- A. reduction in immunosuppression doses
 - B. more potent immunosuppressive regimens
 - C. cidofovir use
 - D. leflunomide use
 - E. ciprofloxacin therapy
91. The following formulas are true **EXCEPT**

- A. sodium requirement = $0.6 \times \text{weight in kg} \times (125 - \text{serum sodium in mEq/L})$
- B. blood anion gap = $[\text{Na}] - [\text{Cl} + \text{HCO}_3]$
- C. blood anion gap = $[\text{Na} - \text{Cl} - \text{HCO}_3]$
- D. urine anion gap = $([\text{urine Na} + \text{urine K}^+] - \text{urine Cl}^-)$
- E. estimated GFR = $0.43 \times \text{height in m} / \text{serum creatinine in mg/dL}$

1.(B).

2.(B).

3.(D). The ultra-filtrate, which is cell free, contains all of the substances in plasma (electrolytes, glucose, phosphate, urea, creatinine, peptides, and low-molecular-weight proteins except proteins having a molecular weight of ≥ 68 kDa [such as albumin and globulins]).

4.(C). Creatinine is affected by sex, height, muscle mass, bilirubin, and red blood cell hemolysis, while the concentration of blood urea nitrogen is affected by state of hydration and nitrogen balance.

5.(B). Membranous nephropathy result from epithelial cell injury, epithelial cells, in conjunction with basement membrane, allow filtration of plasma solutes but retard passage of cells and plasma proteins. Disease related to these cells is typified by the presence of sub epithelial deposits and flattening of the foot processes that engage the basement membrane, resulting in disruption of the filtration barrier and proteinuria.

6.(C). False negative results can occur in the presence of formalin (used as a urine preservative) or high urinary concentrations of ascorbic acid (i.e., in patients with vitamin C intake >2000 mg/day).

7.(A). Rhabdomyolysis can occurs secondary to viral myositis, crush injury, severe electrolyte abnormalities (hypernatremia, hypophosphatemia), hypotension, disseminated intravascular coagulation, toxins (drugs, venom), metabolic disorders of muscles, and prolonged seizures. While other distractors are causes of hemolysis and hemoglobinuria.

8.(A). Hematuria from within the glomerulus is often associated with urinary microscopic findings of RBC casts, and deformed urinary RBCs (particularly acanthocytes), brown, cola- or tea-colored, or burgundy urine, and proteinuria >100 mg/dL via dipstick. Hematuria originating within the tubular system may be associated with the presence of leukocytes or renal tubular casts. Lower urinary tract sources of hematuria may be associated with gross hematuria that is bright red or pink, terminal hematuria (gross hematuria occurring at the end of the urine stream), blood clots, normal urinary RBC morphology, and minimal proteinuria on dipstick (<100 mg/dL).

9.(E). Renal biopsy is indicated for children with persistent low serum complement C3.

10.(D). Gross hematuria often occurs within 1-2 days of onset of an upper respiratory or gastrointestinal infection, in contrast to the longer latency period observed in acute post infectious glomerulonephritis, and may be associated with loin pain. Proteinuria is often <1000 mg/24 hr in patients with asymptomatic microscopic hematuria. Mild to moderate hypertension is most often seen in patients with nephritic or nephrotic

syndrome, but is rarely severe enough to result in hypertensive emergencies. Normal serum levels of C3 in IgA nephropathy help to distinguish this disorder from post streptococcal glomerulonephritis. Serum IgA levels have no diagnostic value because they are elevated in only 15% of pediatric patients.

11.(E). Recurrent macroscopic hematuria is a good prognostic indicator.

12.(D). Angiotensin-converting enzyme inhibitors and angiotensin II receptor antagonists are effective in reducing proteinuria and retarding the rate of disease progression when used individually or in combination. Corticosteroids reduce proteinuria and improve renal function in those patients with a glomerular filtration rate >60 mL/min/m². To date, additional immunosuppression with cyclophosphamide or azathioprine has not appeared to be effective, but further randomized clinical trials are in progress. Tonsillectomy has been used as treatment for IgA nephropathy in many countries including Japan. Performing a tonsillectomy in the absence of significant tonsillitis in association with IgA nephropathy is currently not recommended until appropriate prospective, controlled trials have been performed and demonstrate efficacy. Patients with IgA nephropathy may undergo successful kidney transplantation. Although recurrent disease is frequent, allograft loss caused by IgA nephropathy occurs in only 15-30% of patients.

13.(A). All patients with AS have asymptomatic microscopic hematuria, which may be intermittent in girls and younger boys. Progressive proteinuria, often exceeding 1 g/24 hr., is common by the 2nd decade of life and can be severe enough to cause nephrotic syndrome. Bilateral sensor neural hearing loss, which is never congenital, Ocular abnormalities, which occur in 30-40% of patients with X-linked AS, include anterior lenticonus (extrusion of the central portion of the lens into the anterior chamber), macular flecks, and corneal erosions. Leiomyomatosis of the esophagus, tracheobronchial tree, and female genitals in association with platelet abnormalities has been reported, but is rare.

14.(B). Absence of epidermal basement membrane staining for the $\alpha 5$ chain of type IV collagen in male hemizygotes and discontinuous epidermal basement membrane staining in female heterozygotes on skin biopsy is pathognomonic for X-linked AS and can preclude diagnostic renal biopsy.

15.(E). Rare cases of TBMD progress, and such patients develop significant proteinuria, hypertension, or renal insufficiency. [heterozygous mutations in the COL4A3 and COL4A3 genes, which encode the $\alpha 3$ and $\alpha 4$ chains of type IV collagen present in the GBM, result in TBMD while homozygous mutations in these same genes result in Autosomal recessive Alport syndrome].

16.(D). Four differences in APSGN follow throat or skin infections:

Post streptococcal GN commonly follows streptococcal pharyngitis during cold-weather months and streptococcal skin infections or pyoderma during warm-weather months. Epidemics of nephritis have been described in association with throat (serotypes M1, M4, M25, and some strains of M12) and skin (serotype M49) infections.

The typical patient develops an acute nephritic syndrome 1-2 wk. after an antecedent streptococcal pharyngitis or 3-6 wk. after a streptococcal pyoderma.

The antistreptolysin O titer is commonly elevated after a pharyngeal infection but rarely increases after streptococcal skin infections.

17.(E). Acute nephritic syndrome 90%, nephrotic syndrome 10-20%, acute renal failure 50%, the antistreptolysin O titer is commonly elevated after a pharyngeal infection but rarely increases after streptococcal skin infections, hypertension 70%.

18.(D). Renal biopsy is considered when hematuria and proteinuria, diminished renal function, and/or a low C3 level persist more than 2 mo. after onset.

19.(A). Acute GN can occur after certain fungal, rickettsial, protozoan, parasitic, or viral diseases (influenza and parvovirus) that is why the terms APSGN and acute post infectious GN are used synonymously. Early systemic antibiotic therapy for streptococcal throat and skin infections does not eliminate the risk of GN. Family members of patients with acute GN, especially young children, should be considered at risk and be cultured for group A β -hemolytic streptococci and treated if positive. Family pets, particularly dogs, have also been reported as carriers. A 10 day course of systemic antibiotic therapy with penicillin is recommended to limit the spread of the nephritogenic organisms, antibiotic therapy does not affect the natural history of APSGN, and recurrences are extremely rare.

20.(C). MN also associated with chronic hepatitis B infection and congenital syphilis, other chronic infections, including malaria, which is likely the most common cause of nephrotic syndrome worldwide. Certain medications, such as penicillamine and gold, or chronic factor replacement in patients with hemophilia. Rare causes such as neuroblastoma, or other idiopathic systemic diseases.

21.(A). A subset of patients with MN present with a major venous thrombosis, commonly renal vein thrombosis, this well-known complication. Approximately 20% of children have hypertension at presentation, most patients also have microscopic hematuria and only rarely present with gross hematuria. MN is most common in the 2nd decade of life, but it can occur at any age, including infancy. The disease usually manifests as nephrotic syndrome and accounts for 2-6% of all cases of childhood nephrotic syndrome.

22.(E). Type II MPGN (dense deposit disease) is less common than type I membranoproliferative glomerulonephritis, not to be mediated by immune complexes, demonstrate an associated systemic disease called partial lipodystrophy, serum C3 complement levels are low in the majority of cases (while in membranous nephropathy is normal), Patients present in equal proportions with nephrotic syndrome, acute nephritic syndrome, or persistent asymptomatic microscopic hematuria and proteinuria (while in membranous nephropathy usually manifest as nephrotic syndrome).

23.(B). Renal disease in childhood SLE is present in up to 80% of patients while approximately 50% of patients with HSP develop renal manifestations

- 24.(D). Renal biopsy should be performed in all patients with SLE because histopathologic findings are used to determine the selection of specific immunosuppressive therapies.
- 25.(E). Plasmapheresis is ineffective in lupus nephritis unless there is accompanying thrombotic thrombocytopenic purpura or antineutrophilic cytoplasmic antibody associated disease.
- 26.(D). Spontaneous and complete resolution of the nephritis typically occurs in the majority of patients with mild initial manifestations (isolated hematuria with insignificant proteinuria), and the severity of the systemic manifestations does not correlate with the severity of the nephritis.
- 27.(D). Plasmapheresis may also benefit patients with ANCA-associated crescentic GN, in particular those with the most severe renal dysfunction at presentation. The possible benefits of plasmapheresis in other forms of RPGN are unclear.
- 28.(E).
- 29.(E).
- 30.(E). Malignant hypertension.
- 31.(C). Kidney biopsies are only rarely performed in HUS because the diagnosis is usually established by clinical criteria and the risks of biopsy are significant during the active phase of the disease. The organisms that cause HUS may be rapidly cleared, therefore, the stool culture is often negative in patients who have diarrhea-associated HUS, Partial thromboplastin and prothrombin times are usually normal, the Coombs test is negative, with the exception of pneumococci-induced HUS, where the Coombs test is usually positive.
- 32.(D). Eculizumab is an anti-C5 antibody that inhibits complement activation, a pathway that contributes to active disease in some forms of atypical familial HUS, Eculizumab is FDA approved for the treatment of atypical HUS, while initial reports suggested that eculizumab provided benefit in patients with diarrhea associated HUS, subsequent systematic analysis showed no benefit from either plasma exchange or eculizumab.
- 33.(A). Nephrotic syndrome--- penicillamine while interstitial nephritis--- penicillin
- 34.(E). Less-common causes of cortical necrosis include malaria, extensive burns, snakebites, infectious endocarditis, and medications (e.g., nonsteroidal anti-inflammatory agents). Acute renal cortical necrosis has also been reported to occur in systemic lupus erythematosus-associated antiphospholipid antibody syndrome.
- 35.(A). These include hemolytic uremic syndrome, hydronephrosis, polycystic kidney disease, Wilms tumor, and intrarenal abscess or hematoma. All patients should be evaluated for congenital and acquired hypercoagulable states.
- 36.(B). Hypercalciuria can accompany conditions resulting in hypercalcemia, such as hyperparathyroidism, vitamin D intoxication, immobilization, and sarcoidosis. Hypercalciuria may be associated with Cushing syndrome, corticosteroid therapy, tubular dysfunction secondary to Fanconi syndrome (Wilson disease, oculocerebrorenal syndrome), Williams syndrome, distal renal tubular acidosis, or

Bartter syndrome. Oral thiazide diuretics can normalize urinary calcium excretion by stimulating calcium reabsorption in the proximal and distal tubules.

37.(C). Polyuria caused by a urinary concentrating defect, renal tubular acidosis, and proteinuria associated with the glomerular lesions. Approximately 20-30% of patients with sickle cell disease develop proteinuria, Nephrotic-range proteinuria with or without clinically apparent nephrotic syndrome occurs in up to 30% of patients with SSN, and when present generally heralds progressive renal failure.

38.(E). Mitral valve prolapse is seen in approximately 12% of children; aortic and coronary artery aneurysms and aortic valve insufficiency are noted in affected adults. It is a systemic disorder with possible cyst formation in multiple organs (liver, pancreas, spleen, brain) and the development of saccular cerebral aneurysms.

39.(C). The hemorrhagic cystitis can occur in response to chemical toxins (cyclophosphamide, penicillin's, busulfan, thiotepea, dyes, insecticides), viruses (adenovirus types 11 and 21 and influenza A), radiation, and amyloidosis. The polyoma BK virus, present latently in immunocompetent hosts, is associated with the development of drug-induced cystitis in immunosuppressed patients.

40.(A).

41.(A). Findings on urine culture, intravenous pyelography, voiding cystourethrography, and cystoscopy are normal in most patients, hematuria generally resolves within 48 hr. after cessation of exercise, exercise hematuria is less common in females and can be associated with dysuria.

42.(B). Dilute urine or a large volume of urine output or in disease states in which the predominant urinary protein is not albumin cause false-negative test results.

43.(E).

44.(C). Defined contributing factors include a temperature $>38.3^{\circ}\text{C}$ (101°F), exercise, dehydration, cold exposure, heart failure, seizures, or stress.

45.(C). The absence of proteinuria (dipstick negative or trace for protein; and a normal ratio of urinary protein [mg/dL] to urinary creatinine [mg/dL] = $[\text{uPr}/\text{uCr}] < 0.2$) on the first morning urine sample for 3 consecutive days confirms the diagnosis of orthostatic proteinuria, children with this condition are usually asymptomatic; hematuria, hypertension, hypoalbuminemia, edema, and renal dysfunction are absent, the cause of orthostatic proteinuria is unknown, although altered renal hemodynamics and partial left renal vein obstruction in the upright, lordotic position have been proposed as possible causes.

46.(A). Infections causes secondary nephrotic syndrome is : Endocarditis, Hepatitis B, C,HIV-1, infectious mononucleosis, malaria, syphilis (congenital and secondary), toxoplasmosis, schistosomiasis, filariasis.

47.(E).

48.(C). Vascular stasis from hemoconcentration,intravascular volume depletion , increased platelet number and aggregability, increase in hepatic production of fibrinogen,and low level of antithrombin III, cause hypercoagulable state.

- 49.(A). A spot urine protein : creatinine ratio should be >2.0, serum complement levels are normal, a renal biopsy is not routinely performed if the patient fits the standard clinical picture of MCNS, and the urinalysis reveals 3+ or 4+ proteinuria.
- 50.(E). Symptomatic volume overload, hypertension, heart failure, and pulmonary edema, is a potential complication of parenteral albumin therapy, particularly when administered as rapid infusions.
- 51.(C). Steroid-resistant nephrotic syndrome is usually caused by focal segmental glomerulosclerosis (80%), minimal change nephrotic syndrome, or membranoproliferative glomerulonephritis.
- 52.(D). Following close contact with varicella infection, give immune compromised children on immunosuppressive agents varicella-zoster immune globulin if available.
- 53.(D). To minimize the psychologic effects of the condition and its therapy, children with idiopathic nephrotic syndrome should not be considered chronically ill and should participate in all age-appropriate childhood activities.
- 54.(B). Congenital nephrotic syndrome may be classified as primary or as secondary to a number of etiologies such as in utero infections (cytomegalovirus, toxoplasmosis, syphilis, hepatitis B and C, HIV).
- 55.(E). Renal transplantation is the definitive treatment of congenital nephrotic syndrome, though recurrence of the nephrotic syndrome has been reported to occur after transplantation.
- 56.(A). Hyperkalemia RTA---- Trimethoprim/sulfamethoxazole.
- 57.(D). Ocular presentations include photophobia, retinopathy, and impaired visual acuity. Patients also can develop hepatosplenomegaly, and delayed sexual maturation.
- 58.(D). Cysteamine, which binds to cystine and converts it to cysteine. Oral cysteamine does not achieve adequate levels in ocular tissues, so additional therapy with cysteamine eye drops is required. Patients with growth failure that does not improve with cysteamine might benefit from treatment with growth hormone. Kidney transplantation is a viable option.
59. (A).The urine pH is acidic (<5.5) because distal acidification mechanisms are intact in these patients.
- 60.(E). Hypercalciuria, hypocitraturia, and sensorineural deafness are seen in distal RTA while phosphate and massive bicarbonate wasting are characteristic of proximal RTA.
- 61.(B). Medullary sponge kidney is characterized by cystic dilation of the terminal portions of the collecting ducts as they enter the renal pyramids, Ultrasonographically, patients often have medullary nephrocalcinosis, complications include nephrolithiasis, pyelonephritis, hyposthenuria (inability to concentrate urine), and distal RTA.
- 62.(B). Type IV RTA occurs as the result of impaired aldosterone production (hypoaldosteronism) or impaired renal responsiveness to aldosterone (pseudohypoaldosteronism), proximal RTA pathogenesis is massive bicarbonate wasting. Acidosis results because aldosterone has a direct effect on the H⁺/ATPase responsible for hydrogen secretion. In addition, aldosterone is a potent stimulant for potassium secretion in the collecting tubule; consequently, lack of aldosterone results

in hyperkalemia. This further affects acid–base status by inhibiting ammoniagenesis and, thus, H⁺ excretion.

63.(E). Urinary indices in patients with Fanconi syndrome demonstrate varying degrees of aminoaciduria, and elevated urinary sodium or potassium; while in type IV RTA elevated urinary sodium levels with inappropriately low urinary potassium levels reflect the absence of aldosterone effect.

64.(B). The most common pattern of inheritance in congenital NDI is an X-linked recessive disorder, mutations in the AQP2 gene have been identified in patients with the rarer autosomal dominant and recessive forms, Irritability and crying are common features, patients with the secondary form generally present later in life, primarily with hypernatremia and polyuria.

65.(B).

66.(E). Blood pressure is usually normal. Patients with the antenatal form can have severe salt wasting, resulting in dehydration and hypotension.

67.(B). Urinary calcium levels are typically elevated, as are urinary potassium and sodium levels, nephrocalcinosis resulting from Hypercalciuria may be seen on ultrasound examination. Serum renin, aldosterone, and prostaglandin E levels are often markedly elevated; hypomagnesaemia is seen in a minority of patients but is more common in Gitelman syndrome. Renal biopsy is rarely performed to diagnose this condition.

68.(E).

69.(B). The urinary calcium level is usually very low (in contrast to the elevated urinary calcium level often seen in Bartter syndrome), hypokalemia, aldosterone levels usually normal, prostaglandin E secretion is not elevated.

70.(A). Patients with JN typically present with polyuria, anemia that is seemingly disproportionate to the degree of renal insufficiency, JN often have characteristic small cysts in the corticomedullary region.

71.(D). Patients whose urine shows an elevated specific gravity (>1.020), elevated urine osmolality (UOsm>500 mOsm/kg), low urine sodium (UNa< 20 mEq/L), and fractional excretion of sodium <1% (<2.5% in neonates) most likely have prerenal AKI. Those with a specific gravity of <1.010, low urine osmolality (UOsm< 350 mOsm/kg), high urine sodium (UNa> 40 mEq/L), and fractional excretion of sodium >2% (>10% in neonates), red blood cell casts most likely have intrinsic AKI and low urine osmolality (UOsm< 350 mOsm/kg) in Post renal AKI.

72.(E). Mannitol may be effective in prevention of pigment (myoglobin, hemoglobin) induced renal failure.

73.(B). Calcium gluconate counteracts the potassium-induced increase in myocardial irritability but does not lower the serum potassium level.

74.(B). Hypocalcemia is primarily treated by lowering the serum phosphorus level, phosphate binders should be orally administered to bind any ingested phosphate and increase GI phosphate excretion. Calcium should not be given i.v except in cases of tetany to avoid deposition of calcium salts into tissues.

75.(B). Persistent hyperkalemia.

76.(D). Intermittent hemodialysis is useful in patients with relatively stable hemodynamic status, while continuous renal replacement therapy is useful in patients with unstable hemodynamic status, concomitant sepsis, or multiorgan failure in the intensive care setting, hyperosmolar dialysate is infused into the peritoneal cavity via a surgically or percutaneously placed peritoneal dialysis catheter, urea and creatinine clearance more with intermittent hemodialysis than peritoneal dialysis.

77.(D). After 5 yr of age, acquired diseases (various forms of glomerulonephritis including lupus nephritis) and inherited disorders (familial juvenile nephronophthisis, Alport syndrome) predominate.

78.(B). Hyperkalemia is the result of CKD not a cause.

79.(C). The ultimate goal for children with ESRD is successful kidney transplantation. It is recommended that plans for renal replacement therapy be initiated when a child reaches stage 4 CKD. Hemodialysis performed 3 times weekly. Peritoneal dialysis use cycler-driven therapy reduction in the number of dialysis catheter connections and disconnections (which decreases the risk of peritonitis).

80.(D). Optimal outcomes result when the child weighs ≥ 10 kg.

81.(C).

82.(A). Owing to the high risk of developing Wilms tumor, patients with Denys-Drash syndrome should undergo bilateral nephrectomy before transplantation.

83.(C).

84.(A).

85.(D). Infections need to be identified and treated before renal transplantation are; Hepatitis B and Hepatitis C

86.(B).

87.(A). Central to many current pediatric immunosuppressive regimens is a calcineurin inhibitor (cyclosporine or tacrolimus) in combination with steroids and an adjunctive anti proliferative agent (azathioprine, sirolimus, or MMF), other for induction therapies.

88.(C). Gastrointestinal and hematologic side effects occur in mycophenolate mofetil, but not in cyclosporine.

89.(D).

90.(B). That is why reducing immunosuppression is the main form of therapy, and cidofovir and leflunomide are used as adjunctive therapies.

91.(E). Schwartz formula for estimated GFR = $0.43 \times \text{height in cm} / \text{serum creatinine in mg/dL}$.

Urologic Disorders in Infants and Children QUESTIONS

QAHTAN ALOBAIDY

1. Glomerular filtration rate in full term neonate is
 - A. 15 mL/min/1.73 m²
 - B. 25 mL/min/1.73 m²
 - C. 35 mL/min/1.73 m²
 - D. 45 mL/min/1.73 m²
 - E. 55 mL/min/1.73 m²

2. Potter syndrome (bilateral renal agenesis) is characterized by all the following **EXCEPT**
 - A. widely separated eyes
 - B. polyhydramnios
 - C. low set ears
 - D. broad nose
 - E. limb anomalies

3. Death in neonates with bilateral renal agenesis is due to
 - A. renal failure
 - B. hypertension
 - C. pulmonary insufficiency
 - D. congenital heart disease
 - E. sepsis

4. The **MOST** common cause of an abdominal mass in the newborn is
 - A. polycystic kidney disease
 - B. neuroblastoma
 - C. renal vein thrombosis
 - D. hydronephrosis
 - E. multicystic dysplastic kidney

5. One of the complications of the horseshoe kidney is
 - A. renal abscess
 - B. neuroblastoma
 - C. bladder outlet obstruction
 - D. familial renal dysplasia

- E. multicystic dysplastic kidney
6. All the following are risk factors for urinary tract infection **EXCEPT**
- A. male gender
 - B. tight clothing (underwear)
 - C. pinworm infestation
 - D. poor toilet training
 - E. voiding dysfunction
7. Pyelonephritis usually requires total or partial nephrectomy in
- A. renal abscess
 - B. perinephric abscess
 - C. xanthogranulomatous pyelonephritis
 - D. pyelonephritic scarring
 - E. acute lobar nephronia
8. Symptoms of cystitis include
- A. fever
 - B. polyuria
 - C. nausea
 - D. suprapubic pain
 - E. urinary retention
9. According to the clinical guidelines, urine sample for diagnosis of urinary tract infection in children 2-24 mo should be from
- A. an adhesive collection bag
 - B. mid-stream sample
 - C. urine bag
 - D. catheterized sample
 - E. all of the above
10. In pyelonephritis, a high risk of renal scarring is associated with an elevated
- A. erythrocyte sedimentation rate
 - B. C-reactive protein
 - C. procalcitonin
 - D. white blood cell count
 - E. serum ferritin
11. Acute cystitis should be treated promptly to prevent possible progression to pyelonephritis.
- Of the following, the **LEAST** effective drug is
- A. trimethoprim-sulfamethoxazole

- B. cephalixin
- C. ciprofloxacin
- D. nitrofurantoin
- E. amoxicillin

12. All the following are indications for admission to hospital in patients with urosepsis **EXCEPT**

- A. dehydration
- B. unable to drink fluids
- C. neonatal period
- D. vomiting
- E. microscopic hematuria

13. Regarding vesicoureteral reflux VUR (the retrograde flow of urine from the bladder to the ureter and kidney).

All the following are true **EXCEPT**

- A. it is usually congenital
- B. it may cause hypertension in children
- C. the mean age at VUR resolution is 10yr
- D. severity is graded according to voiding cystourethrogram
- E. reimplantation of the ureters corrects the condition

14. Vesicoureteral reflux is usually discovered during evaluation for a UTI, which of the following is **TRUE**

- A. autosomal dominant inheritance
- B. males are mainly affected
- C. antenatal hydronephrosis mainly seen in females
- D. constipation is a rare association
- E. average age at diagnosis is less than 1 year

15. Regarding the voiding cystourethrogram (VCUG) for diagnosis of vesicoureteral reflux (VUR).

Of the following, the **MOST** appropriate statement is

- A. low-pressure VUR is occurring during voiding
- B. high-pressure VUR is occurring during bladder filling
- C. high-pressure VUR is significantly less likely to resolve spontaneously
- D. contrast VCUG is significantly exposed to less radiation than radionuclide cystogram
- E. contrast VCUG provides more anatomic information

16. The antenatal hydronephrosis is graded by the trimester and the antero-posterior diameter of the renal pelvis.

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

- A. vesicoureteral reflux
- B. ureteropelvic junction obstruction
- C. transient hydronephrosis
- D. posterior urethral valve
- E. ureterovesical junction obstruction

17. Ureteropelvic junction obstruction is the most common obstructive lesion in childhood.

Of the following, the **MOST** correct statement is

- A. it is usually caused by extrinsic stenosis
- B. it mainly occurs on the right side
- C. there is a female preponderance
- D. voiding cystourethrogram (VCUG) is necessary
- E. it is usually bilateral

18. Prompt surgical repair in ureteropelvic junction obstruction is indicated in infants with all the following **EXCEPT**

- A. solitary kidney
- B. abdominal mass
- C. bilateral severe hydronephrosis
- D. hematuria after minimal trauma
- E. diminished function in the involved kidney

19. Ureter that drains outside the bladder is referred to as an ectopic ureter.

Of the following, which statement is **TRUE** regarding ectopic ureter?

- A. male: female ratio is 2: 1
- B. it mostly drains into the cervix in girls
- C. it mostly drains into the vas deferens in boys
- D. urinary tract infection (UTI) is uncommon
- E. nephrectomy is indicated if renal function is poor

20. Ureterocele is a cystic dilation of the terminal ureter.

Of the following, the **MOST** appropriate statement is

- A. it is more common in boys
- B. it cannot be diagnosed prenatally
- C. it is associated with ureteral duplication in girls
- D. it commonly causes bladder outlet obstruction
- E. the orthotopic type extends into the urethra

21. Prune-belly syndrome, also called triad syndrome or Eagle-Barrett syndrome, is characterized by

- A. female predominance
- B. polydramnios
- C. small bladder
- D. cardiac abnormalities (in 10% of cases)
- E. all the above

22. Posterior urethral valves, is the most common cause of severe obstructive uropathies in children.

Of the following, the **MOST** appropriate statement

- A. its incidence 1 in 40,000 boys
- B. vesicoureteral reflux occurs in 5% of patients
- C. the urinary stream is strong
- D. Foley catheter should be avoided
- E. it is a diverticulum in the penile urethra

23. Classic bladder exstrophy is characterized by

- A. hypospadias
- B. narrow based gait
- C. normal upper urinary tracts
- D. posteriorly displaced anus
- E. upward displaced umbilicus is

24. Neuropathic bladder dysfunction in children is usually congenital resulting from neural tube defects or other spinal abnormalities.

All the following are options in the treatment **EXCEPT**

- A. botulinum toxin
- B. cholinergic drugs
- C. cutaneous vesicostomy
- D. antimicrobial prophylaxis
- E. clean intermittent catheterization

25. Complications of augmentation cystoplasty includes all the following **EXCEPT**

- A. bladder calculi
- B. metabolic alkalosis
- C. malignant neoplasm
- D. urinary tract infection
- E. spontaneous perforation

26. In children up to the age of 14 yr, the mean bladder capacity in ounces is equal to the age (in years) plus

- A. 2
- B. 3

- C. 4
- D. 5
- E. 6

27. The **MOST** common cause of daytime incontinence is

- A. an overactive bladder
- B. infrequent voiding
- C. detrusor–sphincter dyssynergia
- D. bladder outlet obstruction
- E. behavioral

28. Staccato urinary stream is seen in

- A. hypospadias
- B. meatal stenosis
- C. ureteral ectopia
- D. posterior urethral valves
- E. non neurogenic bladder

29. Pollakiuria is characterized by

- A. dysuria
- B. nocturia
- C. occurrence at 7-10 yr of age
- D. daytime incontinence
- E. voiding every 10-15 min during the day

30. Family history in nocturnal enuresis is positive in

- A. 10%
- B. 30%
- C. 50%
- D. 70%
- E. 90%

31. The pathogenesis of nocturnal enuresis is

- A. defective sleep arousal
- B. nocturnal polyuria
- C. genetic factors
- D. overactive bladder
- E. multifactorial

32. The **MOST** effective way of treatment in older children with nocturnal enuresis is

- A. motivational therapy
- B. conditioning therapy

- C. desmopressin acetate
 - D. oxybutynin chloride
 - E. imipramine
33. All the following are contraindications to circumcision in neonates **EXCEPT**
- A. hypospadias
 - B. chordee without hypospadias
 - C. dorsal hood deformity
 - D. small penis
 - E. penile torsion
34. The most common cause of micropenis is failure of the hypothalamus to produce an adequate amount of gonadotropin-releasing hormone.
Of the following, the syndrome associated with micropenis is
- A. Kallmann syndrome
 - B. fetal hydantoin syndrome
 - C. Apert syndrome
 - D. de Lange syndrome
 - E. Holt-Oram syndrome
35. Priapism of high-flow type **MOST** commonly follows
- A. sickle cell disease
 - B. perineal trauma
 - C. sildenafil ingestion
 - D. leukemia
 - E. all the above
36. The **MOST** common male urethral anomaly associated with prune-belly syndrome is
- A. congenital urethral fistula
 - B. urethral duplication
 - C. megalourethra
 - D. urethral hypoplasia
 - E. urethral atresia
37. The consequences of cryptorchidism include all the following **EXCEPT**
- A. infertility
 - B. hydrocele
 - C. inguinal hernia
 - D. testicular malignancy
 - E. poor testicular growth
38. The **MOST** common cause of testicular pain in a 12-yr-old boy is

- A. epididymitis
 - B. testicular tumor
 - C. testicular torsion
 - D. scrotal hematoma
 - E. incarcerated inguinal hernia
39. The risk of renal stone formation increases in the presence of
- A. low urine pH
 - B. urinary citrate
 - C. urinary magnesium
 - D. urinary glycosaminoglycan
 - E. urinary osteopontin
40. Irritative symptoms of dysuria, urgency, and frequency usually indicate a calculus in the
- A. urethra
 - B. bladder
 - C. distal ureter
 - D. middle part of ureter
 - E. ureteropelvic junction
41. Uric acid stones occur in
- A. inflammatory bowel disease
 - B. corticosteroids therapy
 - C. vitamin D excess
 - D. distal renal tubular acidosis
 - E. hyperoxaluria
42. The **MOST** accurate study in a child with suspected renal stone is
- A. renal ultra-sonogram
 - B. plain radiograph of the abdomen
 - C. an unenhanced spiral CT scan
 - D. radioisotope studies
 - E. magnetic resonance urography
43. The normal values for 24-hr urine calcium is
- A. <4mmol/1.73 m²
 - B. >4mmol/1.73 m²
 - C. <4 mg/kg
 - D. >4 mg/kg
 - E. = 4 mg/kg

44. Secondary hyperoxaluria can occur in patients with
- A. furosemide therapy
 - B. sarcoidosis
 - C. pyridoxine deficiency
 - D. primary hyperparathyroidism
 - E. immobility
45. Cystinuria is characterized by
- A. X-linked recessive inheritance
 - B. decreased cystine urinary excretion
 - C. renal tubular acidosis
 - D. alkaline urine
 - E. faint radiopaque renal calculi
46. Struvite calculi (secondary to urinary tract infections) are **MOST** likely caused by
- A. Escherichia coli
 - B. Proteus
 - C. Klebsiella
 - D. Pseudomonas
 - E. Staph. aureus
47. Nephrocalcinosis refers to calcium deposition within the renal tissue. All of the following can cause nephrocalcinosis **EXCEPT**
- A. furosemide
 - B. distal RTA
 - C. hypoparathyroidism
 - D. hyperoxaluria
 - E. Cushing syndrome
48. Dietary treatment of renal or ureteral calculi include
- A. increased dietary intake of sodium
 - B. reduced dietary intake in potassium
 - C. increased protein diet
 - D. encourage lemon juice intake
 - E. calcium restriction
49. Which of the following drugs can precipitate and form renal stones?
- A. acetazolamide
 - B. indanivir
 - C. probenecid
 - D. theophylline
 - E. vitamin C

Urologic Disorders in Infants and Children ANSWERS

QAHTAN ALOBAIDY

- 1.(B).
- 2.(B). Oligohydramnios.
- 3.(C). Death is due to pulmonary insufficiency from pulmonary hypoplasia rather than renal failure.
- 4.(E).
- 5.(E). Wilms tumors are 4 times more common in children with horseshoe kidneys than in the general population, stone disease and hydronephrosis secondary to ureteropelvic junction obstruction are other potential late complications.
- 6.(A). Female gender and uncircumcised male.
- 7.(C).
- 8.(D). Symptoms include dysuria, urgency, frequency, suprapubic pain, incontinence, and malodorous urine. Cystitis does not cause fever and does not result in renal injury; malodorous urine is not specific for a UTI.
- 9.(D).
- 10.(C). Others distracters are nonspecific markers of inflammation.
- 11.(D). Nitrofurantoin should not be used routinely in children with a febrile UTI because it does not achieve significant renal tissue levels.
- 12.(E). Microscopic hematuria is common in acute cystitis, but micro hematuria alone does not suggest UTI.
- 13.(C). The mean age at VUR resolution is 6yr.
- 14.(A). Idiopathic VUR appears to be an autosomal dominant inherited trait with variable penetrance, 80% are females, primary VUR also may be discovered during evaluation for antenatal hydronephrosis, in this select population, 80% of affected children are male. Bladder and bowel dysfunction (constipation) may be present in 50% of children with reflux and the average age at diagnosis is 2-3 yr.
- 15.(E). VUR occurring during bladder filling is termed low-pressure VUR; VUR during voiding is termed high-pressure VUR. VUR in children with low-pressure VUR is significantly less likely to resolve spontaneously than in children who exhibit only high-pressure VUR. Radiation exposure during a radionuclide cystogram is significantly less than that from a contrast VCUG.
- 16.(C).
- 17.(D). VCUG is necessary because 10-15% of patients have ipsilateralvesicoureteral reflux.
- 18.(D).

