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Dedication
To those who are special through the ages and in present;
Those who give us inspiration;
Those who give us the spiritual power when we are near defeat;
Those who served the whole world;
We dedicate our humble work.

Special thanks are extended to our colleagues, ALI ALMUSAWI & ALI HASHIM for their help in designing the cover of this book.

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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 all written 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.

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## Contents

<table>
<thead>
<tr>
<th>SECTION 1</th>
<th>The Profession of Pediatrics</th>
</tr>
</thead>
<tbody>
<tr>
<td>QUESTIONS</td>
<td>.......................................................... 1</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>.......................................................... 4</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>SECTION 2</th>
<th>Growth and Development</th>
</tr>
</thead>
<tbody>
<tr>
<td>QUESTIONS</td>
<td>.......................................................... 5</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>.......................................................... 12</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>SECTION 3</th>
<th>Behavioral Disorders</th>
</tr>
</thead>
<tbody>
<tr>
<td>QUESTIONS</td>
<td>.......................................................... 16</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>.......................................................... 22</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>SECTION 4</th>
<th>Psychiatric Disorders</th>
</tr>
</thead>
<tbody>
<tr>
<td>QUESTIONS</td>
<td>.......................................................... 25</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>.......................................................... 34</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>SECTION 5</th>
<th>Psychosocial Issues</th>
</tr>
</thead>
<tbody>
<tr>
<td>QUESTIONS</td>
<td>.......................................................... 38</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>.......................................................... 45</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>SECTION 6</th>
<th>Pediatric Nutrition and Nutritional Disorders</th>
</tr>
</thead>
<tbody>
<tr>
<td>QUESTIONS</td>
<td>.......................................................... 48</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>.......................................................... 56</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>SECTION 7</th>
<th>Fluids and Electrolytes</th>
</tr>
</thead>
<tbody>
<tr>
<td>QUESTIONS</td>
<td>.......................................................... 60</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>.......................................................... 65</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>SECTION 8</th>
<th>The Acutely Ill or Injured Child</th>
</tr>
</thead>
<tbody>
<tr>
<td>QUESTIONS</td>
<td>.......................................................... 67</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>.......................................................... 78</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>SECTION 9</th>
<th>Human Genetics and Dysmorphology</th>
</tr>
</thead>
<tbody>
<tr>
<td>QUESTIONS</td>
<td>.......................................................... 83</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>.......................................................... 93</td>
</tr>
<tr>
<td>SECTION</td>
<td>Topic</td>
</tr>
<tr>
<td>-----------</td>
<td>--------------------------------------</td>
</tr>
<tr>
<td>10</td>
<td>Metabolic Disorders</td>
</tr>
<tr>
<td>11</td>
<td>Fetal and Neonatal Medicine</td>
</tr>
<tr>
<td>12</td>
<td>Adolescent Medicine</td>
</tr>
<tr>
<td>13</td>
<td>Immunology</td>
</tr>
<tr>
<td>14</td>
<td>Allergy</td>
</tr>
<tr>
<td>15</td>
<td>Rheumatic Diseases of Childhood</td>
</tr>
<tr>
<td>16</td>
<td>Infectious Diseases</td>
</tr>
<tr>
<td>17</td>
<td>The Digestive System</td>
</tr>
<tr>
<td>18</td>
<td>The Respiratory System</td>
</tr>
<tr>
<td>19</td>
<td>The Cardiovascular System</td>
</tr>
<tr>
<td>SECTION 20</td>
<td>Hematology</td>
</tr>
<tr>
<td>------------</td>
<td>------------</td>
</tr>
<tr>
<td>QUESTIONS</td>
<td>..................................................</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>..................................................</td>
</tr>
<tr>
<td>SECTION 21</td>
<td>Oncology</td>
</tr>
<tr>
<td>QUESTIONS</td>
<td>..................................................</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>..................................................</td>
</tr>
<tr>
<td>SECTION 22</td>
<td>Nephrology and Urology</td>
</tr>
<tr>
<td>QUESTIONS</td>
<td>..................................................</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>..................................................</td>
</tr>
<tr>
<td>SECTION 23</td>
<td>Endocrinology</td>
</tr>
<tr>
<td>QUESTIONS</td>
<td>..................................................</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>..................................................</td>
</tr>
<tr>
<td>SECTION 24</td>
<td>Neurology</td>
</tr>
<tr>
<td>QUESTIONS</td>
<td>..................................................</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>..................................................</td>
</tr>
<tr>
<td>SECTION 25</td>
<td>Dermatology</td>
</tr>
<tr>
<td>QUESTIONS</td>
<td>..................................................</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>..................................................</td>
</tr>
<tr>
<td>SECTION 26</td>
<td>Orthopedics</td>
</tr>
<tr>
<td>QUESTIONS</td>
<td>..................................................</td>
</tr>
<tr>
<td>ANSWERS</td>
<td>..................................................</td>
</tr>
</tbody>
</table>
SECTION 1 The profession of Pediatrics

QUESTIONS

ZUHAIR ALMUSAWI

1. Infants who are smaller size and relatively underweight at birth because of maternal malnutrition have increased rates of all the following EXCEPT
   A. coronary heart disease
   B. stroke
   C. type 1 diabetes mellitus
   D. obesity
   E. osteoporosis

2. The MOST common cause of death in the age group (1 to 24 years) in the United States in 2010 was
   A. accidents (unintentional injuries)
   B. assaults (homicide)
   C. suicide
   D. malignant neoplasms
   E. congenital malformations

3. The second MOST common cause of death in children (1 to 4 years of age) in the United States in 2010 was
   A. homicide
   B. congenital malformations
   C. chromosomal abnormalities
   D. malignant neoplasms
   E. diseases of the heart

4. Professionalism in pediatrics include the following EXCEPT
   A. pediatrician must treat all persons with respect
   B. pediatrician must listen attentively
   C. pediatrician must seek to learn from errors
   D. pediatricians must work cooperatively and communicate effectively with patients and their families
   E. pediatrician should take a decision when faced with new or complex responsibilities
5. All the following diseases can be diagnosed by genetic screening, even though the manifestations of the disease process do not appear until later in life EXCEPT
   A. polycystic kidney disease
   B. Huntington disease
   C. breast cancer
   D. cystic fibrosis
   E. hemochromatosis

6. In which of the following conditions, a cure is possible, but failure is not uncommon?
   A. cancer with a poor prognosis
   B. cystic fibrosis
   C. severe epidermolysis bullosa
   D. chronic respiratory failure
   E. muscular dystrophy

7. All the following regarding grief are correct EXCEPT
   A. grief has been defined as the emotional response caused by a loss, including pain, distress, and physical and emotional suffering
   B. it is a normal adaptive human response to death
   C. complicated grief is common
   D. parents, who have had closure sessions with the attending staff, are more likely to resolve their grief
   E. sharing accurate and truthful information with a dying child is beneficial

8. All the following are true regarding children's concepts of death and disease EXCEPT
   A. for children, up to 2 years of age, death is seen as separation, and there is probably no concept of death
   B. the associated behaviors in grieving children, up to 2 years of age, include protesting and difficulty of attachment to other adults
   C. children from 3 to 5 years of age have trouble grasping the meaning of the illness and the permanence of the death
   D. in children age 6 to 11 years of age, the finality of death is not realized
   E. in adolescents (>12 years of age), death is a reality and is seen as universal and irreversible
9. The following are true regarding cultural beliefs and values about death and dying EXCEPT
   A. some ethnic groups expect the clinical team to speak with the head of the family outside the patient's presence
   B. most families believe, dying at home can bring the family bad luck
   C. some families believe that the patient's spirit will become lost if the death occurs in the hospital
   D. families differ in the idea of organ donation and the acceptance of autopsy
   E. decisions, rituals, and withholding of palliative or lifesaving procedures that could harm the child should be addressed
SECTION 1 The profession of Pediatrics
ANSWERS
ZUHAIR ALMUSAWI

1. (C). Type 2 diabetes mellitus.
2. (A).
3. (B).
4. (E). Self-awareness/knowledge of limits includes recognition of the need for guidance and supervision when faced with new or complex responsibilities. The pediatrician also must be insightful regarding the impact of his or her behavior on others and cognizant of appropriate professional boundaries.
5. (D).
6. (A).
7. (C). Complicated grief, a pathologic manifestation of continued and disabling grief, is rare.
8. (D). In children age 6 to 11 years of age, the finality of death gradually comes to be understood.
9. (B). For some families, dying at home can bring the family bad luck; whereas others believe that the patient's spirit will become lost if the death occurs in the hospital.
1. Infants born small for gestational age, or prematurely, usually exhibit catch-up growth in the
   A. first 6 months
   B. second 6 months
   C. first birthday
   D. second year of life
   E. third year of life

2. The daily weight gain in the first 3–4 months of life is
   A. 10–20 gm
   B. 20–30 gm
   C. 30–40 gm
   D. 40–50 gm
   E. 50–60 gm

3. Regarding the language skills, a 2-year-old child can
   A. count
   B. name colors
   C. define words
   D. name pictures
   E. know body parts

4. The average length of an infant at 1 year of age is
   A. 10 inches
   B. 20 inches
   C. 30 inches
   D. 40 inches
   E. 50 inches

5. Head circumference (HC) increase in the first 3 months of life by
   A. 0.5 cm per month
   B. 1 cm per month
   C. 1.5 cm per month
   D. 2 cm per month
6. The child who walks backward, scribbles, and uses spoon and fork has achieved the developmental age of
   A. 9 mo
   B. 12 mo
   C. 15 mo
   D. 18 mo
   E. 21 mo

7. The child who can hop on one foot, copies circles, and brushes teeth without help has achieved the developmental age of
   A. 2 yr
   B. 3 yr
   C. 4 yr
   D. 5 yr
   E. 6 yr

8. Regarding speech production, the routine use of sentence sequences; conversational give and-take is usually achieved by age of
   A. 3 yr
   B. 4 yr
   C. 5 yr
   D. 6 yr
   E. 7 yr

9. The clue to need a help for a child who does not respond to his or her name is by age of
   A. 6 months
   B. 8 months
   C. 10 months
   D. 12 months
   E. 14 months

10. The clue to need a help for a child who does not respond correctly to “Give me” or “Sit down” or “Come here” when spoken without gestural cues is by age of
    A. 15 months
    B. 18 months
    C. 21 months
D. 24 months
E. 28 months

11. Regarding the gross motor skills, a 2-year-old child can
   A. heel-to-toe walk
   B. balance on each foot 6 sec
   C. walk up and down stairs
   D. balance well on each foot
   E. walk steps alternating feet

12. The clue to need help by age of 24 months, if the child
   A. speech is not understood by familiar listeners
   B. speech is not understood by unfamiliar listeners
   C. has not begun to ask questions (using where, what, why)
   D. is not using short sentences (e.g., “Daddy went bye-bye”)
   E. does not show understanding of spatial concepts: on, in, under, front, and back

13. The child begins to understand right and left by age of
   A. 3 yr
   B. 4 yr
   C. 5 yr
   D. 6 yr
   C. 7 yr

14. The difficulty in putting words together, limited vocabulary, or inability to use language in a socially appropriate way is defined as a disorder of
   A. fluency
   B. reception
   C. expression
   D. resonance
   E. articulation

15. The **MOST** common type of cerebral palsy is
   A. ataxic cerebral palsy
   B. spastic cerebral palsy
   C. dystonic cerebral palsy
   D. dyskinetic cerebral palsy
   E. choreoathetotic cerebral palsy
16. The age by which the child can pull to stand, starting to pincer grasp, and plays pat-a-cake is
   A. 6 mo
   B. 7 mo
   C. 8 mo
   D. 9 mo
   E. 10 mo

17. The American Academy of Pediatrics recommends the use of validated standardized developmental screening tools at three of the health maintenance visits, these are at
   A. 2 months, 6 months, and 12 months
   B. 6 months, 12 months, and 18 months
   C. 9 months, 18 months, and 30 months
   D. 12 months, 18 months, and 24 months
   E. 12 months, 24 months, and 36 months

18. The Denver II assesses the development of children from birth to 6 years of age, it includes the following domains EXCEPT
   A. language
   B. intelligence
   C. gross motor
   D. personal-social
   E. fine motor–adaptive

19. In infants, inferences about vision may be made by physical examination of the eye and assessment of
   A. language
   B. intelligence
   C. gross motor
   D. personal-social
   E. fine motor–adaptive

20. Autism screening is recommended for all children at age of
   A. 12 to 18 months
   B. 18 to 24 months
   C. 24 to 36 months
   D. 36 to 48 months
   E. 48 to 60 months
21. Because of the significant association of lead intoxication with poverty, the Centers for Disease Control and Prevention (CDC) recommends blood lead screening at
   A. 6 and 12 months
   B. 12 and 24 months
   C. 24 and 36 months
   D. 36 and 48 months
   E. 48 and 60 months

22. Low birth weight or premature infants are screened for anemia at birth and again at the age of
   A. 2 months
   B. 4 months
   C. 6 months
   D. 8 months
   E. 10 months

23. Centers for Disease Control and Prevention (CDC) recommend the blood lead level that may cause learning problems is
   A. 1 to 5 μg/dL
   B. 5 to 10 μg/dL
   C. 10 to 15 μg/dL
   D. 15 to 20 μg/dL
   E. 25 to 30 μg/dL

24. The American Academy of Pediatrics (AAP) recommends the fasting total cholesterol level is elevated when serum cholesterol is
   A. ≥150 mg/dL
   B. ≥175 mg/dL
   C. ≥200 mg/dL
   D. ≥225 mg/dL
   E. ≥250 mg/dL

25. All the following are cholesterol risk screening recommendations EXCEPT
   A. family history of obesity
   B. personal history of diabetes
   C. family history of heart disease
   D. family history of high cholesterol
   E. personal history of high blood pressure
26. The **BEST** car safety issues for a 3-year-old child is
   A. vehicle seat belt
   B. rear-facing safety seat
   C. forward-facing car seat
   D. lap and shoulder seat belts
   E. belt-positioning booster seat

27. The body mass index (screening tool for children and adolescents to identify those overweight or at risk for being overweight) is defined as
   A. body weight in pounds divided by height in inches
   B. body weight in kilograms divided by height in meters squared
   C. body weight in grams divided by height in centimeters squared
   D. body weight in kilograms multiplied by height in meters squared
   E. body weight in grams multiplied by height in centimeters squared

28. Primary amenorrhea should be considered for any female adolescent who has not reached menarche by
   A. 13 years or has not done so within 3 years of thelarche
   B. 14 years or has not done so within 2 years of thelarche
   C. 14 years or has not done so within 3 years of thelarche
   D. 15 years or has not done so within 2 years of thelarche
   E. 15 years or has not done so within 3 years of thelarche

29. The child who can transfer objects from hand to hand and babbles has achieved the developmental age of
   A. 4 months
   B. 6 months
   C. 8 months
   D. 10 months
   E. 12 months

30. Older school-age children who begin to participate in competitive sports should have a careful evaluation of the
   A. vision
   B. hearing
   C. sexual maturity
   D. cardiovascular system
   E. orthopedic deformities
31. Stranger anxiety (infants normally become insecure about separation from the primary caregiver) develops in age between
   A. 6 and 9 months
   B. 9 and 18 months
   C. 18 and 24 months
   D. 24 and 36 months
   E. 36 and 48 months
1. (A). Infants born small for gestational age, or prematurely, ingest more breast milk or formula and, unless there are complications that require extra calories, usually exhibit catch-up growth in the first 6 months. These infants should be fed on demand and provided as much as they want unless they are vomiting (not just spitting up).

2. (B).
   - Weight loss in first few days: 5%–10% of birth weight
   - Return to birth weight: 7–10 days of age
   - Double birth weight: 4–5 months
   - Triple birth weight: 1 year
   - Daily weight gain:
     * 20–30 g for first 3–4 month
     * 15–20 g for rest of the first year

3. (E).

4. (C).
   - Average length: 20 in. at birth, 30 in. at 1 year
   - At age 4 years, the average child is double birth length or 40 in.

5. (D). Average HC: 35 cm at birth (13.5 in.). HC increases: 1 cm per month for first year (2 cm per month for first 3 months, then slower).

6. (C). Developmental milestones of a 15-month-old child are walks backward, scribbles, stacks two blocks, uses spoon and fork, helps in housework, says three to six words, and follows commands.

7. (C). Developmental milestones of a 4-year-old child are balances well on each foot, hops on one foot, copies circles, draws person with three parts, brushes teeth without help, dresses without help, names colors, and understands adjectives.

8. (B).
   - 1 yr -------- one to three words
   - 2 yr -------- two- to three-word phrases
   - 3 yr -------- routine uses of sentences
   - 4 yr -------- routine use of sentence sequences; conversational give and-take
   - 5 yr -------- complex sentences; extensive use of modifiers, pronouns, and prepositions
9. (C). Also clues to when a child with a communication disorder needs help, at 10 months, the child’s sound-making is limited to shrieks, grunts, or sustained vowel production.

10. (C). Clues to When a Child with a communication disorder needs help according to age are:
   - At 12 months, the child’s babbling or speech is limited to vowel sounds.
   - By 15 months, the child does not respond to “no,” “bye-bye,” or “bottle.”
   - By 15 months, the child does not imitate sounds or words.
   - By 18 months, the child is not consistently using at least six words with appropriate meaning.
   - By 21 months, the child does not respond correctly to “Give me . . . ,” “Sit down,” or “Come here” when spoken without gestural cues.
   - By 23 months, two-word phrases that are spoken as single units (e.g., “whatszit,” “thankyou,” “allgone”) have not emerged.

11. (C).
   - walk up and down stairs ---- 2 years
   - walk steps alternating feet ---- 3 years
   - balance well on each foot ---- 4 years
   - heel-to-toe walks ---- 5 years
   - balance on each foot 6 sec ---- 6 years

12. (A).

13. (D). Developmental milestones of a 6-year-old child are balances on each foot 6 sec, copies triangle, draws person with six parts, defines words, and begins to understand right and left.

14. (C).
   - Speech disorders include articulation, fluency, and resonance disorders.
     * Articulation disorders include difficulties producing sounds in syllables or saying words incorrectly to the point that other people cannot understand what is being said.
     * Fluency disorders include problems such as stuttering, the condition in which the flow of speech is interrupted by abnormal stoppages, repetitions (st-st-stuttering), or prolonging sounds and syllables (ssssstuttering).
     * Resonance or voice disorders include problems with the pitch, volume, or quality of a child’s voice that distract listeners from what is being said.
   - Language disorders can be either receptive or expressive.
     * Receptive disorders refer to difficulties understanding or processing language.
     * Expressive disorders include difficulty putting words together, limited vocabulary, or inability to use language in a socially appropriate way
15. (B).

- Spastic cerebral palsy: the most common form of cerebral palsy, it accounts for 70%–80% of cases. It results from injury to the upper motor neurons of the pyramidal tract. It may occasionally be bilateral. It is characterized by at least two of the following: abnormal movement pattern, increased tone, or pathologic reflexes (e.g., Babinski response, hyperreflexia).
- Dyskinetic cerebral palsy: occurs in 10%–15% of cases. It is dominated by abnormal patterns of movement and involuntary, uncontrolled, recurring movements.
- Ataxic cerebral palsy: accounts for <5% of cases. This form results from cerebellar injury and features abnormal posture or movement and loss of orderly muscle coordination or both.
- Dystonic cerebral palsy: also uncommon. It is characterized by reduced activity and stiff movement (hypokinesia) and hypotonia.
- Choreoathetotic cerebral palsy: rare now that excessive hyperbilirubinemia is aggressively prevented and treated. This form is dominated by increased and stormy movements (hyperkinesia) and hypotonia.

16. (D). Developmental milestones of a 9-month-old infant are pulls to stand, gets into sitting position, starting to pincer grasp, bangs two blocks together, waves bye-bye, plays pat-a-cake, says Dada and Mama, but nonspecific, and two-syllable sounds.

17. (C).

18. (B).

19. (C). Inferences about vision may be made by examining gross motor milestones (children with vision problems may have a delay) and by physical examination of the eye. Inferences about hearing are drawn from asking parents about responses to sound and speech and by examining speech and language development closely.

20. (B). Although there are several tools, many pediatricians use the Modified Checklist for Autism in Toddlers (M-CHAT).

21. (B).

22. (B). Children are screened for anemia at ages when there is a higher incidence of iron deficiency anemia. Infants are screened at birth and again at 4 months if there is a documented risk, such as low birth weight or prematurity. Healthy term infants usually are screened at 12 months of age because this is when a high incidence of iron deficiency is noted.

23. (B). Centers for Disease Control and Prevention (CDC) recommends environmental investigation at blood lead levels of 20 μg/dL on a single visit or
persistent 15 μg/dL over a 3-month period, levels of 5 to 10 μg/dL may cause learning problems.

24.(C). The American Academy of Pediatrics (AAP) recommends dyslipidemia screening in the context of regular health care for at-risk populations by obtaining a fasting lipid profile. The recommended screening levels are the same for all children 2 to 18 years. Total cholesterol of less than 170 mg/dL is normal, 170 to 199 mg/dL is borderline, and greater than 200 mg/dL is elevated.

25.(A).

Cholesterol Risk Screening Recommendations;
Risk screening at ages 2, 4, 6, 8, 10 and annually in adolescence:
1. Children and adolescents who have a family history of high cholesterol or heart disease
2. Children whose family history is unknown
3. Children who have other personal risk factors: obesity, high blood pressure, or diabetes

Universal screening at ages 9–11 and ages 18–20

26.(C). Toddlers and preschoolers over age 2 or who have outgrown the rear-facing car seat should use a forward-facing car seat with harness for as long as possible, up to the highest weight or height recommended by the manufacturer.

27.(B).

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

29.(B). Developmental milestones of a 6-month-old infant are sits alone, transfers object hand to hand, feeds self, holds bottle, and babbles.

30.(D). Older school-age children who begin to participate in competitive sports should have a comprehensive sports history and physical examination, including a careful evaluation of the cardiovascular system. The American Academy of Pediatrics 4th edition sports preparticipation form is excellent for documenting cardiovascular and other risks.

31.(B).
1. Most infants cry little during the first 2 weeks of life, gradually increasing to 3 hours per day by
   A. 2 weeks
   B. 4 weeks
   C. 6 weeks
   D. 8 weeks
   E. 12 weeks

2. A 2-month-old infant presented with daily paroxysms of continuous crying for 3-5 hours starting after 6-8 pm associated with facial grimacing, leg flexion, and passing flatus, on examination no abnormality was detected apart from mild abdominal distension.
   Of the following, the MOST likely cause is
   A. hunger
   B. wet diaper
   C. fussiness
   D. colic
   E. overfeeding

3. After full evaluation of an infant with excessive crying, an organic etiology was found in less than
   A. 5% of infants
   B. 10% of infants
   C. 15% of infants
   D. 20% of infants
   E. 25% of infants

4. One of the following is a dangerous soothing technique used to calm a crying infant
   A. swaddling
   B. white noise
   C. gentle vibration
   D. slow rhythmic rocking
   E. placing the infant on a vibrating clothes dryer
5. One of the following may be effective in calming a crying infant with colic
   A. diphenhydramine
   B. phenobarbital
   C. simethicone
   D. lactase
   E. singing

6. Regarding swaddling, one of the following is correct
   A. swaddling is effective if practiced during a crying episode
   B. swaddling is effective if practiced before a crying episode
   C. there is no place for swaddling to calm a crying infant
   D. swaddling may interfere with vascular supply
   E. swaddling exposes the infant to rickets

7. A 3-year-old child has recurrent attacks of screaming, vomiting, and biting other children, these attacks last 2-5 minutes and occurs once or twice weekly, the child looks well between the attacks.
   Of the following, the **MOST** likely cause is
   A. autism
   B. traumatic brain injury
   C. cognitive impairment
   D. temper tantrums
   E. ketotic hypoglycemia

8. A 30-month-old apparently well child presented with stomping, hitting, head banging, and falling down for few minutes every few days.
   Of the following, the **MOST** important investigation to be done is
   A. brain MRI
   B. serum ferritin
   C. skeletal survey
   D. EEG
   E. serum electrolytes

9. Diagnosis of attention deficit/hyperactivity disorder (ADHD) in children up to the age of 16 years requires the presence of at least
   A. 4 symptoms of inattention or 4 symptoms of hyperactivity-impulsivity for at least 4 months in two or more environments
   B. 5 symptoms of inattention or 5 symptoms of hyperactivity-impulsivity for at least 5 months in two or more environments
C. 6 symptoms of inattention or 6 symptoms of hyperactivity-impulsivity for at least 6 months in two or more environments
D. 6 symptoms of inattention or 6 symptoms of hyperactivity-impulsivity for at least 3 months
E. 6 symptoms of inattention or 6 symptoms of hyperactivity-impulsivity for at least 6 months

10. One of the following is not a feature of attention deficit/hyperactivity disorder (ADHD)
   A. ADHD is multifactorial in origin
   B. Positron emission tomography have shown structural and functional differences
   C. prenatal exposure to nicotine increases the risk of ADHD.
   D. female to male ratio is 2 to 6:1
   E. girls often present with inattentive symptoms

11. All the following medical conditions should be considered as possible causes for a child's hyperactivity and distractibility EXCEPT
   A. sleep disordered breathing
   B. seizure disorders
   C. substance use
   D. hypothyroidism
   E. lead intoxication

12. An 8-year-old male child presented with failing to pay close attention to details, appearing to not listen when spoken to directly, and failing to follow through on instructions or finish assigned work, he also appears restless, leaving a seat when expected to remain seated, and running excessively in inappropriate situations. Of the following, the first-line agent for treatment of this child is
   A. atomoxetine
   B. guanfacine
   C. clonidine
   D. daizepam
   E. methylphenidate

13. Common side effects of stimulant medications include
   A. appetite suppression
   B. gastrointestinal tract symptoms
   C. sedation
14. The entire process of toilet training need not be hurried and it can take
   A. 2 months
   B. 4 months
   C. 6 months
   D. 8 months
   E. 10 months

15. For most children with enuresis, the only test recommended is
   A. blood sugar
   B. serum electrolytes
   C. urinalysis
   D. abdominal sonography
   E. bladder function tests

16. Children with secondary nocturnal enuresis may have
   A. UTI
   B. diabetes mellitus
   C. diabetes insipidus
   D. hypercalcemia
   E. psychosocial stressor

17. Pharmacotherapy for nighttime enuresis includes desmopressin acetate and, rarely, tricyclic antidepressants.
   All the following are true regarding desmopressin EXCEPT
   A. It decreases urine production
   B. It is proved to be safe
   C. It is started at 0.2 mg per dose
   D. It is not curative
   E. It has a relapse rate of 40%

18. Encopresis is the regular, voluntary or involuntary passage of feces into a place other than the toilet after the age of
   A. 2 years
   B. 3 years
   C. 4 years
   D. 5 years
19. A 5-year-old child presented with chronic constipation and soiling, the parents give a history of delayed passage of meconium. On examination, he has an empty rectum and tight sphincter.

Of the following, the MOST likely cause of constipation is

A. hypothyroidism
B. Hirschsprung disease
C. functional constipation
D. celiac disease
E. drugs abuse

20. All the following are features of rapid eye movement (REM) EXCEPT

A. REM sleep is characterized by active, awake-like electroencephalography (EEG)
B. Neonates typically begin their sleep cycle in REM sleep
C. Muscle twitches and facial grimaces are common in REM sleep of neonates
D. REM sleep comprises up to 25% of total sleep time in newborns
E. The amount of REM sleep shifts toward the last third of the night as the child grows

21. A polysomnogram consists of an all-night observation and recording performed in a sleep laboratory, it is used to detect

A. excessive limb movements
B. primary insomnia
C. circadian rhythm disorders
D. uncomplicated parasomnias
E. behaviorally based sleep problems

22. An 8-year-old obese hyperactive child presented with history of snoring and excessive daytime sleepiness and poor school performance.

Of the following, the MOST important diagnostic test is

A. polysomnography
B. lateral x-ray of head and neck
C. fibro-optic nasopharyngeal examination
D. CT scan of head and neck
E. MRI of head and neck
23. Rarely children with insomnia are treated pharmacologically. Of the following, the **MOST** successfully used drug is
   A. chlorpheniramine
   B. diazepam
   C. phenobarbitone
   D. melatonin
   E. lorazepam
1. (C). Most infants cry little during the first 2 weeks of life, gradually increasing to 3 hours per day by 6 weeks and decreasing to 1 hour per day by 12 weeks.

2. (D). Colic often is diagnosed using Wessel’s rule of threes—crying for more than 3 hours per day, at least 3 days per week, for more than 3 weeks. Colicky crying is often described as paroxysmal and may be characterized by facial grimacing, leg flexion, and passing flatus.

3. (A). Fewer than 5% of infants evaluated for excessive crying have an organic etiology. Because the etiology of colic is unknown, this syndrome may represent the extreme of the normal phenomenon of infant crying. Nonetheless evaluation of infants with excessive crying is warranted.

4. (E). Avoidance of dangerous soothing techniques, such as shaking the infant or placing the infant on a vibrating clothes dryer (which has resulted in injury from falls), should be stressed.

5. (E). Medications, including phenobarbital, diphenhydramine, alcohol, simethicone, dicyclomine, and lactase, are of no benefit in reducing colic and should be avoided.

6. (B). Infants who have been tightly swaddled for sleep and rest during the first weeks of life often calm to swaddling during a crying episode; this is not true for infants who have not experienced swaddling before a crying episode.

7. (D). A temper tantrum, defined as out-of-control behavior, including screaming, stomping, hitting, head banging, falling down, and other violent displays of frustration, can include breath-holding, vomiting, and serious aggression, including biting. Tantrums are seen most often when the young child experiences frustration, anger, or simple inability to cope with a situation. Temper tantrums can be considered normal behavior in 1- to 3-year-old children, when the temper tantrum period is of short duration and the tantrums are not manipulative in nature.

8. (B). The most likely diagnosis is temper tantrum in which laboratory studies screening for iron deficiency anemia and lead exposure are important. Other laboratory and imaging studies are performed only when the history and physical examination suggest a possible underlying etiology. Some children with excessive tantrums should have a formal developmental evaluation.

9. (C). Diagnosis of children up to the age of 16 years requires the presence of at least 6 symptoms of inattention or 6 symptoms of hyperactivity-impulsivity for at least 6 months in two or more environments. Children 17 years of age and older
must exhibit at least 5 symptoms of inattention or at least 5 symptoms of hyperactivity-impulsivity.

10. (D). The male to female ratio is 2 to 6:1, with greater male predominance for the hyperactive/impulsive and combined types. Girls often present with inattentive symptoms and are more likely to be underdiagnosed or to receive later diagnoses.

11. (D). Hyperthyroidism.

12. (E). Stimulant medications (methylphenidate or amphetamine compounds) are the first-line agents for treatment of ADHD due to extensive evidence of effectiveness and safety.

13. (A). Common side effects include appetite suppression and sleep disturbance with stimulant medications, gastrointestinal tract symptoms with atomoxetine, and sedation with alpha agonists. These side effects usually can be managed by careful adjustment of medication dosage and timing. Screening (by history and exam) for cardiac disease and monitoring of cardiac status is prudent given concerns raised by a retrospective study suggesting extremely rare but slightly increased odds of sudden cardiac death in those taking stimulant medication.

14. (C). Toilet training usually begins after the second birthday and is achieved at about 3 years of age in middle-class white U.S. populations. Toilet training between 12 and 18 months of age continues to be accepted in lower-income families.

15. (E). For most children with enuresis, the only laboratory test recommended is a clean catch urinalysis to look for chronic urinary tract infection (UTI), renal disease, and diabetes mellitus.

16. (E).

17. (E). This treatment must be considered symptomatic, not curative, and has a relapse rate of 90% when the medication is discontinued.

18. (C).

19. (B).

20. (D). REM sleep comprises up to 50% of total sleep time in newborns and gradually decreases to 25% to 30% by adolescence.

21. (A). A polysomnogram is used to detect Obstructive sleep apnea (OSA), excessive limb movements, and seizure disorder. This consists of an all-night observation and recording performed in a sleep laboratory. Polysomnography is not indicated in children with primary insomnia (difficulty initiating or maintaining sleep), circadian rhythm disorders, uncomplicated parasomnias, or behaviorally based sleep problems.

22. (A). Obstructive sleep apnea (OSA) is usually detected by polysomnography.

23. (D). Melatonin (dose, 2.5 to 10 mg) has soporific properties useful in treating delayed sleep phase syndrome. It has been used successfully in both children with
normal development and those with developmental delays. Melatonin is available without prescription in stores that sell dietary supplements.
1. The somatoform disorders are groups of disorders in which physical symptoms are inconsistent and cannot be explained by a medical condition. Of the following, the **MOST** vulnerable group for these disorders is
   A. infants of both sex
   B. male toddlers
   C. young children of female gender
   D. adolescent girls
   E. adolescent boys

2. A 12-year-old boy has unexplained recurrent attacks of severe abdominal pain over the past 2 years, not resolving during holidays; physical examination and lab investigations are unremarkable. Of the following, the risk factor that is **LEAST** likely associated with this disorder is
   A. emotional distress
   B. history of marital conflict
   C. child maltreatment whether emotional, sexual, or physical abuse
   D. history of physical illness
   E. school attendance

3. Symptoms of somatoform disorders are variable during childhood. Of the following, the symptom that is often present during early childhood is
   A. recurrent abdominal pain
   B. headaches
   C. neurologic symptoms
   D. insomnia
   E. fatigue

4. You are evaluating a 5-year-old girl who has multiple unexplained physical complaints. Which of the following symptoms is **LEAST** likely to occur in this age as criteria for this disorder?
   A. headache, abdominal pain, back pain, and chest pain
   B. nausea, vomiting, and diarrhea
   C. sexual indifference
   D. double vision
E. difficulty swallowing or lump in throat

5. Nonepileptic seizures are a subtype of conversion disorder that resembles true epileptic seizures. Of the following, the **MOST** characteristic criteria of these seizures is
   A. there is no electroencephalographic abnormalities
   B. most cases have a protracted course
   C. the course of the disease is refractory
   D. there is a high incidence of recurrence
   E. antiepileptic medication is the cornerstone of treatment

6. Although the course of the nonepileptic seizures is often benign, there are some prognostic factors that may influence the outcome. Of the following, the characteristic feature that carry poor prognosis is
   A. symptoms of paralysis and blindness
   B. presence of tremor
   C. acute onset
   D. above-average intelligence
   E. presence of an identifiable stressor

7. Pain disorder is a somatoform disorder characterized by pain as the predominant physical symptom. Of the following, the **MOST** frequently encountered pain for this disorder is
   A. recurrent chest pain
   B. recurrent abdominal pain
   C. musculoskeletal pain
   D. headache
   E. complex regional pain syndrome type I

8. A 16-year-old girl complains of recurrent attacks of headache over the past 6 months; she has a fear of having brain tumor in spite of normal clinical and radiological findings; she has an underlying depression disorder. Of the following, the **MOST** likely diagnosis is
   A. undifferentiated somatoform disorder
   B. conversion disorder
   C. pain disorder
   D. body dysmorphic disorder
   E. hypochondriasis
9. Suicidal thoughts may be associated with some somatoform disorders. Which of the following disorder has a higher rate of suicidal ideation and attempts
   A. undifferentiated somatoform disorder
   B. conversion disorder
   C. pain disorder
   D. body dysmorphic disorder
   E. hypochondriasis

10. Chronic fatigue syndrome (CFS) is usually associated with comorbid psychiatric disorders.
   Of the following, the psychiatric disorder that is often associated with CFS is
   A. anxiety
   B. depression
   C. obsessive compulsive disorder (OCD)
   D. social phobia
   E. delusion

11. In chronic fatigue syndrome (CFS) with comorbid depression and anxiety, antidepressants can be useful.
   Of the following, the MOST useful antidepressant drug in CFS is
   A. fluoxetine
   B. sertraline
   C. citalopram
   D. clomipramine
   E. bupropion

12. Munchausen syndrome by proxy (MBP) is a form of factitious disorder by proxy, where a parent (usually mother) mimics symptoms in his or her child.
   Of the following, the symptom that is LEAST likely presented is
   A. diarrhea
   B. respiratory arrest
   C. seizures
   D. failure to thrive
   E. psychosis

13. Panic disorder is usually associated with some psychiatric comorbidities; however, there is a high incidence of panic attacks with some medical conditions.
   Of the following, the medical condition that is associated with a high incidence of panic attacks is
A. asthma
B. rheumatoid arthritis
C. common variable immune deficiency
D. sickle cell anemia
E. inflammatory bowel disease

14. Generalized anxiety disorder (GAD) is characterized by 6 or more months of persistent, out of proportion worry and anxiety. Of the following, children have less prominent anxiety symptoms in
A. school performance
B. competitive sports
C. non-competitive sports
D. celebration activity
E. autonomic arousal

15. A 5-year-old boy is brought by her mother complaining of frequent attacks of anger, sadness, and exaggerated startle response over the past 3 weeks; the mother states he has prolonged periods of sitting alone as he doesn’t like to be with the others; he has been developed those symptoms immediately after experiencing a bad car accident with his father before 3 weeks. Of the following, the MOST likely diagnosis is
A. acute posttraumatic stress disorder (PTSD)
B. chronic PTSD
C. delayed onset PTSD
D. acute stress disorder
E. separation anxiety disorder

16. Posttraumatic stress disorder (PTSD) is characterized by re-experiencing a traumatic event that threatened live. It may occur both in children and adults. Of the following, the symptom that is MORE likely seen in children than in adults is
A. difficulty falling or staying asleep
B. outbursts of anger
C. estrangement from others
D. exaggerated startle response
E. flashbacks of daydream quality

17. A 7-year-old child is having multiple complains of abdominal pain, headache, and fatigue; examination and investigations are normal. The mother states that her
son is so attached to her and is experiencing these symptoms when she decided to join him to primary school.

Of the following, the MOST likely diagnosis is

A. school phobia
B. social phobia
C. true phobia related to schoolwork
D. situational type specific phobia
E. separation anxiety disorder

18. You are meeting a 6-year-old girl who is going to join a primary school. The parents states that their child has some antisocial problems and they have a concern regarding school phobia and attendance.

Of the following, the statement about school phobia that should be included in your discussion is

A. affected children usually do not have antisocial tendencies
B. girls are more likely affected than boys
C. there is strong association with social class, intelligence, or academic ability.
D. the oldest member of a family of few children is more likely to be affected
E. truancy is highly associated with young children with higher levels of fear

19. Major depressive disorder (MDD) symptoms include either depressed mood or loss of interest in nearly all activities; it’s severity is variable.

Of the following, the LEAST likely symptoms that are seen in mild cases of MDD is

A. irritability, restlessness, or boredom
B. sudden drop in grades
C. change in appetite with or without weight changes
D. sleep disturbance with somatic complaints
E. Psychotic symptoms which are mood-congruent

20. Family history is the strongest single factor for developing major depressive disorder (MDD) and comorbid disorders are frequently encountered.

Of the following, the MOST likely disorder that is associated with MDD is

A. dysthymic disorder
B. anxiety disorders
C. substance abuse
D. disruptive behavior disorders
E. bipolar disorder
21. Major Depressive disorder (MDD) is being increasingly seen in offspring of depressed parents; it has many forms that may be seen in the same family. Of the following, the **MOST** common depressive disorder in children and adolescent is
   A. dysthymic disorder
   B. atypical depression
   C. adjustment disorder with depressed mood
   D. seasonal affective disorder
   E. depressive disorder not otherwise specified

22. Atypical depression is a form of major depressive disorder (MDD) characterized by all the following **EXCEPT**
   A. insomnia
   B. increased appetite
   C. weight gain
   D. interpersonal rejection sensitivity
   E. mood reactivity

23. A 9-year-old boy has periods of mania alternating with depression.
   Of the following, the hallmark of mania associated with this disorder is
   A. excessive talking
   B. excessive giggling
   C. racing thoughts
   D. decreased need for sleep
   E. grandiosity

24. Obsessive-compulsive disorder (OCD) may be inherited. Other factors such as infection may account for small percentage of childhood-onset OCD. Which of the following infections may account for childhood-onset OCD?
   A. staphylococcus aureus infection
   B. staphylococcus epidermidis infection
   C. group A beta-hemolytic streptococcal infection
   D. chlamydia trachomatis infection
   E. klebsiella pneumoniae infection

25. The combination of medications and cognitive-behavioral therapy (CBT) has shown the best treatment for patients with obsessive-compulsive disorder (OCD). Of the following, the **BEST** initial treatment for children with OCD is
   A. fluoxetine
B. clomipramine
C. risperidone
D. anterior capsulotomy
E. limbic leucotomy

26. Pervasive developmental disorders, also known as autism spectrum disorders (ASDs), consist of five disorders. The hallmark of these disorders is
A. onset is in infancy and preschool years
B. impaired communication and social interaction
C. mental retardation
D. aggression
E. sleep dysregulation

27. Autism spectrum disorders (ASDs) are seen with equal prevalence among all racial and ethnic groups. Of the following, the disorder that is only prevalent in girls is
A. autism
B. Asperger syndrome
C. childhood disintegrative disorder
D. Rett syndrome
E. pervasive developmental disorder not otherwise

28. Stimulant drugs are useful for hyperactivity and inattention associated with autism spectrum disorders (ASDs). Of the following, the BEST response of these drugs is seen with
A. autism
B. Asperger syndrome
C. childhood disintegrative disorder
D. Rett syndrome
E. pervasive developmental disorder not otherwise

29. Sleep dysregulation is commonly seen in autism spectrum disorders (ASDs) that can be treated by some medications. Of the following, the first medication for sleep dysregulation is
A. melatonin
B. guanfacine
C. clonidine
D. haloperidol
E. risperidone
30. You are meeting parents of a 3-year-old boy who has normal developmental milestone and social interaction; they have older child with autism; they have a concern about having this young kid with autism as well. You state that most children with autism have clinical manifestations by the age of
   A. second or third year of life
   B. preschool
   C. school
   D. adolescence
   E. no age predilection

31. Comorbidities are commonly encountered in children with autism, especially other psychiatric disorders. Of the following, the MOST commonly associated disease with autism is
   A. mental retardation
   B. seizure disorder
   C. anxiety disorders
   D. obsessive compulsive disorder
   E. attention-deficit/hyperactivity disorder

32. The earliest studies of autism suggested a relatively poor prognosis, with only a small number of individuals being able to function independently as adults. Of the following, the factor that carry POOR outcome is
   A. early onset
   B. higher IQ
   C. better language skills
   D. good communication
   E. average nonverbal cognitive skills

33. Schizophrenia generally presents in adolescence or early adulthood. However, it may appear in children. Which of the following is true regarding childhood-onset schizophrenia?
   A. it usually indicates a mild form of schizophrenia
   B. it is more prevalent in girls
   C. hallucinations are mainly tactile misperceptions
   D. negative symptoms are most frequent in early childhood and later adolescence
   E. children with high IQs often have more negative and fewer positive symptoms
34. The paranoid type of schizophrenia shows prominent hallucinations and delusions with relatively normal cognition. Of the following, the **MOST** commonly encountered type of delusions is
   A. erotomanic
   B. grandiose
   C. jealous
   D. somatic
   E. persecutory

35. A 13-year-old boy has auditory hallucinations and delusions with lack of social interactions that lasts for 3 weeks. As these symptoms persist for less than 6 months and do not meet the criteria of schizophrenia. Of the following, the **MOST** likely diagnosis is
   A. brief psychotic disorder
   B. schizophreniform disorder
   C. psychotic disorder not otherwise specified
   D. Schizoaffective disorder
   E. major depression with psychotic features

36. Treatment of schizophrenia is based on a multimodal approach, including use of antipsychotic medications. Of the following, the drug that is used as a 1st line medication in schizophrenia treatment is
   A. haloperidol
   B. chlorpromazine
   C. lithium
   D. risperidone
   E. clozapine

37. The course of illness for schizophrenia is characterized by exacerbations and remissions of psychotic symptoms. The outcome is variable depending on some prognostic factors. Of the following, the factor that carries a good prognosis is
   A. age younger than 13 years
   B. poor premorbid function
   C. presence of visual or auditory hallucinations
   D. presence of marked negative symptoms
   E. positive family history of schizophrenia
1. (D). Adolescent girls tend to report nearly twice as many functional somatic symptoms as adolescent boys, whereas prior to puberty the ratio is equal.

2. (E). Somatic disorders usually resolve when eliminating the provoking factor. Symptoms due to academic problems usually resolve during holidays and summer season, which is not the case in this scenario.

3. (A). In early childhood, symptoms often include recurrent abdominal pain. Later headaches, neurologic symptoms, insomnia, and fatigue are more common.

4. (C). Sexual or reproductive symptom is commonly encountered during adolescence, so the diagnosis of somatization disorder is unusual in children.

5. (A). Most cases resolve within 3 months of diagnosis. Referring to non-classic presentations for seizures as spells can help avoid medicalization of these symptoms. The course of the condition is often benign. Only 20% to 25% of patients experience a recurrence.

6. (B). Poor prognostic characteristics include tremor and pseudo seizures. Symptoms of paralysis, aphony, blindness; acute onset; above-average intelligence; presence of an identifiable stressor; and early diagnosis and psychiatric treatment are good prognostic features.

7. (B). Recurrent abdominal pain is the most common recurrent pain complaint of childhood and accounts for 2% to 4% of pediatric office visits.

8. (E). Hypochondriasis is the preoccupation with the fear of having a serious disease based on misinterpretation of bodily symptoms and functions. Body dysmorphic disorder is a preoccupation with an imagined or slight defect in physical appearance that causes clinically significant distress or impairment in functioning.

9. (D). Body dysmorphic disorder (BDD) is also associated with high rates of suicidal ideation and attempts, with 28% of sufferers having attempted suicide.

10. (B). CFS refers to a condition of severe, disabling fatigue associated with self-reported limitations in concentration and short-term memory, sleep disturbance, and musculoskeletal aches and pains that lasts for at least 6 months. It is often associated with depression and can be incapacitating.

11. (E). In chronic fatigue syndrome with comorbid depression and anxiety, a more activating antidepressant, like bupropion, can be useful. Stimulants may also be helpful in CFS.
12. (E). Simulation of psychiatric disorders is rare.
13. (A). Panic disorder is a common anxiety disorder characterized by the presence of recurrent, unexpected panic attacks. Patients with asthma have a high incidence of panic attacks.
14. (E). In children and adolescents, the specific symptoms of autonomic arousal are less prominent, and symptoms are often related to event or activity.
15. (D). Acute stress disorder is characterized by the same signs and symptoms as PTSD but occurs immediately after a traumatic event. If impaired function persists after 1 month, the diagnosis is PTSD.
16. (E). Flashbacks are more daydream quality than the sudden intrusive events seen in adults and complaint of restriction of effect and numbing are less frequent.
17. (E). Separation anxiety disorder (SAD) is seen in children and adolescents who express vague somatic symptoms to avoid or refuse to go to school. Patients may have a valid or an irrational concern about a parent or have had an unpleasant experience in school. School phobia is one of a range of reasons for school nonattendance; unlike anxious school refusers, truants hide their school nonattendance from their parents.
18. (A). Boys and girls are equally affected and there is no association with social class, intelligence, or academic ability. The youngest in a family of several children is more likely to be affected as well as children of older parents. Truancy is generally associated with older adolescents with lower levels of fear. Unlike anxious school refusers, truants hide their school nonattendance from their parents.
19. (E). Psychotic symptoms, seen in severe cases of major depression, are generally mood-congruent (e.g., derogatory auditory hallucinations, guilt associated delusional thinking).
20. (B). Dysthymic disorder and anxiety disorders (prevalence 30% and 80%, respectively), substance abuse (20% to 30%), and disruptive behavior disorders (10% to 20%) are frequent comorbid disorders in depressed children and adolescents. Twenty percent of patients diagnosed with depression develop bipolar disorder (BD).
21. (C). In adjustment disorder with depressed mood, symptoms start within 3 months of an identifiable stressor (e.g., loss of a relationship), with distress in excess of what would be expected and interference with social, occupational, or school functioning.
22. (A). Hypersomnia rather than insomnia is a feature of atypical depression.
23. (D). Bipolar disorder is the most likely diagnosis. A decreased need for sleep is a hallmark of mania. There are no other diagnoses where a child has a greatly
decreased amount of total sleep (compared with age-appropriate norms) and is not fatigued.

24.(C). Streptococcal infection causing inflammation in the basal ganglia and is a part of a condition referred to as pediatric autoimmune neuropsychiatric disorders associated with streptococcal (also known as PANDAS) infection. Early antibiotic therapy may help treat these cases.

25.(A). Selective serotonin reuptake inhibitors (SSRI), like fluoxetine, are generally thought to show a favorable risk-to-benefit ratio in OCD. If an SSRI trial is unsuccessful, clomipramine can be tried next. Combination therapy using an SSRI with an antipsychotic medication (risperidone or another atypical antipsychotic) is especially useful with specific comorbidities (e.g., tic disorders). Deep brain stimulation of the basal ganglia, through surgically implanted electrodes and surgical interventions are reserved for very severe cases or highly refractory cases.

26.(B). Hallmarks of these disorders include impaired communication and impaired social interaction as well as stereotypic behaviors, interests, and activities. Mental retardation is common, with a few children showing remarkable isolated abilities (savant or splinter skills).

27.(D).

28.(B).

29.(A).

30.(A). If no clinical manifestations of the disorder are present by 3 years of age, Rett syndrome or childhood disintegrative disorder needs to be considered.

31.(A). Mental retardation is seen in up to 80% of cases.

32.(A).

33.(D). Childhood-onset schizophrenia usually indicates a more severe form of schizophrenia. Boys tend to be affected about twice as often as girls. Positive symptoms include hallucinations (auditory or visual misperceptions that occur without external stimuli) and delusions. Negative symptoms include a lack of motivation and social interactions and flat effect. Children with high IQs often show more positive and fewer negative symptoms.

34.(E). People with this type of delusion believe that they (or someone close to them) are being mistreated, or that someone is spying on them or planning to harm them.

35.(A). To meet criteria for diagnosing schizophrenia, clinical symptoms should be present for at least 6 months. If symptoms are present for less than 1 month, the condition is called a brief psychotic disorder. If symptoms are present for more than 1 month but less than 6 months, a diagnosis of schizophreniform disorder is made. Schizoaffective disorder is diagnosed when a person has clear symptoms of
schizophrenia for at least 2 weeks without active symptoms of depression or mania.

36.(D). First-line drugs are atypical antipsychotics (e.g., risperidone, olanzapine, quetiapine, aripiprazole, ziprasidone, and paliperidone). Second-line medications are typical antipsychotics (e.g., haloperidol, thiothixene, chlorpromazine, trifluoperazine, loxapine, and molindone). Antipsychotics can be augmented with lithium or another mood stabilizer. Clozapine or electroconvulsive therapy is generally reserved for resistant cases.

37.(C).
1. You are meeting parents of a 1.5-year-old boy who has suboptimal growth. He has been delivered prematurely at the gestational age of 32 weeks. Examination reveals a healthy child with growth parameters below normal for age. You reassure the parents that their child will catch his normal growth with time.

Of the following, the growth parameter that should be corrected at this age is

A. weight
B. height
C. body mass index
D. head circumference
E. arm circumference

2. Failure to thrive (FTT) is a descriptive term given to malnourished infants and young children who fail to meet expected standards of growth.

Of the following, the growth parameter that is usually not affected by malnutrition is

A. weight
B. height
C. weight for height ratio
D. head circumference
E. arm circumference

3. You are discussing failure to thrive (FTT) with medical students. You mention that FTT is most often used to describe malnutrition related to environmental or psychosocial causes. An important statement that should be included in your discussion is

A. FTT is often diagnosed by weight that falls below the 25th percentile for age
B. a weight crossing one major percentile lines on the growth height should be evaluated for FTT chart over time is considered abnormal
C. a weight of less than 60% of the median weight for the height of the child
D. small subset of the population naturally falls below the 3rd percentile but usually have normal weight for height
E. wide fluctuations in percentile position can occur in normal children during adolescence
4. An 11-month-old girl infant is found to have low weight and height consistent with FTT. She was exclusively breast feeding till the age of 4 month, and then artificial milk-formula was added. She has had a normal growth pattern till the age of 6 month when the mother introduced juices and cereals. Examination is unremarkable apart from significant decline of normal growth pattern. Of the following, the MOST likely cause is
   A. failed breast feeding
   B. improper formula preparation
   C. congenital syndromes
   D. congenital infections
   E. celiac disease

5. During starvation, the body slows metabolic processes and growth to minimize the need for nutrients. With the rapid reinstitution of feeding after starvation refeeding syndrome may occur. Of the following, the major changes that may occur with refeeding syndrome typically affect
   A. electrolytes
   B. serum proteins
   C. hormones
   D. liver enzymes
   E. WBC count

6. The physical abuse of children affects children of all ages. Serious injuries, such as head or abdominal trauma, are more likely to be inflicted by
   A. mothers
   B. fathers
   C. old brothers
   D. grand mothers
   E. caregivers

7. You are evaluating a 2-year-old boy with multiple bruises. Physical examination is unremarkable apart from multiple bruising areas. Lab investigations including coagulation profile are normal. Of the following, bruises that are LEAST likely suggestive of physical abuse is
   A. bruises over the neck
   B. looped extension cord marks on the body
   C. bruises over bony prominences
   D. bruising of the torso
E. bruises over the ears

8. Burns are commonly seen in child abuse. Approximately 10% of children hospitalized with burns are victims of abuse.
Of the following, inflicted burn can be MOST commonly the result of
   A. contact with hot iron
   B. contact with radiators
   C. cigarette application
   D. contact with matchsticks
   E. scalding injuries

9. You are discussing physical child abuse with medical students; you state that fractures are common presentation and those that should raise suspicion for abuse include fractures that are unexplained, occurring in young, non-ambulatory children, or involve multiple bones.
Of the following, the site of the fracture that is LESS specific for abuse is
   A. rib
   B. scapula
   C. skull
   D. radius
   E. vertebra

10. The physical abuse of children by parents affects children of all ages. It is estimated that 1% to 2% of children are physically abused during childhood and that significant number of them are fatally injured each year.
Of the following, the second LEADING cause of mortality from physical abuse is
   A. rib fractures
   B. head trauma
   C. abdominal injury
   D. hot tap water burn
   E. trunk bruising

11. Child sexual abuse is the involvement of children in sexual activities that violates societal taboos. Most perpetrators are adults or adolescents who are known to the child and who have real or perceived power over the child.
Of the following, perpetrators who are LESS often involved in child sexual abuse are
   A. fathers
   B. mothers
12. Sexual abuse should be considered in children who have behavioral problems, although no behavior is pathognomonic. Which of the following behavior should raise the suspicion of sexual abuse?
   A. aggressive
   B. obsessive-compulsive
   C. dissociative
   D. depressive
   E. hypersexuality

13. You are meeting parents of a 5-year-old boy; the mother is wondering about the normal sexual behavior of her child. Of the following, the sexual behavior that is considered **HIGHLY** unusual in this age group is
   A. touching his genitals in public
   B. showing his genitals to others
   C. undressing in public
   D. pretending to be a member of the opposite sex
   E. imitating intercourse or other adult sexual behaviors

14. Gender identity disorder (GID) is characterized by intense and persistent cross-gender identification and discomfort with one’s own sex. In early school-age children, the manifestation that is **LEAST** likely considered as GID is
   A. dressing as a member of the opposite sex (i.e., cross dressing)
   B. strong belief that one is the opposite sex
   C. exclusive preference for cross sex roles
   D. playing with toys designed for the opposite sex
   E. strong preference for cross playmates

15. You are meeting a couple who are going to adopt a 1-year-old girl; they are asking you about medical investigations that are needed for the child before adoption. Of the following, the **LEAST** likely required investigation for the adoption of this girl is
   A. neonatal screening tests
   B. immunization history
C. screening tests for sexually transmitted diseases
D. cognitive tests
E. complete blood count

16. A 32-week gestational age male neonate is born to a 30-year-old mother due to abruptio placentae. The baby has been developed intracranial hemorrhage in the first few hours after birth. The mother has a history of using multiple drugs and substances during her pregnancy.
Of the following, the **MOST** likely offending substance
   A. alcohol
   B. codeine
   C. cigarettes
   D. diazepam
   E. cocaine

17. You are evaluating a 2-year-old boy who is experiencing intimate partner violence (IPV) between his parents.
Of the following, the **MOST** likely problem that may be seen in such a child is
   A. conduct disorder
   B. low self-esteem
   C. regression
   D. temper tantrums
   E. disrupted attachment and routines around eating and sleeping

18. Bullying is a form of aggression in which a child repeatedly and intentionally intimidates, harasses, or physically harms another child. Children who are being bullied are more likely to have
   A. suicidal ideation
   B. smoking habit
   C. alcohol abuse
   D. serious violent offenses
   E. raping crimes

19. Youth violence is a problem in urban, suburban, and rural communities and affects children across race and gender. Which subset of children who has more severe violence behavior that continues into adulthood?
   A. adolescents
   B. school-aged
   C. children of all ages with intermittent violence
20. Although understanding risk factors for violence is crucial for developing prevention strategies, the risk factors do not predict whether a particular individual will become violent.

For children who begin their violence early in life, the strongest risk factor for violence is

A. male gender  
B. early substance abuse  
C. poverty  
D. antisocial behavior  
E. hyperactivity

21. A 14-year-old boy develops a youth violence that has been started 6 months ago. The parents are asking about the factors that may increase the risks for serious violence.

Of the following, the **MOST** important risk factor for development of serious youth violence in this boy is

A. male gender  
B. substance abuse  
C. antisocial delinquent friends  
D. low IQ  
E. aggression

22. Dating violence and date rape are common. Adolescent women experience higher rates of sexual assault than any other age group. Substance abuse during the sexual assault is common.

Which of the following substance is commonly used in the episode of sexual assault?

A. alcohol  
B. benzodiazepines  
C. cocaine  
D. marijuana  
E. morphine

23. The child’s reaction to the divorce is influenced by the child’s age and developmental level.
Of the following, the **MOST** likely feeling that preschool children may have when their parents are divorced is

A. fear of abandonment  
B. anger and rejection  
C. depression  
D. unsecure  
E. irritability and listlessness

24. A mother of 4 children is having a relatively long-period business trip in near future and she has to leave her children with their grandmother; she is worrying about how her kids will experience this separation. The children are aged 14, 8, 5, and 2 year old consecutively.

The child who is often having the **MOST** difficult adjustment to a separation from his mother is

A. the youngest child  
B. the child of 8-year-old  
C. the child of 5-year-old  
D. the adolescent  
E. no one will experience any separation

25. The father of a 7-year-old child has been died in a car accident. The mother has been noted many reactions in her child after the father death.

Of the following, the child’s reaction that would need immediate evaluation is

A. yearning to be with the dead father  
B. expression a wish to die so that he can visit the dead father  
C. desire to commit suicide  
D. poor academic performance  
E. lack of enjoyment with activities
1. (D). For prematurity; weight corrections are needed until 24 months of age, height corrections until 40 months of age, and head circumference corrections until 18 months of age.

2. (D). In children with FTT, malnutrition initially results in wasting (deficiency in weight gain). Stunting (deficiency in linear growth) generally occurs after months of malnutrition, and head circumference is spared except with chronic, severe malnutrition.

3. (D). FTT is often diagnosed by weight that falls or remains below the 3rd percentile for age; decreases, crossing two major percentile lines on the growth chart over time; or is less than 80% of the median weight for the height of the child. In the first few years of life, large fluctuations in percentile position can occur in normal children.

4. (E). FTT due to celiac disease may occur after introduction of solid foods containing gluten. All other factors may cause FTT during the neonatal period.

5. (A). Changes in serum electrolyte concentrations and the associated complications are collectively termed the refeeding syndrome. These changes typically affect phosphorus, potassium, calcium, and magnesium.

6. (B). Although mothers are most frequently reported as the perpetrators of physical abuse, serious injuries, such as head or abdominal trauma, are more likely to be inflicted by fathers or maternal boyfriends.

7. (C). Bruises suggestive of abuse include those that are patterned, such as a slap mark on the face or looped extension cord marks on the body. Bruises in healthy children generally are distributed over bony prominences; bruises that occur in an unusual distribution, such as isolated to the torso, ears or neck, should raise concern.

8. (E). Inflicted burns can be the result of contact with hot objects (irons, radiators, or cigarettes) but more commonly the result of scalding injuries.

9. (D).

10. (C). Abusive head trauma is the leading cause of mortality and morbidity from physical abuse.

11. (B). Perpetrators are more often male than female.

12. (E).

13. (E). If this behavior is occurring, the child should be evaluated for exposure to inappropriate sexual material and possible sexual abuse. Stating that one wants to
be a member of the opposite sex and pretending to be a member of the opposite sex are not unusual behaviors in this age group.
14.(D). By this age, dressing as a member of the opposite sex and, particularly, stating a desire to be the opposite sex are uncommon, but playing with toys designed for the opposite sex remains common.
15.(D). Although adopted children have a higher rate of school, learning, and behavioral problems, much of this increase is likely to be related to biologic and social influences before the adoption.
16.(E). Fetal alcohol syndrome (FAS) characterized by in utero and postnatal growth retardation, microcephaly, intellectual disability, and a characteristic dysmorphic facial appearance. Cigarette smoking during pregnancy is associated with lower birth weight and increased child behavioral problems. Use of cocaine in the perinatal period has been associated with prematurity, intracranial hemorrhages, and abruptio placentae.
17.(E). Older children may have conduct disorders, poor school performance, low self-esteem, or other nonspecific behaviors. Infants and young toddlers are at risk for disrupted attachment and routines around eating and sleeping. Preschoolers may show signs of regression, irritable behavior, or temper tantrums.
18.(A). Psychosocial consequences of being bullied include depression and suicidal ideation. Children who bully others are more likely to be involved with other problem behaviors, such as smoking and alcohol use.
19.(E). These children tend to be more serious offenders, perpetrate more crimes, and more often continue their violence into adulthood. Most adolescent violence ends by young adulthood.
20.(B). For children who begin their violence early in life, the strongest risk factors are early substance abuse (<12 years of age) and perpetration of nonviolent, serious crimes during childhood.
21.(C). For children who begin their violence during adolescence, individual risk factors are less important, whereas factors related to peer groups are most important. Gang membership, associating with antisocial or delinquent friends, being unpopular in school, and having weak ties to conventional peer groups are important risk factors for adolescent-onset violence.
22.(A). Alcohol use is common in episodes of adolescent sexual assault, occurring in approximately 50% of cases.
23.(A). Feeling unsecured with irritability and listlessness is mostly seen in infants. Anger and rejection are encountered in school-aged children. Adolescents may respond to the divorce by acting out, becoming depressed, or experiencing somatic symptoms.
24. (A). Children between 6 months and 3 to 4 years of age often have the most difficulty adjusting to a separation from their primary caregiver. Older children have cognitive and emotional skills that help them adjust.