19.(E). This anomaly is 3 times as common in girls as in boys. In girls, approximately 35% of these ureters enter the urethra at the bladder neck, 35% enter the urethra vaginal septum, 25% enter the vagina, and a few drain into the cervix, uterus, Gartner duct, or a urethral diverticulum. In boys, ectopic ureters enter the posterior urethra (above the external sphincter) in 47%, the prostatic utricle in 10%, the seminal vesicle in 33%, the ejaculatory duct in 5%, and the vas deferens in 5%. UTI is common because of urinary stasis.

20.(C). Ureterocele is much more common in girls than in boys. Affected children usually are discovered by prenatal ultrasonography. Rarely, large ectopic ureteroceles can cause bladder outlet obstruction. Ureterocele may be ectopic, in which case the cystic swelling extends through the bladder neck into the urethra, or orthotopic, in which case the ureterocele is entirely within the bladder.

21.(D). 95% of affected children are male, oligohydramnios, very large bladder.

22.(D). Foley (balloon) catheter should not be used, because the balloon can cause severe bladder spasm, which can produce severe ureteral obstruction, affecting 1 in 8,000 boys. Vesicoureteral reflux occurs in 50% of patients and the urinary stream is weak. The urethral valves are tissue leaflets fanning distally from the prostatic urethra to the external urinary sphincter.

23.(C). The umbilicus is displaced downward, the anus is displaced anteriorly in both sexes, and the wide separation of the pubic rami causes a characteristic broad-based gait but no significant disability. In boys, there is complete epispadias with dorsal chordee; girls also have epispadias, with separation of the 2 halves of the clitoris and wide separation of the labia.

24.(B). Treatment includes reduction of bladder pressure with anticholinergic drugs (e.g., oxybutynin, 0.2 mg/kg/24 hr in 2 or 3 divided doses).

25.(B). The enteric mucosal surface in contact with the urine absorbs ammonium, chloride, and hydrogen ions and loses potassium; hyperchloremic metabolic acidosis can result, possibly requiring medical treatment.

26.(A).

27.(A).

28.(E). In posterior urethral valves the urinary stream is weak. Hypospadias is typically ventral deflection or severe splaying. Meatal stenosis; if the meatus is pinpoint, boys void with a forceful, fine stream that goes a great distance. Ureteral ectopia characterized by constant urinary dripping all day.

29.(E). This condition is termed the daytime frequency syndrome of childhood or Pollakiuria; without dysuria, UTI, daytime incontinence, or nocturia. The most common age for these symptoms to occur is 4-6 yr.

30.(C).

31.(E).

32.(B). This form of therapy has a reported success of 30-60%, although the relapse rate is significant, while motivational therapy; there is no evidence that this approach is

beneficial, Pharmacologic therapy is intended to treat the symptom of enuresis and thus is regarded as second line and is not curative.

33.(E). In most cases, penile development is normal, and the condition is unrecognized until circumcision is performed or the foreskin is retractable

34.(A). Others syndromes associated with syndactyly.

35.(B). Non ischemic (high-flow) priapism most commonly follows perineal trauma, such as a straddle injury, while others are causes of ischemic (venoocclusive, low-flow) priapism.

36.(C).

37.(B). Retractable testes may be misdiagnosed as undescended testes.

38.(C).

39.(A). That's why treatment of renal or ureteral calculus by urinary alkalization (the urine pH should be ≥ 6.5). Inorganic (e.g., citrate, magnesium) and organic (e.g., glycosaminoglycan's, osteopontin) substances are known to inhibit stone formation. Organic inhibitory compounds adsorb to the surface of the crystal, thereby inhibiting crystal growth and nucleation.

40.(C). If the stone passes into the bladder, the child usually is asymptomatic. If the stone is in the urethra, dysuria and difficulty voiding can result, particularly in boys.

41.(A). Others distracters are causes of calcium stones.

42.(C). This study takes only a few minutes to perform, has 96% sensitivity and specificity in delineating the number and location of calculi, and demonstrates whether the involved kidney is hydronephrotic or not.

43.(C). Hypercalciuria if >4 mg/kg/24 hr.

44. (C). Others distracters are causes of Hypercalciuria.

45.(E). Autosomal recessive disorder of the epithelial cells of the renal tubule that prevents absorption of the 4 dibasic amino acids (cystine, ornithine, arginine, lysine) and results in excessive urinary excretion of these products. The only known complication of this familial disease is the formation of calculi, because of the low solubility of cystine; the patients usually have acidic urine, which leads to a higher rate of precipitation.

46.(B). Caused by urea-splitting organisms (most often *Proteus* spp., and occasionally *Klebsiella* spp., *Escherichia coli*, *Pseudomonas* spp., and others).

47.(C). Hyperparathyroidism.

48.(D). An excellent source of citrate (inhibitor of calcium stones) is lemonade, reduction in dietary intake of sodium and increased potassium intake is indicated, low-protein diets reduce urinary calcium and oxalate excretion, and calcium restriction in children should be avoided.

49.(B). All the others distractors increase the risk of stone formation but do not themselves precipitate as stones.

1. The least likely cause of Infectious vulvovaginitis is
 - A. Escherichia coli
 - B. Staphylococcus aureus
 - C. Haemophilus influenza
 - D. Enterobius vermicularis
 - E. Candida spp.
2. Amoxicillin-clavulanate is the first-line treatment in vulvovaginal infections caused by
 - A. Streptococcus pyogenes
 - B. Staphylococcus aureus
 - C. Haemophilus influenza
 - D. Shigella
 - E. Chlamydia trachomatis
3. Ultra potent topical corticosteroids is the first-line therapy in
 - A. labial agglutination
 - B. lichen sclerosus
 - C. psoriasis
 - D. atopic dermatitis
 - E. seborrheic dermatitis
4. All the following are first-line treatment of diaper dermatitis **EXCEPT**
 - A. frequent diaper changes
 - B. increasing diaper free period
 - C. frequent bathing
 - D. antifungal treatment
 - E. zinc oxide
5. Vulvar dermatologic condition in children which necessitate evaluation of thyroid function is
 - A. lichen sclerosus
 - B. psoriasis
 - C. vitiligo
 - D. seborrheic dermatitis
 - E. atopic dermatitis

6. The gold standard diagnostic investigation for precocious puberty is
- A. Tanner maturing rate
 - B. left wrist x-ray
 - C. serum luteinizing hormone levels
 - D. gonadotropins measurement
 - E. brain MRI
7. Lack of development of the breast is considered delayed and warrants endocrinology evaluation by the age of
- A. 10 yr
 - B. 11 yr
 - C. 12 yr
 - D. 13 yr
 - E. 14 yr
8. Amastia (complete absence of the breast) associated with all the following **EXCEPT**
- A. Poland syndrome
 - B. ectodermal dysplasia
 - C. Crohn disease
 - D. congenital adrenal hypoplasia
 - E. hypogonadotropic hypogonadism
9. Of the following, the **MOST** likely cause of bloody nipple discharge in infants is
- A. chronic nipple irritation
 - B. ducts of Montgomery
 - C. hypothalamic tumors
 - D. Intraductal cysts
 - E. mammary duct ectasia
10. The **MOST** common cause of breast pain in adolescents is
- A. benign fibroadenoma
 - B. exercise
 - C. mastitis
 - D. lipoma
 - E. Intraductal papilloma
11. Of the following, the **MOST** common solid mass seen in adolescent girls is
- A. lipoma
 - B. fibroadenoma
 - C. lymphangioma
 - D. hematoma
 - E. hamartoma

12. The **MOST** common tumor metastasized to the breast is
- A. rhabdomyosarcoma
 - B. lymphoblastic leukemia
 - C. neuroblastoma
 - D. lymphoma
 - E. cystosarcomaphyllodes
13. The imaging modality of choice for breast abnormalities in the pediatric population is
- A. mammography
 - B. CT scan
 - C. ultrasonography
 - D. MRI
 - E. PET scan
14. All the following may cause hirsutism **EXCEPT**
- A. hydrochlorothiazide
 - B. acetazolamide
 - C. cyclophosphamide
 - D. anabolic steroids
 - E. penicillamine
15. Chocolate cysts are seen in
- A. functional cysts
 - B. endometriomas
 - C. cyst adenomas
 - D. teratomas
 - E. ovarian carcinoma
16. 5-yr survival is 100% in the following germ cell tumors
- A. gonadoblastoma
 - B. embryonal carcinoma
 - C. choriocarcinoma
 - D. dysgerminoma
 - E. endodermal sinus tumor
17. The **MOST** common ovarian malignancy is
- A. gonadoblastoma
 - B. embryonal carcinoma
 - C. choriocarcinoma
 - D. dysgerminoma
 - E. endodermal sinus tumor

18. The tumor marker (α -fetoprotein) is used for the diagnosis of all the following ovarian cancers **EXCEPT**

- A. Immature teratoma
- B. embryonal carcinoma
- C. mixed germ cell
- D. dysgerminoma
- E. endodermal sinus tumor

19. Pathogenesis of leiomyosarcoma is thought to be correlated with

- A. Epstein-Barr virus
- B. haemophilus influenzae
- C. enterococcus
- D. nocardia
- E. Yersinia

20. Approved treatment for condyloma acuminata of the vulva in children include all the following **EXCEPT**

- A. topical trichloroacetic acid
- B. local cryotherapy
- C. electro cautery
- D. laser ablation
- E. sinecatechins ointment

21. Hemihematometra is described as

- A. accumulation of mucus or nonsanguineous fluid in the vagina
- B. atretic segment of vagina with menstrual fluid accumulation
- C. accumulation of serous fluid in the fallopian tube
- D. one cervix associated with 2 uterine horns
- E. the result of failure of 1 müllerian duct to descend

22. The **MOST** sensitive and specific imaging technique used for evaluating müllerian anomalies is

- A. ultrasound
- B. hysterosalpingogram
- C. sonohysterography (saline-infusion sonography)
- D. MRI
- E. CT scan

23. The **MOST** common structural uterine anomaly is

- A. uterine septum
- B. bicornuate uterus
- C. unicornuate uterus

- D. uterine didelphys
- E. arcuate uterus

24. An imperforate hymen is characterized by all the following **EXCEPT**
- A. the incidence is approximately 1 in 1,000
 - B. normal secondary sex characters
 - C. more often it is diagnosed at the time of menarche
 - D. cannot be diagnosed in the newborn period
 - E. primary amenorrhea

- 1.(E).
- 2.(B). In cases of treatment failure in H influenza or non-encapsulated H. influenzae, amoxicillin–clavulanate is recommended.
- 3.(B).
- 4.(D). If diaper dermatitis persists after these conservative measures, or if the classic satellite lesions of candida are present, treatment with an antifungal can decrease the inflammation.
- 5.(C). Although diagnosis is clinical, there is an association with other autoimmune or endocrine disorders (hypothyroidism, Graves' disease, Addison disease, pernicious anemia, and insulin-dependent diabetes mellitus) and workup should include evaluation for at least thyroid dysfunction.
- 6.(D). Measurement of gonadotropins after gonadotropin-releasing hormone or gonadotropin-releasing hormone–agonist stimulation.
- 7.(D).
- 8.(D). Congenital adrenal hyperplasia
- 9.(E).
- 10.(B). Physiologic swelling and tenderness occur on a cyclic basis, most commonly during the premenstrual phase, and are secondary to hormonal stimulation and resulting proliferative changes. Hormonal imbalance can cause exaggerated responses in the breast tissue, especially in the upper and outer quadrants. Nodularity, poorly localized tenderness, and a soreness radiating to the axilla and arm are usual accompanying findings.
- 11.(B).
- 12.(A). Breast tumors also may be the first manifestation of relapse (extra medullary) in acute lymphoblastic leukemia.
- 13.(C). Because the dense breast tissue of the adolescent obstructs the visualization of a palpable mass, mammography is not advised for this age group.
- 14.(C). Cause alopecia.
- 15.(B).
- 16.(A).
- 17.(A).
- 18.(D). In dysgerminoma use E2, estradiol, hCG (human chorionicgonadotropin), and LDH (lactate dehydrogenase) as a tumor markers.
- 19.(A).

20.(E). Some products used to treat skin lesions in adults have not been approved for children, including provider application of podophyllin resin and home application of imiquimod, podofilox, and sinecatechins ointment.

21.(B).

22.(D). Because it can image nearly all reproductive structures, blood flow, external contours, junctional zone resolution on T2-weighted images, and associated renal and other anomalies. MRI also has a high correlation with surgical findings because of its multiplanar capabilities and high spatial resolution.

23.(A).

24.(D). In the newborn period and early infancy, it may be diagnosed by a bulging membrane caused by a mucocolpos from maternal estrogen stimulation of the vaginal mucosa.

The Endocrine System QUESTIONS

ZUHAIR ALMUSAWI

1. Main causes of hypocalcemia in nephrotic syndrome in children are the following **EXCEPT**
 - A. decreased activity of renal α -hydroxylase
 - B. lowered serum albumin
 - C. decreased intestinal absorption of calcium
 - D. increased urinary losses of cholecalciferol-binding globulin
 - E. prednisone therapy

2. The following criteria are needed for the diagnosis of syndrome of inappropriate secretion of antidiuretic hormone (SIADH) **EXCEPT**
 - A. excessive urinary sodium concentration
 - B. hyponatremia with normal serum osmolality
 - C. elevated urine osmolality
 - D. normal renal, adrenal, and thyroid function
 - E. absence of volume depletion

3. The following are main features of diabetic ketoacidosis (DKA) **EXCEPT**
 - A. hyperglycemia (glucose usually >300 mg/dL)
 - B. ketonemia (serum ketones >3 mmol/L)
 - C. ketonuria
 - D. venous pH <7.20
 - E. serum $\text{HCO}_3^- < 15$ mEq/L

4. What percent of newly diagnosed diabetics present with symptoms of diabetic ketoacidosis (DKA)?
 - A. 10%
 - B. 20%
 - C. 30%
 - D. 40%
 - E. 50%

5. Typical insulin dosage requirement in prepubertal children after the "honeymoon" period is
 - A. 0.4 U/kg/day
 - B. 0.5 U/kg/day
 - C. 0.6 U/kg/day

- D. 0.7 U/kg/day
- E. 1 U/kg/day

6. The following features suggest constitutional delay as a cause of short stature **EXCEPT**

- A. no signs or symptoms of systemic disease
- B. bone age delayed beyond the height age
- C. period of poorest growth often occurring between the ages of 18 and 30 months
- D. parental or sibling history of delayed development
- E. height predictions consistent with family characteristics

7. Congenital goiter is seen in which percent of newborns with congenital hypothyroidism?

- A. 10%
- B. 20%
- C. 30%
- D. 40%
- E. 50%

8. During a routine physical examination, a solitary thyroid nodule is palpated in asymptomatic 10-year-old child.

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

- A. thyroid carcinoma
- B. thyroid adenoma
- C. thyroid abscess
- D. thyroid cyst
- E. subacute thyroiditis

9. The pituitary gland is located in a saddle-shaped cavity of the

- A. frontal bone
- B. maxillary bone
- C. sphenoid bone
- D. ethmoid bone
- E. temporal bone

10. Physiologic factors play a role in stimulating and inhibiting growth hormone (GH).

One of the following inhibit GH release

- A. sleep
- B. exercise
- C. hyperglycemia
- D. acute illness

- E. fasting
11. Corticotropin-releasing hormone (CRH) and ACTH release are inhibited by
- A. arginine vasopressin
 - B. oxytocin
 - C. angiotensin II
 - D. atrial natriuretic peptide
 - E. cholecystokinin
12. The **MOST** common cause of acquired hypopituitarism is
- A. craniopharyngioma
 - B. eosinophilic granuloma (histiocytosis)
 - C. tuberculosis
 - D. toxoplasmosis
 - E. meningitis
13. Indications for GH treatment to promote growth include the following **EXCEPT**
- A. GH deficiency
 - B. chronic renal failure before transplantation
 - C. celiac disease
 - D. Prader-Willi syndrome
 - E. Noonan syndrome
14. Reported side effects of GH treatment include the following **EXCEPT**
- A. pseudotumor cerebri
 - B. slipped capital femoral epiphysis
 - C. gynecomastia
 - D. worsening of scoliosis
 - E. 6-fold increase in the risk for type 1 diabetes
15. The following stimulate vasopressin (VP) secretion **EXCEPT**
- A. hyperosmolality
 - B. hypovolemia
 - C. hypotension
 - D. hyperglycemia
 - E. nausea
16. Acquired nephrogenic diabetes insipidus (NDI) is associated with the following **EXCEPT**
- A. lithium
 - B. vancomycin
 - C. amphotericin

- D. methicillin
- E. rifampin

17. Neonates and young infants with central diabetes insipidus are often best treated with

- A. fluid therapy
- B. vasopressin analogs
- C. thiazide diuretics
- D. indomethacin
- E. amiloride

18. Hyponatremia in cerebral salt wasting is accompanied by the following **EXCEPT**

- A. elevated urinary sodium excretion
- B. low urine output
- C. hypovolemia
- D. normal or high uric acid
- E. suppressed vasopressin

19. Broad forehead, hypertelorism, small chin, long philtrum, camptodactyly, and fetal finger pads are features of the following genetic overgrowth syndrome

- A. Perlman syndrome
- B. Sotos syndrome
- C. Weaver syndrome
- D. Beals syndrome
- E. Beckwith-Wiedemann syndrome (BWS)

20. Precocious puberty in girls is defined by the onset of secondary sexual characteristics before the age of

- A. 7 yr
- B. 8 yr
- C. 9 yr
- D. 10 yr
- E. 11 yr

21. The **MOST** common brain lesion causing central precocious puberty is

- A. postencephalitic scar
- B. tuberculous meningitis
- C. hypothalamic hamartoma
- D. tuberous sclerosis
- E. hydrocephalus

22. All the following are true in premature thelarche **EXCEPT**

- A. most often appears in the 1st 2 yr of life
- B. growth and osseous maturation are usually normal
- C. menarche occurs at the expected age
- D. may be unilateral
- E. often persists till puberty

23. Premature adrenarche has traditionally applied to the appearance of sexual hair before the age of 8 yr in girls or 9 yr in boys without other evidence of maturation.

It is characterized by the following **EXCEPT**

- A. more frequent in girls
- B. higher prevalence in african-american
- C. axillary hair generally appears earlier
- D. adult-type axillary odor is common
- E. affected children are often slightly advanced in height

24. The **MOST** accurate test of thyroid function is

- A. T4
- B. free T4
- C. T3
- D. thyroglobulin
- E. TSH

25. Peak serum concentrations of TSH at birth in full-term infants reach

- A. 20 mU/L
- B. 40 mU/L
- C. 60 mU/L
- D. 80 mU/L
- E. 100 mU/L

26. The thyroid hormones are transported in plasma bound to thyroxine-binding globulin (TBG), a glycoprotein synthesized in the liver. TBG binds approximately 70% of T4 and 50% of T3. TBG level increase with administration of

- A. estrogens
- B. androgens
- C. glucocorticoids
- D. nicotinic acid
- E. l-asparaginase

27. The **MOST** common cause of permanent congenital hypothyroidism is

- A. dyshormonogenesis
- B. thyroid dysgenesis
- C. iodine deficiency

- D. defect of iodide transport
- E. Pendred syndrome

28. The cord serum T4 is decreased in proportion to gestational age and birthweight. The serum T4 gradually increases and enters the T4 range seen in term infants by the age of

- A. 2 wk
- B. 4 wk
- C. 6 wk
- D. 8 wk
- E. 10 wk

29. Approximately 10% of infants with congenital hypothyroidism have associated congenital anomalies.

Of the following, the **MOST** common are

- A. nervous system anomalies
- B. lungs anomalies
- C. eye anomalies
- D. cardiac anomalies
- E. genitourinary anomalies

30. The normal level of serum thyroid-stimulating hormone in first week of term Infants is up to

- A. 4.4 mIU/L
- B. 9.5 mIU/L
- C. 13.6 mIU/L
- D. 17.6 mIU/L
- E. 27.0 mIU/L

31. Retardation of osseous development in congenitally hypothyroid infants can be shown radiographically at birth in approximately

- A. 20%
- B. 40%
- C. 60%
- D. 80%
- E. 100%

32. Levothyroxine (l-T4) given orally is the treatment of choice in congenital hypothyroidism.

The recommended initial starting dose is

- A. 2-5 µg/kg/day
- B. 4-7 µg/kg/day

- C. 5-9 µg/kg/day
 - D. 8-12 µg/kg/day
 - E. 10-15 µg/kg/day
33. The **MOST** common cause of acquired hypothyroidism is
- A. craniopharyngioma
 - B. Hashimoto thyroiditis
 - C. meningoencephalitis
 - D. drug-induced
 - E. irradiation
34. Antithyroid antibodies develop and subclinical or overt hypothyroidism occurs in the following conditions **EXCEPT**
- A. Down syndrome
 - B. Turner syndrome
 - C. type 1 diabetes mellitus
 - D. Sjögren syndrome
 - E. Williams syndrome
35. Risk for subclinical hypothyroidism is seen in children with
- A. chronic hepatitis B infection
 - B. chronic hepatitis C infection
 - C. chronic hepatitis D infection
 - D. hepatitis A infection
 - E. hepatitis E infection
36. The first clinical manifestation of acquired hypothyroidism is
- A. poor schoolwork
 - B. goiter
 - C. deceleration of growth
 - D. weight gain
 - E. constipation
37. One of the following is not routinely done in children with suspected hypothyroidism
- A. sonography
 - B. serum free T4
 - C. TSH
 - D. antithyroglobulin
 - E. antiperoxidase antibodies

38. Chronic lymphocytic thyroiditis is the most common cause of acquired hypothyroidism, with or without goiter.

All the following are true **EXCEPT**

- A. more common in girls
- B. peak incidence during adolescence
- C. most of the affected children are asymptomatic
- D. clinical course is constant
- E. familial clusters are common

39. Thyroid peroxidase antibodies are absent in the following thyroiditis syndrome

- A. Hashimoto thyroiditis
- B. painless sporadic thyroiditis
- C. painful subacute thyroiditis
- D. acute suppurative thyroiditis
- E. Riedel thyroiditis

40. Endemic cretinism is the most serious consequence of iodine deficiency; it includes two different but overlapping syndromes: a neurologic type and a myxedematous type.

The neurologic syndrome is characterized by the following **EXCEPT**

- A. intellectual disability
- B. deaf-mutism
- C. disturbances in gait
- D. patellar hyperreflexia
- E. delayed pubertal development

41. All the following conditions are associated with Graves' disease **EXCEPT**

- A. type 1 diabetes mellitus
- B. vitiligo
- C. iron deficiency anemia
- D. alopecia areata
- E. celiac disease

42. Adverse reaction reported exclusively with propylthiouracil is

- A. severe liver disease
- B. agranulocytosis
- C. lupus-like polyarthritis syndrome
- D. glomerulonephritis
- E. antineutrophilic cytoplasmic antibody–positive vasculitis

43. A painless nodule in the thyroid or in the neck is the usual presentation of childhood thyroid cancer.

Of the following, the **MOST** common site of distant metastasis is

- A. mediastinum
- B. long bones
- C. skull
- D. brain
- E. lungs

44. Hypocalcemia is common in neonates between 12 and 72 hr of life, especially in infants with

- A. birth asphyxia
- B. sepsis
- C. exchange transfusion
- D. hypomagnesemia
- E. hyperbilirubinemia

45. Aplasia or hypoplasia of the parathyroid glands is often associated with the DiGeorge/velocardiofacial syndrome. All the following regarding this syndrome are true **EXCEPT**

- A. occurs in 1 in 4,000 newborns
- B. caused by a deletion of chromosome 22q11.2
- C. hypocalcemia is transitory in the majority
- D. hypocalcemia can have its onset later in life
- E. conotruncal defects of the heart occur in 75% of cases

46. There is a spectrum of parathyroid deficiencies with clinical manifestations varying from no symptoms to those of complete and longstanding deficiency.

Early manifestations include

- A. tingling of the hands and feet
- B. muscular pain and cramps
- C. carpopedal spasms
- D. convulsions
- E. delayed irregular teeth eruption

47. Laboratory findings in hypoparathyroidism include the following **EXCEPT**

- A. low serum calcium (5-7 mg/dL)
- B. elevated serum phosphorus (7-12 mg/dL)
- C. elevated serum alkaline phosphatase
- D. level of 1,25(OH)₂D₃ is usually low
- E. normal serum magnesium

48. Episodic symptomatic hypocalcemia occurs in the Kenny-Caffey syndrome, the latter is characterized by the following **EXCEPT**

- A. medullary stenosis of the long bones

- B. short stature
- C. delayed closure of the fontanel
- D. advanced bone age
- E. eye abnormalities

49. Type Ia accounts for the majority of patients with pseudohypoparathyroidism (PHP), they have the following skeletal abnormalities **EXCEPT**

- A. brachydactyly
- B. dimpling of the dorsum of the hand
- C. the three middle fingers are the same length
- D. bowing
- E. thickening of the calvaria

50. Parathyroid crisis is manifested by the following **EXCEPT**

- A. serum calcium levels >15 mg/dL
- B. progressive polyuria
- C. azotemia
- D. stupor
- E. coma

51. The **MOST** consistent and characteristic radiographic finding of primary hyperparathyroidism is

- A. gross trabeculation of the skull
- B. generalized rarefaction
- C. cysts
- D. resorption of subperiosteal bone
- E. radiographic signs of rickets

52. All the following are causes of infantile hypercalcemia **EXCEPT**

- A. subcutaneous fat necrosis
- B. metaphyseal chondrodysplasia
- C. lactase/disaccharidase deficiency
- D. blue diaper syndrome
- E. proximal renal tubular acidosis

53. Glucocorticoids play a major role in immune regulation. They increase circulating

- A. polymorphonuclear cell
- B. monocytes
- C. eosinophils
- D. lymphocytes
- E. basophils

54. Glucocorticoids readily penetrate the blood–brain barrier and have direct effects on brain metabolism. They have the following actions **EXCEPT**

- A. decrease brain edema
- B. insomnia
- C. increased irritability
- D. impaired memory
- E. improved ability to concentrate

55. Patients with mineralocorticoid deficiency can develop the following **EXCEPT**

- A. weight loss
- B. hypotension
- C. metabolic alkalosis
- D. hyponatremia
- E. hyperkalemia

56. The **MOST** common cause of primary adrenal insufficiency in infancy is

- A. congenital adrenal hyperplasia
- B. isolated glucocorticoid deficiency
- C. X-linked adrenal hypoplasia congenita
- D. Lemli-opitz syndrome
- E. adrenoleukodystrophy

57. The clinical presentation of adrenal insufficiency depends on the age of the patient, the usual presentation of adrenal insufficiency in infancy is

- A. ketosis
- B. hyperpigmentation
- C. orthostatic hypotension
- D. hypoglycemia
- E. hypernatremia

58. The **MOST** definitive test for adrenal insufficiency is measurement of

- A. blood sugar
- B. cortisol before and after administration of ACTH
- C. serum sodium
- D. arterial blood gases
- E. urinary excretion of sodium and chloride

59. Patients with autoimmune Addison disease must be closely observed for the development of other autoimmune disorders.

Of the following, the **MOST** commonly associated disorder in children is

- A. alopecia
- B. vitiligo

- C. chronic active hepatitis
- D. type 1 diabetes mellitus
- E. hypoparathyroidism

60. High-dose glucocorticoids (the equivalent of >10 times physiologic cortisol secretion) can be administered without requiring a subsequent tapering for

- A. 5 day
- B. 7 day
- C. 10 day
- D. 14 day
- E. 21 day

61. Classic 21-hydroxylase deficiency occurs in approximately 1 in 15,000- 20,000 births in most populations. Of affected infants, the salt-losing form constitute about

- A. 40%
- B. 50%
- C. 60%
- D. 70%
- E. 80%

62. The signs and symptoms of congenital adrenal hyperplasia (salt wasting form of the disease) caused by 21-hydroxylase deficiency typically first develop in affected infants at approximately

- A. 1 week of age
- B. 2 weeks of age
- C. 3 weeks of age
- D. 4 weeks of age
- E. 6 weeks of age

63. Significantly virilized females with congenital adrenal hyperplasia (CAH) usually undergo surgery between

- A. 2- 6 mo of age
- B. 7-12 mo of age
- C. 13-24 mo of age
- D. 5-9 year of age
- E. 10-14 year of age

64. Cortisol deficiency in congenital adrenal hyperplasia (salt wasting form of the disease) is treated with hydrocortisone

- A. 5-10 mg/m²/24 hr
- B. 10-15 mg/m²/24 hr
- C. 15-20 mg/m²/24 hr

- D. 20-25 mg/m²/24 hr
- E. 25-30 mg/m²/24 hr

65. In older children with Cushing syndrome, in addition to obesity, a common early manifestation is

- A. purplish striae on abdomen
- B. short stature
- C. hypertension
- D. hyperglycemia
- E. osteoporosis

66. All the following tests are of diagnostic utility in the diagnosis of Cushing syndrome **EXCEPT**

- A. midnight cortisol levels
- B. nighttime salivary cortisol levels
- C. glucose tolerance test
- D. urinary excretion of free cortisol
- E. dexamethasone suppression test

67. In primary aldosteronism, one of the following is low

- A. serum PH
- B. serum carbon dioxide
- C. serum sodium
- D. serum chloride
- E. serum calcium

68. Clinical manifestations of pheochromocytomas include the following **EXCEPT**

- A. hypertension
- B. convulsions
- C. pulmonary edema
- D. obesity
- E. good appetite

69. Calcification within the adrenal glands may occur in a wide variety of situations, some serious and others of no obvious consequence.

Adrenal calcifications are bilateral in

- A. neuroblastomas
- B. ganglioneuromas
- C. Wolman disease
- D. cortical carcinomas
- E. pheochromocytomas

70. The major causes of incidentalomas are
- A. benign adenomas
 - B. pheochromocytomas
 - C. adrenocortical carcinoma
 - D. hemorrhagic cysts
 - E. myelolipomas
71. Congenital hypogonadotropic hypogonadism include
- A. gonadal dysgenesis
 - B. Klinefelter syndrome (47,XXY)
 - C. Noonan syndrome
 - D. cystic fibrosis
 - E. Laurence-Moon- Biedl syndrome
72. Acquired hypogonadotropic hypogonadism include
- A. chemotherapy
 - B. malnutrition
 - C. cystic fibrosis
 - D. infection (e.g., mumps)
 - E. infarction (testicular torsion)
73. Klinefelter syndrome is the most common sex chromosomal aneuploidy in males, all the following features are true **EXCEPT**
- A. 80% of them have a 47, xxy chromosome complement
 - B. diagnosis is rarely made before puberty
 - C. psychiatric disorders may be apparent long before defects in sexual development
 - D. few complete high school
 - E. approximately 80% of adults have gynecomastia
74. True gynecomastia is characterized by the presence of a palpable fibroglandular mass (located concentrically beneath the nipple and areolar region) at least
- A. 0.5 cm in diameter
 - B. 1 cm in diameter
 - C. 1.5 cm in diameter
 - D. 2 cm in diameter
 - E. 2.5 cm in diameter
75. Weaker associations with gynecomastia are seen in
- A. spironolactone
 - B. alkylating agents
 - C. ketoconazole

- D. cimetidine
- E. opiates

76. Significant gynecomastia is seen in

- A. hyperthyroidism
- B. Klinefelter syndrome
- C. 17-ketosteroid reductase deficiency
- D. 11 β -hydroxylase deficiency
- E. Peutz-Jeghers syndrome

77. All prepubertal gynecomastia cases, as well as pubertal cases with suspicious features, should be investigated.

Of the following, the initial laboratory evaluation should include

- A. karyotype
- B. dehydroepiandrosterone sulfate
- C. liver function tests
- D. thyroid function tests
- E. renal function tests

78. Many patients with Turner syndrome are recognizable at birth because of

- A. loose skin folds at the nape of the neck
- B. cubitus valgus
- C. webbing of the neck
- D. widely spaced nipples
- E. hyperconvex fingernails

79. The **MOST** common cardiac defect in Turner syndrome is

- A. aortic coarctation
- B. isolated nonstenotic bicuspid aortic valves
- C. aortic stenosis
- D. mitral valve prolapse
- E. anomalous pulmonary venous drainage

80. The 47,XXX (trisomy) chromosomal constitution is the most frequent extra-X chromosome abnormality in females, occurring in almost 1 in 1,000 liveborn females.