25. (C). A plan or desire to commit suicide is uncommon and would need immediate evaluation.
1. There is an association between a long lactation of 12 to 23 months (cumulative lactation of all pregnancies) and a significant maternal reduction of all the following EXCEPT
   A. hypertension
   B. hyperlipidemia
   C. cardiovascular disease
   D. diabetes
   E. cervical cancer

2. The MOST objective indicator of adequate breast milk intake by the infant is
   A. voiding pattern
   B. stooling pattern
   C. rate of weight gain
   D. feeding frequency
   E. state of hydration

3. The American Academy of Pediatrics recommends vitamin D supplementation 400 IU/day for breastfed infants starting
   A. soon after birth
   B. after 1 month
   C. after 2 month
   D. after 3 month
   E. after 4 month

4. Permanent contraindication to breast feeding is recommended in
   A. active tuberculosis of the mother
   B. galactosemia of the infant
   C. Varicella of the mother
   D. H1N1 influenza of the mother
   E. Herpes simplex infection of the breast

5. Of the following breast milk components, which component is affected (reduced) after breast milk freezing and pasteurization?
   A. protein
6. The highest osmolality and renal solute load are seen in
   A. breast milk
   B. breast milk after freezing and pasteurizing
   C. standard formula
   D. soy formula
   E. hypoallergenic formula

7. Daily juice intake for toddlers and young children should be limited to
   A. 2 oz
   B. 4 oz
   C. 6 oz
   D. 8 oz
   E. 10 oz

8. A 5-year-old boy presented with obesity, syndactyly, infantile genitalia, and mental retardation. Fundoscopy revealed retinal degeneration, the parents are first cousin.
   Of the following, the MOST likely cause of his obesity is
   A. Alström syndrome
   B. Laurence-Moon-Bardet-Biedl
   C. Fröhlich syndrome
   D. Prader-Willi syndrome
   E. Carpenter syndrome

9. One of the activities for overweight/obesity prevention in less than 2 year-old-child is to limit watching television TV to
   A. no TV
   B. <1 hour
   C. <2 hours
   D. <3 hours
   E. <4 hours

10. To prevent obesity, families need to be counseled on age-appropriate and healthy eating patterns which include all the following EXCEPT

A. promotion of breastfeeding  
B. smaller bowls should be used  
C. children should never eat directly from a bag or box  
D. no juices or soda should be the rule  
E. food as a reward should be encouraged

11. The greatest consequence of undernutrition is  
   A. stunted growth  
   B. recurrent infections  
   C. death  
   D. intellectual disability  
   E. physical disability

12. Photosensitivity dermatitis in nutritional deficiency disorders of childhood is usually caused by deficiency of  
   A. niacin  
   B. zinc  
   C. vitamin A  
   D. riboflavin  
   E. vitamin C

13. Smooth tongue in nutritional deficiency disorders of childhood is usually caused by deficiency of  
   A. iron  
   B. copper  
   C. biotin  
   D. iodine  
   E. zinc

14. The body of the malnourished child may have compensated for micronutrient deficiencies with lower metabolic and growth rates, and refeeding may unmask these deficiencies. Refeeding syndrome is characterized by  
   A. dehydration  
   B. hyperphosphatemia  
   C. hypermagnesemia  
   D. hypokalemia  
   E. hypocalcemia
15. When nutritional rehabilitation has begun for a malnourished child, all the following are true **EXCEPT**
   A. caloric intake can be increased 10% to 20% per day
   B. caloric intake is increased until catch-up growth is initiated
   C. protein needs are increased as anabolism begins
   D. vitamin and mineral intake is provided
   E. iron supplements are recommended

16. A 10-month-old infant from rural area presented with pallor, irritability, inability to move his legs, and bleeding gums. The infant is on fresh cow’s milk from birth.
   Of the following, the **MOST** likely cause of his condition is deficiency of
   A. vitamin K
   B. vitamin B12
   C. vitamin C
   D. copper
   E. zinc

17. A 6-month-old infant with protein-calorie malnutrition receiving boiled milk from birth presented with anorexia, apathy, vomiting, restlessness, progressive pallor, dyspnea, and cyanosis. The baby is noticed to cry without sound.
   Of the following, the **MOST** likely cause of his condition is deficiency of
   A. thiamine
   B. riboflavin
   C. niacin
   D. zinc
   E. vitamin K

18. All the following matchings' between drugs and vitamin and nutrient deficiency states are true **EXCEPT**
   A. sulfonamides = folate
   B. phenytoin= vitamins D
   C. antibiotics= vitamin K
   D. isoniazid= vitamin B₁₂
   E. digitalis= calcium

19. A 4-year-old diabetic child from a low socioeconomic family presented with angular stomatitis; glossitis; cheilosis; seborrheic dermatitis around the nose and mouth; reduced tearing, and photophobia.
Of the following, the **MOST** likely cause of his condition is deficiency of
A. thiamine  
B. riboflavin  
C. niacin  
D. zinc  
E. vitamin K

20. The 4 d (dermatitis, dementia, diarrhea, and death) are characteristic features in the deficiency of
A. thiamine (B1)  
B. riboflavin (B2)  
C. niacin (B3)  
D. pyridoxine (B6)  
E. B12

21. A 14-month-old child presented with vomiting, diarrhea, failure to thrive, listlessness, hyperirritability, and seizures. His hemoglobin is 6 gm/dl, blood film shows hypochromic microcytic anemia, and he was fed goat’s milk from birth. Of the following, the **MOST** likely cause of his condition is deficiency of
A. thiamine (B1)  
B. riboflavin (B2)  
C. niacin (B3)  
D. pyridoxine (B6)  
E. B12

22. The dietary sources of one of the following vitamins are from animal products only
A. thiamine (B1)  
B. riboflavin (B2)  
C. niacin (B3)  
D. pyridoxine (B6)  
E. B12

23. The clinical manifestations of vitamin A deficiency in humans appear as a group of ocular signs termed xerophthalmia. Of the following, the **EARLIEST** manifestation is
A. night blindness  
B. xerosis of the conjunctiva and cornea  
C. corneal ulceration
D. keratomalacia  
E. corneal scar

24. A 2-year-old child presented with severe clinical manifestations of vitamin A deficiency including xerophthalmia. Of the following, the suggested therapeutic vitamin A doses are
A. oral 5,000–10,000 U/kg/day for 5 days or until recovery  
B. 200,000 U/day orally, q4–6mo  
C. 100,000 U/day for 3 days, then 50,000 U/day for 14 days  
D. oral 500,000 U/day for 3 days; then 50,000 U/day for 14 days; then 10,000–20,000 U/day for 2 mo  
E. 7,500–15,000 U/day IM, followed by oral 5000–10,000 U/day for 10 days

25. A 6-week-old preterm infant presented with progressive pallor and peripheral edema, his blood film revealed anemia, elevated reticulocyte count, and thrombocytosis. Of the following, the BEST management of this preterm is to
A. reassure the parents that it is a transient event  
B. give packed cell blood  
C. give folic acid  
D. give vitamin E  
E. repeat the CBC 2 weeks later

26. The best measure of vitamin D status is the level of
A. 25-(OH)-D  
B. 1,25-(OH)2-D  
C. 24,25-(OH)2-D  
D. serum phosphorus  
E. serum calcium

27. All the following plasma factors in the cascade of blood coagulation factors depend on vitamin K for synthesis and for post-translational conversion of their precursor proteins EXCEPT
A. II  
B. VII  
C. VIII  
D. IX  
E. X
28. The major minerals are those that require intakes of more than 100 mg/day and contribute at least 0.1% of total body weight.

All the following are essential major minerals EXCEPT

A. calcium  
B. phosphorus  
C. magnesium  
D. iron  
E. sulfur

29. The calcium equivalent of 1 cup of milk (about 300 mg of calcium) is

A. 1/2 cup of plain yogurt  
B. 3/4 cup of plain yogurt  
C. 1 cup of plain yogurt  
D. 1 1/4 cup of plain yogurt  
E. 1 1/2 cup of plain yogurt

30. Osteoporosis that occurs in childhood is related to all the following EXCEPT

A. protein-calorie malnutrition  
B. vitamin C deficiency  
C. zinc deficiency  
D. steroid therapy  
E. immobilization

31. The absorption of nonheme iron is influenced by the composition of consumed foods. Inhibitors of nonheme iron absorption include

A. ascorbic acid  
B. bran  
C. meat  
D. fish  
E. poultry

32. The percent intestinal absorption of iron from iron-fortified cow’s milk formula is

A. 4%  
B. 10%  
C. 14%  
D. 20%  
E. 24%

33. Oral treatment of iron deficiency anemia should be continued for
A. 2 months  
B. 3 months  
C. 4 months  
D. 5 months  
E. 6 months

34. On a global basis, widespread occurrence of zinc deficiency is attributed to  
   A. poor zinc bioavailability secondary to phytic acid  
   B. low intake  
   C. diarrhea  
   D. pneumonia  
   E. acrodermatitis enteropathica

35. Clinical manifestations of mild zinc deficiency include  
   A. growth faltering  
   B. delayed sexual maturation  
   C. mood changes  
   D. hepatosplenomegaly  
   E. periorificial erythematous, scaling dermatitis

36. Zinc is relatively nontoxic. Excess intake produces all the following **EXCEPT**  
   A. abdominal pain  
   B. headache  
   C. night blindness  
   D. vertigo  
   E. seizures
1. (E). Cumulative lactation of more than 12 months also correlates with reduced risk of ovarian and breast cancer.
2. (C). Rate of weight gain provides the most objective indicator of adequate milk intake. Total weight loss after birth should not exceed 7%, and birth weight should be regained by 10 days.
3. (A). The American Academy of Pediatrics recommends vitamin D supplementation (400 IU/day starting soon after birth), and, when needed, fluoride after 6 months for breastfed infants.
4. (B). Maternal infection with human immunodeficiency virus (HIV) is considered a contraindication for breastfeeding in developed countries. When the mother has active tuberculosis, syphilis, or varicella, restarting breastfeeding may be considered after therapy is initiated. If a woman has herpetic lesions on her breast, nursing and contact with the infant on that breast should be avoided. Women with genital herpes can breastfeed. Proper hand-washing procedures should be stressed.
There are limited numbers of medical contraindications for breastfeeding, including pediatric metabolic disorders such as galactosemia, and infants with phenylketonuria, although infants with the latter may alternate breastfeeding with special protein-free or modified formulas.
5. (A). The following components of breast milk are reduced after breast milk freezing and pasteurization, protein, IgA, SlgA, IgG, lactoferrin, lysozyme, and lipases.
6. (E). 290 mOsm/L and 125–175 mOsm/L in hypoallergenic formula, while they are 253 mOsm/L and 75 mOsm/L in breast milk.
7. (B). Juice intake for toddlers and young children should be limited to 4 oz, and juice intake for children 7 to 18 years of age should be limited to 8 oz/day. Water and milk are the recommended drinks during the day.
8. (B).
9. (A). Limit screen time (television, computer games/Internet, video games) to <1–2 hours per day (no TV for child <2 yr of age).
10. (E). Children should never be forced to eat when they are not willing, and overemphasis on food as a reward should be avoided. “Choose My Plate” by the U.S. Department of Agriculture can provide parents with a general guideline for the
types of foods to be offered on a regular basis, including fruits, vegetables, grains, protein, and dairy.

11. (C). The greatest consequence of undernutrition is death, but significant intellectual and physical disability exists in many who survive.

12. (A).

13. (A).

14. (D). Nutritional rehabilitation can be complicated by refeeding syndrome, which is characterized by fluid retention, hypophosphatemia, hypomagnesemia, and hypokalemia. Careful monitoring of laboratory values and clinical status with severe malnutrition is essential.

15. (E). Iron supplements are not recommended during the acute rehabilitation phase, especially for children with kwashiorkor, for whom ferritin levels are often high. Additional iron may pose an oxidative stress; iron supplementation is associated with higher morbidity and mortality.

16. (C). Infantile scurvy is manifested by irritability, bone tenderness with swelling, and pseudoparalysis of the legs. The disease may occur if infants are fed unsupplemented cow’s milk in the first year of life or if the diet is devoid of fruits and vegetables. Subperiosteal hemorrhage, bleeding gums and petechiae, hyperkeratosis of hair follicles, and a succession of mental changes characterize the progression of the illness. Anemia secondary to bleeding, decreased iron absorption, or abnormal folate metabolism is also seen in chronic scurvy.

17. (A). Infantile beriberi occurs between 1 and 4 months of age in breastfed infants whose mothers have a thiamine deficiency (alcoholism), in infants with protein-calorie malnutrition, in infants receiving unsupplemented hyperalimentation fluid, and in infants receiving boiled milk. Acute wet beriberi with cardiac symptoms and signs predominates in infantile beriberi. Anorexia, apathy, vomiting, restlessness, and pallor progress to dyspnea, cyanosis, and death from heart failure. Infants with beriberi have a characteristic aphonic cry; they appear to be crying, but no sound is uttered. Other signs include peripheral neuropathy and paresthesias.

18. (D).

- Phenytoin, phenobarbital = Vitamins D, K, folate
- Mineral oil = Vitamins A, D, E, K
- Isoniazid = Vitamin B6
- Antacids = Iron, phosphate, calcium
- Digitalis = Magnesium, calcium
- Penicillamine = Vitamin B6

19. (B). Ariboflavinosis is characterized by an angular stomatitis; glossitis; cheilosis; seborrheic dermatitis around the nose and mouth; and eye changes that include reduced tearing, photophobia, corneal vascularization, and the formation of
cataracts. Subclinical riboflavin deficiencies have been found in diabetic subjects, children in families with low socioeconomic status, children with chronic cardiac disease, and infants undergoing prolonged phototherapy for hyperbilirubinemia.

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

21. (D). Goat’s milk is deficient in vitamin B6. Dietary deprivation or malabsorption of vitamin B6 in children results in hypochromic microcytic anemia, vomiting, diarrhea, failure to thrive, listlessness, hyperirritability, and seizures. Children receiving isoniazid or penicillamine may require additional vitamin B6 because the drug binds to the vitamin. Vitamin B6 is unusual as a water-soluble vitamin in that very large doses (≥500 mg/day) have been associated with a sensory neuropathy.

22. (E). Dietary sources of vitamin B12 are animal products only. Strict vegetarians should take vitamin B12 supplement.

23. (A). The earliest symptom is night blindness, which is followed by xerosis of the conjunctiva and cornea.

24. (A).

25. (D). Deficient preterm infants at 1 to 2 months of age have hemolytic anemia characterized by an elevated reticulocyte count, an increased sensitivity of the erythrocytes to hemolysis in hydrogen peroxide, peripheral edema, and thrombocytosis. All the abnormalities are corrected after oral, lipid, or water-soluble vitamin E therapy.

26. (A). The serum calcium usually is normal but may be low; the serum phosphorus level usually is reduced, and serum alkaline phosphatase activity is elevated. When serum calcium levels decline to less than 7.5 mg/dL, tetany may occur. Levels of 24,25-(OH)2-D are undetectable, and serum 1,25-(OH)2-D levels are commonly less than 7 ng/mL, although 1,25-(OH)2-D levels also may be normal. The best measure of vitamin D status is the level of 25-(OH)-D.

27. (C). Plasma factors II (prothrombin), VII, IX, and X in the cascade of blood coagulation factors depend on vitamin K for synthesis and for post-translational conversion of their precursor proteins. The post-translational conversion of glutamyl residues to carboxyglutamic acid residues of a prothrombin molecule creates effective calcium-binding sites, making the protein active. Other vitamin K–dependent proteins include proteins C, S, and Z in plasma and γ-carboxyglutamic acid–containing proteins in several tissues. Bone contains a major vitamin K–dependent protein, osteocalcin, and lesser amounts of other glutamic acid–containing proteins.

28. (D). There are seven essential major minerals: calcium, phosphorus, magnesium, sodium, potassium, chloride, and sulfur.
29. **(B)**. Calcium intake can come from a variety of sources, with dairy products providing the most common and concentrated source. The calcium equivalent of 1 cup of milk (about 300 mg of calcium) is 3/4 cup of plain yogurt, 1.5 oz of cheddar cheese, 2 cups of ice cream, 4/5 cup of almonds, or 2.5 oz of sardines. Other sources of calcium include some leafy green vegetables (broccoli, kale, collards); lime-processed tortillas; calcium-precipitated tofu; and calcium-fortified juices, cereals, and breads.

30. **(C)**. Osteoporosis that occurs in childhood is related to protein-calorie malnutrition, vitamin C deficiency, steroid therapy, endocrine disorders, immobilization and disuse, osteogenesis imperfecta, or calcium deficiency (in premature infants).

31. **(B)**. Enhancers of nonheme iron absorption are ascorbic acid, meat, fish, and poultry. Inhibitors are bran, polyphenols (including the tannates in tea), and phytic acid, a compound found in legumes and whole grains.

32. **(A)**. The percent intestinal absorption of the small amount of iron in human milk is 10%; 4% is absorbed from iron-fortified cow’s milk formula and from iron-fortified infant dry cereals.

33. **(D)**. Treatment of iron deficiency anemia includes changes in the diet to provide adequate iron and the administration of 2 to 6 mg iron/kg/24 hr (as ferrous sulfate) divided bid or tid. Reticulocytosis is noted within 3 to 7 days of starting treatment. Oral treatment should be continued for 5 months.

34. **(A)**. On a global basis, poor bioavailability secondary to phytic acid is thought to be a more important factor than low intake in the widespread occurrence of zinc deficiency.

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

36. **(C)**. Excess intake produces nausea, emesis, abdominal pain, headache, vertigo, and seizures.
1. Total body water (TBW) as a percentage of body weight varies with age. Which of the following is true regarding TBW?
   A. TBW is about 60% of birth weight for a term infant
   B. premature infants have a higher TBW content than term infants
   C. during the first year of life, TBW decreases to about 50% of body weight
   D. by the end of puberty TBW in males is 50% of body weight
   E. by the end of puberty TBW in females is 65% of body weight

2. The MOST abundant cation in the intracellular fluid is
   A. sodium
   B. chloride
   C. potassium
   D. proteins
   E. phosphate

3. Proper cell functioning requires close regulation of plasma osmolality and intravascular volume. Which of the following statements is TRUE?
   A. maintenance of a normal osmolality depends on regulation of sodium balance
   B. control of volume status depends on regulation of potassium balance
   C. the plasma osmolality is controlled between 295 and 305 mosm/kg
   D. a small increase in the plasma osmolality stimulates thirst
   E. the secretion of antidiuretic hormone decreases with increasing plasma osmolality

4. Maintenance fluids are composed of a solution of water, glucose, sodium, potassium, and chloride. How much of the normal caloric needs of the patient is provided by glucose in maintenance fluids?
   A. 5%
   B. 10%
   C. 15%
   D. 20%
   E. 25%
5. Volume depletion without renal insufficiency may cause a disproportionate increase in the BUN, with little or no change in the creatinine concentration. This increase in the BUN may be absent or blunted in a child with
   A. iron deficiency anemia
   B. growth failure
   C. poor protein intake
   D. gastrointestinal bleed
   E. glucocorticoids intake

6. Which of the following findings is considered abnormal on urinalysis of a patient with dehydration?
   A. hyaline casts
   B. granular casts
   C. few white blood cells
   D. few red blood cells
   E. proteinuria of 150 mg/dl

7. Rapid correction of hyponatremia (>12 mEq/L/24 hr) should be avoided because of the remote risk of
   A. central pontine myelinolysis
   B. cerebral edema
   C. brain herniation
   D. acute tubular necrosis
   E. hypocalcemia

8. The MOST common indication for long-term parenteral nutrition is
   A. short bowel syndrome
   B. intractable diarrhea syndromes
   C. intestinal pseudo-obstruction
   D. inflammatory bowel disease
   E. immunodeficiency

9. The MOST concerning complication of long-term parenteral nutrition is
   A. nutritional deficiencies
   B. cholestatic liver disease
   C. hyperglycemia
   D. azotemia
   E. hyperammonemia
10. Pseudohyponatremia is a laboratory artifact that is present when the plasma contains high concentrations of
   A. potassium
   B. zinc
   C. protein
   D. ascorbic acid
   E. chloride

11. Classification of true hyponatremia is based on the patient’s
   A. conscious level
   B. volume status
   C. renal function
   D. serum osmolality
   E. parathyroid hormone level

12. Hyponatremia with high osmolality occurs in
   A. hyperglycemia
   B. SIADH
   C. glucocorticoid deficiency
   D. hypothyroidism
   E. water intoxication

13. Which of the following conditions lead to euvolemic hyponatremia?
   A. congestive heart failure
   B. cirrhosis
   C. nephrotic syndrome
   D. water intoxication
   E. hypoalbuminemia

14. Hypokalemia is common in children, with most cases related to
   A. gastroenteritis
   B. low intake
   C. renal losses
   D. receiving aggressive doses of β-adrenergic agonists
   E. anorexia nervosa

15. A specific sign of severe dehydration is
   A. dry mucous membrane
   B. oliguria
C. reduced and mottled cutaneous perfusion
D. thirsty
E. slight reduction in skin turgor

16. Oral rehydration therapy can be used to treat dehydration in patients with
   A. oliguria
   B. uncontrollable vomiting
   C. coma
   D. inability to drink
   E. gastric or intestinal distention

17. In treatment of hyperkalemia, which measure removes potassium from the body?
   A. sodium bicarbonate administration (IV)
   B. loop diuretic (IV or PO)
   C. insulin and glucose (IV)
   D. β-Agonist (albuterol via nebulizer)
   E. IV calcium

18. Which of the following is a cause of metabolic acidosis with normal anion gap?
   A. lactic acidosis (shock)
   B. diarrhea
   C. ketoacidosis
   D. kidney failure
   E. inborn errors of metabolism

19. Hypokalemia, metabolic alkalosis, hypercalciuria, and nephrocalcinosis are seen in
   A. Bartter syndrome
   B. Gitelman syndrome
   C. Cushing syndrome
   D. Addison disease
   E. IgA nephropathy

20. Electrocardiographic changes of flattened T wave, depressed ST segment, and appearance of U wave is seen in
   A. hyperkalemia
   B. hyponatremia
   C. hypomagnesaemia
21. The combination of hypokalemia and metabolic acidosis is characteristic of
   A. distal renal tubular acidosis
   B. gastric losses
   C. aldosterone excess
   D. diuretics
   E. Gitelman syndrome

22. Children with pseudohypoaldosteronism type 1 have
   A. hypokalemia
   B. metabolic alkalosis
   C. hypernatremia
   D. volume depletion
   E. decreased aldosterone levels
1. (B). The fetus has a high TBW, which gradually decreases to about 75% of birth weight for a term infant. Premature infants have a higher TBW content than term infants. Because fat has low water content, and muscle has high water content, by the end of puberty TBW in males remains at 60%, but it decreases to 50% of body weight in females.

2. (C). Sodium and chloride are the dominant cation and anion in the ECF. Potassium is the most abundant cation in the ICF, and proteins, organic anions, and phosphate are the most plentiful anions in the ICF.

3. (D). Maintenance of a normal osmolality depends on control of water balance. Control of volume status depends on regulation of sodium balance. The plasma osmolality is tightly controlled between 285 and 295 mOsm/kg through regulation of water intake and urinary water losses. A small increase in the plasma osmolality stimulates thirst. Urinary water losses are regulated by the secretion of antidiuretic hormone (ADH), which increases in response to an increasing plasma osmolality.

4. (D). This percentage is enough to prevent the development of starvation ketoacidosis and diminishes the protein degradation that would occur if the patient received no calories.

5. (C). The increase in the BUN may be absent or blunted in a child with poor protein intake because urea production depends on protein degradation.

6. (E). With dehydration, a urinalysis may show hyaline and granular casts, a few white blood cells and red blood cells, and 30 to 100 mg/dL of proteinuria.

7. (A). Overly rapid correction of hyponatremia (>12 mEq/L/24 hr) should be avoided because of the remote risk of central pontine myelinolysis.

8. (A). Short bowel syndrome is the most common indication for long-term PN; it may be caused by a congenital gastrointestinal anomaly or acquired after necrotizing enterocolitis.

9. (B). It can lead to cirrhosis and liver failure. Current PN decreases the risk of liver disease by including reduced amounts of hepatotoxic amino acids. The best preventive strategy is early use of the gastrointestinal tract, even if only trophic feeds are tolerated.

10. (C). Pseudohyponatremia is a laboratory artifact that is present when the plasma contains high concentrations of protein or lipid.

11. (B). Classification of true hyponatremia is based on the patient’s volume status.

12. (A). All the other conditions associated with low osmolality.
13. (D). All the other conditions associated with hypervolemic hyponatremia.
14. (A). Hypokalemia is common in children, with most cases related to gastroenteritis.
15. (C).
16. (A). IV therapy may still be required for patients with severe dehydration; patients with uncontrollable vomiting; patients unable to drink because of extreme fatigue, stupor, or coma; or patients with gastric or intestinal distention.
17. (B). Sodium bicarbonate administration (IV), insulin and glucose (IV), and β-agonist (albuterol via nebulizer) shift potassium intracellularly to decrease the risk of life-threatening arrhythmias while IV calcium is cardiac membrane stabilization.
18. (B). Other options are causes of metabolic acidosis with increased anion gap.
19. (A). Bartter syndrome and Gitelman syndrome are autosomal recessive disorders resulting from defects in tubular transporters. Both disorders are associated with hypokalemia and metabolic alkalosis. Bartter syndrome is usually associated with hypercalciuria, often with nephrocalcinosis; children with Gitelman syndrome have low urinary calcium losses, but hypomagnesemia secondary to urinary losses.
20. (E). Electrocardiographic (ECG) changes include a flattened T wave, a depressed ST segment, and the appearance of a U wave, which is located between the T wave (if still visible) and P wave.
21. (A). The combination of Hypokalemia and metabolic acidosis is characteristic of diarrhea, distal renal tubular acidosis, and proximal renal tubular acidosis. A concurrent metabolic alkalosis is characteristic of gastric losses, aldosterone excess, diuretics, Bartter syndrome, or Gitelman syndrome.
22. (D). Children with pseudohypoaldosteronism type 1 have hyperkalemia, metabolic acidosis, and salt wasting, leading to hyponatremia and volume depletion; aldosterone levels are elevated.
1. During initial assessment and resuscitation of an acutely injured (traumatized) child after applying of ABCDE approach. Of the following, an important extra action is
   A. assessment of O₂ saturation
   B. assessment of hydration status
   C. protection of cervical spine
   D. medical management of shock
   E. preparation to mechanical ventilation

2. In 2010, the biggest change that the American Heart Association revised in resuscitation for unresponsive adults, children, or an infant's is to
   A. start chest compressions immediately
   B. activate emergency medical service first
   C. attach the AED machine before initiating cardioversion shock
   D. start vaso-active medications sooner after resuscitation
   E. start with airway and breathing

3. Signs of inadequate tissue perfusion in a shocked patient, include all the following EXCEPT
   A. pallor
   B. warm skin
   C. poor pulses
   D. abnormal mentation
   E. oliguria

4. All the following are tools for assessment of central nervous system perfusion EXCEPT
   A. responsiveness scale (AVPU)
   B. recognition of parents or caregivers
   C. pupil size
   D. posturing
   E. blood pressure
5. A 2-month-old baby boy presented with severe pallor, cold extremities and poor pulses, you suspected hemorrhage, after administration of O2. Of the following, the MOST important next step is to start
   A. isotonic crystalloids
   B. packed red blood cells transfusion
   C. FFP
   D. inotropes
   E. chest compression

6. The MOST important bed side test done during initial resuscitation of an acutely ill infant are O2 saturation measurement and
   A. serum calcium
   B. blood glucose
   C. blood gases
   D. blood urea
   E. serum sodium

7. You are in an out-of-hospital setting facing a situation of unresponsive child; knowing little about the patient and obligated to intubate the child. The endotracheal tube size approximately equals to
   A. child’s mid-index phalanx
   B. child’s mid-fifth phalanx
   C. 4 + (patient age in years/4)
   D. child's weight + 6
   E. 3 + (patient age in years/4)

8. You are a 4th year pediatric board trainee, asked to explain the mechanism of airway compromise during loss of consciousness to a medical student. The BEST statement you would like to use is that it’s due to
   A. narrow air ways during childhood and its rapid collapse
   B. accumulation of thick secretion and inability to expectorate it
   C. soft laryngeal cartilage and loss of it's tone
   D. underdevelopment of nasal and oral pharynx
   E. loss of muscle tone allowing the mandible and tongue to move back

9. You are in pediatric ward when you called to check a 10-month-old baby girl who was admitted for (high grade fever) and now she is not responding to voice. You find her, poorly perfused with weak pulse, rate of 50/min with no breathing efforts.
Of the following, the **BEST** action is to
A. attach cardiac and respiratory monitor
B. open the airway by head tilt - chin lift maneuver
C. start mouth to mouth breathing
D. start chest compression
E. apply face mask O2

10. The **BEST** second step in question 9 scenario is to
A. open the airway by head tilt - chin lift maneuver
B. start mouth to mouth breathing
C. use bag-mask ventilation
D. order for atropine IV administration
E. administer 2ml/kg of 25% glucose

11. A 5\textsuperscript{th} year medical student is asking you about the main difference between defibrillation and cardioversion.
Of the following, the **BEST** answer is
A. synchronization
B. use of premedication
C. type of paddle used
D. attachment to patient
E. type of electrical current

12. A fullterm baby delivered by normal vaginal delivery suffers birth asphyxia (Grade II-III hypoxic-ischemic encephalopathy), had been intubated without difficulty, now he is on ventilator; the staff nurse asking you about the type of sedation to be used.
Of the following, the **BEST** answer is
A. midazolam and fentanyl
B. midazolam and atracurum
C. midazolam and morphine
D. morphine alone
E. no sedation

13. A 5-month-old baby boy who is a known case of reflux nephropathy presented with high grade fever, toxicity, tachypnea, and increased work of breathing. O2 saturation in room air was 83%. You started O2 flow by face mask with Fio2 of 0.30 (30% fractional inspired O2) and admitted the baby to PICU. His CBC shows shift to left with increased acute phase reactant. Arterialized capillary blood gas analysis
shows Pao₂ 65 mmhg and Pco₂ 38 mmhg; chest X-ray shows increased vascular markings and his blood culture is awaited.

Of the followings, the MOST likely diagnosis is

A. uro-sepsis
B. systemic inflammatory response syndrome (SIRS)
C. acute lung injury (ALI)
D. acute respiratory distress syndrome (ARDS)
E. respiratory failure

14. Taking in consideration that the heated humidified high-flow nasal cannula had failed to improve oxygenation and improve symptoms for the patient in above scenario (question 13).

Of the following, the MOST effective method of treatment is

A. continuous positive airway pressure (CPAP)
B. conventional mechanical ventilation (CMV)
C. high-frequency oscillatory ventilation (HFOV)
D. extracorporeal membrane oxygenation (ECMO)
E. bag and mask ventilation

15. You are treating a 5-month-old baby boy with severe acute bronchiolitis. Mother is concerned about the prognosis of such an illness. Your BEST response to her is to say

A. severe form mostly needs admission to intensive care unit with guarded prognosis
B. they are mostly recovered and only fewer than 1% dies because of respiratory failure
C. all patients I have seen recovers without long term complications
D. it is a self limiting illness and O2 is the only effective treatment
E. those patients never develops respiratory failure

16. All the following may play role in prevention of respiratory failure EXCEPT

A. active immunization to H. influenzae type b
B. active immunization to group A beta-hemolytic streptococcus
C. passive immunization with respiratory syncytial virus immunoglobulins
D. active immunization to pertussis
E. compliance with appropriate therapies for asthma

17. What is the type of shock in methemoglobinemia?

A. cardiogenic shock
B. hypovolemic shock
C. distributive shock
D. dissociative shock
E. obstructive shock

18. Massive pulmonary embolism causes
   A. cardiogenic shock
   B. hypovolemic shock
   C. distributive shock
   D. dissociative shock
   E. obstructive shock

19. Severe hemorrhage or diarrhea can cause
   A. cardiogenic shock
   B. hypovolemic shock
   C. distributive shock
   D. dissociative shock
   E. septic shock

20. Of the following, the investigation that is useful in all types of shock is
   A. echocardiography
   B. arterial blood gases and lactate
   C. electrolytes
   D. mixed venous oxygen saturation
   E. bacterial and viral cultures

21. Of the following, the test that may help in differentiating septic shock from other types of shock is
   A. lactate
   B. electrolytes
   C. mixed venous oxygen saturation
   D. renal function test
   E. complete blood picture

22. In distributive shock (like septic shock), the systemic inflammatory response syndrome (SIRS) may precede the shock. All the following may be part of the syndrome **EXCEPT**
   A. temperature greater than 38°C
   B. heart rate greater than two standard deviations above normal for age
C. temperature less than 36°C  
D. tachypnea  
E. white blood count immature form greater than 5%

23. An 8-month-old bottle fed baby boy from rural area had history of fever and frequent bowel motions for 3 days. On examination he found to have severe dehydration, tachycardia, tachypnea, hypotension, with good volume pulse and high grade fever. Weight was on 3rd centile.

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

A. start low dose dopamin infusion  
B. start high dose dopamin infusion  
C. give bolus of normal saline  
D. start norepinephrine  
E. give bolus of colloid

24. Of the following, the **MOST** common cause of death in children above 1 year of age is

A. sepsis  
B. suffocation  
C. drowning  
D. motor vehicle accidents  
E. intentional injury

25. A child wearing helmet during riding bicycle is an example of which strategy in trauma prevention in childhood?

A. primary strategy  
B. secondary strategy  
C. tertiary strategy  
D. combined strategy  
E. automatic strategy

26. The **MOST** effective strategy in trauma prevention in childhood is

A. primary strategy  
B. secondary strategy  
C. tertiary strategy  
D. combined strategy  
E. automatic strategy
27. An 18-month-old boy brought to emergency department, suffering from non-fatal drowning. Mother describes the episode as he was swimming with his sibs at home when they suddenly shouted for help. Sooner she arrived and removed him from swimming pool; he was unconscious when she started resuscitation. He responded well within few minutes and regain consciousness. Now he is alert with normal vitals.

Of the following the MOST appropriate next action is to

A. admit to general ward for 24 hours
B. send for neuro-imaging
C. observe for 6-12 hours
D. start prophylactic antibiotics
E. reassure and send home

28. In non-fatal drowning, the outcome of drowning is determined by the success of immediate resuscitation efforts and the severity of the hypoxic-ischemic injury to the brain.

All the following are considered as unfavorable prognostic markers EXCEPT

A. need for CPR for more than 25 minutes
B. continuing CPR at the hospital
C. Glasgow Coma Scale of 5 or less
D. development of seizure
E. coma for 24 hours

29. Of the following, the COMMONEST cause of poisoning in childhood is with

A. cosmetics
B. iron
C. antihistamin
D. carbon monoxide
E. aspirin

30. Of the following, the COMMONEST cause of fatal poisoning in children is/are

A. analgesics
B. iron
C. alcohol
D. organophosphorous
E. cyanide

31. A poisoned child can exhibit any one of following basic clinical patterns EXCEPT

A. coma
32. A mother of an 11-month-old baby girl who had ingested a tasteless alkali, initial endoscopy revealed full-thickness liquefaction necrosis of the esophagus. She underwent successful dilatation of esophageal stricture after 3 weeks from the accident. Mother is asking about any other future risk of this condition. Of the following, the MOST appropriate explanation include

A. as she passed the acute illness successfully; no other complications is expected
B. the likelihood of recurrence of esophageal stricture is high
C. she needs long term follow-up as there is risk of esophageal carcinoma
D. there is possibility of re-infection of affected site
E. long-term esophageal motility disorder is expected and need follow-up

33. An 18-month-old girl brought to ER department after ingesting a small button battery left by father after repair of his watch. She was sitting comfortably with no complaint apart from mild saliva drooling, chest X-ray shows a radio-opaque rounded shadow in the middle of esophagus. Of the following, the BEST action is

A. send the child home with advice to come after 12-24 hours
B. advise to give a plenty of milk and fluid to move it down
C. use a Folly's catheter to pull the foreign body up
D. arrange for endoscopic interference
E. keep in ER room under observation for 24 hours

34. A 20-month-old toddler with suspected poisoning, his blood gas analysis shows high anion gap metabolic acidosis and hyperosmolar load. All the following are possible causes of this poisoning EXCEPT

A. metformin
B. ethylene glycol
C. phenformin
D. iron
E. salicylate

35. Measuring random blood glucose is part of screening clue in toxicological diagnosis. All the following can cause hypoglycemia EXCEPT
A. ethanol  
B. isoniazid  
C. insulin  
D. propranolol  
E. salicylate

36. All the following investigations/laboratory measurers are indicated as screening clue in toxicological diagnosis EXCEPT  
   A. ECG  
   B. blood gas analysis  
   C. urine screen  
   D. stool screen  
   E. blood quantitative assay

37. A 2-year-old girl brought to ER because of decreased level of consciousness. The family stated that, she complained from abdominal pain yesterday and grandmother gave her traditional medication. Her vitals remain normal, but she is responding to painful stimuli only and her pupils are constricted. 
All the following is considered as supportive therapy for a suspect case of poisoning EXCEPT  
   A. protecting and maintaining the airway  
   B. glucose (1 g/kg intravenously)  
   C. naloxone  
   D. glucagon  
   E. 100% oxygen

38. A 2-year-old boy brought to emergency department with history of taking many tablets of panadol (acetaminophen) 30 minutes earlier; you were the most senior doctor in the team. 
Your BEST advice regarding gastric decontamination is to use/do  
   A. syrup of ipecac  
   B. gastric lavage  
   C. activated charcoal  
   D. sorbitol as cathartic  
   E. polyethylene glycol for whole-bowel irrigation

39. Charcoal that is used for gastric decontamination in case of acute poisoning is effective against  
   A. caustics
40. Multiple-dose activated charcoal is considered in patient has ingested a life-threatening amount of
   A. theophylline
   B. azithromycin
   C. ibuprofen
   D. acetaminophen
   E. sodium valprate

41. Enhanced elimination by alkalinization of urine may be helpful in toxic doses or ingestion of
   A. methotrexate
   B. acetaminophen
   C. oral hypoglycemic agents
   D. phenobarbitone
   E. metoprolol

42. The specific antidote for acetaminophen poisoning is
   A. flumazenil
   B. glucagon
   C. sodium bicarbonate
   D. deferoxamine
   E. N-acetylcysteine

43. Of the following the specific antidote for organophosphates poisoning is
   A. atropine
   B. BAL
   C. naloxone
   D. methylene blue
   E. pralidoxime

44. Many medications are subjected to therapeutic monitoring for drug toxicity (measurement of drug level) due to their potential side effects. Of the following, the **LEAST** one in the list is
   A. amikacin
B. digoxin  
C. acetaminophen  
D. vancomycin  
E. vincristine

45. A 6-year-old child, a known case of ALL on maintenance therapy came for bone marrow aspiration.  
Of the following, the **BEST** medication used for sedation with the least side effects is  
A. midazolam  
B. dexmedetomidine  
C. ketamine  
D. chloral hydrate  
E. propofol

46. A full-term newborn baby suspected to have total anomalous pulmonary venous drainage elected for CT angio study.  
Of the following, the **BEST** choice for medication that is causing sedation without respiratory depression is  
A. midazolam  
B. dexmedetomidine  
C. ketamine  
D. fentanyl  
E. propofol
1. (C). Protection of the cervical spine also should be initiated at this step in any child with traumatic injury or who presents with altered mental status of uncertain etiology.

2. (A). Spending more time in evaluation of pulse by a lay person may lead to delayed in starting resuscitation. Now the principal of CAB is applied (circulation, airway, and breathing).

3. (B). Signs of poor perfusion also include cool skin, oliguria and delayed capillary refill.

4. (E). It is useful in assessment of circulation.

5. (A). After ABCDE resuscitation/evaluation, administration of high flow O2 and starting crystalloid is next step in a shocked child. A total of 1-3 shots (in duration of 15-60 min) can be given to restore the circulation; maintain the minimum systolic blood pressure to deliver adequate flow to the vital organs, and to produce adequate urine output. If hemorrhage is known or highly suspected, administration of packed red blood cells is appropriate next step. In critical situation waiting for cross matching may endanger the life of the patient, using of O negative universal donor is allowed. FFP can be given after frequent blood transfusions or when the patient becomes coagulopathic. Inotropes used if fluid resuscitation and drugs that increases the peripheral circulation failed.

6. (B). Neonates and infants have limited glycogen stores, which may become rapidly depleted during shock and lead to hypoglycemia. Alternatively, high levels of endogenous and exogenous catecholamines may cause a relative insulin-resistant state that can result in serum hyperglycemia. Shock may also cause alterations in available levels of serum ionized calcium (2nd appropriate answer), despite normal total serum calcium levels. Hypocalcemia in the shock state is due to impaired parathyroid hormone function, decreased hepatorenal vitamin D hydroxylation, and end-organ resistance.

7. (A). Option C is also true in the case of knowing more about the patient, like the age and weight. Option D is used in neonate to estimate the length of endotracheal tube.

8. (E). In children, airway patency often is compromised by a loss of muscle tone, allowing the mandibular block of tissue, including the tongue, bony mandible, and the soft surrounding tissues, to rest against the posterior pharyngeal wall.
9. (D). The goal in resuscitating a pediatric patient following a cardiopulmonary arrest should be to optimize cardiac output and tissue oxygen delivery, which may be accomplished by using artificial ventilation and chest compression and by the judicious administration of pharmacologic agents that what is called APLS (advanced pediatric life support) which is usually done in hospital setting or when there is good medical facilities and assistants.

Chest compressions should be initiated if a pulse cannot be palpated or if the heart rate is less than 60 beats/min with signs of poor systemic perfusion.

10. (A). The sequence of resuscitation usually follows CAB schedule, so after chest compression you need to

- Open the airway by head tilt - chin lift maneuver.
- Use effective bag and mask ventilation with 100% O2 (effective means that the chest movements is evidently raised with bagging) if ventilation is required for short period, but in case of long period ventilation is required, endotracheal intubation is indicated.
- Use medications, like glucose, IV fluid, atropine, calcium, sodium bicarbonate, epinephrine, lidocaine, adenosine or amiodarone as indicated per each case.

11. (A). Cardioversion requires a lower starting dose and synchronization of the discharge to the patient's electrocardiogram to prevent discharging during a susceptible period (which may convert supraventricular tachycardia to ventricular tachycardia or fibrillation which is called R on T), and it is mainly used for SVT and VT with pulse; while prompt electrical defibrillation is indicated when ventricular fibrillation or pulseless ventricular tachycardia is noted.

12. (E). For a fullterm baby without CNS depression (fighting ventilator), you need to use a combination of sedative (dexmedetomidine; lorazepam or midazolam) and analgesic (morphine or fentanyl). In babies with CNS depression no need for sedation.

13. (C). ARDS and ALI are a non-cardiogenic pulmonary edema which can be triggered by a variety of insults, including sepsis, pneumonia, shock, burns, or traumatic injury, all resulting in inflammation and increased vascular permeability leading to pulmonary edema. For definition of ALI you need to have the following four clinical features: acute onset, bilateral pulmonary edema, no clinical evidence of elevated left atrial pressure (no cardiac cause), and a ratio of Pao2 to Fio2 ≤ 300 mm Hg regardless of the level of positive end-expiratory pressure (PEEP). ARDS is a subset of ALI with more severe hypoxemia (Pao2/Fio2 of ≤ 200 mm Hg). In the above scenario the diagnosis of ALI is base on triggering factor which is the urosepsis; evidence of pulmonary edema (clinically and radiologically) and Pao2/Fio2 ratio is equal to 217 mmhg.
14. **(A).** For such a scenario, the use of noninvasive ventilation methods such as simple mask, heated humidified high-flow nasal cannula, continuous positive airway pressure, biphasic positive airway pressure, or negative pressure ventilation is usually sufficient. If the patient having hypercarbia with its symptomatology (respiratory fatigue or somnolence); invasive ventilation may be needed (option B, C, D). Option E is indicated if the patient developed apnea.

15. **(B).**

16. **(B).** Active immunization to Pneumococcus plus other measures mentioned in the question options participate in decreasing the incidence of respiratory failure.

17. **(D).** Dissociative shock refers to conditions in which tissue perfusion is normal, but cells are unable to use oxygen because the hemoglobin has an abnormal affinity for oxygen, preventing its release to the tissues, it is also seen in carbon monoxide poisoning.

18. **(E).**

19. **(B).**

20. **(B).** Following initial stabilization (including glucose administration if hypoglycemia is present); all patients with shock may benefit from determination of a baseline arterial blood gas and blood lactate level to assess the impairment of tissue oxygenation. Echocardiography may benefit in cardiogenic and obstructive shock; s. electrolytes in hypovolemic shock, and cultures in distributive shock.

21. **(C).** Measurement of mixed venous oxygen saturation aids in the assessment of the adequacy of oxygen delivery. In contrast to other forms of shock, patients with sepsis often have high mixed venous saturation values because of impairment of mitochondrial function and inability of tissues to extract oxygen.

22. **(E).** It also may include white blood count greater than 12,000 cells/mm³; less than 4000 cells/mm³, or greater than 10% immature forms.

23. **(C).** This baby suffering from septic (warm) shock, in those patients the pulse volume is usually good but the signs of SIRS (systemic inflammatory response syndrome) is evident. After ABC resuscitation and O2 therapy for respiratory support (acute lung injury), the choice is to use crystalloid as volume expanders. Most acutely ill children with signs of shock may safely receive, and usually benefit greatly from, a 20-mL/kg bolus of an isotonic crystalloid over 5 to 15 minutes. This dose may be repeated until a response is noted. Care must be exercised in treating cardiogenic shock with volume expansion because the ventricular filling pressures may rise without improvement of the cardiac performance. Carefully monitoring cardiac output or central venous pressure guides safe volume replacement. If no response; 2nd step is to start inotropes and the choice will be according to the patient status.
24.(D). Fifty two % of deaths above 1 yr of life caused by motor vehicle crashes. Most remaining unintentional injury-related deaths were the result of drowning (15%), poisoning (9%), burns (5%), and suffocation (4%).

25.(B). Primary strategies (preventing the event) as education, secondary strategies (minimizing the severity of injury) as wearing helmet, and tertiary strategies (minimizing long term impact) as hospital care and treatment.

26.(D). The most successful approaches to preventing injury have combined strategies (education, environmental changes, and engineering changes focused on the host, agent, and environment in all three time phases). Automatic strategies require no action on the part of the population and often change the environment (speed bumps) or involve engineering (child-resistant pill bottles, air bags).

27.(C). Children who begin breathing spontaneously and awaken before arrival to an emergency department and have significant episode, these children still require careful observation for pulmonary complications over the subsequent 6 to 12 hours.

28.(E). Coma for more than 72 hours in addition to development of fixed and dilated pupils is also considered as poor prognostic markers.

29.(A). The most common agents ingested by young children include cosmetics, personal care products, analgesics (like acetaminophen and ibuprofen), and cleaning solutions (like soap and bleach). Aspirin use is limited nowadays.

30.(A). Fatal childhood poisonings are commonly caused by analgesics, antihistamines, sedative/hypnotics, fumes/gases and, vapors.

31.(E). A poisoned child can exhibit any one of six basic clinical patterns: coma, direct toxicity, metabolic acidosis, heart rhythm aberrations, gastrointestinal symptoms, and seizures.

32.(C).

33.(D). Ingested button batteries also may produce a caustic mucosal injury. Batteries that remain in the esophagus may cause esophageal burns and erosion and should be removed with an endoscope.

34.(D). The MNEMONIC of (MUDPILES) usually refers to Methanol, metformin*, Uremia*, Diabetic ketoacidosis*, Paraldehyde, phenformin*, Isoniazid, iron, Lactic acidosis (cyanide, carbon monoxide), Ethanol, ethylene glycol*, Salicylates*, starvation, seizures; all causing high anion gap metabolic acidosis. Those with asterisk refer to hyperosmolar condition.

35.(E). Usually causes hyperglycemia.

36.(D). Routine screening tests include; specific toxin-drug assays; measurement of arterial blood gases and electrolytes, osmoles, and glucose; and calculation of the anion or osmolar gap. A full 12-lead electrocardiogram should be part of the initial evaluation in all patients suspected of ingesting toxic substances. Urine screens for
drugs of abuse or to confirm suspected ingestion of medications in the home may be revealing.

37. (D). If the level of consciousness is depressed, and a toxic substance is suspected, the management should be directed toward maintaining the airway, establishing effective breathing, and supporting the circulation, all other options in questions (except D), are considered as important supportive measures.

38. (C). Option A had been panned; gastric lavage is not routinely used and its efficacy is not clear. Option D, has no role in the management of the poisoned patient. Whole-bowel irrigation using polyethylene glycol as a nonabsorbable cathartic may be effective for toxic ingestion of sustained-release or enteric-coated drugs. Single-dose activated charcoal decreases drug absorption when used within 1 hour of ingestion; however, it has not been shown to improve outcome. Thus, it should be used selectively in the management of a poisoned patient.

39. (D). Charcoal is ineffective against caustic or corrosive agents, hydrocarbons, heavy metals (arsenic, lead, mercury, iron, and lithium), glycols, and water-insoluble compounds.

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

41. (A). And also in salicylate.

42. (E). Flumazenil for benzodiazepine; glucagon for β-blocking agents; sodium bicarbonate for cyclic antidepressants; deferoxamine for iron and oxygen for carbon monoxide.

43. (E). Atropine is the physiologic antidote by blocking acetylcholine while pralidoxime is the specific antidote by disrupting phosphotriesterase bond.

44. (E). Also methotrexate and cyclosporines requires therapeutic monitoring for drug toxicity.

45. (B). Ketamine has numerous side effects (Dissociative reactions, tachycardia, hypertension, increased bronchial secretions, emergent delirium, hallucinations; increases intracranial pressure) while dexmedetomidine is the least one (may cause bradycardia).

46. (B).
1. All these facts are true about human genome EXCEPT
   A. genetic material contain 3.1 billion nitrogen bases
   B. there is about 21000 genes
   C. active DNA constitute about 60% of the whole DNA
   D. junk DNA does not involve in protein formation
   E. gene function as template for mRNA which change to protein

2. Genetic diseases caused by change or (mutation) in DNA sequence.
   The frame shift mutation refers to
   A. change in a single DNA base
   B. change in amino acid of a particular protein
   C. increase in chromosomal number
   D. loss or addition of one or more DNA bases
   E. structural chromosomal defect

3. All the following are true in autosomal dominant disorder EXCEPT
   A. sex chromosomes are spared
   B. single copy of affect gene can cause the disease
   C. 50% chance of passing affected gene to next generation
   D. male-to-male transmission occurs
   E. male: female ratio usually 2:1

4. The following are examples of autosomal dominant disorders EXCEPT
   A. neurofibromatosis II
   B. Huntington disease
   C. Marfan syndrome
   D. hereditary angioneurotic edema
   E. Friedreich ataxia

5. Parents to a 2-year-old boy with achondroplasia, father is 45 years and mother is 35 years old and no one of them manifest the disease.
   Of the following, the MOST likely explanation is
   A. incomplete penetrance of one parent
B. variable expressivity of affected boy  
C. spontaneous mutation of affected boy  
D. advanced maternal age  
E. double homozygous

6. A 10-month-old baby with achondroplasia; developing recurrent apneas.  
Of the following, the **MOST** likely cause is  
   A. dental malocclusion  
   B. narrowing of the foramen magnum  
   C. hydrocephalus  
   D. obstructive apnea  
   E. lumbar lordosis

7. Neurofibromatosis Type 1 regarded as one of the most common AD disorders  
   but many affected individuals have features so mild that they are never diagnosed.  
   Of the following, the **MOST** likely cause of such phenomena is  
   A. incomplete penetrance  
   B. variable expressivity  
   C. uniparental disomy  
   D. missense mutation  
   E. double homozygous

8. Clinical symptoms in Marfan Syndrome (MFS) mostly involve three systems:  
   cardiac, ophthalmologic, and skeletal. The treatment that is slowing the aortic  
   dilatation and may prevent aortic dissection is  
   A. losartan  
   B. atenolol  
   C. enalapril  
   D. lisinopril  
   E. spironolactone

9. Chromosome analysis should be ordered for children with  
   A. metabolic disorders  
   B. mental retardation  
   C. recalcitrant epilepsy  
   D. conotruncal anomalies  
   E. cleft palate

10. One of the following is an example of an x-linked recessive disorder
A. cystic fibrosis (CF)
B. congenital adrenal hyperplasia
C. Gaucher disease
D. adrenoleukodystrophy
E. sickle cell disease

11. Of the following, the MOST common type of X-linked inheritance is
   A. Rett syndrome
   B. color blindness
   C. adrenoleukodystrophy
   D. hemophilia A
   E. fragile X syndrome

12. All the following are the rules of X-Linked recessive Inheritance EXCEPT
   A. trait is higher in males than in females
   B. each son of a carrier female has a one in two chance of being affected
   C. trait is transmitted from affected males to half of their daughters
   D. father to son transmission is never occur
   E. carrier females may show mild expression of the gene

13. Which of the following is an X-linked dominant inheritant disease?
   A. Duchenne muscular dystrophy
   B. Gaucher disease
   C. Angelman syndrome
   D. cat eye syndrome
   E. Rett syndrome

14. A 2-year-old girl with history of delayed speech, lonely play, and special hand movement (hand washing posture), the parents are healthy relative couple.
   The genetic cause of this disorder is MOST likely due to
   A. carrier mother
   B. carrier father
   C. new mutation
   D. chromosomal disorder
   E. X-linked recessive disorder

15. Polygenic inheritance (multifactorial) disorders result from the interplay of genetic and environmental factors.
   All the following considered as polygenic inheritance EXCEPT
A. cleft lip and palate
B. spina bifida
C. childhood asthma
D. atherosclerosis
E. hairy ear

16. An affected mother with hypertrophic pyloric stenosis (HPS), became pregnant. She came to you to explain the possibility of future affection of her kids. Of the following, the **MOST** likely true explanation is
   A. same like general population (5 times more possible in sons)
   B. daughter is more likely to be affected with 7-10% possibility
   C. sons are likely to be affected with about 30% and daughter 1-2%
   D. both sons and daughter are equally affected by 20% possibility
   E. sons are more by about 20% and daughter by 7%

17. A family consulted you; they have a long history of illnesses in their progeny and ancestors, they have 3 kids; 2 boys and one girl, the latter is complaining from episodic vomiting, seizures, and recurrent attacks of limb weaknesses. The older adolescent boy is wheel chaired due to severe stroke and the young boy is doing well. The maternal grandmother has diabetes and she is visiting the ophthalmologist for treatment of squint; the mother is on treatment for heart failure and having some hearing problem. Of the following, the **MOST** helpful study to confirm diagnosis is
   A. X-chromosome
   B. mitochondrial DNA
   C. chromosomal karyotype
   D. FISH DNA of chromosome no. 15
   E. PCR of nuclear DNA

18. The **MOST** common genetic defect of Prader-Willi syndrome (PWS) is
   A. deletion of maternal chromosome 15 (15q11)
   B. deletion of paternal chromosome 15 (15q11)
   C. paternal uniparental disomy of chromosome 15
   D. maternal uniparental disomy of chromosome 15
   E. isochromosome

19. All the following are features of Prader-Willi syndrome **EXCEPT**
   A. postnatal growth delay
   B. prenatal onset hypotonia
   C. infantile obesity
20. Disorders caused by expansion of trinucleotide repeats include all the following EXCEPT
   A. Fragile X syndrome
   B. ataxia telangiectasia
   C. Huntington disease
   D. myotonic dystrophy
   E. Friedreich ataxia

21. Fragile X syndrome (FRAX) is the most common cause of inherited intellectual disability. The individual who manifests the disease may have the number of trinucleotide repeat of CGG in the region Xq 27 of
   A. 25 to 35
   B. 50 to 75
   C. 100 to 125
   D. 175
   E. 225

22. Teratogenic agents are chemical, physical, or biologic agents that have the potential to damage embryonic tissue and result in congenital malformations. The rate of birth defects caused by teratogens is about
   A. 1%
   B. 6%
   C. 20%
   D. 50%
   E. 75%

23. Premarital screening for variable genetic disorders is mainly depending on ethnic group from which the couple is originated. People whose ancestors originated in the Mediterranean basin may be screened for
   A. Gaucher syndrome
   B. cystic fibrosis
   C. familial dysautonomia
   D. thalassemia
   E. sickle cell anemia

24. Of the following, the first trimester non-invasive screening test with good detection rate for aneuploidy is
A. ultrasonic detection of nuchal translucency
B. amniocentesis
C. chorionic villous sampling
D. quad screen
E. maternal testing for PAPP-A (pregnancy associated plasma protein)

25. A 24-year-old pregnant lady in 2nd trimester comes to your clinic worried about her exposure to CXR done for suspicion of pneumonia last week; she is highly concerned about possibility of congenital anomalies.
   Of the following, the **BEST** advice to her is that
   A. she should go for medical abortion
   B. she should suit the ordering doctor
   C. there is a considerable risk of microcephaly, and skeletal malformations
   D. there is weak possibility and need to followed by serial U/S
   E. the dose of radiation is negligible and unlikely to cause any effect

26. The growing promising technique that is still in the research stage and being piloted in high-risk pregnancies for prenatal diagnosis is
   A. screening anatomy sonogram at 18 weeks’ gestation
   B. fetal sonogram at first trimester for detection of nuchal translucency
   C. quad test
   D. cell-free fetal DNA in maternal blood
   E. free β-HCG and PAPP-A (pregnancy associated plasma protein)

27. Consanguinity may increase the chance that a child will be born with a rare autosomal recessive (AR) condition. The risk of first cousins producing a child with an AR disorder is
   A. 1 in 16
   B. 1 in 32
   C. 1 in 64
   D. 1 in 128
   E. similar to general population

28. Of the following, the study that is **MOST** useful in diagnosis of an aneuploidy is
   A. chromosomal karyotype
   B. fluorescent in situ hybridization
   C. microarray comparative genomic hybridization
   D. direct DNA analysis
   E. phenotypic study
29. The **MOST** useful way for confirmation of genetic defect in velocardiofacial (DiGeorge) syndrome is
   A. chromosomal karyotype
   B. fluorescent in situ hybridization
   C. microarray comparative genomic hybridization
   D. direct DNA analysis
   E. phenotypic study

30. A young lady had aborted recently, part of consoling, you told her that there is possibility of chromosomal anomalies in an aborted fetus, that is actually true and it is equal to be about
   A. 99%
   B. 75%
   C. 50%
   D. 30%
   E. 1%

31. A medical student is asking about the meaning of balanced translocation in parents who have baby with Down syndrome.
   Of the following, the **TRUE** statement is
   A. the actual genetic material is equal to 45 chromosomes
   B. the chromosomes are more in one of the parents
   C. affected parent is subclinical case of Down syndrome
   D. percentage of being Down is more if father is affected
   E. the lost genetic material is ribosomal DNA

32. Children with Down syndrome are most likely diagnosed clinically in the neonatal period.
   All the following are characteristic facial features **EXCEPT**
   A. large protruding tongue
   B. flattened occiput
   C. hypoplastic midface
   D. flattened nasal bridge
   E. down slanting of palpebral fissures

33. Of the following, the test /study that is required to be done periodically in persons with Down syndrome (DS) is
   A. visual assessment
   B. auditory testing
C. echocardiography
D. thyroid function test
E. EEG study

34. A 15-day-old baby with Down syndrome, during routine blood testing found to have high leukocyte count (34,000), the baby otherwise is asymptomatic. Of the following, the **MOST** appropriate advice/action is
   A. to refer the baby to a hematologist as it may be the prodrome of leukemia
   B. collect blood culture and treat the baby with empiric antibiotic therapy
   C. do septic screen and wait for the results
   D. reassure the parents and advice for follow-up
   E. proceed for bone marrow examination

35. A young parents with Down syndrome baby with Robertsonian translocation. Both did a chromosomal karyotype study. Of the following the **MOST** likely result is
   A. father with balanced translocation
   B. mother with balanced translocation
   C. both are normal
   D. both with balanced translocation
   E. mother with robertsonian translocation

36. All the following are recognized associations in infant with Down syndrome **EXCEPT**
   A. polycythemia
   B. leukemoid reaction
   C. neonatal jaundice
   D. cataract
   E. acute lymphoblastic leukemia

37. A fullterm baby with multiple congenital anomalies, his blood film reported nuclear projections in neutrophils. Of the following the **MOST** likely diagnosis is
   A. Down syndrome
   B. Williams syndrome
   C. Cat eye syndrome
   D. Edward syndrome
   E. Patau syndrome
38. All the following are characteristic features of Turner syndrome **EXCEPT**
   A. mildly affected IQ
   B. low-set ears
   C. triangular face
   D. webbing of the neck
   E. puffiness of the hands

39. You are evaluating a neonate with multiple dysmorphic features, you find him small for gestational age, microcephalic, single nostril, postaxial polydactyly and rocker-bottom feet.
   Of the following the **MOST** likely diagnosis is
   A. Patau syndrome
   B. Edward syndrome
   C. Turner syndrome
   D. Cri du chat syndrome
   E. Angelman syndrome

40. You are evaluating a small for gestational age neonate; you find the following features: hypertonia, prominent occiput, short sternum, and clenching of fists.
   A. Down syndrome
   B. Patau syndrome
   C. Edward syndrome
   D. Turner syndrome
   E. Cri du chat syndrome

41. Of the following, the one which carries the higher rate of congenital heart disease is
   A. Turner syndrome
   B. Williams syndrome
   C. Down syndrome
   D. Cri du Chat syndrome
   E. Klinefelter syndrome

42. All the following are characteristic features of Williams syndrome **EXCEPT**
   A. neonatal hypercalcemia
   B. elfin facies
   C. blue irides
   D. normal IQ
   E. striking personality
43. You are evaluating a 2-month-old baby boy with recurrent chest infections, cleft palate, right sided aortic arch, and absent thymic shadow on CXR. He could be nominated as having one of the following syndromes EXCEPT
   A. velocardiofacial syndrome
   B. conotruncal anomaly face syndrome
   C. Shprintzen syndrome
   D. DiGeorge syndrome
   E. Pierre Robin syndrome
1.(C). Active DNA constitutes only 2% of whole DNA. The junk DNA is not involve in protein formation, but most likely serves some regulatory function.