It is characterized by the following **EXCEPT**

- A. maternal meiotic nondisjunction is the most common cause
- B. normal female phenotype
- C. sexual development and menarche are delayed
- D. tall and gangly
- E. behavior disorders

81. The **MOST** common cardiac defect in Noonan syndrome is
- A. aortic coarctation
 - B. pulmonary valvular stenosis
 - C. aortic stenosis
 - D. mitral valve prolapse
 - E. anomalous pulmonary venous drainage
82. Familial isolated gonadotropin deficiency associated with anosmia is
- A. Noonan syndrome
 - B. Bloom syndrome
 - C. Werner syndrome
 - D. Kallmann syndrome
 - E. Prader-Willi
83. Children with type 2 DM often seek medical care because of
- A. weight loss
 - B. excessive weight gain
 - C. polyuria
 - D. polydipsia
 - E. anorexia
84. The current criteria for the diagnosis of type 1 and type 2 DM is a fasting blood glucose that exceeds
- A. 110 mg/dL
 - B. 115 mg/dL
 - C. 120 mg/dL
 - D. 125 mg/dL
 - E. 130 mg/dL
85. The clearest evidence of a role for viral infection in human type 1 DM is seen in
- A. acquired rubella infection
 - B. live-virus rubella immunization
 - C. enteroviral infection
 - D. congenital rubella syndrome
 - E. mumps infection
86. The following factors are implicated in the pathogenesis of type 1 DM **EXCEPT**
- A. prenatal influences
 - B. diet in infancy
 - C. viral infections
 - D. excessive exposure to certain infections
 - E. psychologic stress

87. Falsely low HbA_{1c} levels are noted in the following conditions **EXCEPT**

- A. aplastic anemia
- B. pure red cell aplasia
- C. blood transfusions
- D. cirrhosis
- E. renal disease treated with erythropoietin

88. Subjects with type 1 DM have evidence for celiac disease in

- A. 5-10%
- B. 11-15%
- C. 16-20%
- D. 21-25%
- E. 26-30%

89. Diabetic ketoacidosis (DKA) is the end result of the metabolic abnormalities resulting from a severe deficiency of insulin or insulin effectiveness.

DKA is characterized by the following **EXCEPT**

- A. ketonuria
- B. normal ion gap
- C. decreased serum bicarbonate
- D. decreased pH
- E. elevated effective serum osmolality

90. To continue the insulin infusion in treatment of DKA without causing hypoglycemia, glucose must be added to the infusion. We typically recommend that glucose be added as a 10% solution when the serum glucose has decreased

- A. <100 mg/dL
- B. <150 mg/dL
- C. <200 mg/dL
- D. <250 mg/dL
- E. <300 mg/dL

91. Diabetic ketoacidosis (DKA) may be arbitrarily classified as mild, moderate, or severe. Venous pH in severe DKA is

- A. <7.05
- B. <7.10
- C. <7.15
- D. <7.20
- E. <7.25

92. Any child with diabetic ketoacidosis (DKA) can be easily transitioned to oral intake and subcutaneous insulin when the following criteria are found **EXCEPT**

- A. total CO₂ >15 mEq/L
- B. pH >7.25
- C. sodium stable between 135 and 145 mEq/L
- D. no emesis
- E. no dehydration

93. Cerebral edema complicating DKA remains the major cause of morbidity and mortality in children and adolescents with T1 DM.

All the following are true **EXCEPT**

- A. early bolus administration of insulin is a risk factor
- B. high volumes of fluid is a risk factor
- C. the incidence of cerebral edema in children with DKA has not changed over the past 15-20 yr
- D. radiographic imaging is frequently helpful in making the diagnosis of cerebral edema
- E. its etiology remains unknown

94. Nonketotic hyperosmolar coma is characterized by the following **EXCEPT**

- A. severe hyperglycemia
- B. nonketotic acidosis
- C. severe dehydration
- D. hypothermia
- E. positive babinski signs

95. HbA_{1c} values may be spuriously elevated in

- A. thalassemia
- B. sickle cell disease
- C. iron deficiency anemia
- D. leukemia
- E. aplastic anemia

96. Microvascular complications of DM include

- A. nephropathy
- B. accelerated coronary artery disease
- C. cerebrovascular disease
- D. peripheral vascular disease
- E. peripheral neuropathies

97. Guidelines suggest that prepubertal T1DM patients should commence screening for retinopathy

- A. at diagnosis
- B. 1 yr after diagnosis

- C. 3yr after diagnosis
- D. 5 yr after diagnosis
- E. 10yr after diagnosis

98. Guidelines suggest that diabetic patients should commence screening for celiac disease

- A. at diagnosis
- B. 1 yr after diagnosis
- C. 3 yr after diagnosis
- D. 5 yr after diagnosis
- E. 10 yr after diagnosis

99. Transient elevation of urinary albumin can occur in the following conditions **EXCEPT**

- A. hypoglycemia
- B. strenuous exercise
- C. urinary tract infections
- D. heart failure
- E. acute febrile illness

100. Signs of insulin resistance or conditions associated with insulin resistance in T2 DM in children include the following **EXCEPT**

- A. acanthosis nigricans
- B. hypertension
- C. dyslipidemia
- D. increased waist : hip ratio
- E. polycystic ovary syndrome

101. The **MOST** commonly used and the only FDA-approved oral agent for the treatment of T2 DM in children and adolescents is

- A. thiazolidinediones
- B. pramlintide
- C. metformin
- D. sulfonylureas
- E. acarbose

102. Donohue syndrome is characterized by the following **EXCEPT**

- A. intrauterine growth restriction
- B. fasting hypoglycemia
- C. acanthosis nigricans
- D. postprandial hyperglycemia
- E. profound resistance to insulin

103. One of the following do not appear to be of concern in cystic fibrosis–related diabetes (CFRD)

- A. ketoacidosis
- B. macrovascular complications
- C. microvascular complications
- D. frequent infections
- E. energy needs

104. When diabetes and thyroid disease coexist, the possibility of autoimmune adrenal insufficiency should be considered. It may be heralded by the followings **EXCEPT**

- A. increasing insulin requirements
- B. increasing pigmentation of the skin
- C. salt craving
- D. asthenia
- E. postural hypotension

105. Therapy with one of the following drugs usually results in significant insulin resistance leading to glucose intolerance and overt diabetes

- A. streptozotocin
- B. rodenticide Vacor
- C. high-dose oral steroid
- D. cyclosporin
- E. tacrolimus

The Endocrine System

ANSWERS

ZUHAIR ALMUSAWI

- 1.(A). Decreased activity of renal α -hydroxylase (which is involved in converting the less-active 25-hydroxyvitamin D into the more active 1,25-(OH)₂ D) is a cause in renal insufficiency.
- 2.(B). Hyponatremia with reduced serum osmolality.
- 3.(D). Venous pH <7.30.
- 4.(C). 30%. The percentage is higher in younger children (<5 years old).
- 5.(B). Prepubertal children generally require about 0.5 U/kg/day. During the middle of puberty, dosages often exceed 1 U/kg/day, whereas postpubertal individuals require 0.75-1.0 U/kg/day. Athletes or those with a low caloric intake may require less insulin. For most children with type 1 diabetes, there are a variety of strategies and combinations of insulin that are currently used. There is a trend toward three or more doses of insulin each day. In addition, more children are on insulin pump therapy.
- 6.(B). Bone age delayed up to 2-4 years but consistent with height age
- 7.(B). Congenital goiter is seen in only 20% of newborns with congenital hypothyroidism.
- 8.(A). In children with a solitary nodule, about 30-40% has a carcinoma, 20-30% has an adenoma, and the remainder will have thyroid abscess, thyroid cyst, multinodular goiter, Hashimoto thyroiditis, subacute thyroiditis, or nonthyroidal neck mass.
- 9.(C). The pituitary gland is located at the base of the skull in a saddle-shaped cavity of the sphenoid bone called the sella turcica.
- 10.(C). Sleep, exercise, physical stress, trauma, acute illness, puberty, fasting, and hypoglycemia stimulate the release of GH whereas hyperglycemia, hypothyroidism, and glucocorticoids inhibit GH release.
- 11.(D). Arginine vasopressin, oxytocin, angiotensin II, and cholecystokinin stimulate release of CRH and ACTH, whereas atrial natriuretic peptide and opioids inhibit release of CRH and ACTH.
- 12.(A). Any lesion that damages the hypothalamus, pituitary stalk, or anterior pituitary can cause pituitary hormone deficiency. Because such lesions are not selective, multiple hormonal deficiencies are usually observed. Diabetes insipidus is more frequent in acquired than in congenital hypopituitarism. The most common lesion is the craniopharyngioma.
- 13.(C). The FDA has approved 8 indications for GH treatment to promote growth. They are GH deficiency, Turner syndrome, chronic renal failure before transplantation, idiopathic short stature, small-for-gestational age short stature, Prader-Willi syndrome, *SHOX* gene abnormality, and Noonan syndrome.

14.(E). Recent studies indicate that GH treatment is associated with a 6-fold increase in the risk for type 2 diabetes and no significant increase in the risk for type 1 diabetes.

15.(D). Regulation of vasopressin (VP) secretion and serum osmolality. Hyperosmolality, hypovolemia, and hypotension are sensed by osmosensors, volume sensors, and barosensors, respectively. These stimulate both VP secretion and thirst. VP, acting on the kidney, causes increased reabsorption of water (antidiuresis). Thirst causes increased water ingestion. The results of these dual negative feedback loops cause a reduction in hyperosmolality or in hypotension or hypovolemia. Additional stimuli for VP secretion include nausea, hypoglycemia, and pain.

16.(B). Acquired NDI can result from hypercalcemia or hypokalemia and is associated with lithium, demeclocycline, foscarnet, clozapine, amphotericin, methicillin, and rifampin.

17.(A). Neonates and young infants are often best treated solely with fluid therapy, given their requirement for large volumes (3 L/m²/24 hr) of nutritive fluid.

18.(B). Cerebral salt wasting appears to be the result of hypersecretion of atrial natriuretic peptide and is seen primarily with central nervous system disorders including brain tumors, head trauma, hydrocephalus, neurosurgery, cerebrovascular accidents, and brain death. Hyponatremia is accompanied by elevated urinary sodium excretion (often >150 mEq/L), excessive urine output, hypovolemia, normal or high uric acid, suppressed vasopressin, and elevated atrial natriuretic peptide concentrations (>20 pmol/L). Thus, it is distinguished from SIADH, in which normal or decreased urine output, euvolemia, only modestly elevated urine sodium concentration, and an elevated vasopressin level occur.

19.(C).

20.(B). Precocious puberty is defined by the onset of secondary sexual characteristics before the age of 8 yr in girls and 9 yr in boys.

21.(C). Hypothalamic hamartomas are the most common brain lesion causing central precocious puberty.

22.(E). Breast development may regress after 2 yr, often persists for 3-5 yr, and is rarely progressive.

23.(C). Hair appears on the mons and labia majora in girls and perineal and scrotal area in boys; axillary hair generally appears later.

24.(E). Serum TSH levels are the most accurate test of thyroid function. Serum TSH levels are elevated in primary hypothyroidism and suppressed in hyperthyroidism.

25.(C). At birth, there is an acute release of TSH; peak serum concentrations reach 60 mIU/L 30 min following delivery in full-term infants. A rapid decline occurs in the ensuing 24 hr and a more gradual decline over the next 5 days to <10 mIU/L. The acute increase in TSH produces a dramatic increase in levels of T4 to approximately 16 µg/dL and of T3 to approximately 300 ng/dL in about 4 hr. This T3 seems largely derived from increased peripheral conversion of T4 to T3. T4 levels gradually decrease during the 1st 2 wk of life to 12 µg/dL.

26.(A). TBG levels increase in pregnancy, in the newborn period, with hepatitis, and with administration of estrogens (oral contraceptives), selective estrogen receptor modulators, heroin or methadone, mitotane, 5-fluorouracil, and perphenazine, and they decrease with androgens, anabolic steroids, glucocorticoids, nicotinic acid, and l-asparaginase.

27.(B). Some form of thyroid dysgenesis (aplasia, hypoplasia, or an ectopic gland) is the most common cause of permanent congenital hypothyroidism, accounting for 80-85% of cases.

28.(C). The cord serum T4 is decreased in proportion to gestational age and birthweight. The postnatal TSH surge is reduced, and the more premature, verylow-birthweight infants with complications of prematurity, such as respiratory distress syndrome, actually experience a decrease in serum T4 in the 1st wk of life. As these complications resolve, the serum T4 gradually increases so that generally by 6 wk of life it enters the T4 range seen in term infants.

29.(D).

30.(D). Thyroid-stimulating hormone levels: premature Infants (28-36 wk); 1st wk of life 0.7-27.0 mIU/L; term Infants: birth to 4 days 1.0-17.6 mIU/L; 2-20 wk 0.6-5.6 mIU/L; 5 mo-20 yr 0.5-5.5 mIU/L.

31.(C). Retardation of osseous development can be shown radiographically at birth in approximately 60% of congenitally hypothyroid infants and indicates some deprivation of thyroid hormone during intrauterine life. The distal femoral and proximal tibial epiphyses, normally present at birth, are often absent.

32.(E). The recommended initial starting dose is 10-15 µg/kg/day (totaling 37.5-50.0 µg/ day for most term infants). The starting dose can be tailored to the severity of hypothyroidism. Rapid normalization of thyroid function has been demonstrated to be important in achieving optimal neurodevelopmental outcome.

33.(B). The most common cause of acquired hypothyroidism is chronic lymphocytic (Hashimoto) thyroiditis.

34.(E). Williams syndrome is associated with subclinical hypothyroidism; this does not appear to be autoimmune, as antithyroid antibodies are negative.

35.(B). Children with chronic hepatitis C infection are at risk for subclinical hypothyroidism; this does not appear to be autoimmune, because antithyroid antibodies are negative.

36.(C). Deceleration of growth is usually the first clinical manifestation, but this sign often goes unrecognized.

37.(A). Sonography is not indicated unless there is a suspicion of a thyroid nodule on neck palpation. In such cases, ultrasound examination is the most accurate study to confirm the presence of a nodule and determine if other, smaller nodules are present. In addition, an ultrasound examination can determine the nodule dimensions, texture (solid vs cystic nature), and presence or absence of other features that might influence a decision to undertake fine-needle aspiration, such as microcalcifications, blurred

margins, “taller-than-wide” shape, intranodular vascular flow, and pathologic-appearing adjacent lymph nodes.

38.(D). The clinical course is variable. The goiter might become smaller or might disappear spontaneously, or it might persist unchanged for years while the patient remains euthyroid.

39.(D).

40.(E). Affected persons are goitrous but euthyroid, have normal pubertal development and adult stature, and have little or no impaired thyroid function.

41.(C). Pernicious anemia.

42.(A). Severe liver disease, including liver failure requiring transplantation, has been reported exclusively with propylthiouracil.

43.(E).

44.(A). Hypocalcemia is common in neonates between 12 and 72 hr of life, especially in premature infants, in infants with asphyxia, and in infants of diabetic mothers (early neonatal hypocalcemia).

45.(E). Associated abnormalities of the third and fourth pharyngeal pouches are common; these include conotruncal defects of the heart in 25%, velopharyngeal insufficiency in 32%, cleft palate in 9%, renal anomalies in 35%, and aplasia of the thymus with severe immunodeficiency in 1%.

46.(B). Muscular pain and cramps are early manifestations; they progress to numbness, stiffness, and tingling of the hands and feet.

47.(C). The serum level of alkaline phosphatase is normal or low.

48.(D). Delayed bone age.

49.(C). The 2nd metacarpal is involved least often. As a result, the index finger occasionally is longer than the middle finger. Likewise, the 2nd metatarsal is only rarely affected.

50.(B). Progressive oliguria.

51.(D). The most consistent and characteristic radiographic finding is resorption of subperiosteal bone, best seen along the margins of the phalanges of the hands.

52.(E). Distal renal tubular acidosis.

53.(A). High doses of glucocorticoids deplete monocytes and lymphocytes, especially T cells. They do so at least in part by inducing cell-cycle arrest in the G1 phase and by activating apoptosis through glucocorticoid receptor-mediated effects. The effects on lymphocytes are primarily exerted on T-helper 1 cells and hence on cellular immunity, whereas the T-helper 2 cells are spared, leading to a predominantly humoral immune response.

54.(E). There is an increase in irritability and emotional lability, with an impairment of memory and ability to concentrate.

55.(C). Patients with mineralocorticoid excess can develop hypertension, hypokalemia, and metabolic alkalosis.

56.(A). The most common causes of adrenocortical insufficiency in infancy are the salt-losing forms of congenital adrenal hyperplasia. Approximately 75% of infants with 21-

hydroxylase deficiency, almost all infants with lipid adrenal hyperplasia, and most infants with a deficiency of 3 β -hydroxysteroid dehydrogenase manifest saltlosing symptoms in the newborn period because they are unable to synthesize either cortisol or aldosterone.

57.(D). Hyperkalemia, hyponatremia, and hypoglycemia are prominent presenting signs of adrenal insufficiency in infants.

58.(B). The most definitive test for adrenal insufficiency is measurement of serum levels of cortisol before and after administration of ACTH; resting levels are low and do not increase normally after administration of ACTH.

59.(E). Patients with autoimmune Addison disease must be closely observed for the development of other autoimmune disorders. In children, hypoparathyroidism is the most commonly associated disorder, and it is suspected if hypocalcemia and elevated phosphate levels are present.

60.(B).

61.(D). Approximately 70% of affected infants have the salt-losing form, whereas 30% have the simple virilizing form of the disorder.

62.(B). Progressive weight loss, anorexia, vomiting, dehydration, weakness, hypotension, hypoglycemia, hyponatremia, and hyperkalemia. Typically first develop in affected infants at approximately 10-14 days of age. Without treatment, shock, cardiac arrhythmias, and death may occur within days or weeks.

63.(A). Significantly virilized females usually undergo surgery between 2-6 mo of age. If there is severe clitoromegaly, the clitoris is reduced in size, with partial excision of the corporal bodies and preservation of the neurovascular bundle; however, moderate clitoromegaly may become much less noticeable even without surgery as the patient grows. Vaginoplasty and correction of the urogenital sinus usually are performed at the time of clitoral surgery; revision in adolescence is often necessary.

64.(C). Cortisol deficiency is treated with glucocorticoids. This often requires larger glucocorticoid doses than are needed in other forms of adrenal insufficiency, typically 15-20 mg/m²/24 hr of hydrocortisone daily administered orally in 3 divided doses. Affected infants usually require dosing at the high end of this range. Double or triple doses are indicated during periods of stress, such as infection or surgery.

65.(B). In older children, in addition to obesity, short stature is a common presenting feature. Gradual onset of obesity and deceleration or cessation of growth may be the only early manifestations.

66.(C). The glucose tolerance test is often abnormal but is of no diagnostic utility. Levels of serum electrolytes are usually normal, but potassium may be decreased, especially in patients with tumors that secrete ACTH ectopically.

67.(D). Hypokalemia occurs frequently. Serum pH and the carbon dioxide and sodium concentrations may be elevated and the serum chloride and magnesium levels decreased. Serum levels of calcium are normal, even in children who manifest tetany.

68.(D). Patients have a good appetite but because of the hypermetabolic state may not gain weight, and severe cachexia may develop.

69.(C). Infants with Wolman disease, a rare lipid disorder caused by a deficiency of lysosomal acid lipase, have extensive bilateral calcifications of the adrenal glands.

70.(A). Benign, hormonally inactive adrenocortical adenomas make up the majority of incidentalomas.

71.(E).

- Follicle-stimulating hormone (FSH) and luteinizing hormone (LH) resistance
- Mutations in steroid synthetic pathways
- Gonadal dysgenesis
- Klinefelter syndrome (47,XXY)
- Noonan syndrome (PTPN-11 gene mutation in many cases)
- Cystic fibrosis (infertility)

are congenital causes of hypergonadotropic hypogonadism (primary hypogonadism; testes).

72.(B).

- Anorexia nervosa
- Drug use
- Malnutrition
- Chronic illness, especially Crohn disease
- Hyperprolactinemia
- Pituitary tumors
- Pituitary infarction
- Infiltrative disorders (e.g., histiocytosis, sarcoidosis)
- Hemosiderosis and hemochromatosis
- Radiation

are acquired causes of hypogonadotropic hypogonadism (secondary hypogonadism; hypothalamic-pituitary).

73.(D). Verbal cognitive defects and underachievement in reading, spelling, and mathematics are common. By late adolescence, many boys with Klinefelter syndrome have generalized learning disabilities, most of which are language based. Despite these difficulties, most complete high school.

74.(A).

75.(E). Weaker associations are seen with a large number of other medications and drugs of abuse, including opiates, alcohol, and marijuana, although the association with marijuana may not be as strong as previously thought.

76.(B). Significant gynecomastia is seen in 50% of adolescents with Klinefelter syndrome.

77.(D). No laboratory evaluation is indicated in routine cases with no other associated abnormality but all prepubertal cases, as well as pubertal cases with suspicious features, should be investigated; initial laboratory evaluation should include thyroid function tests (to rule out hyperthyroidism), testosterone, estradiol, human chorionic gonadotropin, luteinizing hormone, and prolactin levels.

78.(A). Many patients with Turner syndrome are recognizable at birth because of a characteristic edema of the dorsa of the hands and feet and loose skinfolds at the nape of the neck. Low birthweight and decreased birth length are common.

79.(B). Regardless of the age, all patients with Turner syndrome at the time of diagnosis need comprehensive cardiovascular evaluation by a cardiologist specializing in congenital heart disease. Complete cardiologic evaluation, including echocardiography, reveals isolated nonstenotic bicuspid aortic valves in one third to one half of patients. In later life, bicuspid aortic valve disease can progress to dilation of the aortic root. Less-frequent defects include aortic coarctation (20%), aortic stenosis, mitral valve prolapse, and anomalous pulmonary venous drainage.

80.(C). Sexual development and menarche are normal. Most pregnancies have resulted in normal infants.

81.(B). The phenotype differs from Turner syndrome in several respects. Cognitive impairment is often present, the cardiac defect is most often pulmonary valvular stenosis or an atrial septal defect rather than an aortic defect, normal sexual maturation usually occurs but is delayed by 2 yr on average, and premature ovarian failure has been reported.

82.(D).

83.(B). The presentation of T2DM is typically more insidious than that with T1DM. In contrast to patients with T1DM who are usually ill at the time of diagnosis and whose presentation rarely spans more than a few weeks, children with T2DM often seek medical care because of excessive weight gain and fatigue as a result of insulin resistance and/or the incidental finding of glycosuria during routine physical examination.

84.(D). It should be noted that a fasting blood glucose that exceeds 125 mg/dL (6.9 mmol/L) is the accepted criterion for the diagnosis of diabetes.

85.(D). The clearest evidence of a role for viral infection in human T1DM is seen in congenital rubella syndrome. Prenatal infection with rubella is associated with β -cell autoimmunity in up to 70%, with development of T1DM in up to 40% of infected children.

86.(D). A number of factors, including prenatal influences, diet in infancy, viral infections, lack of exposure to certain infections even psychologic stress, are implicated in the pathogenesis of T1DM, but their exact role and the mechanism by which they trigger or aggravate autoimmunity remains uncertain.

87.(A). Falsely low HbA1c levels are noted in hemolytic anemias, pure red cell aplasia, blood transfusions, and anemias associated with hemorrhage, cirrhosis, myelodysplasias, or renal disease treated with erythropoietin.

88.(A). Fifteen to 30% of subjects with T1DM have elevated thyroid-stimulating hormone (TSH) and antithyroid antibodies and close to 5-10% have evidence for celiac disease. These diseases share common genes and likely the same interplay between environmental and immunologic factors.

89.(B). An increased ion gap.

90.(C). To continue the insulin infusion without causing hypoglycemia, glucose must be added to the infusion. We typically recommend that glucose be added as a 5% solution when the serum glucose has decreased <300 mg/dL and as a 10% solution when the serum glucose has decreased <200 mg/dL. The insulin infusion can also be lowered from the initial maximal rate if, despite the above outlined interventions, the serum glucose falls further.

91.(C).

92.(B). Any child can be easily transitioned to oral intake and subcutaneous insulin when DKA has resolved (total CO₂ >15 mEq/L; pH >7.30; sodium stable between 135 and 145 mEq/L; no emesis). The first dose of short acting subcutaneous insulin is given with a meal, and the insulin drip is discontinued approximately 30 min later.

93.(D). Radiographic imaging is frequently unhelpful in making the diagnosis of cerebral edema.

94.(D). This syndrome is characterized by severe hyperglycemia (blood glucose >800 mg/dL), absence of or only slight ketosis, nonketotic acidosis, severe dehydration, depressed sensorium or frank coma, and various neurologic signs that may include grand mal seizures, hyperthermia, hemiparesis, and positive Babinski signs.

95.(A). Depending on the method used for determination, HbA1c values may be spuriously elevated in thalassemia (or other conditions with elevated hemoglobin F) and spuriously lower in sickle cell disease (or other conditions with high red blood cell turnover).

96.(A). Complications of DM can be divided into 3 major categories: (1) microvascular complications, specifically, retinopathy and nephropathy; (2) macrovascular complications, particularly accelerated coronary artery disease, cerebrovascular disease, and peripheral vascular disease; and (3) neuropathies, both peripheral and autonomic, affecting a variety of organs and systems. In addition, cataracts may occur more frequently.

97.(D). Screening for retinopathy commenced after 5 yr duration in prepubertal children, after 2 yr in pubertal children.

98.(A). Screening for thyroid disease and celiac disease commenced at diagnosis.

99.(A). Short-term hyperglycemia, strenuous exercise, urinary tract infections, marked hypertension, heart failure, and acute febrile illness can cause transient elevation urinary albumin excretion. There is marked day-to-day variability in albumin excretion, so at least 2 of 3 collections done in a 3-6 mo period should show elevated levels before microalbuminuria is diagnosed and treatment is started.

100.(D).

101.(C). The most commonly used and the only FDA-approved oral agent for the treatment of T2DM in children and adolescents is metformin. Renal function must be assessed before starting metformin as impaired renal function has been associated with potentially fatal lactic acidosis.

Significant hepatic dysfunction is also a contraindication to metformin use, although mild elevations in liver enzymes may not be an absolute contraindication. The usual

starting dose is 500 mg once daily. This may be increased to a maximum dose of 2,000 mg/day. Abdominal symptoms are common early in the course of treatment, but in most cases they will resolve with time.

102.(C). Extreme insulin resistance, acanthosis nigricans, abnormalities of the teeth and nails, and pineal hyperplasia are features of Rabson-Mendenhall syndrome.

103.(B). Macrovascular complications do not appear to be of concern in CFRD, perhaps because of the shortened life span of these patients.

104.(A). Decreasing insulin requirements.

105.(C). High-dose oral or parenteral steroid therapy usually results in significant insulin resistance leading to glucose intolerance and overt diabetes. The immunosuppressive agents cyclosporin and tacrolimus are toxic to β cells, causing IDDM in a significant proportion of patients treated with these agents. Their toxicity to pancreatic β cells was 1 of the factors that limited their usefulness in arresting ongoing autoimmune destruction of β cells. Streptozotocin and the rodenticide Vacor are also toxic to β cells, causing diabetes.

The Nervous System

QUESTIONS

ZUHAIR ALMUSAWI

1. The skull tends to assume a square or boxlike shape in
 - A. Klinefelter syndrome
 - B. neurofibromatosis
 - C. storage disorder
 - D. chronic subdural hemorrhages
 - E. hydrocephalus

2. Smell can be tested reliably at
 - A. 32nd wk of gestation
 - B. birth
 - C. three months of age
 - D. six months of age
 - E. nine months of age

3. Contraindications to performing a lumbar puncture include the following **EXCEPT**
 - A. suspected mass lesion of the brain
 - B. suspected mass lesion of the spinal cord
 - C. symptoms and signs of impending cerebral herniation
 - D. skin infection at the site of the lumbar puncture
 - E. thrombocytopenia with a platelet count $<50 \times 10^9/L$

4. An elevated polymorphonuclear (PMN) cells count in cerebrospinal fluid (CSF) suggests
 - A. tuberculous meningitis
 - B. early phase of aseptic meningitis
 - C. fungal meningitis
 - D. demyelinating diseases
 - E. brain or spinal cord tumor

5. Xanthochromia of cerebrospinal fluid (CSF) suggests the following **EXCEPT**
 - A. bloody tap
 - B. subarachnoid hemorrhage
 - C. carotenemia
 - D. hyperbilirubinemia
 - E. markedly elevated CSF protein

6. Cranial CT is a valuable diagnostic tool in the evaluation of the following conditions **EXCEPT**

- A. skull fractures
- B. intracranial hemorrhages
- C. acute infarcts
- D. hydrocephalus
- E. impending herniation

7. An approach to imaging of the spine in patients with cutaneous lesions is indicated in the following conditions **EXCEPT**

- A. hairy patch
- B. subcutaneous mass or lipoma
- C. dermal sinus
- D. coccygeal pits
- E. scarlike lesions

8. Regarding myelomeningocele, all the following are true **EXCEPT**

- A. risk of recurrence after one affected child is 3-4%
- B. maternal periconceptional use of folic acid supplementation reduces the incidence of neural tube defects (NTDs) in pregnancies at risk by at least 50%
- C. anticonvulsant valproic acid causes NTDs in approximately 1-2% of pregnancies when administered during pregnancy
- D. hydrocephalus in association with a type II Chiari malformation develops in at least 80% of patients with myelomeningocele
- E. the lower the deformity is in the neuraxis (sacrum), the more likely is the risk of hydrocephalus

9. Mobius syndrome is characterized by the following **EXCEPT**

- A. bilateral facial weakness
- B. paralysis of the abducens nerve
- C. hypoplasia or agenesis of brainstem nuclei
- D. feeding difficulties
- E. mental retardation

10. Dandy-Walker malformation is characterized by the following **EXCEPT**

- A. cystic dilatation of the fourth ventricle
- B. herniation of the cerebellar tonsils through the foramen magnum
- C. hypoplasia of the cerebellar vermis
- D. hydrocephalus
- E. an enlarged posterior fossa

11. Familial (autosomal recessive) microcephaly is characterized by the following **EXCEPT**

- A. slanted forehead
- B. prominent nose and ears
- C. mild or borderline mental retardation
- D. prominent seizures
- E. surface convolitional markings of the brain

12. Causes of communicating hydrocephalus include

- A. achondroplasia
- B. aqueductal stenosis
- C. Chiari malformation
- D. Dandy-Walker malformation
- E. Klippel-Feil syndrome

13. Nonobstructive or communicating hydrocephalus may follow the following conditions **EXCEPT**

- A. a subarachnoid hemorrhage
- B. pneumococcal meningitis
- C. tuberculous meningitis
- D. leukemic infiltrates
- E. a vein of Galen malformation

14. Regarding the CSF, all the following are true **EXCEPT**

- A. approximately 25% of CSF originates from extrachoroidal sources
- B. in a normal child, about 20 mL/hr of CSF is produced
- C. total volume of CSF approximates 150 mL in an infant
- D. normally, CSF flows from the lateral ventricles through the foramina of Monro into the 3rd ventricle
- E. CSF is absorbed primarily by the arachnoid villi

15. Trigoncephaly is a skull deformity that is a direct result of premature fusion of the following

- A. sagittal suture
- B. coronal suture
- C. lambdoid suture
- D. metopic suture
- E. multiple sutures

16. Crouzon syndrome is characterized by the following **EXCEPT**

- A. an autosomal dominant inheritance
- B. brachycephaly

- C. ocular proptosis
 - D. hypoplasia of the maxilla
 - E. syndactyly
17. Minor risk factors for recurrence of febrile seizure include the following **EXCEPT**
- A. complex febrile seizure
 - B. age <1 yr
 - C. family history of febrile seizures
 - D. male gender
 - E. lower serum sodium
18. There are several predictors of epilepsy after febrile seizures, the highest percent of risk factor for subsequent epilepsy after febrile seizure is
- A. complex febrile seizure, any type
 - B. fever <1 hr before febrile seizure
 - C. recurrent febrile seizures
 - D. focal complex febrile seizure
 - E. family history of epilepsy
19. Dravet syndrome is characterized by all the following **EXCEPT**
- A. the most severe of the phenotypic spectrum of febrile seizures
 - B. its onset is in the 1st yr of life
 - C. seizures subsequently start to occur without fever
 - D. developmental delay
 - E. an autosomal dominant inheritance
20. Electroencephalogram (EEG) in febrile seizure is characterized by the following **EXCEPT**
- A. an EEG need not normally be performed in first simple febrile seizure
 - B. an abnormal EEG could predict the future recurrence of febrile seizures or epilepsy
 - C. spikes during drowsiness are often seen in children with febrile seizures
 - D. an EEG performed within 2 wk of a febrile seizure often have nonspecific slowing
 - E. an EEG should be used to delineate the type of epilepsy
21. Absence seizures are **MOST** often initially treated with
- A. ethosuximide
 - B. valproate
 - C. lamotrigine
 - D. acetazolamide
 - E. clonazepam

22. Benign myoclonic epilepsies are often best treated with
- A. clonazepam
 - B. lamotrigine
 - C. topiramate
 - D. valproate
 - E. benzodiazepines
23. Gingival hyperplasia, coarsening of the facies, hirsutism, and cerebellovestibular symptoms (nystagmus and ataxia) are adverse effects of
- A. carbamazepine
 - B. lamotrigine
 - C. phenytoin
 - D. valproic acid
 - E. succinimides
24. Rickets is a potential side effect from all the following **EXCEPT**
- A. phenytoin
 - B. valproate
 - C. phenobarbital
 - D. primidone
 - E. carbamazepine
25. Epilepsy surgery is often used to treat refractory epilepsy of a number of etiologies including the following **EXCEPT**
- A. cortical dysplasia
 - B. tuberous sclerosis
 - C. polymicrogyria
 - D. degenerative problems
 - E. Sturge-Weber syndrome
26. The **MOST** common cause of neonatal seizures is
- A. vascular events
 - B. intracranial infections
 - C. brain malformations
 - D. hypoxic-Ischemic encephalopathy
 - E. metabolic disturbances
27. All the following are true regarding diazepam in neonatal seizures **EXCEPT**
- A. diazepam is highly lipophilic
 - B. it is cleared very quickly
 - C. recurrence of seizures is more than other anticonvulsants
 - D. it carries a risk of apnea and hypotension