2.(D). Genetic disease leads to abnormal protein formation or stopping forming of a particular protein which usually results from a point mutation of single DNA base. When that point mutation changes a codon and the resulting amino acid that goes into the protein is referred to as a missense mutation. When the point mutation changes the codon to a “stop” codon so that transcription stops prematurely is called nonsense mutation; while frameshift mutation often stems from the loss or addition of one or more DNA bases; this causes a shift in how the DNA is transcribed and generally leads to premature stop codons.

3.(E). No sex preference.

4.(E). AR.

5.(C). Approximately 80% of patients with achondroplasia have a spontaneous mutation and it is known in some cases associated with advanced paternal age (>35 years of age).

6.(B). Dental malocclusion, obstructive apnea, and hearing loss due to middle ear dysfunction are common in later childhood while Hydrocephalus and central apnea may occur because of narrowing of the foramen magnum and compression of the brainstem and may present a life-threatening complication in infancy.

7.(B). The penetrance of NF1 is 100% (the disease is present whenever the mutant gene is present), while the expression is extremely variable (the disease can present with mild to severe symptoms), both represent features of AD disorders.

8.(A). Losartan, an angiotensin II receptor antagonist that lowers levels of TGF-β and may prevent aneurysms in patients with MFS.

9.(B). Chromosome analysis should be ordered for children with multiple congenital anomalies (as in major trisomies), the involvement of one major organ system and the presence of multiple dysmorphic features (like conotruncal anomalies with another dysmorphic features as cleft palate but alone it is not an indication), or the presence of mental retardation.

10.(D). X-linked recessive.

11.(B). Is the most common of X-linked disease with incidence of 1:12; males, Rett syndrome 1:10,000–1:15,000; girls, Hemophilia A 1:5000–1:10,000; males, Fragile X syndrome 1:4000; males, while adrenoleukodystrophy is uncommon.
12.(C). Trait is transmitted from affected males to all of their daughters. Each son of a carrier female has a one in two chance of being affected and trait is passed from carrier females, who may show mild expression of the gene, to half of their sons, who are more severely affected.

13.(E). Duchenne is X-linked recessive, Gaucher is AR, Angelman is uniparental disomy, cat eye is a chromosome duplication syndrome.

14.(C). In Rett syndrome, females are normal at birth, but later in the first year of life develop microcephaly and developmental regression and plateau. About 50% of patients develop seizures. Girls often are diagnosed with autism and, by 2 years of age, adopt a handwashing posture that causes them to lose all purposeful hand movements. Cases represent new mutations or parental gonadal mosaicism.

15.(E). Hairy ears still of unknown type of inheritance, it is thought to be Y-linked disorder.

16.(E). The rule in HPS is that the inheritance is always more in the male side. In general population it is five times more likely to occur in males than in females. When a child with HPS is born, the recurrence risk in future progeny is 5% to 10% for males and 1.5% to 2% for females. In adulthood, the risk of an affected male having an affected child is markedly increased over the general population: 4% of sons and 1% of daughters of such men would be likely to be affected. Even more striking is the risk to children born to affected females: 17% to 20% of sons and 7% of daughters are affected.

17.(B). Human cells contain non-nuclear DNA; a single chromosome is present in each mitochondrion, and mutations within this DNA are associated with a group of diseases. Virtually all mitochondria are supplied by the oocyte, which means that mtDNA is maternally derived. The mtDNA mutation may be present in a few or many mitochondria. When the fertilized egg divides, mitochondria are distributed randomly. If an abundance of mutant mitochondria exists in tissue that has high energy requirements (brain, muscle, and liver), clinical symptoms occur. MELAS (mitochondrial encephalomyopathy with lactic acidosis and strokelike episodes) is an example of a mitochondrial disorder. Normal in early childhood, individuals affected with MELAS develop episodic vomiting, seizures, and recurrent cerebral insults that resemble strokes between 5 and 10 years of age. In families in which MELAS occurs, a range of symptoms is seen in first-degree relatives, including progressive external ophthalmoplegia, hearing loss, cardiomyopathy, and diabetes mellitus.

18.(B). 70% of individuals with PWS have a small deletion of paternal chromosome 15 (15q11) and in 20% have maternal UPD of chromosome 15. In Angleman syndrome the reverse is true.
19. (C). Early in life, affected infants are so hypotonic that they cannot consume enough calories to maintain their weight. After first year of life children develop a voracious appetite and obesity develops.
20. (B). It includes also spinocerebellar ataxias.
21. (E). Unaffected individuals who have no family history of FRAX have 0 to 45 CGG repeats (most have 25 to 35). In individuals with FRAX, the number of repeats is greater than 200; such people are said to have a full mutation. Between these two categories, a third group has 56 to 200 repeats; always phenotypically normal, these individuals are permutation carriers.
22. (B). Known teratogens include drugs (prescription and nonprescription); intrauterine infections (rubella); maternal diseases, such as diabetes mellitus; and environmental substances, such as alcohol and heavy metals. Knowledge of teratogenic agents and their effect on the developing fetus is important, because limiting exposure to these agents is an effective way to prevent birth defects.
23. (D). People of Ashkenazi Jewish background may choose to be screened for heterozygosity for a panel of AR disorders, including Tay-Sachs disease, Niemann-Pick disease, Bloom syndrome, Canavan disease, Gaucher syndrome, cystic fibrosis, Fanconi anemia, and familial dysautonomia. People of African-American ancestry may choose to be screened for sickle cell anemia. People whose ancestors originated in the Mediterranean basin may be screened for thalassemia.
24. (A). During the first trimester, measurement of a fluid collection of the posterior neck of the developing fetus is termed nuchal translucency. An increase in the nuchal translucency is a marker for chromosomal anomalies as well as genetic and structural abnormalities in the fetus. This association provided a noninvasive first trimester marker with detection rate for aneuploidy approached 70%. The quad screen is done in the second trimester. B & C options are both invasive and considerable risk of fetal loss. Option E along with free β-HCG and fetal nuchal translucency increases detection rate up to 90%.
25. (E). Exposure to radiation causes microcephaly, mental retardation and skeletal malformations, these effects were manifested if the dose of radiation near 25 rad, while the dose from routine radiologic diagnostic examinations is in the millirad range (CXR=25 millirad).
26. (D). All are true and established screening prenatal tests but the evolving non-invasive (need only maternal blood sample) which can check fetal genome is D.
27. (C). In some cultures, it is common for relatives to mate. This relatedness or consanguinity does not increase the likelihood of offspring having any particular single genetic disorder, but it may increase the chance that a child will be born with a rare autosomal recessive (AR) condition, as the mutated gene segregates through that family. Generally, the closer the relation between the partners, the greater the
chance that the couple shares one or more mutated genes in common; increasing the risk that offspring will have an AR disorder.

28.(A). Aneuploidy means problems with the chromosomal numbers, the basic and easiest and most useful test is chromosomal karyotype by using a cells which is able to divide as blood lymphocyte.

29.(B). Fluorescent in situ hybridization (FISH) allows the identification of the presence or absence of a specific region of DNA. This technique is useful in Prader-Willi syndrome and Angelman syndrome, in which a deletion in a segment of 15q11.2 occurs, and in velocardiofacial (DiGeorge) syndrome, which is associated with a deletion of 22q11.2.

30.(C). Fifty percent of spontaneous abortuses have chromosomal abnormalities, the most common being 45,X (TS); an estimated 99% of 45,X fetuses are spontaneously aborted. The fetal loss rate for Down syndrome, the most viable of the autosomal aneuploidies, approaches 80%.

31.(E). Simply in balanced translocation (the parental side) the genetic material is normal in amount but abnormal in construction when the long arms (q) of two acrocentric chromosomes (numbers 13, 14, 15, 21, or 22) fuse at the centromeres, and the short arms (p), containing copies of ribosomal RNA, are lost. By this way no extra genetic material in parents. In robertsonian translocation (after conception), the chromosome which contains the genetic material of 2 chromosomes in the parent with balanced translocation, will be transmitted as one chromosome which leads to increase genetic material in offspring.

32.(E). Up slanting palpebral fissures, epicanthal folds and large protruding tongue are additional facial features. Down slanting palpebral fissures is seen in Noonan syndrome.

33.(D). Four percent to 18% of infants with DS are found to have congenital hypothyroidism, which is identified as part of the newborn screening program. Acquired hypothyroidism is a more common problem. Thyroid function testing must be monitored periodically during the child’s life.

34.(D). Some infants with DS show a leukemoid reaction, with markedly elevated white blood cell counts. Although this resembles congenital leukemia, it is a self-limited condition, resolving on its own over the first month of life.

35.(C). If the child has a robertsonian translocation, chromosomal analysis of both parents must be performed. In approximately 65% of cases, the translocation is found to have arisen de novo (i.e., spontaneously, with both parents having normal karyotypes), and in 35% of cases, one parent has a balanced translocation. The robertsonian translocation occurs in about 4.5% of cases of Down syndrome. The most common robertsonian translocation leading to DS involves chromosomes 14 and 21; standard nomenclature is 46,XX,t(14q21q) or 46,XY,t(14q21q).
36. (E). In children with DS younger than 2 years of age, the type is generally acute megakaryoblastic leukemia; in individuals older than 3 years of age, the types of leukemia are similar to those of other children, with acute lymphoblastic leukemia being the predominant type.
37. (E).
39. (A). In Patau syndrome midline facial defects such as cyclopia (single orbit), cebocephaly (single nostril), and cleft lip and palate are common, as are midline central nervous system anomalies, such as alobar holoprosencephaly.
40. (C). In Edward syndrome, you find also rocker-bottom feet, hypoplastic nails, and characteristic clenching of fists—the second and fifth digits overlap the third and fourth digits.
41. (B). In Turner syndrome about 50%, Williams syndrome 80% (supravalvar aortic-valve and pulmonic-valve stenosis and peripheral pulmonic stenosis being the most common anomalies), Down syndrome 50%, Low rate in Cri du Chat syndrome (around 15-20%) and as in general population with Klinefelter syndrome.
42. (D). Williams syndrome resulted from small deletion of chromosome 7q. Moderate intellectual disability (average IQ in the 50 to 60 range) is common, but developmental testing reveals strength in personal social skills and deficiencies in cognitive areas. Individuals with Williams syndrome often have a striking personality (cocktail party personality). However, approximately 10% of children with Williams syndrome have features of autism spectrum disorder. Patients occasionally have unusual musical ability (about 20% have absolute or perfect pitch).
43. (E). All except E represent a continuum of findings, virtually due to the chromosome 22q deletion. Common features include clefting of the palate with velopharyngeal insufficiency; conotruncal cardiac defects (including truncus arteriosus, ventriculoseptal defect, tetralogy of Fallot, and right sided aortic arch); and a characteristic facial appearance, including a prominent nose and a broad nasal root. Speech and language difficulties are common, as is mild intellectual impairment. About 70% have immunodeficiencies, largely related to T-cell dysfunction. A wide spectrum of psychiatric disturbances, including schizophrenia and bipolar disorder, has been seen in more than 33% of affected adults.
ZUHAIR ALMUSAWI

1. Metabolic disorders should be considered in all neonates presenting with the following EXCEPT
   A. lethargy
   B. poor tone
   C. poor feeding
   D. hyperthermia
   E. seizures

2. Any organ or system can be injured by toxic accumulation of any of the metabolites involved in inborn errors. Of the following, the metabolic disease causing hepatic failure is
   A. glycogen storage disease I
   B. glycogen storage disease III
   C. glycogen storage disease IV
   D. mucopolysaccharidosis I
   E. mucopolysaccharidosis II

3. Clinical laboratory testing can define the metabolic derangement. Vacuolated lymphocytes and metachromatic granules are seen in
   A. lysosomal disorders
   B. organic acidurias
   C. nonketotic hyperglycemia
   D. pyruvate carboxylase deficiency
   E. isovaleric acidemia

4. The prognosis of glycogen storage diseases is good in
   A. type 1a: von Gierke
   B. type 1b
   C. type II: Pompe
   D. type IV: Andersen
   E. type V: McArdle
5. A 5-month-old infant presented with symmetrical profound muscle weakness, tachypnea, tachycardia, and poor feeding. His chest x-ray showed cardiomegaly and his ECG revealed short PR interval.

Of the following, the **MOST** likely glycogen storage disease is
- A. type 1a: von Gierke
- B. type 1b
- C. type II: Pompe
- D. type IV: Andersen
- E. type V: McArdle

6. Galactosemia is an autosomal recessive disease characterized by all the following **EXCEPT**
- A. hyperbilirubinemia
- B. disorders of coagulation
- C. hyperglycemia
- D. glycosuria
- E. aminoaciduria

7. Renal tubular dysfunction in galactosemia may be evidenced by a
- A. normal–anion-gap hypochloremic metabolic acidosis
- B. high–anion-gap hyperchloremic metabolic acidosis
- C. normal–anion-gap hyperchloremic metabolic acidosis
- D. high–anion-gap hypochloremic metabolic acidosis
- E. normal–anion-gap hypokalemic metabolic acidosis

8. Individuals homozygous for galactokinase deficiency usually develop
- A. severe neonatal Escherichia coli sepsis
- B. cataracts after the neonatal period
- C. learning disorders
- D. disordered renal tubular function
- E. hyperbilirubinemia

9. Phenylketonuria (PKU) is characterized by all the following **EXCEPT**
- A. autosomal recessive inheritance
- B. primarily affects the brain
- C. occurs in 1 in 1,000 persons
- D. affected infants are normal at birth
- E. untreated infants develops severe mental retardation in infancy
10. Transient tyrosinemia of the newborn responds to treatment with
   A. ascorbic acid
   B. vitamin B6
   C. vitamin B12
   D. special medical formula
   E. vitamin E

11. A constant threat of homocystinuria is
   A. dislocated ocular lenses
   B. scoliosis
   C. pectus excavatum
   D. mental retardation
   E. arterial thromboses

12. A 3-week-old neonate presented with poor feeding, vomiting, and tachypnea with profound depression of the central nervous system, associated with alternating hypotonia and hypertonia, opisthotonos, and seizures. Laboratory manifestations reveal hypoglycemia and metabolic acidosis with high anion gap.
   Of the following, the **MOST** likely diagnosis is
   A. tyrosinemia
   B. phenylketonuria
   C. homocystinuria
   D. maple syrup urine disease
   E. arginosuccinate lyase (ASL) deficiency

13. During episodes of symptomatic hyperammonemia, all the following treatment options can be used **EXCEPT**
   A. intravenous glucose
   B. sodium benzoate
   C. hemodialysis
   D. hemofiltration
   E. peritoneal dialysis

14. Cystinuria is a disorder of renal tubular transport of the following amino acids **EXCEPT**
   A. cystine
   B. lysine
   C. tryptophan
D. arginine  
E. ornithine

15. A first cousin parents brought their 3-week-old neonate with fever, vomiting, and diarrhea, on examination he is tachypnic and lethargic, lab. Investigations revealed ketoacidosis, hyperglycinemia, neutropenia, thrombocytopenia, hyperammonemia and hypoglycemia.  
Of the following, the **MOST** likely diagnosis is  
A. propionic acidemia  
B. glutaric acidemia  
C. biotinidase deficiency  
D. holocarboxylase deficiency  
E. ornithine carbamoyltransferase (OTC) deficiency

16. Sweaty feet odor is seen in untreated infants with  
A. isovaleric acidemia  
B. propionic acidemia  
C. glutaric acidemia  
D. biotinidase deficiency  
E. holocarboxylase deficiency

17. Sudden infant death syndrome is reported in infants with  
A. very long chain acyl-CoA dehydrogenase (VLCAD)  
B. long-chain hydroxyacyl-CoA dehydrogenase (LCHAD)  
C. medium-chain acyl-CoA dehydrogenase (MCAD)  
D. short-chain acyl-CoA dehydrogenase (SCAD)  
E. hydroxymethylglutaryl-CoA lyase deficiency

18. A relative parents brought their underweight 6-month-old female infant who has high foreheads, flat orbital ridges, widely open fontanelles, hepatomegaly, hypotonia, and nystagmus; she had history of recurrent seizures.  
Of the following, the **MOST** likely diagnosis is  
A. carnitine deficiency  
B. Zellweger syndrome  
C. Refsum disease  
D. neonatal adrenoleukodystrophy  
E. malonic aciduria
19. Mucopolysaccharidoses (MPS) affect many solid organs, the cornea is clear but there are retinal degeneration and papilledema in
   A. MPS I (Hurler)
   B. MPS II (Hunter)
   C. MPS III (Sanfilippo)
   D. MPS IV (Morquiro)
   E. MPS VI (Maroteaux-Lamy)

20. Mucopolysaccharidoses (MPS) affect many solid organs, hepatosplenomegaly or hepatomegaly is usual finding, but the liver and spleen are normal in
   A. MPS I (Hurler)
   B. MPS II (Hunter)
   C. MPS III (Sanfilippo)
   D. MPS VI (Maroteaux-Lamy)
   E. MPS VII (Sly)

21. Mucopolysaccharidoses (MPS) affect many solid organs, the brain is spared in
   A. MPS I (Hurler)
   B. MPS II (Hunter)
   C. MPS III (Sanfilippo)
   D. MPS VI (Maroteaux-Lamy)
   E. MPS VII (Sly)

22. Lipidoses affect many solid organs, optic atrophy is seen in
   A. Glucosylceramide lipidosis (Gaucher 1)
   B. Glucosylceramide lipidosis 2 (Gaucher 2)
   C. Sphingomyelin lipidosis B (Niemann-Pick B)
   D. GM2 gangliosidosis (Tay-Sachs)
   E. Galactosylceramide lipidosis (Krabbe)

23. Lipidoses affect many solid organs, cherry-red spots is seen in
   A. Glucosylceramide lipidosis (Gaucher 1)
   B. Glucosylceramide lipidosis 2 (Gaucher 2)
   C. Sphingomyelin lipidosis B (Niemann-Pick B)
   D. GM2 gangliosidosis (Tay-Sachs)
   E. Galactosylceramide lipidosis (Krabbe)
24. Foam cell and sea-blue histiocytes in marrow are seen in
   A. Niemann-Pick C
   B. Glucosylceramide lipidosis (Gaucher 1)
   C. GM2 gangliosidosis (Tay-Sachs)
   D. Metachromatic leukodystrophy
   E. Fabry disease

25. Lipidoses are autosomal recessive disorders, while it is X-linked in
   A. Niemann-Pick C
   B. Glucosylceramide lipidosis (Gaucher 1)
   C. GM2 gangliosidosis (Tay-Sachs)
   D. Metachromatic leukodystrophy
   E. Fabry disease

26. Vertical ophthalmoplegia, dystonia, cataplexy, and seizures are seen in
   A. Niemann-Pick C
   B. Glucosylceramide lipidosis (Gaucher 1)
   C. GM2 gangliosidosis (Tay-Sachs)
   D. Metachromatic leukodystrophy
   E. Fabry disease

27. Mitochondrial disorders may affect as many as
   A. 1 in 25 people
   B. 1 in 250 people
   C. 1 in 2500 people
   D. 1 in 25000 people
   E. 1 in 250000 people
1. (D). Hypothermia.

2. (C). Hepatic failure is seen in the following metabolic diseases:
   - Citrin deficiency
   - Galactosemia
   - Hereditary fructose intolerance
   - Tyrosinemia type 1 (fumarylacetoacetate hydrolase deficiency)
   - GSD IV (slowly evolving)

3. (A). Organ-specific evaluation is indicated for specific symptoms (e.g., cranial magnetic resonance imaging for coma or seizures; echocardiography for cardiomyopathy; cerebrospinal fluid amino acids by column chromatography if nonketotic hyperglycemia is suspected). Thrombocytopenia and neutropenia are seen in organic acidurias; vacuolated lymphocytes and metachromatic granules are seen in lysosomal disorders.

4. (E).

5. (C).

6. (C). Galactosemia is an autosomal recessive disease caused by deficiency of galactose-1-phosphate uridylyltransferase. Clinical manifestations are most striking in a neonate who, when fed milk, generally exhibits evidence of liver failure (hyperbilirubinemia, disorders of coagulation, hypoglycemia), disordered renal tubular function (acidosis, glycosuria, aminoaciduria), and cataracts. The neonatal screening test must have a rapid turn around time because affected infants may die in the first week of life. Affected infants are at increased risk for severe neonatal Escherichia coli sepsis.

7. (C).

8. (B). Individuals homozygous for galactokinase deficiency usually develop cataracts after the neonatal period, whereas heterozygous individuals may be at risk for cataracts as adults.

9. (C). Phenylketonuria (PKU), an autosomal recessive disease, primarily affects the brain and occurs in 1 in 10,000 persons. Classic PKU is the result from a defect in the hydroxylation of phenylalanine to form tyrosine.

10. (A). Tyrosinemia is identified in neonatal screening programs using tandem mass spectrometry methods to detect elevated tyrosine and/or succinylacetone. Elevated tyrosine levels also occur as a nonspecific consequence of severe liver
disease or transient tyrosinemia of the newborn, which responds to ascorbic acid treatment.

11. (E). Major arterial or venous thromboses are a constant threat.

12. (D). Although MSUD does have intermittent-onset and late-onset forms, clinical manifestations of the classic form typically begin within 1 to 4 weeks of birth. Poor feeding, vomiting, and tachypnea commonly are noted, but the hallmark of the disease is profound depression of the central nervous system, associated with alternating hypotonia and hypertonia (extensor spasms), opisthotonos, and seizures. The urine has the odor of maple syrup.

13. (E). During episodes of symptomatic hyperammonemia, protein intake is reduced, and intravenous glucose is given in sufficient quantity to suppress catabolism of endogenous protein. Ammonia can be eliminated by use of the alternate pathway agents, sodium benzoate and sodium phenylacetate, which are excreted in the urine as conjugates of glycine and glutamine. Arginine, which is usually deficient, is supplied. When ammonia levels are very high (>1000 μM) or refractory to therapy, direct removal of ammonia using hemodialysis or hemofiltration, but not peritoneal dialysis, is required.

14. (C). Intestinal transport of tryptophan is impaired in Hartnup syndrome; pellagra-like symptoms result from this deficiency. Diagnosis is based on the amino acid pattern in urine. Treatment with tryptophan is successful.

15. (A). Propionic acidemia and methylmalonic acidemia are identified by neonatal screening with tandem mass spectrometry methods. The clinical manifestations of both of these disorders in the neonatal period consist of tachypnea, vomiting, lethargy, coma, intermittent ketoacidosis, hyperglycinemia, neutropenia, thrombocytopenia, hyperammonemia, and hypoglycemia. If these disorders are not identified by neonatal screening, intermittent episodes of metabolic acidosis occur. Crises occur during periods of catabolic stress, such as fever, vomiting, and diarrhea; they also may occur without an apparent precipitating event. During periods of neutropenia, the risk of serious bacterial infection is increased. Failure to thrive and impaired development are common.

16. (A). Isovaleric acidemia results from a block in the catabolism of leucine. Its clinical manifestations are similar to those of defects in the propionate pathway. The strong odor of isovaleric acid results in sweaty feet odor in untreated infants. Therapy involves restricting the intake of leucine, and providing glycine as an alternate pathway therapy that conjugates isovaleric acid and is then excreted in the urine.

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

18. (B). Zellweger syndrome, an autosomal recessive disease (1:100,000 births), is also called cerebrohepatorenal syndrome. Peroxisomes are virtually absent, as are normal peroxisomal functions, which include the oxidation of very long chain fatty acids. Affected infants have high foreheads, flat orbital ridges, widely open fontanelles, hepatomegaly, and hypotonia. Other anomalies are common. Failure to thrive, seizures, and nystagmus develop early, and death occurs within the first year.

19. (B).
20. (D).
21. (D).
22. (E).
23. (D).
24. (A).
25. (E).
26. (A).
27. (C).
1. Acute polyhydramnios is usually associated with
   A. cleft palate
   B. hydrops fetalis
   C. premature labor
   D. esophageal atresia
   E. meningomyelocele

2. A newly delivered newborn baby admitted to the neonatal care unit with respiratory distress; physical examination revealed clubfeet, characteristic compressed facies, low-set ears, and diminished chest wall size; abdominal ultrasound reveals bilateral renal agenesis; ultrasound during pregnancy show amniotic fluid index ≤2 cm.
   Of the following, the MOST likely cause of death in this condition is
   A. sepsis
   B. asphyxia
   C. heart failure
   D. renal insufficiency
   E. respiratory failure

3. Prolonged rupture of the membranes is associated with an increased risk of chorioamnionitis and preterm birth. The latent period is
   A. ≥12 hours
   B. ≥24 hours
   C. ≥36 hours
   D. ≥48 hours
   E. ≥72 hours

4. In multiple gestations, the birth order of twins affects morbidity by increasing the risk of the second-born twin for all the following EXCEPT
   A. anemia
   B. birth trauma
   C. birth asphyxia
   D. breech position
E. respiratory distress syndrome

5. Twin-to-twin transfusion syndrome suggested by
   A. weight differences of 20% and hemoglobin differences of 1 g/dL
   B. weight differences of 20% and hemoglobin differences of 3 g/dL
   C. weight differences of 20% and hemoglobin differences of 5 g/dL
   D. weight differences of 20% and hemoglobin differences of 7 g/dL
   E. weight differences of 20% and hemoglobin differences of 9 g/dL

6. Perinatal mortality rate is expressed as number of deaths per 1000 live births and it’s refers to fetal deaths occurring from
   A. the 20th week of gestation until the 7th day after birth
   B. the 20th week of gestation until the 28th day after birth
   C. the 32th week of gestation until the 7th day after birth
   D. the 32th week of gestation until the 28th day after birth
   E. the 36th week of gestation until the 7th day after birth

7. Very low birth weight (VLBW) infants, defined as infants having birth weights of less than
   A. 750 gm
   B. 100 gm
   C. 1500 gm
   D. 2000 gm
   E. 2500 gm

8. The major cause of neonatal mortality in preterm infant is
   A. birth trauma
   B. birth asphyxia
   C. respiratory distress syndrome
   D. meconium aspiration pneumonia
   E. persistent pulmonary hypertension

9. The major cause of neonatal mortality in fullterm infant is
   A. birth asphyxia
   B. chronic lung disease
   C. necrotizing enterocolitis
   D. intraventricular hemorrhage
   E. respiratory distress syndrome
10. All the following are problems of small for gestational age infants **EXCEPT**
   A. anemia
   B. hypothermia
   C. hypoglycemia
   D. perinatal asphyxia
   E. pulmonary hemorrhage

11. In neonatal examination, the pupillary response to light is present at gestational age of
   A. 28 weeks
   B. 30 weeks
   C. 32 weeks
   D. 34 weeks
   E. 36 weeks

12. The palmar grasp is gone by 4 months of age and present by gestational age of
   A. 28 weeks
   B. 30 weeks
   C. 32 weeks
   D. 34 weeks
   E. 36 weeks

13. The rooting reflex is present by gestational age of
   A. 30 weeks
   B. 32 weeks
   C. 34 weeks
   D. 36 weeks
   E. 38 weeks

14. A bifid uvula suggests a
   A. laryngomalacia
   B. submucosal cleft
   C. chromosomal defect
   D. tracheoesophageal fistula
   E. neurological abnormalities

15. You are on call at the delivery room, a newly delivered newborn presented to you with pallor, irregular respiration, and bradycardia; after initial resuscitation (warming, drying, and stimulation), heart rate still less than 100/min.
Of the following, the **NEXT** step in management is
A.  intravenous naloxone  
B.  intravenous epinephrine  
C.  endotracheal intubation  
D.  external cardiac massage  
E.  positive pressure ventilation with 100% O₂

16. In the delivery room and after prolonged labor you received a newborn baby with central and peripheral cyanosis, heart rate 90/min, weak cry, arms and legs well flexed, and withdrawal motion. The Apgar score is
A.  4  
B.  5  
C.  6  
D.  7  
E.  8

17. The Apgar score is a good method for assessing the need to resuscitate a newborn, all the following parameters are evaluated **EXCEPT**
A.  heart rate  
B.  respiration  
C.  temperature  
D.  color of body  
E.  reflex irritability

18. A 2-hour-old newborn admitted to neonatal care unit with respiratory distress, heart sounds heard mainly on the right side, and scaphoid abdomen; O₂ saturation at room air 68%; chest radiograph was ordered.
Of the following, the **BEST** way of O₂ administration is by
A.  CPAP  
B.  head box  
C.  face mask  
D.  nasal cannula  
E.  endotracheal intubation

19. Severe cold injury in an infant is manifested by all the following **EXCEPT**
A.  acidosis  
B.  bradycardia  
C.  hypoglycemia  
D.  blue skin color
20. The characteristic radiographic finding of bronchopulmonary dysplasia (BPD) is
   A. air bronchograms
   B. spongelike appearance
   C. fluid in the lung fissures
   D. reticulogranular pattern
   E. increased anteroposterior diameter

21. All the following are features of cephalhematoma **EXCEPT**
   A. soft swelling
   B. subperiosteal hemorrhage
   C. occasionally extend across suture lines
   D. association with underlying skull fracture
   E. disappearance within 2 weeks – 3 months

22. Neonatal hypocalcemia is considered when serum ionized calcium level is
   A. ≤3.5 mg/dL
   B. ≤4.5 mg/dL
   C. ≤5.5 mg/dL
   D. ≤6.5 mg/dL
   E. ≤7.5 mg/dL

23. Late neonatal hypocalcemia (after 3 days of life) is often the result of
   A. hypercalcitoninemia
   B. sodium bicarbonate therapy
   C. transient hypoparathyroidism
   D. phosphate release from cell necrosis
   E. high phosphate-containing milk ingestion

24. All the following matching regarding maternal diseases and their effect on fetus
   or neonate are true **EXCEPT**
   A. severe diabetes mellitus ----- macrosomia
   B. Graves disease --------- transient thyrotoxicosis
   C. antiphospholipid syndrome --------- thrombophilia
   D. systemic lupus erythematosus ----- congenital heart block
   E. idiopathic thrombocytopenic purpura ----- thrombocytopenia
25. The teratogenic effect of vitamin D is
   A. hypoplastic nails
   B. enamel hypoplasia
   C. cutis laxa syndrome
   D. facial and ear anomalies
   E. supravalvular aortic stenosis

26. Ebstein anomaly can be induced by teratogenic effect of
   A. lithium
   B. valproate
   C. streptomycin
   D. thalidomide
   E. radioactive iodine

27. In congenital infection, the organism that is usually identified by specific fetal production of antibodies (IgM or increasing titer of IgG) is
   A. CMV
   B. rubella
   C. gonorrhea
   D. parvovirus
   E. M. tuberculosis

28. A 7-month-old infant presented with recurrent seizures; on examination there are hydrocephalus and chorioretinitis; skull film revealed intracerebral calcifications.
   Of the following, the MOST likely diagnosis is
   A. syphilis
   B. HSV infection
   C. CMV infection
   D. congenital rubella
   E. congenital toxoplasmosis

29. A 3-month-old infant present with history of low birth weight, early-onset jaundice, and seizures; on examination there are hepatosplenomegaly and hydrocephalus; skull films reveal diffuse cortical calcifications.
   Of the following, the primary method of diagnosis is
   A. PCR
   B. culture
   C. brain CT
D. serology  
E. cerebrospinal fluid

30. Congenital toxoplasmosis occurs through vertical transmission of Toxoplasma gondii by transplacental transfer of the organism from the mother to the fetus. All the following statements are true EXCEPT
   A. treatment ordered for symptomatic and asymptomatic cases
   B. transmission rate varies directly with gestational age
   C. organism transfer occurs after an acute maternal infection
   D. fetal infection commonly occur after reactivation of disease
   E. the severity of fetal disease varies inversely with the gestational age

31. A 9-month-old infant presents with history of early-onset jaundice and growth retardation; on examination there are cataracts, hepatosplenomegaly, and purpuric skin lesions; CBC revealed thrombocytopenia; echo study showed peripheral pulmonary artery stenosis. Of the following, the MOST likely diagnosis is
   A. congenital syphilis
   B. HSV infection
   C. CMV infection
   D. congenital rubella
   E. congenital toxoplasmosis

32. The MOST common characteristic abnormality associated with congenital rubella is
   A. growth retardation
   B. early-onset jaundice
   C. blueberry muffin rash
   D. radiolucent bone disease
   E. cataract

33. Infants with congenital rubella should be isolated while in the hospital and kept away from susceptible pregnant women when sent home for about
   A. 1 yr
   B. 2 yr
   C. 3 yr
   D. 4 yr
   E. 5 yr
34. Recent congenital rubella infection is usually detected by measurement of
   A. rubella-specific IgA
   B. rubella-specific IgD
   C. rubella-specific IgG
   D. rubella-specific IgM
   E. rubella-specific IgE

35. A full-term infant presents with history of low birth weight, microcephaly, hepatosplenomegaly, and chorioretinitis; investigations reveals thrombocytopenia and abnormal liver enzymes; skull films revealed intracranial calcifications.
   Of the following, the **MOST** likely diagnosis is
   A. syphilis
   B. HSV infection
   C. CMV infection
   D. congenital rubella
   E. congenital toxoplasmosis

36. The **MOST** common congenital infection and the leading cause of sensorineural hearing loss is
   A. syphilis
   B. HSV infection
   C. CMV infection
   D. congenital rubella
   E. congenital toxoplasmosis

37. Congenital CMV infection proved when CMV detected within the first
   A. 3 wks after birth
   B. 6 wks after birth
   C. 9 wks after birth
   D. 12 wks after birth
   E. 15 wks after birth

38. Ganciclovir is antiviral agent used in severely symptomatic congenital CMV infection to decrease the progression of
   A. hearing loss
   B. chorioretinitis.
   C. hepatosplenomegaly
   D. blueberry muffin rash
   E. periventricular calcifications
39. All the following are features of congenital CMV infection **EXCEPT**
   A. deafness
   B. microcephaly
   C. chorioretinitis
   D. blueberry muffin rash
   E. diffuse cortical calcifications

40. Regarding herpes simplex virus (HSV) infection, all the following statements are true **EXCEPT**
   A. 70% to 85% of neonatal infections are caused by HSV-1
   B. acquired shortly before or during passage through the birth canal
   C. the risk to an infant born vaginally to a mother with a primary genital herpes is about 33% to 50%
   D. the risk to an infant born to a mother with a reactivated infection is less than 5%
   E. 75% of infants are born to mothers with no previous history or clinical findings consistent with HSV infection

41. Most infants with herpes simplex virus (HSV) infection are normal at birth, and symptoms of infection develop at
   A. 5 to 10 days of life
   B. 11 to 15 days of life
   C. 16 to 20 days of life
   D. 21 to 25 days of life
   E. 26 to 30 days of life

42. Infants who weigh less than 1500 g or who are born before 28 weeks’ gestational age should be screened for retinopathy of prematurity (ROP) when they are
   A. 2 wks of age or >32 wks’ corrected gestational age
   B. 4 wks of age or >34 wks’ corrected gestational age
   C. 8 wks of age or >36 wks’ corrected gestational age
   D. 12 wks of age or >38 wks’ corrected gestational age
   E. 16 wks of age or >40 wks’ corrected gestational age

43. The treatment of choice for neonatal herpes simplex virus (HSV) infections is
   A. observation
   B. vaccination
   C. acyclovir
D. IV immunoglobulin
E. ganciclovir

44. A 3-week-old infant presented with poor feeding, maculopapular rash over the palms and soles and around the mouth, and bloody nasal discharge; on examination there are hepatosplenomegaly and lymphadenopathy; CBC reveals hemolytic anemia and thrombocytopenia. Of the following, the **MOST** likely diagnosis is
   A. congenital syphilis
   B. HSV infection
   C. CMV infection
   D. congenital rubella
   E. congenital toxoplasmosis

45. The drug of choice for treatment of congenital syphilis is
   A. penicillin
   B. tetracycline
   C. erythromycin
   D. metronidazole
   E. cephalosporine

46. The late manifestation of congenital syphilis (may not become apparent until many years after birth) is
   A. snuffles
   B. osteochondritis
   C. hemolytic anemia
   D. interstitial keratitis
   E. hepatosplenomegaly

47. A 7-day-old infant, a product of normal vaginal delivery, presented with history of clear, watery discharge from both eyes that became purulent; on examination there is conjunctival hyperemia and chemosis. Of the following, the recommended treatment is
   A. oral erythromycin
   B. topical tetracycline
   C. topical silver nitrate
   D. topical erythromycin
   E. intramuscular ceftriaxone
48. A 4-week-old infant presents with history of conjunctivitis and repetitive cough; on examination temperature 37°C, respiratory rate 68/min, and rales on auscultation; chest radiograph show hyperinflation with diffuse infiltrates. Of the following, the treatment of choice is
   A. ampicillin
   B. cefotaxim
   C. tetracyclin
   D. vancomycin
   E. erythromycin

49. The treatment of neonatal chlamydial conjunctivitis is
   A. oral erythromycin
   B. topical tetracycline
   C. topical silver nitrate
   D. topical erythromycin
   E. eye irrigations with saline solution

50. In neonatal sepsis, the infection may be acquired through the transplacental or transcervical routes or through ascending infection. Of the following, the organism that is acquired by transplacental route is
   A. Klebsiella
   B. Escherichia coli
   C. group B streptococci
   D. Haemophilus influenzae
   E. Listeria monocytogenes

51. The unexplained additional risk factor for early-onset neonatal sepsis is
   A. male sex
   B. preterm birth
   C. maternal fever
   D. fetal tachycardia
   E. prolonged rupture of the membranes (>24 hours)

52. The mainstay of treatment for neonatal sepsis is
   A. high-dose penicillin
   B. cefotaxime and amikacin
   C. ampicillin and gentamicin
   D. ampicillin and cefotaxime
   E. vancomycin and amikacin
53. The **MOST** appropriate treatment for neonatal group B streptococcal meningitis is
   A. meropenem  
   B. high-dose penicillin  
   C. ampicillin and gentamicin  
   D. ampicillin and cefotaxime  
   E. vancomycin and amikacin

54. In late-onset sepsis (8 to 28 days), the **LEAST** common focal infection result from hematogenous seeding is
   A. arthritis  
   B. meningitis  
   C. pneumonia  
   D. osteomyelitis  
   E. urinary tract infection

55. Repetitive, rhythmic contractions of muscle groups of the limbs, face, or trunk are characteristic of
   A. myoclonic seizure  
   B. focal tonic seizure  
   C. focal clonic seizure  
   D. generalized tonic seizure  
   E. progression movements

56. The treatment of bronchopulmonary dysplasia (BPD) include all the following **EXCEPT**
   A. tracheotomy  
   B. good hydration  
   C. dexamethasone  
   D. mechanical ventilation  
   E. prophylaxis against respiratory syncytial virus

57. Retinopathy of prematurity (ROP) is caused by the acute and chronic effects of oxygen toxicity on the developing blood vessels of the premature infant’s retina. To avoid the ROP, arterial oxygen level in premature infants should be kept between
   A. 10 and 30 mm Hg  
   B. 30 and 50 mm Hg  
   C. 50 and 70 mm Hg
D. 70 and 90 mm Hg  
E. 90 and 100 mm Hg

58. Seizures noted in the delivery room often are caused by
   A. hypoglycemia  
   B. drug withdrawal  
   C. intraventricular hemorrhage  
   D. hypoxic-ischemic encephalopathy  
   E. direct injection of local anesthetic agents

59. Sudden onset, short duration seizure that appears on day 1 to 3 of life in a well appearing infant that do not recur may be the result of
   A. hypoglycemia  
   B. drug withdrawal  
   C. benign familial seizures  
   D. subarachnoid hemorrhage  
   E. direct injection of local anesthetic agents

60. The **MOST** correct statement consistent with grade 3 intraventricular hemorrhage (IVH) is
   A. IVH is confined to the germinal matrix  
   B. blood noted in the ventricle with ventricular dilation  
   C. blood noted in the ventricle without ventricular enlargement  
   D. blood in dilated ventricles and in the cerebral cortex contiguous with the ventricle  
   E. blood in dilated ventricles and in the cerebral cortex distant from the ventricle

61. In intraventricular hemorrhage (IVH), posthemorrhagic hydrocephalus may be managed with serial daily lumbar punctures, an external ventriculostomy tube, or a permanent ventricular-peritoneal shunt.  
   Implementation of the shunt often is delayed because of the
   A. risk of kinking  
   B. risk of infection  
   C. high RBC content of the hemorrhagic ventricular fluid  
   D. high protein content of the hemorrhagic ventricular fluid  
   E. high glucose content of the hemorrhagic ventricular fluid

62. In neonate, intraventricular hemorrhage (IVH) is unusual to occur after
   A. 5th day of life
B. 10\textsuperscript{th} day of life  
C. 15\textsuperscript{th} day of life  
D. 20\textsuperscript{th} day of life  
E. 25\textsuperscript{th} day of life

63. A 40-day-old infant product of difficult vaginal delivery, presented with repeated vomiting, pallor, and recurrent seizures. On examination, occipitofrontal circumference was 41 cm. Lab study showed; Hb level, 7 gm/dl. You suspect intracranial hemorrhage.  
Of the following, the **MOST** likely site of hemorrhage is  
A. subdural  
B. subarachnoid  
C. periventricular  
D. intraventricular  
E. intraparenchymal

64. All the following statements regarding subarachnoid hemorrhage are true EXCEPT  
A. may be spontaneous  
B. associated with hypoxia  
C. needs surgical evacuation  
D. seizures are a common presenting manifestation  
E. caused by bleeding from a cerebral arteriovenous malformation

65. Hypoxic-ischemic encephalopathy in the preterm infant is characterized by  
A. cerebral edema  
B. cortical necrosis  
C. subarachnoid hemorrhage  
D. periventricular leukomalacia  
E. involvement of the basal ganglia

66. In hypoxic-ischemic encephalopathy, refractory seizures begin  
A. 12 to 24 hours after birth  
B. 24 to 48 hours after birth  
C. 48 to 72 hours after birth  
D. 72 to 96 hours after birth  
E. 96 to 120 hours after birth
67. Hypoxic-ischemic encephalopathy stage 2 is characterized by all the following EXCEPT
A. miosis
B. seizures
C. hypotonia
D. active Moro reflex
E. hyperactive tendon reflexes

68. Necrotizing enterocolitis (NEC) occurs in 1 to 3 per 1000 live births but the incidence of NEC in neonates admitted to the neonatal intensive care unit is
A. 1% - 8%
B. 10% - 18%
C. 20% - 28%
D. 30% - 38%
E. 40% - 48%

69. The MOST common characteristic cardiac abnormality associated with congenital rubella is
A. aortic stenosis
B. atrial septal defect
C. ventricular septal defect
D. patent ductus arteriosus
E. pulmonary valve stenosis

70. Early clinical sign of necrotizing enterocolitis (NEC) is
A. lethargy
B. bilious emesis
C. abdominal wall erythema
D. abdominal distension
E. increased episodes of apnea

71. In necrotizing enterocolitis (NEC), abdominal perforation may be manifested by development of
A. bilious emesis
B. abdominal wall erythema
C. increased episodes of apnea
D. bluish discoloration of the abdomen
E. disseminated intravascular coagulation
72. The earliest radiographic finding in necrotizing enterocolitis (NEC) is
   A. intestinal ileus
   B. pneumoperitoneum
   C. intrahepatic venous gas
   D. fixed dilated bowel loop
   E. pneumatosis intestinalis

73. The pathognomonic radiographic finding in necrotizing enterocolitis (NEC) is
   A. intestinal ileus
   B. pneumoperitoneum
   C. intrahepatic venous gas
   D. fixed dilated bowel loop
   E. pneumatosis intestinalis

74. In necrotizing enterocolitis (NEC), even with aggressive and appropriate medical management, 25% - 50% of infants require surgical intervention. Of the following, the MOST obvious indication for surgical intervention is
   A. abdominal mass
   B. pneumoperitoneum
   C. intestinal obstruction
   D. rapid onset and progression of pneumatosis
   E. rapid clinical deterioration despite medical therapy

75. In neonatal hyperbilirubinemia, bilirubin is produced by the catabolism of hemoglobin in the reticuloendothelial system, one gram of hemoglobin produces
   A. 5 mg of bilirubin
   B. 15 mg of bilirubin
   C. 25 mg of bilirubin
   D. 35 mg of bilirubin
   E. 45 mg of bilirubin

76. Compared with adults (bilirubin production 3 mg/kg/24 hr), newborns have greater rate of bilirubin production which is about
   A. 6-10 mg/kg/24 hr
   B. 11-15 mg/kg/24 hr
   C. 16-20 mg/kg/24 hr
   D. 21-25 mg/kg/24 hr
   E. 26-30 mg/kg/24 hr
77. Unconjugated bilirubin binds to albumin on specific bilirubin binding sites; in a newborn each 1 g of albumin binds to
   A. 4.5 mg of bilirubin
   B. 6.5 mg of bilirubin
   C. 8.5 mg of bilirubin
   D. 10.5 mg of bilirubin
   E. 12.5 mg of bilirubin

78. Physiologic jaundice is the result of many factors that are normal physiologic characteristics of newborns which include all the following EXCEPT
   A. increased RBC mass
   B. shortened RBC life span
   C. delayed meconium passage
   D. lower ligandin concentration
   E. lower glucuronosyl transferase concentration

79. Oxygen concentrations that are toxic to the neonatal lung is greater than
   A. 20%
   B. 40%
   C. 60%
   D. 80%
   E. 100%

80. All the following are clinical manifestations of bronchopulmonary dysplasia (BPD) EXCEPT
   A. poor growth
   B. oxygen dependence
   C. right sided heart failure
   D. pulmonary hypertension
   E. compensatory metabolic acidosis

81. Crigler-Najjar syndrome characterized by all the following EXCEPT
   A. autosomal recessive
   B. severe direct hyperbilirubinemia
   C. Type I often led to kernicterus
   D. Type II responds to phenobarbital
   E. permanent deficiency of glucuronosyltransferase
82. Gilbert disease, caused by a mutation of the promoter region of glucuronosyl transferase, is characterized by development of
   A. anemia
   B. cholestasis
   C. kernicterus
   D. hepatosplenomegaly
   E. indirect hyperbilirubinemia

83. All the following are radiographic features of transient tachypnea of the newborn **EXCEPT**
   A. overaeration
   B. fluid in the lung fissures
   C. reticulogranular pattern
   D. small pleural effusion
   E. prominent central vascular marking

84. The incidence of meconium-stained amniotic fluid in term and post-term deliveries is
   A. 5%
   B. 15%
   C. 25%
   D. 35%
   E. 45%

85. All the following statements regarding meconium aspiration pneumonia are true **EXCEPT**
   A. it’s common in the breech presentation infants
   B. small airway obstruction causing a ball-valve effect
   C. chemical pneumonitis develop within 24 to 48 hours
   D. meconium is usually aspirated into the lung in utero
   E. prevented by suction the infant’s oropharynx before delivering the rest of the body

86. All the following are recognized features of idiopathic apnea **EXCEPT**
   A. it is a disease of premature infants
   B. occurs during the second week of life
   C. absence of any other identifiable disease
   D. usually resolves by 36 to 40 weeks of postconceptual age
   E. methylxanthines are the mainstay of pharmacologic treatment
87. Of the following, the **MOST** common etiology of conjugated hyperbilirubinemia is
   A. biliary atresia  
   B. galactosemia  
   C. choledochal cyst  
   D. hyperalimentation cholestasis  
   E. α1-Antitrypsin deficiency

88. The presence of dark color urine and gray-white (acholic) stools with jaundice after the second week of life strongly suggest
   A. galactosemia  
   B. biliary atresia  
   C. choledochal cyst  
   D. neonatal hepatitis  
   E. α1-Antitrypsin deficiency

89. The **MOST** important therapy for primary pulmonary hypertension of the newborn (PPHN) is
   A. diuretic  
   B. dopamine  
   C. inhaled nitric oxide  
   D. assisted ventilation  
   E. surfactant replacement

90. The incidence of kernicterus in term infants increases as serum bilirubin levels exceed
   A. 15 mg/dL  
   B. 20 mg/dL  
   C. 25 mg/dL  
   D. 30 mg/dL  
   E. 35 mg/dL

91. Early signs of kernicterus are noted after day 4 of life, the earliest clinical manifestations is
   A. fever  
   B. lethargy  
   C. bulging fontanelle  
   D. opisthotonic posturing  
   E. paralysis of upward gaze
92. Which of the following sequelae of kernicterus is resolved in surviving infants?
   A. spasticity
   B. nerve deafness
   C. choreoathetosis
   D. enamel dysplasia
   E. discoloration of teeth

93. In term infants, phototherapy is initiated when indirect bilirubin levels are between
   A. 10 and 12 mg/dL
   B. 13 and 15 mg/dL
   C. 16 and 18 mg/dL
   D. 19 and 21 mg/dL
   E. 22 and 24 mg/dL

94. The **MOST** characteristic complication of phototherapy in infants with direct-reacting hyperbilirubinemia is
   A. lethargy
   B. dehydration
   C. skin bronzing
   D. macular-papular skin rash
   E. potential for retinal damage

95. An unusual complication of exchange transfusion is
   A. infection
   B. hypoglycemia
   C. vessel perforation
   D. thrombocytopenia
   E. necrotizing enterocolitis

96. Polycythemia is an excessively high hematocrit, which may lead to blood hyperviscosity; it is defined as central hematocrit of
   A. ≥50%
   B. ≥55%
   C. ≥60%
   D. ≥65%
   E. ≥70%
97. All the following are risk factors for polycythemia **EXCEPT**
   A. trisomy 13
   B. prematurity
   C. adrenogenital syndrome
   D. neonatal hyperthyroidism
   E. infants of diabetic mothers

98. Coagulation factors do not pass through the placenta to the fetus, and newborn infants have relatively low level of factor
   A. I
   B. V
   C. VII
   D. VIII
   E. XIII

99. The normal term newborn have significant
   A. low fibrinogen levels
   B. prolonged bleeding time
   C. low fibrin degradation products
   D. prolonged prothrombin time (PT)
   E. prolonged partial thromboplastin time (PTT)

100. The classic hemorrhagic disease of the newborn (HDN) usually occurs between 3 and 7 days of life, early HDN occurs at the 1st day if mother receive
    A. aspirin
    B. heparin
    C. tegretol
    D. rifampin
    E. methyldopa

101. Spontaneous pneumothorax is seen in fewer than 1% of deliveries and may be associated with malformations of the
    A. liver
    B. brain
    C. heart
    D. kidney
    E. esophagus
102. A 12-hour-old term infant delivered by elective cesarean section after uneventful pregnancy presented with tachypnea, grunting, and retractions; oxygen saturation at room air 84% which respond to 40% oxygen (oxygen saturation 95%). Of the following, the MOST likely diagnosis is
   A. congenital pneumonia
   B. congenital heart syndrome
   C. respiratory distress syndrome
   D. meconium aspiration syndrome
   E. transient tachypnea of the newborn

103. Hemorrhagic disease of the newborn (HDN) is prevented by administration of single dose of vitamin K (1 mg)
   A. oral
   B. intravenous
   C. intratracheal
   D. intramuscular
   E. subcutaneous

104. All the following may predispose to hemorrhagic disease of the newborn (HDN) that occur 1 to 3 months after birth, particularly among breastfed infants EXCEPT
   A. hepatitis
   B. cystic fibrosis
   C. biliary atresia
   D. antibiotic use
   E. maternal warfarin

105. The risks of hemorrhage in an infant with neonatal thrombocytopenia born to a mother with idiopathic thrombocytopenic purpura (ITP) may be lessened by cesarean section and by treatment of the mother with
   A. splenectomy
   B. anti-D therapy
   C. corticosteroids
   D. platelet transfusion
   E. intravenous immunoglobulin

106. The treatment of an affected infant with neonatal thrombocytopenia born to a mother with idiopathic thrombocytopenic purpura (ITP) is
   A. conservative
B. splenectomy
C. anti-D therapy
D. corticosteroids
E. platelet transfusion

107. The diagnosis of a neonatal pneumothorax may be confirmed by
   A. chest MRI
   B. chest CT scan
   C. chest radiograph
   D. chest ultrasound
   E. chest transillumination

108. In infants with isoimmune thrombocytopenia or with neonatal thrombocytopenia born to a mother with idiopathic thrombocytopenic purpura (ITP), without treatment, thrombocytopenia resolves spontaneously after birth within
   A. 1 mo
   B. 4 mo
   C. 6 mo
   D. 8 mo
   E. 12 mo

109. The predominant site of hematopoiesis in the last trimester is
   A. liver
   B. spleen
   C. yolk sac
   D. lymph node
   E. bone marrow

110. During intrauterine life, hemoglobin concentration increases from 8 to 10 g/dL at 12 weeks to 16.5 to 18 g/dL at
   A. 34 wks
   B. 36 wks
   C. 38 wks
   D. 40 wks
   E. 42 wks
111. The diagnosis of primary pulmonary hypertension of the newborn (PPHN) is confirmed by
   A. echocardiography
   B. chest radiography
   C. electrocardiography
   D. cardiac catheterization
   E. pulmonary function test

112. The life span of fetal and neonatal RBCs is
   A. 50 to 70 days
   B. 70 to 90 days
   C. 90 to 110 days
   D. 110 to 130 days
   E. 130 to 150 days

113. The mean corpuscular volume of fetal and neonatal RBCs is
   A. 80 to 90 fl
   B. 90 to 100 fl
   C. 100 to 110 fl
   D. 110 to 120 fl
   E. 120 to 130 fl

114. Early clamping of umbilical cord may lead to all the following EXCEPT
   A. anemia
   B. less tachypnea
   C. cardiac murmur
   D. poor peripheral perfusion
   E. improves glomerular filtration

115. The physiologic anemia noted in term infants at age of
   A. 2 to 3 mo
   B. 3 to 4 mo
   C. 4 to 5 mo
   D. 5 to 6 mo
   E. 6 to 7 mo

116. As a complication of RDS, rupture of the alveolar epithelial lining lead to gas dissection along the interstitial space and the peribronchial lymphatics that’s may produce
A. pneumothorax  
B. pneumomediastinum  
C. tension pneumothorax  
D. subcutaneous emphysema  
E. pulmonary interstitial emphysema  

117. All the following statements regarding ABO incompatibility are true EXCEPT  
A. it became more severe with subsequent pregnancies  
B. it may affect the firstborn infant of A or B blood type  
C. it is less severe than hemolysis in Rh-sensitized pregnancy  
D. it is the most common cause of neonatal hyperbilirubinemia requiring therapy  
E. it usually does not cause fetal disease other than extremely mild anemia  

118. Erythroblastosis fetalis classically is caused by Rh blood group incompatibility, which of the following is not antigenic in Rh antigen system?  
A. C  
B. D  
C. E  
D. d  
E. e  

119. The chest radiograph of primary pulmonary hypertension of the newborn (PPHN) usually reveals  
A. hyperinflation  
B. normal lung fields  
C. small pleural effusion  
D. bilateral lung infiltration  
E. prominent vascular marking  

120. Sensitization of the mother carrying an Rh-positive fetus prevented by anti-Rh-positive immune globulin (RhoGAM) 300 μg which given after birth within  
A. 24 hours  
B. 48 hours  
C. 72 hours  
D. 96 hours  
E. 120 hours
121. Which of the following is immune mediated hemolysis in the newborn?
   A. thalassemia
   B. pyruvate kinase
   C. ABO incompatibility
   D. hereditary spherocytosis
   E. glucose-6-phosphate dehydrogenase deficiency

122. Primary pulmonary hypertension of the newborn (PPHN) is characterized by severe hypoxemia, without evidence of parenchymal lung or structural heart disease.
   Of the following, the congenital heart disease that is initially indistinguishable from PPHN is
   A. aortic stenosis
   B. tricuspid atresia
   C. transposition of great arteries
   D. total anomalous venous return
   E. hypoplastic left heart syndrome

123. The initial evaluation of infants with hemolysis include all the following EXCEPT
   A. blood type
   B. reticulocyte count
   C. direct Coombs test
   D. complete blood count
   E. hemoglobin electrophoresis

124. Arterial blood gas analysis (BGA) should perform in all infants with significant respiratory distress, whether or not cyanosis is perceived.
   All the following are true about a normal values of arterial BGA in term infants EXCEPT
   A. pH = 7.35 - 7.40
   B. $\text{Paco}_2 = 35 - 40 \text{ mm Hg}$
   C. $\text{Pao}_2 = 55 - 60 \text{ mm Hg at 30 minutes of life}$
   D. $\text{Pao}_2 = 75 \text{ mm Hg at 4 hours of life}$
   E. $\text{Pao}_2 = 80 \text{ mm Hg at 24 hours of life}$

125. Cyanosis becomes evident clinically when the unsaturated hemoglobin is equal or more than
   A. 1 gm
B. 3 gm  
C. 5 gm  
D. 7 gm  
E. 9 gm

126. Metabolic acidosis defined as a reduced pH (<7.25) and bicarbonate concentration (<18 mEq/L) accompanied by a normal or low Pco₂ level.  
Of the following, the **LAST** line of therapy is  
A. intravenous fluids therapy  
B. increasing inspired oxygen concentration  
C. administration of inotropic or vasoactive drug  
D. slow intravenous sodium bicarbonate infusion  
E. applying of continuous positive airway pressure

127. Respiratory acidosis defined as an elevated Pco₂ level and reduced pH without a reduction in the bicarbonate concentration.  
Of the following, the **MOST** appropriate treatment is  
A. assisted ventilation  
B. intravenous fluids therapy  
C. increasing inspired oxygen concentration  
D. administration of inotropic or vasoactive drug  
E. slow intravenous sodium bicarbonate infusion

128. Sufficient surfactant is produced by type II cells in the lung, is secreted into the alveolar lumen, and is excreted into the amniotic fluid by gestational age of  
A. 28 to 30 wks  
B. 30 to 32 wks  
C. 32 to 34 wks  
D. 34 to 36 wks  
E. 36 to 38 wks

129. Which of the following in amniotic fluid indicates fetal pulmonary maturity?  
A. surfactant proteins  
B. lecithin concentration  
C. neutral lipids concentration  
D. lecithin to sphingomyelin ratio  
E. phosphatidylglycerol concentration
130. In addition to prematurity, all the following are risk factors for respiratory distress syndrome (RDS) **EXCEPT**
   A. male sex
   B. asphyxia
   C. maternal diabetes
   D. being the first-born of twins
   E. delivery of a previous preterm infant with RDS

131. The incidence of respiratory distress syndrome (RDS) increases with decreasing gestational age, RDS develops in 30% to 60% of infants between gestational ages of
   A. 24 – 26 wks
   B. 26 - 28 wks
   C. 28 - 32 wks
   D. 32 - 34 wks
   E. 34 - 36 wks

132. In extremely immature infants (26-30 wks), RDS may develop immediately in the delivery room; while in some more mature infants (34 wks), signs of RDS may develop
   A. 1 to 2 hours after birth
   B. 3 to 4 hours after birth
   C. 5 to 6 hours after birth
   D. 7 to 8 hours after birth
   E. 9 to 10 hours after birth

133. In respiratory distress syndrome (RDS), early chest radiographic finding is
   A. overaeration
   B. whiteout lung
   C. air bronchogram
   D. ground-glass haze
   E. fluid in the lung fissures
1. (C). Polyhydramnios may be acute and associated with premature labor, maternal discomfort, and respiratory compromise. More often, polyhydramnios is chronic and is associated with diabetes, immune or nonimmune hydrops fetalis, multiple gestation, trisomy 18 or 21, and major congenital anomalies. Anencephaly, hydrocephaly, and meningomyelocele are associated with reduced fetal swallowing of amniotic fluid. Esophageal and duodenal atresia as well as cleft palate interferes with swallowing and gastrointestinal fluid dynamics.

2. (E). Potter syndrome characteristic by diminished chest wall size accompanied by pulmonary hypoplasia and, often, pneumothorax. Uterine compression in the absence of amniotic fluid retards lung growth, and patients with this condition die of respiratory failure rather than renal insufficiency.

3. (B).

4. (A).

5. (C). Twin-to-twin transfusion syndrome is associated with a high mortality and is seen only in monozygotic twins who share a common placenta and have an arteriovenous connection between their circulations. The fetus on the arterial side of the shunt serves as the blood donor, resulting in fetal anemia, growth retardation, and oligohydramnios for this fetus. The recipient, or venous-side twin, is larger or discordant in size, is plethoric and polycythemic, and may show polyhydramnios. Weight differences of 20% and hemoglobin differences of 5 g/dL suggest the diagnosis.

6. (B).

7. (C). Low birth weight (LBW) infants, defined as infants having birth weights of less than 2500 gm.

8. (C). The major causes of neonatal mortality in preterm infant are respiratory distress syndrome/bronchopulmonary dysplasia (chronic lung disease), severe immaturity, congenital anomalies, infection, intraventricular hemorrhage, and necrotizing enterocolitis.

9. (A). The major causes of neonatal mortality in fullterm infant are birth asphyxia, birth trauma, congenital anomalies, infection, macrosomia, meconium aspiration pneumonia, and persistent pulmonary hypertension.

11. (A).
12. (A).
13. (B).
14. (B).
15. (E).
16. (C).
17. (C). Muscle tone.
18. (E). Evidence of a diaphragmatic hernia and a low Apgar score indicate that immediate endotracheal intubation is required. If a mask and bag are used, gas enters the lung and the stomach, and the latter may act as an expanding mass in the chest that compromises respiration.
19. (D). Severe cold injury in an infant is manifested by acidosis, hypoxia, hypoglycemia, apnea, bradycardia, pulmonary hemorrhage, and a pink skin color. The color is caused by trapping of oxygenated hemoglobin in the cutaneous capillaries.
20. (B). The radiographic appearance of BPD is characterized initially by lung opacification and subsequently by development of cysts accompanied by areas of overdistention and atelectasis, giving the lung a spongelike appearance.
21. (C). This is a feature of caput succedaneum.
22. (A). Total serum calcium levels of less than 7 mg/dL and ionized calcium levels of less than 3 to 3.5 mg/dL are considered hypocalcemia.
23. (E). Late neonatal hypocalcemia, or neonatal tetany, often is the result of ingestion of high phosphate-containing milk or the inability to excrete the usual phosphorus in commercial infant formula. While other distracters may be responsible for early neonatal hypocalcemia associated with asphyxia.
25. (E).
   - Tetracycline --- enamel hypoplasia
   - Penicillamine --- cutis laxa syndrome
   - Vitamin D --- supravalvular aortic stenosis
   - Vitamin A --- facial and ear anomalies and congenital heart disease
   - Phenytoin---hypoplastic nails, intrauterine growth restriction, and typical facies
26. (A).
   - Valproate ---- spina bifida
   - Streptomycin ---- deafness
Lithium ---- Ebstein anomaly
Thalidomide ---- phocomelia
Radioactive iodine ---- fetal hypothyroidism

27. (D). Evaluation of patients thought to have a congenital infection should include attempts to isolate the organism by culture (for rubella, CMV, HSV, gonorrhea, and M. tuberculosis), to identify the antigen of the pathogen (for hepatitis B and Chlamydia trachomatis), to identify the pathogen’s genome with polymerase chain reaction (PCR), and to identify specific fetal production of antibodies (IgM or increasing titer of IgG for Toxoplasma, syphilis, parvovirus, HIV, or Borrelia).

28. (E). Also affected infants with congenital toxoplasmosis tend to be small for gestational age, develop early-onset jaundice, have hepatosplenomegaly, and present with a generalized maculopapular rash. Seizures are common, and skull films may reveal diffuse cortical calcifications in contrast to the periventricular pattern observed with CMV.

29. (D). In congenital toxoplasmosis serologic tests are the primary means of diagnosis. IgG-specific antibodies achieve a peak concentration 1 to 2 months after infection and remain positive indefinitely. For infants with seroconversion or a fourfold increase in IgG titers, specific IgM antibody determinations should be performed to confirm disease. Especially for congenital infections, measurement of IgA and IgE antibodies can be useful to confirm the disease.

30. (D). Fetal infection rarely can occur after reactivation of disease in an immunocompromised pregnant mother.

31. (D).

32. (E). The most common characteristic abnormalities associated with congenital rubella include ophthalmologic (cataracts, retinopathy, and glaucoma), cardiac (patent ductus arteriosus and peripheral pulmonary artery stenosis), auditory (sensorineural hearing loss), and neurologic (behavioral disorders, meningoencephalitis, and mental retardation) conditions.

33. (A). Infants with congenital rubella are chronically and persistently infected and tend to shed live virus in urine, stools, and respiratory secretions for 1 yr.

34. (D). Detection of rubella-specific IgM antibody usually indicates recent infection. Measurement of rubella-specific IgG over several months can be confirmatory. Rubella virus can be isolated from blood, urine, CSF, and throat swab specimens.

35. (C). More than 90% of infants who have congenital CMV infection exhibit no clinical evidence of disease at birth. Approximately 10% of infected infants are small for gestational age and have symptoms at birth. Findings include microcephaly, thrombocytopenia, hepatosplenomegaly, hepatitis, intracranial calcifications, chorioretinitis, and hearing abnormalities. Some infants can present
with a blueberry muffin appearance as the result of dermal erythropoiesis. Skull films may reveal periventricular calcifications.

36. (C).
37. (A).
38. (A).
39. (E). Skull films may reveal periventricular calcifications in CMV in contrast to diffuse cortical calcifications pattern observed with congenital toxoplasmosis.
40. (A). HSV-2 accounts for 90% of primary genital herpes. About 70% to 85% of neonatal herpes simplex infections are caused by HSV-2.
41. (A). Symptoms of neonatal HSV infection include disseminated disease involving multiple organ systems (liver and lungs); localized infection to the CNS; or localized infection to the skin, eyes, and mouth.
42. (B). 4 weeks of age or more than 34 weeks’ corrected gestational age, whichever comes first.
43. (C). Parenteral acyclovir is the treatment of choice for neonatal HSV infections. Acyclovir should be administered to all infants suspected to have infection or diagnosed with HSV.
44. (A). In syphilis, severe consolidated pneumonia may be present at birth, and there may be laboratory findings consistent with a glomerulonephritis. CSF evaluation may reveal a pleocytosis and elevated protein. More than 90% of symptomatic infants exhibit radiographic abnormalities of the long bones consistent with osteochondritis and perichondritis.
45. (A). Parenteral penicillin is the preferred drug of choice for treatment of syphilis. Penicillin G for 10 to 14 days is the only documented effective therapy for infants who have congenital syphilis and neurosyphilis.
46. (D). Some manifestations of disease may not become apparent until many years after birth, such as interstitial keratitis, eighth cranial nerve deafness, Hutchinson teeth, bowing of the shins, frontal bossing, mulberry molars, saddle nose, rhagades, and Clutton joints.
47. (E). Recommended treatment for isolated infection, such as ophthalmia neonatorum, is one intramuscular dose of ceftriaxone. Infants with gonococcal ophthalmia should receive eye irrigations with saline solution at frequent intervals before discharge. Topical antibiotic therapy alone is inadequate and is unnecessary when recommended systemic antimicrobial therapy is given. Infants with gonococcal ophthalmia should be hospitalized and evaluated for disseminated disease (sepsis, arthritis, meningitis).
48. (E). Chlamydia trachomatis, is the most common reportable sexually transmitted infection. In infected infants, the risk of conjunctivitis is 25% to 50%, and the risk of pneumonia is 5% to 20% (Pneumonia in a young infant can occur
between 2 and 19 weeks of age and is characterized by an afebrile illness with a repetitive staccato cough, and tachypnea. Wheezing is uncommon).

49.(A). Infants with conjunctivitis and pneumonia are treated with oral erythromycin for 14 days. Topical treatment of conjunctivitis is ineffective and unnecessary. The recommended topical prophylaxis with silver nitrate, erythromycin, or tetracycline for all newborns for the prevention of gonococcal ophthalmia does not prevent neonatal chlamydial conjunctivitis.

50.(E). The bacteria responsible for ascending infection of the fetus are common bacterial organisms of the maternal genitourinary tract, such as group B streptococci, Escherichia coli, Haemophilus influenzae, and Klebsiella. Herpes simplex virus (HSV)-1 or, more often, HSV-2 also causes ascending infection that at times may be indistinguishable from bacterial sepsis. Syphilis and Listeria monocytogenes are acquired by transplacental infection.

51.(A). Risk factors for early-onset sepsis include vaginal colonization with group B streptococci, prolonged rupture of the membranes (>24 hours), amnionitis, maternal fever or leukocytosis, fetal tachycardia, and preterm birth. African American race and male sex are unexplained additional risk factors for neonatal sepsis.

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

53.(B). High-dose penicillin (250,000 to 450,000 U/kg/24 hr) is appropriate for group B streptococcal meningitis.

54.(C). Late-onset sepsis (8 to 28 days) usually occurs in a healthy full-term infant who was discharged in good health from the normal newborn nursery. In addition to bacteremia, hematogenous seeding may result in focal infections, such as meningitis (in 75% of cases), osteomyelitis (group B streptococci, Staphylococcus aureus), arthritis (gonococcus, S. aureus, Candida albicans, gram-negative bacteria), and urinary tract infection (gram-negative bacteria).

55.(C). Focal clonic seizure is characterized by repetitive, rhythmic contractions of muscle groups of the limbs, face, or trunk; may be unilateral or multifocal; may appear synchronously or asynchronously in various body regions; cannot be suppressed by restraint.

56.(B). Severe chest retractions produce negative interstitial pressure that draws fluid into the interstitial space. Together with cor pulmonale, these chest retractions cause fluid retention, necessitating fluid restriction and the administration of diuretics.
57. (C). Although there is no absolutely safe Pao₂ level, it is wise to keep the arterial oxygen level between 50 and 70 mm Hg in premature infants.

58. (E).

- Seizures noted in the delivery room often are caused by direct injection of local anesthetic agents into the fetal scalp, severe anoxia, or congenital brain malformation.
- Hypoxic-ischemic encephalopathy, a common cause of seizures in the full-term infant, usually occurs 12 to 24 hours after a history of birth asphyxia.
- Intraventricular hemorrhage (IVH) is a common cause of seizures in premature infants and often occurs between 1 and 3 days of age.
- Seizures caused by hypoglycemia often occur when blood glucose levels decline to the lowest postnatal value (at 1 to 2 hours of age or after 24 to 48 hours of poor nutritional intake).
- Seizures after the first 5 days of life may be the result of infection or drug withdrawal.

59. (D).

60. (B). Grade 1 IVH is confined to the germinal matrix; grade 2 is an extension of grade 1, with blood noted in the ventricle without ventricular enlargement; grade 3 is an extension of grade 2 with ventricular dilation; and grade 4 has blood in dilated ventricles and in the cerebral cortex, either contiguous with or distant from the ventricle.

61. (D).

62. (A). Most periventricular hemorrhages and IVHs in neonate occur in the first 3 days of life. It is unusual for IVH in neonate to occur after day 5 of life.

63. (A). The subdural hematoma does not always cause symptoms immediately after birth; with time, however, the RBCs undergo hemolysis and water is drawn into the hemorrhage because of the high oncotic pressure of protein, resulting in an expanding symptomatic lesion. Anemia, vomiting, seizures, and macrocephaly may occur in an infant who is 1 to 2 months of age and has a subdural hematoma.

64. (C). Treatment is directed at the seizure and the rare occurrence of posthemorrhagic hydrocephalus. The treatment of all symptomatic subdural hematomas is surgical evacuation.

65. (D). Hypoxic-ischemic encephalopathy in the term infant is characterized by cerebral edema, cortical necrosis, and involvement of the basal ganglia, whereas in the preterm infant it is characterized by periventricular leukomalacia. Both lesions may result in cortical atrophy, mental retardation, and spastic quadriplegia or diplegia.

66. (A).
67. (D). Weak Moro reflex.
68. (A).
69. (D). Patent ductus arteriosus and peripheral pulmonary artery stenosis.
70. (D). Early clinical signs of NEC include abdominal distention, feeding intolerance/increased gastric residuals, emesis, rectal bleeding, and occasional diarrhea. As the disease progresses, patients may develop marked abdominal distention, bilious emesis, ascites, abdominal wall erythema, lethargy, temperature instability, increased episodes of apnea/bradycardia, disseminated intravascular coagulation, and shock.
71. (D). With abdominal perforation, the abdomen may develop a bluish discoloration.
72. (A). The earliest radiographic finding is intestinal ileus, often associated with thickening of the bowel loops and air-fluid levels.
73. (E). The pathognomonic radiographic finding is pneumatosis intestinalis caused by hydrogen gas production from pathogenic bacteria present between the subserosal and muscularis layers of the bowel wall. Radiographic findings also may include a fixed or persistent dilated loop of bowel, intrahepatic venous gas, and pneumoperitoneum seen with bowel perforation.
74. (B). The decision to perform surgery is obvious when the presence of a pneumoperitoneum is observed on abdominal radiograph. Other, not so obvious indications for surgical intervention include rapid clinical deterioration despite medical therapy, rapid onset and progression of pneumatosis, abdominal mass, and intestinal obstruction.
75. (D).
76. (A). This increased production is caused, in part, by an increased RBC mass (higher hematocrit) and a shortened erythrocyte life span of 70 to 90 days compared with the 120-day erythrocyte life span in adults.
77. (C). If the binding sites become saturated or if a competitive compound binds at the site, displacing bound bilirubin, free bilirubin becomes available to enter the central nervous system.
78. (C). Delayed passage of meconium, which contains bilirubin, is not a normal physiologic characteristic of newborns, may contribute to the enterohepatic recirculation of bilirubin.
79. (B). Oxygen-mediated lung injury results from the generation of superoxides, hydrogen peroxide, and oxygen free radicals, which disrupt membrane lipids.
80. (E). The clinical manifestations of BPD are oxygen dependence, hypercapnia with a compensatory metabolic alkalosis, pulmonary hypertension, poor growth, and development of right sided heart failure.
81. (B). Crigler-Najjar syndrome is a serious, rare, autosomal recessive, permanent deficiency of glucuronosyltransferase that results in severe indirect hyperbilirubinemia. Type II responds to enzyme induction by phenobarbital, producing an increase in enzyme activity and a reduction of bilirubin levels. Type I do not respond to phenobarbital and manifests as persistent indirect hyperbilirubinemia, often leading to kernicterus.

82. (E). In the presence of another icterogenic factor (hemolysis), more severe jaundice may develop.

83. (C). Air bronchograms and a reticulogranular pattern are not seen; their presence suggests another pulmonary process, such as RDS or pneumonia.

84. (B).

85. (D). Aspiration of amniotic fluid contaminated with particulate meconium may occur in utero in a distressed, gasping fetus; more often, meconium is aspirated into the lung immediately after delivery.

86. (B). Occurs during the first week of life.

87. (D).

- Common; hyperalimentation cholestasis, CMV infection, TORCH, inspissated bile from prolonged hemolysis, neonatal hepatitis, and sepsis.
- Uncommon; hepatic infarction, inborn errors of metabolism (galactosemia, tyrosinemia), cystic fibrosis, biliary atresia, choledochal cyst, α1-Antitrypsin deficiency, neonatal iron storage disease, Alagille syndrome (arteriohepatic dysplasia), and Byler disease

88. (B).

89. (D). The most important therapy for PPHN is assisted ventilation. Reversible mild pulmonary hypertension may respond to conventional assisted ventilation. Patients with severe PPHN do not always respond to conventional therapy.

90. (C). Kernicterus usually is noted when the bilirubin level is excessively high for gestational age. It usually does not develop in term infants when bilirubin levels are less than 20 to 25 mg/dL, but the incidence increases as serum bilirubin levels exceed 25 mg/dL.

91. (B).

- The earliest clinical manifestations of kernicterus are lethargy, hypotonia, irritability, poor Moro response, and poor feeding. A high-pitched cry and emesis also may be present.
- Later signs include bulging fontanelle, opisthotonic posturing, pulmonary hemorrhage, fever, hypertonicity, paralysis of upward gaze, and seizures.
92. (A). Spasticity resolves in surviving infants, who may manifest later nerve deafness, choreoathetoid cerebral palsy, mental retardation, enamel dysplasia, and discoloration of teeth as permanent sequelae.

93. (C). Phototherapy is initiated in premature infants when bilirubin is at lower levels, to prevent bilirubin from reaching the high concentrations necessitating exchange transfusion. Blue lights and white lights are effective in reducing bilirubin levels.

94. (C). Complications of phototherapy include an increased insensible water loss, diarrhea, and dehydration. Additional problems are macular-papular red skin rash, lethargy, masking of cyanosis, nasal obstruction by eye pads, and potential for retinal damage.

95. (D). Complications of exchange transfusion include problems related to the blood (transfusion reaction, metabolic instability, or infection), the catheter (vessel perforation or hemorrhage), or the procedure (hypotension or necrotizing enterocolitis [NEC]). Unusual complications include thrombocytopenia and graft-versus-host disease.

96. (D). Polycythemia is an excessively high hematocrit (≥65%), which may lead to hyperviscosity that produces symptoms related to vascular stasis, hypoperfusion, and ischemia.

97. (B). Infants at special risk for polycythemia are term and post-term small for gestational age infants, infants of diabetic mothers, infants with delayed cord clamping, and infants with neonatal hyperthyroidism, adrenogenital syndrome, trisomy 13, trisomy 18, trisomy 21, twin-to-twin transfusion syndrome (recipient), or Beckwith-Wiedemann syndrome.

98. (C). Coagulation factors do not pass through the placenta to the fetus, and newborn infants have relatively low levels of the vitamin K–dependent factors II, VII, IX, and X. Contact factors XI and XII, prekallikrein, and kininogen also are lower in newborns than in adults. Fibrinogen (factor I); plasma levels of factors V, VIII, and XIII; and platelet counts are within the adult normal range.

99. (E). Because of the transient, relative deficiencies of the contact and vitamin K–dependent factors, the PTT, which is dependent on factors XII, IX, VIII, X, V, II, and I, is prolonged in the newborn period. Preterm infants have the most marked prolongation of the PTT (50 to 80 seconds) compared with term infants (35 to 50 seconds) and older, more mature infants (25 to 35 seconds). The PT is only slightly prolonged in term infants (13 to 20 seconds) compared with preterm infants (13 to 21 seconds) and more mature patients (12 to 14 seconds).

100. (D). Hemorrhage on the first day of life resulting from a deficiency of the vitamin K–dependent factors often is associated with administration to the mother of drugs that affect vitamin K metabolism in the infant (maternal warfarin or
antibiotic, e.g., isoniazid or rifampin) therapy and mothers receiving phenobarbital and phenytoin).

101. (D).

102. (E). Transient tachypnea of the newborn (TTN) is a self-limited condition. Cyanosis, when present, usually requires treatment with supplemental oxygen in the range of 30% to 40%. TTN usually is noted in larger premature infants and in term infants born by precipitous delivery or cesarean section without prior labor.

103. (D).

104. (E). Bleeding also may occur 1 to 3 months after birth, particularly among breastfed infants. Vitamin K deficiency in breastfed infants also should raise suspicion about the possibility of vitamin K malabsorption resulting from cystic fibrosis, biliary atresia, hepatitis, or antibiotic suppression of the colonic bacteria that produce vitamin K.

105. (C). Neonatal thrombocytopenia in infants born to women with idiopathic thrombocytopenic purpura (ITP) also is a result of placental transfer of maternal IgG antibodies. In ITP, these autoantibodies are directed against all platelet antigens; mother and newborn may have low platelet counts.

106. (D). Prednisone and intravenous immunoglobulin.

107. (C). The diagnosis of a neonatal pneumothorax may be based on unequal transillumination of the chest and may be confirmed by chest radiograph.

108. (A).

- Isoimmune thrombocytopenia; without treatment, thrombocytopenia resolves during the first month of life as the maternal antibody level declines.
- Neonatal thrombocytopenia born to a mother with idiopathic thrombocytopenic purpura (ITP); thrombocytopenia resolves spontaneously during the first month of life as maternal-derived antibody levels decline.

109. (E). Embryonic hematopoiesis begins by the 20th day of gestation and is evidenced as blood islands in the yolk sac. In midgestation, erythropoiesis occurs in the liver and spleen; the bone marrow becomes the predominant site in the last trimester.

110. (D). Fetal red blood cell (RBC) production is responsive to erythropoietin; the concentration of this hormone increases with fetal hypoxia and anemia.

111. (A). The diagnosis is confirmed by echocardiographic examination, which shows elevated pulmonary artery pressures and sites of right-to-left shunting. Echocardiography also rules out structural congenital heart disease and transient myocardial dysfunction.

112. (B).
113. (D). Delayed clamping may increase the risk of polycythemia and jaundice, but it improves glomerular filtration. Early clamping may lead to anemia, a cardiac murmur, poor peripheral perfusion, and less tachypnea.

114. (E). Physiologic anemia is a normal process that does not result in signs of illness and does not require any treatment. It is a physiologic condition believed to be related to several factors, including increased tissue oxygenation experienced at birth, shortened RBC life span, and low erythropoietin levels.

115. (A). Physiologic anemia is a normal process that does not result in signs of illness and does not require any treatment. It is a physiologic condition believed to be related to several factors, including increased tissue oxygenation experienced at birth, shortened RBC life span, and low erythropoietin levels.

116. (E). Rupture of the alveolar epithelial lining may produce pulmonary interstitial emphysema as gas dissects along the interstitial space and the peribronchial lymphatics. Extravasation of gas into the parenchyma reduces lung compliance and worsens respiratory failure. Gas dissection into the mediastinal space produces a pneumomediastinum, occasionally dissecting into the subcutaneous tissues around the neck, causing subcutaneous emphysema. Alveolar rupture adjacent to the pleural space produces a pneumothorax.

117. (A). ABO incompatibility with sensitization usually does not cause fetal disease other than extremely mild anemia. It may produce hemolytic disease of the newborn, which is manifested as significant anemia and hyperbilirubinemia. Because many mothers who have blood group O have IgG antibodies to A and B before pregnancy, the firstborn infant of A or B blood type may be affected. In contrast to Rh disease, ABO hemolytic disease does not become more severe with subsequent pregnancies. Hemolysis with ABO incompatibility is less severe than hemolysis in Rh-sensitized pregnancy, either because the anti-A or anti-B antibody may bind to nonerythrocytic cells that contain A or B antigen or because fetal erythrocytes have fewer A or B antigenic determinants than they have Rh sites. With the declining incidence of Rh hemolytic disease, ABO incompatibility has become the most common cause of neonatal hyperbilirubinemia requiring therapy—currently accounting for approximately 20% of clinically significant jaundice in the newborn.

118. (D). The Rh antigen system consists of five antigens: C, D, E, c, and e; the d type is not antigenic. In most Rh-sensitized cases, the D antigen of the fetus sensitizes the Rh-negative (d) mother, resulting in IgG antibody production during the first pregnancy.

119. (B).

120. (C). Prevention of sensitization of the mother carrying an Rh-positive fetus is possible by treating the mother during gestation (>28 weeks’ gestational age) and within 72 hours after birth with anti-Rh-positive immune globulin (RhoGAM).

121. (C). Other distracters are non-immune mediated hemolysis in the newborn.
122. (D). Total anomalous venous return associated with obstruction of blood flow may produce a clinical picture that involves severe hypoxia and that is initially indistinguishable from PPHN; however, a chest radiograph reveals severe pulmonary venous engorgement and a small heart. Echocardiography or cardiac catheterization confirms the diagnosis.

123. (E). A complete blood count, blood smear, reticulocyte count, blood type, and direct Coombs test should be performed in the initial evaluation of all infants with hemolysis. Reduced hemoglobin levels, reticulocytosis, and a blood smear characterized by polychromasia and anisocytosis are expected with isoimmune hemolysis. Spherocytes commonly are observed in ABO incompatibility. The determination of the blood type and the Coombs test identify the responsible antigen and antibody in immunologically mediated hemolysis.

In the absence of a positive Coombs test and blood group differences between the mother and fetus, other causes of nonimmune hemolysis must be considered. RBC enzyme assays, hemoglobin electrophoresis, or RBC membrane tests (osmotic fragility, spectrin assay) should be performed.

124. (E). Arterial blood gas analysis, $P_{aO_2} = 90$ mm Hg at 24 hours of life. Capillary blood gas determinations are useful in determining blood pH and the $P_{aco_2}$ level but may result in falsely low blood $P_{aO_2}$ readings.

125. (C). Anemia may interfere with the perception of cyanosis. Jaundice also may interfere with the appearance of cyanosis.

126. (D). The management of metabolic acidosis may include, as in the sequence of therapy for hypoxia, increasing the inspired oxygen concentration; applying continuous positive airway pressure nasally; or initiating mechanical ventilation using positive end-expiratory pressure. Patients with hypotension produced by hypovolemia require fluids and may need inotropic or vasoactive drug support. If metabolic acidosis persists despite specific therapy, sodium bicarbonate (1 mEq/kg/dose) may be given by slow intravenous infusion.

127. (A). Respiratory acidosis may be caused by pulmonary insufficiency or central hypoventilation. Most disorders producing respiratory distress can lead to hypercapnia. Treatment involves assisted ventilation but not sodium bicarbonate.

128. (D). The timing of surfactant production in quantities sufficient to prevent atelectasis depends on an increase in fetal cortisol levels that begins between 32 and 34 weeks of gestation.

129. (D). The concentration of lecithin in amniotic fluid indicates fetal pulmonary maturity. Because the amount of lecithin is difficult to quantify, the ratio of lecithin (which increases with maturity) to sphingomyelin (which remains constant during gestation) (L/S ratio) is determined. A L/S ratio of 2:1 usually indicates pulmonary maturity. The presence of minor phospholipids, such as phosphatidylglycerol, also
is indicative of fetal lung maturity and may be useful in situations in which the L/S ratio is borderline or possibly affected by maternal diabetes, which reduces lung maturity.

130. **(D)**. Other risk factors in addition to prematurity include delivery of a previous preterm infant with RDS, maternal diabetes, hypothermia, fetal distress, asphyxia, male sex, white race, being the second-born of twins, and delivery by cesarean section without labor.

131. **(C)**. Infants at greatest risk for RDS are premature and have an immature L/S ratio.

132. **(B)**. This correlates with the initial release of stored surfactant at the onset of breathing accompanied by the ongoing inability to replace the surfactant owing to inadequate stores.

133. **(D)**. Atelectasis is well documented by radiographic examination of the chest, which shows a ground-glass haze in the lung surrounding air-filled bronchi (the air bronchogram). Severe RDS may show an airless lung field (whiteout) on a radiograph, even obliterating the distinction between the atelectatic lungs and the heart.
1. The leading cause of death in adolescents is
   A. unintentional injury
   B. homicide
   C. suicide
   D. malignant neoplasm
   E. cardiac disease

2. Hematocrit is normally higher in adolescent boys than girls; this difference in value is mainly due to
   A. greater androgenic stimulation of the bone marrow in boys
   B. loss through menstruation in girls
   C. dietary preference of both sexes
   D. earlier growth spurt of girls
   E. difference in physiology of utilization of minerals between both sexes

3. Completion of the Tanner stages in females should take 4 to 5 years. The peak growth spurt usually occurs during
   A. sexual maturity rating I-II of breast development
   B. sexual maturity rating II –III of breast development
   C. sexual maturity rating III-IV of breast development
   D. one year after menarche
   E. just after pubarche

4. Determination of physical growth and development of puberty in adolescent girls is dependent on the rating of
   A. adrenarche and menarche
   B. menarche
   C. thelarche and pubarche
   D. thelarche and menarche
   E. ovarian maturation

5. You are meeting a mother of comparatively healthy twins (male and female), her concern that, the male adolescent is not growing (especially the height) as his sister
is doing. Your initial evaluation shows normal growth parameters for both. The next appropriate step is to

A. refer the male adolescent to pediatric endocrinologist
B. refer the female adolescent to pediatric endocrinologist
C. explain that it could be related to familial short stature
D. explain that it could be related to female adolescent growth spurt
E. send for assessment of bone age

6. You are evaluating sexual maturity rating (SMR) of the breast of an adolescent female; you find a small mound of the breast and papilla with enlargement of areolar diameter. The SMR of breast will be

A. stage I
B. stage II
C. stage III
D. stage IV
E. stage V

7. You are evaluating a preadolescent boy by using an orchidometer. The expected testicular volume at this stage should be less than

A. 2 ml
B. 4 ml
C. 6 ml
D. 9 ml
E. 10 ml

8. A 14-year-old girl presents to endocrinology clinic complaining of amenorrhea; you find breast development of stage III SMR. Of the following, the MOST appropriate next action is to

A. do pelvic examination to rule out an outflow obstruction
B. send for FSH and LH
C. send for TSH
D. send for chromosomal study
E. give her follow up visit

9. An 8-year-old girl is brought by her mother to your clinic with high concern about breast mass; you find a well nourished; normally developed girl with asymmetrical; tender pea-size right breast mass just beneath the nipple. Of the following, the MOST appropriate action is to

A. reassure the mother
B. send for hormonal assay  
C. send for U/S  
D. send for mammography  
E. do FNA (fine needle aspiration) cytology  

10. An 11-year-old adolescent girl presents with a complaint of odorless/colorless vaginal discharge. Locally you find a well estrogenized vulva and hymen without erythema or excoriation. 

Of the following, the **MOST** appropriate action is to  
A. take cultures and consider abuse  
B. inform as this is the first signs of menarche  
C. send for hormonal assessment  
D. send for pelvic U/S  
E. reassure the patient  

11. A 12-year-old introverted boy complaining of gynecomastia, he informs you that he feels embarrassed because of his breast size in front of his beers. You find a 5 cm, round, freely mobile, mildly tender, and firm mass immediately beneath the areola. His sexual maturity rating was stage III. 

Of the following the **MOST** appropriate action is to  
A. refer him to psychiatrist  
B. reassure him as this is a self limiting mass  
C. prescribe bromocriptine  
D. refer to cosmetic surgery  
E. use new technique of botocks injection  

12. You are explaining the etiology of anorexia nervosa to a mother of affected girl. Your statement about the etiology of anorexia nervosa is **MOSTLY**  
A. unknown  
B. social  
C. environmental  
D. psychological  
E. biologic  

13. A 16-year-old adolescent girl referred from dietician for further evaluation. She had increasing weight loss of about 30% of body weight in spite of management in the last 2 months. You find a well negotiating girl, accepting her weight, she was wearing an oversize multilayer cloths. There was lanugo-hair over the face, erosion of enamel of teeth, a well tolerated bradycardia and hypothermia in addition to cyanosis of hands and feet.
Of the following, the **MOST** likely diagnosis is

A. a missed case of cyanotic heart disease
B. hypothyroidism
C. inflammatory bowel disease
D. malabsorption syndrome
E. anorexia nervosa

14. The **BEST** treatment plan to be chosen for the above scenario (question 13) is

A. refer back to dietician with titled diagnosis
B. admit the girl and start voluntary NG feeding
C. give a set of advices and follow-up appointment
D. arrange for gastrostomy feeding
E. send for electrolytes and renal function assessment
1. (A). Motor vehicle injuries and other unintentional injuries account for more than 75% of all deaths in this age group.

2. (A). It represents physiological change during adolescents. Alkaline phosphatase levels in boys and in girls increase during puberty because of rapid bone turnover, especially during the growth spurt.

3. (C). It usually occurs 1 year after thelarche at sexual maturity rating stage III to IV breast development and just before the onset of menstruation (menarche).

4. (C). Understanding these terms is important in understanding sexual maturity rating (SMR) of an adolescent girl. Thelarche is a breast budding under the areola and adrenarche or (pubarche) is a fine straight pubic hair over the mons pubis. These changes mark the sexual maturity rating, or Tanner staging during puberty of an adolescent female. In a male adolescent is mainly determined by the size of testes and adrenarche.

5. (D). After assessment of both growth parameters and SMR for each one of them with the normal results according to age of each one, the most likely explanation is the earlier growth spurt that occurs in the adolescent female.

6. (B). This is breast bud stage. In stage I, the breasts are preadolescent. There is elevation of the papilla only. In stage III, there is further enlargement of breast and areola with no separation of their contours. In stage IV there is a projection of the areola and papilla to form a secondary mound above the level of the breast while in stage V; the breasts resemble those of a mature female as the areola has recessed to the general contour of the breast.

7. (B).

8. (E). Primary amenorrhea is the complete absence of menstruation by 16 years of age in the presence of breast development or by 14 years of age in the absence of breast development. So this girl does not fit the definition of primary amenorrhea. Regarding other options, it could be true if another piece of information is given in the question like recurrent abdominal pain in option A, hoarse voice for option C, short stature for option D, and etc.

9. (A). Actually this is a picture of normal variant of puberty, and the description is typical. U/S is preferred than mammography in adolescents with breast mass to avoid radiation exposure. Breast carcinoma is extremely rare in this group.

10. (E). This is physiologic leukorrhea, due to ovarian estrogen stimulation of the uterus and vagina usually occurs in peripubertal girls (sexual maturity rating stage
III) who often complain of vaginal discharge. If there are symptoms (odor/color/pruritis), cultures should be obtained and sexual abuse is considered. In these circumstances, vaginal cultures can be obtained without a speculum because sexually transmitted infections are vaginal until menarche, when cervical infections are the rule.

11.(C). Breast enlargement in boys is usually benign and self-limited condition. It’s noted in 50% to 60% of boys during early adolescence. Typical size is between 1- to 3-cm. Reassurance is usually the only treatment required. If the condition worsens and is associated with psychological morbidity, it may be treated with bromocriptine. Surgical treatment with reduction mammoplasty can be helpful with massive hypertrophy.

12.(A). Actually it is unknown but involves interaction of all the mentioned above options.

13.(E). This is typical presentation of anorexia nervosa; usually they have fear of being overweight (in spite of their miserable weight). Except option A all other options are in differential diagnosis, in addition to gastroesophageal reflux, peptic ulcer, chronic diarrhea, increased energy demands, hypothalamic lesions, hyperthyroidism, diabetes mellitus, and Addison disease. Psychiatric disorders also need to be considered (e.g., drug abuse, depression, obsessive-compulsive disorders).

14.(B). If we look at the scenario we will find most of indications for admission is met; like weight loss >25% ideal body weight, bradycardia, hypothermia and failure of outpatient treatment. Other indications for admission are risk of suicide, dehydration, hypokalemia and dysrhythmias. Treatment requires a multidisciplinary approach, including a feeding program as well as individual and family therapy. Feeding is accomplished through voluntary intake of regular foods, nutritional formula orally or by nasogastric tube.
1. The major components of host defense include anatomic barriers and the innate and adaptive immune systems. The adaptive immune system is made up of
   A. T and B lymphocytes
   B. acute-phase proteins
   C. cytokines
   D. complement
   E. cellular components

2. Failure to thrive, diarrhea, malabsorption, and infections with opportunistic infections (i.e., fungi, Candida sp, Pneumocystis jiroveci [carinii]) suggest
   A. B-cell defects
   B. complement defects
   C. T-cell defects
   D. neutrophil defects
   E. NK-cell deficiency

3. Delayed separation of umbilical cord, absence of pus at site (s) of infection, and poor wound healing suggest
   A. B-cell defects
   B. complement defects
   C. T-cell defects
   D. neutrophil defects
   E. NK-cell deficiency

4. Selective IgA deficiency is defined as serum IgA levels less than 10 mg/dL with normal levels of other immunoglobulins. The diagnosis cannot be confirmed until the patient is at least
   A. 2 years of age
   B. 4 years of age
   C. 6 years of age
   D. 8 years of age
   E. 10 years of age
5. A 13-month-old male presented with vomiting, fever, and irritability. On examination his temperature was 40\(^\circ\)c, lethargic, with atrophied tonsils, clear tympanic membrane, clear chest and neck stiffness. The parents gave past history of pneumonia at the age of 6 and 9 months for which he was admitted to hospital and treated with i.v. antibiotics for 10 days. Of the following, the MOST likely diagnosis is
   A. agammaglobulinemia
   B. common variable immunodeficiency
   C. transient hypogammaglobulinemia of infancy
   D. IgA deficiency
   E. hyper-IgM syndrome

6. An 8-month-old infant presented with chronic diarrhea, persistent right ear discharge, oral candidiasis responding poorly to antifungal drugs, and failure to thrive. On examination the infant has exfoliative erythroderma, lymphadenopathy, and hepatosplenomegaly. Peripheral blood showed eosinophilia and elevated IgE. The parents are young first cousin and he is their first baby. Of the following, the MOST likely diagnosis is
   A. DiGeorge anomaly
   B. Hyper-IgM syndrome
   C. Omenn syndrome
   D. Reticular dysgenesis
   E. Bare lymphocyte syndrome

7. DiGeorge syndrome, also known as velocardiofacial syndrome or CATCH 22 syndrome. One of the following IS NOT a feature of this syndrome
   A. cardiac anomalies
   B. abnormal facies
   C. thymic hypoplasia
   D. cleft lip
   E. hypocalcemia

8. A 15-month-old male child presented with thrombocytopenic purpura, atopic dermatitis, and recurrent infections caused by cytomegalovirus and P. jiroveci. Immunoglobulin assay revealed elevated IgE and IgA with decreased IgM levels. Of the following, the MOST likely diagnosis is
   A. Wiskott-Aldrich syndrome
   B. X-linked lymphoproliferative syndromes
   C. Nijmegen breakage syndrome
   D. Omenn syndrome
E. common variable immunodeficiency

9. An 8-year-old child having coarse facial features seen by dentist because of failure to shed primary teeth and visited an orthopedician due to frequent fractures of lower limbs, now presented with skin abscesses and eczema. Of the following, the **MOST** likely diagnosis is
   A. Nijmegen breakage syndrome
   B. Omenn syndrome
   C. common variable immunodeficiency
   D. hyper-IgE syndrome
   E. cartilage-hair hypoplasia (short limbed dwarf)

10. Neutropenia (for white children 1 year of age or older) is defined as an absolute neutrophil count (ANC) less than
   A. 2500/mm3
   B. 2000/mm3
   C. 1500/mm3
   D. 1000/mm3
   E. 500/mm3

11. A 15-month-old child presented with repeated skin and pulmonary infections started shortly after birth, his blood film showed severe neutropenia with impressive monocytosis, he is the third sibling of a first cousin healthy parents, the second sibling had the same problem and died due to acute myeloid leukemia. Of the following, the **MOST** likely diagnosis is
   A. cyclic neutropenia
   B. Kostmann syndrome
   C. Shwachman-Diamond syndrome
   D. benign congenital neutropenia
   E. autoimmune neutropenia

12. A 5-month-old infant presented with reluctance to feed, lethargy, abdominal distension, and severe gingivitis. The parents are healthy relatives and gave history of non-purulent omphalitis with separation of umbilical stump at the age of 2 months. His peripheral smear showed neutrophil count 24,000/mm3. Of the following, the **MOST** likely diagnosis is
   A. lazy leukocyte syndrome
   B. myeloperoxidase deficiency
   C. hyperimmunoglobulin E (Job syndrome)
   D. chronic granulomatous disease
13. Pancreatic insufficiency, metaphyseal chondrodysplasia, autosomal recessive inheritance, defective chemotaxis, and neutropenia are features of
   A. lazy leukocyte syndrome
   B. leukocyte adhesion deficiency
   C. Chédiak-Higashi syndrome
   D. chronic granulomatous disease
   E. Shwachman-Diamond syndrome

14. A 5-month-old infant presented with reluctance to feed, lethargy, abdominal distension, and severe gingivitis. The parents are healthy relatives and give history of non-purulent omphalitis with separation of umbilical stump at the age of 2 months. His peripheral smear showed neutrophil count 24,000/mm3.
   Of the following, the **MOST** effective life saving measure is
   A. i.v. antibiotics
   B. i.v.immunoglobulin
   C. stem cell transplantation
   D. integrins infusion
   E. selectins infusion

15. An 18-month-old male child presented with failure to thrive and chronic diarrhea. On examination he has dermatitis, generalized lymphadenopathy, hepatosplenomegaly, and deep seated infection in the inguinal area. His lab. investigations revealed anemia and hypergammaglobulinemia.
   Of the following, the **MOST** likely diagnosis is
   A. lazy leukocyte syndrome
   B. myeloperoxidase deficiency
   C. hyperimmunoglobulin E (Job syndrome)
   D. chronic granulomatous disease
   E. leukocyte adhesion deficiency

16. Viral infections associated with neutropenia include
   A. varicella
   B. parainfluenza
   C. respiratory syncyial
   D. hepatitis B
   E. mumps
17. Autosomal recessive inheritance; oculocutaneous albinism, neuropathy, giant neutrophilic cytoplasmic inclusions; malignancy, and neutropenia are features of one of the following phagocytic disorders
   A. chronic granulomatous disease
   B. Chédiak-Higashi syndrome
   C. hyperimmunoglobulin E (Job syndrome)
   D. glucose-6-phosphate dehydrogenase deficiency
   E. lazy leukocyte syndrome

18. Light microscopy of neutrophils for the presence of giant granules can help to diagnose
   A. chronic granulomatous disease
   B. Chédiak-Higashi syndrome
   C. hyperimmunoglobulin E (Job syndrome)
   D. glucose-6-phosphate dehydrogenase deficiency
   E. lazy leukocyte syndrome

19. C5, C6, C7, and C8 deficiency usually lead to
   A. recurrent meningococcal infections
   B. encapsulated bacterial infections
   C. glomerulonephritis
   D. atypical HUS
   E. hereditary angioedema

20. A 7-year-old child presented with severe stridor and abdominal pain, he gives history of recurrent episodes of nonpruritic edema lasting 48 to 72 hours, which usually triggered by stress, or anxiety.
Of the following, the MOST likely cause is deficiency of
   A. factor H
   B. factor I
   C. C1 inhibitor
   D. properdin
   E. C3

21. A 7-year-old child presented with severe stridor and abdominal pain, he gives history of recurrent episodes of nonpruritic edema lasting 48 to 72 hours, which usually triggered by stress, or anxiety.
Of the following, the MOST effective treatment is
   A. purified C1-inhibitor
   B. epinephrine
C. antihistamines  
D. corticosteroids  
E. fresh frozen plasma
1. (A). The innate immune system includes soluble factors, including acute-phase proteins, cytokines, chemokines, and complement, as well as cellular components, including neutrophils, monocytes/macrophages, and natural killer (NK) cells. The adaptive immune system is made up of T and B lymphocytes and their effector molecules.

2. (C). Failure to thrive, diarrhea, malabsorption, and infections with opportunistic infections (i.e., fungi, Candida sp, Pneumocystis jiroveci [carinii]) suggest T-cell immunodeficiency. Recurrent viral infections can result from T-cell or NK-cell deficiency. Deep-seated abscesses and infections with Staphylococcus aureus, Serratia marcescens, and Aspergillus suggest a disorder of neutrophil function, such as chronic granulomatous disease (CGD). Delayed separation of the umbilical cord, especially in the presence of omphalitis and later onset periodontal disease, in addition to poorly formed abscesses, indicates leukocyte adhesion deficiency.

3. (D).
   - Recurrent dermatologic infections with bacteria such as *Staphylococcus, Pseudomonas, and Escherichia coli*, and fungi such as *Aspergillus*
   - Subcutaneous, lymph node, lung, and liver abscesses
   - Pulmonary infections common, including abscess and pneumatocele formation, contributing to chronic disease
   - Bone and joint infection common
   - Delayed separation of umbilical cord
   - Absence of pus at site(s) of infection
   - Poor wound healing

All the above suggest neutrophil defects

4. (B). The diagnosis cannot be confirmed until the patient is at least 4 years of age when IgA levels should reach adult levels. Selective IgA deficiency occurs in approximately 1 in 500 individuals. Most patients with selective IgA deficiency are asymptomatic. In others it is associated with recurrent sinopulmonary infections, IgG2 subclass deficiency, specific antibody deficiency, food allergy, autoimmune disease, or celiac disease. IgA deficiency occurs in families, suggesting autosomal inheritance.

5. (A).
6. (C).
7. **(D).** CATCH 22 syndrome (cardiac anomalies, abnormal facies, thymic hypoplasia, cleft palate, and hypocalcemia), is the result of dysmorphogenesis of the third and fourth pharyngeal pouches, resulting in hypoplasia of the thymus required for T-cell maturation. Most, but not all, patients with DiGeorge syndrome have a defect on chromosome 22q11.2. DiGeorge syndrome is classically characterized by hypocalcemic tetany, conotruncal and aortic arch anomalies, and increased infections. The diagnosis is established by fluorescent in situ hybridization or a polymerase chain reaction with a DNA probe to detect deletions in chromosome 22q11.2.

8. **(A).** Wiskott-Aldrich syndrome is an X-linked disorder characterized by thrombocytopenia, eczema, defects in cell-mediated and humoral immunity, and a predisposition to lymphoproliferative disease. It is caused by mutations of the gene on chromosome Xp11.22 coding for the Wiskott-Aldrich syndrome protein (WASP), expressed in lymphocytes, platelets, and monocytes. Opportunistic infections and autoimmune cytopenias become problematic in older children. Isolated X-linked thrombocytopenia also results from mutations of the identical gene. One third of patients with Wiskott- Aldrich syndrome die as a result of hemorrhage, and two thirds die as a result of recurrent infection caused by bacteria, cytomegalovirus, P. jiroveci (carinii), or herpes simplex virus. Stem cell transplantation has corrected the immunologic and hematologic problems in some patients.

9. **(D).**

10. **(C).** The normal neutrophil count varies with age. Neutropenia is defined as an absolute neutrophil count (ANC) less than 1500/mm³ for white children 1 year of age or older. African American children normally have lower total white blood cell and neutrophil counts.

11. **(B).** Severe congenital neutropenia (Kostmann syndrome) is an autosomal recessive disorder in which myeloid cells fail to mature beyond the early stages of the promyelocyte due to mutations in the HAX-1 gene. The peripheral blood may show an impressive monocytosis.

12. **(E).** In leukocyte adhesion deficiency type I (LAD-I), infants lacking the β2 integrin CD18 exhibit the condition early in infancy with failure of separation of the umbilical cord (often 2 months after birth) with attendant omphalitis and sepsis. The neutrophil count usually is greater than 20,000/mm³ because of failure of the neutrophils to adhere normally to vascular endothelium and to migrate out of blood to the tissues. Cutaneous, respiratory, and mucosal infections occur. Children with this condition often have severe gingivitis. Sepsis usually leads to death in early childhood. This disorder is transmitted as an autosomal recessive trait. Stem cell transplantation can be lifesaving.
13. (E).
14. (C). Children with this condition often have severe gingivitis. Sepsis usually leads to death in early childhood. This disorder is transmitted as an autosomal recessive trait. Stem cell transplantation can be lifesaving.
15. (D). Patients characteristically have lymphadenopathy, hepatosplenomegaly, hypergammaglobulinemia, dermatitis, failure to thrive, anemia, chronic diarrhea, and abscesses. Infections occur in the lungs, the middle ear, gastrointestinal tract, skin, urinary tract, lymph nodes, liver, and bones. Granulomas are prominent and may obstruct the pylorus or ureters.
16. (D). Viral infections associated with neutropenia include
- Measles
- Hepatitis B
- HIV
- Rubella
- Cytomegalovirus
- Influenza
- Epstein-Barr virus
17. (B).
18. (B). Neutrophil chemotactic defects can be excluded by the presence of neutrophils at the site of infection. Flow cytometry for the presence of adhesion molecules, such as CD18 or CD15, can help diagnose leukocyte adhesion defects. CGD can be diagnosed by the flow cytometry-based test using dihydrorhodamine 123 or the nitroblue tetrazolium test. Light microscopy of neutrophils for the presence of giant granules can help diagnose Chédiak-Higashi syndrome.
19. (A).
20. (C).
- C1 inhibitor= Angioedema
- Factor H= Glomerulonephritis, atypical HUS
- Factor I= Recurrent infections, glomerulonephritis
- C3= Recurrent bacterial infections, rare glomerulonephritis, or SLE
- Properdin= Recurrent infections, severe meningococcal infection
21. (A). Prophylactic administration of fresh frozen plasma before surgery can prevent angioedema, but administration during an acute episode may exacerbate the episode. Angioedema of the airway can present as an acute emergency, necessitating a tracheostomy because administration of epinephrine, antihistamines, or corticosteroids is ineffective in reversing this type of angioedema. Purified C1-inhibitor is available and can be used prophylactically (before surgery) and during acute episodes of angioedema.
1. Type I Hypersensitivity reactions are triggered by the binding of antigen to high-affinity IgE receptors on the surface of tissue mast cells, circulating basophils, or both.

Of the following, a recognized example of type I reaction is

A. Rh hemolytic anemia
B. Goodpasture syndrome
C. allergic asthma
D. serum sickness
E. hypersensitivity pneumonitis

2. A classic example of type III (immune complex) reactions is

A. serum sickness
B. TB skin test reactions
C. contact dermatitis (neomycin)
D. graft versus-host disease
E. allergic rhinitis

3. A family history of allergic disease is often present in atopic patients. Multiple genes predispose to atopy. If one parent has allergies, the risk that a child will develop an allergic disease is

A. 15%
B. 25%
C. 35%
D. 50%
E. 75%

4. The prominent creases under the lower eyelids in an atopic child are called

A. allergic salute
B. allergic shiners
C. pityriasis alba
D. Dennie–Morgan folds
E. keratosis pilaris
5. Extreme eosinophilia suggests
   A. drug reaction
   B. allergic rhinitis
   C. atopic dermatitis
   D. asthma
   E. inflammatory bowel disease

6. In vivo skin testing for identifying allergen-specific IgE is done by introducing allergen into the skin via a prick/puncture or intradermal injection. This prompts the development of a central wheal and erythematous flare that are measured after the allergen has been placed by
   A. 15 to 20 minutes
   B. 30 to 60 minutes
   C. 12 to 24 hours
   D. 24 to 48 hours
   E. 48 to 72 hours

7. Disorders associated with elevated IgE include all the following EXCEPT
   A. atopic dermatitis (eczema)
   B. tissue-invasive helminthic infections
   C. Chediak-Higashi syndrome
   D. Hodgkin disease
   E. idiopathic nephrotic syndrome

8. All the following are features of in vivo skin testing for identifying allergen-specific IgE EXCEPT
   A. less expensive
   B. greater sensitivity
   C. wide allergen selection
   D. not suppressed by antihistamines
   E. results available immediately

9. One of the following is an aggravating factor for asthma
   A. gastroesophageal reflux
   B. viral infections
   C. smoke
   D. exercise
   E. emotions
10. One of the following monitor asthma and airway inflammation directly
   A. spirometry
   B. exhaled nitric oxide analysis
   C. radioallergosorbent test (RAST)
   D. fluorescent enzyme immunoassay (FEIA)
   E. enzymelinked immunosorbent assay (ELISA)

11. A chest radiograph should be performed with the first episode of asthma. Repeat chest radiographs are not needed with new episodes unless there is
   A. tachycardia
   B. poor air movement
   C. agitation
   D. pulsus paradoxus
   E. high grade fever

12. Inhaled corticosteroids are the most effective anti-inflammatory medications for the treatment of chronic, persistent asthma and are the preferred therapy when initiating long-term control therapy. Early intervention with inhaled corticosteroids have the following advantages EXCEPT
   A. reduces morbidity
   B. alter the natural history of asthma
   C. reduces airway hyperreactivity
   D. reduces the need for rescue bronchodilator therapy
   E. reduces the risk of death from asthma

13. Essential actions for reducing exposure to dust mites include
   A. encase pillow and mattress in allergen-impermeable encasement
   B. avoid sleeping or lying on upholstered furniture
   C. minimize number of stuffed toys in child’s bedroom
   D. reduce indoor humidity to <50%
   E. remove carpets from bedroom and play areas

14. Overuse of β2-agonists implies inadequate control; a change in medications may be warranted. Of the following, poor control is suggested by the
   A. use of more than 4 puffs per day
   B. use of more than 6 puffs per day
   C. use of more than 8 puffs per day
   D. use of more than 10 puffs per day
E. use of more than 12 puffs per day

15. A child with intermittent asthma has asthma symptoms less than
   A. two times per week
   B. three times per week
   C. four times per week
   D. five times per week
   E. six times per week

16. Daily long-term control therapy is recommended for infants and young children 0 to 4 years of age who had in the previous year
   A. two or more episodes of wheezing that lasted more than 1 day
   B. three or more episodes of wheezing that lasted more than 1 day
   C. four or more episodes of wheezing that lasted more than 1 day
   D. five or more episodes of wheezing that lasted more than 1 day
   E. six or more episodes of wheezing that lasted more than 1 day

17. The only inhaler corticosteroid (ICS) with FDA approved labeling for children <4 years of age is
   A. beclomethasone HFA
   B. budesonide nebulizer suspension
   C. mometasone DPI
   D. triamcinolone acetonide
   E. flunisolide HFA

18. Risk factors for persistent asthma include all the following EXCEPT
   A. atopic dermatitis
   B. allergic conjunctivitis
   C. elevated total serum IgE levels (first year of life)
   D. peripheral blood eosinophilia >4% (2–3 yr of age)
   E. food and inhalant allergen sensitization

19. The hallmarks of allergic rhinitis are the following EXCEPT
   A. thick rhinorrhea
   B. nasal congestion
   C. sneezing
   D. pruritus of the eyes, nose, and ears
   E. postnasal drip
20. The **MOST** common form of nonallergic rhinitis in children is
   A. noninfectious (vasomotor) rhinitis
   B. nonallergic rhinitis with eosinophilia
   C. physical rhinitis
   D. infectious rhinitis
   E. rhinitis medicamentosa

21. CHARGE association include all the following **EXCEPT**
   A. cataract
   B. congenital heart disease
   C. choanal atresia
   D. genitourinary defects
   E. ear anomalies

22. Nasal polyps are rare in children younger than 10 years of age but, if present, warrant evaluation for an underlying disease process, such as
   A. celiac disease
   B. ulcerative colitis
   C. unilateral choanal atresia
   D. primary ciliary dyskinesia
   E. bronchopulmonary dysplasia

23. Intranasal corticosteroids are the most potent pharmacologic therapy for treatment of allergic and nonallergic rhinitis. These topical agents work to reduce inflammation, edema, and mucus production but they are less helpful for symptoms of
   A. nasal congestion
   B. rhinorrhea
   C. itching
   D. sneezing
   E. conjunctival injection

24. Antihistamines are the medications used most frequently to treat allergic rhinitis. They are less helpful in treating
   A. sneezing
   B. rhinorrhea
   C. ocular itching
   D. nasal congestion
   E. nasal itching
25. The **MOST** common reported adverse effect of first-generation antihistamines is
   A. sedation
   B. blurred vision
   C. urinary retention
   D. dry mouth
   E. tachycardia

26. Nasal congestion of rhinitis is usually relieved by
   A. diphenhydramine
   B. hydroxyzine
   C. pseudoephedrine
   D. cetirizine
   E. loratadine

27. Children with allergic rhinitis have symptoms of reactive airways disease/asthma in approximately
   A. 30%
   B. 40%
   C. 50%
   D. 60%
   E. 70%

28. Approximately 35% to 40% of infants and young children with moderate to severe atopic dermatitis have coexisting food allergies. The most common cause of food-induced eczematous reactions is allergy of
   A. wheat
   B. egg
   C. fish
   D. cow milk protein
   E. soy protein

29. The clinical manifestations of atopic dermatitis vary with age. In infants, atopic dermatitis usually spares
   A. face
   B. scalp
   C. cheeks
   D. diaper area
   E. extensor surfaces of the extremities
30. Many conditions share signs and symptoms of atopic dermatitis, differential diagnosis of atopic dermatitis include all the following metabolic disorders EXCEPT
   A. zinc deficiency
   B. pyridoxine deficiency
   C. niacin deficiency
   D. multiple carboxylase deficiency
   E. tuberous sclerosis

31. Topical anti-inflammatory agents, including corticosteroids and immunomodulators, are the cornerstone of therapy for acute flares and prevention of relapses in atopic dermatitis.
   All the following statements are true EXCEPT
   A. Ointments generally are preferred over creams and lotions
   B. The least potent corticosteroid that is effective should be used
   C. Low-potency, nonfluorinated corticosteroids should be used on the face
   D. Reduced efficacy of topical corticosteroids usually related to glucocorticoid resistance
   E. Higher potency corticosteroids may be necessary to diminish the dermatitis flare but should be used for limited periods

32. More than 90% of patients with atopic dermatitis have colonization of lesional skin with Staphylococcus aureus, and uninvolved skin has colonization of more than
   A. 5%
   B. 15%
   C. 25%
   D. 50%
   E. 75%

33. An important step in the management of atopic dermatitis is to identify and avoid allergens and irritants.
   Of the following, a COMMON irritant is
   A. soaps
   B. wool
   C. food allergen
   D. dust mites
   E. pollens
34. What is the **MOST** common physical urticaria?
   A. dermatographism  
   B. cholinergic urticaria  
   C. cold urticaria  
   D. delayed pressure urticaria  
   E. solar urticaria

35. The absence of dermal symptoms does not exclude the diagnosis of anaphylaxis, because patients who present with cutaneous symptoms (urticaria, angioedema, flushing, and warmth) are
   A. 50%  
   B. 60%  
   C. 70%  
   D. 80%  
   E. 90%

36. Avoidance of triggering agents is important in the management of urticaria and angioedema.
   Of the following, the **MAINSTAY** of pharmacologic treatment is
   A. H₂ antihistamines  
   B. tricyclic antidepressants  
   C. corticosteroids  
   D. leukotriene receptor blocker  
   E. second-generation H₁ antihistamines

37. Anaphylaxis is a medical emergency; prompt recognition and immediate treatment are crucial.
   Of the following, the **MAINSTAY** of therapy is early administration of
   A. corticosteroids  
   B. antihistamines  
   C. H₂-receptor antagonists  
   D. intramuscular epinephrine  
   E. bronchodilators

38. The **MOST** common medication frequently implicated in serum sickness is
   A. gentamycin  
   B. ceftiazone  
   C. penicillin  
   D. carbamazepine
E. furosemide

39. Food allergy or hypersensitivity reactions are the result of immune reactions to glycoproteins and develop in genetically predisposed individuals. In older children and adults, the item which account for most reactions is
   A. peanuts
   B. cow’s milk
   C. eggs
   D. soybean
   E. wheat

40. Most food allergy or hypersensitivity reactions typically outgrown, those that persist include
   A. milk
   B. soy
   C. egg
   D. shellfish
   E. wheat

41. The standard method for diagnosis of food allergy which can be performed to determine whether a child can eat the food safely is
   A. oral food challenges
   B. serum specific IgE assay
   C. prick skin test
   D. patch testing
   E. endoscopy and biopsy

42. Drug reactions to penicillins and cephalosporins are the most common allergic drug reactions encountered in the pediatric population. Risk factors for drug reactions include all the following EXCEPT
   A. previous drug exposure
   B. increasing age (>20 years of age)
   C. higher dose
   D. intermittent repeated exposure
   E. atopic background

43. Tinnitus after a single, small dose of aspirin is an example of the following adverse drug reactions
   A. pseudoallergic
B. idiosyncratic
C. intolerance
D. pharmacologic adverse effect
E. drug toxicity

44. Skin testing protocols are standardized for penicillin and are well described for other agents, such as local anesthetics, muscle relaxants, vaccines, and insulin. Penicillin skin testing is helpful for
   A. patients in acute need of treatment
   B. IgE-mediated reactions
   C. non-IgE-mediated reactions
   D. patients with a history consistent with serum sickness
   E. patients with a history consistent with desquamative-type reactions

45. The risk of a child who has reacted positively to penicillin skin testing suffering an allergic reaction to a cephalosporin antibiotic is less than
   A. 2%
   B. 4%
   C. 6%
   D. 10%
   E. 20%
1. (C). Anaphylaxis, urticaria, allergic rhinitis, and allergic asthma are examples of Type I reactions.

2. (A). The administration of large amounts of antigen leads to serum sickness, a classic example of a type III reaction. Other type III–mediated reactions include hypersensitivity pneumonitis and some vasculitic syndromes.

3. (B). The risk that a child will develop an allergic disease is 25%. If both parents have allergies, the risk increases to 50% to 70%. Similar atopic diseases tend to occur in families.

4. (D). Dermatologic findings of atopy include hyperlinearity of the palms and soles, white dermatographism, pityriasis alba, prominent creases under the lower eyelids (Dennie–Morgan folds or Dennie lines), and keratosis pilaris (asymptomatic horny follicular papules on the extensor surfaces of the arms).

5. (A). Extreme eosinophilia suggests a nonallergic disorder such as infections with tissue-invasive parasites, drug reactions, or malignancies.

6. (A).

7. (C). Other disorders associated with elevated IgE include
   - allergic disease
   - hyperimmunoglobulin-e syndromes
   - allergic bronchopulmonary aspergillosis
   - Wiskott–Aldrich syndrome
   - bone marrow transplantation
   - bullous pemphigoid

8. (D). Antihistamines may interfere with skin test results.

9. (A). Exacerbating factors include viral infections, exposure to allergens and irritants (e.g., smoke, strong odors, fumes), exercise, emotions, and change in weather/humidity. Nighttime symptoms are common. Rhinosinusitis, gastroesophageal reflux, and nonsteroidal anti-inflammatory drugs (especially aspirin) can aggravate asthma.

10. (B). Two novel forms of monitoring asthma and airway inflammation directly include exhaled nitric oxide analysis and quantitative analysis of expectorated sputum for eosinophilia.

11. (E). Repeat chest radiographs are not needed with new episodes unless there is fever (suggesting pneumonia) or localized findings on physical examination.
12. (B). Early intervention with inhaled corticosteroids reduces morbidity but does not alter the natural history of asthma.
13. (A). The other distracters are desirable actions and not essential.
14. (C). Use of more than one metered dose inhaler canister per month or more than eight puffs per day suggests poor control.
15. (A).
16. (C).
17. (B). For children < 4 years of age: The safety and efficacy of ICSs in children < 1 year has not been established. Children < 4 years of age generally require delivery of ICS (budesonide and fluticasone HFA) through a face mask that should fit snugly over nose and mouth and avoid nebulizing in the eyes. Wash face after each treatment to prevent local corticosteroid side effects.
18. (B). Allergic rhinitis.
20. (D).
22. (D). Cystic fibrosis and primary ciliary dyskinesia.
23. (E).
24. (D).
25. (A).
26. (C). Decongestants, taken orally or intranasally, may be used to relieve nasal congestion. Oral medications, such as pseudoephedrine and phenylephrine, are available either alone or in combination with antihistamines.
27. (D).
28. (B).
29. (D).
31. (D). Reduced efficacy of topical corticosteroids may be related to disease severity rather than glucocorticoid resistance.
32. (E).
33. (A). Common irritants include soaps, detergents, fragrances, chemicals, smoke, and extremes of temperature and humidity.
34. (A). Dermatographism means “writing on the skin” and is easily diagnosed by firmly scratching the skin with a blunt point, such as the wooden tip of a cotton swab or tongue depressor. It is characterized by an urticarial reaction localized to the site of skin trauma. It has been suggested that trauma induces an IgE-mediated reaction causing histamine to be released from the mast cells.
35. (E).
36. (E).
37.(D). Early administration of intramuscular epinephrine is the mainstay of therapy and should be given at the same time that basic measures of cardiopulmonary resuscitation are being performed.

38.(C). Common inciting agents include blood products and foreign proteins, such as antithymocyte globulin and antivenoms. Medications frequently implicated include penicillin, sulfonamides, minocycline, cefaclor, hydantoins, and thiazides.

39.(A). In children, cow’s milk, eggs, peanuts, soybean, wheat, tree nuts, fish, and shellfish cause 90% of IgE-mediated reactions. In older children and adults, peanuts, tree nuts, fish, and shellfish account for most reactions.

40.(D). Milk, soy, egg, and wheat typically outgrown while peanut, tree nuts, seeds, and shellfish typically persist.

41.(A). Oral food challenges remain the standard of diagnosis and can be performed to determine whether a child can eat the food safely.

42.(E). An atopic background does not predispose an individual to the development of drug reactions but may indicate a greater risk of serious reaction.

43.(C).
- Pseudoallergic = Anaphylactoid reaction after radiocontrast media
- Idiosyncratic = Hemolytic anemia in a patient with G6PD deficiency after primaquine therapy
- Intolerance = Tinnitus after a single, small dose of aspirin
- Drug toxicity = Hepatotoxicity from methotrexate
- Pharmacologic adverse effect = Dry mouth from antihistamines; tremor from albuterol

44.(B). Penicillin allergy should be evaluated when the individual is well and not in acute need of treatment. Penicillin skin testing is helpful for IgE-mediated reactions because of its negative predictive value; only 1% to 3% of patients with negative skin tests have a reaction, which is mild, when re-exposed to penicillin. Skin testing for penicillin should be performed using the major determinant, penicilloyl polyllysine (available as Pre-Pen), and minor determinants, which include penicillin G, penicilloate, and penilloate. Skin testing to penicillin does not predict non-IgE-mediated reactions. For patients with a history consistent with serum sickness or desquamative-type reactions, skin testing should not be performed, and penicillin should be avoided indefinitely.