- E. it is currently recommended as a first-line agent

28. Neuro-imaging is warranted in a child with headache in the following conditions

EXCEPT

- A. abnormal neurologic examination
- B. afternoon headache
- C. headache in children <6 yr old
- D. brief cough headache
- E. migrainous headache in the child with no family history of migraine

29. Tension-type headaches (TTH) are characterized by the following **EXCEPT**

- A. diffuse in location
- B. not affected by activity
- C. throbbing quality
- D. mild to moderate in severity
- E. less frequently associated with nausea and photophobia

30. The following are complications of neurofibromatosis (type 1) NF-1 **EXCEPT**

- A. learning disability
- B. cataracts
- C. seizures
- D. precocious puberty
- E. scoliosis

31. Definite tuberous sclerosis complex (TSC) is diagnosed when at least 2 major or 1 major plus 2 minor features are present, all the following are minor features **EXCEPT**

- A. shagreen patch
- B. cerebral white matter migration lines
- C. multiple dental pits
- D. gingival fibromas
- E. bone cysts

32. The facial port-wine stain of Sturge-Weber syndrome (SWS) is characterized by the following **EXCEPT**

- A. present at birth
- B. tends to be unilateral
- C. always involves the upper face and eyelid
- D. its distribution is consistent with the ophthalmic division of the trigeminal nerve
- E. most children with facial port-wine stain have SWS

33. All the following are features of Von Hippel–Lindau (VHL) disease **EXCEPT**

- A. its incidence is around 1: 36,000
 - B. fifty percent have a de novo gene mutation
 - C. hemangioblastoma of the spinal cord may be found
 - D. renal carcinoma is the most common cause of death
 - E. pheochromocytoma is a frequent association
34. PHACE syndrome denotes to the following **EXCEPT**
- A. posterior fossa malformations
 - B. hemangiomas
 - C. anal anomalies
 - D. coarctation of the aorta
 - E. eye abnormalities
35. The current antiepileptic drug (AEDs) of choice for primary generalized tonic-clonic seizures in children is
- A. phenytoin
 - B. carbamazepine
 - C. phenobarbital
 - D. valproate
 - E. topiramate
36. The **MOST** common type of cerebral palsy (CP) associated with seizures is
- A. spastic hemiplegia
 - B. spastic quadriplegia
 - C. spastic diplegia
 - D. hypotonic CP
 - E. ataxic CP
37. The following are manifestations of increased intracranial pressure in an infant **EXCEPT**
- A. increasing head circumference
 - B. bulging fontanel
 - C. failure to thrive
 - D. persistent vomiting
 - E. setting sun sign
38. Characteristic features of pseudotumor cerebri include the following **EXCEPT**
- A. headache
 - B. stiff neck
 - C. papilledema
 - D. fatigue
 - E. abnormal CSF profile

39. How often are EEGs abnormal in healthy children?
- A. 5%
 - B. 10%
 - C. 15%
 - D. 20%
 - E. 25%
40. The **MOST** common precipitant of status epilepticus in children is
- A. CNS infection
 - B. fever
 - C. medication change
 - D. trauma
 - E. metabolic cause
41. A 9-month-old infant can do all the following **EXCEPT**
- A. pulls to stand
 - B. pincer grasp
 - C. plays pat-a-cake
 - D. imitates sounds
 - E. comes when called
42. A premature infant blinks in response to a bright light at
- A. 26 wk of corrected gestational age
 - B. 28 wk of corrected gestational age
 - C. 30 wk of corrected gestational age
 - D. 32 wk of corrected gestational age
 - E. 34 wk of corrected gestational age
43. The **MOST** common cause of anatomic megalencephaly is
- A. Sotos syndrome
 - B. benign familial megalencephaly
 - C. Simpson-Golabi-Behmel syndrome
 - D. fragile X syndrome
 - E. Weaver syndrome
44. Craniosynostosis is defined as premature closure of the cranial sutures. Of the following, the **MOST** common form of craniosynostosis is
- A. frontal plagiocephaly
 - B. occipital plagiocephaly
 - C. scaphocephaly
 - D. trigonocephaly
 - E. turriccephaly

45. The following are risk factors for deformational plagiocephaly **EXCEPT**
- A. female sex
 - B. firstborn child
 - C. congenital torticollis
 - D. developmental delay
 - E. exclusive bottle feeding
46. Major risk factors for recurrence of febrile seizure include
- A. fever 38-39°C
 - B. family history of febrile seizures
 - C. family history of epilepsy
 - D. complex febrile seizure
 - E. lower serum sodium at time of presentation
47. In patients with febrile seizures, one of the following risk factors has the highest risk for subsequent epilepsy
- A. recurrent febrile seizures
 - B. fever <1 hr before febrile seizure
 - C. neurodevelopmental abnormalities
 - D. family history of epilepsy
 - E. complex febrile seizures
48. The majority of patients who had prolonged febrile seizures and encephalopathy after vaccination and who had been presumed to have suffered from vaccine encephalopathy (seizures and psychomotor regression occurring after vaccination and presumed to be caused by it) turn out to have
- A. generalized epilepsy with febrile seizures plus (GEFS+)
 - B. temporal lobe epilepsy secondary to mesial temporal sclerosis
 - C. myoclonic astatic seizures
 - D. Dravet syndrome
 - E. focal febrile seizures plus epilepsy variant
49. Higher risk of recurrence of the febrile seizure is associated with lower serum
- A. sodium
 - B. potassium
 - C. chloride
 - D. calcium
 - E. magnesium
50. A history of personality change in a patient with seizure could suggest the following as a cause of seizure
- A. intracranial tumor

- B. degenerative disease
- C. metabolic disease
- D. stimulants drugs
- E. congenital brain dysfunction

51. Guidelines on the evaluation of a first unprovoked nonfebrile seizure, the following studies are recommended in specific clinical situations

- A. head CT
- B. head MRI
- C. spinal tap
- D. ECG
- E. EEG

52. Drug therapy should be based on the type of seizure and the epilepsy syndrome as well as on other individual factors.

Of the following, the drug of first choice for focal seizures and epilepsies is

- A. carbamazepine
- B. ethosuximide
- C. valproate
- D. lamotrigine
- E. clobazam

53. Weight gain and alopecia are side effects of the following antiepileptic drugs AEDs

- A. primidone
- B. valproic acid
- C. oxcarbazepine
- D. lamotrigine
- E. acetazolamide

54. The ketogenic diet is absolutely contraindicated in

- A. primary carnitine deficiency
- B. myoclonic–astatic epilepsy
- C. tuberous sclerosis complex
- D. Rett syndrome
- E. infantile spasms

55. Discontinuation of antiepileptic drugs (AEDs) is usually indicated when children are free of seizures for at least

- A. 1 yr
- B. 1.5 yr
- C. 2 yr
- D. 2.5 yr

E. 3 yr

56. The following factors are associated with a higher risk of seizure relapse after antiepileptic drugs (AEDs) withdrawal **EXCEPT**

- A. younger age of epilepsy onset
- B. longer duration of epilepsy
- C. presence of multiple seizure types
- D. need to use more than 1 AED
- E. abnormal EEG before medication is discontinued

57. Sudden unexpected death in epilepsy (SUDEP) is the most common epilepsy related mortality in patients with chronic epilepsy. All the following are risk factors **EXCEPT**

- A. polypharmacology
- B. female gender
- C. age younger than 16 yr
- D. long duration of epilepsy
- E. frequent seizures

58. There are 5 main neonatal seizure types: subtle, clonic, tonic, spasms, and myoclonic.

One of the following seizures is frequently not associated with electrographic discharges

- A. spasms
- B. focal clonic
- C. subtle
- D. focal tonic
- E. generalized myoclonic

59. Subtle seizures include all the following **EXCEPT**

- A. blinking
- B. mouthing
- C. fluctuations in heart rate
- D. hypotension episodes
- E. apnea

60. The following are features of Aicardi syndrome **EXCEPT**

- A. coloboma of the iris
- B. retinal lacunae
- C. agenesis of the corpus callosum
- D. severe seizures
- E. aminoaciduria

61. Many inborn errors of metabolism cause generalized convulsions in the newborn period. Prominent hiccups, persistent generalized seizures, and lethargy rapidly leading to coma are features of

- A. propionic academia
- B. maple syrup urine disease
- C. nonketotic hyperglycinemia
- D. Leigh disease
- E. neonatal adrenoleukodystrophy

62. The initial drug used to control acute neonatal seizures is usually

- A. diazepam
- B. midazolam
- C. phenobarbital
- D. lorazepam
- E. phenytoin

63. Intravenous phenytoin is not widely used to control acute neonatal seizures because of all the following **EXCEPT**

- A. reduced solubility
- B. severe local cutaneous reactions
- C. interaction with other drugs
- D. possible cardiac toxicity
- E. not possible to mix with saline solutions

64. Status epilepticus is a medical emergency that should be anticipated in any patient who presents with an acute seizure. It is defined as continuous seizure activity or recurrent seizure activity without regaining of consciousness lasting for more than

- A. 5 min
- B. 15 min
- C. 30 min
- D. 45 min
- E. 60 min

65. The following studies are needed for all patients with status epilepticus **EXCEPT**

- A. glucose
- B. magnesium
- C. complete blood count
- D. MRI
- E. CT scan

66. Currently, the level of the evidence for refractory treatment of status epilepticus is strongest for

- A. midazolam
- B. propofol
- C. levetiracetam
- D. phenytoin
- E. Phenobarbital

67. All the following are treatment options for breath-holding spells **EXCEPT**

- A. education and reassurance of the parents
- B. iron therapy
- C. atropine sulfate
- D. antiepileptic drug therapy
- E. providing more care and interest for the patient

68. Migraine is the most frequent type of recurrent headache that is brought to the attention of parents and primary care providers, but it remains under recognized and undertreated, particularly in children.

Migraine is characterized by all the following **EXCEPT**

- A. episodic attacks
- B. moderate to severe in intensity
- C. focal in location on the head
- D. constant quality
- E. may be associated with nausea and vomiting

69. The aura associated with migraine is a neurologic warning that a migraine is going to occur.

Of the following, the **LEAST** common type of typical auras is

- A. sensory aura
- B. dysphasic aura
- C. visual aura
- D. vertigo
- E. distortion

70. The following are indications for neuroimaging in a child with a headache **EXCEPT**

- A. abnormal neurologic examination
- B. headache worst on first awakening
- C. brief cough headache
- D. frontal headache
- E. migrainous headache in the child with no family history of migraine

71. Café-au-lait macules are not specific for NF-1; they may be seen in the following **EXCEPT**

- A. Noonan syndrome

- B. ataxia telangiectasia
- C. Fanconi anemia
- D. Gaucher disease
- E. Hurler syndrome

72. The **MOST** frequent lesion associated with neurofibromatosis type 2 NF-2 is

- A. bilateral vestibular schwannomas
- B. intracranial
- C. cataract
- D. retinal hamartoma
- E. skin plaque

73. Slow, writhing, continuous, and involuntary movements are called

- A. stereotypies
- B. tics
- C. tremor
- D. athetosis
- E. chorea

74. A slowly progressive ataxia that involves the lower extremities to a greater degree than the upper extremities. The Romberg test result is positive; the deep-tendon reflexes are absent (particularly at the ankle), and the plantar response is typically extensor (Babinski sign).

Of the following, the **MOST** likely cause of this ataxia is

- A. ataxia-telangiectasia
- B. Friedreich ataxia
- C. abetalipoproteinemia
- D. Roussy-Levy disease
- E. Ramsay Hunt syndrome

75. The **MOST** common acquired cause of chorea in childhood is

- A. Huntington disease
- B. paraneoplastic choreas
- C. benign hereditary chorea
- D. Sydenham chorea
- E. chorea gravidarum

76. Prematurity is a major cause of

- A. spastic diplegia
- B. spastic quadriplegia
- C. spastic hemiplegia
- D. athetoid CP

- E. dyskinetic CP
77. The **MOST** severe form of CP is
- A. spastic diplegia
 - B. spastic quadriplegia
 - C. spastic hemiplegia
 - D. athetoid CP
 - E. dyskinetic CP
78. All the following can be used for treatment of cerebral palsy (CP) **EXCEPT**
- A. benzodiazepines
 - B. baclofen
 - C. dantrolene
 - D. botulinum toxin
 - E. hyperbaric oxygen
79. Infants with Tay-Sachs disease (TSD) are characterized by the following **EXCEPT**
- A. convulsions
 - B. blindness
 - C. deafness
 - D. cherry-red spots
 - E. microcephaly
80. Excessive irritability and crying, unexplained episodes of hyperpyrexia, vomiting, and difficulty feeding are the symptoms of the following neuro-degenerative disease
- A. Tay-Sachs disease (TSD)
 - B. Sandhoff disease
 - C. juvenile GM2 gangliosidosis
 - D. Krabbe disease (KD)
 - E. metachromatic leukodystrophy
81. Presenting symptoms in pediatric multiple sclerosis (MS) include the following **EXCEPT**
- A. hemiparesis
 - B. optic neuritis
 - C. ataxia
 - D. dysarthria
 - E. encephalopathy
82. The **MOST** common focal presentation of arterial ischemic stroke is
- A. hemiparesis
 - B. acute visual deficit

- C. speech deficit
- D. sensory deficit
- E. balance deficit

83. The **MOST** common cause of childhood subarachnoid and intraparenchymal hemorrhagic stroke (HS) is

- A. moyamoya disease/syndrome
- B. arteriovenous malformations
- C. cerebral sinovenous thrombosis
- D. hemolytic uremic syndrome
- E. idiopathic thrombocytopenic purpura

84. Bilateral cortical visual dysfunction, encephalopathy, and seizures are seen in

- A. global hypoxic–ischemic encephalopathy
- B. herpes encephalitis
- C. multiple sclerosis
- D. hypertensive encephalopathy
- E. hypoglycemia

85. The imaging studies are usually normal in the following stroke-like disorders of children

- A. alternating hemiplegia
- B. inborn errors of metabolism
- C. hypertensive encephalopathy
- D. hypoglycemia
- E. global hypoxic–ischemic encephalopathy

86. Small vessel childhood primary angiitis of the CNS (SVcPACNS) is usually presented with

- A. arterial ischemic stroke
- B. seizures
- C. cognitive dysfunction
- D. personality changes
- E. loss of social control

87. The diagnosis of diffuse CNS infections depends on examination of cerebrospinal fluid (CSF) obtained by lumbar puncture (LP). The cerebrospinal fluid (CSF) protein may reach 3,000 (mg/dL) in

- A. acute bacterial meningitis
- B. tuberculous meningitis
- C. fungal meningitis
- D. amebic (naegleria) meningoencephalitis

- E. subdural empyema
88. The organism causing CNS infection is never seen on direct examination of CSF in
- A. tuberculous meningitis
 - B. acute bacterial meningitis
 - C. partially treated bacterial meningitis
 - D. fungal meningitis
 - E. amebic (naegleria) meningoencephalitis
89. T-lymphocyte defects (congenital or acquired by chemotherapy, AIDS, or malignancy) are associated with an increased risk of infections of the CNS with
- A. *Listeria monocytogenes*
 - B. *Streptococcus pneumoniae*
 - C. *Neisseria meningitidis*
 - D. *Haemophilus influenzae* type b
 - E. coagulase-negative Staphylococci
90. The CSF leukocyte count in normal healthy neonates may reach
- A. 10 leukocytes/mm³
 - B. 15 leukocytes/mm³
 - C. 20 leukocytes/mm³
 - D. 25 leukocytes/mm³
 - E. 30 leukocytes/mm³
91. The following is an alternative treatment for *L. monocytogenes* meningitis
- A. vancomycin
 - B. cefotaxime
 - C. chloramphenicol
 - D. intravenous trimethoprim-sulfamethoxazole
 - E. meropenem
92. Reversible gallbladder pseudolithiasis, detectable by abdominal ultrasonography during treatment of bacterial meningitis is usually caused by
- A. ceftazidime
 - B. cefotaxime
 - C. ceftriaxone
 - D. meropenem
 - E. chloramphenicol
93. Data support the use of intravenous dexamethasone, 0.15 mg/kg/dose given every 6 hr for 2 days, in the treatment of children older than 6 wk with acute bacterial meningitis caused by *H. influenzae* type b. for the following reasons **EXCEPT**

- A. shorter duration of fever
- B. lower CSF protein
- C. lower mortality
- D. lower CSF lactate levels
- E. reduction in sensorineural hearing loss

94. Seizures are common during the course of bacterial meningitis. Immediate therapy for seizures includes intravenous diazepam (0.1- 0.2 mg/kg/dose) or lorazepam (0.05- 0.10 mg/kg/dose).

After immediate management of seizures, patients should receive the following anticonvulsant to reduce the likelihood of recurrence

- A. valproic acid
- B. phenytoin
- C. phenobarbital
- D. lorazepam
- E. carbamazepine

95. The **MOST** common neurologic sequelae of bacterial meningitis is

- A. recurrent seizures
- B. delay in acquisition of language
- C. hearing loss
- D. cognitive impairment
- E. visual impairment

96. The **MOST** common cause of viral meningoencephalitis are

- A. arboviruses
- B. enteroviruses
- C. herpes simplex virus (HSV) type 1
- D. varicella-zoster virus
- E. mumps

97. The following viruses have clinical manifestations similar to that of the enteroviruses with the exception of more severe MRI lesions of the cerebral cortex and at times an absence of a CSF pleocytosis

- A. arboviruses
- B. parechoviruses
- C. herpes simplex virus (HSV) type 1
- D. rabies virus
- E. Epstein-Barr virus

98. Detection of viral DNA or RNA by polymerase chain reaction is the test of choice in the diagnosis of CNS infection caused by

- A. arboviruses
- B. parechoviruses
- C. West Nile virus (WNV)
- D. rabies virus
- E. Epstein-Barr virus

99. The predominant organisms causing brain abscesses in children are

- A. aerobic and anaerobic streptococci
- B. *Streptococcus pneumoniae*
- C. *Enterococcus faecalis*
- D. *bacteroides* spp
- E. *Haemophilus aphrophilus*

100. A brain abscess can be treated with antibiotics without surgery in the following conditions **EXCEPT**

- A. abscess is <2 cm in diameter
- B. illness is of short duration (<2 wk)
- C. lesion is located in the posterior fossa
- D. no signs of increased intracranial pressure
- E. child is neurologically intact

101. Idiopathic intracranial hypertension, also known as pseudotumor cerebri, is a clinical syndrome that mimics brain tumors and is characterized by the following **EXCEPT**

- A. increased intracranial pressure ≥ 280 mm Hg in nonobese, nonsedated children
- B. normal cerebrospinal fluid (CSF) cell count and protein content
- C. normal to slightly decreased ventricular size
- D. normal ventricular anatomy and position documented by MRI
- E. papilledema

102. The following are hematologic causes of childhood pseudotumor cerebri **EXCEPT**

- A. Wiskott-Aldrich syndrome
- B. megaloblastic anemia
- C. polycythemia
- D. sickle cell disease
- E. Fanconi anemia

103. The **MOST** common presenting complaint of intramedullary spinal cord tumors is

- A. gait disturbance
- B. sensory deficits
- C. scoliosis
- D. back pain

E. urinary urgency

104. The **MOST** common involved segments in transverse myelitis (TM) are in the

- A. cervical region
- B. thoracic region
- C. lumbar region
- D. lumbo-sacral region
- E. sacral region

- 1.(D). In chronic subdural hemorrhages, the skull tends to assume a square or boxlike shape, because the long-standing presence of fluid in the subdural space causes enlargement of the middle fossa.
- 2.(A). Although not a routine component of the examination, smell can be tested reliably as early as the 32nd wk of gestation by presenting a stimulus and observing for an alerting response, withdrawal, or both. Care should be taken to use appropriate stimuli, such as coffee or peppermint, as opposed to strongly aromatic substances (e.g., ammonia inhalants) that stimulate the trigeminal nerve. Each nostril should be tested individually by pinching shut the opposite side.
- 3.(E). Thrombocytopenia with a platelet count $<20 \times 10^9/L$.
- 4.(B). Normal CSF contains up to $5/mm^3$ white blood cells (WBCs), and a newborn can have as many as $15/mm^3$. Polymorphonuclear (PMN) cells are always abnormal in a child, but $1-2/mm^3$ may be present in a normal neonate. An elevated PMN count suggests bacterial meningitis or the early phase of aseptic meningitis. CSF lymphocytosis can be seen in aseptic, tuberculous, or fungal meningitis; demyelinating diseases; brain or spinal cord tumor; immunologic disorders, including collagen vascular diseases; and chemical irritation (following myelogram, intrathecal methotrexate).
- 5.(A). Normal CSF contains no red blood cells (RBCs); thus, their presence indicates a traumatic tap or a subarachnoid hemorrhage. Progressive clearing of the blood between the first and last samples indicates a traumatic tap. Bloody CSF should be centrifuged immediately. A clear supernatant is consistent with a bloody tap, whereas xanthochromia (yellow color that results from the degradation of hemoglobin) suggests a subarachnoid hemorrhage. Xanthochromia may be absent in bleeds <12 hr old, particularly when laboratories rely on visual inspection rather than spectroscopy. Xanthochromia can also occur in the setting of hyperbilirubinemia, carotenemia, and markedly elevated CSF protein.
- 6.(C). CT is less useful for diagnosing acute infarcts in children, because radiographic changes might not be apparent for up to 24 hr.
- 7.(D). Imaging of the spine is not indicated in patients with simple dimples (<5 mm, <25 mm from anal verge) or coccygeal pits.
- 8.(E). Generally, the lower the deformity is in the neuraxis (sacrum), the less likely is the risk of hydrocephalus.
- 9.(E). The immobile, dull facies might give the incorrect impression of mental retardation; the prognosis for normal development is excellent in most cases.

10.(B). Chiari malformation is the most common malformation of the posterior fossa and hindbrain. It consists of herniation of the cerebellar tonsils through the foramen magnum.

11.(C). Familial (autosomal recessive) microcephaly is characterized by severe mental retardation.

12.(A). Aqueductal stenosis, Chiari malformation, Dandy-Walker malformation, and Klippel-Feil syndrome are causes of non-communicating hydrocephalus.

13.(E). A vein of Galen malformation can expand to become large and, because of its midline position, obstruct the flow of CSF.

14.(C). The total volume of CSF approximates 50 mL in an infant and 150 mL in an adult.

15.(D).

Sagittal= Dolicocephaly or scaphocephaly (boat-shaped)

Coronal= Unilateral: plagiocephaly Bilateral: brachycephaly, acrocephaly

Lambdoid= Lambdoid/occipital plagiocephaly; right side affected in 70% of cases

Multiple= Oxycephaly

16.(E). Apert syndrome is characterized by syndactyly of the 2nd, 3rd, and 4th fingers, which may be joined to the thumb and the 5th finger.

17.(B). Age <1 yr, duration of fever <24 hr, and fever 38-39°C are major risk factors for recurrence of febrile seizure.

18.(D). Focal complex febrile seizure has 29% risk for subsequent epilepsy.

19.(E). This syndrome is usually caused by a new mutation, although rarely it is inherited in an autosomal dominant manner. The mutated gene is located on 2q24-31 and encodes for SCN1A, the same gene mutated in GEFS+ spectrum. However, in Dravet syndrome the mutations lead to loss of function and thus to a more severe phenotype.

20.(B). An EEG would not predict the future recurrence of febrile seizures or epilepsy even if the result is abnormal.

21.(A). Absence seizures are most often initially treated with ethosuximide, which is as effective as and less toxic than valproate and more effective than lamotrigine.

22.(D). Benign myoclonic epilepsies are often best treated with valproate, particularly when patients have associated generalized tonic-clonic and absence seizures. Benzodiazepines, clonazepam, lamotrigine, and topiramate are alternatives for the treatment of benign myoclonic epilepsy.

23.(C).

24.(B). Potential side effects are rickets from phenytoin, phenobarbital, primidone, and carbamazepine (enzyme inducers that reduce 25-hydroxy-vitamin D level by inducing its metabolism) and hyperammonemia from valproate.

25.(D). Epilepsy surgery is often used to treat refractory epilepsy of a number of etiologies including cortical dysplasia, tuberous sclerosis, polymicrogyria, hypothalamic hamartoma, and hemispheric syndromes, such as Sturge-Weber syndrome, hemimegalencephaly, Rasmussen encephalitis, and Landau-Kleffner syndrome. Patients

with intractable epilepsy resulting from metabolic or degenerative problems are not candidates for resective epilepsy surgery.

26.(D). Hypoxic-ischemic encephalopathy is the most common cause of neonatal seizures, accounting for 50-60% of patients. Seizures secondary to this encephalopathy occur within 12 hr of birth.

27.(E). Because of the respiratory and blood pressure limitations and because the intravenous preparation contains sodium benzoate and benzoic acid, it is currently not recommended as a first-line agent.

28.(B). Neuroimaging is warranted when the neurologic examination is abnormal or unusual neurologic features occur during the migraine; when the child has headaches that awaken him or her from sleep or that are present on first awakening and remit with upright posture; when the child has brief headaches that only occur with cough or bending over; and when the child has migrainous headache with an absolutely negative family history of migraine or its equivalent (e.g., motion sickness, cyclic vomiting).

29.(C). TTH are mild to moderate in severity, are diffuse in location, are not affected by activity (although the patient may not feel like being active), and are nonthrobbing (often described as a constant pressure).

30.(B). Cataracts are found in 60-81% of patients with NF-2.

31.(A). Shagreen patch is a major feature.

32.(E). It is important to note that not all children with facial port-wine stain have SWS. In fact, the overall incidence of SWS has been reported to be 8-33% in those with a port-wine stain.

33.(B). Around 80% of individuals with VHL disease have an affected parent, and around 20% have a de novo gene mutation.

34.(C). Arterial anomalies.

35.(D). Class I evidence demonstrates that phenytoin, carbamazepine, phenobarbital, primidone, valproate, topiramate, oxcarbazepine, and lamotrigine are effective for the treatment of primary generalized tonic-clonic seizures. On the basis of efficacy and tolerability, valproate and lamotrigine have emerged as the current drugs of choice for grand mal seizures.

36.(A).

37.(D). The manifestations of increased intracranial pressure in an infant as compared with an older child are:

Infant: Increasing head circumference, delayed closure of the fontanel, suture separation, bulging fontanel, failure to thrive, macrocephaly, setting sun sign, and shrill cry.

Older child: Headache (especially in the early morning, awakening the child from sleep, or association with vomiting), nausea, persistent vomiting, personality/mood changes, lethargy, anorexia, fatigue, somnolence, diplopia as a result of sixth-nerve palsy or third-nerve palsy with uncal herniation, and papilledema.

38.(E). Normal CSF profile with the exception of an elevated opening pressure.

39.(B). Approximately 10% of "normal" children have mild, nonspecific abnormalities in background activity.

40.(B). Fever/infection (36%) ,CNS infection (5%) , medication change (20%) , trauma (4%) , unknown (9%) , cerebrovascular (3%) , metabolic (8%) , ethanol/drug-related (2%) , congenital (7%) , tumor (1%) , and anoxia (5%).

41.(E). Walks with 1 hand held, releases an object on command, comes when called, and 1-2 meaningful words are milestones of 12-month-old child.

42.(B). At 28 wk of corrected gestational age, a premature infant blinks in response to a bright light, and at 32 wk, the infant maintains eye closure until the light source is removed. A normal 37 wk infant turns the head and eyes toward a soft light, and a term infant is able to fix on and follow a target, such as the examiner's face.

43.(B).

44.(C). Premature closure of the sagittal suture produces a long and narrow skull, or scaphocephaly, the most common form of craniosynostosis.

45.(A). Male sex.

46.(A). Risk factors for recurrence of febrile seizure include

MAJOR

- Age <1 yr
- Duration of fever <24 hr
- Fever 38-39°C (100.4-102.2°F)

MINOR

- Family history of febrile seizures
- Family history of epilepsy
- Complex febrile seizure
- Daycare
- Male gender
- Lower serum sodium at time of presentation

47. (C). Risk factor for subsequent epilepsy

* simple febrile seizure 1%

* recurrent febrile seizures 4%

* complex febrile seizures (more than 15 min duration or recurrent within 24 hr) 6%

* fever<1 hr before febrile seizure 11%

* family history of epilepsy 18%

* complex febrile seizures (focal) 29%

* neurodevelopmental abnormalities 33%

48.(D). The majority of patients who had prolonged febrile seizures and encephalopathy after vaccination and who had been presumed to have suffered from vaccine encephalopathy(seizures and psychomotor regression occurring after vaccination and presumed to be caused by it) turn out to have Dravet syndrome mutations, indicating that their disease is caused by the mutation and not secondary to

the vaccine. This has raised doubts about the very existence of the entity termed vaccine encephalopathy.

49.(A). A low sodium level is associated with higher risk of recurrence of the febrile seizure within the following 24 hr.

50.(A). A history of personality change or symptoms of increased intracranial pressure can suggest an intracranial tumor. Similarly, a history of cognitive regression can suggest a degenerative or metabolic disease. Certain medications such as stimulants or antihistamines, particularly sedating ones, can precipitate seizures. A history of prenatal or perinatal distress or of developmental delay can suggest etiologic congenital or perinatal brain dysfunction.

51.(C). Spinal tap is considered in patients with suspected meningitis or encephalitis, in children without brain swelling or papilledema, and in children in whom a history of intracranial bleeding is suspected without evidence of such on head CT.

52.(A). In general, the drugs of first choice for focal seizures and epilepsies are oxcarbazepine and carbamazepine; for absence seizures, ethosuximide; for juvenile myoclonic epilepsy, valproate and lamotrigine; for Lennox-Gastaut syndrome, clobazam, valproate, topiramate, lamotrigine, and, most recently, as add on, rufinamide; and for infantile spasms, adrenocorticotrophic hormone (ACTH).

53.(B). Side effects of valproic acid: weight gain, hyperammonemia, tremor, alopecia, and menstrual irregularities. Serious: hepatic and pancreatic toxicity.

54.(A). The ketogenic diet is absolutely contraindicated in carnitine deficiency (primary); carnitine palmitoyltransferase I or II deficiency; carnitine translocase deficiency; β -oxidation defects; medium-chain acyl dehydrogenase deficiency; long-chain acyl dehydrogenase deficiency; short-chain acyl dehydrogenase deficiency; long-chain 3-hydroxyacyl-coenzyme A deficiency; medium-chain 3-hydroxyacyl-coenzyme A deficiency; pyruvate carboxylase deficiency; and porphyrias.

55.(C). Discontinuation of AEDs is usually indicated when children are free of seizures for at least 2 yr.

56.(A). Older age of epilepsy onset.

57.(B). Male gender.

58.(C). Spasms, focal clonic, focal tonic, and generalized myoclonic seizures are, as a rule, associated with electrographic discharges (epileptic seizures), whereas motor automatisms, the subtle, generalized tonic and multifocal myoclonic episodes are frequently not associated with discharges and thus are thought to often represent release phenomena with abnormal movements secondary to brain injury rather than true epileptic seizures.

59.(D). Subtle seizures include transient eye deviations, nystagmus, blinking, mouthing, abnormal extremity movements (rowing, swimming, bicycling, pedaling, and stepping), and fluctuations in heart rate, hypertension episodes, and apnea. Subtle seizures occur more commonly in premature than in full-term infants.

60.(E). Brain malformations account for 5-10% of neonatal seizure cases. An example is Aicardi syndrome, which affects girls only and consists of retinal lacunae, agenesis of

the corpus callosum, and severe seizures including subsequent infantile spasms with hypsarrhythmia that is sometimes initially unilateral on EEG.

61.(C). Nonketotic hyperglycinemia, an intractable condition characterized by markedly elevated plasma and CSF glycine levels, prominent hiccups, persistent generalized seizures, and lethargy rapidly leading to coma.

62.(D). The initial drug used to control acute seizures is usually lorazepam. Lorazepam is distributed to the brain very quickly and exerts its anticonvulsant effect in <5 min. It is not very lipophilic and does not clear out from the brain very rapidly. Its action can last 6-24 hr. Usually, it does not cause hypotension or respiratory depression. The dose is 0.05 mg/kg (range: 0.02-0.10 mg/kg) every 4-8 hr.

63.(E). It is not possible to mix phenytoin or fosphenytoin with dextrose solutions.

64.(A).

65.(D). Lumbar puncture, comprehensive toxicologic screens, MRI, and other laboratory tests are performed depending on clinical suspicion and need.

66.(A). Currently, the level of the evidence for refractory treatment is strongest for midazolam and valproate, followed by propofol and pentobarbital/thiopental, followed by levetiracetam, phenytoin/fosphenytoin, lacosamide, topiramate, and phenobarbital.

67.(E). All parents should be taught not to provide secondary gain when the episodes occur, because this can reinforce the episodes.

68.(D). Migraine is characterized by episodic attacks that may be moderate to severe in intensity, focal in location on the head, have a throbbing quality, and may be associated with nausea, vomiting, light sensitivity, and sound sensitivity. Compared to adults, pediatric migraine is shorter in duration and has a bilateral, often bifrontal, location. Migraine can also be associated with an aura that may be typical (visual, sensory, or dysphasic) or atypical (i.e., hemiplegic, "Alice in Wonderland" syndrome).

69.(B). Dysphasic auras are the least-common type of typical aura and have been described as an inability or difficulty to respond verbally.

70.(D). When the headache is mostly in the occipital area.

71.(E). Hunter syndrome.

72.(A). Bilateral vestibular schwannomas 90-95%.

73.(D).

74.(B). Friedreich ataxia is inherited as an autosomal-recessive disorder involving the spinocerebellar tracts, dorsal columns in the spinal cord, the pyramidal tracts, and the cerebellum and medulla. The onset of ataxia is somewhat later than in ataxiatelangiectasia, but usually occurs before age 10 yr. The ataxia is slowly progressive and involves the lower extremities to a greater degree than the upper extremities. The Romberg test result is positive; the deep-tendon reflexes are absent (particularly at the ankle), and the plantar response is typically extensor (Babinski sign). Patients develop a characteristic explosive, dysarthric speech, and nystagmus is present in most children.

- 75.(D). Sydenham chorea (St. Vitus dance) is the most common acquired chorea of childhood. It occurs in 10-20% of patients with acute rheumatic fever, typically weeks to months after a group A β -hemolytic streptococcal infection. Peak incidence is at age 8-9 yr, with a female predominance of 2:1.
- 76.(A). Prematurity, ischemia, infection, and endocrine/metabolic (e.g., thyroid) are major causes of spastic diplopia.
- 77.(B). Spastic quadriplegia is the most severe form of CP because of marked motor impairment of all extremities and the high association with intellectual disability and seizures.
- 78.(E). Hyperbaric oxygen has not been shown to improve the condition of children with CP.
- 79.(E). Macrocephaly becomes apparent by 1 yr of age and results from the 200-300 fold normal content of GM2 ganglioside deposited in the brain.
- 80.(D). The symptoms of KD become evident in the 1st few mo of life and include excessive irritability and crying, unexplained episodes of hyperpyrexia, vomiting, and difficulty feeding. In the initial stage of KD, children are often treated for colic or milk allergy with frequent formula changes.
- 81.(E). Encephalopathy is less common and suggests consideration of ADEM or possibly neuromyelitis optica (NMO).
- 82.(A). The most common focal presentation is hemiparesis, but acute visual, speech, sensory, or balance deficits also occur. Children with these presentations require urgent neuroimaging and consultation with a child neurologist, as emergency interventions may be indicated.
- 83.(B). Arteriovenous malformations are the most common cause of childhood subarachnoid and intraparenchymal HS and may occur anywhere.
- 84.(D). The posterior reversible leukoencephalopathy syndrome is seen in children with hypertension, often in the context of an acute rise in blood pressure. Posterior regions are selectively involved, possibly resulting in symptoms of bilateral cortical visual dysfunction in addition to encephalopathy and seizures.
- 85.(A).
- 86.(B). Seizures are a hallmark of SVcPACNS, as more than 80% of children with SVcPACNS present with seizures; all seizure types are seen.
- 87.(B). In tuberculous meningitis, CSF protein is 100-3,000; may be higher in presence of block, while in other options, it may reach 500.
- 88.(A). Acid-fast organisms almost never seen on smear. Organisms may be recovered in culture of large volumes of CSF. Mycobacterium tuberculosis may be detected by PCR of CSF.
- 89.(A).
- 90.(E). Normal healthy neonates may have as many as 30 leukocytes/mm³ (usually <10), but older children without viral or bacterial meningitis have <5 leukocytes/mm³ in the CSF; in both age groups there is a predominance of lymphocytes or monocytes.

- 91.(D). If *L. monocytogenes* infection is suspected, as in young infants or those with a T-lymphocyte deficiency, ampicillin (200 mg/kg/24 hr, given every 6 hr) should also be given because cephalosporins are inactive against *L. monocytogenes*. Intravenous trimethoprim sulfamethoxazole is an alternative treatment for *L. monocytogenes*.
- 92.(C). Ceftriaxone may cause reversible gallbladder pseudolithiasis, detectable by abdominal ultrasonography. This is usually asymptomatic but may be associated with emesis and upper right quadrant pain.
- 93.(C). Among children with meningitis caused by *H. influenza* type b, corticosteroid recipients have a shorter duration of fever, lower CSF protein and lactate levels, and a reduction in sensorineural hearing loss. Data in children regarding benefits, if any, of corticosteroids in the treatment of meningitis caused by other bacteria are inconclusive.
- 94.(B). After immediate management of seizures, patients should receive phenytoin (15-20 mg/ kg loading dose, 5 mg/kg/24 hr maintenance) to reduce the likelihood of recurrence. Phenytoin is preferred to phenobarbital because it produces less CNS depression and permits assessment of a patient's level of consciousness. Serum phenytoin levels should be monitored to maintain them in the therapeutic range (10-20 µg/mL).
- 95.(C). Sensorineural hearing loss is the most common sequela of bacterial meningitis and, usually, is already present at the time of initial presentation. It is a result of cochlear infection and occurs in as many as 30% of patients with pneumococcal meningitis, 10% with meningococcal, and 5-20% of those with *H. influenzae* type b meningitis. Hearing loss may also be caused by direct inflammation of the auditory nerve. All patients with bacterial meningitis should undergo careful audiologic assessment before or soon after discharge from the hospital. Frequent reassessment on an outpatient basis is indicated for patients who have a hearing deficit.
- 96.(B). Enteroviruses are the most common cause of viral meningoencephalitis. As of 2014, more than 70 serotypes of these small RNA viruses have been identified.
- 97.(B). Parechoviruses may be an important cause of aseptic meningitis or encephalitis in infants. The clinical manifestations are similar to that of the enteroviruses with the exception of more severe MRI lesions of the cerebral cortex and at times an absence of a CSF pleocytosis.
- 98.(B). Detection of viral DNA or RNA by polymerase chain reaction is the test of choice in the diagnosis of CNS infection caused by HSV, parechovirus, and enteroviruses, respectively. CSF serology is the diagnostic test of choice for WNV.
- 99.(A). The predominant organisms causing brain abscesses in children are aerobic and anaerobic streptococci (60-70% of the cases) with *Streptococcus milleri* gp (*Streptococcus anginosus*, *Streptococcus constellatus*, and *Streptococcus intermedius*) being increasingly isolated from surgically drained brain abscesses.
- 100.(C). Surgery is indicated when the abscess is >2.5 cm in diameter, gas is present in the abscess, the lesion is multiloculated, the lesion is located in the posterior fossa, or a fungus is identified. Associated infectious processes, such as mastoiditis, sinusitis, or a

periorbital abscess, may require surgical drainage. The duration of antibiotic therapy depends on the organism and response to treatment but is usually 4-6 wk.

101.(A). Increased intracranial pressure ≥ 280 mm Hg in sedated or obese children; ≥ 250 mm Hg in nonobese, nonsedated children.

102.(B). Iron-deficiency anemia.

103.(D). With the exception of the uncommon malignant glial tumors of the spinal cord, which tend to present precipitously, intramedullary spinal cord tumors present in a very insidious manner. Back pain related to the level of the tumor is a common presenting complaint. It is likely that this pain will awaken the child from sleep and improve as the day progresses.

104.(B).

1. Gowers sign is fully expressed by the age of
 - A. 3 yr
 - B. 5 yr
 - C. 7 yr
 - D. 9 yr
 - E. 11 yr

2. Duchenne muscular dystrophy (DMD) is the most common hereditary neuromuscular disease affecting all races and ethnic groups.
All the following are features of DMD **EXCEPT**
 - A. scoliosis
 - B. contracture
 - C. fasciculation
 - D. cardiomyopathy
 - E. intellectual impairment

3. The characteristic features of Duchenne muscular dystrophy (DMD) are progressive weakness, intellectual impairment, hypertrophy of the calves, and proliferation of connective tissue in muscle.
Of the following, the **BEST** initial test for diagnosis is
 - A. PCR
 - B. muscle biopsy
 - C. electromyography
 - D. serum creatine kinase
 - E. nerve conduction study

4. In Duchenne muscular dystrophy (DMD), ambulation is important not only for postponing the psychologic depression but also for postponing
 - A. lordosis
 - B. scoliosis
 - C. kyphosis
 - D. ankles contracture
 - E. pseudohypertrophy

5. Becker muscular dystrophy (BMD) is a disease that is fundamentally similar to Duchenne muscular dystrophy (DMD).

Of the following, the **MOST** recognized feature of BMD is

- A. cardiomyopathy
- B. longer ambulation time
- C. calf pseudohypertrophy
- D. elevated creatine kinase
- E. genetic defect at the Xp21.2 locus

6. Emery-Dreifuss Muscular Dystrophy (scapuloperoneal or scapulohumeral muscular dystrophy) is a rare X-linked recessive dystrophy.

Of the following, the **MOST** characteristic feature is

- A. myotonia
- B. facial weakness
- C. pseudohypertrophy
- D. intellectual impairment
- E. dilated cardiomyopathy

7. Myotonic muscular dystrophy (Steinert disease) is the second most common muscular dystrophy in North America, Europe, and Australia, inherited as an autosomal dominant trait.

Of the following, the **LEAST** recognized feature is

- A. cataract
- B. myotonia
- C. low serum IgG
- D. cardiomyopathy
- E. intellectual impairment

8. All the following myopathies are characterized by proximal muscle wasting **EXCEPT**

- A. Becker dystrophy
- B. myotonic dystrophy
- C. Duchenne dystrophy
- D. central core myopathy
- E. hypothyroid myopathy

9. Limb-girdle muscular dystrophies (LGMDs) are a heterogeneous group of progressive hereditary muscular dystrophies that mainly affect muscles of the hip and shoulder girdles.

Of the following, the presenting complaint may be

- A. low back pain
- B. cardiomyopathy
- C. difficulty in swallowing
- D. characteristic facies
- E. intellectual impairment

10. The term congenital muscular dystrophy is misleading because all muscular dystrophies are genetically determined.

A distinguishing feature of the congenital dystrophies from other muscular dystrophies, is a high association with malformations of

- A. liver
- B. lung
- C. brain
- D. bone
- E. kidney

11. All the following may induce myopathy **EXCEPT**

- A. steroid use
- B. hypothyroidism
- C. hyperthyroidism
- D. hyperaldosteronism
- E. hypoparathyroidism

12. In periodic paralysis, all the following are precipitating factors **EXCEPT**

- A. heavy fat meal
- B. licorice ingestion
- C. emotional stress
- D. hyperthyroidism
- E. amphotericin B use

13. In spinal muscular atrophy (SMA) type 1, all the following are spared **EXCEPT**

- A. heart
- B. diaphragm
- C. intelligence
- D. anal sphincter
- E. extraocular muscles

14. Muscle denervation is any loss of nerve supply regardless of the cause.

Of the following, the **MOST** specific clinical sign of denervation is

- A. myotonia
- B. weakness
- C. fatigability
- D. fasciculation
- E. abnormal sensation

15. The **MOST** definitive diagnostic test in Werdnig-Hoffmann disease is

- A. EMG
- B. muscle biopsy

- C. serum creatine kinase
- D. molecular genetic test
- E. motor nerve conduction study

16. In Guillain-Barré syndrome, the paralysis usually follows a nonspecific gastrointestinal or respiratory infection by approximately 10 days.

Of the following, the **MOST** likely respiratory infection that triggers the disease is

- A. Chlamydia trachomatis
- B. Staphylococcal aureus
- C. Haemophilus influenzae
- D. Mycoplasma pneumoniae
- E. Streptococcal pneumonia

17. All the following vaccines may induce Guillain-Barré syndrome (GBS) **EXCEPT**

- A. OPV
- B. Rota
- C. Rabies
- D. Influenza
- E. Conjugated meningococcal vaccine

18. In Guillain-Barré syndrome, the onset is gradual and progresses over days or weeks. The maximal severity of weakness is usually reached by

- A. 2 wk
- B. 4 wk
- C. 6 wk
- D. 8 wk
- E. 10 wk

19. In Guillain-Barré syndrome, respiratory effort must be monitored to prevent respiratory failure and respiratory arrest.

Of the following, the **MOST** recognized sign of impending respiratory failure is

- A. dysphagia
- B. tachypnea
- C. tachycardia
- D. altered sensorium
- E. vasomotor instability

20. Miller-Fisher syndrome (MFS) consists of acute external and occasionally internal ophthalmoplegia, ataxia, and areflexia.

Of the following, the **MOST** likely cranial nerve involved is

- A. 3rd
- B. 4th

- C. 5th
- D. 6th
- E. 7th

21. The least significant finding in Miller-Fisher syndrome (MFS) is

- A. ataxia
- B. areflexia
- C. lower limbs weakness
- D. internal ophthalmoplegia
- E. external ophthalmoplegia

22. Guillain Barré syndrome is an autoimmune disorder often considered a postinfectious polyneuropathy involving mainly motor but also sensory and sometimes autonomic nerves.

All the following are required in diagnosis **EXCEPT**

- A. CSF study
- B. muscle biopsy
- C. electromyography
- D. sural nerve biopsy
- E. motor nerve conduction study

23. The best diagnostic test in Guillain-Barré syndrome (GBS) is

- A. CSF study
- B. electromyography
- C. sural nerve biopsy
- D. serum creatine kinase
- E. motor nerve conduction study

24. Patients in early stages of Guillain-Barré syndrome should be admitted to the hospital for observation because the ascending paralysis can rapidly involve respiratory muscles during the next 24 hr.

Of the following, the **MOST** effective treatment is

- A. steroids
- B. plasmapheresis
- C. antibiotics for C. jejuni
- D. immunosuppressive drugs
- E. intravenous immunoglobulin (IVIG)

25. The last function to be recovered in Guillain-Barré syndrome (GBS) is

- A. sensation
- B. bulbar muscles
- C. anal sphincters

- D. tendon reflexes
- E. respiratory muscles

26. Bell palsy is an acute unilateral peripheral facial nerve palsy that is not associated with other cranial neuropathies or brainstem dysfunction.

Of the following, the **MOST** traditional treatment is

- A. acyclovir
- B. prednisone
- C. laser therapy
- D. physiotherapy
- E. surgical decompression

27. Bell palsy usually develops abruptly about 2 wk after a systemic viral infection.

Of the following, the **MOST** common viral cause is

- A. Mumps virus
- B. Cytomegalovirus
- C. Epstein-Barr virus
- D. Herpes simplex virus
- E. Human herpes virus 6

28. Charcot-Marie-Tooth disease, the most common genetically determined neuropathy.

Of the following, the **MOST** severely affected nerve is

- A. radial
- B. facial
- C. femoral
- D. peroneal
- E. mandibular

29. All the following are preserved in Charcot-Marie-Tooth disease **EXCEPT**

- A. intelligence
- B. tibial nerves
- C. axial muscles
- D. cranial nerves
- E. anal sphincter

30. Of the following, the **LEAST** recognized presentation of Charcot-Marie-Tooth disease is

- A. foot drop
- B. claw hand
- C. pes cavus deformities
- D. stork-like contour legs

- E. tripping over their own feet

31. Myasthenia gravis is a chronic autoimmune disease of neuromuscular blockade.

Of the following, the **MOST** characteristic feature is

- A. fatigue
- B. myalgia
- C. fasciculation
- D. sensory symptom
- E. abnormal pupillary response

32. Myasthenia gravis should be differentiated from other causes of neuromuscular blockade as organophosphate chemicals, botulism, and tick paralysis.

Of the following, the **MOST** specific diagnostic test is

- A. muscle biopsy
- B. electromyography
- C. anti-AChR antibodies
- D. serum creatine kinase
- E. nerve conduction study

33. The earliest and **MOST** constant sign of myasthenia gravis is

- A. ptosis
- B. dysphagia
- C. slurred speech
- D. difficult chewing
- E. poor head control

34. The Creatine kinase (CK) which is found in only 3 organs and may be separated into corresponding isozymes: MM for skeletal muscle, MB for cardiac muscle, and BB for brain.

Of the following, the CK level is characteristically elevated in

- A. Myasthenia gravis
- B. Guillain-Barré syndrome
- C. Werdnig-Hoffmann disease
- D. Duchenne muscular dystrophy
- E. Emery-Dreifuss Muscular Dystrophy

35. Myotubular myopathy, maturational arrest of fetal muscle during the myotubular stage of development at 8-15 wk of gestation.

All the following are a characteristic features **EXCEPT**

- A. high arch palate
- B. cardiomyopathy
- C. polyhydramnios

- D. undescended testes
- E. decrease fetal movement

36. Dolichocephalic head is a feature of

- A. Central core myopathy
- B. Myotubular myopathy
- C. Nemaline rod myopathy
- D. Becker muscular dystrophy
- E. Emery-Dreifuss Muscular Dystrophy

37. Malignant hyperthermia is an acute hypermetabolic syndrome that is triggered by inhalational anesthetic agents and succinylcholine.

Of the following, which myopathy is consistently associated with malignant hyperthermia?

- A. Central core myopathy
- B. Myotubular myopathy
- C. Nemaline rod myopathy
- D. Becker muscular dystrophy
- E. Emery-Dreifuss Muscular Dystrophy

38. Congenital Guillain-Barré syndrome, manifest as generalized hypotonia, weakness, and areflexia in an affected neonate, fulfilling all electrophysiologic and CSF criteria and in the absence of maternal neuromuscular disease.

Of the following, the **MOST** effective treatment is

- A. steroids
- B. reassurance
- C. plasmapheresis
- D. immunosuppressive drugs
- E. intravenous immunoglobulin (IVIG)

39. All the following are features of chronic inflammatory demyelinating polyradiculoneuropathies (CIPDs) **EXCEPT**

- A. motor deficits in 94%
- B. sensory paresthesias in 64%
- C. cranial nerve involvement in <30%
- D. CSF shows pleocytosis and high protein
- E. hyporeflexia or areflexia is almost universal

Neuromuscular Disorders

ANSWERS

KHALID ALAARJI

1. **(B).** An early Gower's sign is often evident by age 3 yr and is fully expressed by age 5 or 6 yr.
2. **(C).** Fasciculations of muscle, which are often best seen in the tongue, are a sign of denervation is seen in spinal muscular atrophy.
3. **(D).** The diagnosis should be confirmed by blood PCR or muscle biopsy in every case.
4. **(B).** Ambulation is important not only for postponing the psychologic depression but also because scoliosis usually does not become a major complication as long as a patient remains ambulatory, even for as little as 1 hr per day; scoliosis often becomes rapidly progressive after confinement to a wheelchair.
5. **(B).** In BMD, boys remain ambulatory until late adolescence or early adult life. The other distracters occur in both DMD and BMD.
6. **(E).** Also characterized by contractures of elbows and ankles that develop early, and muscle becomes wasted in a scapulohumeroperoneal distribution. The serum CK value is only mildly to moderately elevate.
7. **(D).** Cardiac involvement is usually manifested as heart block in the Purkinje conduction system and arrhythmias (and sudden death) rather than as cardiomyopathy, unlike most other muscular dystrophies.
8. **(B).** The distal distribution of muscle wasting in myotonic dystrophy is an exception to the general rule of myopathies having proximal and neuropathies having distal distribution patterns.
9. **(A).** Low back pain may be a presenting complaint because of the lordotic posture resulting from gluteal muscle weakness. The other distracters are less likely to be occurring.
10. **(C).** Brain malformations, particularly disorders of cortical development such as lissencephaly/pachygyria and polymicrogyria, often complicated by severe epilepsy.
11. **(E).** Most patients with primary hyperparathyroidism develop weakness, fatigability, fasciculation, and muscle wasting that is reversible after removal of the parathyroid adenoma.
12. **(A).** Heavy carbohydrate meal.
13. **(B).** In severe infantile form, also known as Werdnig-Hoffmann disease or SMA type 1, there is diaphragmatic involvement, even late.
14. **(D).** Fasciculations of muscle, which are often best seen in the tongue, are a sign of denervation. Sensory abnormalities indicate neuropathy. Fatigable weakness is characteristic of neuromuscular junctional disorders. Myotonia is specific for a few myopathies.

- 15.(D). The serum creatine kinase level may be normal but more commonly is mildly elevated in the hundreds. Results of motor nerve conduction studies are normal, except for mild slowing in terminal stages of the disease. EMG shows fibrillation potentials and other signs of denervation of muscle. A secondary mitochondrial DNA depletion is sometimes demonstrated in the muscle biopsy of infants with SMA. The molecular genetic test of the SMN gene provides definite confirmation of the diagnosis.
- 16.(D). The original infection might have caused only gastrointestinal (especially *Campylobacter jejuni*, but also *Helicobacter pylori*) or respiratory tract (especially *Mycoplasma pneumoniae*) symptoms.
- 17.(B).
- 18.(B). Weakness usually begins in the lower extremities and progressively involves the trunk, the upper limbs, and, finally, the bulbar muscles, a pattern known as Landry ascending paralysis. The onset is gradual and progresses over days or weeks; the process plateaus in 1-28 days. Weakness can progress to inability or refusal to walk and later to flaccid tetraplegia. Maximal severity of weakness is usually reached by 4 wk after onset.
- 19.(A). Dysphagia and facial weakness are often impending signs of respiratory failure.
- 20.(D).
- 21.(C). Although areflexia is seen in MFS, patients do not have significant lower extremity weakness compared with Guillain Barré syndrome.
- 22.(B). Muscle biopsy is not usually required for diagnosis; specimens appear normal in early stages and show evidence of denervation atrophy in chronic stages.
- 23.(A). The dissociation between high CSF protein and a lack of cellular response in a patient with an acute or subacute polyneuropathy is diagnostic of Guillain-Barré syndrome.
- 24.(E). IVIG administered for 2, 3, or 5 days. A commonly recommended protocol is IVIG 0.4 g/kg/day for 5 consecutive days, but some studies suggest that larger doses are more effective (1g/kg/day for 2 consecutive days) and related to improved outcome.
- 25.(D). The clinical course is usually benign, and spontaneous recovery begins within 2-3 wk. The tendon reflexes are usually the last function to recover. Improvement usually follows a gradient opposite the direction of involvement: bulbar function recovering first, and lower extremity weakness resolving last.
- 26.(B). Oral prednisone (1 mg/kg/day for 1 wk, followed by a 1 wk taper) started within the 1st 3-5 days results in improved outcome and is a traditional treatment, its efficacy confirmed in a recent long-term prospective study in the United Kingdom.
- 27.(D). Active or reactivation of herpes simplex or varicella-zoster virus may be the most common cause of Bell palsy.
- 28.(D). The peroneal and tibial nerves are the earliest and most severely affected.
- 29.(B). The peroneal and tibial nerves are the earliest and most severely affected. Children with the disorder are often described as being clumsy, falling easily, or tripping over their own feet. Muscles of the anterior compartment of the lower legs become

wasted, and the legs have a characteristic stork-like contour. The muscular atrophy is accompanied by progressive weakness of dorsiflexion of the ankle and eventual footdrop. The process is bilateral but may be slightly asymmetric. Pes cavus deformities invariably develop as a result of denervation of intrinsic foot muscles, further destabilizing the gait.

30.(B). Atrophy of muscles of the forearms and hands is usually not as severe as that of the lower extremities, but in advanced cases contractures of the wrists and fingers produce a claw hand.

31.(A). Fasciculations of muscle, myalgias, and sensory symptoms do not occur. Pupillary responses to light are preserved.

32.(B). Myasthenia gravis is one of the few neuromuscular diseases in which electromyography (EMG) is more specifically diagnostic than a muscle biopsy. A decremental response is seen to repetitive nerve stimulation; the muscle potentials diminish rapidly in amplitude until the muscle becomes refractory to further stimulation. Anti-AChR antibodies should be assayed in the plasma but are inconsistently demonstrated.

33.(A). In juvenile autoimmune myasthenia gravis, unilateral or bilateral but usually asymmetrical ptosis and some degree of extraocular muscle weakness are the earliest and most constant signs.

34.(D). Serum CK determination is not a universal screening test for neuromuscular disease because many diseases of the motor unit are not associated with elevated enzymes. The CK level is characteristically elevated in certain diseases, such as Duchenne muscular dystrophy, and the magnitude of increase is characteristic for particular diseases.

35.(B). At birth, affected infants have a thin muscle mass involving axial, limb girdle, and distal muscles; severe generalized hypotonia; and diffuse weakness. Respiratory efforts may be ineffective, requiring ventilatory support. The testes are often undescended. Facial muscles may be weak, but infants do not have the characteristic facies of myotonic dystrophy. Ptosis may be a prominent feature. Ophthalmoplegia is observed in a few cases. The palate may be high. Myotubular myopathy is not associated with cardiomyopathy (mature cardiac muscle fibers normally have central nuclei). The tongue is thin, but fasciculations are not seen. Tendon stretch reflexes are weak or absent.

36.(C). Nemaline rod myopathy; Neonatal, infantile, and juvenile forms of the disease are known. The neonatal form is severe and usually fatal because of respiratory failure since birth. In the infantile form, generalized hypotonia and weakness, which can include bulbar-innervated and respiratory muscles and a very thin muscle mass, are characteristic. The head is dolichocephalic, and the palate high arched or even cleft. Muscles of the jaw may be too weak to hold it closed.

37.(A). Mutations in the RYR1 gene are the cause of both central core myopathy and malignant hyperthermia.

38.(B). Treatment might not be required, and there is gradual improvement over the 1st few months and no evidence of residual disease by 1 yr of age.

39.(D). Patients are usually severely weak and can have a flaccid tetraplegia with or without bulbar and respiratory muscle involvement. Cerebrospinal fluid (CSF) shows no pleocytosis and protein is variably normal or mildly elevated.

1. Persistent deviation of an eye in an infant requires evaluation at the age of
 - A. 1 mo
 - B. 3 mo
 - C. 6 mo
 - D. 9 mo
 - E. 12 mo

2. The **MOST** widely used visual acuity test for preschool children is
 - A. Tumbling E test
 - B. Snellen letters
 - C. Lea symbols
 - D. Allen figures
 - E. HOTV test

3. Children may have problems with going to sleep in a dark room (which may be mistaken for a behavioral problem)
Of the following, the **MOST** likely cause is
 - A. dyslexia
 - B. diplopia
 - C. nyctalopia
 - D. amaurosis
 - E. amblyopia

4. The **MOST** common cause of a dilated unreactive pupil is
 - A. internal ophthalmoplegia
 - B. tonic pupil
 - C. ocular trauma
 - D. pharmacologic blockade
 - E. Hutchinson pupil

5. Children with aniridia should be screened using renal ultrasonography every 3-6 mo until approximately 5 yr of age if there is an 11p13 region deletion because the child is at risk for
 - A. rhabdomyosarcoma
 - B. neuroblastoma
 - C. Wilms tumor

- D. gonadoblastoma
 - E. lymphoblastic leukemia
6. Horner syndrome is an important cause of
- A. dyscoria
 - B. corectopia
 - C. microcoria
 - D. aniridia
 - E. anisocoria
7. Waardenburg syndrome is characterized by all the following **EXCEPT**
- A. an autosomal dominant inheritance
 - B. lateral displacement of the inner canthi
 - C. median white forelock
 - D. defective hearing
 - E. hyperpigmentation of the skin
8. Cat's-eye reflex is seen in all the following **EXCEPT**
- A. juvenile xanthogranuloma
 - B. persistent hyperplastic primary vitreous
 - C. cicatricial retinopathy of prematurity
 - D. retinoschisis
 - E. larval granulomatosis
9. Hypertropia is seen in
- A. 3rd nerve palsy
 - B. 4th nerve palsy
 - C. 5th nerve palsy
 - D. 6th nerve palsy
 - E. 7th nerve palsy
10. Bilateral facial palsy is a distinctive feature of
- A. Möbius syndrome
 - B. Brown syndrome
 - C. Parinaud syndrome
 - D. Duane syndrome
 - E. Bardet-Biedl syndrome
11. Spasmus nutans is a special type of acquired nystagmus in childhood. In its complete form, it is characterized by the triad of head nodding, torticollis and
- A. pendular nystagmus
 - B. latent nystagmus

- C. Seesaw nystagmus
- D. downbeat nystagmus
- E. gaze-paretic nystagmus

12. All the following are causes of blepharitis **EXCEPT**

- A. Molluscum virus
- B. Phthirus pubis
- C. Staphylococcus epidermidis
- D. Staphylococcus aureus
- E. Streptococci

13. All the following are options in the treatment of congenital nasolacrimal duct obstruction **EXCEPT**

- A. nasolacrimal massage
- B. cleansing of the lids with cold water
- C. topical antibiotic
- D. systemic antibiotics
- E. probing with topical anesthesia

14. All the following agents are used as prophylaxis to prevent neonatorum ophthalmia **EXCEPT**

- A. 0.5% erythromycin drops
- B. 1% silver nitrate drops
- C. povidone iodine (2% solution)
- D. single dose of ceftriaxone
- E. saline irrigation

15. Cicatricial adhesion between the conjunctiva of the lid and the globe is called

- A. dermoid cyst
- B. pterygium
- C. pinguecula
- D. symblepharon
- E. parinaudoculoglandular syndrome

16. Keratoconus is characterized by progressive thinning and bulging of the central cornea (cone shaped), which of the following is a sign of keratoconus?

- A. Munson sign
- B. Dalrymple sign
- C. Stellwag sign
- D. von Graefe sign
- E. Pseudohypopyon sign

17. Epibulbar dermoids are choristomas found in 75% of
- A. Marfan syndrome
 - B. craniosynostosis
 - C. Alport syndrome
 - D. Goldenhar syndrome
 - E. Cogan syndrome
18. All the following are options in the treatment of dendritic keratitis **EXCEPT**
- A. trifluridine
 - B. topical ganciclovir
 - C. systemic acyclovir
 - D. cycloplegic agent
 - E. topical corticosteroids
19. The **MOST** serious organism which can rapidly destroy stromal tissue and lead to corneal perforation is
- A. Neisseria gonorrhoea
 - B. Pseudomonas aeruginosa
 - C. Staphylococcus
 - D. Streptococcus
 - E. Yersinia
20. Several metabolic diseases produce distinctive corneal changes in childhood. Fine opacities radiating in a whorl or fan-like pattern is seen in
- A. cystinosis
 - B. mucopolysaccharidosis
 - C. gangliosidosis
 - D. Fabry disease
 - E. Wilson disease
21. The ectopia lentis (displacement of the lens) is often downward and forward, and the lens tends to be small and round in
- A. Marfan syndrome
 - B. Homocystinuria
 - C. Weill-Marchesani syndrome
 - D. Sulfite oxidase deficiency
 - E. Ehlers-Danlos syndrome
22. Anterior uveitis is caused by
- A. toxoplasmosis
 - B. toxocariasis
 - C. cytomegalovirus

- D. rubella
- E. brucellosis

23. The risk factors associated with retinopathy of prematurity (ROP) are not fully known, but prematurity and the associated retinal immaturity at birth represent the major factors. Contributory factors include all the following **EXCEPT**

- A. oxygenation
- B. apnea
- C. heart disease
- D. hypercarbia
- E. polycythemia

24. Retinopathy of prematurity (ROP) are classified into 5 stages. The stage which is characterized by the presence of a ridge and development of extraretinal fibrovascular tissue is

- A. stage 1
- B. stage 2
- C. stage 3
- D. stage 4
- E. stage 5

25. Retinoblastoma is the most common primary malignant intraocular tumor of childhood. It is unusual for a child to present with a retinoblastoma after the age of

- A. 1 yr
- B. 2 yr
- C. 3 yr
- D. 4 yr
- E. 5 yr

26. The initial sign in the majority of patients with retinoblastoma is

- A. strabismus
- B. leukocoria
- C. pseudohypopyon
- D. hyphema
- E. vitreous hemorrhage

27. During the last decade there has been a dramatic shift in the treatment of retinoblastomas, by markedly reduced use of

- A. external beam radiation
- B. systemic chemotherapy
- C. laser therapy
- D. cryotherapy

E. brachytherapy

28. Cherry-red spots (a bright to dull red spot at the center of the macula surrounded and accentuated by a grayish white or yellowish halo) are seen in all the following

EXCEPT

- A. Tuberous sclerosis
- B. Tay-Sachs disease
- C. Sandhoff disease
- D. Sulfatide lipidosis
- E. Niemann-Pick disease

29. Phakomas are seen in all the following **EXCEPT**

- A. Sturge-Weber syndrome
- B. von Hippel-Lindau disease
- C. von Recklinghausen disease
- D. Niemann-Pick disease
- E. Tuberous sclerosis

30. The first sign in hypertensive retinopathy is

- A. retinal edema
- B. flame-shaped hemorrhages
- C. cotton-wool spots
- D. papilledema
- E. irregular narrowing of the arteriole

31. Retinopathy in subacute bacterial endocarditis is present in approximately

- A. 5% of cases
- B. 20% of cases
- C. 40% of cases
- D. 60% of cases
- E. 80% of cases

32. Vascular tortuosity, arterial and venous occlusions, "salmon patches," refractile deposits, pigmented lesions, arteriolarvenous anastomoses, and neovascularization (with "sea-fan" formations); is retinopathy of

- A. Iron deficiency anemia
- B. polycythemia vera
- C. leukemia
- D. sickling disorders
- E. β -thalassemia

33. Optic neuritis is inflammation or demyelination of the optic nerve with attendant impairment of function, all the following may cause optic neuritis **EXCEPT**
- A. lead poisoning
 - B. chloramphenicol
 - C. vincristine
 - D. methylprednisolone
 - E. hydroxychloroquine
34. The symptoms of infantile glaucoma include the classic triad of
- A. photophobia ,conjunctival injection, and blepharospasm
 - B. tearing, photophobia , and blepharospasm
 - C. tearing, photophobia , and ocular enlargement
 - D. corneal enlargement, photophobia , and blepharospasm
 - E. tearing, corneal edema , and blepharospasm
35. Of the following, the **MOST** common benign tumor of the orbit is
- A. hemangioma
 - B. rhabdomyosarcoma
 - C. lymphosarcoma
 - D. metastatic neuroblastoma
 - E. teratoma
36. Treatment of hyphema include all the following **EXCEPT**
- A. bed rest
 - B. nonsteroidal anti-inflammatory drugs
 - C. cycloplegic agent
 - D. topical steroids
 - E. systemicsteroids

- 1.(C).
- 2.(A). The tumbling E test, in which the child indicates which direction the E is facing, is the most widely used visual acuity test for preschool children.
- 3.(C). Nyctalopia, or night blindness, is vision that is defective in reduced illumination.
- 4.(D). The most common cause of a dilated unreactive pupil is purposeful or accidental instillation of a cycloplegic agent, particularly atropine and related substances.
- 5.(C).
- 6.(E). Anisocoria means inequality of the pupils. Dyscoria is abnormal shape of the pupil, and corectopia is abnormal pupillary position. Microcoria (congenital miosis) appear as a small pupil that does not react to light or accommodation and that dilates poorly, if at all, with medication. The term aniridia is a misnomer because iris tissue is usually present, although it is hypoplastic.
- 7.(E). Waardenburg syndrome is characterized principally by pigmentary disturbances (usually a median white forelock and patches of hypopigmentation of the skin).
- 8.(A).
- 9.(B).
- 10.(A).
- 11.(A). The nystagmus is characteristically very fine, very rapid, horizontal, and pendular; it is often asymmetric, sometimes unilateral. Signs usually develop within the 1st yr or 2 of life. Components of the triad may develop at various times. In many cases, the condition is benign and self-limited, usually lasting a few months, sometimes years.
- 12.(E).
- 13.(B). Cleansing of the lids with warm water.
- 14.(E). The eye should be irrigated initially with saline every 10-30 min, gradually increasing to 2-hr intervals until the purulent discharge has cleared, this is as part of treatment but not as prevention. An infant born to a woman who has untreated gonococcal infection should receive a single dose of ceftriaxone, 50 mg/kg (maximum 125 mg) IV or IM, in addition to topical prophylaxis.
- 15.(D).
- 16.(A). Signs of keratoconus include Munson sign (bulging of the lower eyelid on looking downward) and the presence of a Fleischer ring (a deposit of iron in the epithelium at the base of the cone). Myogenic retraction of the upper lid occurs in thyrotoxicosis, in which it is associated with 3 classic signs: a staring appearance (Dalrymple sign), infrequent blinking (Stellwag sign), and lag of the upper lid on downward gaze (von Graefe sign).