45.(A). It is believed that the first-generation cephalosporins (e.g., cephalexin) are more likely than second-generation (e.g., cefuroxime) or third-generation (cefpodoxime) cephalosporins to be cross-reactive. This is due to the chemical similarity of side chains of the β-lactam ring between penicillin and first-generation cephalosporins.
1. The rheumatic diseases of childhood are characterized by autoimmunity and inflammation, which may be localized or generalized.

Of the following, the classical rheumatic disease of childhood is

A. scleroderma
B. Behçet syndrome
C. juvenile rheumatoid arthritis
D. Sjögren syndrome
E. all the above

2. Fever in rheumatic diseases is caused by cytokine release and can take many forms. Regular temperature spikes, once or twice a day is seen in

A. scleroderma
B. Behçet syndrome
C. systemic-onset juvenile rheumatoid arthritis
D. Sjögren syndrome
E. Kawasaki disease

3. Morning stiffness is seen in which of the following pediatric diseases?

A. rheumatic fever
B. systemic lupus erythematosus
C. leukemia
D. gonococcemia
E. Kawasaki disease

4. Erythema marginatum is seen in which of the following pediatric arthritis syndromes?

A. rheumatic fever
B. systemic lupus erythematosus
C. lyme disease
D. gonococcemia
E. Kawasaki disease

5. Erosive arthritis in the following pediatric arthritis syndromes, is mostly seen in

A. systemic lupus erythematosus
6. A 14-year-old adolescent girl presented with palpable purpuric rash on the buttocks and lower extremities with arthritis of ankles and knees. Investigation as follow; erythrocyte sedimentation rate, C-reactive protein, and white blood cell count are elevated, the platelet count is normal, urinalysis show evidence of hematuria.

Of the following, the **MOST** common immune complex associated with this disease is

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

7. Henoch-Schönlein purpura (HSP) is the **MOST** common systemic vasculitis of childhood.

Arthritis occurs in

A. 20%  
B. 40%  
C. 60%  
D. 80%  
E. 100%

8. Anti-neutrophil cytoplasmic antibody-associated vasculitis is seen in all the following **EXCEPT**

A. wegener granulomatosis  
B. polyarteritis nodosa  
C. Churg-Strauss syndrome  
D. microscopic polyangiitis  
E. Henoch-Schönlein purpura

9. A 3-year-old-boy presented with fever 6 days ago associated with periungual desquamation. Examination showed a strawberry tongue, cervical lymphadenopathy, truncal polymorphous rash.

The rash in this patient occurs in
A. 20%
B. 40%
C. 60%
D. 80%
E. 100%

10. Criteria for diagnosis of Kawasaki disease include all the following EXCEPT
A. fever of >5 days duration
B. bilateral suppurative conjunctivitis
C. periungual desquamation
D. dry fissured lips
E. polymorphous rash

11. An 8-year-old girl complains from morning stiffness and pain of 4 joints in the last 6 months with uveitis. On physical examination, signs of inflammation are present, including joint tenderness, erythema, and effusion, but no fever or weight loss. Laboratory result was: ANA positive, rheumatoid factor negative. Of the following, the MOST commonly involved joint is
A. knee
B. ankle
C. hip
D. wrist
E. toe

12. The MOST common form of juvenile idiopathic arthritis (JIA) is
A. polyarticular juvenile idiopathic arthritis
B. oligoarticular juvenile idiopathic arthritis
C. systemic-onset juvenile idiopathic arthritis
D. juvenile ankylosing spondylitis
E. psoriatic arthritis

13. The MOST common radiologic finding in the early stages of juvenile idiopathic arthritis (JIA) is
A. fusion of C1-4
B. erosions of bony articular surfaces
C. normal bone x-ray
D. evidence of bony proliferation
E. periarticular osteopenia
14. The treatment of juvenile idiopathic arthritis (JIA) focuses on suppressing inflammation, preserving and maximizing function, preventing deformity, and preventing blindness.

Of the following, the **FIRST** option in the treatment of JIA is

A. systemic corticosteroid
B. intra-articular corticosteroid
C. hydroxychloroquine
D. nonsteroidal anti-inflammatory drug
E. infliximab

15. Of the following rheumatic diseases of childhood, shawl sign is seen in

A. Kawasaki disease
B. juvenile dermatomyositis
C. Henoch-Schönlein purpura
D. systemic lupus erythematosus
E. systemic-onset juvenile idiopathic arthritis

16. A 9-year-old girl has a raised erythematous rash on the cheeks; effusion and tenderness of the small joints of the hands; with history of recurrent mouth and nasal sores, GUE show proteinuria (>500 mg/24 h). All the following hematologic abnormalities are suspected in this patient **EXCEPT**

A. hemolytic anemia with reticulocytosis
B. leukopenia (<4000 on two occasions)
C. low levels of C3 and normal C4
D. lymphopenia (<1500 on two occasions)
E. thrombocytopenia (<100,000/mm3)

17. A 10-year-old female with history of insidious onset of fatigue, malaise, and progressive muscle weakness, accompanied by low-grade fevers and scaly, red plaques are found across the knuckles, she is not able to sit up from a supine position, investigations revealed: elevated muscle enzymes (ALT, AST, LDH, CPK, and aldolase), EMG abnormalities: fasciculations, needle insertion irritability, and high-frequency discharges.

Of the following, the **MOST** serious complication that can occurs in this patient is the development of

A. calcinosis
B. infections
C. avascular necrosis
D. myocardial infarction
E. accelerated atherosclerosis

18. The musculoskeletal pain syndromes that is **MOSTLY** associated with mitral valve prolapse is
   A. fibromyalgia
   B. growing pains
   C. benign hypermobility
   D. Ehlers-Danlos syndrome
   E. myofascial pain syndromes
1. (C). The classic rheumatic diseases of children include juvenile idiopathic arthritis (JIA), formerly called juvenile rheumatoid arthritis, systemic lupus erythematosus (SLE), and juvenile dermatomyositis (JDM). Other distracters are rare in childhood.

2. (C). While hectic fever, without periodicity or pattern, is commonly found in vasculitides such as Kawasaki disease but also occurs in children with underlying infection. Other rheumatic illnesses cause low-grade fevers.

3. (B). Morning stiffness is seen in systemic lupus erythematosus and juvenile idiopathic arthritis.

4. (A).
   - Systemic lupus erythematosus: butterfly; discoid rash
   - Lyme disease: erythema migrans
   - Gonococcemia: palms/soles, papulopustules
   - Kawasaki disease: diffuse maculopapular (nonspecific), desquamation

5. (E).

6. (A). Henoch-Schönlein purpura (HSP) is a vasculitis of unknown etiology characterized by inflammation of small blood vessels with leukocytic infiltration of tissue, hemorrhage, and ischemia. Diagnostic biopsy reveals granulocytes in the walls of arterioles or venules and IgA deposits in vessel wall.

7. (D). Arthritis occurs in 80% of patients with HSP and is most common in the lower extremities, particularly the ankles and knees. The arthritis is acute and very painful with refusal to bear weight. Joint swelling can be confused with peripheral edema seen with the rash of HSP.

8. (E).

9. (D). Kawasaki disease (KD) is a vasculitis of unknown etiology that is characterized by multisystem involvement and inflammation of small to medium-sized arteries with resulting aneurysm formation. A rash, which can vary in appearance, occurs in 80% of children with KD and may be particularly accentuated in the inguinal area and on the chest.

10. (B). Bilateral nonsuppurative conjunctivitis.

11. (A). Oligoarticular juvenile idiopathic arthritis is defined as the presence of arthritis in fewer than five joints within 6 months of diagnosis. The arthritis is found in medium-sized to large joints; the knee is the most common joint involved, followed by the ankle and the wrist. It is unusual for small joints, such as the fingers.
or toes, to be involved, although this may occur. Neck and hip involvement is uncommon.

12.(B). This is the most common form of JIA, accounting for approximately 50% of cases.

13.(C). Over time, periarticular osteopenia, resulting from decreased mineralization, is most commonly found. Growth centers may be slow to develop, whereas there may be accelerated maturation of growth plates or evidence of bony proliferation. Erosions of bony articular surfaces may be a late finding. If the cervical spine is involved, fusion of C1-4 may occur, and atlantoaxial subluxation may be demonstrable.

14.(D). Naproxen, sulindac, ibuprofen, indomethacin, and others have been used successfully. Systemic corticosteroid medications, such as prednisone and prednisolone, should be avoided but reserved for the most extreme circumstances.

15.(B). A classic juvenile dermatomyositis rash occurs on the face and across the cheeks but also can be found on the shoulders and back (shawl sign).

16.(C). This is a case of systemic lupus erythematosus. Excessive circulating antibodies and immune complexes also lead to the consumption of complement proteins, with low levels of C3 and C4 and decreased complement function as measured by CH50.

17.(A). This is a case of juvenile dermatomyositis. Other distracters are long-term complications of systemic lupus erythematosus.

18.(D). Excessive skin elasticity, easy bruisability, or mitral valve prolapse suggests Ehlers-Danlos syndrome or Marfan syndrome.
1. A 16-month-old infant present with high fever, cough, coryza, and conjunctivitis; after 3 days a macular rash begins on the head and spreads over most of the body. On examination; cervical lymphadenopathy, splenomegaly, and temperature 40°C. CBC reveals WBC count 3000/mm³

The severity of this illness is **MOST** likely related to the extent of

- A. rash
- B. fever
- C. cough
- D. leukopenia
- E. splenomegaly

2. In acute measles, the World Health Organization recommends routine administration for 2 days of

- A. zinc
- B. folic acid
- C. vitamin A
- D. vitamin C
- E. vitamin D

3. The **MOST** common complication of measles infection is

- A. otitis media
- B. myocarditis
- C. encephalomyelitis
- D. interstitial pneumonia
- E. subacute sclerosing panencephalitis

4. What is the **MOST** common site of skeletal tuberculosis?

- A. spine
- B. femur
- C. hip joint
- D. humerus
- E. knee joint
5. Ali’s mother worried about her 23-month-old baby who has chronic renal failure, his sister developed measles 2 days ago.
Of the following, the **MOST** appropriate action is
   A. isolation
   B. reassurance
   C. measles vaccine within 72 hr
   D. immunoglobulin within 12 days
   E. serologic test for IgM antibodies

6. A 3-year-old child present with anorexia, malaise, low-grade fever, and maculopapular rash begins on the face and spreads to the body for 3 days. On examination there is retroauricular and posterior occipital lymphadenopathy. CBC show WBC, 4500/mm$^3$ and platelet count, 110000/mm$^3$.
Of the following, the **MOST** likely diagnosis is
   A. rubella
   B. roseola
   C. measles
   D. scarlet fever
   E. Kawasaki disease

7. Hind’s mother brought her 2-year-old daughter to you in the primary health center, Hind’s aunt developed rubella 2 weeks ago and they visited her yesterday. Hind’s mother worried about herself and her daughter.
Of the following, the **BEST** advice to Hind’s mother is
   A. isolation
   B. reassurance
   C. rubella vaccine within 72 hr
   D. immunoglobulin within 4 days
   E. serologic test for IgM antibodies

8. A 9-month-old infant have remittent fever for the last 4 days with nasal congestion and mild cough, today he developed maculopapular rash after subsidence of the fever.
Of the following, the **MOST** likely diagnosis is
   A. rubella
   B. roseola
   C. measles
   D. scarlet fever
   E. Kawasaki disease
9. A 6-year-old child present with low grade fever, pharyngitis and mild conjunctivitis. After about one week he developed facial rash with circumoral pallor followed by a pruritic reticulated rash over the body that waxes and wanes (recur with exercise and bathing).
Of the following, the **MOST** likely cause is
   A. adenovirus
   B. measles virus
   C. parvovirus B19
   D. Epstein-Barr virus
   E. herpesvirus type 6

10. The treatment of latent tuberculosis is
   A. observation
   B. isoniazid only
   C. rifampicin only
   D. rifampicin and isoniazid
   E. rifampicin, isoniazid, and pyrazinamide

11. A 6-year-old boy present with fever, malaise, anorexia, and rash which appear as small red papules and vesicles on an erythematous base; which ulcerate and crusted mainly on the trunk, head, and face.
Of the following, the **LEAST** common neurologic complication of this disease is
   A. tremor
   B. nystagmus
   C. encephalitis
   D. cerebellar ataxia
   E. optic neuritis

12. A 7-year-old child, a known case of chronic eczema, exposed to chickenpox 2 days ago.
Of the following, the **MOST** appropriate action is
   A. observation for 2wk
   B. VZIG administration
   C. acyclovir administration
   D. varicella vaccine administration
   E. serologic test for IgM antibodies

13. In chickenpox, acyclovir is considered in all the following **EXCEPT**
   A. chronic cutaneous disease
B. chronic pulmonary disease  
C. short course corticosteroids  
D. long-term salicylate therapy  
E. unvaccinated persons < 12 yr

14. In meningitis, the characteristic sign of increased intracranial pressure with brain herniation is  
A. vomiting  
B. headache  
C. papilledema  
D. bulging fontanelle  
E. bradycardia with hypertension

15. A 5-year-old child (who had neurosurgical procedure before 1 month) presented with fever, headache, repeated vomiting, and nuchal rigidity; Kernig and Brudzinski are positive; cerebrospinal fluid findings are (leukocytes 550/µL with PMNs predominate), protein (120 mg/dL), and glucose (44 mg/dL); serum glucose (118 mg/dL). Of the following, the MOST common organism that cause this condition is  
A. Staphylococcus  
B. Neisseria meningitidis  
C. Haemophilus influenzae  
D. Pseudomonas aeruginosa  
E. Streptococcus pneumoniae

16. Of the following, the LEAST common cause of neonatal bacterial meningitis is  
A. Klebsiella  
B. Enterobacter  
C. Escherichia coli  
D. Group B streptococcus  
E. Listeria monocytogenes

17. Dexamethasone used as adjunctive therapy in bacterial meningitis to diminish the incidence of hearing loss and neurologic deficits resulting from  
A. Listeria monocytogenes  
B. Haemophilus influenzae type b  
C. Gram-negative bacilli  
D. Neisseria meningitidis  
E. Streptococcus pneumoniae
18. A 9-year-old child presented with history of fall from a height, he developed otorrhea and basilar skull fracture; 2 wk later he developed fever, headache, repeated vomiting, and nuchal rigidity; cerebrospinal fluid findings are (leukocytes 1600/µL with PMNs predominate), protein (140 mg/dL), and glucose (39 mg/dL); serum glucose (109 mg/dL).

Of the following, the **MOST** common organism causing this condition is
- A. Staphylococcus
- B. Neisseria meningitidis
- C. Haemophilus influenzae
- D. Pseudomonas aeruginosa
- E. Streptococcus pneumoniae

19. A 4-year-old child presented with history of fever, headache, lethargy, abnormal behavior, and seizures; CSF examination reveal (leukocytes 350/µL with mononuclear cells predominate), protein (70 mg/dL), and glucose (69 mg/dL); serum glucose (98 mg/dL); brain imaging show temporal lobe abnormalities.

Of the following, the **MOST** likely cause of this condition is
- A. Adenovirus
- B. Enterovirus
- C. Haemophilus influenzae
- D. Epstein-Barr virus
- E. Herpes simplex virus

20. Of the following, the **MOST** common cause of acute encephalitis is
- A. Enterovirus
- B. Adenovirus
- C. Epstein-Barr virus
- D. Herpes simplex virus
- E. Human immunodeficiency virus

21. All the following matching regarding prevention of encephalitis are true **EXCEPT**
- A. enteroviral encephalitis -- no action
- B. influenza encephalitis ----- influenza vaccination
- C. rabies ----------- preexposure or postexposure vaccination
- D. arboviral encephalitis ---------- avoid mosquito-borne exposures
- E. HSV encephalitis in infant of mother with active genital lesion --- IV acyclovir

22. Of the following, the **MOST** common viral cause of common cold is
- A. influenza
23. Allergic rhinitis is characterized by all the following EXCEPT
   A. nasal polyps
   B. allergic shiners
   C. absence of fever
   D. transverse crease on the nasal bridge
   E. swollen, erythematous nasal turbinates

24. Of the following, the MOST common bacterial cause of pharyngitis is
   A. Francisella tularensis
   B. group A streptococcus
   C. group C streptococcus
   D. Chlamydia pneumoniae
   E. Arcanobacterium haemolyticum

25. The incidence of streptococcal pharyngitis increased in
   A. toddlers
   B. preschool-age children
   C. school-age children
   D. late adolescence
   E. adulthood

26. A 16-year-old adolescent presented with history of acute pharyngitis; followed by thrombosis of the internal jugular vein and septic pulmonary embolism. Throat culture reveals Fusobacterium necrophorum. Of the following, the MOST likely diagnosis is
   A. trench mouth
   B. ludwig angina
   C. vincent angina
   D. vincent infection
   E. Lemierre syndrome

27. In acute pharyngitis, antibiotic therapy should be started promptly in children with all the following EXCEPT
   A. scarlet fever
B. past family history of rheumatic fever
C. positive rapid test for group A streptococcus
D. symptomatic pharyngitis whose sibling has documented streptococcal pharyngitis
E. symptomatic pharyngitis and living in an area experiencing an epidemic of acute rheumatic fever or poststreptococcal glomerulonephritis

28. Regarding paranasal sinuses, all the following are true EXCEPT
   A. ethmoidal sinuses are pneumatized at birth
   B. sphenoid sinuses are present by 5 years of age
   C. frontal sinuses begin to develop at 7 years of age
   D. maxillary sinuses are pneumatized at 2 years of age
   E. frontal sinuses are not completely developed until adolescence

29. A 7-year-old child presented with persistent mucopurulent rhinorrhea, nasal stuffiness, headache and cough, mainly at night preceded by history flue like illness; on examination there are facial swelling and tenderness. Of the following, the major predisposing factor for the development of this condition is
   A. allergy
   B. nasal polyps
   C. cystic fibrosis
   D. common cold
   E. nasal foreign body

30. The first-line therapy of sinusitis in children is
   A. cefixime
   B. levofloxacin
   C. clindamycin
   D. cefpodoxime
   E. amoxicillin-clavulanate

31. In pulmonary tuberculosis, most infectious patients become noninfectious after starting effective treatment within
   A. 2 weeks
   B. 3 weeks
   C. 4 weeks
   D. 5 weeks
   E. 6 weeks
32. The region of lymphadenopathy that's affected by tuberculosis and known as Ghon complex is
   A. hilar
   B. axillary
   C. cervical
   D. supraclavicular
   E. submandibular

33. A 16-month-old child presented with fever, poor feeding, and ear discomfort which interfere with normal sleep but you are an uncertain about the diagnosis of acute otitis media.
   Of the following, the first-line therapy of this condition is
   A. observation
   B. cefdinir
   C. amoxicillin
   D. ceftriaxone
   E. amoxicillin-clavulanate

34. A 7-year-old boy presented with earache and aural discharge mainly in left side, he recently was diving in swimming pool; on examination he is afebrile with tenderness on movement of the pinna.
   Of the following, the MOST likely diagnosis is
   A. otitis media
   B. otitis externa
   C. cholesteatoma
   D. acute mastoiditis
   E. otitis media with effusion

35. A 7-year-old boy presented with earache and aural discharge mainly in left side, he recently was diving in swimming pool; on examination he is afebrile with tenderness on movement of the pinna.
   Of the following, the MOST likely cause of this condition is
   A. Moraxella catarrhalis
   B. group A streptococcus
   C. Pseudomonas aeruginosa
   D. Streptococcus pneumonia
   E. nontypable Haemophilus influenzae
36. A 2-year-old child presented with recurrent attacks of stridor and harsh cough, sudden in onset mainly at night with no fever; most of the time the stridor relieved during the trip to hospital. Of the following, the **MOST** likely diagnosis is
   A. epiglottitis
   B. vascular ring
   C. spasmodic croup
   D. subglottic stenosis
   E. laryngotracheobronchitis

37. The **MOST** common cause of laryngotracheobronchitis (croup) is
   A. adenoviruses
   B. parainfluenza viruses
   C. group A streptococcus
   D. respiratory syncytial virus
   E. Haemophilus influenza type b

38. A 16-month-old child presented with history of a flu-like illness followed by harsh cough, hoarseness, inspiratory stridor, low-grade fever, and respiratory distress. Of the following, the **MOST** characteristic radiographic finding is
   A. thumb sign
   B. steeple sign
   C. hyperinflated lung
   D. bilateral lung infiltrate
   E. ground glass appearance

39. Treatment with erythromycin in the paroxysmal stage of pertussis
   A. is of no benefit
   B. lessen symptom severity
   C. shorten the course of illness
   D. decreases the potential for spread to others
   E. eradicates nasopharyngeal carriage of organisms

40. A 3-year-old child presented with increased nasal secretions and low-grade fever for 1 week followed by paroxysmal coughing with post tussive emesis, the child was vaccinated with 1st 2 doses at one week and two month. Of the following, the **MOST** helpful diagnostic test for this condition is
   A. serologic tests
B. lymphocytosis
C. chest radiograph
D. polymerase chain reaction
E. direct fluorescent antibody staining

41. Regarding complications of pertussis, the MOST permanent disability is a result of
   A. seizures
   B. pneumonia
   C. otitis media
   D. malnutrition
   E. encephalopathy

42. Of the following, the LEAST frequent cause of bronchiolitis is
   A. coronaviruses
   B. parainfluenza viruses
   C. Mycoplasma pneumoniae
   D. respiratory syncytial virus
   E. human metapneumovirus

43. Monthly injections of palivizumab, a RSV-specific monoclonal antibody, initiated just before the onset of the RSV season confers some protection from severe RSV disease. Palivizumab is indicated for some infants under 2 years old with all the following EXCEPT
   A. asthma
   B. chronic lung disease
   C. very low birth weight
   D. hemodynamically significant cyanotic CHD
   E. hemodynamically significant acyanotic CHD

44. A 4-year-old child admitted to hospital with history of high fever, cough, and tachypnea; chest radiograph reveals lobar pneumonia treated with I.V. ceftriaxone with partial response; 7 days later, the fever recurs with toxicity, new chest radiograph reveals cavitary lesion with an air-fluid level. Of the following, the BEST management is
   A. to order CT scan
   B. to add vancomycin
   C. surgical interference
   D. to add clindamycin
E. to continue on the same antibiotic for 14 days

45. A 14-year-old adolescent boy presented with malaise, dry cough, and mild shortness of breath for 1 week. Frontal chest radiograph shows a diffuse pattern of increased interstitial markings, including Kerley lines. Of the following, the **MOST** likely micro-organism causing this condition is
   A. Chlamydia trachomatis
   B. Ureaplasma urealyticum
   C. Mycoplasma pneumoniae
   D. Respiratory syncytial virus
   E. Streptococcus pneumoniae

46. A 6-year-old child presented with high fever, cough, and chest pain. Frontal chest radiograph shows airspace consolidation, which obliterates the silhouette of the heart border on the left. Of the following, the recommended treatment is
   A. amoxicillin
   B. ceftriaxone
   C. clindamycin
   D. vancomycin
   E. azithromycin

47. The **MOST** common etiologic agent in a 4-year-old child with pneumonia is
   A. influenza viruses
   B. human metapneumovirus
   C. Respiratory syncytial virus
   D. Streptococcus pneumoniae
   E. Haemophilus influenzae (type b)

48. A 6-year-old child with surgically repaired cyanotic CHD with prosthetic valve presented with sudden attack of unexplained fever, malaise, and weight loss; on examination there are palpable spleen, tachycardia, and a new heart murmur. Of the following, the **MOST** likely etiologic pathogen of this condition is
   A. Viridans streptococci
   B. Staphylococcus aureus
   C. Streptococcus pneumoniae
   D. Gram-negative enteric bacilli
   E. Hemolytic streptococci group A
49. In high-risk patients, to prevent infective endocarditis, prophylactic antibiotics are required before and during all dental procedures and invasive procedures of the respiratory tract, infected skin, or muscle.

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

A. amoxicillin  
B. ceftriaxone  
C. cindamycin  
D. azithromycin  
E. benzathine penicillin

50. Modified Duke clinical criteria for diagnosis of infective endocarditis are two major criteria or one major and three minor criteria or five minor criteria.

Of the following, a major criteria is

A. Osler nodes  
B. fever >38°C  
C. Janeway lesions  
D. new valvular regurgitation  
E. predisposing heart condition

51. One of the mechanisms of infectious diarrhea is increasing cyclic adenosine monophosphate levels which mainly occurred by

A. Shigella  
B. Rotavirus  
C. Salmonella  
D. Vibrio cholerae  
E. Yersinia

52. A 9-month-old infant presented with low grade fever, repeated vomiting for 3 days followed by frequent loose bowel motions; the stool test was negative for blood and leukocytes, and there is no history to suggest contaminated food ingestion.

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

A. rotavirus infection  
B. traveler’s diarrhea  
C. lactose intolerance  
D. Entameba histolytica  
E. enteropathogenic E. Coli
53. All the following matching are true **EXCEPT**
   A. Shigella dysenteriae ------- febrile seizures
   B. Campylobacter jejuniis ----- traveler’s diarrhea
   C. Clostridium difficile ------- antibiotic-associated diarrhea
   D. enterohemorrhagic (EHEC) ------ hemolytic uremic syndrome
   E. enteropathogenic (EPEC) ------ epidemics of diarrhea in newborn nurseries

54. In gastroenteritis, a large inoculums is required to cause infection with
   A. Shigella
   B. Salmonella
   C. E. coli O157:H7
   D. Giardia lamblia
   E. Entameba histolytica

55. A 3-year-old child presented with high fever, diarrhea, and seizure; stool examination is foul smelling and positive for mucus, blood, and leukocytes. Of the following, the **MOST** likely diagnosis is
   A. giardiasis
   B. shigellosis
   C. amebic dysentery
   D. salmonella infection
   E. Campylobacter infection

56. In the United States, the **MOST** common bacterial food-borne cause of diarrhea is
   A. Shigella
   B. Campylobacter
   C. E. coli O157:H7
   D. Listeria monocytogenes
   E. nontyphoidal Salmonella

57. Colitis with minimal fecal leukocytes occur in infection with
   A. Shigella
   B. C. jejuni
   C. Salmonella
   D. E. histolytica
   E. invasive E. coli
58. In patients with diarrhea, bacterial stool culture is recommended in all the following EXCEPT
   A. fever
   B. profuse diarrhea
   C. traveler’s diarrhea
   D. pseudomembranous colitis
   E. hemolytic uremic syndrome

59. Positive blood culture in bacterial enteritis is expected in
   A. Shigella
   B. Campylobacter jejuni
   C. Yersina enterocolitica
   D. Listeria monocytogenes
   E. nontyphoidal Salmonella

60. Which of the following is a defective virus that occurs either as coinfection with HBV or superinfection in chronic HBsAg carriers?
   A. hepatitis A virus
   B. hepatitis B virus
   C. hepatitis C virus
   D. hepatitis D virus
   E. hepatitis E virus

61. Regarding serological tests of hepatitis B virus (HBV) infection and it’s significance, all the following matching are true EXCEPT
   A. HBsAg ------ acute or chronic infection with HBV
   B. Anti-HBs ------ HBV infection during the window phase
   C. HBeAg ------ acute HBV
   D. Anti-HBe ------ a low degree of infectivity during the carrier state
   E. HBsAg and HBeAg in absence of anti-HBe ------ high risk of transmissibility

62. Ali’s mother is worried about her 10-month-old infant who is exposed to his sister with hepatitis A viral infection.
   Of the following, the BEST advice is to
   A. reassure the mother
   B. give HAV vaccine
   C. give immunoglobulin
   D. do liver function test
   E. give both HAV vaccine and immunoglobulin
63. A newly delivered full term newborn to HBsAg-positive mothers, all the following are true regarding neonatal management **EXCEPT**
   A. HBV vaccine within 12 hours of birth
   B. hepatitis B immunoglobulin within 12 hours of birth
   C. testing for HBsAg within 12 hours of birth
   D. subsequent vaccine doses at 1 and 6 months of age
   E. testing for anti-HBs at 9 to 15 months of age

64. The **MOST** prevalent hepatitis virus infection in HIV-infected persons is
   A. hepatitis B virus
   B. hepatitis C virus
   C. hepatitis D virus
   D. hepatitis E virus
   E. hepatitis G virus

65. A good predictor for severe hepatocellular injury and progression to fulminant hepatic failure is
   A. serum albumin
   B. 5α-nucleotidase
   C. prothrombin time
   D. conjugated bilirubin
   E. alanine and aspartate aminotransferase

66. In hepatitis A virus infection, the **MOST** common physical finding that is characteristic of the icteric phase in addition to jaundice is
   A. lymphadenopathy
   B. petechial rash
   C. splenomegaly
   D. tender hepatomegaly
   E. abdominal distension

67. The treatment of acute hepatitis is largely supportive and involves rest, hydration, and adequate nutrition.
   All the following are indications for hospitalization **EXCEPT**
   A. fever
   B. dehydration
   C. severe vomiting
   D. prolonged prothrombin time
   E. signs of hepatic encephalopathy
68. A 2-year-old girl presented with fever, urgency, dysuria, and abdominal pain. Of the following, the **MOST** common etiological pathogen is
   A. Proteus
   B. Klebsiella
   C. Enterococcus
   D. Pseudomonas
   E. Escherichia coli

69. In general, 8% of girls and 2% of boys have a UTI by 11 years of age, but approximately 75% of infants with bacteriuria are male in age group of
   A. < 3 months
   B. 3 - 6 months
   C. 6 - 9 months
   D. 9 – 12 months
   E. >1 year

70. The classic signs of UTI such as urgency, dysuria, frequency, and abdominal or back pain begin in children at age of
   A. 1 year
   B. 2 years
   C. 3 years
   D. 4 years
   E. 5 years

71. The diagnosis of UTI in infants requires the presence of both pyuria and single pathogenic organism of at least
   A. 25,000 CFU/mL
   B. 50,000 CFU/mL
   C. 75,000 CFU/mL
   D. 100,000 CFU/mL
   E. 125,000 CFU/mL

72. Urinalysis showing leukocyturia suggests urinary tract infection if there is
   A. ≥1 white blood cells [WBCs]/mm$^3$
   B. ≥ 5 white blood cells [WBCs]/mm$^3$
   C. ≥10 white blood cells [WBCs]/mm$^3$
   D. ≥15 white blood cells [WBCs]/mm$^3$
   E. ≥20 white blood cells [WBCs]/mm$^3$
73. All the following can differentiate lower UTI from upper UTI EXCEPT
   A. high ESR
   B. high fever
   C. leukocytosis
   D. urine culture
   E. costovertebral tenderness

74. Renal scarring is a late effect of UTI, it’s mostly identified by
   A. KUB
   B. DMSA scan
   C. ultrasonography
   D. voiding cystourethrogram
   E. urinary dipstick test for leukocyte esterase

75. A 21-day-old infant presented with poor weight gain, decreased feeding, fever, and jaundice; urinalysis showing leukocyturia of 20 WBCs/mm³; total bilirubin level, 13 mg/dl and direct bilirubin level, 4 mg/dl
   Of the following, the MOST appropriate treatment is
   A. oral cefixime
   B. IV gentamicin
   C. oral cephalaxine
   D. oral amoxicillin plus clavulanic acid
   E. oral trimethoprim sulfamethoxazole

76. A 2-year-old child presented with fever, irritability, and inability to walk; on examination there is erythema, swelling, warmth, and tenderness over the lower thigh; radiographs of the left knee showed focal destruction of the distal femoral metaphysis and periosteal reaction.
   Of the following, the MOST common etiological pathogen of this condition is
   A. anaerobes
   B. Staphylococcus aureus
   C. Group A streptococcus
   D. Streptococcus pneumonia
   E. Mycobacterium tuberculosis

77. Of the following, the MOST common etiological pathogen of osteomyelitis in children with sickle cell disease is
   A. anaerobes
   B. Salmonella
C. Pseudomonas aeruginosa
D. Streptococcus pneumonia
E. Haemophilus influenzae type b

78. The MOSTLY involved joint in suppurative arthritis is
   A. hip
   B. knee
   C. ankle
   D. elbow
   E. shoulder

79. The hallmark of the radiological findings in chronic osteomyelitis is
   A. sequestra
   B. Brodie abscess
   C. bone destruction
   D. periosteal elevation
   E. loss of the periosteal fat line

80. All the following matching regarding pathogens of osteomyelitis and their recommended treatment are true EXCEPT
   A. Kingella kingae ------ oxacillin
   B. Streptococcus pyogenes ------ penicillin G
   C. Pseudomonas aeruginosa ------ ceftazidime
   D. Streptococcus pneumonia ------ ceftriaxone
   E. Methicillin-resistant staphylococcus aureus ------ cefazolin

81. In osteomyelitis, which of the following joints when involved necessitate surgical drainage
   A. hip
   B. knee
   C. ankle
   D. elbow
   E. shoulder

82. A 3-year-old child presented with limping, painful movement of lower limbs with both thighs held in external rotation and flexion; he had previous history of gastroenteritis.
   Of the following, the MOST common causative pathogenic organism is
   A. S. aureus
B. N. gonorrhoeae
C. Group A streptococci
D. Shigella flexneri
E. Haemophilus influenzae type b

83. A 3-month-old infant presented with fever, irritability, and excessive crying especially during a diaper change; on examination, there is erythema, swelling, and tenderness over the left hip joint. Of the following, the initial diagnostic procedure of choice for evaluation of this condition is
   A. MRI
   B. CT scan
   C. ultrasound
   D. radiograph
   E. radionuclide scans

84. Respiratory syncytial virus (RSV) is a primary cause of bronchiolitis, the second common cause is
   A. rhinoviruses
   B. adenoviruses
   C. coronaviruses
   D. parainfluenza viruses
   E. human metapneumovirus
1. (A). In measles the rash fades in the same pattern, and illness severity is related to the extent of the rash. It may be petechial or hemorrhagic (black measles). As the rash fades, it undergoes brownish discoloration and desquamation.

2. (C). High-dose vitamin A supplementation has been shown to improve the outcome of infants with measles in developing countries.

3. (A). Otitis media is the most common complication of measles infection. Interstitial (measles) pneumonia can occur, or pneumonia may result from secondary bacterial infection with Streptococcus pneumoniae, Staphylococcus aureus, or group A streptococcus. Myocarditis and mesenteric lymphadenitis are infrequent complications. Encephalomyelitis occurs in 1 to 2 per 1000 cases and usually occurs 2 to 5 days after the onset of the rash. Subacute sclerosing panencephalitis is a late neurologic complication of slow measles infection that is characterized by progressive behavioral and intellectual deterioration and eventual death. It occurs in approximately 1 in every 1 million cases of measles, an average of 8 to 10 years after measles. There is no effective treatment.

4. (A). Tuberculosis of the spine, Pott’s disease, is the most common skeletal site followed by the hip as well as fingers and toes (dactylitis).

5. (C). Susceptible household contacts with a chronic disease or who are immunocompromised should receive postexposure prophylaxis with measles vaccine within 72 hours of measles exposure or immunoglobulin within 6 days of exposure.

6. (A). Rubella, also known as German measles or 3-day measles, is caused by a single-stranded RNA virus with a glycolipid envelope and is a member of the togavirus family. Humans are the only natural host.

7. (B). Rubella virus is most contagious through direct or droplet contact with nasopharyngeal secretions from 2 days before until 5 to 7 days after rash onset, although virus may be present in nasopharyngeal secretions from 7 days before until 14 days after the rash.

8. (B). Roseola is characterized by high fever (often >40° C) with an abrupt onset that lasts 3 to 5 days. A maculopapular, rose-colored rash erupts coincidentally with defervescence, although it may be present earlier. The rash usually lasts 1 to 3 days but may fade rapidly and is not present in all infants with HHV-6 infection. Upper respiratory symptoms, nasal congestion, erythematous tympanic membranes, and cough may occur. Gastrointestinal symptoms are described
9. (C). Erythema infectiosum (fifth disease) is caused by the human parvovirus B19, a single-stranded DNA virus producing a benign viral exanthem in healthy children.

10. (B). Latent tuberculosis describes the asymptomatic stage of infection with M. tuberculosis. The tuberculin skin test (TST) is positive, but the chest radiograph is normal, and there are no signs or symptoms of illness. To prevent reactivation of these latent bacilli, therapy with a single agent (usually isoniazid for 9 months) is suggested.

11. (E). Neurologic complications of chickenpox (varicella) include postinfectious encephalitis, cerebellar ataxia, nystagmus, and tremor. Less common neurologic complications include Guillain-Barre syndrome, transverse myelitis, cranial nerve palsies, optic neuritis, and hypothalamic syndrome.

12. (B). Passive immunity can be provided by VZIG, which is indicated within 96 hours of exposure for susceptible individuals at increased risk for severe illness. Administration of VZIG does not eliminate the possibility of disease in recipients and prolongs the incubation period up to 28 days. Other indication of VZIG in a severe form of neonatal varicella may develop in newborns of mothers with varicella (but not shingles) occurring 5 days before to 2 days after delivery. The fetus is exposed to a large inoculum of virus but is born before the maternal antibody response develops and can cross the placenta. These infants should be treated as soon as possible with varicellazoster immunoglobulin (VZIG) or intravenous immunoglobulin if VZIG is unavailable, to attempt to prevent or ameliorate the infection.

13. (E). Routine oral administration of acyclovir is not recommended in otherwise healthy children with varicella. The decision to use antiviral medications, the route, and duration of treatment depend on host factors and the risk for severe infection or complications. Early therapy with antivirals (especially within 24 hours of rash onset) in immunocompromised persons is effective in preventing severe complications, including pneumonia, encephalitis, and death from varicella. Acyclovir or valacyclovir may be considered in those at risk of severe varicella, such as unvaccinated persons older than 12 years; those with chronic cutaneous or pulmonary disease; receiving short course, intermittent, or aerosolized corticosteroids; or receiving long-term salicylate therapy. The dose of acyclovir used for VZV infections is much higher than that for HSV.

14. (E). Symptoms of increased intracranial pressure include headache, diplopia, and vomiting; a bulging fontanelle may be present in infants. Ptosis, sixth nerve palsy, anisocoria, bradycardia with hypertension, and apnea are signs of increased intracranial pressure with brain herniation. Papilledema is uncommon, unless there is occlusion of the venous sinuses, subdural empyema, or brain abscess.
15. **(A).** Staphylococcal meningitis occurs primarily after neurosurgery or penetrating head trauma.

16. **(E).** Most common; Group B streptococcus, Escherichia coli, Klebsiella, and Enterobacter
Less common; Listeria monocytogenes, Coagulase-negative staphylococci, Enterococcus faecalis, Citrobacter diversus, Salmonella, Pseudomonas aeruginosa, and Haemophilus influenzae types a, b, c, d, e, f, and nontypable.

17. **(B).** Dexamethasone (0.6 to 0.8 mg/kg daily in two to three divided doses for 2 days), as adjunctive therapy initiated just before or concurrently with the first dose of antibiotics, significantly diminishes the incidence of hearing loss and neurologic deficits resulting from H. influenzae meningitis. Adjuvant steroids do not appear to decrease mortality or hospital stay in studies in the developed world.

18. **(E).** A CSF leak (fistula), resulting from congenital anomaly or following a basilar skull fracture, increases the risk of meningitis, especially that caused by S. pneumoniae.

19. **(E).** A temporal lobe focus on EEG or brain imaging is the characteristic feature of herpes simplex virus (HSV) infection.

20. **(A).**

21. **(E).** There are no specific preventive measures for HSV encephalitis except for cesarean section for mothers with active genital lesions.

22. **(B).** The viruses primarily associated with colds are rhinoviruses and, less commonly, coronaviruses. Other viruses that cause common cold symptoms include respiratory syncytial virus and, less commonly, influenza, parainfluenza, and adenoviruses.

23. **(E).** Pale, edematous, nasal turbinate mucosa.

24. **(B).** Group A streptococci (Streptococcus pyogenes). Other bacterial organisms less often associated with pharyngitis include group C streptococcus, Arcanobacterium haemolyticum, and Francisella tularensis. Chlamydophila pneumoniae is associated with lower respiratory disease but also causes sore throat. Mycoplasma pneumonia is associated with atypical pneumonia and may cause mild pharyngitis without distinguishing clinical manifestations.

25. **(C).** Streptococcal pharyngitis is relatively uncommon before 2 to 3 years of age, but the incidence increases in young school-age children and then declines in late adolescence and adulthood.

26. **(E).** **Vincent infection or trench mouth** is a fulminant form of acute necrotizing ulcerative gingivitis with synergistic infection with certain spirochetal organisms, notably Treponema vincentii, with anaerobic Selenomonas and Fusobacterium. **Vincent angina** refers to a virulent form of anaerobic pharyngitis; gray pseudomembranes are found on the tonsils. **Ludwig angina** is a mixed anaerobic
bacterial cellulitis of the submandibular and sublingual regions. Although often applied to any infection of the sublingual or submandibular region, the term originally was reserved for a rapidly spreading bilateral cellulitis of the sublingual and submandibular spaces. It typically is due to spreading from a periapical abscess of the second or third mandibular molar. It also has been associated with tongue piercing. A propensity for rapid spread, glottic and lingual swelling, and consequent airway obstruction makes prompt intervention imperative

27. (B). Prompt treatment indicated if there is past personal history of rheumatic fever or a recent family history of rheumatic fever.

28. (D). The maxillary sinuses are present at birth and become pneumatized at 4 years of age.

29. (D). The common cold is the major predisposing factor for developing sinusitis at all ages. Other risk factors include allergy, cystic fibrosis, immunodeficiency, human immunodeficiency virus (HIV) infection, nasogastric or nasotracheal intubation, immotile cilia syndrome, nasal polyps, and nasal foreign body.

30. (E). Amoxicillin-clavulanate for 10 to 14 days is recommended as first-line therapy of sinusitis in children. High-dose therapy is recommended for children at increased risk for resistant bacteria (antibiotic treatment in the preceding 1 to 3 months, day care attendance, age <2 years, high rates of antimicrobial resistance locally). Levofloxacin is recommended for children with type I hypersensitivity to penicillins. Clindamycin plus a third generation cephalosporin (cefixime, cefpodoxime) is recommended for children with non-type I hypersensitivity.

31. (A). In pulmonary tuberculosis, most infectious patients become noninfectious within 2 weeks of starting effective treatment, and many become noninfectious within several days.

32. (A). Primary pulmonary tuberculosis in older infants and children is usually an asymptomatic infection. Often the disease is manifested by a positive TST with minimal abnormalities on the chest radiograph, such as an infiltrate with hilar lymphadenopathy or Ghon complex. Lymphadenopathy is common in primary pulmonary disease. Hilar lymphadenopathy may compress the bronchi or trachea. The most common extrathoracic sites of lymphadenitis are the cervical, supraclavicular, and submandibular areas (scrofula).

33. (C). The recommended first-line therapy for most children with a certain diagnosis of acute OM or those with an uncertain diagnosis but who are younger than 2 years of age or have fever greater than 39°C or otalgia is amoxicillin (80 to 90 mg/kg/day in two divided doses). Children with an uncertain diagnosis who are older than 2 years of age may be observed if appropriate follow-up can be arranged.
Recommended next-step treatments include high-dose amoxicillin-clavulanate (amoxicillin 80 to 90 mg/kg/day), cefuroxime axetil, cefdinir, or ceftriaxone (50 mg/kg intramuscularly in daily doses for 1 to 3 days).

34. (B). Otitis externa, also known as swimmer’s ear, is defined by inflammation and exudation in the external auditory canal in the absence of other disorders, such as otitis media or mastoiditis. The most common bacterial pathogens are Pseudomonas aeruginosa, especially in association with swimming in pools or lakes, and Staphylococcus aureus.

35. (C). The most common bacterial pathogens are Pseudomonas aeruginosa, especially in association with swimming in pools or lakes, and Staphylococcus aureus.

36. (C). Spasmodic croup describes sudden onset of croup symptoms, usually at night, but without a significant upper respiratory tract prodrome. These episodes may be recurrent and severe but usually are of short duration. Spasmodic croup has a milder course than viral croup and responds to relatively simple therapies, such as exposure to cool or humidified air. The etiology is not well understood and may be allergic.

37. (B).

38. (B). In laryngotracheobronchitis (croup), the anteroposterior radiographs of the neck often show the diagnostic subglottic narrowing of croup known as the steeple sign (the tapered narrowing of the immediate subglottic airway).

39. (C). Treatment during the catarrhal phase eradicates nasopharyngeal carriage of organisms within 3 to 4 days and may lessen symptom severity. Treatment in the paroxysmal stage does not alter the course of illness but decreases the potential for spread to others.

40. (D). The diagnosis of pertussis depends on isolation of B. pertussis or detection of its nucleic acids. Culture on specialized media is usually accomplished during the early phases of illness on specimens from nasopharyngeal swabs or aspirates but can be difficult to accomplish given the organism’s fastidious nature. Polymerase chain reaction, although potentially lacking sensitivity in previously immunized individuals, is likely as sensitive as culture and is available in many clinical laboratories.

41. (E). The most frequent complication is pneumonia caused by B. pertussis itself or resulting from secondary bacterial infection with Streptococcus pneumoniae, Haemophilus influenzae, and Staphylococcus aureus. Most children recover normal pulmonary function with complete healing of the respiratory epithelium. Most permanent disability is a result of encephalopathy.

42. (C). Respiratory syncytial virus (RSV) is a primary cause of bronchiolitis, followed in frequency by human metapneumovirus, parainfluenza viruses, influenza viruses,
adenoviruses, rhinoviruses, coronaviruses, and, infrequently, Mycoplasma pneumoniae.

43. (A).

44. (D). Lung abscess usually caused by certain virulent organisms (Anaerobic bacteria usually predominate, along with various streptococci, Escherichia coli, Klebsiella pneumoniae, Pseudomonas aeruginosa, and Staphylococcus aureus). Diagnostic bronchoscopy may be indicated to exclude a foreign body and obtain microbiologic specimens. Lung abscesses usually respond to appropriate antimicrobial therapy with clindamycin, penicillin G, or ampicillin-sulbactam.

45. (C). Atypical pneumonia describes patterns typically more diffuse or interstitial than lobar pneumonia. M. pneumonia and Chlamydia pneumonia are principal causes.

46. (A). The most likely pathogen causing lobar pneumonia is Streptococcus pneumoniae.

*Recommended treatment; Ampicillin or penicillin IV; amoxicillin PO.
*Alternative treatment; Ceftriaxone, cefotaxime, clindamycin or vancomycin IV; Cefuroxime, cefpodoxime, levofloxacin, or linezolid PO.

47. (C). In age group (3 months to 5 year); the common pathogens (in approximate order of frequency) are respiratory syncytial virus, other respiratory viruses (parainfluenza viruses, influenza viruses, human metapneumovirus adenoviruses), S. pneumoniae, H. influenzae (type b, nontypable).

48. (B). Viridans streptococci are the principal causes of infective endocarditis in children with congenital heart diseases without previous surgery. Staphylococcus aureus and coagulase-negative staphylococci are important causes of endocarditis, especially following cardiac surgery and in the presence of prosthetic cardiac and endovascular materials.

49. (A). In most cases, oral amoxicillin 50 mg/kg (maximum dose, 2 g) taken 30 to 60 minutes before the procedure is the recommended regimen. Clindamycin or azithromycin are alternative regimens indicated for most patients allergic to β-lactams. Prolonged or continuous antibiotic prophylaxis is not recommended.

50. (D).

**Major Criteria**

1. Positive blood cultures;
   *Two or more separate cultures positive with typical organisms for infective endocarditis.
   *Two or more positive cultures of blood drawn more than 12 hours apart or 4 positive blood cultures irrespective of timing of obtaining specimen.
   *A positive blood culture for Coxiella burnetii positive IgG titer >1:800.

2. Evidence of endocardial involvement.
3- Positive findings on echocardiogram (vegetation on valve or supporting structure, abscess, new valvular regurgitation).

**Minor Criteria**
1- Predisposition—predisposing heart condition or injection drug use.
2- Fever—temperature >38° C (>100.4° F).
3- Vascular phenomena (major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial hemorrhage, conjunctival hemorrhages, Janeway lesions).
4- Immunologic phenomena (glomerulonephritis, Osler nodes, Roth spots, rheumatoid factor).
5- Microbiologic evidence (positive blood culture result, but not meeting major criteria, or serologic evidence of active infection with organism consistent with infective endocarditis).

51. *(D).*
*Secretory mechanism --- Cholera, toxigenic Escherichia coli (EPEC, ETEC); carcinoid, Clostridium difficile, and cryptosporidiosis (in AIDS)*

52. *(A).* Rotavirus is the most frequent cause of diarrhea during the winter months. Vomiting may last 3 to 4 days, and diarrhea may last 7 to 10 days. Dehydration is common in younger children. Primary infection with rotavirus may cause moderate to severe disease in infancy but is less severe later in life. This illness is much less common in areas where infants receive the rotavirus vaccine.

53. *(B).* Enterotoxigenic (ETEC) causes 40% to 60% of cases of traveler’s diarrhea.

54. *(B).* A large inoculum, of 1000 to 10 billion organisms, is required because Salmonella is killed by gastric acidity.

55. *(B).* Dysentery is enteritis involving the colon and rectum, with blood and mucus, possibly foul smelling stools, and fever. Shigella is the prototype cause of dysentery, which must be differentiated from infection with EIEC, EHEC, E. histolytica (amebic dysentery), C. jejuni, Y. enterocolitica, and nontyphoidal Salmonella.

56. *(E).* In the United States, the **MOST** common bacterial food-borne causes (in order of frequency) are nontyphoidal Salmonella, Campylobacter, Shigella, E. coli O157:H7, Yersinia, Listeria monocytogenes, and V. cholerae. The most common parasitic food-borne causes are Cryptosporidium parvum and Cyclospora cayetanensis.

57. *(D).* Stool specimens should be examined for mucus, blood, and leukocytes, which indicate colitis in response to bacteria that diffusely invade the colonic mucosa, such as Shigella, Salmonella, C. jejuni, and invasive E. coli. Patients
infected with Shiga toxin-producing E. coli and E. histolytica generally have minimal fecal leukocytes.

58. (C). Bacterial stool cultures are recommended for patients with fever, profuse diarrhea, and dehydration or if HUS or pseudomembranous colitis is suspected.

59. (E). Positive blood cultures are uncommon with bacterial enteritis except for S. typhi (typhoid fever), nontyphoidal Salmonella, and E. coli enteritis in very young infants.

60. (D). HDV, also known as the delta agent, is a defective virus that requires HBV for spread and causes either coinfection with HBV or superinfection in chronic HBsAg (hepatitis B surface antigen) carriers.

61. (B).

- Anti-HBs ------ lifelong immunity and is also a marker of immunization.
- Anti-HBc ------ HBV infection during the window phase.

62. (C). Unvaccinated household and sexual contacts of persons with HAV should receive postexposure prophylaxis as soon as possible and within 2 weeks of the last exposure. A single dose of HAV vaccine at the age-appropriate dose is preferred for persons 12 months to 40 years of age. Immunoglobulin (0.02 mL/kg) given intramuscularly is preferred for children under 12 months of age, persons over 40 years of age, and immunocompromised persons.

63. (C). Infants born to HBsAg-positive mothers should receive HBV vaccine and hepatitis B immunoglobulin (HBIG) (0.5 mL) within 12 hours of birth, with subsequent vaccine doses at 1 month and 6 months of age followed by testing for HBsAg and anti-HBs at 9 to 15 months of age.

Infants born to mothers whose HBsAg status is unknown should receive vaccine within 12 hours of birth. If maternal testing is positive for HBsAg, the infant should receive HBIG as soon as possible (no later than 1 week of age).

64. (E).

65. (C). Alanine aminotransferase and aspartate aminotransferase levels are elevated and generally reflect the degree of parenchymal inflammation. Alkaline phosphatase, 5α-nucleotidase, and total and direct (conjugated) bilirubin levels indicate the degree of cholestasis, which results from hepatocellular and bile duct damage.

66. (D).

67. (A).

68. (E). Escherichia coli, ascending from bowel flora, accounts for 90% of first UTI infections and 75% of recurrent UTI infections. Over 90% of nephritogenic E. coli possess P fimbriae that bind to uroepithelial cells and P blood group antigens. Individuals with high-level expression of P1 blood group antigen are predisposed to
pyelonephritis and bacteremia, as well as recurrent UTIs. Other bacteria commonly causing infection include Klebsiella, Proteus, Enterococcus, and Pseudomonas.

69.(A). Approximately 8% of girls and 2% of boys have a UTI by 11 years of age. The lifetime incidence of UTI in females is about 30% compared to only 1% in males. Approximately 75% of infants younger than 3 months of age with bacteriuria are male compared with only 10% between 3 and 8 months of age. After 12 months of age, UTI in healthy children usually is seen in girls.

70.(B).

71.(B).

72.(C). Urinalysis showing pyuria (leukocyturia of >10 white blood cells [WBCs]/mm$^3$) suggests infection, but also is consistent with urethritis, vaginitis, nephrolithiasis, glomerulonephritis, and interstitial nephritis.

73.(D). The diagnosis of a UTI is confirmed by a positive urine culture, but this does not distinguish between upper tract and lower tract infection. Fever and abdominal pain may occur with either lower or upper UTI, although high fever, costovertebral tenderness, high erythrocyte sedimentation rate, leukocytosis, and bacteremia each suggest upper tract involvement. DMSA scan is sensitive for detecting acute pyelonephritis but not commonly used.

74.(B). A technetium-99m DMSA scan can identify acute pyelonephritis and is most useful to define renal scarring as a late effect of UTI.

75.(B). Neonates with UTI are treated for 10 to 14 days with parenteral antibiotics because of the higher rate of bacteremia.

76.(B). Staphylococcus aureus is responsible for more than 80% of acute skeletal infections. Other common causes include Group A streptococcus and Streptococcus pneumoniae. Neisseria meningitidis, Mycobacterium tuberculosis, Bartonella henselae, Actinomycetes spp., and anaerobes are less common causes.

77.(B). Sickle cell disease and other hemoglobinopathies predispose to osteomyelitis caused by Salmonella and S. aureus.

78.(B). The joints of the lower extremity are most often involved: the knees (40%), the hips (20%), and the ankles (14%). Small joints, such as those of the hand, usually are involved after penetrating trauma or closed fist injuries.

79.(A). Plain radiographs can demonstrate soft tissue swelling such as the loss of the periosteal fat line within the first 3 days of symptoms, but bony lesions such as periosteal elevation and bone destruction are absent until after 10 to 14 days of symptoms. Brodie abscess is a subacute intraosseous abscess that does not drain into the subperiosteal space and is classically located in the distal tibia. Sequestra, portions of avascular bone that have separated from adjacent bone, frequently are
covered with a thickened sheath, or involucrum, both of which are hallmarks of chronic osteomyelitis

80.(E).

- Methicillin-sensitive Staphylococcus aureus ---- nafcillin (or oxacillin) or cefazolin.
- Methicillin-resistant Staphylococcus aureus ---- clindamycin or vancomycin or Linezolid.

81.(A). Lack of improvement after 48 hours indicates that surgical drainage may be necessary or that an unusual pathogen may be present. Surgical drainage is indicated for extensive or severe disease, if the disease is chronic or atypical, the hip joint is involved, or sequestrum or spinal cord compression is present.

82.(D). Reactive arthritis is typically symmetric, polyarticular, and usually involves the large joints, especially the hips. Patients may have had a preceding episode of gastroenteritis or urethritis. Pathogenic organisms causing arthritis in children are Yersinia enterocolitica, Campylobacter jejuni, Shigella flexneri, Salmonella, Group A streptococci, Neisseria meningitidis, Coccidioides immitis, and Rubella virus.

83.(C). Ultrasound is especially useful for identifying joint effusions and is the initial diagnostic procedure of choice for evaluation of suppurative infections of the hip.

84.(E). Respiratory syncytial virus (RSV) is a primary cause of bronchiolitis, followed in frequency by human metapneumovirus, parainfluenza viruses, influenza viruses, adenoviruses, rhinoviruses, coronaviruses, and, infrequently, Mycoplasma pneumoniae.
1. The **MAJOR** cause of inadequate pancreatic digestive function in children in developed countries is
   A. mumps
   B. cystic fibrosis
   C. Shwachman-Diamond syndrome
   D. Pearson syndrome
   E. malnutrition

2. Pancreatic enzymes efficacy is increased by the use of
   A. ceftriaxone
   B. metoclopramide
   C. omeprazole
   D. aspirin
   E. carbamazepine

3. Most cases of acute pancreatitis in children are due to
   A. hypertriglyceridemia
   B. biliary microlithiasis
   C. trauma
   D. medications
   E. viral infections

4. Rectal examination is helpful in children with which of the following?
   A. acute diarrhea
   B. chronic diarrhea
   C. anal itching
   D. jaundice
   E. vomiting

5. Hepatic synthetic function can be assessed by
   A. total and direct bilirubin
   B. alanine aminotransferase
   C. albumin level
   D. γ-glutamyltransferase
6. A 6-year-old female presented with history of abdominal pain after each meal and at bed time, burning in nature associated with sour taste in her mouth. Of the following, the **MOST** likely cause is
   A. functional abdominal pain
   B. esophageal reflux
   C. duodenal ulcer
   D. celiac disease
   E. lactose intolerance

7. A common cause of recurrent abdominal pain in children is
   A. duodenal ulcer
   B. pneumonia
   C. functional abdominal pain
   D. obstructive uropathy
   E. congenital intestinal malformation

8. Which drug is useful for the prophylaxis and treatment of motion sickness?
   A. dimenhydrinate
   B. ondansetron
   C. dexamethasone
   D. propranolol
   E. cyproheptadine

9. Micrognathia and cleft palate are common associations with which of the following?
   A. osteogenesis imperfecta
   B. ectodermal dysplasia
   C. Pierre Robin syndrome
   D. mandibulofacial dysostosis
   E. Crouzon syndrome

10. Typically the first erupted teeth are
    A. upper central incisors
    B. lower central incisors
    C. lateral incisors
    D. first molars
    E. bicuspids
11. Delayed eruption of teeth may occur in association with
   A. hyperthyroidism
   B. osteopetrosis
   C. Neiman pick disease
   D. Turner syndrome
   E. scurvy

12. Persistent oral thrush in a thirty five-day-old healthy infant is **MOST** likely due to
   A. chronic diarrhea
   B. broad-spectrum antibiotic use
   C. diabetes
   D. use of dirty bottles
   E. infected nipples

13. Which of the following factors is involved in causing infantile GER?
   A. solid diet
   B. vertical body position
   C. long, narrow esophagus
   D. small, noncompliant stomach
   E. frequent small volume feedings

14. Pill ulcers occur when certain medications are swallowed without sufficient liquids, allowing prolonged direct contact of the pill with the esophageal mucosa. Of the following, the **MOST** accused one is
   A. acetazolamide
   B. methyldopa
   C. carbamazepine
   D. retinol
   E. tetracycline

15. A common presentation of malrotation during the first month of life is
   A. lethargy
   B. abdominal distention
   C. bloody diarrhea
   D. bilious vomiting
   E. hypoglycemia
16. Beckwith-Wiedmann syndrome is caused by an imprinting disorder and characterized by gigantism, macroglossia, microcephaly, visceromegaly, and
   A. gastroschisis
   B. omphalocele
   C. Hirschsprung disease
   D. Meckel diverticulum
   E. anorectal malformations

17. True manifestations of inflammatory bowel disease are
   A. colonic involvement in Crohn’s disease
   B. whole gut involvement in ulcerative colitis
   C. pyoderma gangrenosum in Crohn’s disease
   D. fissures and skin tags in Crohn’s disease.
   E. erythema nodosum in ulcerative colitis

18. A 1-day-old full-term neonate experiences coughing, frothing of the mouth and cyanosis during attempts of breastfeeding. Because of concern about aspiration, an attempt is made to pass a nasogastric tube, but the tube meets resistance and cannot be advanced adequately.
   Of the following, the MOST likely diagnosis is
   A. choanal atresia
   B. esophageal atresia
   C. gastroesophageal reflux disease
   D. pyloric atresia
   E. Zenker’s diverticulum

19. A healthy 3-year-old boy reports to his mother that he accidentally swallowed a small plastic toy and is brought to the emergency department for evaluation. The mother recalls that the other toys in the playset are all 1 to 2 cm in length and have no sharp edges. On examination, the child appears to be in no distress and is playing in the examination room with his sister. His vital signs are normal.
   Of the following, the BEST next step is
   A. barium swallow and meal imaging
   B. observation
   C. chest and abdominal radiograph series
   D. upper endoscopy to remove the toy
   E. surgical consultation as the object could cause a bowel obstruction
20. An 18-month-old girl presented with a 6 month history of diarrhea, abdominal distension, failure to thrive, irritability, and decreased appetite. Of the following, the **MOST** likely diagnosis is
   A. giardiasis
   B. toddler diarrhea
   C. Crohn’s disease
   D. celiac disease
   E. lactase deficiency

21. The diagnosis of celiac disease is confirmed by
   A. ultra sound abdomen
   B. barium study
   C. duodenal biopsy
   D. serum B_{12} level
   E. TTG antibody

22. Which of the following infections is a common cause of acute abdominal pain?
   A. spontaneous bacterial peritonitis
   B. cholecystitis
   C. pneumonia (lower lobes)
   D. urinary tract infection
   E. bacterial typhlitis

23. Which of the following is a common cause of chronic diarrhea in infancy?
   A. congenital chloridorrhea
   B. acrodermatitis enteropathica
   C. abetalipoproteinemia
   D. intractable diarrhea syndrome
   E. post-infectious secondary lactase deficiency

24. One of the following studies may be helpful in discovering the cause behind small bowel atresia
   A. immunoglobulin assay
   B. chloride sweat test
   C. liver function tests
   D. serum electrolytes
   E. complete blood count
25. The following features are more common in Crohn's disease than ulcerative colitis
   A. fever
   B. weight loss
   C. mouth ulceration
   D. abdominal pain
   E. risk of cancer

26. An 18-month-old boy presented with sudden onset of vomiting; crampy abdominal pain; his knees draw up, and cries out and exhibits pallor with a colicky pattern occurring every 15 to 20 minutes.
   Of the following, the MOST likely diagnosis is
   A. gastroenteritis
   B. acute appendicitis
   C. pancreatitis
   D. urinary tract infection
   E. intussusception

27. Cow's milk protein intolerance is characterized by
   A. abdominal tenderness
   B. streaks of bloody mucus in stools
   C. abdominal distention
   D. vomiting
   E. peripheral eosinophilia

28. All the following are sensitive indices of synthetic function of the liver EXCEPT
   A. serum albumin
   B. prothrombin time
   C. partial thromboplastin time
   D. factor V level
   E. serum amylase

29. The Alvarado/MANTRELS rule for diagnosis of acute appendicitis gives 2 point for
   A. migration of pain to the right lower quadrant
   B. anorexia
   C. rebound pain
   D. WBC shift to greater than 75% neutrophils
   E. tenderness in the right lower quadrant
30. The presence of hepatocyte giant cells in liver biopsy is characteristic of
   A. extrahepatic biliary atresia
   B. neonatal hepatitis
   C. α1-Antitrypsin deficiency
   D. Alagille syndrome
   E. tyrosinemia

31. An 18-month-old boy presented with jaundice since long time, on examination; he has systolic murmur; hypertelorism; deep-set eyes, prominent forehead, and a pointed chin.
   Of the following, the MOST likely diagnosis is
   A. Alagille syndrome
   B. Aagenaes Syndrome
   C. Niemann-Pick disease
   D. Zellweger syndrome
   E. Wolman disease

32. Paucity of bile ducts in the portal triads in liver biopsy is a unique finding in
   A. Alagille syndrome
   B. Aagenaes Syndrome
   C. Niemann-Pick disease
   D. Zellweger syndrome
   E. Wolman disease
1. (B). Cystic fibrosis is the main cause in 95%; less common causes of pancreatic insufficiency are Shwachman-Diamond syndrome and Pearson syndrome in developed countries and severe malnutrition in developing countries.