17.(D). A dermoid usually appears as a well-circumscribed rounded or oval, gray or pinkish-yellow mass with a dry surface from which short hairs may protrude. It may affect only the superficial layers of the cornea, although full-thickness involvement is common.

18.(E). Topical use of corticosteroids causes exacerbation of superficial herpetic disease of the eye and may lead to corneal perforation; eye drops combining steroids and antibiotics are therefore to be avoided in treatment of red eye unless there are clear-cut indications for their use and close supervision during therapy.

19.(B).

20.(D).

21.(C). In most cases of Marfan syndrome, the lens is displaced superiorly and temporarily, it is almost always bilateral and relatively symmetric. In homocystinuria, the lens is usually displaced inferiorly and sometimes nasally.

22.(E). The causes of posterior uveitis are numerous; the more common are toxoplasmosis, histoplasmosis, cytomegalic inclusion disease, sarcoidosis, syphilis, tuberculosis, and toxocariasis.

23.(E). Anemia and the need for transfusion are thought by some to be contributory factors.

24.(C).

25.(C).

26.(B).

27.(A). Because those children who are irradiated during their 1st yr of life are 2-8 times more likely to develop second cancers as those irradiated after 1 yr of age. Patients treated with radiation tend to develop brain tumors and sarcomas of the head and neck. Secondary cataracts can also develop from radiation.

28.(A).

29.(D).

30.(E).

31.(C). The lesions include hemorrhages, hemorrhages with white centers (Rothspots), papilledema, and, rarely, embolic occlusion of the central retinal artery.

32.(D).

33.(D). Treatment of optic neuritis include high-dose intravenous methylprednisolone may help to speed the visual recovery in young adults, and it may prevent the development of multiple sclerosis in those at risk.

34.(B). The symptoms of infantile glaucoma include the classic triad of epiphora (tearing), photophobia (sensitivity to light), and blepharospasm (eyelid squeezing). Each can be attributed to corneal irritation. Signs of glaucoma include corneal edema, corneal and ocular enlargement, and conjunctival injection.

35.(A). Among malignant neoplasms, rhabdomyosarcoma, lymphosarcoma, and metastatic neuroblastoma are the most frequent.

36.(B). All nonsteroidal anti-inflammatories and aspirin must be avoided.

1. Purulent otorrhea is a sign of all the following conditions **EXCEPT**
 - A. otitis externa
 - B. otitis media with perforation of the tympanic membrane
 - C. drainage from the middle ear through a patent tympanostomy tube
 - D. skull base fracture
 - E. drainage from first branchial cleft sinus

2. Bloody otorrhea is associated with all the following **EXCEPT**
 - A. neoplasm
 - B. trauma
 - C. foreign body
 - D. branchial cleft sinus
 - E. blood dyscrasia

3. The **MOST** common cause of dizziness in young children is
 - A. cholesteatoma in the mastoid or middle ear
 - B. eustachian tube middle ear disease
 - C. labyrinthitis
 - D. vestibular neuronitis
 - E. benign paroxysmal vertigo

4. The **MOST** common infectious cause of congenital sensorineural hearing loss is
 - A. Measles virus
 - B. Cytomegalovirus
 - C. Toxoplasma gondii
 - D. Rubella virus
 - E. Treponema pallidum

5. The **MOST** common cause of bacterial meningitis that results in sensorineural hearing loss after the neonatal period is
 - A. H. influenza
 - B. E. Coli
 - C. Streptococcus pneumoniae
 - D. Listeria monocytogenes
 - E. Staphylococcus aureus

6. Of the following, the congenital infectious pathogen implicated in sensorineural hearing loss in children is
- A. Measles virus
 - B. Mumps virus
 - C. Streptococcus pneumoniae
 - D. Lymphocytic choriomeningitis virus
 - E. H. influenza
7. All the following are causes of sudden sensorineural hearing loss **EXCEPT**
- A. autoimmune disease
 - B. Epstein Barr virus infection
 - C. thromboembolic event
 - D. trauma
 - E. Rubella virus infection
8. All the following are included in the normal flora of the external ear canal **EXCEPT**
- A. Coagulase negative staphylococcus
 - B. Micrococcus
 - C. Diphtheroids
 - D. Pseudomonas aeruginosa
 - E. E. coli
9. Of the following, the major trigger for otitis externa is
- A. eczema
 - B. trauma
 - C. excessive wetness
 - D. foreign body
 - E. previous infection
10. The **MOST** common cause of otitis externa is
- A. Pseudomonas aeruginosa
 - B. Enterobacter aerogenes
 - C. Proteus mirabilis
 - D. Streptococci
 - E. Diphtheroids
11. The predominant symptom of otitis externa is
- A. tenderness by pressure on the tragus
 - B. otalgia
 - C. itching
 - D. hearing loss

- E. otorrhea
12. The prominent sign of acute otitis externa is
- A. white cerumen
 - B. swollen and tender canal
 - C. opaque tympanic membrane
 - D. otorrhea
 - E. periauricular lymphadenopathy
13. Necrotizing malignant otitis externa is probable with the finding of
- A. white cerumen
 - B. swollen and tender canal
 - C. facial paralysis
 - D. otorrhea
 - E. periauricular lymphadenopathy
14. The **MOST** common causative organism of necrotizing otitis externa is
- A. *Pseudomonas aeruginosa*
 - B. *Enterobacter aerogenes*
 - C. *Proteus mirabilis*
 - D. Streptococci
 - E. Diphtheroids
15. A major physical examination point to differentiate externa otitis from mastoiditis and otitis media is
- A. visualization of tympanic membrane
 - B. pus discharge from ear canal
 - C. pain on manipulation of the auricle
 - D. periauricular lymphadenopathy
 - E. hearing loss
16. Of the following, the treatment of choice for otomycosis is topical
- A. polymyxin
 - B. ciprofloxacin
 - C. clotrimazole
 - D. hydrocortisone
 - E. neomycin
17. The **MOST** effective prophylaxis for recurrent otitis externa is
- A. instillation of dilute alcohol immediately after bathing

- B. avoidance of swimming during the episode
 - C. ear protection
 - D. topical neomycin
 - E. use of hair dryer after swimming
18. The peak incidence of otitis media (OM) is in the age of
- A. 1-2 yr of life
 - B. 4-5 yr of life
 - C. 6-7 yr of life
 - D. 9-11 yr of life
 - E. 13-15 yr of life
19. The leading reason for physician visits and for use of antibiotics among children is
- A. urinary tract infection
 - B. lower respiratory tract infection
 - C. otitis media
 - D. infectious gastroenteritis
 - E. lymphadenitis
20. The mainstay of pain management for acute otitis media is
- A. acetaminophen
 - B. topical lidocaine
 - C. tympanostomy
 - D. myringotomy
 - E. narcotic analgesics
21. An important predictor for the development of recurrent and chronic otitis media is
- A. gender
 - B. race
 - C. age
 - D. genetic background
 - E. bottle feeding
22. The **MOST** likely reason for the higher rate of otitis media in infants is
- A. early exposure to unusual organisms
 - B. bottle feeding
 - C. oral appreciation of infancy
 - D. less developed immunologic defenses

- E. high incidence of respiratory tract infections
23. Middle ear aspirates in children with bronchiolitis regularly contain
- A. Respiratory syncytial virus
 - B. Parainfluenza virus
 - C. Streptococcus pneumonia
 - D. Pseudomonas aeruginosa
 - E. Staphylococcus aureus
24. Using standard culture techniques, the pathogens typically found in acute otitis media with effusion are recoverable in
- A. 10%
 - B. 30%
 - C. 50%
 - D. 70%
 - E. 90%
25. The reason behind progressive decline in the occurrence of otitis media as children grow older is
- A. frequent exposure to the causative organisms
 - B. improved immune response
 - C. progressive reduction in eustachian tube wall compliance
 - D. the more-use of over-the counter medications
 - E. reduced incidence of respiratory tract infections
26. Otitis media is a universal finding in
- A. Down syndrome
 - B. cleft lip
 - C. Patau syndrome
 - D. Ig A deficiency
 - E. cleft palate
27. Symptoms of acute otitis media include the following **EXCEPT**
- A. irritability
 - B. change in sleep habit
 - C. upper respiratory tract symptoms
 - D. fever
 - E. facial pain

28. Of the following, the **LEAST** sensitive and specific symptom of acute otitis media is
- A. pulling at the ear
 - B. pain
 - C. pus discharge
 - D. hearing loss
 - E. fullness in the ear
29. The **MOST** sensitive finding of tympanic membrane in determining middle ear effusion is
- A. contour
 - B. translucency
 - C. color
 - D. mobility
 - E. structural changes
30. The **MOST** common finding of the tympanic membrane in middle ear effusion is
- A. bulging
 - B. impairment of mobility
 - C. opacification
 - D. white color
 - E. reduced translucency
31. The **MOST** specific finding of the tympanic membrane in acute otitis media is
- A. bulging
 - B. impairment of mobility
 - C. perforation
 - D. pale color
 - E. reduced translucency
32. Purulent otorrhea of a recent onset is indicative of
- A. acute otitis externa
 - B. chronic otitis externa
 - C. acute otitis media
 - D. chronic otitis media
 - E. otitis media with effusion
33. The following features specify non-typeable H. influenza otitis media **EXCEPT**
- A. purulent conjunctivitis

- B. often occurs in multiple family members
 - C. often affects infants
 - D. ineffective topical ocular antibiotics
 - E. bulging of tympanic membrane
34. "Watchful waiting" and analgesics use in otitis media treatment is the right choice in
- A. a four-month-old infant having presumed diagnosis of acute otitis media
 - B. a three-year-old girl having unilateral acute otitis media without otorrhea
 - C. a one-year-old infant having bilateral acute otitis media with otorrhea
 - D. a twelve-year-old girl having unilateral acute otitis media and fever of 39⁰ c
 - E. an eighteen-month-old boy having bilateral acute otitis media without otorrhea
35. The **MOST** crucial aspect in the treatment of acute otitis media is
- A. choosing the right antibiotic
 - B. skillful examination of the ear
 - C. culture of middle ear aspirate
 - D. accurate diagnosis
 - E. analgesics use
36. Of the following, the recommended first line treatment of acute otitis media is
- A. amoxicillin
 - B. cefdinir
 - C. cefuroxime
 - D. clindamycin
 - E. cefpodoxime
37. All the following are at greatest risk of harboring resistant bacteria in otitis media **EXCEPT**
- A. those younger than 2 years of age
 - B. those in regular contact with large group of children
 - C. those who recently have received antimicrobial treatment
 - D. those who have bilateral otitis media with toxicity
 - E. those who are immune compromised
38. Second line treatment for acute otitis media includes all the following agents **EXCEPT**

- A. trimethoprim
- B. cefdinir
- C. amoxicillin-clavulanate
- D. cefuroxime
- E. ceftriaxone

39. Indications for myringotomy in children with acute otitis media include all the following **EXCEPT**

- A. severe refractory pain
- B. hyperpyrexia
- C. facial paralysis
- D. immune compromised
- E. failure of first course of antibiotics

40. The **MOST** common etiologic organism behind chronic suppurative otitis media is

- A. Klebsiella
- B. Proteus
- C. Pseudomonas
- D. E. Coli
- E. Candida

41. Gradenigo syndrome is the triad of

- A. suppurative otitis media, paralysis of the internal rectus muscle, and pain in the contralateral orbit
- B. suppurative otitis media, paralysis of the external rectus muscle, and pain in the ipsilateral orbit
- C. acute otitis media, paralysis of the external rectus muscle, and pain in the ipsilateral orbit
- D. acute otitis media, paralysis of the external rectus muscle, and pain in the contralateral orbit
- E. suppurative otitis media, paralysis of the internal rectus muscle, and pain in the ipsilateral orbit

42. Bezold abscess refers to

- A. neck abscess that is originated from mastoiditis involving the temporal bone
- B. mastoid bone abscess that is originated from mastoiditis
- C. orbital cavity abscess that is originated from mastoiditis involving the temporal bone

- D. brain abscess that is originated from mastoiditis involving the temporal bone
 - E. paranasal abscess that is originated from mastoiditis
43. All the following are common organisms in all variants of acute mastoiditis **EXCEPT**
- A. *S. Pneumoniae*
 - B. non-typable *H. Influenza*
 - C. *Pseudomonas aeruginosa*
 - D. *Klebsiella* spp.
 - E. group A *Streptococcus*
44. Otitic hydrocephalus (a complication of chronic otitis media) is commonly associated with
- A. meningitis
 - B. lateral sinus thrombosis
 - C. subdural abscess
 - D. brain abscess
 - E. facial paralysis

- 1.(D).
- 2.(D).
- 3.(B).
- 4.(B).
- 5.(C).
- 6.(D).
- 7.(E).
- 8.(E). The normal flora of the external canal consists mainly of aerobic bacteria and includes Coagulase-negative staphylococci, *Corynebacterium* (diphtheroids), *Micrococcus*, and, occasionally, *Staphylococcus aureus*, Viridans streptococci, and *Pseudomonas aeruginosa*.
- 9.(C). Excessive wetness (swimming, bathing, increased environmental humidity) and dryness (dry canal skin and lack of cerumen) are the major triggers.
- 10.(A). External otitis (swimmer's ear) is caused most commonly by *P. aeruginosa*.
- 11.(B).
- 12.(D). Edema of the ear canal, erythema, and thick clumpy otorrhea are prominent signs of the acute disease.
- 13.(C). Rarely, facial paralysis, other cranial nerve abnormalities, vertigo, and/or sensorineural hearing loss are present. If these occur, necrotizing (malignant) otitis externa is probable.
- 14.(A).
- 15.(C).
- 16.(C). A fungal infection of the external auditory canal, or otomycosis, is characterized by fluffy white debris, sometimes with black spores seen; treatment includes cleaning and application of antifungal solutions such as clotrimazole or nystatin; other antifungal agents include m-cresyl acetate, gentian violet, and thimerosal.
- 17.(A). The most effective prophylaxis is instillation of dilute alcohol or acetic acid (2%) immediately after swimming or bathing.
- 18.(A). The peak incidence and prevalence of OM is during the 1st 2 yr of life. More than 80% of children will have experienced at least 1 episode of OM by the age of 3 yr.
- 19.(C). Otitis media is a leading reason for physician visits and for use of antibiotics and figures importantly in the differential diagnosis of fever. It often serves as the sole or the main basis for undertaking the most frequently performed operations in infants and young children; myringotomy with insertion of tympanostomy tubes and adenoidectomy. OM is also the most common cause of hearing loss in children.

- 20.(A). Acetaminophen and Ibuprofen are the mainstay of pain management in acute otitis media.
- 21.(C). The age of onset of OM is an important predictor of the development of recurrent and chronic OM, with earlier age of onset having an increased risk for exhibiting these difficulties later in life.
- 22.(D). Less developed immunologic defenses and less-favorable eustachian tubal factors involving both the structure and function of the tube.
- 23.(C). Acute otitis media is a known complication of bronchiolitis; middle-ear aspirates in children with bronchiolitis regularly contain bacterial pathogens, suggesting that respiratory syncytial virus is rarely, if ever, the sole cause of their acute otitis media.
- 24.(B).
- 25.(C). Progressive reduction in tubal wall compliance with increasing age may explain the progressive decline in the occurrence of OM as children grow older.
- 26.(E). In children with cleft palate, where OM is a universal finding, a main factor underlying the chronic middle-ear inflammation appears to be impairment of the opening mechanism of the eustachian tube.
- 27.(E).
- 28.(A).
- 29.(D).
- 30.(B).
- 31.(A).
- 32.(C).
- 33.(E). This is a common finding in acute otitis media regardless the causative organism.
- 34.(B). Younger children, children with otorrhea, and children with bilateral acute otitis media have a significantly enhanced benefit from antimicrobial therapy in comparison to older children, children without otorrhea, or children with unilateral acute otitis media.
- 35.(D).
- 36.(A). Amoxicillin 80-90 mg/kg/day in 2 divided doses for 10 days.
- 37.(D). Those who have bilateral otitis media with toxicity.
- 38.(A). Drugs chosen for second-line treatment should be effective against β -lactamase-producing strains of non typeable H. influenzae and M. catarrhalis and against susceptible and most non- susceptible strains of S. pneumoniae. Only 4 antimicrobial agents meet these requirements: amoxicillin-clavulanate, cefdinir, cefuroxime axetil, and intramuscular ceftriaxone.
- 39.(E). Myringotomy should be considered as third-line therapy in patients that have failed 2 courses of antibiotics for an episode of AOM.
- 40.(C). The most common etiologic organisms are P. aeruginosa and S. aureus; however, the typical acute otitis media bacterial pathogens may also be the cause, especially in younger children or in the winter months.

41. **(B)**. Suppurative otitis media, paralysis of the external rectus muscle, and pain in the ipsilateral orbit.
42. **(A)**. Neck abscess that is originated from mastoid bone infection involving the temporal bone.
43. **(D)**.
44. **(B)**.

The Skin QUESTIONS

ZUHAIR ALMUSAWI, ZAHRA ALMUSAWI

1. The apocrine glands are located in the following areas **EXCEPT**
 - A. axillae
 - B. upper chest
 - C. areolae
 - D. perianal
 - E. genital

2. Nail growth is relatively slow; complete fingernail regrowth takes
 - A. 2 mo
 - B. 4 mo
 - C. 6 mo
 - D. 8 mo
 - E. 10 mo

3. The presenting symptom of Behçet disease is
 - A. erythema nodosum
 - B. recurrent aphthous stomatitis
 - C. genital ulcerations
 - D. perianal ulceration
 - E. purpuric lesions

4. One of the following may support a diagnosis of drug eruption
 - A. neutrophilia
 - B. basophilia
 - C. eosinophilia
 - D. lymphocytosis
 - E. monocytosis

5. In the use of topical medication, consideration of vehicle is as important as the specific therapeutic agent. Acute weeping lesions respond best to
 - A. ointments
 - B. gels
 - C. solutions
 - D. wet compresses
 - E. pastes

6. Topical antibiotics have been used for many years to treat local cutaneous infections. Of the following, the **MOST** effective topical agents currently available is

- A. polysporin
- B. bacitracin
- C. fusidic acid
- D. gentamycin
- E. tetracycline

7. Topical corticosteroids are potent anti-inflammatory agents and effective antipruritic agents.

Medium-potency topical corticosteroids include

- A. betamethasone
- B. fluocinonide
- C. halcinonide
- D. clobetasol
- E. hydrocortisone butyrate

8. Systemic adverse effects of high-potency and superpotent topical steroids occur with long-term use and include

- A. telangiectasia
- B. acneiform eruptions
- C. purpura
- D. cataracts
- E. increased hair growth

9. A 3-day-old neonate presented with numerous firm, yellow-white, 1-2 mm papules and pustules with a surrounding erythematous flare in several sites of the body surface but the palms and soles were spared. Intralesional contents demonstrated eosinophils in Wright-stained smears and culture was sterile.

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

- A. pyoderma
- B. erythema toxicum
- C. candidiasis
- D. herpes simplex
- E. transient neonatal pustular melanosis

10. Redundant skin over the posterior part of the neck is common in the following syndromes **EXCEPT**

- A. Ehlers-Danlos
- B. Turner
- C. Noonan
- D. Down

E. Klippel-Feil

11. Hypohidrotic ectodermal dysplasia is characterized by the following **EXCEPT**

- A. dry skin
- B. sparse scalp hair
- C. normal sexual hair growth
- D. anodontia
- E. delayed sexual development

12. Capillary malformations (CMs) are present at birth. The most effective treatment for CM is with the pulsed-dye laser. Therapy can begin

- A. in infancy
- B. after 5 yr
- C. after 10 yr
- D. during puberty
- E. after puberty

13. A 3-day-old neonate presented with unvarying red purple hue restricted to left lower limb with atrophic underlying subcutaneous tissue. The lesions become more pronounced during changes in environmental temperature, physical activity, or crying.

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

- A. reticulate capillary malformation
- B. cutis marmorata telangiectatica congenital
- C. harlequin color change
- D. cutis marmorata
- E. cutis verticis gyrata

14. A 25-day-old neonate presented with bright red, protuberant, compressible, and sharply demarcated lesion on right cheek.

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

- A. strawberry hemangioma
- B. cavernous hemangioma
- C. superficial infantile hemangioma
- D. deep infantile hemangioma
- E. tufted angioma

15. All the following treatments can be used in Kasabach-Merritt phenomenon **EXCEPT**

- A. corticosteroids
- B. aminocaproic acid
- C. vincristine
- D. interferon- α
- E. propranolol

16. In the usual patient with infantile hemangiomas (IH) who has no serious complications or extensive growth resulting in tissue destruction and severe disfigurement, treatment consists of

- A. expectant observation
- B. pulsed-dye laser therapy
- C. topical timolol solution
- D. oral propranolol
- E. oral corticosteroids

17. In a disfiguring, life or vision threatening, or ulcerated infantile hemangiomas IH; the first-line treatment is

- A. oral corticosteroids
- B. oral propranolol
- C. intralesional corticosteroid injection
- D. vincristine
- E. interferon- α

18. Evaluate for PHACES in any patient with facial hemangioma involving significant area of face, one of the following is not a feature of PHACES

- A. posterior fossa abnormalities
- B. arterial cerebrovascular abnormalities
- C. coarctation of the aorta
- D. ear abnormalities
- E. sternal abnormalities

19. Ataxia-telangiectasia is transmitted as an autosomal recessive trait; the characteristic telangiectasias develop at approximately 3 yr of age, first on

- A. bulbar conjunctivae
- B. nasal bridge
- C. malar areas
- D. external ears
- E. upper anterior chest

20. The association of spindle cell hemangiomas with nodular enchondromas in the metaphyseal or diaphyseal cartilaginous portion of long bones is known as

- A. Angiokeratoma Corporis Diffusum
- B. Osler-Weber-Rendu Disease
- C. Angiokeratoma of Mibelli
- D. Kaposiform Hemangioendothelioma
- E. Maffucci syndrome

21. All the following are risk factors for development of melanoma **EXCEPT**

- A. xeroderma pigmentosum
- B. dark complexion
- C. excessive sun exposure
- D. giant congenital nevus
- E. immunosuppression

22. An 8-year-old female child presented with a solitary, asymptomatic, smooth, dome shaped, blue-gray papule 8 mm in diameter on the dorsal aspect of her hand.

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

- A. nevus spilus
- B. nevus of ota
- C. Spitz nevus
- D. blue nevus
- E. epidermal nevus

23. The Peutz-Jeghers syndrome is characterized by melanotic macules on the lips and mucous membranes and by gastrointestinal (GI) polyposis.

Of the following, The **MOST** constant feature is

- A. diffuse hyperpigmentation of the nails
- B. buccal mucosal macules
- C. jejunal polyposis
- D. melena
- E. episodic abdominal pain

24. The following syndrome has a strong association with Café-au-lait spots

- A. Turner syndrome
- B. McCune-Albright syndrome
- C. Russell-Silver syndrome
- D. Noonan syndrome
- E. Rubinstein-Taybi syndrome

25. The hallmark of Waardenburg type 1 is the

- A. white forelock
- B. depigmented skin
- C. deafness
- D. heterochromia irides
- E. unibrow (synophrys)

26. The following is a feature of generalized (nonsegmental) vitiligo

- A. often occurs in the face
- B. usually not accompanied by other autoimmune diseases
- C. progressive, with flare-ups

- D. involves hair compartment soon after onset
- E. usually responsive to autologous grafting

27. The **MOST** common agent implicated in the etiology of erythema multiforme (EM) is

- A. Mycoplasma pneumonia
- B. Herpes simplex virus (HSV) labialis
- C. Herpes simplex virus HSV genitalis
- D. Streptococcus pneumonia
- E. Staphylococcus epidermidis

28. Erythema multiforme (EM) is characterized by an abrupt, symmetric cutaneous eruption, **MOST** commonly on the

- A. face
- B. trunk
- C. legs
- D. palms and soles
- E. extensor upper extremities

29. Stevens-Johnson syndrome is defined as affected body surface area

- A. <5%
- B. <10%
- C. <15%
- D. <20%
- E. <30%

30. All the following are advised in the treatment of Stevens-Johnson syndrome **EXCEPT**

- A. topical ocular steroid
- B. topical oral anesthetics
- C. systemic antibiotics for documented urinary or cutaneous infections
- D. prophylactic systemic antibiotics
- E. IV immunoglobulin

31. Toxic epidermal necrolysis is characterized by the following **EXCEPT**

- A. widespread blister formation
- B. skin tenderness
- C. target lesions
- D. sudden onset
- E. confluent erythema

32. Severe mucosal blistering; GI involvement; laryngeal involvement (airway obstruction); and urologic involvement are the extracutaneous clinical features of

- A. epidermolysis bullosa simplex– generalized (AD)
 - B. epidermolysis bullosa simplex–Dowling- Meara (AD)
 - C. junctional epidermolysis bullosa–herlitz (AR)
 - D. dominant dystrophic epidermolysis bullosa (AD)
 - E. recessive dystrophic epidermolysis bullosa–hallopeau- siemens (AR)
33. Pemphigus vulgaris (PV) is a rare autoimmune blistering disorder, best treated initially with
- A. IVIG
 - B. methotrexate
 - C. azathioprine
 - D. cyclophosphamide
 - E. systemic methylprednisolone
34. Dermatitis herpetiformis eruption is characterized by the following **EXCEPT**
- A. symmetric
 - B. intensely pruritic
 - C. mostly on knees and elbows
 - D. usual mucous membranes involvement
 - E. occasional hemorrhagic lesions on the palms and soles
35. Most cases of drug-induced linear IgA dermatosis are related to
- A. vancomycin
 - B. anticonvulsants
 - C. ampicillin
 - D. cyclosporine
 - E. captopril
36. Diaper dermatitis can be treated with following measures **EXCEPT**
- A. frequent changing of the diapers
 - B. overwashing
 - C. application of a protective barrier agent
 - D. application of topical sucralfate
 - E. application of 2.5% topical hydrocortisone
37. One of the **MOST** common causes of allergic contact dermatitis is
- A. neomycin
 - B. topical antihistamines
 - C. topical anesthetics
 - D. topical corticosteroids
 - E. ethylenediamine

38. Pityriasis alba occurs mainly in children and causes lesions that are
- A. hyperpigmented
 - B. well-defined
 - C. severely erythematous
 - D. finely scaly
 - E. intensely pruritic
39. Seborrheic dermatitis is a chronic inflammatory disease most common in infancy and adolescence, it is characterized by the following **EXCEPT**
- A. manifest in 1st mo of life
 - B. self-resolves by 1 yr
 - C. cradle cap
 - D. pruritis
 - E. post inflammatory pigmentary changes
40. Initial management for infantile seborrheic dermatitis is
- A. conservative
 - B. low-potency topical corticosteroids
 - C. topical antifungal
 - D. topical calcineurin inhibitors
 - E. oral antifungal agents
41. Phototoxic drug eruptions can be caused by
- A. nalidixic acid
 - B. tetracyclines
 - C. chlorothiazides
 - D. sulfonamides
 - E. barbiturates
42. The **MOST** common photosensitive reaction seen in children is acute sunburn, effective treatment of the desquamative phase is with
- A. topical corticosteroids
 - B. bland emollient
 - C. cool compresses
 - D. aloe vera products
 - E. calamine lotion
43. Guttate psoriasis can be confused with the following **EXCEPT**
- A. viral exanthems
 - B. secondary syphilis
 - C. nummular dermatitis
 - D. pityriasis rosea

E. pityriasis lichenoides chronica (PLC)

44. The treatment of psoriasis should be viewed as a 4-tier process.

The second tier of therapy is

- A. phototherapy
- B. high-potency corticosteroids
- C. tar preparations
- D. methotrexate
- E. infliximab

45. The generalized eruption of pityriasis rosea resembles a number of other diseases.

Of the following, the **MOST** important is

- A. secondary syphilis
- B. drug eruptions
- C. viral exanthems
- D. guttate psoriasis
- E. nummular dermatitis

46. Ichthyosis vulgaris is the most common of the disorders of keratinization; with an incidence of 1/250 live births. Onset generally occurs in the 1st yr of life.

Scaling is **MOST** prominent on the

- A. extensor aspects the legs
- B. abdomen
- C. neck
- D. face
- E. scalp

47. In X-Linked ichthyosis, scaling is **MOST** pronounced on the

- A. sides of the neck
- B. elbow flexures
- C. knee flexures
- D. palms
- E. soles

48. A keloid is a sharply demarcated, benign, dense growth of connective tissue that forms in the dermis after trauma.

Of the following, the **LEAST** characteristic feature of keloid is

- A. firm
- B. raised
- C. pink
- D. rubbery
- E. pruritic

49. A 10-year-old child with anterior uveitis developed a firm, smooth, erythematous papules that gradually enlarge to form annular plaques with a papular border and a normal, slightly atrophic central area several centimeters in size on the dorsum of the hands and feet.

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

- A. tinea corporis
- B. rheumatoid nodules
- C. granuloma annulare
- D. necrobiosis lipoidica
- E. xanthomas

50. A 12-year-old child presented with brawny edema of the face and neck that spreads rapidly to involve the thorax and arms. The face acquires a waxy, mask-like appearance. The involved areas feel indurated, woody, nonpitting, and are not sharply demarcated from normal skin.

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

- A. scleredema of buschke
- B. scleroderma
- C. morphea
- D. dermatomyositis
- E. subcutaneous fat necrosis

51. Beaded papules on the eyelids is the classic sign of

- A. pseudoxanthoma elasticum
- B. Ehlers-danlos syndrome
- C. cutis laxa
- D. lipoid proteinosis
- E. anetoderma

52. Cutis laxa is characterized by all the following **EXCEPT**

- A. widespread folds of lax skin
- B. characteristic facial features
- C. hypermobility of the joints
- D. normal tensile strength of the skin
- E. hoarse cry

53. Classic Ehlers-Danlos syndrome is characterized by the following **EXCEPT**

- A. post-term birth
- B. skin hyperelasticity
- C. easy bruising
- D. severe joint hypermobility
- E. subcutaneous spheroids

54. Urticaria pigmentosa is the most common form of mastocytosis. It can be confused with the following **EXCEPT**

- A. drug eruptions
- B. herpes simplex
- C. pigmented nevi
- D. insect bites
- E. bullous impetigo

55. The etiology is unknown in 30-50% of pediatric cases of erythema nodosum; one of the **MOST** common known etiologies in children is

- A. group A streptococcal infection
- B. cat-scratch disease
- C. leprosy
- D. mycoplasma
- E. brucellosis

56. A rare but potentially life threatening complication of subcutaneous fat necrosis is

- A. hyperkalemia
- B. hypercalcemia
- C. acidosis
- D. septicemia
- E. seizures

57. Histopathologic changes in sclerema neonatorum consist of

- A. increased size of fat cells
- B. fat necrosis
- C. inflammation
- D. giant cells
- E. calcium crystals

58. Infiltrative or destructive disorders that may produce atrophy of sweat glands by pressure or scarring include the following **EXCEPT**

- A. scleroderma
- B. acrodermatitis chronica atrophicans
- C. burns
- D. ichthyoses
- E. Sjögren syndrome

59. The following drugs may cause hyperhidrosis **EXCEPT**

- A. antipyretics
- B. antiemetics
- C. insulin

- D. opiates
- E. ciprofloxacin

60. The following disorders are associated with anhidrosis by unknown mechanisms **EXCEPT**

- A. dehydration
- B. uremia
- C. cirrhosis
- D. Addison disease
- E. hypothyroidism

61. Apocrine bromhidrosis develops after puberty as a result of the formation of short-chain fatty acids and ammonia by the action of the following bacteria on axillary apocrine sweat

- A. anaerobic diphtheroids
- B. group A streptococcal infection
- C. *S. aureus*
- D. *S. epidermidis*
- E. *Pseudomonas aerogenosa*

62. Nail pitting or grooves are usually seen in the following hair disorders

- A. telogen effluvium
- B. trichotillomania
- C. tinea capitis
- D. alopecia areata
- E. traction alopecia

63. Anagen effluvium is an acute, severe, diffuse inhibition of growth of anagen follicles, resulting in loss of >80-90% of scalp hair. All the following are causes of anagen effluvium **EXCEPT**

- A. radiation
- B. alkylating agents
- C. thallium
- D. thiouracil
- E. hypervitaminosis D

64. Nail anomalies are common in certain congenital disorders; large nails are seen in

- A. Rubinstein-Taybi syndrome
- B. Ellis-van Creveld syndrome
- C. ectodermal dysplasias
- D. trisomy 18 syndrome
- E. Turner syndrome

65. Terry nails are characterized by a white ground glass appearance of the entire or the proximal end of the nail and a normal pink distal 1-2 mm of the nail; this finding is associated with

- A. leprosy
- B. tuberculosis
- C. Hodgkin disease
- D. arsenic poisoning
- E. hypoalbuminemia

66. Yellow nail syndrome manifests as thickened, excessively curved, slow-growing yellow nails without lunulae.

Associated systemic diseases include the following **EXCEPT**

- A. bronchiectasis
- B. recurrent bronchitis
- C. chylothorax
- D. chronic hepatitis
- E. focal edema of the limbs and face

67. Onycholysis indicates separation of the nail plate from the distal nail bed; drugs which may cause onycholysis include

- A. meropenem
- B. vancomycin
- C. adriamycin
- D. indomethacin
- E. cyclophosphamide

68. Beau lines are transverse grooves in the nail plate. They are usually indicative of periodic trauma or episodic shutdown of the nail matrix secondary to the following systemic diseases **EXCEPT**

- A. mumps
- B. measles
- C. pneumonia
- D. celiac disease
- E. hand-foot-and-mouth disease

69. Nail changes may be particularly associated with various other diseases.

The following matchings are true **EXCEPT**

- A. psoriasis : pitting, yellow-brown discoloration, and thickening
- B. lichen planus : violaceous papules in the proximal nail bed
- C. Darier disease: red or white streaks that extend longitudinally
- D. alopecia areata: Transverse rows of fine pits
- E. acrodermatitis enteropathica: subungual parakeratotic scaling and thickening

70. Periungual fibromas that appear in late childhood should suggest a diagnosis of

- A. mucous cysts
- B. junctional nevi
- C. Lichen planus
- D. tuberous sclerosis
- E. subungual exostoses

71. Angular cheilitis is characterized by inflammation and fissuring at the corners of the mouth, often with associated erosion, maceration, and crusting.