2. (C). Use of H2 receptor antagonists or proton-pump inhibitors can increase the efficacy of pancreatic enzymes by enhancing their release from the microspheres and reducing inactivation by acid.

3. (D). In children, most cases are idiopathic or due to medications. Some cases are caused by pancreatic sufficient cystic fibrosis, hypertriglyceridermia, biliary microlithiasis, trauma, or viral infection. Collagen vascular disorders and parasite infestations are responsible for the remainder.

4. (B). A rectal examination, including inspection for fissures, skin tags, abscesses, and fistulous openings, should be performed for children with history suggesting constipation, GI bleeding, abdominal pain, chronic diarrhea, and suspicion of inflammatory bowel disease (IBD).

5. (C). Tests of liver dysfunction include total and direct bilirubin, alanine aminotransferase, and aspartate aminotransferase for evidence of hepatocellular injury, and γ-glutamyltransferase or alkaline phosphatase for evidence of bile duct injury. Hepatic synthetic function can be assessed by coagulation factor levels, prothrombin time, and albumin level.

6. (B).

7. (C). Common causes of recurrent abdominal pain include functional abdominal pain, irritable bowel syndrome, lactose intolerance, inflammatory bowel disease, constipation, celiac disease and eosinophilic esophagitis.

8. (A). Anticholinergics (e.g., scopolamine) and antihistamines (e.g., dimenhydrinate) are useful for the prophylaxis and treatment of motion sickness.

9. (C). Osteogenesis imperfecta is associated with abnormal dentin and risk of caries. Children with ectodermal dysplasias commonly have malformed or missing teeth. Pierre Robin syndrome is associated with micrognathia and cleft palate. Disorders resulting in facial dysmorphism can have a profound effect on dental occlusion and mandibular function. Examples include mandibulofacial dysostosis, Crouzon syndrome, conditions associated with dwarfism, and others.
10. (B). The lower central incisors are typically the first to erupt, followed by the upper central incisors, lateral incisors, first molars, and bicuspids.

11. (B). Delayed eruption may occur in association with hypopituitarism, hypothyroidism, osteopetrosis, Gaucher disease, Down syndrome, cleidocranial dysplasia, and rickets.

12. (E). Thrush is common in healthy neonates. The organism may be acquired in the birth canal or from the environment. Persistent infection is common in breastfed infants as a result of colonization or infection of the mother’s nipples. Thrush in healthy older patients can occur, but should suggest the possibility of an immunodeficiency, broad-spectrum antibiotic use, or diabetes.

13. (D). Factors involved in causing infantile GER include liquid diet; horizontal body position; short, narrow esophagus; small, noncompliant stomach; frequent, relatively large-volume feedings; and an immature lower esophageal sphincter (LES).

14. (E). Tetracyclines and nonsteroidal anti-inflammatory drugs [NSAIDs].

15. (D). About 60% of children with malrotation present with symptoms of bilious vomiting during the first month of life. The remaining 40% present later in infancy or childhood.

16. (B).

17. (D). UC involves only the colon. CD can include the entire gut from mouth to anus. Extraintestinal manifestations of UC include primary sclerosing cholangitis, arthritis, uveitis, and pyoderma gangrenosum. Extraintestinal manifestations of CD include arthritis, erythema nodosum, and uveitis or iritis.

18. (B).

19. (B). Removal is usually not necessary if the object is small and it will most likely pass without complication. Endoscopy is emergent for removal of any esophageal foreign body if symptomatic or if the ingested foreign body is a suspected disc battery in the esophagus or multiple magnets located in the upper tract.

20. (D).

21. (C). Symptoms can begin at any age when gluten-containing foods are given. Diarrhea, abdominal bloating, failure to thrive, irritability, decreased appetite, and ascites caused by hypoproteinemia are classic. An endoscopic small bowel biopsy is essential to confirm the diagnosis and should be performed while the patient is still taking gluten.

22. (D). Appendicitis, viral or bacterial gastroenteritis/mesenteric adenitis, and urinary tract infection are common infectious cause of acute abdominal pain.

23. (E). Common causes include post-infectious secondary lactase deficiency, cow's milk/soy protein intolerance, chronic nonspecific diarrhea of infancy (toddler's diarrhea), celiac disease, and cystic fibrosis.
24. (B). Laboratory evaluation for cystic fibrosis is indicated in cases of small bowel atresia. Other laboratory studies are not specific for atresia, but a complete blood count, serum electrolytes, liver functions, and amylase should be measured to identify dehydration, pancreatitis, and other complications.

25. (E). Clinical manifestations depend on the region of involvement. UC involves only the colon, whereas CD can include the entire gut from mouth to anus.

26. (E). An infant with intussusception has sudden onset of crampy abdominal pain; the infant’s knees draw up, and the infant cries out and exhibits pallor with a colicky pattern occurring every 15 to 20 minutes.

27. (B) There is no abdominal tenderness or distention and no vomiting. If these are present, other diagnoses, such as intussusception or volvulus, should be considered. Peripheral eosinophilia generally is not present on complete blood count, which nevertheless should be performed to rule out an associated iron deficiency anemia.

28. (E). In addition to monitoring prothrombin time and partial thromboplastin time, many centers measure factor V serially as a sensitive index of synthetic function.

29. (E). The Alvarado/ MANTRELS rule is scored by 1 point for each of the following: migration of pain to the right lower quadrant, anorexia, nausea/ vomiting, rebound pain, temperature of at least 37.3°C, and WBC shift to greater than 75% neutrophils; 2 points are given for each of tenderness in the right lower quadrant and leukocytosis greater than 10,000/cmm. Children with a score of 4 or less are unlikely to have appendicitis; a score of 7 or greater increases the likelihood that the patient has appendicitis.

30. (B). If liver biopsy is performed, the presence of hepatocyte giant cells is characteristic.

31. (A).

32. (A). Alagille syndrome is characterized by chronic cholestasis with the unique liver biopsy finding of paucity of bile ducts in the portal triads.
1. A full-term infant has approximately 25 million alveoli; an adult nearly 300 million alveoli. The growth of new alveoli occurs during the first 2 years of life and is complete by the age of
   A. 4 years
   B. 6 years
   C. 8 years
   D. 10 years
   E. 12 years

2. Gas exchange depends on alveolar ventilation, pulmonary capillary blood flow, and the diffusion of gases across the alveolar-capillary membrane. PaO₂ does not improve with supplemental oxygen in
   A. asthma
   B. bronchopulmonary dysplasia
   C. pneumonia
   D. cyanotic heart disease
   E. atelectasis

3. Hyperresonant chest percussion is elicited in one of the following disease processes
   A. consolidation
   B. bronchospasm
   C. pneumothorax
   D. atelectasis
   E. interstitial process

4. Chronic cough is defined as a daily cough lasting longer than
   A. 2 weeks
   B. 3 weeks
   C. 4 weeks
   D. 6 weeks
   E. 12 weeks
5. Magnetic resonance imaging (MRI) is useful in visualizing
   A. cardiac anatomy
   B. pulmonary parenchymal lesions
   C. pleural masses
   D. bronchiectasis
   E. mediastinal lesions

6. Ultrasonography is the imaging procedure of choice for assessing
   A. empyema
   B. mediastinal lesions
   C. pulmonary parenchymal lesions
   D. congenital pulmonary malformations
   E. great vessel anatomy

7. The Pco$_2$ from a capillary sample is similar to that from arterial blood. The Pco$_2$ in venous samples is approximately higher than arterial Pco$_2$ by
   A. 4 mm Hg
   B. 6 mm Hg
   C. 8 mm Hg
   D. 10 mm Hg
   E. 12 mm Hg

8. Measurement of lung volumes and airflow rates using spirometry are important in assessing pulmonary disease. These measures are compared to predicted values which rely mostly on patient
   A. height
   B. age
   C. gender
   D. race
   E. weight

9. Supplemental O$_2$ may be delivered by a variety of face-mask systems ranging from a simple face mask, which can provide 30% to 40% O$_2$, to a nonrebreather mask with reservoir that can provide nearly
   A. 60% O$_2$
   B. 70% O$_2$
   C. 80% O$_2$
   D. 90% O$_2$
   E. 100% O$_2$
10. The acceptable O\textsubscript{2} saturation depends on the patient and clinical situation. Generally, supplemental O\textsubscript{2} should be administered to achieve goal saturation level above
   A. 80%
   B. 85%
   C. 90%
   D. 93%
   E. 95%

11. Chest physiotherapy is most beneficial in children with
   A. foreign body inhalation
   B. cystic fibrosis
   C. asthma
   D. pneumonia
   E. atelectasis

12. An acute life-threatening event (ALTE) is defined as any unexpected and frightening change in condition characterized by apnea, color change (usually blue or pale), sudden limpness, choking, or gagging.
   Of the following, the **MOST** likely cause is
   A. breath-holding spells
   B. intracranial bleeding
   C. pertussis
   D. meningitis
   E. gastroesophageal reflux

13. The decreased incidence of SIDS during the past two decades explained partly by
   A. widely advocated supine sleeping position
   B. ECG monitoring to detect prolonged Q-T interval
   C. avoiding soft bedding
   D. decreasing maternal cigarette smoking
   E. avoiding parents to share beds with their infants

14. the most common cause of obstructive sleep apnea syndrome (OSA) in young children is
   A. obesity
   B. craniofacial malformations
   C. glossoptosis
D. neuromuscular diseases
E. adenotonsillar hypertrophy

15. The **MOST** common non-infectious cause of inspiratory stridor in infants is
   A. subglottic stenosis
   B. laryngeal web
   C. airway hemangioma
   D. rhinitis
   E. laryngomalacia

16. Barking cough is not a feature in
   A. laryngotracheobronchitis
   B. laryngitis
   C. spasmodic croup
   D. epiglottitis
   E. bacterial tracheitis

17. A newborn presented with respiratory distress relieved by crying. Of the following, the **MOST** likely cause is
   A. choanal atresia
   B. micrognathia
   C. macroglossia
   D. laryngeal web
   E. congenital subglottic stenosis

18. Croup, or laryngotracheobronchitis, is the most common infection of the middle respiratory tract. Of the following, The **MOST** common cause of croup is
   A. influenza virus
   B. parainfluenza virus
   C. adenovirus
   D. rhinovirus
   E. human metapneumovirus

19. An 18-month-old child presented with a second attack of sudden onset of harsh barking cough, hoarseness, and inspiratory stridor at night without a significant upper respiratory tract prodrome. Of the following, The **MOST** likely diagnosis is
   A. bacterial tracheitis
B. epiglottitis  
C. spasmodic croup  
D. laryngotracheobronchitis  
E. foreign body aspiration

20. Vocal cord paralysis is an important cause of laryngeal dysfunction. Paralysis may be unilateral or bilateral and is more often caused by  
   A. Arnold-Chiari malformation  
   B. hydrocephalus  
   C. intracranial hemorrhage  
   D. damage to the recurrent laryngeal nerve  
   E. stroke

21. The **MOST** common foreign bodies aspirated by young children are  
   A. nuts  
   B. small toys  
   C. coins  
   D. rubber balloons  
   E. handwatch batteries

22. Kartagener syndrome is the triad of  
   A. situs inversus, pansinusitis, and bronchiectasis  
   B. dextrocardia, pansinusitis, and bronchiectasis  
   C. situs inversus, recurrent otitis media, and bronchiectasis  
   D. situs inversus, recurrent otitis media, and infertility  
   E. situs inversus, pansinusitis, and asthma

23. The primary cause of bronchiolitis is  
   A. human metapneumovirus  
   B. parainfluenza viruses  
   C. rhinoviruses  
   D. coronaviruses  
   E. respiratory syncytial virus

24. It may be difficult to differentiate asthma from bronchiolitis by physical examination, but all the following may be helpful **EXCEPT**  
   A. age of presentation  
   B. fever  
   C. personal history of asthma
D. family history of asthma
E. wheezing

25. Bronchiolitis treatment consists of supportive therapy including all the following EXCEPT
   A. respiratory monitoring
   B. bronchodilators
   C. control of fever
   D. hydration
   E. upper airway suctioning

26. Monthly injections of palivizumab, a RSV-specific monoclonal antibody, initiated just before the onset of the RSV season confers some protection from severe RSV disease. Palivizumab is indicated for some infants under 2 years old with
   A. spasmodic croup
   B. very low birth weight
   C. cyanotic congenital heart disease
   D. GERD
   E. recurrent pneumonia

27. The MOST common bacterial cause of lobar pneumonia in children of any age outside the neonatal period is
   A. M. pneumoniae
   B. Chlamydia pneumoniae
   C. Streptococcus pneumoniae
   D. Staphylococcus aureus
   E. Hemophilus influenza

28. The MOST common cause of afebrile pneumonia in infants 1 to 3 months of age is
   A. Chlamydia trachomatis
   B. Mycoplasma hominis
   C. Ureaplasma urealyticum
   D. Cytomegalovirus
   E. Bordetella pertussis

29. Infectious agents that commonly cause community-acquired pneumonia vary by age.
    Of the following, the MOST common pathogen in 3 months to 5 years age group is
A. respiratory syncytial virus (RSV)
B. Chlamydia trachomatis
C. Mycoplasma pneumoniae
D. group A streptococcus,
E. Staphylococcus aureus

30. Pneumonia in older patients with cystic fibrosis is usually caused by
   A. Pseudomonas aeruginosa
   B. Staphylococcus aureus
   C. Chlamydia trachomatis
   D. Mycoplasma pneumoniae
   E. Hemophilus influenza

31. Immunizations have had a great impact on the incidence of pneumonia caused
    by all the following **EXCEPT**
    A. pertussis
    B. diphtheria
    C. measles
    D. Haemophilus influenza type b
    E. Mycoplasma pneumoniae

32. In general viral pneumonias are associated more often with
   A. chills
   B. dyspnea
   C. wheezing
   D. higher fever
   E. auscultatory findings of lung consolidation

33. Mild eosinophilia is characteristic of infantile pneumonia caused by
    A. Pseudomonas aeruginosa
    B. Staphylococcus aureus
    C. Chlamydia trachomatis
    D. Mycoplasma pneumoniae
    E. Hemophilus influenza

34. Blood cultures should be performed on hospitalized children to attempt to
    diagnose a bacterial cause of pneumonia. Blood cultures are positive in
    A. 10% to 20% of bacterial pneumonia
    B. 20% to 30% of bacterial pneumonia

228
C. 30% to 40% of bacterial pneumonia  
D. 40% to 50% of bacterial pneumonia  
E. 50% to 60% of bacterial pneumonia

35. If cold agglutinins are present in peripheral blood samples.  
Of the following, the **MOST** suspected pathogen of pneumonia is  
A. Pseudomonas aeruginosa  
B. Staphylococcus aureus  
C. Chlamydia trachomatis  
D. Mycoplasma pneumoniae  
E. Hemophilus influenza

36. Most children recover from pneumonia rapidly and completely, although radiographic abnormalities may return to normal in  
A. 1 to 2 weeks  
B. 2 to 4 weeks  
C. 4 to 6 weeks  
D. 6 to 8 weeks  
E. 8 to 10 weeks

37. Unilateral hyperlucent lung, or Swyer-James syndrome, is a focal sequela of severe necrotizing pneumonia in which all or part of a lung has increased translucency radiographically; it has been linked to  
A. influenza virus  
B. parainfluenza virus  
C. adenovirus type 21  
D. rhinovirus  
E. human metapneumovirus

38. Pulmonary edema is the seepage of fluid into the alveolar and interstitial spaces.  
Of the following, the **MOST** likely cause is  
A. sepsis  
B. acute respiratory distress syndrome  
C. left ventricular dysfunction  
D. tracheal foreign body aspiration  
E. severe obstruction from hypertrophied tonsils and adenoids
39. Acute respiratory failure occurs when the pulmonary system is unable to maintain adequate gas exchange to meet metabolic demands. Of the following, the **MOST** frequent cause is
   A. bronchiolitis
   B. asthma
   C. pneumonia
   D. upper airway obstruction
   E. sepsis/ARDS

40. One of the early signs of hypoxic respiratory failure is
   A. nasal flaring
   B. grunting
   C. diaphoresis
   D. tachypnea
   E. agitation

41. Hypercarbic respiratory failure can occur in
   A. interstitial lung disease
   B. croup
   C. bronchiolitis
   D. fungal pneumonia
   E. sepsis

42. Definitive diagnosis of pulmonary arterial hypertension is made by
   A. ECG
   B. echocardiography
   C. CXR
   D. cardiac catheterization
   E. cardiac MRI

43. The diagnostic test of choice for pulmonary embolism is a
   A. CT angiogram of the chest
   B. ventilation-perfusion scans
   C. Doppler or compression ultrasonography
   D. pulmonary angiography
   E. measurement of D-dimers
44. The sweat chloride test, which is still the standard diagnostic test for cystic fibrosis (CF), is positive if sweat chloride more than
   A. 40 mEq/L
   B. 50 mEq/L
   C. 60 mEq/L
   D. 70 mEq/L
   E. 80 mEq/L

45. Which approximate percent of patients with CF are born with intestinal obstruction caused by inspissated meconium (meconium ileus)?
   A. 10%
   B. 20%
   C. 30%
   D. 40%
   E. 50%

46. The diagnosis of CF should be seriously considered in any infant presenting with failure to thrive, cholestatic jaundice, chronic respiratory symptoms, or electrolyte abnormalities in the form of
   A. hypernatremia, hypochloremia, metabolic alkalosis
   B. hypernatremia, hypochloremia, metabolic alkalosis
   C. hyponatremia, hyperchloremia, metabolic alkalosis
   D. hyponatremia, hypochloremia, metabolic acidosis
   E. hypernatremia, hyperchloremia, metabolic alkalosis

47. All the following are indications for sweat testing **EXCEPT**
   A. chronic or recurrent cough
   B. chronic or recurrent pneumonia
   C. recurrent bronchiolitis
   D. recurrent otitis media
   E. hemoptysis

48. False negative results on sweat testing is seen in
   A. edema
   B. eczema
   C. hypothyroidism
   D. dehydration
   E. malnutrition
49. The treatment of CF is multifactorial, but it is primarily directed toward the gastrointestinal and pulmonary complications. Presently, there is no effective single cure for cystic fibrosis. Regarding treatment, all the following are true **EXCEPT**

A. 7% hypertonic saline delivered by nebulizer decrease the viscosity of mucus
B. antibiotics are selected based on organisms identified by sputum culture
C. common infecting organisms are Pseudomonas aeruginosa and *Staphylococcus aureus*
D. fat should be withheld from the diet when significant steatorrhea exists
E. fat-soluble vitamins (A, D, E, and K) are recommended

50. Transaminase elevation is common in patients with CF, and progressive cirrhosis resulting in portal hypertension is seen in

A. 1% to 3% of patients
B. 4% to 6% of patients
C. 7% to 10% of patients
D. 11% to 14% of patients
E. 15% to 20% of patients

51. The main reason for surgical correction of pectus excavatum is generally to improve

A. appearance (cosmetic reasons)
B. cardiac function
C. exercise tolerance
D. obstructive defects
E. vital capacity

52. Factors predisposing to secondary pneumothorax (underlying cause identified) include all the following **EXCEPT**

A. asthma
B. cystic fibrosis
C. trauma to the chest
D. severe necrotizing pneumonia
E. tall thin males

53. In patients with recurrent or persistent pneumothoraces, sclerosing the pleural surfaces to obliterate the pleural space (pleurodesis) may be necessary. This can be done by instilling sclerosis agents like
A. rifadin  
B. clindamycin  
C. doxycycline  
D. cephalexin  
E. clarithromycin

54. Parapneumonic effusion/empyema is the most common effusion in children. Most parapneumonic effusions are due to pneumonia caused by  
A. Pseudomonas aeruginosa  
B. Chlamydia trachomatis  
C. Mycoplasma pneumoniae  
D. Streptococcus pneumoniae  
E. Hemophilus influenzae

55. The analysis of pleural fluid is useful in differentiating a transudate from an exudate. Transudative pleural effusions have all the following criteria EXCEPT  
A. low specific gravity (<1.015)  
B. low protein content (<2.5 g/dL)  
C. low lactate dehydrogenase activity(<200 IU/L)  
D. low white blood cell (WBC) count  
E. low glucose level (<40 mg/dL)
1. (C). The growth of new alveoli occurs during the first 2 years of life and is complete by 8 years of age. After this time, lung volume increases primarily by increase in alveolar dimensions, with new alveoli rarely formed.

2. (D). PaO₂ does not improve with supplemental oxygen in cyanotic heart disease, pulmonary arteriovenous malformation, and pulmonary edema.

3. (B).
- Consolidation=Dull
- Pneumothorax=Resonant
- Atelectasis=Dull
- Interstitial process=Normal

4. (B). Common causes of chronic cough are asthma, postnasal drip (allergic rhinitis, sinusitis), and postinfectious tussive syndromes. It can also be caused by gastroesophageal reflux disease, swallowing dysfunction (infants), anatomic abnormalities (tracheoesophageal fistula, tracheomalacia), and chronic infection. Persistent cough may also be caused by exposure to irritants (tobacco and wood stove smoke) or foreign body aspiration, or it may be psychogenic in origin.

5. (A). Magnetic resonance imaging (MRI), useful in visualizing cardiac and great vessel anatomy, is less useful for evaluation of pulmonary parenchymal lesions.

6. (A). Ultrasonography can be used to delineate some intrathoracic masses and is the imaging procedure of choice for assessing parapneumonic effusion/empyema. It is useful for assessing diaphragmatic motion in small children.

7. (B). The Pco₂ in venous samples is approximately 6 mm Hg higher than arterial Pco₂. The ratio of the serum bicarbonate concentration to Pco₂ determines the pH. Capillary or venous samples should not be used to assess oxygenation.

8. (A). During the forced expiratory maneuver, forced vital capacity (FVC), forced expired volume in the first second (FEV1), and forced expiratory flow (FEF) rates are measured. These are compared to predicted values based on patient age, gender, and race, but rely mostly on height.

9. (E).

10. (C). Generally, supplemental O₂ should be administered to achieve a goal saturation level above 90%. Normal oxygen saturation is greater than 95%. It is unnecessary to achieve 100% saturation, especially if this requires potentially toxic levels of inspired O₂ for extended periods of time.
11. (B). Chest physiotherapy is most beneficial in children with chronic airway secretions, especially those with CF.
12. (E). Gastroesophageal reflux and laryngospasm are the most common causes of ALTE, and they are associated with emesis, choking, or gagging.
13. (A). The widely advocated supine sleeping position explains, in part, the decreased incidence of SIDS during the past two decades.
14. (E).
15. (E). Laryngomalacia (floppy larynx) is the most common cause of inspiratory stridor in infants and may be aggravated by swallowing problems and gastroesophageal reflux.
16. (D).
17. (A).
18. (B). The most common causes of croup are parainfluenza viruses (types 1, 2, 3, and 4) and respiratory syncytial virus.
19. (C). Spasmodic croup describes sudden onset of croup symptoms, usually at night, but without a significant upper respiratory tract prodrome. These episodes may be recurrent and severe but usually are of short duration. Spasmodic croup has a milder course than viral croup and responds to relatively simple therapies, such as exposure to cool or humidified air. The etiology is not well understood and may be allergic.
20. (D).
21. (A). The most common foreign bodies aspirated by young children are food (especially nuts) and small toys.
22. (A).
23. (E). Respiratory syncytial virus (RSV) is a primary cause of bronchiolitis, followed in frequency by human metapneumovirus, parainfluenza viruses, influenza viruses, adenoviruses, rhinoviruses, coronaviruses, and, infrequently, Mycoplasma pneumoniae.
24. (E). Bronchiolitis occurs primarily in the first year of life and is accompanied by fever, whereas asthma usually presents in older children with previous wheezing episodes typically unaccompanied by fever unless a respiratory tract infection is the trigger for the asthma exacerbation.
25. (B). Bronchodilators and corticosteroids are seldom effective and are not generally recommended.
26. (B). Palivizumab is indicated for some infants under 2 years old with chronic lung disease, very low birth weight, and those with hemodynamically significant cyanotic and acyanotic congenital heart disease.
27. (C).
28. (A). Chlamydia trachomatis and less commonly Mycoplasma hominis, Ureaplasma urealyticum, and cytomegalovirus (CMV) cause a similar respiratory syndrome in infants 1 to 3 months of age with subacute onset of an afebrile pneumonia; cough and hyperinflation are the predominant signs.

29. (A). Infectious agents that commonly cause community-acquired pneumonia vary by age. Most common causes are respiratory syncytial virus (RSV) in infants, other respiratory viruses (parainfluenza viruses, influenza viruses, human metapneumovirus, adenoviruses) in children younger than 5 years old, and Mycoplasma pneumonia in children older than age 5 years.

30. (A). Pneumonia in patients with cystic fibrosis usually is caused by Staphylococcus aureus in infancy and Pseudomonas aeruginosa or Burkholderia cepacia in older patients.

31. (E).

32. (C). In general viral pneumonias are associated more often with cough, wheezing, or stridor; fever is less prominent than with bacterial pneumonia. Mucosal congestion and upper airway inflammation suggest a viral infection.

33. (C). Mild eosinophilia is characteristic of infant C. trachomatis pneumonia.

34. (A).

35. (D).

36. (D).

37. (C). Severe adenovirus pneumonia may result in bronchiolitis obliterans and unilateral hyperlucent lung, or Swyer-James syndrome.

38. (C). Pulmonary edema is most commonly due to heart failure from left ventricular or biventricular dysfunction.

39. (A). Respiratory failure is frequently caused by bronchiolitis (often caused by respiratory syncytial virus), asthma, pneumonia, upper airway obstruction, and sepsis/ARDS. Respiratory failure requiring mechanical ventilation develops in 7% to 21% of patients hospitalized for respiratory syncytial virus.

40. (D). Early signs of hypoxic respiratory failure include tachypnea and tachycardia in attempt to improve minute ventilation and cardiac output and to maintain delivery of oxygenated blood to the tissues.

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

42. (D). Definitive diagnosis is made by cardiac catheterization, but echocardiography may confirm the presence of right ventricular hypertrophy,
ventricular dysfunction, interventricular septal flattening, and tricuspid insufficiency, which can be used to estimate the pulmonary artery pressures.

43.(A).

44.(C). It is positive (elevated sweat chloride > 60 mEq/L) in 99% of patients with CF.

45.(A).

46.(B).

47.(D).

48.(A).

False negative
- edema
- poor technique/inadequate sweat collection
- atypical cystic fibrosis (unusual gene mutations—uncommon)

49.(D). Even with optimal pancreatic enzyme replacement, stool losses of fat and protein may be high. Fat should not be withheld from the diet, even when significant steatorrhea exists.

50.(A).

51.(A).

52.(E). Spontaneous primary pneumothorax (no underlying cause) occurs in teenagers and young adults, more commonly in tall, thin males and smokers. Factors predisposing to secondary pneumothorax (underlying cause identified) include barotrauma from mechanical ventilation, asthma, cystic fibrosis, trauma to the chest, and severe necrotizing pneumonia.

53.(C).

54.(D). Most parapneumonic effusions are due to pneumonia caused by Streptococcus pneumoniae, group A streptococci, or Staphylococcus aureus.

55.(E). Exudates are characterized by high specific gravity and high protein (>3 g/dL) and lactate dehydrogenase (>250 IU/L) levels. They may also have a low pH (<7.2); low glucose level (<40 mg/dL); and a high WBC count with many lymphocytes or polymorphonuclear leukocytes.
1. the **MOST** appropriate action regarding the management of the child in above scenario is to
   A. send for renal ultrasound
   B. send for antinuclear antibody
   C. adjust or stop medications
   D. give non-steroidal anti-inflammatory medications
   E. collect blood culture and start antibiotics

2. The **LEAST** cardiac finding in Kawasaki disease is
   A. coronary artery aneurysm
   B. valvular insufficiency
   C. coronary artery thrombosis
   D. myocardial infarction
   E. myocarditis

3. All the following matching of cardiac manifestations of systemic diseases are true **EXCEPT**
   A. Duchenne dystrophy ......................... cardiomyopathy
   B. Pompe disease ............................... short PR interval
   C. Marfan syndrome ......................... coronary ischemia
   D. systemic lupus erythematosus .... Libman-Sacks endocarditis
   E. neurofibromatosis ......................... pulmonic stenosis

4. The **MOST** common form of congenital heart disease associated with Down syndrome is
   A. patent ductus arteriosus
   B. coaractation of aorta
   C. atrioventricular septal defect
   D. ventricular septal defect
   E. atrial septal defect
5. **VACTERL** association refers to cardiac malformation (usually VSD) and all the following malformations **EXCEPT**
   A. vertebral
   B. anal
   C. tracheoesophageal
   D. respiratory
   E. limbs anomalies

6. The pulse pressure is determined by subtracting the diastolic pressure from the systolic pressure, it is normally below 50 mm Hg or half the systolic pressure, whichever is less. A wide pulse pressure may be seen in
   A. mitral insufficiency
   B. pericardial tamponade
   C. aortic stenosis
   D. anemia
   E. heart failure

7. Inspection is an integral part of cardiac examination. All the following are important aspects to look for in a child with heart disease **EXCEPT**
   A. perioral cyanosis
   B. nutritional status
   C. pallor
   D. clubbing of fingers
   E. dysmorphic features

8. You are measuring blood pressure in an adolescent, which site you would use for measurement?
   A. right arm
   B. left arm
   C. left thigh
   D. right thigh
   E. 4 limbs

9. A single and loud $S_2$ is heard in
   A. TOF
   B. pulmonary atresia
   C. severe aortic stenosis
   D. d-TGA
   E. pulmonary hypertension
10. Of the following, an ejection systolic murmur is heard in
   A. mitral regurgitation
   B. aortic regurgitation
   C. mitral prolapse
   D. ventricular septal defect
   E. atrial septal defect

11. The murmur that is described as "easily heard but not loud" is equal to grade
   A. grade I
   B. grade II
   C. grade III
   D. grade IV
   E. grade V

12. A high pitch murmur is indicative of a/an
   A. large defect
   B. mild obstruction across the valve
   C. restrictive defect
   D. low pressure gradient
   E. innocent murmur

13. You are evaluating a well 4-year-old child in out-patient clinic, you heard a grade-III murmur at lower left sternal border which decreased in intensity when the child is upright; it is of vibratory quality.
   Of the following, the BEST action is to
   A. check 4 limbs blood pressure
   B. order chest X-ray
   C. arrange for an appointment of echocardiography
   D. nominate as Still's murmur
   E. order an ECG

14. On a good inspiratory film, the cardiothoracic ratio in an infant should be less than
   A. 50%
   B. 55%
   C. 60%
   D. 65%
   E. 70%
15. On anterio-posterior chest X-ray film; the major component of right cardiac silhouette is
   A. brachiocephalic vein
   B. inferior vena cava
   C. right atrium
   D. right ventricle
   E. pulmonary artery

16. A 6-year-old boy brought from crowded place during hot season to emergency department with history of collapse to ground after being pale. He lost consciousness with brief tonic movement for about one minute. On examination he was conscious, H.R 70/min, and B.P 60/45 mmhg.
   Of the following, the **MOST** likely diagnosis is
   A. vasovagal syncope
   B. long QT syndrome
   C. hypertrophic obstructive cardiomyopathy
   D. seizure disorder
   E. pulmonary hypertension

17. The family of the child that is presented in question (16) is worried and asking about any investigation needed. Your **BEST** response is
   A. reassurance and discharge to home
   B. ECG
   C. EEG
   D. serum electrolytes
   E. neuroimaging study

18. Of the following, the **MOST** common cause of chest pain in pediatric population is
   A. ischemic pain
   B. severe cough
   C. reflux esophgitis
   D. vaso-occlusive crisis
   E. muscle strain

19. Classically when reading the ECG, we look to P wave to be upright in leads
   A. I and AVR
   B. II and AVF
   C. I and AVF
D. strip of lead II
E. AVL

20. A 4th year medical student asked you for an abnormal cardiac rhythm he felt in a 5-year-old well child. It was varying with respiration, of equal volumes and the rate was 120/min.
Your answer that, this is a/an
A. sinus tachycardia
B. supraventricular tachycardia
C. sinus arrhythmia
D. atrial extrasystoles
E. atrial flutter

21. You are a resident in NICU when called to see a 7-day-old boy who had an abnormal ECG that shows different P wave morphology (upright, notched or biphasic), PR intervals were variable and QRS was normal. He was hemodynamically stable.
Of the following, the MOST likely diagnosis is
A. Wenckebach heart block
B. Mobitz I heart block
C. complete heart block
D. wandering atrial pacemaker
E. supraventricular tachycardia

22. You are attending duty in emergency department, when a 2-week-old baby girl brought with tachycardia. She was mottled with weak pulse and hypotension. The 12-leads ECG shows HR of 242/min, narrow QRS and variable P wave morphology and position.
Of the following, the MOST likely diagnosis is
A. wandering atrial pacemaker
B. sinus tachycardia
C. supraventricular tachycardia
D. atrial flutter
E. ventricular fibrillation

23. The BEST treatment option for the baby in the previous scenario is
A. bag of ice water to face
B. adenosine
C. digoxin
D. sotalol  
E. synchronized cardioversion

24. An 8-year-old asthmatic child on oral theophylline, referred to cardiology clinic because of irregular heart beat. He was hemodynamically stable; his ECG showed premature, wide QRS complexes with large inverted T wave, treadmill ECG testing showed disappearance of ventricular contractions. 
Of the following, the **BEST** action is to  
A. reassure parents and discontinue medication  
B. send for drug level  
C. send for echocardiography  
D. start oral amiodarone  
E. admit and observe for 24 hours

25. Which of the following is a cyanotic congenital heart disease?  
A. atroventricular septal defect  
B. severe aortic stenosis  
C. coarctation of aorta  
D. pulmonary atresia  
E. patent ductus arteriosus

26. An 8-month-old baby boy who is a known case of RSV induced wheeze on salbutamol inhalational therapy, found to have loud cardiac murmur and echocardiography shows small perimemberanous VSD.  
Of the following, the **BEST** advice to the parents is to  
A. reassure and continue on salbutamol  
B. stop salbutamol and refer to pediatric cardiologist  
C. start diuretics therapy  
D. start afterload reducing agent  
E. refer for cardiac closure device

27. A 3-month-old baby boy who is a known case of large VSD with failure to gain weight has fussiness, excessive sweating, grade III/VI systolic murmur at LLSB with faint diastolic murmur at apex and loud S2. ECG showed right ventricular hypertrophy. He is on anti-failure therapy (diuretics, afterload reducing agent and digoxin). His mother is describing attacks of cyanosis with crying.  
Of the following, the **BEST** action is to  
A. escalate antifailure therapy  
B. advice for serial echocardiography
C. refer to pediatric cardiologist  
D. refer for CT angiography scan  
E. add a potent diuretics

28. Of the following, the **LEAST** common type of ASD is  
A. patent foramen ovale  
B. secundum  
C. primum  
D. sinus venosis  
E. complete atrioventricular canal

29. The type of VSD which is nearest to the arterial valves (aorta or pulmonary) is called  
A. perimembranous  
B. supracrisal  
C. muscular  
D. inlet  
E. apical

30. You are evaluating a 7-day-old full-term baby girl with improving respiratory distress due to congenital pneumonia, the parents concern about echocardiography which shows small PDA.  
Of the following, your **BEST** answer is  
A. almost all PDAs discovered in this time will be closed  
B. we need to do another echo after few weeks  
C. no need for follow up as these defects rarely causes symptoms  
D. starting diuretic therapy will improve the outcome  
E. giving 3 days course of parenteral iboprunen

31. A 10-week-old infant presented with tachypnea, tachycardia, and faint cardiac murmur with load S₂, saturation was 94% with 2 liters of nasal O₂, cardiomegaly on chest X-ray, ECG shows biventricular hypertrophy, biphasic P wave with left superior axis. You are discussing the possibilities with a senior colleague.  
Of the following, the **MOST** likely diagnosis is  
A. large VSD  
B. tricuspid atresia  
C. endocardial cushing defect  
D. left hypoplastic heart syndrome  
E. right hypoplastic heart syndrome
32. A 3-year-old girl referred from health center with history of recurrent chest infections and cardiac murmur. On examination, she found to be under weight, having wide pulse pressure, and continuous, machine-like murmur heard at the left infraclavicular area, radiating to the back.
Of the following, the MOST likely diagnosis is
   A. peripheral pulmonary stenosis
   B. coaractation of aorta
   C. tricuspid regurgitation
   D. patent ductus arteriosus
   E. aortic regurgitation

33. A 4-year-old boy observed by his parents to have exertional dyspnea and easy fatigability. On examination found a well-child with no dysmorphic features with grade III systolic murmur at the second left intercostal space that radiates to the back. S2 is widely split, and a heave is felt at the lower left sternal border.
Of the following, the MOST likely diagnosis is
   A. peripheral pulmonary stenosis
   B. patent ductus arteriosus
   C. aortic stenosis
   D. pulmonic stenosis
   E. coaractation of aorta

34. The echocardiography done for the patient mentioned in the question 33 shows severe subvalvular muscular pulmonic stenosis.
Of the following, the BEST option for management is
   A. surgical repair
   B. balloon valvoplasty
   C. diuretics
   D. afterload reducing agents
   E. watchful waiting according to symptoms

35. A young parents consulted you about the result of echocardiography for their 9-month-asymptomatic baby with mild pulmonary stenosis.
Of the following, the MOST appropriate response is that
   A. we need to do follow him up by serial echocardiography
   B. we need to arrange catheterization for him
   C. the problem is not progressive especially if it is mild
   D. the option of balloon valvuloplasty is valid if became more severe
   E. the surgery is the last option for treatment
36. A 12-year-old adolescent boy complaining from leg discomfort with exercise, headache, and frequent epistaxis. A continuous murmur heard throughout the chest and back. He was well with good body built but the blood pressure found to be high.

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

A. check pulses in all extremities
B. do complete neurological examination
C. send for MRI of lower spine
D. send for renal ultrasound
E. send for echocardiography

37. You are evaluating a 9-year-old child with coaractation of aorta.

Of the following, the **MOST** likely associated anomaly is

A. hypoplastic aortic arch
B. ventricular septal defect
C. coaractation of abdominal aorta
D. corrected transposition of great arteries
E. abnormal aortic valve

38. A 6-day-old baby boy presented with sudden severe respiratory distress and grunting. Heart rate was 180/min, RR 80/min, B.P 50/40, O₂ saturation 75% with 0.6 FiO₂. No evident murmur heard but evident hepatomegaly. Chest X-ray shows cardiomegaly and increased lung plethora.

All the following lesions can have similar presentation **EXCEPT**

A. total anomalous pulmonary venous drainage
B. hypoplastic left heart syndrome
C. coaractation of aorta
D. single ventricle
E. d-TGA

39. The echocardiography of the baby in question 38 shows severe coaractation of aorta.

Of the following, the **BEST** action is to do/give

A. urgent balloon angioplasty
B. infusion of prostaglandin E1
C. diuretics
D. inotropes
E. surgery
40. The **MOST** common cyanotic lesion presenting in the newborn period is
   A. tetralogy of Fallot
   B. d-transposition of great arteries
   C. l-transposition of great arteries
   D. pulmonary atresia
   E. hypoplastic left heart syndrome

41. Tetralogy of Fallot is composed of ventricular septal defect (VSD), overriding aorta, right ventricular hypertrophy, and **COMMONLY** with
   A. subvalvular pulmonary stenosis
   B. valvular pulmonary stenosis
   C. supravalvular pulmonary stenosis
   D. atretic pulmonary valve
   E. absent pulmonary valve

42. In tetralogy of Fallot, the coronary anatomy is required by surgeon (just to avoid its injury during surgical repair). The associated coronary anomalies in TOF is about
   A. 1%
   B. 5%
   C. 10%
   D. 15%
   E. 20%

43. The management of hypoxic (Tet) spells include all the following **EXCEPT**
   A. O₂ administration
   B. knee-chest position
   C. sodium bicarbonate infusion
   D. morphine sulfate IM
   E. phenylephrine IV

44. A concerned parents of a 2-month-old baby boy who recently diagnosed as tetralogy of Fallot asking about the timing of operation. You told them that the main decisive factor in timing of surgical repair depends on the
   A. degree of pulmonary stenosis
   B. occurrence of cyanotic spells
   C. degree of right ventricular hypertrophy
   D. degree of nail clubbing and O₂ saturation
   E. development of heart failure
45. A 5-year-old girl who had recently operated successfully for TOF, she had dental decay and want to attend dental clinic. The correct statement regarding subacute bacterial endocarditis prophylaxis
   A. no need for prophylaxis
   B. it is indicated until 12 months after complete repair
   C. 2 years of prophylaxis is indicated
   D. an 18 months of prophylaxis is usually enough
   E. should be continued for 6 months after complete repair

46. A fullterm baby boy presented with cyanosis and tachypnea, his echocardiography showed d-Transposition of great arteries with ASD and PDA, he is on prostaglandin E1 infusion. His mother was much concerned about the timing of operation.
   Of the following, the BEST answer is that
   A. it depends on clinical progress of the disease
   B. usually performed in first 2 months of life
   C. we will perform a serial echocardiography to assess the ventricular function
   D. it depends on type of surgery we are going to do
   E. it's better to be done in first 2 wks of life

47. All the following congenital heart diseases may present without murmur, but the one which always present without cardiac murmur is
   A. coaractation of aorta
   B. tricuspid atresia
   C. total anomalous pulmonary venous return
   D. transposition of great arteries
   E. hypoplastic left heart syndrome

48. Subclavian artery-to-pulmonary shunt (Blalock-Taussig procedure, B-T shunt) is a famous palliative surgery in pediatric cardiology. It can be helpful in
   A. pulmonary atresia
   B. d-TGA
   C. hypoplastic left heart syndrome
   D. total anomalous pulmonary venous drainage
   E. truncus arteriosus

49. The use of prostaglandin E1 may be useful in all of the following lesions EXCEPT
   A. d-TGA
B. hypoplastic left heart syndrome  
C. tricuspid atresia  
D. truncus arteriosus  
E. pulmonary atresia

50. You are evaluating a 5-year-old girl with non-operated complex cyanotic congenital heart disease (CCHD) in cardiology clinic, she is complaining of fatigue, tightness of the chest and numbness of limbs. She is hemodynamically stable but lab. test shows HTC of 68%.

Of the following, the MOST appropriate next step in treatment is

A. phlebotomy  
B. assessing iron level  
C. rehydration  
D. watchful waiting  
E. refer to neurologist

51. Of the following ECG findings, the one that is BEST match pericarditis (with or without effusion) is

A. decrease rate  
B. electrical alternans  
C. wide QRS voltage  
D. short PR interval  
E. depressed ST segments

52. Of the following, the complication that is related to right-left shunt in cyanotic congenital heart disease is

A. gingival hypertrophy  
B. retarded growth  
C. CNS stroke  
D. lung abscess  
E. clubbing

53. The major determinants of cardiac function include all the following EXCEPT

A. heart rate  
B. left end diastolic volume  
C. cardiac contractility  
D. afterload  
E. stroke volume
54. In a fetus, all the following are possible causes of heart failure **EXCEPT**
   A. fetal-maternal transfusion
   B. SVT
   C. atrioventricular valve insufficiency
   D. large mixing defects
   E. teratomas

55. The principle cause that is lead to the development of heart failure in infants with left-right shunt after 2 mo
   A. the shunt across the defect is increased
   B. the pulmonary flow to systemic flow is less than 2-1
   C. the flow across the defect tend to be less restrictive in this age
   D. the pulmonary vascular resistance is deceased in this age
   E. this is the age when early pulmonary hypertension is usually developed

56. In predominantly left sided heart failure, all the following are recognized findings **EXCEPT**
   A. hepatomegaly
   B. tachypnea
   C. orthopnea
   D. wheezing
   E. pulmonary edema

57. A previously well 2-month-old baby presented to ER with tachypnea, examination shows HR of 170/min, good peripheral pulses, normal blood pressure and \( O_2 \) saturation was 89%. The cardiac murmur was grade II at LLSB with faint diastolic murmur at apex. Chest X-ray shows CT ratio of 72% and echocardiography shows large VSD. You admitted the baby and started supportive care (semi-sitting position and \( O_2 \)).
   Of the following, the **BEST** option of treatment is to start
   A. loop diuretics
   B. loop diuretics and afterload reducing agent
   C. loop diuretics, afterload reducing agent and digoxin
   D. loop diuretics, afterload reducing agent and spironolactone
   E. inotropes plus option D

58. Of the following, the **LEAST** clinical finding in acute rheumatic fever is
   A. migratory arthritis
   B. pancarditis
C. subcutaneous nodule
D. erythema marginatum
E. Sydenham chorea

59. Of the following, the criteria of rheumatic fever which does not need evidence of recent group A streptococcal disease is
   A. migratory arthritis
   B. Sydenham chorea
   C. pancarditis
   D. subcutaneous nodule
   E. erythema marginatum

60. All the following are minor criteria for acute rheumatic fever EXCEPT
   A. fever (39°- 40°C)
   B. prolonged PR interval
   C. elevated CRP
   D. previous rheumatic fever
   E. arthralgia

61. A 6-year-old boy who had diagnosed as acute rheumatic fever with arthritis and carditis. Parents were so worried about heart lesion.
   Of the following, the MOST appropriate statement describing the prognosis of heart disease
   A. actually it is a chronic disease which may lead to valve stenosis
   B. the chance of cure is minimal and long term prophylaxis is indicated
   C. most patients will recover that illness after proper treatment
   D. prednisolone therapy can cure almost all the cardiac lesions
   E. oral prophylaxis is indicated for 5 years

62. In dilated cardiomyopathy the usual presenting sign/symptom is
   A. chest pain
   B. cardiogenic shock
   C. sudden death
   D. syncope
   E. tachypnea

63. The echocardiographic description of massive atrial dilation is seen in
   A. restrictive cardiomyopathies
   B. dilated cardiomyopathies
C. hypertrophic cardiomyopathies  
D. viral myocarditis  
E. noncompaction cardiomyopathies

64. In hypertrophic cardiomyopathy, the drug of choice usually is  
   A. positive inotropes  
   B. diuretics  
   C. afterload reduction  
   D. β-Blockers  
   E. antiarrhythmics

65. A 6-year-old boy who is on maintenance treatment of acute lymphoblastic leukemia with CNS infiltration, received irradiation course in the last month. He developed chest pain which seems to be reduced with sitting position, and dyspnea. By examination you found the HR 140/min, distended neck veins, inspiratory increase in jugular venous pressure, distant heart sounds, pericardial knock, hepatomegaly and mild ascites. Of the following the MOST likely diagnosis is  
   A. restrictive cardiomyopathy  
   B. constrictive pericarditis  
   C. dilated cardiomyopathy  
   D. hypertrophic cardiomyopathy  
   E. viral myocarditis

66. A 5-year-old girl with large ASD who had recently underwent surgical repair, came to you on follow up with fever, on evaluation you heard distant heart sounds and friction rub otherwise, the child is hemodynamically stable. Acute phase reactant was increased with leukocytosis and shift to left. Echocardiography showed mild-moderate pericardial effusion. Of the following, the MOST likely cause is  
   A. cardiac tamponade  
   B. infective endocarditis  
   C. postcardiotomy syndrome  
   D. wound infection  
   E. rheumatic fever

67. You are evaluating a 3-year-old boy with truncus arteriosus; he is on antifailure medications (diuretics and ACEi). Mother said that, the child has painful swelling of his ankle with intermittent red urine.
Of the following, the **MOST** likely diagnosis is

A. uric acid nephropathy  
B. bacterial endocarditis  
C. lupus nephritis  
D. drug induced hemolytic anemia  
E. bacterial sepsis
1. (C). The child is on antifailure therapy; diuretics including loop and thiazide diuretics can cause hyperuricemia which manifest in this child as uric acid nephropathy and arthropathy. Initially you need to do RFT and s.uric acid; stop diuretic and to start allopurinol.

2. (B).

3. (C). Patients with Marfan syndrome may have aortic insufficiency, mitral insufficiency, or dissecting aortic aneurysm.

4. (C). About 60% of persons with Down syndrome have congenital heart disease; of those about 60% are affected by endocardial cushion defect (atrioventricular septal defect).

5. (D). Renal.

6. (D). The wide pulse pressure may be seen with aortopulmonary connections (patent ductus arteriosus, truncus arteriosus, arteriovenous malformations), aortic insufficiency, or relative intravascular volume depletion (anemia, vasodilation with fever or sepsis). A narrow pulse pressure is seen with pericardial tamponade, aortic stenosis, and heart failure.

7. (A). Perioral cyanosis is a common finding, especially in pale infants or when infants and toddlers become cold. Isolated peripheral cyanosis (hands, feet) is associated with normal arterial saturation and increased peripheral extraction of oxygen. Central cyanosis (tongue, lips) is associated with arterial desaturation.

8. (A). Initially, blood pressure is measured in the right arm. If elevated, measurements in the left arm and legs are indicated to rule out coaractation of the aorta.

9. (E). In TOF and pulmonary atresia it will be single but decreased intensity, in severe aortic stenosis it will paradoxically spilt, and in d-TGA it will be single with normal intensity.

10. (E). In mitral regurgitation and ventricular septal defect the murmur are pansystolic, in aortic regurgitation is diastolic, while in mitral prolapse it is late systolic with ejection click, in ASD the murmur result from increase flow through the pulmonary valve. Systolic ejection murmurs require the ejection of blood from the ventricle and may occur with aortic stenosis, pulmonary stenosis, atrial septal defect (ASD), and coaractation of the aorta.

11. (B).
12. (C). In case of high frequency (high pitch) murmur, it is usually indicative of a smaller defect is present (like small VSD) or more severe obstruction (like moderate or severe PS).

13. (D). Still's murmur is one of the innocent cardiac murmurs that manifest between the ages of 3-6 years, it is a systolic ejection murmurs that is usually heard at LLSB or between LLSB and apex, with grades I-III/VI, of vibratory or musical quality and it is intensity decreases in upright position.

14. (B). And less than 50% in older children and adolescents.

15. (C). The component of right cardiac silhouette from top to bottom are superior vena cava then right atrium, while the component of left cardiac silhouette from top to bottom are aorta; pulmonary artery; left atrium; left ventricle. Assessment of the location and size of the heart and cardiac silhouette may suggest a cardiac defect.

16. (A). Vasovagal syncope, usually happens after stimulation (like what happens in given scenario) while in neurocardiogenic syncope, it happens during rest. Long QT syndrome can be excluded by normal ECG, while in hypertrophic obstructive cardiomyopathy; the episode occurs after exercise and associated with chest pain and shortness of breath. A seizure disorder does not fit the prodrome and postictal phase of the given scenario, while the presentation widely differs in pulmonary hypertension (cyanosis after exercise).

17. (B). ECG is mandatory in syncopal disorders to rule out long QT syndromes (measuring corrected QT interval which should be less than 0.45 msec), after that reassurance and anticipation (means trying to avoid similar situation in future) of events are the only discharging massages to the family.

18. (E). Most diagnosable chest pain in childhood is musculoskeletal in origin (osteocondritis and muscle overuse and strain). A significant amount remains idiopathic. Ischemia is associated only in certain situations like coronary artery abnormalities which presents early in life, severe AS or PS and HOCM. Pulmonary causes are also common as asthma (often exercise-induced), severe cough and pneumonia. Reflux esophagitis remains in between the common gastrointestinal causes of chest pain.

19. (C). Looking for sinus rhythm and the normal axis of P wave in lead I and AVF when it should be upright. P wave is inverted in AVR. In dextrocardia P wave in both (lead I and AVF) may be inverted in addition to right axis deviation and upright P wave and QRS complexes in AVR.

20. (C). Sinus arrhythmia is a common finding in children and represents a normal variation in the heart rate associated with breathing. The heart rate increases with inspiration and decreases with expiration, producing a recurring pattern on the
electrocardiogram (ECG) tracing. Sinus arrhythmia is normal and does not require further evaluation or treatment.

21. (D).

22. (C). Supraventricular tachycardia is the most common symptomatic dysrhythmia in pediatric patients (supraventricular is narrow QRS tachycardia while ventricular is wide QRS tachycardia). If there is structural heart disease or the episode is prolonged (>12 hours), there may be alteration in the cardiac output and development of symptoms of heart failure.

23. (E). In hemodynamically stable child the treatment options start with vagal maneuvers then adenosine. Digoxin can be used in patients how are known not to have WPW syndrome as maintenance therapy. Oral sotalol can be used with certain precautions by doses according to age. Synchronized cardioversion is the definite option in the hemodynamically unstable children.

24. (B). Patients with premature ventricular contractions with normal echocardiography and treadmill test can be discharged home and no treatment is required. But in this scenario the adjustment of theophylline dose and or drug level is more important as it may be the cause of this event. Ventricular tachycardia defined as three or more consecutive premature ventricular contractions which need treatment in symptomatic child with synchronized cardioversion shock and in asymptomatic child with lidocaine or amiodarone.

25. (D). Acyanotic congenital heart disease includes left-to-right shunts resulting in an increase in pulmonary blood flow (like PDA, VSD, ASD) and obstructive lesions (like AS, PS, COA), which usually have normal pulmonary blood flow, while pulmonary atresia is a cyanotic congenital heart disease and it is usually a duct-dependent lesion.

26. (A). Inhalational and nebulized B-agonist has no effect on cardiac status. Small left-right lesions usually close spontaneously and does not cause cardiac failure.

27. (C). The baby in this scenario shows evidence of pulmonary hypertension (attacks of cyanosis with crying and his ECG shows right ventricular predominance). Starting sildenafil and arranging for early surgical intervention (either complete correction or easier operation of pulmonary artery banding) is the best action. It should be referred to pediatric cardiologist (again) to take an action.

28. (D). It can be associated with anomalous pulmonary venous return. Secundum is the most common type. Patent foramen ovale is a normal defect at atrial septum which can close spontaneously later on. Complete atrioventricular canal is a separate entity which present more commonly in patients with Down syndrome.

29. (B). Which carries the highest risk of valve regurgitation and usually needs surgical closure even if it is small.
30. (B). Most PDAs discovered in a full-term few days after birth are functional and usually closes within few weeks, so repeating an echocardiography after few weeks is the most reasonable answer. Small PDAs which remains opened; closing them by closure device is controversial. The use of prostaglandin inhibitors for significant PDAs has a role in preterm babies.

31. (C). This scenario shows an infant presented with heart failure sooner after reduction of pulmonary vascular resistant by 8 wks of life which usually happens with large Lt-Rt shunts. Presence of left axis deviation and absence of cyanosis make the diagnosis of complete atrioventricular septal defect (one type of endocardial cushing defects) the most likely one, while in presence of cyanosis, tricuspid atresia is the most likely diagnosis.

32. (D). In between the mentioned options wide pulse pressure can present in aortic regurgitation and PDA, but with AR the murmur is usually diastolic.

33. (D). All the mentioned options have a murmur which radiates to the back except AS which radiate to the neck. Peripheral pulmonary stenosis usually does not cause symptoms. Other options can be differentiated by the character of murmur. Presence of left sternal heave which is associated with right ventricular hypertrophy, support the diagnosis of PS.

34. (A). Surgical repair is required if balloon valvuloplasty is unsuccessful for valvular PS or when subvalvular muscular stenosis is present.

35. (C). Valvular pulmonary stenosis usually does not progress, especially if it is mild.

36. (A). The murmur of coaractation of aorta (COA) is typically best heard in the left interscapular area of the back. If significant collaterals have developed, continuous murmurs may be heard throughout the chest. For the above scenario the best next action is checking the pulse in all extremities and checking the 4 limbs blood pressure. Doing complete neurological assessment is also important but not the first choice. MRI of lumbosacral area to rule out tethered cord. After complete and proper physical examination sending for echocardiography and renal ultrasound will be next most appropriate action.

37. (E). An abnormal aortic valve is present approximately 50% of the time, causing a systolic ejection click and systolic ejection murmur of aortic stenosis. Hypoplastic aortic arches, abnormal aortic valves, and VSDs are frequent associations with infantile type of COA.

38. (D). The presentation of hypoperfusion, right sided volume overload, cyanosis and respiratory distress are common presentation of ductal dependent lesions (option B, C, and also in E in certain situation when no VSD and the ASD is restrictive and small). In option A this is usually the presentation with obstructive
type. In option D the presentation is usually later (it may take 2-3 month) when the symptoms of heart failure develops rather than cyanosis.

39. (B). All the options are true but in order of B; C; D; A and later on E.

40. (B). TOF is the most common cyanotic congenital heart defect, while hypoplastic left heart syndrome is the most common cause of death from cardiac defects in the first month of life. D-TGA (transposition of great arteries) is the most common cyanotic heart defect present in neonatal period.

41. (A). Infundibular or subvalvular is the most common associated form of pulmonary stenosis; other types are recognized but less frequent.

42. (B).

43. (C). The mechanism of Tet spells is secondary to Infundibular spasm and/or decreased systemic vascular resistance (SVR) resulting in increased right-to-left shunting at the ventricular septal defect (VSD), and in diminished pulmonary blood flow. If left untreated, it may result in syncope, seizure, stroke, or death. Morphine sulfate IM is used for relaxing pulmonary infundibulum and phenylephrine IV for increasing SVR. Treating acidosis with sodium bicarbonate may reduce the respiratory center stimulation.

44. (B). The occurrence of cyanotic spell is an indication to proceed with surgical repair.

45. (E). Subacute bacterial endocarditis prophylaxis is indicated until 6 months after complete repair. In case of residual VSD, it should continue until it is closure.

46. (E). The arterial switch usually is performed within the first 2 weeks of life, when the left ventricle can still maintain systemic pressure.

47. (E).

48. (A). The BT shunt mimics the role of the ductus arteriosus. It is often put in place after the ductus closes naturally. It is used for defects that affect the flow of blood from the right ventricle, through the pulmonary artery, and to the lungs, like pulmonary atresia, pulmonary stenosis, and tricuspid atresia. Shunts are usually used for four to five months, until the child outgrows them and a second operation or definitive repair is needed.

49. (D). A temporary measure to keep the ductus arteriosus open in duct-dependent cyanotic congenital heart diseases meanwhile preparing for palliative surgery or definitive repair.

50. (A). In case of symptomatic hyperviscosity in CCHD and PCV >65%, the first assessment is to rule out acute causes as dehydration (which is clearly not the cause in this scenario); otherwise phlebotomy is indicated; gradual withdrawal of 10 ml/kg of blood with replacement with saline should be done. If PCV < 65%, we need to check for iron body status (s.ferritin or iron level), if low they need iron replacement otherwise no need for treatment.
51.(B). Depressed PR interval with increase rate, elevated ST segments, reduced QRS voltage or electrical alternans (variable QRS amplitude) are ECG findings in acute pericarditis which is usually involves all leads.
52.(C). And CNS abscess.
53.(E).
54.(D). Large mixing defects as single ventricle or truncus arteriosus can cause heart failure in neonatal period. Teratomas represent high-output cardiac failure.
55.(D). Reduction in pulmonary vascular resistance leads to increase the shunt across the defect and consequently to increase the pulmonary flow which predispose to heart failure.
56.(A). Distended neck veins, hepatomegaly and edema, are signs of right-sided failure.
57.(D). In case of heart failure with L-R shunt and when no signs of cardiogenic shock, the best option to start with loop diuretics, afterload reducing agent and digoxin. The diuretics are used to reduce the preload and also to avoid pulmonary edema. Afterload reducing agent (as ACEi), starting in a low dose which can be escalated gradually keeping an eye on blood pressure; it reduces angiotensin II which is a potent vasoconstrictor. Spironolactone is usually added to the medical regimen because of its effect on cardiac remodeling. In case of hypotension or cardiogenic shock, inotropes is indicated. It is important not to start the medication in aggressive way, excessive diuretics or ACEi may cause an abrupt reduction in end-diastolic pressure which can further reduce the stroke volume.
58.(C). Subcutaneous nodules are seen predominantly with chronic or recurrent disease.
59.(B). It consists of neurologic and psychiatric signs. It also is uncommon and often presents long after the infection. It may be the only presenting sign.
60.(A). Fever is usually in the range of 38.2°–38.9°C.
61.(C). Most patients with carditis (50-70%) will recover, the remaining need follow up for possibility of development of stenotic valvular lesions.
62.(E). Children with dilated cardiomyopathy present with signs and symptoms of inadequate cardiac output and heart failure as tachypnea and tachycardia, concurrent infectious illness may result in circulatory collapse and shock in children with dilated cardiomyopathies. Hypertrophic cardiomyopathy in infants is present with signs of heart failure; while in older children sudden death may be the initial presentation. Dyspnea, fatigue, chest pain, syncope or near-syncope, and palpitations may be present. The murmur is heard in more than 50% of children referred after identification of an affected family member. Restrictive cardiomyopathies the presentation usually includes dyspnea exacerbated by a respiratory illness, syncope and hepatomegaly.
63.(A). In dilated cardiomyopathies result in left atrial and ventricular dilation, a decreased shortening fraction, and globally depressed contractility. Asymmetrical septal hypertrophy and left ventricular outflow tract obstruction are seen in hypertrophic cardiomyopathies.

64.(D). The problem in hypertrophic cardiomyopathy is diastolic dysfunction were the end diastolic volume is reduced and the stroke volume consequently is reduced also. So the role of medications is to relax the heart muscle and to slow the heart rate so that the heart can pump more efficiently (inotropes is contraindicated). Other options are good for dilated cardiomyopathy while diuretics are helpful in restrictive type.

65.(B). Constrictive pericarditis secondary to radiation exposure with sings of increase in pulmonary pressure. The child received a course of irradiation which leads to option B, other common causes are postsurgical and idiopathic. TB is a rare cause. Presence of Kussmaul sign (inspiratory increase in jugular venous pressure) with predominance of right sided heart signs make the diagnosis is in favor of option B.

66.(C). It is a common complication after an open heart surgery due to immunological phenomena with up to 30% of cardiac surgery. Treatment is usually with anti-inflammatory medications and colchicine.

67.(A).
1. The **LEAST** indicator for an underlying hematological problem is
   A. pallor
   B. fever
   C. petechial skin rash
   D. lymphadenopathy
   E. failure to thrive

2. During intrauterine life, hematopoiesis usually begins by
   A. 3\textsuperscript{rd} week of gestation
   B. 5\textsuperscript{th} week of gestation
   C. 7\textsuperscript{th} week of gestation
   D. 9\textsuperscript{th} week of gestation
   E. 11\textsuperscript{th} week of gestation

3. The primary site of hematopoiesis in the fourth week of gestational age is
   A. liver
   B. spleen
   C. lymph nodes
   D. yolk sac
   E. bone marrow

4. Which of the following symptoms/sings is a manifestation of anemia?
   A. irritability
   B. pharyngitis
   C. bruising
   D. fatigue
   E. gingivitis

5. Which of the following symptoms/signs is a manifestation of polycythemia?
   A. lymphadenopathy
   B. deep vein thrombosis
   C. epistaxis
   D. seizures
   E. oral ulceration
6. A true hematological index for a cord blood sample is
   A. reticulocyte of 1%
   B. leukocyte of 6,000/cmm
   C. hemoglobin of 16.0 gm/dl
   D. neutrophils of 30%
   E. platelets of 90,000/cmm

7. Males, at onset of puberty, maintain a normal hemoglobin value about 1.5-2.0 gm/dl higher than females because of
   A. oxygen affinity of RBC is lower in males
   B. bone marrow production of RBC precursors in higher in males
   C. androgen release at the onset of puberty
   D. medullary and extramedullary site for precursors production is potent in males
   E. the RBC count is higher in males during early embryogenesis

8. An example of macrocytic anemia is
   A. thalassemia
   B. iron deficiency anemia
   C. anemia of chronic disease
   D. glucose 6-phosphate dehydrogenase deficiency
   E. folic acid deficiency

9. The urgency for packed cell transfusion in childhood anemia should be dictated by
   A. absolute level of hemoglobin
   B. extent of functional impairment
   C. age of the patient
   D. gender of the patient
   E. presence or absence of splenomegaly

10. The LEAST helpful historical clue in Iron deficiency anemia is
    A. pica
    B. age
    C. ingestion of cow’s milk
    D. Giardia lamblia infection
    E. intestinal surgery

11. The LEAST helpful historical clue in the diagnosis of hemolytic anemia is
A. mycoplasma infection
B. gallstones
C. splenectomy of a family member
D. chronic infection
E. affected siblings

12. An 8-year-old boy presented with recurrent skin ecchymoses for the last few months. On examination; he has café-au-lait spots, microcephaly and absent thumb.
Of the following, the **MOST** likely diagnosis is
   A. neurofibromatosis
   B. Fanconi anemia
   C. Peutz-Jeghers syndrome
   D. sickle cell anemia
   E. dyskeratosis congenita

13. A 5-year-old boy presented with history of frequent blood transfusion since the end of the first year of life, he has a similarly affected older sibling. On examination: he is pale and jaundiced, with frontal bossing and enlarged liver and spleen.
Of the following, the **MOST** likely diagnosis is
   A. Diamond Blackfan anemia
   B. thalassemia major
   C. Peutz-jeghers syndrome
   D. sickle cell anemia
   E. dyskeratosis congenita

14. A 13-month-old girl presented with progressive pallor for the last 3 months. The mother noticed that she started to eat a non-nutritive and dirty substance. She is on breast feeding till this age. On examination, she is pale, apathetic looking, no LAP, no organomegal, and no skin bleeds.
Of the following, the **MOST** likely diagnosis is:
   A. iron deficiency anemia
   B. folic acid deficiency
   C. B₁₂ deficiency
   D. vitamin E deficiency
   E. aplastic anemia
15. Vitiligo, glossitis, ataxia, and peripheral neuropathy are features of
   A. iron deficiency anemia
   B. vitamin B\textsubscript{12} deficiency
   C. folic acid deficiency
   D. heavy metal intoxication
   E. thalassemia

16. Growth retardation is not evident in anemia of
   A. chronic disease
   B. malnutrition
   C. Fanconi anemia
   D. HIV infection
   E. spherocytosis

17. The individual that is \textbf{least} susceptible to develop iron deficiency anemia is a
   A. 6-year-old girl with juvenile rheumatoid arthritis
   B. 13-year-old girl started menses few months ago
   C. 7-year-old girl with retinoblastoma
   D. 3-year-old boy fed large volume of cow’s milk
   E. 10-month-old breast-fed infant not started solid food yet

18. The \textbf{most} prevalent anemia in the world is
   A. thalassemia
   B. pyruvate kinase deficiency
   C. B\textsubscript{12} deficiency anemia
   D. Diamond Blackfan anemia
   E. iron deficiency anemia

19. Which percent of iron deficient children develop iron deficiency anemia?
   A. 10%
   B. 30%
   C. 50%
   D. 70%
   E. 100%

20. Iron deficiency anemia is less prevalent in breast fed infants because
   A. breast milk contains higher amount of iron than cow’s milk
   B. mothers status of iron is enough for the infant in the first year of life
   C. iron in human milk is more efficiently absorbed
D. fortification of cow’s milk formula is inadequate
E. other supplements in cow’s milk formula competes with iron

21. Which of the following central nervous system (CNS) abnormalities have been linked to iron deficiency anemia?
   A. seizure
   B. ataxia
   C. poor concentration
   D. blindness
   E. sleep disturbance

22. The best diagnostic study for iron deficiency in an otherwise healthy child is
   A. serum ferritin
   B. therapeutic trial
   C. serum iron
   D. blood film
   E. iron binding capacity

23. The earliest response to oral iron, in iron deficiency anemia is
   A. improved neurologic function
   B. reticulocytosis
   C. increased hemoglobin
   D. reduction in iron binding capacity
   E. changes in blood film

24. The increment in hemoglobin after instituting oral iron therapy for iron deficiency anemia is about
   A. 1.1 gm/dl/day
   B. 0.9 gm/dl/day
   C. 0.7 gm/dl/day
   D. 0.5 gm/dl/day
   E. 0.3 gm/dl/day

25. An illogical cause of failure of increment of hemoglobin after starting treatment of iron deficiency anemia is
   A. poor compliance
   B. infection
   C. excessive formula milk intake
   D. wrong dosing
E. ongoing blood loss

26. Which of the following is **TRUE** regarding prevention of Iron deficiency anemia in infants?
   - A. introduction of solid foods at 3 months of age for a bottle fed infant
   - B. addition of iron-containing formula to breast fed infant
   - C. use of special high-calories formula to a bottle fed infant
   - D. addition of iron supplement after 6 months of age to a breast fed infant
   - E. introduction of solid food at 3 months of age for a breast fed infant

27. A common significant feature of blood smear in lead poisoning is
   - A. normochromic RBC
   - B. macrocytic RBC
   - C. basophilic stippling
   - D. target cells
   - E. raised reticulocytes

28. A feature that signifies β-halassemia minor is
   - A. low iron level
   - B. high iron binding capacity level
   - C. absent marrow sideroblasts
   - D. elevated hemoglobin A2
   - E. high red cell distribution width

29. Mentzer index of less than 12.5 suggests thalassemia trait, it represents
   - A. MCV divided by RBC count
   - B. MCH divided by RBC count
   - C. MCHC divided by RBC count
   - D. PCV divided by RBC count
   - E. Hb divided by RBC count

30. Which of the following hemoglobinopathies is transfusion dependent?
   - A. alpha thalassemia trait
   - B. heterozygous beta thalassemia
   - C. hemoglobin H disease
   - D. homozygous β-halassemia
   - E. sickle trait
31. Which of the following results indicates Iron deficiency anemia?
   A. high serum ferritin
   B. high red cell distribution width
   C. increased marrow iron
   D. raised marrow sideroblasts
   E. reduced free erythrocyte protoporphyrin

32. Which of the following viruses can cause aplastic crises in chronic hemolytic disease?
   A. Herpes virus
   B. Parvovirus B19
   C. Cytomegalovirus
   D. Rota virus
   E. T-lymphocyte virus

33. A challenging differential diagnosis of Diamond-Blackfan syndrome is
   A. iron deficiency anemia
   B. thalassemia syndrome
   C. transient erythroblastopenia of childhood
   D. spherocytosis
   E. aplastic anemia

34. Which liver protein plays a key role in iron homeostasis?
   A. albumin
   B. fibronectin
   C. opsonin
   D. hepcidin
   E. haptoglobin

35. The LEAST confirmatory finding of ABO compatibility in a 2-day-old newborn is
   A. positive direct coombs test on infant’s RBCs
   B. positive indirect antiglobulin test on mother’s serum
   C. spherocytes on infant’s blood smear
   D. immature erythroid precursors (erythroblastosis) on infant’s smear
   E. elevated total serum bilirubin

36. The role of corticosteroid in autoimmune hemolytic anemia is to decrease
   A. antibody production
   B. the size of the spleen
C. the clearance of sensitized RBCs in the spleen
D. vascular stability
E. immune response to offending organisms

37. All the following cause autoimmune hemolytic anemia through mechanical damage of the RBCs **EXCEPT**
   A. Kasabach-Merritt syndrome
   B. disseminated intravascular coagulation
   C. malignant hypertension
   D. Mycoplasma pneumonia
   E. hemolytic uremic syndrome

38. A 7-year-old apparently healthy boy presented to the outpatient clinic with mild pallor and slight yellowish discoloration of sclera since early childhood, family was reluctant to seek medical consultation. On examination, he had palpable spleen in addition to pallor and jaundice. His uncle had a history of repeated blood transfusions during childhood that was relieved with splenectomy. Of the following, the **MOST** likely diagnosis at this stage is
   A. autoimmune hemolytic anemia
   B. hereditary spherocytosis
   C. acute leukemia
   D. immune thrombocytopenia
   E. hemolytic uremic syndrome

39. The best treatment option for hereditary spherocytosis is
   A. splenectomy
   B. corticosteroids
   C. immunoglobulin
   D. cyclosporine
   E. dapsone

40. The best clinical indicator of ongoing hemolysis in G6PD is
   A. pulse rate
   B. blood pressure
   C. degree of pallor
   D. urine color
   E. respiratory rate
41. Decisions about transfusion of a child with sickle cell disease should be made on the following bases **EXCEPT**
   A. age of the patient
   B. clinical condition
   C. hemoglobin level
   D. reticulocyte count
   E. presence of crises

42. The **MOST** common type of vaso-occlusive crises in children with sickle cell disease is
   A. acute chest syndrome
   B. pain crises
   C. priapism
   D. overt stroke
   E. silent stroke

43. Parvovirus B19 infection in sickle cell disease is associated with which type of crises?
   A. splenic sequestration
   B. bone pain
   C. priapism
   D. aplastic
   E. stroke

44. Which of the following agents helps in decreasing the frequency of vaso-occlusive crises in children with sickle cell disease?
   A. cyclosporine
   B. azathioprine
   C. hydroxyurea
   D. tacrolimus
   E. methotrexate

45. Beside pallor, the **MOST** common presentation of sickle cell disease during infancy is
   A. dactyliitis
   B. hemolytic crises
   C. splenic sequestration crises
   D. acute chest syndrome
   E. priapism
46. Which of the following clinical conditions **DOES NOT** signify iron overload in children with β-thalassemia major?
   A. heart failure
   B. liver cirrhosis
   C. bronzing of the skin
   D. growth failure
   E. non-immune diabetes mellitus

47. Which of the following criteria is included in the definition of severe aplastic anemia?
   A. reticulocyte count less than 100.000/ μl
   B. neutrophil count less than 500/mm3
   C. platelets count less than 50.000/mm3
   D. bone marrow cellularity less than 50% of normal
   E. lymphocyte count less than 1000/mm3

48. Acquired factors that predispose to thrombosis include all the following **EXCEPT**
   A. nephrotic syndrome
   B. sepsis
   C. abortion
   D. spherocytosis
   E. vasculitis

49. Which of the following conditions causes profound thrombocytopenia?
   A. disseminated intravascular coagulation
   B. liver failure
   C. vitamin k deficiency
   D. sepsis without shock
   E. hemophilia A

50. In disseminated intravascular coagulation, which of the followings is reduced?
   A. prothrombin time
   B. partial thromboplastin time
   C. D-dimer
   D. fibrinogen
   E. bleeding time

51. In liver failure, one of the following usually shows normal value
   A. platelets count
B. prothrombin time  
C. partial thromboplastin time  
D. D-dimer  
E. fibrinogen  

52. A 6-year-old girl presented with recurrent mucocutaneous bleeding episodes since early childhood that was usually stopped spontaneously or by local measures. Her brother also complained from the same problem, initial CBC was normal. Of the following, the **MOST** likely diagnosis is  
   A. hemophilia A  
   B. hemophilia B  
   C. von-Willebrand disease  
   D. disseminated intravascular coagulation  
   E. immune thrombocytopenia  

53. The treatment of choice for most cases of type 1 von-Willebrand disease is  
   A. desmopressin  
   B. vWF-concentrate  
   C. cryoprecipitate  
   D. platelets transfusion  
   E. fresh frozen plasma  

54. One of the following features **DOES NOT** fit hemophilia A  
   A. normal ristocetin-induced platelets agglutination  
   B. prolonged prothrombin time  
   C. prolonged partial thromboplastin time  
   D. low factor VIII activity  
   E. normal factor IX activity  

55. One of the following features **DOES NOT** fit von-Willebrand disease  
   A. normal prothrombin time  
   B. prolonged partial thromboplastin time  
   C. prolonged bleeding time  
   D. low von-Willebrand factor activity  
   E. impaired platelets aggregation  

56. Considering factor replacement therapy, what level of factor VIII is appropriate for mild bleeding?  
   A. 100%
B. 80%
C. 60%
D. 40%
E. 20%

57. One unit per kilogram body weight of factor VIII increase the plasma level by
   A. 10%
   B. 8%
   C. 6%
   D. 4%
   E. 2%

58. One of the following features is **TRUE** about hemophilia inhibitors
   A. IgM antibodies directed against transfused factor
   B. more common in factor IX hemophiliacs
   C. continuous factor infusions are the treatment of choice for high titer inhibitors
   D. recombinant factor VIIa is the treatment of choice for low titer inhibitors
   E. activated prothrombin complex concentrates may cause myocardial infarction

59. The **BEST** treatment option for factor VIII hemophiliacs with high titer inhibitor is
   A. continuous infusion of factor VIII
   B. immune modulators
   C. activated prothrombin complex concentrate
   D. recombinant factor VIIa
   E. high dose steroids

60. A 2-year-old boy presented with recurrent skin and respiratory tract infections since early infancy, he had abnormally eczematous skin of the upper and lower limbs. His initial tests showed normal liver and renal functions with CBC showing low platelets count. He has similarly affected male cousin.
   Of the following, the **MOST** likely diagnosis is
   A. Glanzmann thrombasthenia
   B. Wiskott-Aldrich syndrome
   C. Bernard-Soulier syndrome
   D. immune thrombocytopenia
   E. thrombotic microangiopathy
61. A 5-year-old boy presented to the emergency department with sudden onset of skin ecchymoses and single attack of epistaxis, the history is negative for any significant event except for preceding upper respiratory tract infection. The examination showed an active healthy looking boy with diffuse skin petechial rash and ecchymoses over both lower and upper limbs, no lymphadenopathy and no organomegaly. The initial CBC is as follows: Hb 12.3 gm/dl, WBC 7.600/cmm, N 45%, L 51%, plt 17.000/cmm, and normal looking film except for rarely seen platelets. The most likely diagnosis is:
   A. immune thrombocytopenia
   B. acute leukemia
   C. aplastic anemia
   D. hemophilia A
   E. von-Willebrand disease

62. All the following are treatment options for immune thrombocytopenia **EXCEPT**
   A. steroids
   B. androgen
   C. anti-D
   D. immunoglobulin
   E. splenectomy

63. Splenectomy in acute immune thrombocytopenia is indicated
   A. after failure of steroids therapy
   B. concomitant with immunoglobulin therapy
   C. in life-threatening bleeding
   D. for histopathological study to exclude malignancy
   E. to reduce the rate of platelet transfusions

64. Which of the following tests measures the step from fibrinogen to fibrin conversion?
   A. prothrombin time
   B. partial thromboplastin time
   C. bleeding time
   D. thrombin time
   E. platelet function analyzer

65. Which of the following conditions affects partial thromboplastin time?
   A. disseminated intravascular coagulation
   B. thrombocytopenia

273
C. thrombasthenia
D. hemangioma
E. factor VII deficiency

66. A non-hematological cause of bleeding includes
   A. hepatic failure
   B. Wiskott-Aldrich syndrome
   C. systemic lupus erythematosus
   D. hemophilia
   E. Ehlers-Danlos syndrome

67. Which of the following factors affects prothrombin time?
   A. Factor VII
   B. Factor VIII
   C. Factor IX
   D. Factor XI
   E. Factor XII

68. The following factors affect the common pathway EXCEPT
   A. Factor I
   B. Factor II
   C. Factor V
   D. Factor X
   E. Factor XI

69. Deficiency of which of the following factors is asymptomatic?
   A. factor VII
   B. factor VIII
   C. factor IX
   D. factor XI
   E. factor XII

70. Which of the following results correctly fits the expected transfusion outcome?
   A. packed red blood cell; rise of Hb by 5-7 gm/dl
   B. platelets concentrate; rise of platelets by 80.000 – 100.000/ml
   C. cryoprecipitate; rise of fibrinogen by 50-100 mg/dl
   D. recombinant factor VIII; rise of factor VIII by 0.5%/unit/kg
   E. recombinant factor IX; rise of factor IX by 2%/unit/kg
71. One of the following factors is not available in cryoprecipitate
   A. von-Willebrand factor
   B. Factor XIII
   C. Factor V
   D. Fibrinogen
   E. Factor VIII

72. A 6-year-old boy presented three hours after receiving whole blood transfusion with fever and chills, his urine color is clear and the mother stated that the transfusion process went smoothly.
   Of the following, the **MOST** likely diagnosis is
   A. acute hemolytic transfusion reaction
   B. delayed hemolytic transfusion reaction
   C. allergic transfusion reaction
   D. bacteremia for the transfused blood
   E. febrile non-hemolytic transfusion reaction

73. A mother called you fifteen minutes after starting blood transfusion of her 3-year-old leukemia boy for a concern of being hot, examination showed alert and vitally-stable boy with a temperature of 39.8°C, his urine color is clear.
   Of the following, the **BEST** treatment option now is
   A. stop the transfusion
   B. give antipyretics
   C. check blood compatibility
   D. administer hydrocortisone and diphenhydramine
   E. check for any electrolyte abnormalities

74. A 6-year-old boy presented three hours after receiving blood transfusion with urticaria, pruritus, maculopapular rash, progressively developed facial edema, and respiratory distress.
   Of the following, the **MOST** likely diagnosis is
   A. acute hemolytic transfusion reaction
   B. delayed hemolytic transfusion reaction
   C. allergic transfusion reaction
   D. bacteremia for the transfused blood
   E. febrile non-hemolytic transfusion reaction

75. Unwise action during management of acute hemolytic reaction during blood transfusion of a 7-year-old boy is
   A. stop transfusion
B. intravenous hydration
C. check compatibility
D. administer hydrocortisone and diphenhydramine
E. alkalinize urine
1. (B). Fever is not an indicator for an underlying hematological problem. Jaundice is an indicator together with the above-mentioned distractors.
2. (A). Begins by the 3rd week of gestation.
3. (D). Hematopoiesis begins by 3 weeks of gestation with erythropoiesis in the yolk sac. By 2 months’ gestation, the primary site of hematopoiesis migrates to the liver. By 5 to 6 months’ gestation, the process shifts from the liver to the bone marrow.
5. (D). Irritability, cyanosis, seizures, jaundice, stroke, and headache.
6. (C). Hemoglobin of 16.0 gm/dl, WBCs of 18,000/cmm, neutrophils of 60% and reticulocytes of 5%.
7. (C). Androgen at onset of puberty maintain hemoglobin value about 1.5 – 2 gm/dl higher in males.
8. (E). Folic acid and B12 deficiency are examples of macrocytic anemias.
9. (B). The urgency of diagnostic and therapeutic intervention, especially the use of packed RBC transfusion, should be dictated by the extent of cardiovascular or functional impairment more than the absolute level of hemoglobin.
10. (E). Helpful in megaloblastic anemia.
11. (B). Suggests anemia of chronic disease.
12. (B). Fanconi anemia. Also, absent kidney, small stature, and microphthalmia.
13. (B).
14. (A).
15. (B).
16. (E). Spherocytosis usually is accompanied by normal growth.
17. (C). A, B, D and E are at high risk individuals.
18. (E). The prevalence of iron deficiency, the most common cause of anemia in the world, is about 9% in toddlers, 9% to 11% in adolescent girls, and less than 1% in teenage boys.
19. (B). Thirty percent of children who are iron deficient develop iron deficiency anemia.
20. (C). Breastfed infants are less likely to have iron deficiency than bottle-fed infants because, although there is less iron in breast milk, this iron is more efficiently absorbed.
21. (C). Central nervous system (CNS) abnormalities (apathy, irritability, poor concentration) have been linked to iron deficiency, presumably resulting from alterations of iron-containing enzymes (monoamine oxidase) and cytochromes.

22. (B). In an otherwise healthy child, a therapeutic trial of iron is the best diagnostic study for iron deficiency as long as the child is re-examined and a response is documented.

23. (A). The response to oral iron includes rapid subjective improvement, especially in neurologic function (within 24 to 48 hours) and reticulocytosis (48 to 72 hours); increase in hemoglobin levels (4 to 30 days); and repletion of iron stores (in 1 to 3 months).

24. (E). The usual therapeutic dose of 4 to 6 mg/day of elemental iron induces an increase in hemoglobin of 0.25 to 0.4 g/dL/day (a 1%/day increase in hematocrit).

25. (C). If the hemoglobin level fails to increase within 2 weeks after institution of iron treatment, careful re-evaluation for ongoing blood loss, development of infection, poor compliance, or other causes of microcytic anemia is required.

26. (D). Bottle-fed infants should receive an iron-containing formula until 12 months of age, and breastfed infants older than 6 months of age should receive an iron supplement. The introduction of iron-enriched solid foods at 6 months of age, followed by a transition to a limited amount of cow’s milk and increased solid foods at 1 year, can help prevent iron deficiency anemia.

27. (C).

28. (D). This is the most important feature, others are wrong choices.

29. (A). If the mean corpuscular volume (MCV) divided by the RBC count is less than 12.5 (Mentzer index), the diagnosis is suggestive of thalassemia trait.

30. (D). Beta thalassemia major (Cooley’s anemia).

31. (B). Low serum iron, low serum ferritin, high iron binding capacity, reduced marrow iron stores, decreased marrow sideroblasts and high free erythrocyte protoporphyrin, normal electrophoresis and high red cell distribution width.

32. (B). Human parvovirus B19 infects erythroid precursors and shuts down erythropoiesis.

33. (C). Transient erythroblastopenia of childhood, a normocytic anemia caused by suppression of RBC synthesis, usually appears after 6 months of age in an otherwise normal infant. The recovery is spontaneous.

34. (D). Inflammation causes an increase in the production of hepcidin, interrupting the process of iron release by macrophages and the absorption of iron from the intestines leading to anemia. Haptoglobin carries free hemoglobin released from erythrocytes.

35. (E). Non-specific.

36. (C). Corticosteroids reduce the clearance of sensitized RBCs in the spleen.
37. **(D).** Cold agglutinins.

38. **(B).** Spherocytosis is an autosomal dominant disorder but new mutations are common.

39. **(A).** Splenectomy corrects the anemia and normalizes the RBC survival in patients with hereditary spherocytosis, but the morphologic abnormalities persist.

40. **(D).** Urine color is the best clinical indicator for the ongoing hemolysis.

41. **(A).** Age of the patient is the least effective factor among the above mentioned.

42. **(B).** Pain crisis is the most common type of vasoocclusive event. The pain usually localizes to the long bones of the arms or legs but may occur in smaller bones of the hands or feet in infancy (dactylitis) or in the abdomen.

43. **(D).** In an aplastic crisis, parvovirus B19 infects RBC precursors in the bone marrow and induces transient RBC aplasia with reticulocytopenia and a rapid worsening of anemia because of the very short life span of sickle RBCs.

44. **(C).** Hydroxyurea, which increases hemoglobin F, decreases the number and severity of vasoocclusive events and frequency of acute chest syndrome in children as early as 1 year of age.

45. **(A).** Hand-foot swelling.

46. **(D).** The anemia itself is severe and leads to growth failure.

47. **(B).** Severe aplastic anemia is defined by an absolute reticulocyte count less than 50,000/μL, absolute neutrophil count less than 500/mm3, platelet count less than 20,000/mm3, and bone marrow cellularity on biopsy specimen less than 25% of normal.

48. **(D).** Indwelling catheters, vasculitis, sepsis, immobilization, nephrotic syndrome, coagulopathy, trauma, infection, surgery, inflammatory bowel disease, oral contraceptive agents, pregnancy, and abortion all predispose to thrombosis.

49. **(A).** Liver failure causes normal or reduced platelets count. The other choices do not reduce the platelet count.

50. **(D).** Fibrinogen and platelets are reduced.

51. **(A).** Sometimes reduced but usually normal.

52. **(C).** Inherited as an autosomal dominant trait and rarely as an autosomal recessive trait.

53. **(A).** Type 1 is partial deficiency of VWF and usually the deficiency is mild to moderate.

54. **(B).** The prothrombin time is normal.

55. **(E).** Platelet aggregation is normal, partial thromboplastin time is prolonged or normal and bleeding time is prolonged or normal.

56. **(D).** 40% for factor VIII is appropriate for mild episodes of bleeding whereas 80%-100% is necessary for life threatening bleeding.
57. (E). 1 U/kg body weight of factor VIII increases plasma level 2%, while 1.5 U/kg of factor IX increases plasma level 1%.
58. (E). IgG type; more common in factor VIII; high inhibitors is best treated with factor VIIa; low inhibitors is best treated with continuous factor infusion. Activated prothrombin complex concentrates, used in the past to treat inhibitor patients, paradoxically increased the risks of thrombosis, resulting in fatal complications, such as myocardial infarction.
59. (D). For high titer inhibitors, it is usually necessary to administer a product that bypasses the inhibitor, preferably recombinant factor VIIa.
60. (B). X-linked disorder characterized by hypogammaglobulinemia, eczema, and thrombocytopenia caused by a molecular defect in a cytoskeletal protein common to lymphocytes and platelets.
61. (A). Childhood ITP is caused by an antibody (IgG or IgM) that binds to the platelet membrane.
62. (B). Androgen is used for Fanconi anemia.
63. (C). In acute ITP, only in cases of life-threatening bleeding.
64. (D).
65. (A). Hemophilia; von Willebrand disease; heparin; DIC; deficient factors XII and XI; lupus anticoagulant.
66. (E). Child abuse, vasculitis, telangiectasia and angiodysplasia are other causes of non-hematological bleeding.
68. (E). Affects the intrinsic pathway only.
69. (E). Factor VII, VIII and IX deficiencies cause significant bleeding, whereas factor XI deficiency causes mild bleeding.
70. (C). Packed red blood cell; rise of Hb by 2-3 gm/dl, platelets concentrate; rise of platelets by 30,000 – 50,000/ml, recombinant factor VIII; rise of factor VIII by 2%/unit/kg, and recombinant factor IX; rise of factor IX by 0.5%/unit/kg.
71. (C). Cryoprecipitate contain Factors VIII and XIII, VWF and fibrinogen.
72. (E). Fever during or within 4 hours after end of transfusion, chills, usually from passively transfused cytokines or recipient reaction to leukocytes. Bacteremia is a less possible cause.
73. (B). Treatment of febrile non-hemolytic transfusion reaction includes antipyretics for symptomatic treatment or pretreatment for future transfusions; leukocyte reduction has decreased incidence of febrile reactions. Blood culture if high suspicion for bacterial infection.
74. (C). Urticaria, pruritus, maculopapular rash, edema, respiratory distress and hypotension during or within 4 hours of transfusion; often because recipient has
pre-formed antibodies against donor antigens, occasionally from passive infusion of antibodies from atopic donor. 75.(D). Used for allergic transfusion reaction.
1. Malignancies and age; the **LEAST** likely association is
   A. neuroblastoma peaks during the first two years of life
   B. leukemia peaks during early childhood
   C. retinoblastoma peaks during late childhood
   D. osteosarcoma peaks during adolescence
   E. Hodgkin disease peaks during late adolescence

2. The **BEST** screening radiographic study for a patient with suspicious cervical lymphadenopathy, fever and weight loss is
   A. chest x-ray
   B. chest MRI
   C. abdominal CT scan
   D. neck x-ray
   E. bone scan

3. Metaiodobenzylguanidine (MIBG) scans are useful in the workup of
   A. retinoblastoma
   B. Wilms’ tumor
   C. acute leukemia
   D. neuroblastoma
   E. CNS tumors

4. Rb gene analysis is useful in the workup of
   A. Wilms’ tumor
   B. neuroblastoma
   C. rhabdomyosarcoma
   D. CNS tumors
   E. retinoblastoma

5. Cutaneous manifestations are helpful in suspecting
   A. osteogenic sarcoma
   B. craniopharyngioma
   C. retinoblastoma
6. Diarrhea is a possible manifestation of which malignancy?
   A. Wilms’ tumor
   B. rhabdomyosarcoma
   C. CNS tumors
   D. melanoma
   E. neuroblastoma

7. Posterior mediastinal mass is found in
   A. neuroblastoma
   B. Wilms’ tumor
   C. germ cell tumor
   D. T- cell lymphoma
   E. Hodgkin lymphoma

8. Anterior mediastinal mass is found in
   A. rhabdomyosarcoma
   B. Wilms’ tumor
   C. CNS tumors
   D. lymphoma
   E. neuroblastoma

9. Leukocoria is a manifestation of
   A. neuroblastoma
   B. retinoblastoma
   C. Wilms’ tumor
   D. CNS tumors
   E. leukemia

10. A 2-year- old girl presented with abdominal mass, periorbital ecchymoses and ataxia.
      Of the following, the **MOST** likely diagnosis is
      A. Wilms’ tumor
      B. neuroblastoma
      C. leukemia
      D. rhabdomyosarcoma
      E. brain tumor
11. A 5-year-old boy presented with abdominal mass, hematuria and hypertension. Of the following, the **MOST** likely diagnosis is
   A. Wilms’ tumor
   B. neuroblastoma
   C. leukemia
   D. rhabdomyosarcoma
   E. brain tumor

12. A 5-year-old girl presented with prolonged fever, progressive pallor, arthralgia, and skin ecchymoses. Of the following, the **MOST** likely diagnosis is
   A. Wilms’ tumor
   B. hepatoblastoma
   C. leukemia
   D. rhabdomyosarcoma
   E. brain tumor

13. A 15-year-old boy presented with a 3-month history of cervical swelling, fever, weight loss, and night sweating. Of the following, the **MOST** likely diagnosis is
   A. Wilms’ tumor
   B. hepatoblastoma
   C. Hodgkin lymphoma
   D. rhabdomyosarcoma
   E. brain tumor

14. A 10-year-old girl with newly diagnosed abdominal lymphoma was admitted to the hematology ward with difficult breathing and poor urine output for the last day, her biochemical renal indices (urea and creatinine) are high and the electrolytes were disturbed (hyperkalemia, hyperuricemia and hyperphosphatemia). Of the following, the **MOST** likely diagnosis is
   A. renal infiltration by lymphoma
   B. hemolytic uremic syndrome
   C. tumor lysis syndrome
   D. leukostasis
   E. superior vena cava syndrome
15. Tumor lysis syndrome is associated with which of the following malignancies?
   A. neuroblastoma
   B. Wilms’ tumor
   C. hepatoblastoma
   D. lymphoma
   E. Ewing sarcoma

16. A 3-year-old girl with newly diagnosed paravertebral neuroblastoma, presented to the pediatric emergency department with rapid onset weakness of both lower limbs following a 3-day history of severe back pain.
   Of the following, the MOST possible diagnosis is
   A. brain infiltration
   B. paraneoplastic manifestation
   C. spinal cord compression
   D. hypokalemia
   E. intracranial bleeding

17. A 5-year-old girl diagnosed recently with lymphoma, presented to the emergency room with distended neck veins, plethora, and swollen head and neck.
   Of the following, the MOST likely cause is
   A. lung metastasis
   B. superior vena cava syndrome
   C. pleural effusion
   D. tracheal compression
   E. shock

18. Steroid could be of MINIMAL help in which of the following oncologic event?
   A. neuroblastoma and spinal cord compression
   B. lymphoma and superior vena cava syndrome
   C. leukemia and hyper leukocytosis
   D. Wilms’ tumor and diffuse lung metastasis
   E. brain tumor and increased intracranial pressure

19. Disseminated intravascular coagulation as a consequence of chemotherapy administration is LEAST likely to get benefit from
   A. fresh frozen plasma
   B. cryoprecipitate
   C. antibiotics
   D. intravenous fluids
20. Radiation therapy is indicated in which of the following oncologic emergencies?
   A. spinal cord compression
   B. graft versus host disease
   C. disseminated intravascular coagulation
   D. tumor lysis syndrome
   E. hyper leukocytosis

21. Biochemical abnormalities in tumor lysis syndrome include
   A. hypokalemia
   B. hyperuricemia
   C. hypercalcemia
   D. hypophosphatemia
   E. hypernatremia

22. Which of the following tumors requires biopsy rather than complete surgical resection?
   A. neuroblastoma
   B. Wilms' tumor
   C. lymphoma
   D. hepatoblastoma
   E. pheochromocytoma

23. Almost all the following pediatric malignancies require chemotherapy **EXCEPT**
   A. high grade hepatoblastoma
   B. low grade neuroblastoma
   C. low grade soft tissue sarcoma
   D. high grade lymphoma
   E. low grade osteogenic sarcoma

24. Neo-adjuvant chemotherapy means
   A. chemotherapy administered after resection of the tumor
   B. chemotherapy administered after recurrence of the primary mass
   C. chemotherapy administered while the primary tumor is still present
   D. new chemotherapeutic agents administered along with the classical therapy
   E. supportive agents used in addition to the original chemotherapy
25. Hemorrhagic cystitis is a complication of which of the following chemotherapeutic agents?
   A. vincristine
   B. actinomycin
   C. 6-mercaptopurine
   D. doxorubicin
   E. cyclophosphamide

26. Minimal myelosuppression is reported with which agent?
   A. vincristine
   B. cyclophosphamide
   C. methotrexate
   D. cisplatin
   E. cytosine arabinoside

27. Which of the following agents can cause significant allergic reaction?
   A. methotrexate
   B. asparginase
   C. 6-mercaptopurine
   D. cytosine arabinoside
   E. cyclophosphamide

28. Pulmonary fibrosis is a side effect of
   A. vinblastine
   B. cyclophosphamide
   C. bleomycin
   D. methotrexate
   E. cytosine arabinoside

29. Of the long-term consequences of cancer therapy, cardiomyopathy may be related to
   A. anthracycline
   B. methotrexate
   C. corticosteroids
   D. etoposide
   E. cyclophosphamide
30. A translocation between chromosomes 9 and 22 (BCR-ABL) plays an important role in the development of
   A. acute myeloid leukemia
   B. chronic myeloid leukemia
   C. chronic lymphoid leukemia
   D. Hodgkin’s lymphoma
   E. non-Hodgkin lymphoma

31. Which of the following genotypes can predispose a child to the development of acute leukemia?
   A. Turner syndrome
   B. Neurofibromatosis type 2
   C. Bloom syndrome
   D. WAGR syndrome
   E. Denys-Drash syndrome

32. Which of the following classes of chemotherapeutics carries a particular risk for the development of future leukemia?
   A. alkylating agents
   B. corticosteroids
   C. mitotic inhibitors
   D. anti-tumor antibiotics
   E. topoisomerase II inhibitors

33. The **MOST** common type of leukemia in childhood is
   A. acute lymphoblastic leukemia
   B. acute myeloid leukemia
   C. chronic lymphoid leukemia
   D. chronic myeloid leukemia
   E. juvenile myelomonocytic leukemia

34. The incidence of pediatric acute lymphoblastic leukemia peaks at the age of
   A. infancy
   B. 2-5 years
   C. 6-9 years
   D. 10-13 years
   E. 14-17 years
35. An 11-year-old boy presented with a 1-month history of fever, pallor, and easy skin bruising. His CBC showed: Hb 5.0 gm/dl, WBC 90.000/cmm, Plt 33.000 with many blast cells in the peripheral film. His chest X-ray showed big mediastinal mass. Physical examination showed pale, febrile, ill looking boy with generalized lymph node enlargement, mild respiratory distress and hepatosplenomegaly.

Of the following, the MOST likely diagnosis is
A. precursor B-cell ALL
B. precursor T-cell ALL
C. acute promyelocytic leukemia
D. acute myelomonocytic leukemia
E. Burkitt’s leukemia

36. A common extramedullary site for acute lymphoblastic leukemia is
A. pancreas
B. kidneys
C. lungs
D. testes
E. vertebrae

37. The MOST common chromosomal translocation in pediatric acute lymphoblastic leukemia is
A. t(12;21)
B. t(9;22)
C. t(4;11)
D. t(1;19)
E. t(10;14)

38. The commonest translocation in infantile acute lymphoblastic leukemia is
A. t(12;21)
B. t(9;22)
C. t(4;11)
D. t(1;19)
E. t(10;14)

39. CNS involvement should be excluded at the time of diagnosis of acute lymphoblastic leukemia by
A. EEG
B. lumbar puncture
C. brain CT
40. A 10-day-old newborn presented with pallor, poor oral intake and abdominal distension, physical examination revealed stigmata of trisomy 21, hepatosplenomegaly, and his peripheral blood film showed low hemoglobin, high white cell count, low platelets and many blasts cells. Of the following, the **MOST** likely diagnosis at this stage is
   
   A. acute lymphoblastic leukemia  
   B. transient myeloproliferative syndrome  
   C. acute myeloid leukemia  
   D. chronic myeloid leukemia  
   E. juvenile myelomonocytic leukemia

41. Prophylaxis with oral trimethoprim-sulfamethoxazole is recommended during treatment of acute lymphoblastic leukemia to prevent
   
   A. Pneumocystis jiroveci pneumonia  
   B. pneumococcal pneumonia  
   C. gram negative sepsis  
   D. anaerobic sepsis  
   E. Mycobacterium tuberculosis infection

42. The major short term complication of chemotherapy treatment of childhood leukemia is
   
   A. cardiac dysfunction  
   B. secondary malignancy  
   C. bone marrow suppression  
   D. infertility  
   E. obesity

43. The **LEAST** effective factor in the prognosis of acute lymphoblastic leukemia is
   
   A. gender  
   B. age  
   C. initial WBC count  
   D. cytogenetics  
   E. response to therapy

44. A boy with acute lymphoblastic leukemia is assigned to be of high risk group if he had got which of the following features?
A. being 6-year-old
B. testicular disease at time of diagnosis
C. t(12;21)
D. initial WBC count of 40,000/cmm
E. good response to induction therapy

45. A boy with acute lymphoblastic leukemia is assigned to be of very high risk group if he had got which of the following features?
   A. being thirteen-year-old
   B. hyperdiploid DNA index
   C. t(12;21)
   D. good response to induction therapy
   E. t(9;22)

46. Which of the following features labels a patient with acute lymphoblastic leukemia as a low risk patient?
   A. being fourteen-year-old
   B. initial WBC count of 72,000/cmm
   C. failure to achieve remission after induction therapy
   D. t(12;21)
   E. CNS disease at time of diagnosis

47. The MOST common site of relapse of acute lymphoblastic leukemia is
   A. testes
   B. kidneys
   C. bone marrow
   D. brain
   E. lungs

48. Which of the following viruses plays a role in the etiology of lymphoma?
   A. Cytomegalovirus
   B. Epstein-Barr virus
   C. Rhinovirus
   D. Rota virus
   E. Herpes virus

49. The MOST common clinical presentation of Hodgkin disease in children is painless lymph node enlargement of which region?
   A. mediastinal
B. cervical
C. axillary
D. inguinal
E. abdominal

50. B symptoms in Hodgkin disease are
   A. fever, drenching sweats and poor appetite
   B. fever, skin rash and poor appetite
   C. skin rash, drenching sweats and significant weight loss
   D. fever, drenching sweats and significant weight loss
   E. skin rash, drenching sweats and bone pain

51. A 6-year-old girl presented with a 3-week history of abdominal distension, fever, and periorbital puffiness. Abdominal sonogram showed multiple intestinal related masses suggestive of lymphoma.
   Of the following, the **BEST** diagnostic procedure at this time is
   A. bone marrow aspiration
   B. urinalysis
   C. fine needle aspiration from the mass
   D. peripheral blood film
   E. CT scan of abdomen

52. The **LEAST** likely presenting feature for a newly diagnosed 3-year-old girl with non-Hodgkin’s lymphoma is
   A. fever
   B. pallor
   C. abdominal distension
   D. shortness of breath
   E. jaundice

53. A 6-year-old girl presented with a 3-month history of cervical swelling, fever, and excessive sweating. In the context of suspecting Hodgkin lymphoma.
   Of the following, the **BEST** diagnostic procedure at this time is
   A. bone marrow biopsy
   B. lymph node biopsy
   C. CT neck
   D. Gallium scan
   E. PET scan
54. The **MOST** common histopathological subtype in childhood Hodgkin disease is
   A. lymphocyte predominant
   B. nodular sclerosis
   C. mixed cellularity
   D. lymphocyte depletion
   E. undifferentiated

55. The **MOST** common solid tumors in children are
   A. lymphomas
   B. sarcomas
   C. hepatoblastomas
   D. renal tumors
   E. CNS tumors

56. A unique symptom of slowly growing CNS tumors is
   A. poor school performance
   B. fever
   C. weight loss
   D. acute paralysis
   E. convulsions

57. A common sign of increased intracranial pressure is
   A. second nerve palsy
   B. fifth nerve palsy
   C. sixth nerve palsy
   D. ninth nerve palsy
   E. tenth nerve palsy

58. If an intracranial lesion is suspected, the examination of choice is
   A. CT scan
   B. CSF exam
   C. MRI
   D. PET scan
   E. MIBG

59. During follow up of a surgically resected brain tumor, the examination of choice to distinguish recurrent tumor from radiation necrosis is
   A. computed tomography
   B. magnetic resonance spectroscopy
C. cerebrospinal fluid analysis  
D. positron emission topography  
E. bone scan

60. Of the long-term adverse effects of brain tumor therapy, the one that carries particular significance to infants is  
A. endocrine sequelae  
B. myelodysplasia  
C. ototoxicity  
D. neurocognitive deficits  
E. renal insufficiency

61. Of the following, the **MOST** common brain tumor is  
A. cerebellar astrocytoma  
B. ependymoma  
C. glioblastoma multiforme  
D. craniopharyngioma  
E. optic nerve glioma

62. After resection of a posterior fossa tumor in a 10-year-old boy, he became mute suddenly and his family reported recent behavioral changes in form of irritability and occasional apathy.  
Of the following, the **MOST** likely diagnosis is  
A. posterior fossa syndrome  
B. middle cerebral artery syndrome  
C. cerebellar mutism syndrome  
D. somnolence syndrome  
E. cerebral hyper perfusion syndrome

63. The **MOST** common malignancy in infancy is  
A. Wilms’ tumor  
B. neuroblastoma  
C. rhabdomyosarcoma  
D. Ewing sarcoma  
E. hepatoblastoma

64. Which of the following malignant tumors has a remarkable broad spectrum clinical prognosis ranging from spontaneous regression to death?  
A. Wilms’ tumor
B. rhabdomyosarcoma  
C. Ewing sarcoma  
D. neuroblastoma  
E. hepatoblastoma

65. The **MOST** common clinical presentation for a 2-year-old girl with neuroblastoma is
   A. abdominal mass  
   B. bone pain  
   C. fever  
   D. diarrhea  
   E. poor appetite

66. The **LEAST** likely presentation of neuroblastoma is
   A. weakness of both lower limbs  
   B. Horner syndrome  
   C. secretory diarrhea  
   D. opsomyoclonus  
   E. vomiting

67. Which organ is rarely involved by metastatic neuroblastoma?
   A. liver  
   B. bone marrow  
   C. lung  
   D. bone  
   E. lymph nodes

68. A 6-month-old infant presented with multiple skin nodules and pallor, his CBC showed reduced blood elements with no abnormal cells. Abdominal sonogram showed small right adrenal mass with diffuse liver hypoechogenic lesions. The bone marrow aspiration was highly suggestive of involvement by neuroblastoma. Of the following, the **MOST** likely stage is
   A. stage 1  
   B. stage 2  
   C. stage 3  
   D. stage 4  
   E. stage 4s
69. Neuroblastoma associated diarrhea is likely to be due to
   A. infectious organism
   B. electrolyte disturbances
   C. mechanical effect
   D. catecholamine release
   E. metabolic derangement

70. A strong mimicker of neuroblastoma is
   A. Wilms’ tumor
   B. lymphoma
   C. rhabdomyosarcoma
   D. hepatoblastoma
   E. retinoblastoma

71. The initial treatment of choice for localized neuroblastoma is
   A. radiotherapy
   B. surgical excision
   C. chemotherapy
   D. watchful waiting
   E. bone marrow transplantation

72. An oncologic emergency in neuroblastoma is
   A. spinal cord compression
   B. opsomyoclonus syndrome
   C. pallor
   D. Horner syndrome
   E. bleeding tendency

73. A favorable prognostic factor in neuroblastoma is
   A. lack of cell differentiation
   B. amplification of the MYCN oncogene
   C. lack of hyperdiploidy
   D. metastatic disease at diagnosis
   E. stage 4S

74. WAGR syndrome implies the following
   A. Wilms tumor, aniridia, genitourinary malformation and mental retardation
   B. Wilms tumor, aniridia, gigantism and renal failure
   C. Wilms tumor, anisochromia, genitourinary malformation and rickets
D. Wilms tumor, anisochromia, gigantism and renal failure
E. Wilms tumor, asplenia, genitourinary malformation and rickets

75. The congenital anomalies that are associated with Wilms tumor are
   A. heterochromia, skeletal dysplasia, and cardiac anomalies
   B. cataract, genu varus, and gastrointestinal anomalies
   C. sporadic aniridia, hemihypertrophy, and genitourinary anomalies
   D. coloboma, syndactyly, and lung anomalies
   E. glaucoma, polydactyly, and brain anomalies

76. The **MOST** common presentation of Wilms’ tumor is
   A. fever
   B. hypertension
   C. hematuria
   D. abdominal mass
   E. abdominal pain

77. Evaluation of the inferior vena cava is crucial in the assessment of Wilms’ tumor to detect
   A. tumor vascularity
   B. chemotherapy choice
   C. tumor extension
   D. lymphatic metastasis
   E. portal hypertension

78. A 2-year-old girl presented with abdominal mass and irritability. The mother reported change in the color of urine to red for the last few days. The examination was notable for a flank mass with raised blood pressure. Of the following, the **MOST** likely diagnosis is
   A. neuroblastoma
   B. Wilms’ tumor
   C. lymphoma
   D. rhabdomyosarcoma
   E. hepatoblastoma

79. The **MOST** common soft tissue sarcoma in children is
   A. fibrosarcoma
   B. synovial sarcoma
   C. myofibroma
D. rhabdomyosarcoma  
E. extra osseous Ewing sarcoma

80. Individuals with hereditary retinoblastoma have an increased risk for  
A. rhabdomyosarcoma  
B. osteosarcoma  
C. Wilms’ tumor  
D. neuroblastoma  
E. acute leukemia

81. The **MOST** common site for Ewing sarcoma is  
A. pelvis  
B. humerus  
C. radius  
D. tibia  
E. metacarpals

82. A common site for osteosarcoma is  
A. distal humerus  
B. distal tibia  
C. distal femur  
D. proximal fibula  
E. proximal ulna

83. A possible differentiating symptom between osteosarcoma and Ewing sarcoma is  
A. pain in osteosarcoma  
B. swelling in Ewing sarcoma  
C. fever in osteosarcoma  
D. sweating in Ewing sarcoma  
E. weight loss in Ewing sarcoma

84. Under light microscopy; small, round, blue cell tumors are the hallmark of  
A. osteosarcoma  
B. neurofibroma  
C. acute leukemia  
D. Ewing sarcoma  
E. histiocytosis
85. Embryonal versus alveolar rhabdomyosarcoma, which of the following site is more in favor of alveolar type?
   A. head
   B. neck
   C. genitourinary
   D. para meningeal
   E. lower limb

86. Definitive diagnosis for sarcomas often requires
   A. CT chest
   B. bone scan
   C. bone marrow biopsy
   D. tissue biopsy
   E. PET scan

87. Common site for metastasis of osteosarcoma is
   A. bone
   B. bone marrow
   C. liver
   D. kidneys
   E. lungs

88. The extent of the primary tumor in osteogenic sarcoma should be delineated carefully before starting chemotherapy and this is best established by
   A. local CT
   B. local MRI
   C. local Doppler study
   D. local sonography
   E. local X-ray

89. Which of the following tumors is likely to be misdiagnosed as osteomyelitis?
   A. osteosarcoma
   B. rhabdomyosarcoma
   C. Ewing sarcoma
   D. fibrosarcoma
   E. neurofibroma
1. (C). Embryonal tumors, such as neuroblastoma and retinoblastoma, peak during the first 2 years of life; acute lymphoblastic leukemia peaks during early childhood (ages 2 to 5 years); Osteosarcoma peaks during adolescence; and Hodgkin disease peaks during late adolescence.

2. (A). A chest x-ray (CXR) (posteroanterior and lateral) is the best screening radiographic study for a patient with suspicious cervical lymphadenopathy, fever, and weight loss.

3. (D).

4. (E).

5. (D). Helpful in leukemia, neuroblastoma and Langerhans cell Histiocytosis.


7. (A). Posterior mediastinal mass is found in neuroblastoma, ganglioneuroblastoma and ganglioneuroma.

8. (D). Anterior mediastinal mass is found in leukemia, lymphoma, and germ cell tumor.

9. (B).

10. (B).

11. (A).


13. (C).

14. (C). A common metabolic emergency is tumor lysis syndrome, which occurs during treatment of leukemia and lymphoma where large amounts of phosphate, potassium, and uric acid are released into the circulation from the lysed cells.

15. (D). Leukemia and lymphoma.

16. (C). Spinal cord compression happens after spinal cord infiltration by primary of metastatic disease like neuroblastoma and medulloblastoma.

17. (B). Distended neck veins, plethora, edema of head and neck, cyanosis, proptosis; Horner syndrome are feature of superior vena cava syndrome that is usually associated with lymphoma.

18. (D). Steroid plays a great role in many oncologic emergencies like tumor lysis syndrome, spinal cord compression, superior vena cava syndrome and shock. Respiratory symptoms in Wilms’ tumor are likely to be of metastatic result and steroid is of negligible help.
19. (E). Treatment of DIC should include administration of blood products (fresh frozen plasma and platelets), antibiotic therapy and fluid resuscitation. Leukapheresis is of help in hyper leukocytosis.

20. (A). Radiation therapy in oncologic emergencies has therapeutic role in superior vena cava syndrome, spinal cord compression and increased intracranial pressure.

21. (B). Biochemical abnormalities in TLS include hyperphosphatemia, hypocalcemia, hyperkalemia and hyperuricemia.

22. (C). Almost all pediatric lymphomas are chemosensitive and do not require surgical resection.

23. (B). Chemotherapy is used in almost all cases; with exceptions including low-stage neuroblastoma, Wilms tumor (particularly in infants), and low-grade central nervous system malignancies.

24. (C). Chemotherapy administered while the primary tumor still present is referred to as neo-adjuvant chemotherapy.


27. (B). Allergic reaction is a significant complication of asparginase.

28. (C). Pulmonary fibrosis and pneumonitis.

29. (A). Doxorubicin and daunorubicin, along with radiation may cause cardiomyopathy as a long-term sequela.

30. (B). In chronic myelogenous leukemia and some cases of acute lymphoblastic leukemia, a translocation between chromosomes 9 and 22 results in a fusion gene incorporating parts of two genes, BCR and ABL.

31. (C). Certain constitutional genotypes can predispose a child to the development of acute leukemia. Patients with Down syndrome, Fanconi anemia, Bloom syndrome, ataxia-telangiectasia, Wiskott-Aldrich syndrome, and neurofibromatosis 1, all have an increased risk of acute leukemia.

32. (E). Environmental factors that may increase the risk of leukemia include ionizing radiation and exposure to certain chemotherapy agents, particularly the topoisomerase II inhibitors.

33. (A). Acute lymphoblastic leukemia accounts for 75%, acute myelogenous leukemia (AML) accounts for 15% to 20%, and CML accounts for less than 5% of cases. Other chronic leukemias, including juvenile myelomonocytic leukemia, chronic myelomonocytic leukemia, and chronic lymphocytic leukemia, are rare in childhood.

34. (B). The incidence of acute lymphoblastic leukemia peaks at 2 to 5 years of age and is higher in boys than in girls.
35.(B). T-cell ALL, in particular, is associated with male predominance as well as an older age of peak incidence. The WBC count is usually high and there is a generalized lymphadenopathy with mediastinal widening.
36.(D). The testes and central nervous system (CNS) are common extramedullary sites for acute lymphoblastic leukemia; neurologic symptoms or a painless enlargement of one or both testes may be seen.
37.(A). The t(12;21) translocation is the most common (approximately 20% of all cases) and is associated with a favorable prognosis.
38.(C). The t(4;11) translocation (and other translocations involving the MLL gene on chromosome 11) often occurs in infants and patients with secondary AML and is generally associated with a poor prognosis.
39.(B). Lumbar puncture should always be performed at the time of diagnosis to evaluate the possibility of CNS involvement.
40.(B). Newborns with trisomy 21 may have a condition known as transient myeloproliferative disorder, which produces elevated WBC counts with peripheral blasts, anemia, and thrombocytopenia. It usually resolves with supportive care only, but these children have a significantly increased risk (30%) of developing true acute leukemia (ALL or AML) within the next few months and years of life.
41.(A). Cell-mediated immunosuppression increases the risk of Pneumocystis jiroveci (carinii) pneumonia. Prophylaxis with oral trimethoprim-sulfamethoxazole or aerosolized pentamidine is recommended.
42.(C). The major short term complication of chemotherapy treatment is bone marrow suppression. Long term sequelae include neurocognitive impairment, short stature, obesity, cardiac dysfunction, infertility, second malignant neoplasms, and psychosocial problems.
43.(A). The prognosis of ALL treatment is based on age, initial WBC count, genetic characteristics, and response to induction therapy.
44.(B). High-risk patients are younger than 1 year of age or 10 years of age and older, have an initial WBC count greater than 50,000/mm³, have CNS or testicular disease at diagnosis, or have unfavorable cytogenetics such as t(4;11).
45.(E). Very high-risk patients have a hypodiploid DNA index, a t(9;22) translocation, or fail to achieve remission after 4 weeks of therapy.
46.(D). In general low-risk patients are 1 to 9 years old with an initial WBC count less than 50,000/mm³ and favorable cytogenetic findings such as t(12;21).
47.(C). Relapse of ALL occurs most commonly in the bone marrow but also may occur in the CNS, testes, or other extramedullary sites.
48.(B). The etiologies of lymphoma are unknown, but evidence in many cases suggests that the Epstein-Barr virus (EBV) plays a causal role.
The most common clinical presentation of Hodgkin disease is painless, firm lymphadenopathy often confined to one or two lymph node areas, usually the supraclavicular and cervical nodes. Mediastinal lymphadenopathy producing cough or shortness of breath is another frequent initial presentation.

The presence of one of three B symptoms has prognostic value: fever (>38°C for 3 consecutive days), drenching night sweats, and unintentional weight loss of 10% or more within 6 months of diagnosis.

The diagnosis of lymphoma is established by tissue biopsy or examination of pleural or peritoneal fluid.

Jaundice is an unusual presenting feature except when the LN of the porta hepatitis is involved causing obstructive effect.

The diagnosis ultimately requires a pathologic confirmation from tissue. The other mentioned investigations are used for staging.

Histopathologic subtypes in childhood Hodgkin disease are similar to those in adults; 10% to 15% have lymphocyte predominant, 50% to 60% have nodular sclerosis, 30% have mixed cellularity, and less than 5% have lymphocyte depletion.

CNS tumors are the most common solid tumors in children and are second to leukemia in overall incidence during childhood.

Irritability, anorexia, poor school performance, and loss of developmental milestones all may be signs of slow-growing CNS tumors.

Inability to abduct the eye as the result of sixth cranial nerve palsy is a common sign of increased intracranial pressure.

If an intracranial lesion is suspected, magnetic resonance imaging is the examination of choice.

During follow-up, magnetic resonance spectroscopy can help distinguish recurrent tumor from radiation necrosis.

Long-term adverse effects include neurocognitive deficits, endocrinologic sequelae, decreased bone growth, ototoxicity, renal insufficiency, cataracts, infertility, and second malignant neoplasms (including myelodysplasia). The neurocognitive deficits can be significant, particularly in infants and young children.

Cerebellar astrocytoma represents 20% of the brain tumors, cerebral astrocytoma represents 15%.

Cerebellar mutism syndrome occurs in 25% of patients after resection of a posterior fossa tumor.

Neuroblastoma is the most common extracranial solid tumor of childhood and the most common malignancy in infancy.

Neuroblastoma is remarkable for its broad spectrum of clinical prognosis ranging from spontaneous regression to rapid progression and metastasis resulting in death.
Children with localized disease are often asymptomatic at diagnosis, whereas children with metastases often appear ill and have systemic complaints, such as fever, weight loss, and pain. The most common presentation is abdominal pain or mass.

Paraspinal tumors may invade through the neural foramina and cause spinal cord compression. Horner syndrome sometimes is seen with neck or apical masses. Several paraneoplastic syndromes, including secretory diarrhea, profuse sweating, and opsomyoclonus (dancing eyes and dancing feet), are associated with neuroblastoma.

Neuroblastoma may metastasize to multiple organs, including the liver, bone, bone marrow, and lymph nodes.

A unique category of neuroblastoma, stage 4S, is defined in infants (<1-year-old) with a small primary tumor and metastasis limited to skin, liver, or bone marrow. It is associated with a favorable outcome.

Paraneoplastic syndrome cause secretory diarrhea due to catecholamine release.

The abdominal presentation of neuroblastoma must be differentiated from Wilms tumor, which also presents as an abdominal flank mass. Ultrasound or CT examination usually differentiates the tumors.

The initial treatment of choice for localized neuroblastoma is complete surgical excision.

Spinal cord compression from neuroblastoma may cause an irreversible neurologic deficit and is an oncologic emergency.

Unfavorable biologic factors include lack of cell differentiation, amplification (>10 copies/ cell) of the MYCN oncogene, lack of hyperdiploidy, and mutations of chromosome 1p, 11q, or 6p22.

Patients with the WAGR (Wilms tumor, aniridia, genitourinary malformation, mental retardation) syndrome have Wilms tumor as a manifestation of the syndrome itself, resulting from a germline deletion at chromosome 11p.

Many congenital anomalies are associated with Wilms tumor, including sporadic aniridia, hemihypertrophy, and genitourinary abnormalities.

Most children with Wilms tumor are brought to medical attention because they have an abdominal mass that is discovered by their parents. Although many children do not have complaints at the time that the mass is first noted, associated symptoms may include abdominal pain, fever, hypertension, and hematuria.

Evaluation of the inferior vena cava is crucial because the tumor may extend from the kidney into the vena cava.

These are the presenting features of Wilms’ tumor.
79. *(D).* Rhabdomyosarcoma, the most common soft tissue sarcoma in children, is derived from mesenchymal cells that are committed to skeletal muscle lineage. Less common soft tissue sarcomas include fibrosarcoma, synovial sarcoma, and extraosseous Ewing sarcoma.

80. *(B).* There is a 500-fold increased risk for osteosarcoma for individuals with hereditary retinoblastoma.

81. *(A).* Ewing sarcoma can occur in almost any bone in the body, the femur and pelvis are the most common sites.

82. *(C).* Osteosarcoma often is located at the epiphysis or metaphysis of anatomic sites that are associated with maximum growth velocity (distal femur, proximal tibia, proximal humerus), but any bone may be involved.

83. *(E).* Systemic symptoms (fever and weight loss) are usually evident in Ewing sarcoma. Both Ewing sarcoma and osteosarcoma can present with pain and swelling.

84. *(D).* Under light microscopy, Ewing sarcoma appear as small, round, blue cell tumors.

85. *(E).* Embryonal histologic variant is most common in younger children with head, neck, and genitourinary primary tumors. Alveolar histologic variant occurs in older patients and is seen most commonly in trunk and extremity tumors.

86. *(D).* Definitive diagnosis of sarcomas often requires tissue biopsy.

87. *(E).* Osteosarcoma tends to metastasize to the lung, most commonly, and rarely to other bones. Metastatic evaluation includes a chest CT scan and a bone scan.

88. *(B).* Local MRI (magnetic resonance imaging) is the best.