Of the following, the **LEAST** likely cause is

- A. chapping
- B. chronic lip lickers
- C. excessive salivation
- D. nutritional deficiencies
- E. contact dermatitis to toothpaste

72. Aphthous stomatitis consists of solitary or multiple painful ulcerations occur on the labial, buccal, lingual, sublingual, palatal, or gingival mucosa; in severe, debilitating cases, systemic therapy with all the following may be helpful **EXCEPT**

- A. corticosteroids
- B. colchicine
- C. antibiotics
- D. dapsone
- E. thalidomide

73. A 7-year-old child presented with painless, fluctuant, tense, 7mm, bluish papule on the floor of the mouth.

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

- A. mucocele
- B. Epstein pearls
- C. Fordyce spots
- D. noma
- E. canker sores

74. The **MOST** common lesions that precede nonbullous impetigo are

- A. scabies
- B. burns
- C. insect bites
- D. chickenpox
- E. pediculosis

75. Staphylococcal scalded skin syndrome is caused predominantly by phage group 2 staphylococci, particularly strains 71 and 55, which are present at localized sites of infection.

Of the following, the **MOST** common focus of infection is

- A. nasopharynx
- B. umbilicus
- C. urinary tract
- D. superficial abrasion
- E. conjunctivae

76. Ecthyma resembles nonbullous impetigo in onset and appearance but gradually evolves into a deeper, more chronic infection.

Of the following, the usual causative agent is

- A. *S. aureus*
- B. *S. pneumoniae*
- C. *H. influenzae* type b
- D. *Clostridium perfringens*
- E. group A- β hemolytic streptococcus (GABHS)

77. Ecthyma gangrenosa is a necrotic ulcer covered with a gray-black eschar. It usually occurs in immunosuppressed patients with neutropenia.

Of the following, the usual causative agent is

- A. *S. aureus*
- B. *P. aeruginosa*
- C. *S. pneumoniae*
- D. *H. influenzae* type b
- E. *Clostridium perfringens*

78. Furunculosis is more common in individuals with low

- A. serum zinc level
- B. serum iron level
- C. serum sodium level
- D. serum calcium level
- E. serum phosphorus level

79. Vaccination with BCG characteristically produces a papule approximately

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

80. All the following matching are true **EXCEPT**

- A. Tinea capitis: dermatophyte infection of the scalp
- B. Tinea corporis: infection of the glabrous skin
- C. Tinea cruris: infection of the axilla
- D. Tinea pedis: infection of the toe webs
- E. Tinea unguium: dermatophyte infection of the nail plate

81. A 3-month-old infant presented with an intensely erythematous, confluent plaque with a scalloped border, a sharply demarcated edge, and satellite pustules in the perianal skin, perineum, and inguinal folds.

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

- A. seborrheic dermatitis
- B. atopic dermatitis
- C. primary irritant contact dermatitis
- D. candidal diaper dermatitis
- E. bacterial dermatitis

82. A 4-year-old child presented with a 2 pearly, skin-colored, smooth, dome-shaped, 4 mm papules with a central umbilication on the face.

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

- A. molluscum contagiosum
- B. ectopic sebaceous glands
- C. keratoacanthoma
- D. warty dyskeratoma
- E. cryptococcosis

83. All the following can be used in the treatment of papular urticaria **EXCEPT**

- A. oral antihistamines
- B. cool compresses
- C. potent topical corticosteroids
- D. topical antihistamines
- E. systemic steroids

84. Scabies is transmitted only rarely by fomites because the isolated mite dies within

- A. 2-3 hours
- B. 12-24 hours
- C. 2-3 days
- D. 5-7 days
- E. 10-20 days

85. In an immunocompetent host, scabies is frequently heralded by intense pruritus, particularly at night.

Of the following, the classic lesions of scabies are

- A. bullae
- B. pustules
- C. wheals
- D. red papules
- E. threadlike burrows

86. The treatment of choice for scabies is

- A. oral ivermectin
- B. lindane 1% cream
- C. sulfur ointment 5-10%
- D. permethrin 5% cream
- E. crotamiton 10% cream

87. The treatment of choice for head lice is

- A. spinosad
- B. pyrethroids
- C. malathion
- D. lindane shampoo
- E. benzyl alcohol lotion

88. Comedonal acne, particularly of the central face, is frequently the first sign of pubertal maturation. It occurs in

- A. 20% of adolescents
- B. 40% of adolescents
- C. 60% of adolescents
- D. 80% of adolescents
- E. 100% of adolescents

89. All the following drugs can induce acneiform lesions in susceptible individuals **EXCEPT**

- A. gold
- B. isoniazid
- C. phenytoin
- D. vitamin B₁
- E. phenobarbital

90. Little evidence shows that ingestion of the following foods can trigger acne flares

- A. high carbohydrate foods
- B. high spicy foods
- C. high sweet foods
- D. high fatty foods

E. no certain foods

91. All topical preparations for acne vulgaris must be used for 6-8 wk before their effectiveness can be assessed.

Of the following, the primary topical preparation should be

- A. azelaic acid
- B. salicylic acid
- C. topical retinoid
- D. Benzoyl peroxide
- E. topical clindamycin

92. Antibiotics are indicated for treatment of patients whose acne has not responded to topical medications.

Of the following, the antibiotic that may cause bluish discoloration of the skin and mucous membranes is

- A. tetracycline
- B. doxycycline
- C. minocycline
- D. erythromycin
- E. trimethoprim-sulfamethoxazole

93. Approximately 20% of normal neonates demonstrate acne in the 1st mo of life. Treatment is usually unnecessary. If desired, the lesions can be treated effectively with

- A. azelaic acid
- B. salicylic acid
- C. topical retinoid
- D. benzoyl peroxide
- E. topical clindamycin

94. The nodules **MOST** commonly seen in children are

- A. fibrofolliculomas
- B. epidermoid cysts
- C. pilar cyst
- D. pilomatricoma
- E. trichoepithelioma

95. A 7-year-old child presented with a smooth, pearly, pink, telangiectatic ulcerated papule that enlarge slowly on his face.

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

- A. pyogenic granuloma
- B. nevocellular nevus

- C. epidermal inclusion cyst
- D. dermatofibroma
- E. basal cell carcinoma

96. All the following are features of acrodermatitis enteropathica **EXCEPT**

- A. a rare autosomal recessive disorder
- B. caused by an inability to absorb sufficient zinc from the diet
- C. Initial signs and symptoms usually occur in the 2nd year of life
- D. the cutaneous eruption consists of vesiculobullous, eczematous, dry, scaly, skin lesions
- E. alopecia

97. Ocular manifestations of acrodermatitis enteropathica include all the following **EXCEPT**

- A. photophobia
- B. cataract
- C. conjunctivitis
- D. blepharitis
- E. corneal dystrophy

98. The replacement of elemental zinc for individuals with inherited acrodermatitis enteropathica is equal to

- A. 1 mg/kg/24 hr
- B. 3 mg/kg/24 hr
- C. 5 mg/kg/24 hr
- D. 7 mg/kg/24 hr
- E. 10 mg/kg/24 hr

99. The hair is sparse, thin, and depigmented, sometimes displaying a “flag sign” which is a characteristic finding in

- A. pellagra
- B. scurvy
- C. vitamin A deficiency
- D. kwashiorkor
- E. acrodermatitis enteropathica

100. Facial eruption that frequently follows a butterfly distribution and a dermatitis encircling the neck “Casal’s necklace.” is usually seen in

- A. pellagra
- B. scurvy
- C. vitamin A deficiency
- D. kwashiorkor

E. acrodermatitis enteropathica

101. The **MOST** common risk factor for scurvy in children is

- A. prolonged antibiotics use
- B. psychiatric disease
- C. exclusive breast feeding
- D. chronic diarrhea
- E. cow milk protein allergy

- 1.(B).
- 2.(C). Nail growth is relatively slow; complete fingernail regrowth takes 6 mo, while complete toe nail regrowth requires 12-18 mo.
- 3.(B). Recurrent aphthous stomatitis is present in almost all patients and is commonly the presenting symptom.
- 4.(C). Medication eruptions begin on the trunk 7-10 days after exposure; they spread peripherally and are associated with pruritus and, less commonly, with fever, arthralgia, and lymphadenopathy. Eosinophilia may support a diagnosis of drug eruption but may be absent in the setting of bone marrow suppression. Penicillins, sulfa drugs, cephalosporins, nonsteroidal anti-inflammatory drugs, anticonvulsants, and aminoglycosides are common offenders.
- 5.(D). Acute weeping lesions respond best to wet compresses, followed by lotions or creams.
- 6.(C). Mupirocin, fusidic acid, and retapamulin are the most effective topical agents currently available and are as effective as oral erythromycin in treatment of mild to moderate impetigo.
- 7.(A). Corticosteroids can be divided into 7 different categories on the basis of strength, but for practical purposes, 4 categories can be used: low, moderate, high, and super. Low-potency preparations include hydrocortisone, desonide, and hydrocortisone butyrate. Medium-potency compounds include amcinonide, betamethasone, flurandrenolide, fluocinolone, mometasone furoate, and triamcinolone. High-potency topical steroids include fluocinonide and halcinonide. Betamethasone dipropionate and clobetasol propionate are superpotent preparations and should be prescribed with care.
- 8.(D). Poor growth, cataracts, and suppression of adrenal function.
- 9.(B). The cause of erythema toxicum is unknown. The lesions can mimic pyoderma, candidiasis, herpes simplex, transient neonatal pustular melanosis, and miliaria but can be differentiated by the characteristic infiltrate of eosinophils and the absence of organisms on a stained smear.
- 10.(A). Loose folds of skin must be differentiated from a congenital defect of elastic tissue or collagen such as cutis laxa, Ehlers-Danlos syndrome, or pseudoxanthoma elasticum. Redundant skin over the posterior part of the neck is common in the Turner, Noonan, Down, and Klippel-Feil syndromes and monosomy 1p36; more generalized folds of skin occur in infants with trisomy 18 and short-limbed dwarfism.
- 11.(E). Sexual development is usually normal.
- 12.(A). Therapy can begin in infancy, when the surface area of involvement is smaller.

13.(B).

14.(C). Superficial IHs are bright red, protuberant, compressible, sharply demarcated lesions that may occur on any area of the body. Although sometimes present at birth, they more often appear in the 1st 2 mo of life and are heralded by an erythematous or blue mark or an area of pallor, which subsequently develops a fine telangiectatic pattern before the growth phase.

15.(E).

16.(A). Expectant observation, because almost all lesions regress spontaneously, therapy is rarely indicated. Parents require repeated reassurance and support.

17.(B). In a disfiguring, life- or vision-threatening, or ulcerated IH that is not responding to other treatment, oral propranolol is the first-line treatment in most cases. IHs typically responds with growth arrest and often early signs of involution within a couple weeks of treatment initiation.

18.(D). **PHACES** stands for **p**osterior fossa brain defects such as Dandy-Walker malformation or cerebellar hypoplasia, large segmental facial infantile **h**emangioma, **a**rterial cerebrovascular abnormalities such as aneurysms and stroke, **c**oarctation of the aorta, **e**ye abnormalities. **S**ternal raphe defects such as pits, scars, or **s**upraumbilical raphe are infrequently observed.

19.(A). The characteristic telangiectasias develop at approximately 3 yr of age, first on the bulbar conjunctivae and later on the nasal bridge, malar areas, external ears, hard palate, upper anterior chest, and antecubital and popliteal fossae. Additional cutaneous stigmata include café-au-lait spots, premature graying of the hair, and sclerodermatous changes. Progressive cerebellar ataxia, neurologic deterioration, sinopulmonary infections, and malignancies are also seen.

20.(E).

21.(B). Fair complexion.

22.(D).

23.(B). Buccal mucosal macules are the most constant feature of the disorder; in some families, occasional members may be affected only with the pigmentary changes.

24.(B). All other distracters have weak association.

25.(A). The hallmark of Waardenburg type 1 is the white forelock, which is seen in 20-60% of patients. Only 15% of patients have areas of depigmented skin. Deafness occurs in 9-37%, heterochromia irides in 20%, and unibrow (synophrys) in 17-69% of those affected. Dystopia canthorum (i.e., telecanthus) is seen in all patients with Waardenburg type 1.

26.(C). All other distracters are features of segmental type.

27.(B). HSV labialis and, less commonly, HSV genitalis are implicated in 60-70% of episodes of EM and are believed to trigger nearly all episodes of recurrent EM, frequently in association with sun exposure.

28.(E). Lesions are relatively sparse on the face, trunk, and legs. Lesions can be seen on the palms and soles.

29.(B). SJS is defined as affected body surface area <10%, SJS-TEN overlap syndrome is affected body surface area between 10% and 30%, and TEN is affected body surface area >30%.

30.(D). Systemic antibiotics are indicated for documented urinary or cutaneous infections and for suspected bacteremia (*Staphylococcus aureus* or *Pseudomonas aeruginosa*) because infection is the leading cause of death. Prophylactic systemic antibiotics are not necessary.

31.(C). TEN is defined by (1) widespread blister formation and morbilliform or confluent erythema, associated with skin tenderness; (2) absence of target lesions; (3) sudden onset and generalization within 24-48 hr; (4) histologic findings of full-thickness epidermal necrosis and a minimal-to-absent dermal infiltrate. These criteria categorize TEN as a separate entity from EM.

32.(C).

33.(E). The disease is best treated initially with systemic methylprednisolone 1-2 mg/kg/day. Azathioprine, cyclophosphamide, and methotrexate therapy all have been useful in maintenance regimens.

34.(D). Mucous membranes are usually spared.

35.(A).

36.(B). Overwashing should be avoided because it leads to chapping and a worsening of the dermatitis.

37.(A). Neomycin sulfate is present in many nonprescription topical antibiotic preparations, and thus children are frequently exposed at an early age. It is one of the most common causes of allergic contact dermatitis, and use of combination products of neomycin with other antibiotics, antifungals, or corticosteroids may induce co-reactivity with these chemically-unrelated substances.

38.(D). Pityriasis alba occurs mainly in children and causes lesions that are hypopigmented, ill-defined, round or oval patches. They may be mildly erythematous and finely scaly. Itching is minimal or absent.

39.(D). A greasy, scaly, erythematous papular dermatitis, which is usually nonpruritic in infants, may involve the face, neck, retroauricular areas, axillae, umbilicus, and diaper area.

40.(A). Initial management for infantile seborrheic dermatitis is generally conservative given the self-limited nature of this condition. Emollients, baby oil, gentle shampooing with non-medicated baby shampoo and gentle use of a soft brush to remove scales are usually effective measures.

41.(A). Other distracters cause photoallergic drug eruptions.

42.(B). A bland emollient is effective in the desquamative phase.

43.(C). The differential diagnosis of plaque type psoriasis includes nummular dermatitis, tinea corporis, seborrheic dermatitis, postinfectious arthritis syndromes, and pityriasis rubra pilaris.

44.(A).

45.(A).

- 46.(A). Scaling is most prominent on the extensor aspects of the extremities, particularly the legs. Flexural surfaces are spared, and the abdomen, neck, and face are relatively uninvolved.
- 47.(A). Scaling is most pronounced on the sides of the neck, lower face, preauricular areas, anterior trunk, and the limbs, particularly the legs.
- 48.(E). They may be tender or pruritic.
- 49.(C).
- 50.(A).
- 51.(D). Lipoid proteinosis may be noted initially in early infancy as hoarseness. Skin lesions appear during childhood and consist of yellowish papules and nodules that may coalesce to form plaques. The classic sign is beaded papules on the eyelids. Lesions also occur on the face, forearms, neck, genitals, dorsum of the fingers, and scalp, where they result in patchy alopecia.
- 52.(C). Hyperelasticity and hypermobility of the joints are not present as they are in the Ehlers-Danlos syndrome.
- 53.(A). Premature birth caused by rupture of membranes.
- 54.(B). The differential diagnosis of solitary mastocytomas includes recurrent bullous impetigo, herpes simplex, congenital melanocytic nevi, and juvenile xanthogranuloma. Urticaria pigmentosa can be confused with drug eruptions, postinflammatory pigmentary change, juvenile xanthogranuloma, pigmented nevi, ephelides, xanthomas, chronic urticaria, insect bites, and bullous impetigo. Diffuse cutaneous mastocytoma may be confused with epidermolytic hyperkeratosis.
- 55.(A). Most common etiologies in children include: group A streptococcal infection, *Yersinia enterocolitica* gastroenteritis, medications (cephalosporins, penicillins, macrolides), and inflammatory disorders (inflammatory bowel disease); sarcoidosis should be considered in young adults.
- 56.(B). A rare but potentially life threatening complication is hypercalcemia. It manifests at 1-6 mo of age as lethargy, poor feeding, vomiting, failure to thrive, irritability, seizures, shortening of the QT interval on electrocardiography, or renal failure.
- 57.(A). Histopathologic changes in sclerema neonatorum consist of increases in the size of fat cells and in the width of the fibrous connective tissue septa. In contrast to SCFN, with which this disorder is mostly to be confused, fat necrosis, inflammation, giant cells, and calcium crystals are generally absent.
- 58.(D). Obstruction of sweat glands may occur in miliaria and in a number of inflammatory and hyperkeratotic disorders, such as the ichthyoses, psoriasis, lichen planus, pemphigus, porokeratosis, atopic dermatitis, and seborrheic dermatitis.
- 59.(B). Emetics.
- 60.(E). Diverse disorders that are associated with anhidrosis by unknown mechanisms include dehydration; toxic overdose with lead, arsenic, thallium, fluorine, or morphine; uremia; cirrhosis; endocrine disorders such as Addison disease, diabetes mellitus, diabetes insipidus, and hyperthyroidism.

61.(A).

62.(D).

63.(E). Hypervitaminosis A.

64.(A). Large nails are seen in Pachyonychia congenita, Rubinstein-Taybi syndrome, hemihypertrophy, while smallness or absence of nails are seen in Ectodermal dysplasias, nail-patella, dyskeratosis congenita, focal dermal hypoplasia, cartilage-hair hypoplasia, Ellis-van Creveld, Larsen, epidermolysis bullosa, incontinentia pigmenti, Rothmund-Thomson, Turner, popliteal web, trisomy 13, trisomy 18, Apert, Gorlin-Pindborg, long arm 21 deletion, otopalatodigital, fetal alcohol, fetal hydantoin, elfin facies, anonychia, and acrodermatitis enteropathica.

65.(E).

66.(D). Deficient lymphatic drainage, caused by hypoplastic lymphatic vessels, is believed to lead to the manifestations of this syndrome.

67.(D). Common causes are trauma, long-term exposure to moisture, hyperhidrosis, cosmetics, psoriasis, fungal infection (distal onycholysis), atopic or contact dermatitis, porphyria, drugs (bleomycin, vincristine, retinoid agents, indomethacin, chlorpromazine [Thorazine]), and drug-induced phototoxicity from tetracyclines or chloramphenicol.

68.(D).

69.(E). Patients with acrodermatitis enteropathica may have transverse grooves (Beau lines) and nail dystrophy as a result of periungual dermatitis.

70.(D).

71.(D). Nutritional deficiencies are a less-frequent etiology.

72.(C). Treatment of aphthous stomatitis is palliative. The majority of mild cases do not require therapy. Relief of pain, particularly before eating, may be achieved with the use of a topical anesthetic such as viscous lidocaine or an oral rinse with a combined solution of elixir of diphenhydramine, viscous lidocaine, and an oral antacid.

Caution must be taken to avoid hot food and drink after topical anesthetic use. A superpotent topical corticosteroid in a mucosa-adhering agent may help reduce inflammation, and topical tetracycline mouthwash may also hasten healing. In severe, debilitating cases, systemic therapy with corticosteroids, colchicine, dapsone, or thalidomide may be helpful.

73.(A). Lesions on the floor of the mouth are known as ranulas when the sublingual or submandibular salivary gland ducts are involved. Fluctuations in size are typical, and the lesions may disappear temporarily after traumatic rupture. Recurrence is prevented by surgical excision of the mucus deposit and associated salivary gland(s).

74.(C).

75.(A). Foci of infection include the nasopharynx and, less commonly, the umbilicus, urinary tract, a superficial abrasion, conjunctivae, and blood.

76.(E). The causative agent is usually GABHS; *S. aureus* is also cultured from most lesions but is probably a secondary pathogen.

- 77.(B). Ecthyma gangrenosum occurs in up to 6% of patients with systemic *P. aeruginosa* infection but can also occur as a primary cutaneous infection by inoculation.
- 78.(B). Furunculosis is also more common in individuals with low serum iron levels, diabetes, malnutrition, HIV infection, or other immunodeficiency states.
- 79.(B). Vaccination with BCG characteristically produces a papule approximately 2 wk after vaccination. The papule expands in size, typically ulcerates within 2-4 mo, and heals slowly with scarring.
- 80.(C). Tinea cruris, or infection of the groin, occurs most often in adolescent males and is usually caused by the anthropophilic species *Epidermophyton floccosum* or *T. rubrum*, but occasionally by the zoophilic species *T. mentagrophytes*.
- 81.(D).
- 82.(A). They typically have a central umbilication from which a plug of cheesy material can be expressed. The papules may occur anywhere on the body, but the face, eyelids, neck, axillae, and thighs are sites of predilection.
- 83.(D). Topical antihistamines are potent immunologic sensitizers and have no role in the treatment of insect bite reactions.
- 84.(C).
- 85.(E). Threadlike burrows are the classic lesion of scabies but may not be seen in infants.
- 86.(D). The treatment of choice for scabies is permethrin 5% cream (Elimite) applied to the entire body from the neck down, with particular attention to intensely involved areas, which is also standard therapy.
- 87.(C). Because of resistance of head lice to pyrethroids, malathion 0.5% in isopropanol is the treatment of choice for head lice and should be applied to dry hair until hair and scalp are wet, and left on for 12 hr. A second application 7-9 days after initial treatment may be necessary.
- 88.(D).
- 89.(D). Diagnosis of acne is rarely difficult, although flat warts, folliculitis, and other types of acne (drug induced: glucocorticoid agents, anabolic steroids, gold, dactinomycin, isoniazid, lithium, phenytoin, progestins) may be confused with acne vulgaris. The differential diagnosis includes sarcoidosis, angiofibromas, keratosis pilaris, chloracne, rosacea, and fibrofolliculomas.
- 90.(E). Little evidence shows that ingestion of particular foods can trigger acne flares. When a patient is convinced that certain dietary items exacerbate acne, it is prudent for the patient to omit those foods.
- 91.(C). A topical retinoid should be the primary treatment for acne vulgaris.
- 92.(C). Rarely, minocycline causes dizziness, intracranial hypertension, bluish discoloration of the skin and mucous membranes, hepatitis, a lupus-like syndrome, and drug reaction with eosinophilia and systemic symptoms.
- 93.(D). Topical antifungals and/or benzoyl peroxide.
- 94.(B). Epidermoid cysts are the nodules most commonly seen in children. Such a cyst is a sharply circumscribed, dome-shaped, firm, freely movable, skin-colored nodule

often with a central dimple or punctum that is a plugged, dilated pore of a pilosebaceous follicle.

95.(E). Basal cell carcinoma is very rare in children in the absence of a predisposing condition, such as nevoid basal cell carcinoma syndrome, xeroderma pigmentosum, and nevus sebaceus of Jadassohn, arsenic intake, or exposure to irradiation. The lesions are smooth, pearly, pink, telangiectatic papules that enlarge slowly and may bleed or ulcerate. Sites of predilection are the face, scalp, and upper back.

96.(C). Initial signs and symptoms usually occur in the 1st few mo of life, often after weaning from breast milk to cow's milk.

97.(B).

98.(B). Oral therapy with zinc compounds is the treatment of choice. Replacement for individuals with inherited acrodermatitis enteropathica is with 3 mg/kg/24 hr of elemental zinc found in zinc sulfate, gluconate, or acetate (i.e., 220 mg of zinc sulfate contains 50 mg of elemental zinc). Zinc gluconate carries less risk of gastrointestinal distress.

99.(D). Nails are thin and soft, and hair is sparse, thin, and depigmented, sometimes displaying a "flag sign" consisting of alternating light and dark bands that reflect alternating periods of adequate and inadequate nutrition.

100.(A).

101.(B). In children, the most common risk factors are behavioral or psychiatric disease that results in poor nutrition.

1. Normal full-term newborns can have up to 20-30 degree hip and knee flexion contractures, these contractures tend to resolve by

- A. 1-3 mo
- B. 4-6 mo
- C. 7-9 mo
- D. 10-12 mo
- E. 13-15 mo

2. The ossification centers that are typically present at birth include all the following

EXCEPT

- A. distal femur
- B. proximal tibia
- C. distal humerus
- D. calcaneus
- E. talus

3. The gait of a child become similar to that of an adult at

- A. 3 yr
- B. 5 yr
- C. 7 yr
- D. 9 yr
- E. 11 yr

4. A rocker-bottom foot describes

- A. congenital vertical talus
- B. flexible flatfoot
- C. calcaneovalgus foot
- D. congenital talipes equinovarus
- E. metatarsus adductus

5. Polydactyly is the most common congenital toe deformity, it may be preaxial (great toe) or postaxial (5th toe), and occasionally one of the central toes is duplicated.

Which of the following statements is **TRUE**?

- A. it is seen in approximately 2 in 100,000 births
- B. it is bilateral in 10% of cases
- C. one-third of patients will also have polydactyly of the hand

- D. it may be associated with Turner syndrome
 - E. surgical removal of the extra digit is generally performed after 2 years of age
6. Macrodactyly is seen in the following conditions **EXCEPT**
- A. Proteus syndrome
 - B. Neurofibromatosis
 - C. Tuberous sclerosis
 - D. Klippel-Trenaunay-Weber syndrome
 - E. Alport syndrome
7. Pediatricians must understand normal limb development so as to recognize pathologic conditions during routine and targeted exams.
Of the following, the **TRUE** statement is
- A. normal tibiofemoral angle at birth is 10-15 degrees of physiologic valgus
 - B. physiologic varus up to 12 degrees is reached in between 3 and 4 yr of age
 - C. normal varus of 7 degrees is achieved by 5-8 yr of age
 - D. persistence of varus beyond 2 yr of age may be pathologic
 - E. more than 35% of developmental genu varum and genu valgum cases need intervention
8. In evaluation of concerns relating to the limb, the examination should assess the exact torsional profile which include all the following **EXCEPT**
- A. foot progression angle
 - B. femoral anteversion
 - C. knee – knee distance
 - D. tibial version with thigh–foot angle
 - E. assessment of foot adduction and abduction
9. In-toeing gait most commonly results from excessive femoral anteversion.
Of the following, the **TRUE** statements is
- A. it is more common in boys than girls (2 : 1)
 - B. patellas are pointing outward when the foot is straight
 - C. diagnosis is made by CT
 - D. treatment is predominantly observation
 - E. anteversion >25 degrees is an indication for operative intervention
10. Which torsional deformity can follow a slipped capital femoral epiphysis and increase incidence of degenerative arthritis?
- A. external tibial torsion
 - B. external femoral torsion
 - C. internal tibial torsion
 - D. internal femoral torsion

E. metatarsus adductus

11. A school age child presented with a mass behind the knee. Physical examination reveals a firm mass in the popliteal fossa, medially located and distal to the popliteal crease. The mass is most prominent when the knee is extended. Knee radiographs are normal.

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

- A. osteochondroma
- B. osteochondritis dissecans
- C. popliteal cyst
- D. malignancy
- E. juvenile idiopathic arthritis

12. An adolescent girl presented with pain beneath the patella, walking up and down stairs aggravates the pain; squatting, running, and other vigorous physical activities also exacerbate the pain. There is no history of antecedent trauma and there is no swelling. Pain is often relieved through knee extension.

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

- A. osteochondroma
- B. osteochondritis dissecans
- C. popliteal cyst
- D. patellofemoral pain syndrome
- E. juvenile idiopathic arthritis

13. The proximal femoral ossification center (in the center of the femoral head) appears between

- A. 1st and 3rd mo
- B. 4th and 7th mo
- C. 8th and 11th mo
- D. 12th and 15th mo
- E. 16th and 18th mo

14. There is marked geographic and racial variation in the incidence of developmental dysplasia of the hip (DDH).

All the following are true **EXCEPT**

- A. 1.7 in 1,000 babies in Sweden
- B. 75 in 1,000 in Yugoslavia
- C. 188.5 in 1,000 in a district in Manitoba, Canada
- D. 3.7 in 1,000 babies in Chinese newborn
- E. 0% in African newborn

15. The **MOST** reliable sign of a dislocated hip after the 2nd month of life is

- A. limited hip abduction
- B. apparent shortening of the thigh
- C. proximal location of the greater trochanter
- D. asymmetry of the glutealor thigh folds
- E. positive Ortolani test

16. Transient synovitis (toxic synovitis) is a reactive arthritis and is one of the most common causes of hip pain in young children.

All the following clinical manifestations are true **EXCEPT**

- A. it is most prevalent between 3 and 8 yr of age
- B. 70% of affected children have had a preceding nonspecific upper respiratory tract infection
- C. symptoms often develop acutely
- D. children are usually able to bear weight on the affected limb
- E. usually associated with a high grade fever

17. Legg-Calvé-Perthes disease is a hip disorder of unknown etiology that results from temporary interruption of the blood supply to the proximal femoral epiphysis, leading to osteonecrosis and femoral head deformity.

Of the following, the **MOST** common presenting symptom is

- A. limp of varying duration
- B. pain
- C. failure to ambulate
- D. atrophy of the muscles of the thigh
- E. an apparent leg-length inequality

18. Differential diagnosis of legg-calvé-perthes disease include all the following **EXCEPT**

- A. osteochondromatosis
- B. Schwartz-Jampel syndrome
- C. Marfan syndrome
- D. Maroteaux-Lamy syndrome
- E. Martsolf syndrome

19. Scoliosis is a complex 3-dimensional spinal deformity that is defined in the coronal plane as a curve of at least 10 degrees, measured by the Cobb method, on a posteroanterior (PA) radiograph of the spine.

Which statement is **TRUE** about this disease?

- A. adolescent idiopathic scoliosis (AIC) is 2-10 times more common in males than females
- B. fathers with AIS transmit the disease to 50% of their children
- C. high plasma melatonin levels have been noted in patients with progressive curvatures

- D. girls with AIS have been noted to have a smaller foramen magnum
- E. approximately one-third of girls with AIS have osteopenia

20. The normal thoracic spine has 20-50 degrees of kyphosis as measured from T3 to T12. A thoracic kyphosis in excess of the normal range of values is termed hyperkyphosis.

All the following conditions are associated with hyperkyphosis **EXCEPT**

- A. trauma causing spinal fractures
- B. osteogenesis imperfecta
- C. Marfan syndrome
- D. glycogen storage disease
- E. mucopolysaccharidoses

21. Torticollis, literally meaning twisted neck, is not a diagnosis but rather a manifestation of a variety of underlying conditions.

All the following conditions may cause torticollis **EXCEPT**

- A. positional deformation
- B. Klippel-Feil syndrome
- C. cervical lymphadenitis
- D. supratentorial brain tumor
- E. upper lobe pneumonia

22. Klippel-Feil syndrome includes the classic triad of a low posterior hairline, short neck, and decreased cervical range of motion.

Other associations include the following **EXCEPT**

- A. Sprengel's deformity
- B. congenital scoliosis
- C. genitourinary anomalies
- D. conductive hearing loss
- E. congenital heart disease

23. Polydactyly or duplication of a digit can occur either as a preaxial deformity (involving the thumb) or as a postaxial deformity (involving the small finger)

All the following syndromes associated with polydactyly **EXCEPT**

- A. Carpenter syndrome
- B. Ellis-van Creveld syndrome
- C. Trisomy 18
- D. Orofaciodigital syndrome
- E. Rubinstein-Taybi syndrome

24. Syndactyly is one of the common anomalies observed in the upper limb.

All the following syndromes are associated with syndactyly **EXCEPT**

- A. Carpenter syndrome
- B. Ellis-van Creveld syndrome
- C. Laurence-Moon-Biedl syndrome
- D. Fanconi pancytopenia
- E. Trisomy 13

25. Fractures are the second most common manifestation of child abuse after skin injury (bruises, burns/abrasions).

All of the following fractures suggest nonaccidental injury **EXCEPT**

- A. femur fractures in nonambulatory children
- B. distal femoral metaphyseal corner fractures
- C. posterior rib fractures
- D. scapular spinous process fractures
- E. distal humeral fractures

26. Fractures of the wrist and forearm are very common fractures in children.

All the following statements are true **EXCEPT**

- A. the most common mechanism of injury is a fall on the outstretched hand
- B. an eighty percent of forearm fractures involve the proximal radius and ulna
- C. the majority of forearm fractures are torus or greenstick fractures
- D. a significant malunion of a forearm diaphyseal fracture can lead to a permanent loss of pronation and supination
- E. the AP and lateral radiographs of the forearm and wrist confirm the diagnosis

27. All the following statements are true regarding clavicular fracture **EXCEPT**

- A. most common site for fracture is the junction of the middle and medial 3rd of clavicle
- B. biceps function is important to assess as it is a prognostic indicator for future function
- C. posterior medial clavicular physeal injuries are particularly problematic
- D. fractures heal rapidly usually in 3-6 wk
- E. a palpable mass of callus is usually visible in thin children

28. Toddler fractures occur in young ambulatory children. The age range for this fracture is typically around 1-4 yr.

All the following statement are true **EXCEPT**

- A. children in this age group are usually unable to describe the area of injury well
- B. radiographs may show no fracture
- C. classic symptom is refusal to bear weight
- D. Inflammatory markers may be ordered to rule out infectious processes
- E. fracture is treated with bed rest and analgesia for approximately 2 wk

29. All the following are indications for external fixation in pediatric fractures **EXCEPT**
- A. grades II and III open fractures
 - B. fractures associated with severe burns
 - C. fractures with soft-tissue loss
 - D. pelvic fractures
 - E. fractures with associated head injuries and spasticity
30. The two **MOST** common causative organisms of osteomyelitis in children with sickle cell anemia are
- A. *S. aureus* and streptococci
 - B. *Salmonella* spp. and *S. aureus*
 - C. *S. aureus* and *Pseudomonas aeruginosa*
 - D. Enterobacteriaceae and *S. aureus*
 - E. *S. aureus* and *Bartonella henselae*
31. The second **MOST** common cause of osteomyelitis in children younger than 5 yr of age in some parts of the world is
- A. *S. aureus*
 - B. *Kingella kingae*
 - C. *Pseudomonas aeruginosa*
 - D. Enterobacteriaceae
 - E. *Bartonella henselae*
32. The **MOST** common site of osteomyelitis in children is
- A. femur
 - B. tibia
 - C. humerus
 - D. radius
 - E. ulna
33. Septic arthritis in infants and children has the potential to damage the synovium, adjacent cartilage, and bone causing permanent disability.
- All the following statements regarding its epidemiology are true **EXCEPT**
- A. septic arthritis is more common in young children
 - B. half of all cases occur by 5 yr of age
 - C. adolescents and neonates are at risk of gonococcal septic arthritis
 - D. majority of infections in otherwise healthy children are of hematogenous origin
 - E. infection of joints following penetrating injuries are uncommon
34. Which medical condition is regarded a contraindication for sport participation?
- A. atlantoaxial instability
 - B. splenomegaly

- C. carditis
- D. long-QT syndrome
- E. advanced heart block

35. Adverse effect of anabolic–androgenic steroids include all the following **EXCEPT**

- A. infertility
- B. gynecomastia
- C. female virilization
- D. hypotension
- E. epiphyseal closure

36. Creatine is an amino acid mostly stored in skeletal muscle increasing muscle performance. Its use has been increased when other supplements have been withdrawn from the market.

Of the following, the possible adverse effect include

- A. mouth ulcer
- B. carditis
- C. hepatitis
- D. nephritis
- E. genital sore

37. In competitive swimming, the **MOST** common injuries are of the

- A. neck
- B. shoulder
- C. spine
- D. hip
- E. knee

38. Osteogenesis imperfecta, the most common genetic cause of osteoporosis.

Which of their types is mild and non-deforming?

- A. I
- B. II
- C. III
- D. IV
- E. V

39. The **MOST** severe non-lethal form of osteogenesis imperfecta which results in significant physical disability is type

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

E. V

40. The **MOST** common ocular manifestation in Marfan syndrome is

- A. dislocation of the ocular lens
- B. severe myopia
- C. flat cornea
- D. hypoplastic iris
- E. retinal detachment

41. Current pharmacologic approaches that are considered the standard of care in Marfan syndrome (MFS) are

- A. angiotensine converting enzyme inhibitors
- B. beta blockers
- C. calcium channel blockers
- D. diuretics
- E. angiotensine receptor II antagonists

- 1.(B). Normal full-term newborns can have up to 20-30 degree hip and knee flexion contractures. These contractures tend to resolve by 4-6 mo of age.
- 2.(C). The ossification centers that are typically present at birth are the distal femur, proximal tibia, calcaneus, and talus.
- 3.(C). The gait characteristics of a 7 yr old child are similar to those of an adult.
- 4.(A). Congenital vertical talus has also been described as a rocker-bottom foot or a Persian slipper foot. The plantar surface of the foot is convex, and the talar head is prominent along the medial border of the midfoot. The fore part of the foot is dorsiflexed and abducted relative to the hindfoot, and the hindfoot is in equinus and valgus.
- 5.(C). It is seen in approximately 2 in 1,000 births and is bilateral in 50% of cases. Conditions that may be associated with polydactyly include Ellis-Van Creveld (chondroectodermal dysplasia), longitudinal deficiency of the tibia, and Down syndrome. Treatment is indicated for cosmeses and to allow for fitting with standard shoes. This involves surgical removal of the extra digit, and the procedure is generally performed between 9 and 12 mo of age.
- 6.(E).
- 7.(D). The normal tibiofemoral angle at birth is 10-15 degrees of physiologic varus. The alignment changes to 0 degrees by 18 mo, and physiologic valgus up to 12 degrees is reached in between 3 and 4 yr of age. The normal valgus of 7 degrees is achieved by 5-8 yr of age. Persistence of varus beyond 2 yr of age may be pathologic and is seen in conditions such as Blount disease. Overall, 95% of developmental physiologic genu varum and genu valgum cases resolve with growth.
- 8.(C).
- 9.(D). It occurs more commonly in girls than boys (2: 1) in children 3-6 yr of age. The patellas are pointing inward when the foot is straight, and compensatory external rotation of the tibia is demonstrated. Diagnosis is made clinically on examination; CT can provide objective measurements but is rarely indicated. The torsion usually corrects with growth by 8-10 yr of age. Persistent deformity, unacceptable cosmesis, functional impairment, anteversion >45degrees, and no external rotation beyond neutral are some of the indications for operative intervention.
- 10.(B). External femoral torsion can follow a slipped capital femoral epiphysis. The disorder is associated with an out-toeing gait and increased incidence of degenerative arthritis. If slipped capital femoral epiphysis is detected, it is treated surgically.
- 11.(C). Popliteal cysts, or Baker cysts, are cystic masses filled with gelatinous material that develop in the popliteal fossa, the shallow depression located at the posterior part

of the knee. They are considered rare in children. They most commonly occur in the region of the medial head of the gastrocnemius and semimembranosus. Transillumination of the cyst on physical examination is a simple diagnostic test. Knee radiographs are normal and should be obtained to identify other lesions, such as osteochondromas, osteochondritis dissecans, and malignancies. The diagnosis may be confirmed by ultrasonography, MRI, or aspiration.

12.(D). Patellofemoral pain syndrome (PFPS) is one of the most common causes of knee pain, particularly in adolescent girls. The precise etiology of the knee pain remains unknown and is likely multifactorial.

13.(B). Between the 4th and 7th mo of life, the proximal femoral ossification center (in the center of the femoral head) appears.

14.(D). The incidence of DDH in Chinese and African newborns is almost 0%, whereas it is 1% for hip dysplasia and 0.1% for hip dislocation in white newborns. These differences may be the result of environmental factors, such as child-rearing practices, rather than to genetic predisposition.

15.(A). As the baby enters the 2nd and 3rd mo of life, the soft tissues begin to tighten and the Ortolani and Barlow tests are no longer reliable. Limitation of abduction is the most reliable sign of a dislocated hip in this age group.

16.(E). They are often afebrile or have a low-grade fever $<38^{\circ}\text{C}$ (100.4°F). The hip is not held flexed, abducted, or laterally rotated unless a significant effusion is present.

17.(A). The most common presenting symptom is a limp of varying duration.

18.(C). Differential diagnosis of Legg-Calvé-Perthes disease include: Sick cell disease, other hemoglobinopathies (e.g., thalassemia), chronic myelogenous leukemia, steroid medication, sequela of traumatic hip dislocation, treatment of developmental dysplasia of the hip, septic arthritis, mucopolysaccharidoses, and hypothyroidism.

19.(E). AIS is 2-10 times more common in females than males. Fathers with AIS *transmit* the disease to 80% of their children, but mothers with AIS *transmit* the disease to only 56% of their children. Lower plasma melatonin levels have been noted in patients with progressive curvatures. Girls with AIS have been noted to have a larger foramen magnum.

20.(D). Conditions associated with hyperkyphosis are: Trauma causing spinal fractures, spinal infections resulting from bacterial, tuberculosis, and fungal diseases, metabolic diseases such as osteogenesis imperfecta or osteoporosis, iatrogenic (laminectomy, spinal irradiation), neuromuscular diseases, neoplasms, congenital/developmental, disorders of collagen such as Marfan syndrome, dysplasias such as neurofibromatosis, achondroplasia, and mucopolysaccharidoses.

21.(D). Posterior fossa brain tumor.

22.(D). Other associations include Sprengel's deformity (congenital elevation of the scapula), congenital scoliosis, genitourinary anomalies (25-35%), sensorineural hearing loss (5%), and congenital heart disease (5-10%).

23.(C). Trisomy 13

24.(B). Ellis-van Creveld syndrome associated with polydactyly.

- 25.(E). Proximal humeral fractures.
- 26.(B). Eighty percent of forearm fractures involve the distal radius and ulna, 15% involve the middle third, and the rest are rare fractures of the proximal third of the radius or ulnar shaft.
- 27.(A). The most common site for fracture is the junction of the middle and lateral 3rd of clavicle.
- 28.(E). The fracture is treated with an above-knee cast for approximately 3 wk.
- 29.(D). Not all pelvic fractures only the unstable one.
- 30.(B). *Streptococcus pneumoniae* most commonly causes osteomyelitis in children younger than 24 mo of age and in children with sickle cell anemia, but its frequency has declined because of pneumococcal conjugate vaccines. Cases of *Pseudomonas* infection are related almost exclusively to puncture wounds of the foot, with direct inoculation of *P. aeruginosa* from the foam padding of the shoe into bone or cartilage, which develops as osteochondritis. *Bartonella henselae* can cause osteomyelitis of any bone, but especially in pelvic and vertebral bones.
- 31.(B). *Kingella kingae* may be the second most common cause of osteomyelitis in children younger than 5 yr of age in some parts of the world. The organism is increasingly recognized as a cause of osteomyelitis, spondylodiskitis, and septic arthritis, especially when polymerase chain reaction testing is employed.
- 32.(A). Femur 23-28%, tibia 20-26%, humerus 8-13%, radius and ulna 5-6%.
- 33.(A). Half of all cases occur by 2 yr of age and three-fourths of all cases occur by 5 yr of age.
- 34.(C). Carditis can result in sudden death with exertion. All the other conditions need individual assessment.
- 35.(D). Hypertension.
- 36.(D). Concerns about nephritis in case reports have not been supported by controlled studies.
- 37.(B). Injuries of the shoulder in competitive swimming are most common and generally a result of chronic, overuse. Swimmer's shoulder is a combination of subacromial bursitis and tendinosis of the rotator cuff and long-head of the biceps.
- 38.(A). I: Mild, Nondeforming, II: Lethal perinatal, III: Progressively deforming, IV: Moderately deforming
- 39.(C). Birthweight and length are often low normal. Fractures usually occur in utero. There is relative macrocephaly and triangular facies. Postnatally, fractures occur from inconsequential trauma and heal with deformity. Disorganization of the bone matrix results in a "popcorn" appearance at the metaphyses. The rib cage has flaring at the base, and pectal deformity is frequent. Virtually all type III patients have scoliosis and vertebral compression. Growth falls below the curve by the 1st yr; all type III patients have extreme short stature. Scleral hue ranges from white to blue. Dentinogenesis imperfecta, hearing loss, and kyphoscoliosis may be present or develop over time.
- 40.(A). Dislocation of the ocular lens (ectopia lentis) occurs in approximately 60-70% of patients, although it is not unique to the disorder.

41.(B). β -Blockers have traditionally been considered the standard of care in MFS and multiple small observational studies suggest there is a protective effect on aortic root growth, with the dose typically titrated to achieve a resting heart rate <100 beats/min during submaximal exercise.

Rehabilitation Medicine

QUESTIONS

ZUHAIR ALMUSAWI

1. Children with severe traumatic brain injury (TBI) may experience autonomic dysfunction characterized by all the following **EXCEPT**

- A. elevated temperature
- B. elevated heart rate
- C. elevated respiratory rate
- D. lowered blood pressure
- E. diaphoresis

2. Osteopenia begins immediately after a spinal cord injury (SCI) occurs and plateaus 6-12 mo later. Pathologic fractures occur as a consequence of loss of bone mineral density.

Of the following, the **MOST** common site of fracture is

- A. distal tibia
- B. supracondylar region of the femur
- C. lumbosacral spine
- D. proximal humerus
- E. distal radius

3. Abrupt withdrawal of baclofen may cause

- A. depression
- B. drowsiness
- C. headache
- D. euphoria
- E. seizure

4. Oral medications are often used as an early treatment for generalized spasticity. Which of the following works at the level of skeletal muscle to block calcium release from the sarcoplasmic reticulum?

- A. dantrolene sodium
- B. clonidine
- C. tizanidine
- D. baclofen
- E. diazepam

5. Coadministration of botulinum toxin (BTX) and one of the following drugs should be performed with caution as the effect of the toxin may be potentiated

- A. aminoglycosides
- B. cephalosporines
- C. penicillins
- D. macrolids
- E. sulphonamides

6. Risk factors for birth brachial plexus injury include the following **EXCEPT**

- A. shoulder dystocia
- B. birthweight greater than 4 kg
- C. primiparous mothers
- D. mothers with excessive weight gain
- E. diabetic mothers

7. The baby with birth brachial plexus palsy (BBPP) will start with occupational or physical therapy at approximately

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

8. Prenatal screening is recommended for all pregnant women to detect neural tube defect. If a neural tube defect is present, one of the following is often elevated

- A. human chorionic gonadotropin
- B. α -fetoprotein
- C. estriol
- D. inhibin
- E. prolactin

9. Hindbrain herniation or the Chiari type II malformation is seen in which percent of individuals with myelomeningocele

- A. 10-20%
- B. 20-30%
- C. 40-50%
- D. 60-70%
- E. 80-90%

- 1.(D). Children with severe TBI may experience autonomic dysfunction characterized by elevated temperature, heart rate, respiratory rate, and blood pressure, accompanied by diaphoresis and posturing. Autonomic dysfunction is a diagnosis of exclusion and is associated with poor outcome after acquired brain injury in children.
2. (B). The most common sites of fracture include the supracondylar region of the femur and the proximal tibia.
- 3.(E). Abrupt withdrawal may cause seizures, hallucinations, rebound muscle spasms, and hyperpyrexia.
- 4.(A). Dantrolene sodium works at the level of skeletal muscle to block calcium release from the sarcoplasmic reticulum. Despite its peripheral site of action, dantrolene may induce sedation, although to a lesser degree than other centrally acting agents.
- 5.(A). Coadministration of BTX and aminoglycosides or other agents interfering with neuromuscular transmission (curare-like nondepolarizing blockers, lincosamides, polymyxins, quinidine, magnesium sulfate, anticholinesterases, succinylcholine chloride) should be performed with caution as the effect of the toxin may be potentiated.
- 6.(C). Risk factors for birth brachial plexus injury include prior infants with BBPP, shoulder dystocia, birthweight greater than 4 kg, multiparous mothers, mothers with excessive weight gain, and diabetic mothers. Delivering twins or triplets, as well as cesarean sections, have been described as protective from BBPP.
- 7.(B).
- 8.(B). Prenatal screening is recommended for all pregnant women to detect neural tube defect. A simple blood test is done in the 2nd trimester to evaluate α -fetoprotein, human chorionic gonadotropin, estriol, and inhibin. If a neural tube defect is present, the α -fetoprotein is often elevated and further screening using high-resolution ultrasound is indicated.
- 9.(E).

1. In utero radiation exposure is associated with an excess risk of dying from leukemia before age 10 yr.

Of the following, the **MOST** acceptable risk percent is

- A. 32%
- B. 52%
- C. 75%
- D. 92%
- E. 180%

2. The following diseases are associated with sensitivity to radiation **EXCEPT**

- A. ataxia-telangiectasia
- B. Edward syndrome
- C. Down syndrome
- D. Fanconi anemia
- E. Gardner syndrome

3. The acute effects of radiation therapy (occurring less than 3 mo after therapy begins) are usually related to the area of the body being irradiated.

Of the following, the **MOST** severe acute reactions is

- A. pneumonitis
- B. dermatitis
- C. mucositis
- D. esophagitis
- E. cerebral edema

4. Primary malignancies with the highest cumulative incidence of a second neoplasm are

- A. soft tissues sarcoma
- B. cancers of bone
- C. Hodgkin disease
- D. leukemias
- E. central nervous system (CNS) cancers

5. Children exposed to second-hand tobacco smoke have increased frequency of

- A. middle ear effusions
- B. bacterial respiratory illnesses

- C. otitis externa
 - D. sinusitis
 - E. tonsillitis
6. Children who grow up on farms have elevated rates of
- A. mesothelioma
 - B. leukemia
 - C. skin tumor
 - D. lymphoma
 - E. carcinoma of lungs
7. Arsine gas is colorless, odorless, nonirritating, and highly toxic. Inhalation causes no immediate symptoms. After a latent period of 2-24 hr, exposed individuals experience
- A. hematuria
 - B. sensorimotor peripheral neuropathy
 - C. respiratory failure
 - D. massive hemolysis
 - E. proteinuria
8. Acute ingestion of inorganic mercury salts (typically secondary to ingestion of a button battery) can manifest in a few hours as
- A. tremor
 - B. neuropsychiatric disturbances
 - C. gingivostomatitis
 - D. corrosive gastroenteritis
 - E. necrotizing bronchiolitis
9. Acrodynia, or pink disease, is a rare idiosyncratic hypersensitivity reaction to mercury that occurs predominantly in children exposed to mercurous powders. The symptom complex includes the following **EXCEPT**
- A. generalized pain
 - B. paresthesias
 - C. an acral red-pink rash
 - D. photophobia
 - E. generalized spasticity
10. It is estimated that lead-poisoned children are identified by screening procedures rather than through clinical recognition of lead related symptoms in
- A. 99% of cases
 - B. 79% of cases
 - C. 59% of cases
 - D. 39% of cases

E. 19% of cases

11. Mushrooms are an ideal food because they are

- A. low in calories
- B. high in calories
- C. high in fat
- D. low in fat
- E. low in protein

12. One of the following is not recommended for treatment of Amanita poisoning

- A. oral activated charcoal
- B. forced diuresis
- C. silibinin
- D. intravenous penicillin G
- E. acetylcysteine

13. Mushrooms of the genera *Inocybe* contain muscarine or muscarine-related compounds. These quaternary ammonium derivatives bind to postsynaptic receptors, 'producing an exaggerated cholinergic response.

Of the following, the **MOST** serious complication is

- A. diaphoresis
- B. bradycardia
- C. hypotension
- D. vomiting
- E. bronchospasm

14. Pediatricians are likely to experience unique problems in managing childhood victims of biologic or chemical attack.

The very rapid onset of neuromuscular symptoms after an exposure should lead the clinician to consider

- A. botulism
- B. nerve agent intoxication
- C. chlorine
- D. phosgene
- E. cyanide

15. Pediatricians are likely to experience unique problems in managing childhood victims of biologic or chemical attack.

A delayed onset of respiratory symptoms (days after exposure) is characteristic of

- A. chlorine
- B. phosgene
- C. cyanide

- D. anthrax
- E. sarin

16. Pediatricians are likely to experience unique problems in managing childhood victims of biologic or chemical attack.

Patients suffering from the sudden onset of severe neuromuscular symptoms may have nerve agent intoxication and should be given atropine (0.05 mg/kg) promptly for its antimuscarinic effects. Atropine has the following effects **EXCEPT**

- A. relieves bronchospasm
- B. relieves bradycardia
- C. reduces bronchial secretions
- D. ameliorates diarrhea
- E. improve skeletal muscle paralysis

17. In cases in which the delayed onset of respiratory symptoms may be the result of a terrorist attack, consideration should be given to the empirical administration of an antibiotic effective against anthrax, plague, and tularemia. A reasonable choice is

- A. ciprofloxacin
- B. rifampin
- C. vancomycin
- D. ampicillin
- E. clarithromycin

18. Infection is the most common complication of bite injuries, regardless of the species of biting animal.

The **MOST** common microorganisms associated with cat bites are

- A. Staphylococcus species
- B. Streptococcus species
- C. Eikenella species
- D. Pasteurella species
- E. Proteus species

19. The **MOST** important proposed field treatments for snake bites is

- A. tourniquets
- B. immobilization of injured body part at the level of the heart
- C. ice
- D. incision
- E. suction

20. Specific antivenoms (AV) are available for many venomous creatures of the world, particularly snakes, spiders, and scorpions.

All the following about antivenoms are true **EXCEPT**

- A. AV is capable of neutralizing only circulating venom
- B. it is beneficial to give AV locally at the bite site
- C. most AVs are given intravenously
- D. skin tests are unreliable
- E. intravenous AV should be started slowly

21. One of the following differentiate organophosphate poisoning and methamphetamine intoxication from bark scorpion envenomations

- A. paresthesias
- B. roving eye movements
- C. cranial nerve dysfunction
- D. seizures
- E. hypertensive crisis

1.(D). In utero radiation exposure is associated with a 92% excess risk of dying from leukemia before age 10 yr and a 180% excess risk of dying from other malignant diseases. Even 40 yr later there is a 228% increased relative risk of cancer associated with radiation in utero.

2.(B). Diseases that are associated with sensitivity to radiation are

- Ataxia-telangiectasia
- Basal cell nevoid syndrome
- Cockayne syndrome
- Down syndrome
- Fanconi anemia
- Gardner syndrome
- Nijmegen breakage syndrome
- Usher syndrome

3.(A). The acute effects of therapy (occurring less than 3 mo after therapy begins) are usually related to the area of the body being irradiated (except fatigue, which can begin during this time period). These acute effects include radiation-caused pneumonitis, dermatitis, mucositis and esophagitis, cerebral edema, and swelling of the organ irradiated.

There may be changes in bowel movement patterns. Of these, one of the most severe acute reactions is pneumonitis. It can be manifest within 24 hr of irradiation when there is an exudation of proteinaceous material into the alveoli and intraalveolar edema. Most often, however, radiation pneumonitis begins 2-6 mo after the beginning of radiation with a clinical presentation of fever, cough, congestion, and pruritic pain.

4.(C). Primary malignancies with the highest cumulative incidence of a second neoplasm in the order of frequency are Hodgkin disease (7.6), soft tissuesarcoma (4.0), cancers of bone (3.3), leukemia (2.1), central nervous system (CNS) cancers (2.1), and non-Hodgkin disease lymphoma (1.9).

5.(A). Children exposed to second-hand tobacco smoke have increased frequency of lower respiratory illness, more middle-ear effusions, and more viral respiratory illnesses than unexposed children.

6.(B). Children who grow up on farms have elevated rates of leukemia; pesticides are suspected of playing an etiologic role.

7.(D). After a latent period of 2-24 hr, exposed individuals experience massive hemolysis, malaise, headache, weakness, dyspnea, nausea, vomiting, abdominal pain, hepatomegaly, pallor, jaundice, hemoglobinuria, and renal failure.

8.(D). Acute ingestion of inorganic mercury salts (typically secondary to ingestion of a button battery) can manifest in a few hours as corrosive gastroenteritis, signified by metallic taste, oropharyngeal burns, nausea, hematemesis, severe abdominal pain, hematochezia, acute tubular necrosis, cardiovascular collapse, and death.

9.(E). The symptom complex includes generalized pain, paresthesias, and an acral (hands, feet) rash that may spread to involve the face. The rash typically is red-pink, papular, pruritic, and painful; it may progress to desquamation and ulceration. Morbilliform, vesicular, and hemorrhagic variants have been described. Other important features include anorexia, apathy, photophobia, and hypotonia, especially of the pectoral and pelvic girdles. Irritability, tremors, diaphoresis, insomnia, hypertension, and tachycardia may be present. Some cases initially were diagnosed as pheochromocytoma.

10.(A). It is estimated that 99% of lead-poisoned children are identified by screening procedures rather than through clinical recognition of lead related symptoms.

11.(A). Mushrooms are an ideal food. They are low in calories, fat free, and high in protein, making them a great source of nutrition.

12.(B). Forced diuresis should be avoided, as this increases renal exposure.

13.(E). Respiratory distress caused by bronchospasm and increased bronchopulmonary secretions is the most serious complication.

14.(B). The very rapid onset of neuromuscular symptoms after an exposure should lead the clinician to consider nerve agent intoxication. The nerve agents (tabun, sarin, soman, and VX) are organophosphate analogs of common pesticides that act as potent inhibitors of the enzyme acetylcholinesterase. They are hazardous via ingestion, inhalation, or cutaneous absorption.

15.(D). A delayed onset of respiratory symptoms (days after exposure) is characteristic of several infectious diseases and 1 toxin that might be adapted for sinister purposes by terrorists. Among the most threatening and problematic of these are anthrax, plague, tularemia, and ricin, the latter having garnered considerable media attention in recent years.

16.(E). Pralidoxime (also known as 2-PAM) cleaves the organophosphate moiety from cholinesterase and regenerates intact enzyme if "aging" has not occurred. The effect is most prominent at the neuromuscular junction and leads to improved muscle strength. Its prompt use (at a dose of 25 mg/kg) as an adjunct to atropine is recommended in all serious cases.

17.(A). Ciprofloxacin (10-15 mg/kg IV q12h), levofloxacin (8 mg/kg IV q12h), or doxycycline (2.2 mg/kg IV q12h) is a reasonable choice.

18.(D). *P. multocida* is the predominant species in at least 50% of cat bite wound infections.

19.(B). Constrictive clothing, jewelry, and watches should be removed, and the injured body part should be immobilized in a position of function at the level of the heart. All proposed field treatments for snake bites, such as tourniquets, ice, electric shock,

incision, and suction, have proven problematic, with most being ineffective and deleterious.

20.**(B)**. Skin tests, often recommended by AV manufacturers, are unreliable and should be omitted.

21.**(B)**. Two important ingestions that can be confused with a bark scorpion envenomation are organophosphate poisoning and methamphetamine intoxication. The opsoclonus-like roving eye movements seen in bark scorpion envenomations are not seen in the above ingestions and may help differentiate these conditions.

1. "Normal values" (reference intervals) are difficult to establish within the pediatric population. Many variables should be considered when developing reference intervals. All the following are variables that commonly considered to define reference intervals

EXCEPT

- A. genetic composition
- B. physiologic development
- C. environmental influences
- D. subclinical disease
- E. ethnicity

2. The most commonly used reference range is generally given as the mean of the reference population ± 2 standard deviations (SD).

The term "normal distribution" refers to which of the following?

- A. gaussian distribution
- B. exponential distribution
- C. skewed distribution
- D. uniform distribution
- E. unimodal distribution

3. The closeness of a measured value to a standard or known value is termed as

- A. accuracy
- B. precision
- C. sensitivity
- D. specificity
- E. none of the above

4. You asked to obtain a measurement of serum sodium level in the hospital lab; you obtain a value of 125 mmol/L by repeating the test 5 times. The reference value of serum sodium at that lab is between 135-155 mmol/L.

Your measurement is termed

- A. accurate but not precise
- B. precise but not accurate
- C. neither accurate nor precise
- D. both accurate and precise
- E. sensitive and specific

5. Assume you are going to estimate the prevalence of amoebic dysentery in a small country which harbors a total number of population of 530,000; you find that 57,000 of the population are infected by the disease. The prevalence of this disease is closest to

- A. 5.33%
- B. 7.45%
- C. 10.75%
- D. 20.22%
- E. 25.3%

6. Assume you have the results of PCR tests for *Mycoplasma pneumoniae*; the number of positive tests is 83 collected from a 100 truly-infected persons.

The sensitivity of this test is

- A. 17%
- B. 55%
- C. 60%
- D. 83%
- E. 95%

7. Assume you are evaluating the specificity of the indirect fluorescence antibody test for rapid diagnosis of *Campylobacter enteritis*. The number of normal persons tested is 150 and the test is positive in 30 persons. The specificity of this test is

- A. 20%
- B. 40%
- C. 60%
- D. 70%
- E. 80%

8. You are assigned to evaluate the outcome of fecal occult blood test as a screening test for patients with bowel cancer. The total number of patients who underwent endoscopy is 2030; in those whom the disease is confirmed are 30. The results of fecal occult blood test are as follows: positive in 20 patients who has a confirmed bowel cancer (truly positive), and in 180 patients who don't have the disease (falsely positive); and negative in 10 patients who truly has the disease (falsely negative), and in 1820 patients who are not affected by the disease (truly negative).

All the following statements are true **EXCEPT**

- A. the prevalence of the disease for those population is 1.48%
- B. the sensitivity of the test is 67%
- C. the specificity of the test is 91%
- D. the positive predictive value is 20%
- E. the negative predictive value is 99.5%

9. Rapid HIV antibody testing procedures using a finger stick or venipuncture to obtain whole blood, plasma, or serum, and tests using oral fluid were approved.

All the following regarding this test are true **EXCEPT**

- A. they are simple and accurate as to render the likelihood of an erroneous result by the user negligible
- B. a positive result does not need confirmation by Western blot analysis or immunofluorescence assay
- C. they allow for implementation of antiretroviral therapy for HIV-infected women who have not been tested or are unaware of their HIV status
- D. they significantly reduce the risk of mother-to-child transmission
- E. The median turnaround time for obtaining results from blood collection to patient notification was only 66 minutes

10. Almost all of the diseases detected in neonatal screening programs have a very low prevalence; the strategy is to use the initial screening test to separate a highly suspect group of patients from normal infants (i.e., to increase the prevalence) and then to follow this suspect group aggressively.

Of the following, the disease that has low prevalence rendering it not useful for neonatal screening testing is

- A. phenylketonuria
- B. hypothyroidism
- C. sickle cell disease
- D. cystic fibrosis
- E. neuroblastoma

- 1.(E). Other considerations for further defining reference intervals include partitioning based on sex and age.
- 2.(A). All others are non-normal distribution.
- 3.(A). Accuracy is a measure of the nearness of a test result to the actual value, whereas precision is a measure of the reproducibility of a result.
- 4.(B). Precision is the repeatability of the measurement. The measurement can be precise but not accurate, accurate but not precise, both accurate and precise, or not accurate nor precise.
- 5.(C). Prevalence refers to the percentage of the infected people from the total number of population.
- 6.(D). Sensitivity means the probability that a patient who is disease positive will test positive. $\text{Sensitivity} = \frac{\text{number positive by test} \times 100}{\text{total number positive}}$.
- 7.(E). Specificity (also called the true negative rate) measures the proportion of negatives that are correctly identified as such (e.g., the percentage of healthy people who are correctly identified as not having the condition).

$\text{Specificity} = \frac{\text{number negative by test} \times 100}{\text{total number without disease}}$.

- 8.(D). $\text{PV of a positive test result} = \frac{\text{True-positive results} \times 100}{\text{Total positive results}}$. $\text{PV of a negative test result} = \frac{\text{True-negative results} \times 100}{\text{Total negative results}}$.
- 9.(D). A positive rapid HIV test result is then confirmed by Western blot analysis or immunofluorescence assay. Rapid whole blood HIV testing is now the standard of care for women in labor with undocumented HIV status. Rapid HIV testing can also be used in developing countries. In resource-poor settings, because of the lack of properly equipped laboratories, skilled technologists, and basic resources, such as electricity and water.
- 10.(E). A simple spot test for VMA is not useful in general screening programs because of the low prevalence of neuroblastoma (3 cases/100,000) and the low sensitivity of the test (69%). Sickle cell disease, easily detected using liquid chromatography or isoelectric focusing, can be treated more effectively if it is diagnosed before clinical signs appear. In addition, the results of neonatal screening for cystic fibrosis show that there are clear benefits associated with preclinical diagnosis.

Thanks for Allah



PUBLISHED BOOKS FOR THE AUTHOR

IRAQI ANTI-INFECTIVE DRUG GUIDE	2010
UP TO DATE IN PEDIATRICS FIRST EDITION	2010
UP TO DATE IN PEDIATRICS SECOND EDITION	2011
MCQs IN PEDIATRICS PART 1	2012
PEDIATRICS OSCE STATIONS PART 1	2013
PEDIATRICS OSCE STATIONS PART 2	2015