89. *(C).* Ewing sarcoma is often accompanied by systemic symptoms like fever and weight loss.
1. The **MOST** likely cause of flank mass in neonate is
   A. renal stones
   B. hydronephrosis
   C. glomerulonephritis
   D. interstitial nephritis
   E. acute tubular necrosis

2. Approximately 75% of filtered bicarbonate is reabsorbed in the
   A. juxtaglomerular apparatus
   B. proximal tubule
   C. loop of Henle
   D. distal tubule
   E. glomeruli

3. Active hydrogen ion secretion, which is responsible for the final acidification of the urine, occurs in the
   A. distal convoluted tubule
   B. collecting ducts
   C. loop of Henle
   D. distal tubule
   E. proximal tubule

4. The glomerular filtration rate (GFR) reaches adult levels by the age of
   A. 1 to 2 years
   B. 3 to 4 years
   C. 5 to 6 years
   D. 7 to 8 years
   E. 9 to 10 years

5. Which of the following is secreted by interstitial cells in the renal medulla in response to low oxygen delivery?
   A. ammonia
   B. calcitriol
   C. erythropoietin
D. renin
E. vasopressin

6. All the following are causes of hematuria **EXCEPT**
   A. acute cortical necrosis
   B. urinary tract malformation
   C. trauma
   D. renal vein thrombosis
   E. hemoglobinuria

   The electrolyte disturbance causing polyuria is
   A. hyperkalemia
   B. hypercalcemia
   C. hypocalcemia
   D. hyperphosphatemia
   E. hypophosphatemia

8. Renal disorders in children may represent intrinsic renal diseases (primary) or derive from systemic conditions (secondary).
   Which of the following is a systemic cause of renal disease in children?
   A. polycystic kidney disease
   B. cystinosis
   C. Alport syndrome
   D. focal segmental glomerulosclerosis
   E. congenital nephrotic syndrome

9. The revised Schwartz formula is the following:
   A. \( \text{GFR} = 0.134 \times \text{Ht} / \text{Cr serum} \)
   B. \( \text{GFR} = 0.413 \times \text{Cr serum} / \text{Ht} \)
   C. \( \text{GFR} = 0.134 \times \text{Cr serum} / \text{Ht} \)
   D. \( \text{GFR} = 0.413 \times \text{Ht} / \text{Cr serum} \)
   E. \( \text{GFR} = 0.413 \times \text{Ht} / \text{Creatinine clearance} \)

10. A medical student asks you about the main factor which affects plasma creatinine level. The correct answer is the
    A. degree of dehydration
    B. nutritional state
C. muscle mass
D. presence of catabolism
E. presence of tissue breakdown

11. A 6-year-old boy, he is a known case of nephrotic syndrome, presented with mild edema around the eyes and lower extremities, but urine albumin show trace result for three successive days.
Of the following, the MOST likely cause of false-negative result of protein is
A. extremely alkaline urine
B. delay in reading the test
C. dilute urine
D. glucose in urine
E. bacteriuria

12. On routine checkup of three years old boy, his general urine examination show: nitrite test positive, you think this is false-positive result.
Of the following the MOST likely cause is
A. frequent voiding
B. low urine bacterial count
C. urinary tract obstruction
D. prolonged contact (uncircumcised boys)
E. infection with bacteria unable to generate nitrite

13. Ultrasound reliably assesses all the following EXCEPT
A. kidney size
B. determines degree of dilation
C. differentiates cortex and medulla
D. renal function
E. resistive index

Of the following, the MOST common type of secondary NS in this girl is
A. membranoproliferative glomerulonephritis
B. membranous nephropathy
C. focal segmental glomerulosclerosis
D. minimal change nephrotic syndrome
E. minimal change with mesengial proliferation
15. Minimal change nephrotic syndrome (MCNS) is the most common histologic form of primary nephrotic syndrome (NS) in children. Children 7 to 16 years old with NS have a chance of having MCNS in a percentage of
   A. 50%
   B. 60%
   C. 70%
   D. 80%
   E. 90%

16. In evaluation of 3-year-old boy with minimal change nephrotic syndrome (MCNS), his GUE show: albumin +++ and RBC +.
   In cases of MCNS, microscopic hematuria may be present in up to
   A. 15%
   B. 25%
   C. 35%
   D. 45%
   E. 55%

17. Transient proteinuria can be seen after all the following EXCEPT
   A. vigorous exercise
   B. fever
   C. dehydration
   D. seizures
   E. adrenergic antagonist therapy

18. An adolescent’s urine examination show normal protein excretion while recumbent but significant proteinuria when upright. This condition is characterized by
   A. tubular in nature
   B. more common in short individuals
   C. more common in obese individuals
   D. associated with progressive renal disease
   E. benign course

19. A 5-month-old boy with Fanconi syndrome, send for urine examination.
   Of the following the MOST likely finding is
   A. low-molecular-weight proteins
   B. hexagonal crystals
   C. red blood cell casts
D. WBC cast
E. high calcium

20. A 10-year-old boy he is a known case of steroid dependent nephrotic syndrome presented to the ER with severe generalized edema.
In addition to restricting salt intake, the BEST diuretic to be use is
A. aldactone
B. manitol
C. acetazolamide
D. frusemide
E. hydrochlorothiasid

All the following agents may minimize the risk of clots EXCEPT
A. warfarin
B. lovenox
C. low-dose aspirin
D. clopidogrel
E. dipyridamole

22. Hemolytic uremic syndrome (HUS) is characterized by the triad of microangiopathic hemolytic anemia, thrombocytopenia, and renal injury and is an important cause of acute kidney injury in children.
Evidence of renal injury in HUS is the presence of
A. anemia
B. decreased haptoglobin
C. leukocytosis
D. pyuria
E. increased LDH

23. Painless gross hematuria may be seen with all the following EXCEPT
A. sickle cell trait
B. renal stone
C. Wilms tumor
D. strenuous exercise
E. sickle cell disease

24. A 3-month-old baby boy had history of upper respiratory tract infection before two days ago presented to ER with repeated fit, rapid breathing, face swelling, and
urine output <0.5 ml/kg/h. Investigations result are: Hb 8 g/dl, WBC 20000/UL, platlate 65000/UL, blood urea 180 mg/dl, serum creatinine 1.6 mg/dl.

Of the following, the **MOST** expected laboratory finding in this case is

A. Increased lactate dehydrogenase (LDH)
B. increased haptoglobin
C. Increased direct bilirubin
D. decreased aspartate aminotransferase (AST)
E. low reticulocyte count

25. Verotoxin (VT)-producing Escherichia coli causes hemorrhagic enterocolitis of variable severity and results in hemolytic uremic syndrome in

A. 5% to 15% of affected children
B. 25% to 35% of affected children
C. 45% to 55% of affected children
D. 65% to 75% of affected children
E. 85% to 95% of affected children

26. Hemolytic uremic syndrome presenting without a prodrome of diarrhea (atypical HUS) may occur at any age. It can be secondary to infection with

A. E.coli O157:H7
B. Shigella
C. Streptococcus pneumonia
D. Mycoplasma pneumonia
E. H. influenza

27. A 9-month-old patient diagnosed as hemolytic uremic syndrome (HUS) without active hemorrhage.

All the following are options for treatment **EXCEPT**

A. volume repletion
B. control of hypertension
C. dialysis
D. platelet transfusion
E. red blood cell transfusion

28. Acute kidney injury (AKI) refers to an abrupt decrease in glomerular filtration rate and tubular function. In **MANY** cases of AKI the cause is

A. prerenal
B. postrenal
C. intrinsic
29. Of the following, the **MOST** common intrinsic cause of acute kidney injury (AKI) in childhood is
   A. acute interstitial nephritis
   B. rhabdomyolysis
   C. glomerulonephritis
   D. tumor lysis syndrome
   E. acute tubular necrosis

30. A 9-month-old patient presented with severe dehydration due to frequent bowel motions and repeated vomiting; investigations show high renal indices. Laboratory and clinical evaluation of this case should include
   A. urine sodium (mEq/L) <15
   B. fractional excretion of sodium (%) >2
   C. urine osmolality ~ 300
   D. increased echogenicity in renal ultrasound
   E. WBCs in urinalysis

31. A 4-month-old baby boy did intravenous pyelography (IVP) because high suspicion of ureteropelvic junction obstruction. Post IVP patient became anuric for 24 hr. Blood gas analysis result was: PH 7.2, HCO3 14, pCO2 65, K+7.1, Ca++ 0.8. To hasten removal of potassium from the body is by use of
   A. bicarbonate
   B. beta-agonists
   C. diuretics
   D. insulin/dextrose
   E. all the above

32. All the following agents use in treatment of hypertension in the setting of acute Kidney Injury (AKI) **EXCEPT**
   A. diuretics
   B. calcium channel blockers
   C. vasodilators
   D. angiotensin-converting enzyme inhibitors
   E. none of the above
33. Complications of chronic kidney disease (CKD) **MOSTLY** do not manifest until at least stage of
   A. 1 CKD
   B. 2 CKD
   C. 3 CKD
   D. 4 CKD
   E. 5 CKD

34. Children with end-stage renal disease (ESRD) are typically treated with either dialysis or renal transplantation when glomerular filtration rate is less than
   A. 15 ml/min/1.73 m$^2$
   B. 25 ml/min/1.73 m$^2$
   C. 35 ml/min/1.73 m$^2$
   D. 45 ml/min/1.73 m$^2$
   E. 55 ml/min/1.73 m$^2$

35. Moderate reduction of glomerular filtration rate 30–59 ml/min/1.73 m$^2$ is equal to chronic kidney disease stage
   A. 1
   B. 2
   C. 3
   D. 4
   E. 5

36. A 5-year-old patient had poor growth secondary to chronic kidney disease. Of the following, the **BEST** management of this problem is by
   A. increased caloric intake
   B. correction of acidosis
   C. correction of renal osteodystrophy
   D. recombinant GH
   E. all the above

37. A 10-year-old patient he is known case of end-stage renal disease (ESRD), his mother consult you about his short stature and bony deformities. Your advise is to start treatment by all the following **EXCEPT**
   A. 1,25-dihydroxy vitamin D supplementation
   B. calcium supplementation
   C. dietary phosphorous restriction
   D. alendronate sodium

313
E. phosphate binders

38. All the following are options in dietary adjustment in patient with chronic kidney disease (CKD) **EXCEPT**
   A. increased caloric intake
   B. protein restriction
   C. iron supplementation
   D. dietary phosphorous restriction
   E. low potassium diet

39. Children with kidney transplant generally do well, but have to take immunosuppressive medications associated with a variety of side effects which include all the following **EXCEPT**
   A. nephrotoxicity
   B. cardiovascular complications
   C. increased risk for certain malignancies
   D. sudden death
   E. infections

40. A 9-year-old male presented to ER with severe headache. His blood pressure measurement in the left hand was 160/90 mmHg. Of the following, the **MOST** likely cause of this reading is
   A. endocrine disorders
   B. renal disorders
   C. neurologic disorders
   D. vascular diseases
   E. metabolic syndromes

41. In children, hypertension (HTN) is defined as blood pressure (BP) greater than the 95\textsuperscript{th} percentile for age, gender, and height on at least three different occasions. Stage 1 hypertension is defined as
   A. >99\textsuperscript{th} blood pressure percentile
   B. 95\textsuperscript{th} to 99\textsuperscript{th} blood pressure percentile
   C. 90\textsuperscript{th} to 95\textsuperscript{th} blood pressure percentile
   D. >99\textsuperscript{th} + 5 mm Hg blood pressure percentile
   E. 95\textsuperscript{th} to 99\textsuperscript{th} + 5 mm Hg blood pressure percentile
42. Infant with history of recurrent urinary tract infections, abdominal US show bilateral hydronephrosis and hydroureter. MCUG show bilateral vesicoureteral reflux (VUR) grade 4.
Siblings of this patient may have a risk to have VUR as
  A. 10% to 20%
  B. 30% to 40%
  C. 50% to 60%
  D. 70% to 80%
  E. 90% to 100%

43. Potter syndrome may have all the following EXCEPT
  A. flat face
  B. clubfeet
  C. cardiac abnormalities
  D. pulmonary hypoplasia
  E. renal agenesis

44. A 5-year-old boy presented to ER with severe scrotal pain.
Of the following, the MOST likely cause of his problem is
  A. retractile testes
  B. torsion of the testis
  C. epididymitis
  D. incarcerated hernia
  E. trauma

45. A 3-week-old neonate with abdominal mass.
Of the following the MOST common cause is
  A. ureteropelvic junction obstruction
  B. obstructive megaureter
  C. posterior urethral valves
  D. multicystic renal dysplasia
  E. polycystic kidney diseases

46. Hepatic fibrosis that leads to portal hypertension is a usual finding in
  A. Poland syndrome
  B. VACTERL association
  C. multicystic renal dysplasia
  D. autosomal recessive polycystic kidney diseases
  E. autosomal dominant polycystic kidney diseases
47. A 3-year-old boy is suffering from recurrent renal stones, 24 hr urine collection shows calcium level >4 mg/kg.
Of the following, the **MOST** appropriate treatment is
A. low calcium intake  
B. normal sodium intake  
C. low oxalate intake  
D. loop diuretics  
E. low potassium diet

48. A 7-year-old boy complaining from bed wetting, no history of bladder control previously.
The safe and effective resolution of this problem is
A. anticholinergic  
B. bed wetting alarm  
C. imipramine  
D. DDAVP  
E. anticholinergic and imipramine

49. Boys with hypospadias have undescended testes in
A. 10% of cases  
B. 20% of cases  
C. 30% of cases  
D. 40% of cases  
E. 50% of cases
QAHTAN ALOBAIDY

1. (B). Differential diagnosis of flank mass in neonate are: multicystic dysplasia, urinary tract obstruction (hydronephrosis), polycystic disease, and tumor.
2. (B).
3. (B).
4. (A).
5. (C). Erythropoietin is secreted by interstitial cells in the renal medulla in response to low oxygen delivery and helps regulate bone marrow red blood cell production.
6. (E). Microscopic hematuria, defined as more than 3 to 5 red blood cells (RBCs) per high-power field on freshly voided and centrifuged urine. Hemoglobinuria (hemolysis) causes cola red-colored urine rather than hematuria.
7. (B). The electrolyte disturbance cause polyuria is hypokalemia and hypercalcemia.
8. (B). Other distracters are causes of primary renal disease in children.
9. (D).
10. (C). While blood urea nitrogen is affected by renal function but is greatly altered by all the following (hydration, nutrition, catabolism, and tissue breakdown).
11. (C). While false-positive result for protein is seen in concentrated urine.
12. (D). The nitrite test may detect bacteriuria if the bacteria reduce nitrate to nitrite and have long contact time with the urine. Gross hematuria or prolonged contact (uncircumcised boys) may result in a false-positive nitrite test. Other distracters are causes of false negative results.
13. (D). While radionuclide studies scan define renal size, scars, and renal function/excretion.
14. (B). Membranous nephropathy represents less than 5% of children with primary nephrotic syndrome. It is seen most commonly in adolescent and children with systemic infections, such as hepatitis B, syphilis, malaria, and toxoplasmosis, or on specific medications (gold, penicillamine).
15. (A). While more than 80% of children less than 7 years of age with NS have MCNS.
16. (B).
17. (E). Transient proteinuria can be seen after adrenergic agonist therapy.
18. (E). Postural (orthostatic) proteinuria is a benign condition defined by normal protein excretion while recumbent but significant proteinuria when upright. It is glomerular in nature, more common in adolescents and tall, thin individuals, and not associated with progressive renal disease.
19. (A). Tubular proteinuria is characterized by low-molecular-weight proteins while glomerular proteinuria is characterized by a combination of large- and small-molecular-weight proteins in the urine, variable levels of proteinuria, and often evidence of glomerular disease (hematuria, red blood cell casts, hypertension and renal insufficiency).

20. (D). Nephrotic syndrome edema is treated by restricting salt intake. Severe edema may require the use of loop diuretics. When these therapies do not alleviate severe edema, cautious parenteral administration of 25% albumin (0.5 to 1.0 g/kg intravenously over 1 to 2 hours) with an intravenous loop diuretic usually results in diuresis.

21. (D). Safety and efficacy have not been established in pediatric patients.

22. (D). The evidence of renal injury in hemolytic uremic syndrome include elevated creatinine and presence of hematuria, proteinuria, pyuria, casts on urinalysis.

23. (B). A renal stone causes painful hematuria.

24. (A). The diagnosis is atypical hemolytic uremic syndrome. Laboratory findings include, increased LDH, decreased haptoglobin, increased indirect bilirubin, increased AST and elevated reticulocyte count.

25. (A).

26. (C). Atypical HUS can be secondary to Streptococcus pneumonia and human immunodeficiency virus.

27. (D). Therapy for HUS is supportive, platelet transfusions should be avoided because they may add to the thrombotic microangiopathy and are indicated only by active hemorrhage or in anticipation of a procedure.

28. (D).

29. (E). Tubular injury encompasses the most common causes of intrinsic AKI in children. Tubular injury may occur from hypoxia-ischemia (acute tubular necrosis), infection (sepsis), nephrotoxic agents (medications, contrast, myoglobin) and inflammation (interstitial nephritis).

30. (A). The diagnosis is prerenal cause of acute kidney injury (AKI). Other distracters are laboratory and clinical evaluation of renal cause of acute Kidney Injury.

31. (C). This case is contrast-induced AKI. A large part of acute Kidney Injury treatment involves management of its complications. To hasten removal of potassium from the body is by (diuretics, sodium-potassium exchange resins, and dialysis). Other distracters are used to shift potassium into cells without removal of potassium from the body.

32. (D). Angiotensin-converting enzyme (ACE) inhibitors are usually avoided in the setting of AKI. While in chronic kidney disease; hypertension and proteinuria can be treated with ACE inhibitors or angiotensin receptor blockers.
33.(C).
34.(A). Children with stage 5 CKD (ESRD) are typically treated with either dialysis or renal transplantation.
35.(C).
*Chronic kidney disease stage 1 = glomerular filtration rate >90 ml/min/1.73 m²
*Chronic kidney disease stage 2 = glomerular filtration rate 60-89 ml/min/1.73 m²
*Chronic kidney disease stage 3 = glomerular filtration rate 30-59 ml/min/1.73 m²
*Chronic kidney disease stage 4 = glomerular filtration rate 15-29 ml/min/1.73 m²
*Chronic kidney disease stage 5 = glomerular filtration rate <15 ml/min/1.73 m²
36.(E).
37.(D). Renal osteodystrophy is usually due to secondary hyperparathyroidism. Alendronate sodium is used in adult osteoporosis.
38.(B). Protein intake is typically not restricted in pediatric CKD.
39.(D).
40.(B). Renal disorders of hypertension as: congenital anomalies (renal dysplasia, obstructive uropathy), glomerulonephritis, and acquired injury (renal scarring, acute tubular necrosis).
41.(E). While stage 2 hypertension >99th + 5 mm Hg blood pressure percentile (%).
42.(B). Vesicoureteral reflux (VUR) is the retrograde flow of urine from the bladder up to the ureter or even up to the kidney. VUR may be familial.
43.(C). Cardiac abnormalities is part of VACTERL association.
44.(B). The major cause of the acute scrotum in boys less than 6 years of age is torsion of the testis.
45.(A).
46.(D). Autosomal recessive PKD is characterized by marked bilateral renal enlargement. Interstitial fibrosis and tubular atrophy progress over time. Kidney failure usually occurs in early childhood. Hepatic fibrosis is present and may lead to portal HTN. Bile duct ectasia and biliary dysgenesis occur. Many affected infants display flank masses, hepatomegaly, pneumothorax, proteinuria, and/or hematuria.
47.(C). Idiopathic familial hypercalciuria (IHC) treated by normal calcium intake with low sodium and low oxalate intake. For some children with IHC, potassium citrate or thiazides are required to minimize stone recurrence.
48.(B). Nocturnal enuresis is the involuntary loss of urine during sleep. Medical therapy with anticholinergics, imipramine, or DDAVP may also be used in selected children.
49.(A).
1. The hypothalamus secretes releasing or inhibiting factors to control the anterior pituitary gland. Of the following, the pituitary hormone that is elevated in a hypothalamic disease is
   A. thyroid-stimulating hormone
   B. follicle-stimulating hormone
   C. luteinizing hormone
   D. prolactin
   E. adrenocorticotropic hormone

2. You are evaluating a newborn baby with cleft palate; his mother asks you about any risk of associated congenital hypopituitarism. Of the following, this baby is MOST likely at risk for
   A. hypothyroidism
   B. hypogonadism
   C. hypoprolactinemia
   D. anti-diuretic hormone deficiency
   E. growth hormone deficiency

3. During a routine clinical visit, a healthy asymptomatic 6-year-old boy is found to have fasting blood glucose of 110 mg/dL; he has normal oral glucose tolerance test, no glycosuria, and HgbA1c of 6.4%. This child is MOST likely considered to have
   A. impaired fasting glucose
   B. impaired glucose tolerance
   C. sporadic hyperglycemia
   D. type 1 diabetes mellitus
   E. normal glucose metabolism

4. In insulin-dependent diabetes mellitus, hyperglycemia results when insulin secretion becomes inadequate to enhance peripheral glucose uptake and to suppress hepatic and renal glucose production. Of the following, the sign of more complete insulin deficiency is
A. postprandial hyperglycemia  
B. fasting hyperglycemia  
C. ketogenesis  
D. glycosuria  
E. metabolic acidosis

5. A 5-year-old boy develops vomiting and abdominal pain with signs of dehydration. He has been diagnosed with type 1 diabetes mellitus since the age of 4 year and he is on insulin therapy since that time. Of the following, the sign that differentiate this condition from gastroenteritis is
   A. polyuria  
   B. polydipsia  
   C. abdominal tenderness  
   D. tachypnea  
   E. altered mental status

6. In DKA, electrolyte abnormalities occurs secondary to hyperglycemia and acidosis. Which of the following electrolytes is artificially measured low in DKA?
   A. potassium  
   B. sodium  
   C. phosphorus  
   D. calcium  
   E. bicarbonate

7. A 9-year-old girl develops polyuria, vomiting, abdominal pain, and deep acidotic breathing during an intercurrent illness. She has been diagnosed with type 1 diabetes mellitus 5 years ago. Lab investigations reveal: blood glucose, 600 mg/dL; blood pH, 7.28; serum bicarbonate concentration, 14 mEq/L; and ketones in urine are positive. Of the following, the MOST serious complication that may occur in this girl is
   A. cardiac arrhythmias  
   B. cerebral edema  
   C. acute tubular necrosis  
   D. pulmonary edema  
   E. bowel ischemia

8. You are going to treat a 7-year-old boy with type 1 diabetes mellitus who develops DKA due to a recent infection. Examination reveals severe dehydration
and deep acidotic breathing. Lab investigations shows: blood glucose, 450 mg/dL; blood pH, 7.2; and serum bicarbonate concentration, 14 mEq/L.

Which of the following should be avoided during the treatment of this girl?

A. lactated Ringer solution  
B. glucose  
C. bicarbonate  
D. fast-acting soluble insulin  
E. potassium phosphate

9. Measurement of hemoglobin A1c (HgbA1c) provides a mean for assessing long-term glycemic control for children with diabetes mellitus. In which of the following conditions this measurement is inaccurate for long-term glycemic control?

A. children with hypoalbuminemia  
B. children with cystic fibrosis  
C. children with malabsorption  
D. children with hemoglobinopathies  
E. children with chronic liver disease

10. A 5-year-old girl has been developed type 1 diabetes mellitus 6 months ago. She is on intensive control using multiple daily injections of insulin. A recent blood testing of HgbA1c is 6.5%.

Of the following, the MOST frequent complication that may be encountered at this age is

A. recurrent hypoglycemia  
B. recurrent hyperglycemia  
C. weight loss  
D. genital candidiasis  
E. recurrent urinary tract infections

11. Type 2 DM can be managed with lifestyle modifications, including nutrition therapy, increased exercise, and oral hypoglycemic agent (metformin); however, insulin therapy is necessary in special circumstances.

Of the following, insulin treatment is required when there is

A. fasting hyperglycemia  
B. postprandial hyperglycemia  
C. hyperkalemia  
D. hypophosphatemia  
E. ketonuria
12. Hypoglycemia may be a manifestation of child abuse or Munchausen syndrome by proxy induced by exogenous administration of insulin (i.e., factitious hyperinsulinemia). Of the following, the **MOST** reliable test that confirms the diagnosis of factitious hyperinsulinemia is
   A. very low concentration of C-peptide
   B. very low level of blood glucose
   C. presence of ketones in urine
   D. presence of ketones in serum
   E. high level of insulin concentration

13. Hypoglycemia may result from absence or deficiency of the counter-regulatory hormones (growth hormone and cortisol). It is sometimes difficult to differentiate it from hyperinsulinism. Which of the following suggests hypoglycemia due to absence of counter-regulatory hormones rather than hyperinsulinism?
   A. presence of serum ketones
   B. very low level of blood glucose
   C. absence of ketones in urine
   D. high level of GH and cortisol
   E. high level of insulin concentration

14. The upper-to-lower segment ratio is one of methods for growth assessment that changes with age. Of the following, this ratio becomes 1:1 at the age of
   A. two year
   B. four year
   C. six year
   D. eight year
   E. ten year

15. A 12-year-old male adolescent is found to have short stature; lab tests and radiological study confirms idiopathic growth hormone (GH) deficiency. You decide to start treatment with biosynthetic recombinant DNA–derived GH. This adolescent is especially at high risk for development of
   A. pseudotumor cerebri
   B. leukemia
   C. carpal tunnel syndrome
   D. diabetes mellitus
E. slipped capital femoral epiphysis

16. You are discussing short stature (SS) with medical students. You mention that congenital growth hormone deficiency is one of common causes of SS. An important statement that should be included with your discussion is
   A. infants usually have a normal birth length and weight at term
   B. growth rate accelerates after birth till the age of 2 to 3 years, then declines
   C. these children have a normal weight-to-height ratio
   D. infants are usually thin and slim
   E. microphallus is usually a late presentation in male gender

17. The staging of pubertal changes differs between both genders. The onset of puberty is marked by pubarche and gonadarche.
Of the following, the first normal event of pubertal maturation in boys is
   A. enlargement of the testis
   B. appearance of the pubic hair
   C. thinning of the scrotum
   D. deepening of the voice
   E. appearance of the axillary hair

18. A 15-year-old boy is brought to clinic by his parents because of delayed puberty. His growth chart reveals a height just below but parallel to the 5th centile. Examination is unremarkable. Bone age is consistent with a 12 year chronological age. Lab investigations including hormonal study are normal.
Of the following, the MOST likely diagnosis is
   A. hypothyroidism
   B. Klinefelter syndrome
   C. hypogonadotropic hypogonadism
   D. craniopharyngioma
   E. constitutional growth delay

19. A 16-year-old boy has delayed puberty; he doesn’t develop secondary sexual characteristics; he has a normal stature according to his chronological age. Examination reveals an upper-to-lower segment ratio of 0.8 (normal ratio is more than 0.9).
Of the following, the MOST important test to confirm diagnosis is
   A. plasma gonadotropins
   B. growth hormone stimulation test
   C. serum prolactin
D. brain MRI
E. bone age

20. A 16-year-old boy has delayed puberty and absence of secondary sexual characteristics; he has decreased sense of smell. Examination reveals shortening of the fourth finger. MRI of the brain shows absence of the olfactory bulbs and sulci; abdominal ultrasonography shows absence of the right kidney. Of the following, the MOST likely diagnosis is
   A. Prader-Willi syndrome
   B. Kallmann syndrome
   C. Laurence-Moon-Bardet-Biedl
   D. Klinefelter syndrome
   E. Noonan syndrome

21. A 9-year-old boy develops secondary sexual characteristics manifested by the parents. Examination reveals an enlarged testis of 2.7 cm and presence of pubic and axillary hair. Hormonal assessment reveals increased level of testosterone and gonadotropins. Of the following, an important NEXT step in the management is
   A. abdominal ultrasonography
   B. bone age
   C. bone scan
   D. brain MRI
   E. testicular ultrasonography

22. Thyroid disorders consist of a variety of diseases that require different treatment according to the cause. Of the following, the thyroid disorder that does not require treatment is
   A. congenital hypothyroidism
   B. endemic cretinism
   C. Hashimoto thyroiditis
   D. hyperthyroidism
   E. congenital thyroxine-binding globulin deficiency

23. A 16-year-old female adolescent develops swelling at the anterior aspect of the neck with difficulty of the swallowing. Examination reveals a firm, nontender diffuse goiter with a pebble-like surface. Her mother has had a thyroid disease in early adulthood. Of the following, the MOST valuable test to confirm the diagnosis is
A. thyroid scan  
B. biopsy of the thyroid gland  
C. ultrasonography of the thyroid gland  
D. thyroid function test  
E. serum antithyroid peroxidase

24. An adolescent female develops fever, palpitation and disorientation. Examination shows exophthalmos, goiter, tachycardia and elevated blood pressure. Lab tests reveal elevated T4 and T3 with low TSH.  
Of the following, the **BEST** initial treatment is
   A. beta blockers  
   B. methimazole  
   C. propylthiouracil  
   D. radioiodine I^{131}  
   E. thyroidectomy

25. A 5-day-old male neonate develops poor feeding, irritability and tachycardia; her mother has been developed hyperthyroidism before pregnancy that was treated by radioiodine. The pregnancy course was unremarkable. Lab investigations reveal markedly elevation of T4 and T3.  
Of the following, the **MOST** likely diagnosis is
   A. congenital hyperthyroidism  
   B. primary hyperthyroidism  
   C. secondary hyperthyroidism  
   D. tertiary hyperthyroidism  
   E. hyperthyroidism due to radioiodine treatment

26. You are examining a 10-month-old male infant during a routine visit; you find that he has wide wrists. He is still breast fed. X-ray of the wrist joint shows signs of rickets.  
Of the following, the **BEST** reflection of vitamin D deficiency is
   A. serum calcium  
   B. serum phosphate  
   C. measurement of parathyroid hormone  
   D. serum concentration of 1,25-hydroxyvitamin D  
   E. serum concentration of 25-hydroxyvitamin D
27. A 12-year-old female adolescent is brought to the emergency room because of irritability, paresthesia, and seizure-like state after a psychological trauma. She is conscious with tachypnea and carpopedal spasm. Of the following, the MOST valuable test is the determination of
   A. total serum calcium
   B. ionized calcium
   C. serum pH
   D. serum albumin
   E. serum magnesium

28. Rickets is defined as decreased bone mineralization in growing children. Hypocalcemia may accompany rickets. Of the following, the condition that causes hypocalcemia rather than rickets is
   A. nutritional vitamin D deficiency
   B. biliary atresia
   C. primary hypoparathyroidism
   D. renal failure
   E. familial hypophosphatemia

29. You are examining a 5-day-old neonate with ambiguous genitalia; there are no palpable gonads in the inguinal or genital area; the blood pressure is normal according to the age. Of the following, the NEXT step in the management of this neonate is
   A. pelvic ultrasonography
   B. chromosomal study
   C. measurement of testosterone level
   D. measurement of the plasma concentration of 17-hydroxyprogesterone
   E. measurement of adrenocorticotropic hormone (ACTH)

30. A 3-week-old neonate develops repeated vomiting and dehydration. Examination reveals ambiguous genitalia with fusion of the anterior portion of the labioscrotal folds. Ultrasonography reveals presence of uterus, ovaries, and fallopian tubes. Of the following, the MOST valuable test to establish the diagnosis is
   A. plasma concentration of 17-hydroxyprogesterone
   B. aldosterone level
   C. testosterone level
   D. plasma concentration of 11-deoxycorticisol
   E. plasma concentration of 18-hydroxycorticosterone
31. A 6-year-old boy develops precocious puberty; he has acne, pubic and axillary hair, and penile enlargement with normal testicular size. His blood pressure is elevated. Lab test reveals hypokalemia, and elevated level of deoxycorticosterone. MRI of the brain is normal. 
Of the following, the MOST likely enzyme deficiency for this boy is
   A. 11-hydroxylase
   B. 17-hydroxylase
   C. 18-hydroxylase
   D. 18-oxidase
   E. 21-hydroxylase

32. A 7-year-old boy is going to undergo orthopedic surgery for right total hip replacement; he has been diagnosed with Addison disease 2 years ago and he is on replacement therapy by hydrocortisone since that time. 
Of the following, the MOST important step in the management of this boy is
   A. frequent monitoring of serum electrolytes and glucose
   B. start fludrocortisone
   C. proceed with surgery
   D. increase the maintenance dose of hydrocortisone
   E. rigorous use of intravenous fluids

33. Cushing syndrome in children has many causes, either exogenous or endogenous. 
Of the following, the LEAST likely endogenous cause in children is
   A. adrenal adenoma
   B. adrenal carcinoma
   C. nodular adrenal hyperplasia
   D. ACTH-secreting pituitary microadenoma
   E. ACTH-secreting tumor

34. An ACTH-secreting pituitary microadenoma resulting in bilateral adrenal hyperplasia is termed Cushing disease; this should be distinguished from Cushing syndrome due to various causes. 
Of the following, the BEST test that helps to differentiate between Cushing disease and Cushing syndrome is
   A. a 24-hour urinary cortisol excretion
   B. low-dose dexamethasone suppression test
   C. high dose dexamethasone suppression test
D. late evening salivary cortisol sampling.
E. serum cortisol level
1. (D). Prolactin is the only pituitary hormone that is suppressed by a hypothalamic factor, dopamine. Hypothalamic deficiency leads to a decrease in most pituitary hormone secretions but may lead to an increase in prolactin secretion.

2. (E). Six percent of cases of cleft palate are associated with GH deficiency.

3. (A). A patient is considered to have impaired fasting glucose, if fasting serum glucose concentration is 100 to 125 mg/dL or impaired glucose tolerance if 2-hour plasma glucose following an OGTT is 140 to 199 mg/dL. Sporadic hyperglycemia can occur in children, usually in the setting of an intercurrent illness.

4. (C). Insulin deficiency usually first causes postprandial hyperglycemia and then fasting hyperglycemia. Ketogenesis is a sign of more complete insulin deficiency.

5. (A). The presence of polyuria, despite a state of clinical dehydration, indicates osmotic diuresis and differentiates patients with DKA from patients with gastroenteritis or other gastrointestinal disorders.

6. (B). The measured serum sodium concentration is artificially low, however, because of hyperglycemia. Hyperlipidemia also contributes to the decrease in measured serum sodium.

7. (B).

8. (C). Bicarbonate therapy should be avoided as it increases risk of the development of cerebral edema.

9. (D). Measurements of HgbA1c are inaccurate in patients with hemoglobinopathies. Glycosylated albumin or fructosamine can be used in these cases.

10. (A).

11. (E). If ketonuria or ketoacidosis occurs, insulin treatment is necessary to first achieve adequate glycemic control but may be discontinued within weeks with continuation of oral medications.

12. (A). This diagnosis should be suspected if extremely high insulin concentrations are detected (>100/µU/mL). C-peptide concentrations are low or undetectable, which confirms that the insulin is from an exogenous source.

13. (A). The absence of serum and urine ketones at the time of hypoglycemia is an important diagnostic feature, distinguishing hyperinsulinism from defect in counter regulatory hormone secretion. In contrast to hyperinsulinism, serum and urine ketones are positive at the time of hypoglycemia and free fatty acids are elevated.
14.(E). Infant has an upper-to-lower ratio of 1.7:1, a 1-year-old has a ratio of 1.4:1, and a 10-year-old has a ratio of 1:1.

15.(E). Treatment with GH carries the risk of an increased incidence of slipped capital femoral epiphysis, especially in rapidly growing adolescents.

16.(A). In Infants with congenital GH deficiency, the growth rate slows after birth, most noticeably after age 2 to 3 years. These children have an elevated weight-to-height ratio, appearing chubby and short. Male neonates may have a microphallus.

17.(C). The normal developmental sequence in girls is thelarche followed closely by pubarche and finally menarche, 2 to 3 years later. In boys the first normal event is scrotal thinning followed by the enlargement of testes and by the appearance of pubic hair.

18.(E). Patients with constitutional delay have delayed onset of pubertal development and significant bone age delay. Usually height gain is below, although fairly parallel to, the normal percentiles on the growth curve. The prepubertal nadir, or deceleration before their pubertal growth spurt, is prolonged or protracted. A family history of delayed puberty in a parent or sibling is reassuring.

19.(A). Patients with hypogonadotropic hypogonadism have normal proportions and growth throughout childhood and in early puberty. When these patients reach adulthood, eunuchoid proportions may ensue because their long bones grow for longer than normal, producing an upper-to-lower ratio below the lower limit of normal of 0.9 and an arm span greater than their height.

20.(B).

21.(D). Compared with girls, boys with precocious puberty have a higher incidence of CNS disorders, such as tumors and hamartomas, precipitating the precocious puberty.

22.(E). It is associated with low serum total T4 concentration, normal TSH, and normal serum free T4. It is a euthyroid condition and does not require treatment with thyroid hormone because it is merely a binding protein abnormality. It is commonly X-linked dominant.

23.(E). Neither biopsy nor thyroid scan is indicated, although the thyroid scan with reduced uptake may differentiate hashitoxicosis from Graves' disease.

24.(A). Thyroid storm is a rare medical emergency. Treatment includes reducing the hyperthermia and administering a beta blocker to control the tachycardia, hypertension, and autonomic hyperfunction symptoms.

25.(A). Cure of hyperthyroidism before pregnancy (surgery or radioiodine treatment) limits or curtails T4 production but not the underlying immune disturbance producing thyroid stimulating immunoglobulins (TSIs); thus the infant still may be affected, at least transiently.
26. (E). The serum concentration of 25-hydroxyvitamin D is a better reflection of vitamin D sufficiency than the measurement of 1,25-hydroxyvitamin D.
27. (B). The fraction of ionized calcium is inversely related to plasma pH; alkalosis (resulting from hyperpnea caused by anxiety or from hyperventilation related to physical exertion) can precipitate hypocalcemia by lowering ionized calcium without changing total serum calcium.
28. (C). Primary hypoparathyroidism causes hypocalcemia but does not cause rickets.
29. (A). The first step toward diagnosis is to determine whether the disorder represents virilization of a genetic female (androgen excess) or underdevelopment of a genetic male (androgen deficiency). Inguinal gonads that are evident on palpation usually are testes and indicate that incomplete development of a male phenotype has occurred. Similarly absence of female internal genitalia (detected by ultrasound) implies that müllerian-inhibiting substance was present and secreted by fetal testes.
30. (A). Statistically most virilized females have CAH; 90% of these females have 21-hydroxylase deficiency. The diagnosis is established by measuring the plasma concentration of 17-hydroxyprogesterone and androstenedione, which typically is hundreds of times above the normal range.
31. (A). In 11-hydroxylase deficiency, virilization occurs with salt retention, hypokalemia, and hypertension as a result of the excessive production of deoxycorticosterone, a potent mineralocorticoid. Late-onset CAH is typically noted years after birth with pseudo precocious puberty rather than ambiguous genitalia.
32. (D). In Addison disease, replacement treatment with 10 to 15 mg/m$^2$/24 hours of hydrocortisone is indicated, with supplementation during stress at three times the maintenance dosage or the use of intramuscular hydrocortisone.
33. (E). ACTH-secreting tumor is an extremely rare.
34. (C).
1. The neurologic symptoms evolution provides clues to the underlying process as symptoms may evolve in a progressive, static, or episodic fashion. The disease that evolves subacutely over days or weeks of symptoms is seen in
   A. seizures
   B. stroke
   C. epidural hemorrhage
   D. brain tumor
   E. hereditary neuropathies

2. The **MOST** common cause of static neurologic abnormalities which are observed early in life is
   A. cerebral palsy
   B. demyelinating diseases
   C. autoimmune diseases
   D. vascular diseases
   E. stroke

3. Neurofibromatosis (NF type 1) has characteristic flat, light brown macules, called
   A. café aulait macule
   B. adenoma sebaceum
   C. ash-leaf spots
   D. shagreen patch
   E. nail fibromas

4. A 9-year-old female presented with acne that was resistance to treatment, examination revealed fibrovascular lesions on the nose and malar regions. She gave history of seizures started in early infancy. This skin lesion is called
   A. café aulait macule
   B. adenoma sebaceum
   C. ash-leaf spots
   D. shagreen patch
   E. fibromas
5. The grasp and rooting reflexes are neonatal reflexes that disappear after six months of life, it may reappear later in life with acquired lesions of the
   A. frontal lobe
   B. occipital lobe
   C. parietal lobe
   D. temporal lobe
   E. cerebellum

6. Stroking lateral aspect of sole from heel up results in dorsiflexion of the great toe and fanning of the remaining toes, this reflex disappear at age of
   A. 6–12 mo
   B. 12–18 mo
   C. 18–24 mo
   D. 24–30 mo
   E. 30–36 mo

7. During clinical examination of an infant you suspended his face down by the chest when he is moved toward a table, the arms extend, this reflex appear at
   A. birth
   B. 2-4 mo
   C. 4-6 mo
   D. 6-8 mo
   E. 8-10 mo

8. A medical student during practical final exam was asked by the examiner to do a stroking of the skin along the edge of vertebrae to a 2-week-old neonate, which produces curvature of the spine with concavity on the side of the stroke, this maneuver is called
   A. asymmetrical tonic neck reflex
   B. parachute reflex
   C. placing reflex
   D. Gallant reflex
   E. Moro reflex

9. The usual normal posture of a full term healthy infant is
   A. “fencing” posture assumption
   B. flexion of lower and upper extremities
   C. lower extremities flexion with extension of upper extremities
   D. lower extremities extension with flexion upper extremities
E. extension of the lower and upper extremities

10. Recoil (the readiness with which an arm or leg springs back to its original position after passive stretching and release) is essentially absent in
   A. very premature infant
   B. premature infant
   C. near term infant
   D. term infant
   E. post term infant

11. The purpose of the neurologic examination is to localize or identify the region within the neuraxis from which the symptoms arise. 
To evaluate the integrity of the brainstem you should examine the
   A. mental status
   B. cranial nerves
   C. motor system
   D. sensory system
   E. gait

12. Language function is receptive (understanding speech or gestures) and expressive (speech and use of gestures).
Anterior, expressive aphasia (characterized by sparse, non-fluent language) is called
   A. Broca aphasia
   B. Wernicke aphasia
   C. global aphasia
   D. alexia
   E. agraphia

13. The cranial nerve evaluation assesses brainstem integrity but depends on the stage of brain maturation and the ability to cooperate.
Smell can be assessed in verbal, cooperative children older than
   A. 1 year
   B. 2 years
   C. 3 years
   D. 4 years
   E. 5 years
14. Visual acuity reaches 20/20 at age of
   A. 2 mo
   B. 4 mo
   C. 6 mo
   D. 8 mo
   E. 10 mo

15. During assessment of the neurological status of a 6-year-old boy, he was asked to smile, blow out his cheeks, blink forcibly, and furrow his forehead. The cranial nerve that is evaluated by doing the above maneuver is
   A. III
   B. IV
   C. VI
   D. VII
   E. VIII

16. In motor examination, the power that is able to move against gravity but not against resistance; is graded as
   A. 1
   B. 2
   C. 3
   D. 4
   E. 5

17. A 4-year-old boy is asked to stand up; he arises from lying on the floor by using his arms to climb up his legs and body, this sign reflects
   A. denervation
   B. proximal weakness
   C. corticospinal tract dysfunction
   D. cerebellar lesions
   E. upper motor neuron disease

18. Muscle bulk represents the volume of muscle tissue. Excessive muscle bulk is seen in
   A. neuropathies
   B. spinal muscular atrophy(SMA)
   C. myotonia congenita
   D. Duchene muscular dystrophy
   E. multiple sclerosis
19. Lower limbs injury in a child after car accident, lead to common peroneal nerve palsy, which results in gait abnormality due to weakness of ankle dorsiflexors. The gait that result from such injury is called
   A. shuffling gait
   B. tandem walk
   C. steppage gait
   D. toe walking
   E. waddling gait

20. During assessment of the neurological status of a 5-year-old boy, it revealed exaggerated knee reflex, with spread to contiguous areas. What is the approximate scale of this reflex?
   A. 0
   B. 1
   C. 2
   D. 3
   E. 4

21. A 3-year-old boy presented with fever, anorexia and headache; examination showed positive meningeal irritation; analysis of cerebrospinal fluid (CSF) showed increased lymphocytes and decreased glucose. He has a history of prolonged use of steroid therapy due to nephrotic syndrome. Of the following, the **MOST** likely diagnosis is
   A. partially treated bacterial meningitis
   B. fungal meningitis
   C. brain abscess
   D. parasitic meningitis
   E. viral meningitis

22. Bloody CSF is seen in
   A. multiple sclerosis
   B. leukodystrophy
   C. venous thrombosis
   D. hypertension
   E. herpetic meningoencephalitis

23. Bilateral diffuse slow wave activity (delta frequency) in an electroencephalogram (EEG) of an infant, is **MOSTLY** seen in
   A. generalized seizure
B. metabolic encephalopathy
C. brain tumor
D. brain abscess
E. stroke

24. Electromyography and nerve conduction velocities (NCVs) assess for abnormalities of the neuromuscular apparatus, including anterior horn cells, peripheral nerves, neuromuscular junctions, and muscles. The amplitude of the NCVs signal is diminished in
   A. axonal neuropathies
   B. Guillain-Barré syndrome
   C. botulism
   D. myasthenia gravis
   E. viral meningitis

25. A healthy 10-year-old child presented to ER with sudden, severe onset headache, the study of choice that should be done is
   A. cranial CT
   B. magnetic resonance imaging (MRI)
   C. skull-x-ray
   D. electroencephalogram (EEG)
   E. lumbar puncture (LP)

26. An adolescent female is complaining from chronic progressive headache that is gradually increasing in frequency and severity, her headache are worse when she lying down and in the first awaking
   Of the following, the MOST likely cause is
   A. anxiety
   B. depression
   C. pseudo tumor cerebri
   D. benign occipital epilepsy
   E. substance abuse

27. A 12-year-old female is complaining from stereotyped attacks of bitemporal, severe, pounding pain that are aggravated by activity and last for 1 to 72 hours.
   Of the following, the MOST likely associated diagnosis is
   A. tension headache
   B. migrane headache
   C. secondary headache
D. brain tumor
E. sinusitis

28. Increased intracranial pressure (ICP) should be suspected if the headache and associated vomiting are worse when lying down.

The characteristic of this headache is include all the following EXCEPT

A. awaken the child from sleep
B. remit on arising
C. exacerbate by coughing
D. exacerbate by bending over
E. fourth cranial nerve palsy

29. The study of choice in any of the following conditions (focal neurological deficits, alteration in level of consciousness, or chronic progressive headache pattern) is

A. cranial CT
B. magnetic resonance imaging (MRI)
C. skull-x-ray
D. electroencephalogram (EEG)
E. lumbar puncture (LP)

30. In a patient with severe migraine that is not responding to the first-line measures (an analgesic, rest, and sleep in a quiet dark room), the MOST useful agent that may be considered is

A. propranolol
B. triptan
C. nortriptyline
D. topiramate
E. cyproheptadine

31. Triptans (available in injectable, nasal spray, oral disintegrating, and tablet form) may alleviate migraine symptoms promptly. It acts as

A. monoamine oxidase inhibitor
B. benzodiazepine
C. parasympathomimetic
D. parasympatholytic
E. serotonin receptor agonist
32. The incidence of childhood epilepsy is
   A. 1% to 2%
   B. 3% to 4%
   C. 5% to 6%
   D. 7% to 8%
   E. 9% to 10%

33. The **MOST** common epilepsy syndrome is
   A. benign Rolandic epilepsy
   B. Juvenile myoclonic epilepsy
   C. Infantile spasms
   D. Lennox-Gastaut syndrome
   E. Landau-Kleffner syndrome

34. The electroencephalographic (EEG) of an 8-year-old girl revealed generalized 3-Hz spike-and-wave activity.
   Of the following, the **MOST** likely diagnosis is
   A. atypical absence seizure
   B. benign childhood epilepsy
   C. acquired epileptic aphasia
   D. absence seizure
   E. West syndrome

35. The teacher of a 6-year-old girl noticed that she had frequent attacks of a brief loss of environmental awareness accompanied by eye fluttering and lip smacking.
   Of the following, the first-choice therapy in this condition is
   A. fosphenytoin
   B. phenobarbital
   C. valproic acid
   D. midazolam
   E. ethosuximide

36. A 5-month-old boy presented with brief contractions of the neck, trunk, and arm muscles, followed by a phase of sustained muscle contraction lasting less than 2 seconds. His EEG pattern consists of chaotic high-voltage slow waves, spikes, and polyspikes.
   Of the following, the first-line treatment in this condition is
   A. adrenocorticotropic hormone
   B. phenobarbital
C. valproic acid  
D. midazolam  
E. ethosuximide  

37. Status epilepticus is a neurologic emergency and is defined as ongoing seizure activity or repetitive seizures without return of consciousness for greater than
   A. 15 minutes  
   B. 30 minutes  
   C. 45 minutes  
   D. 60 minutes  
   E. 90 minutes  

38. A 10-year-old boy presented to ER as a status epilepticus, initial management is done by ensuring an adequate airway, breathing, and circulation; benzodiazepine was started but the seizure does not resolve after two doses. A second-line agent that must be administered is
   A. phenobarbital  
   B. valproic acid  
   C. midazolam  
   D. ethosuximide  
   E. phenytoin  

39. The duration of anticonvulsant treatment varies according to seizure type and epilepsy syndrome. For most children, anticonvulsant medications can be weaned off after 2 years without seizures. Of the following, the condition that usually require treatment for life is
   A. simple partial seizure  
   B. complex partial seizure  
   C. juvenile myoclonic epilepsy  
   D. infantile spasm  
   E. Landau-Kleffner syndrome  

40. The disorder of anterior horn cell causing muscle weakness in infants and children is
   A. Guillain-Barré syndrome  
   B. hereditary motor sensory neuropathy  
   C. tick paralysis  
   D. poliomyelitis  
   E. Bell palsy
41. Neuromuscular disease affects any component of the lower motor neuron unit. The distribution of muscle weakness can point toward specific diseases. The proximal muscle weakness is seen in
   A. polyneuropathy
   B. dermatomyositis
   C. hereditary motor sensory neuropathy 2
   D. myotonic dystrophy
   E. distal myopathy

42. A 6-month-old infant presents with severe hypotonia and generalized weakness, his creatine phosphokinase (CK) is mildly elevated, the electromyelogram (EMG) shows fasciculations and fibrillations, muscle biopsy specimens show grouped atrophy. Of the following, the MOST likely finding in this infant is
   A. absent deep tendon reflexes
   B. absent sensation
   C. bilateral ptosis
   D. abnormal cognition
   E. persistent Moro reflex

43. Secondary (acquired) microcephaly is seen in
   A. Angelman syndrome
   B. Prader-Willi syndrome
   C. agenesis of the corpus callosum
   D. pachygyria
   E. craniosynostosis totalis

44. All the following are caused by autonomic nerves dysfunction in Guillain-Barré syndrome EXCEPT
   A. blood pressure changes
   B. tachycardia
   C. urinary incontinence
   D. stool retention
   E. flaccidity

45. Cranial nerve variant of Guillain-Barré syndrome is called Miller Fisher variant; it is manifested with
   A. ataxia, partial ophalmoplegia and areflexia
   B. flaccidity, sensory loss and ataxia
C. areflexia, urinary retention and flaccidity
D. urinary retention, partial opthalmopegia and sensory loss
E. sensory loss, ataxia and urinary retention

46. The first-line treatment of Guillain-Barré syndrome is
   A. physiotherapy
   B. intravenous immunoglobulin (IVIG)
   C. plasma exchange
   D. immunosuppressive drugs
   E. glucocorticoids

47. Chronic inflammatory demyelinating polyneuropathy (CIDP) is an immune-mediated peripheral neuropathy and can affect patients of all ages. The diagnosis is usually by
   A. clinical examination
   B. genetic testing
   C. nerve conduction velocity
   D. magnetic resonance imaging
   E. cerebrospinal fluid study

48. An adolescent patient develops both proximal and distal weakness of the extremities in an episodic, relapsing-remitting pattern; he also experiences numbness and tingling. Of the following, the **MOST** likely diagnosis is
   A. Charcot-Marie-Tooth Disease
   B. juvenile myasthenia
   C. hypothyroidism
   D. Guillain-Barré Syndrome
   E. chronic inflammatory demyelinating polyneuropathy

49. Pescavus deformity of the feet (high-arched feet) is seen in
   A. congenital myasthenia gravis
   B. Duchenne muscular dystrophy
   C. hereditary motor sensory neuropathy
   D. neuropathy chronic inflammatory demyelinating polyneuropathy
   E. juvenile myasthenia
50. A 9-year-old child has repeated history of variable ptosis, diplopia, ophthalmoplegia, and facial weakness. There is also an associated dysphagia and extremity weakness.

Of the following, the distinguished feature of this disease from other neuromuscular disorders is

A. variable ptosis
B. diplopia
C. ophthalmoplegia
D. facial weakness
E. rapid fatigue of muscles over the day

51. You examined two patients attending neurological clinic, one with myotonic dystrophy and the other with Duchenne muscular dystrophy.

Of the following, the MOST distinguishing feature to differentiate them is by the

A. ptosis
B. cardiac dysfunction
C. leg weakness
D. slow muscle relaxation after contraction
E. facial weakness

52. The CT scan report of your patient who complained from hemiparesis is as follow: unilateral clefts within the cerebral hemispheres that extend from the cortical surface to the ventricular cavity.

Of the following, the MOST likely cause of this description is

A. schizencephaly
B. lissencephaly
C. pachgyria
D. polymicrogyria
E. gray matter heterotopias

53. The triad of complete agenesis of the cerebellar vermis, cystic dilation of the fourth ventricle, and enlarged posterior fossa is seen in

A. agenesis of the corpus callosum
B. Dandy-Walker malformation
C. holoprosencephaly
D. hydranencephaly
E. encephalocele
54. Megalencephaly (enlargement of the brain) is seen in
   A. achondroplasia
   B. tuberous sclerosis
   C. fragile x syndrome
   D. osteopetrosis
   E. hypochondroplasia

55. The **MOST** common genetic cause of neonatal hypotonia is
   A. Down syndrome
   B. congenital laxity of ligaments
   C. Ehlers-Danlos syndrome
   D. benign congenital hypotonia
   E. neonatal adrenal leukodystrophy

56. Infant with benign congenital hypotonia is characterized by
   A. presentation at 3 months old
   B. ability to sit
   C. poor manipulative skills
   D. inability kick arms briskly
   E. good verbal skills

57. A child presents with acute, focal neurologic deficits (hemiparesis) with visual, speech, and sensory deficits. Radiological imaging is consistent with hemorrhagic stroke (HS).
   Of the following, the **MOST** common cause of HS is
   A. congenital heart disease
   B. sickle cell anemia
   C. iron deficiency anemia
   D. vasculitis
   E. arterial dissection

58. Ataxia is the inability to make accurate, smooth and coordinated movements, usually due to a dysfunction of the cerebellar pathways.
   Of the following, the **MOST** common cause of acute ataxia in childhood is
   A. strokes
   B. postictal states
   C. multiple sclerosis
   D. posterior fossa tumors
   E. postinfectious acute cerebellar ataxia
59. A child present to you with opsoclonus-myoclonus-ataxia, the **MOST** likely cause that you should vigorously search for is
   A. neuroblastoma
   B. medulloblastoma
   C. ependymoma
   D. cerebellar astrocytoma
   E. brainstem glioma

60. Which of the following is a cause of dystonia?
   A. kernicterus
   B. drug induced
   C. tardive dyskinesia
   D. pelizaeus-merzbacher disease
   E. hypoxic-ischemic encephalopathy

61. Dystonia is characterized by abnormally sustained muscle contraction, causing twisting motion (torsion spasm) and repetitive movements or abnormal postures. The **MOST** common cause of dystonia among children is
   A. Tourette syndrome
   B. electrolyte disturbances
   C. Wilson disease
   D. cerebral palsy
   E. tardive dyskinesia

62. An anxious mother consults you about her healthy looking son who is complaining from rapid, purposeless, involuntary, stereotyped movements that typically involves the face.
   Of the following, the **MOST** likely diagnosis is
   A. myoclonus
   B. tremor
   C. tics
   D. dystonia
   E. athetosis

63. A state of responsiveness to pain but not to other stimuli is
   A. lethargy
   B. obtundation
   C. stupor
   D. coma
E. consciousness

64. The **MOST** common cause of long-term morbidity in a patient with depressed consciousness is
   A. hypoglycemia
   B. hyperosmolality
   C. hypercapnia
   D. hypoxia
   E. hyperammonemia

65. A 4-year-old boy presented to ER with depressed consciousness and focal neurologic deficits, following head injury. Head CT findings shows multifocal low-density areas with punctate hemorrhages
   Of the following, the **MOST** likely diagnosis is
   A. acute subdural hemorrhage
   B. epidural hemorrhage
   C. brain contusion
   D. subarachnoid hemorrhage
   E. intraparenchymal hemorrhage

66. The onset of clinical and radiologic characteristics of post traumatic intracranial hemorrhage occur over minutes to hours in
   A. acute subdural hemorrhage
   B. epidural hemorrhage
   C. intraparenchymal hemorrhage
   D. subarachnoid hemorrhage
   E. brain contusion

67. Which of the following syndromes of post traumatic intracranial hemorrhage cause neck stuffiness?
   A. acute subdural hemorrhage
   B. epidural hemorrhage
   C. intra parenchymal hemorrhage
   D. subarachnoid hemorrhage
   E. brain contusion

68. Hydrocephalus resulting from overproduction of CSF is seen in
   A. craniopharyngioma
   B. aqueductal stenosis
C. choroid plexus papilloma  
D. basilar impression  
E. Dandy-Walker malformation

69. In pseudotumor cerebri; the **MOST** likely mechanism of increased ICP is  
A. idiopathic  
B. blockage of CSF pathways  
C. toxic cerebral edema  
D. increase in cerebral blood flow  
E. multifocal cerebral infarctions

70. Degenerative diseases may affect gray matter (neuronal degenerative disorders), white matter (leukodystrophies), or both gray and white matter.  
Of the following, the **LEAST** likely applicable diagnostic tool of neurodegenerative disorders is  
A. brain biopsy  
B. magnetic resonance imaging  
C. measurement of lysosomal enzymes  
D. measurement of peroxisomal enzymes  
E. measurement of mitochondrial enzymes

71. The treatable degenerative condition that exhibits signs of cerebellar and basal ganglia dysfunction is  
A. Rett syndrome  
B. Wilson disease  
C. adrenoleukodystrophy  
A. mitochondrial diseases  
B. neuronal ceroidlipofuscinoses

72. A patient with a history of recurrent seizures, on examination you find a hypomelanotic macules, acne-like small red nodules over the nose and cheeks, and elevated, rough plaques of skin over the lumbar area.  
Of the following, the **MOST** likely brain lesions in this case is  
A. bilateral acoustic schwannomas  
B. meningiomas  
C. gliomas  
D. subependymal nodules  
E. schwannomas of spinal nerves
73. Nevus flammeus is due to anectasia of superficial venules and may have a much more extensive and even bilateral distribution. It is seen in the following neurocutaneous disorders
   A. Neurofibromatosis types 1
   B. tuberous sclerosis
   C. Sturge-Weber syndrome
   D. von Hippel–Lindau disease
   E. ataxia-telangiectasia

74. A 4-year-old boy presented with an ipsilateral facial port-wine stain involving the ophthalmic division of the trigeminal nerve and glaucoma. Of the following, the **MOST** common associated neurologic abnormality in this boy is
   A. seizures
   B. hemiparesis
   C. stroke-like episodes
   D. headaches
   E. mental retardation

75. The recurrence risk in subsequent pregnancies for neural tube defects is
   A. 1% to 2%
   B. 3% to 4%
   C. 5% to 6%
   D. 7% to 8%
   E. 9% to 10%
1. (D). Progressive symptoms may evolve suddenly (seizures, stroke); acutely over minutes or hours (epidural hemorrhage); sub acutely over days or weeks (brain tumor); or slowly over years (hereditary neuropathies).

2. (A). Static lesions are often caused by congenital brain abnormalities or prenatal/perinatal brain injury. Episodic disorders are characterized by periods of symptoms, followed by partial or complete recovery (demyelinating, autoimmune, vascular diseases).

3. (A). Because the brain and skin have the same embryonic origin (ectoderm), abnormalities of hair, skin, teeth, and nails are associated with congenital brain disorders (neurocutaneous disorders) such as neurofibromatosis (NF type 1) in which café au lait macules (flat, light brown macules) are characteristic.

4. (B). Adenoma sebaceum, fibrovascular lesions that look like acne on the nose and malar regions, nail fibromas, ash-leaf spots (hypopigmented macules), and Shagreen patches (flesh-colored soft plaques with prominent follicular openings) are commonly seen in older children and adults with tuberous sclerosis.

5. (A). The grasp and rooting reflexes are inhibited by maturation of frontal lobes but may reappear later in life with acquired frontal lobe lesions.

6. (B). Babinski reflex (stroking lateral aspect of sole from heel up results in dorsiflexion of the great toe and fanning of the remaining toes).

7. (E). Parachute reflex (Infant is suspended face down by the chest when infant is moved toward a table, the arms extend as if to protect self).

8. (D). Also called trunk incurvation reflex.

9. (B). Posture is the position that a calm infant naturally assumes when placed supine. Ipsilateral extension of the arm and leg with flexion of opposite extremities in a “fencing” posture is seen in symmetrical tonic neck reflex. An infant at 28 weeks of gestation shows an extended posture. By 32 weeks, there is a slight trend toward increase in tone and flexion of the lower extremities. At 34 weeks, the lower extremities are flexed; the upper extremities are extended. The term infant flexes lower and upper extremities.

10. (A).

11. (B). The mental status examination assesses the cerebral cortex. The cranial nerve examination evaluates the integrity of the brain stem. The motor examination evaluates upper and lower motor neuron function. The sensory examination assesses the peripheral sensory receptors and their central

12.(A). Abnormalities of language resulting from cerebral hemisphere disorders are referred to as aphasias. Anterior, expressive, or Broca aphasia is characterized by sparse, nonfluent language. Posterior, receptive, or Wernicke aphasia is characterized by an inability to understand language, with speech that is fluent but nonsensical. Global aphasia refers to impaired expressive and receptive language. Acquired disorders of reading (alexia) or writing (agraphia).

13.(B). Olfactory nerve is evaluated by aromatic substances (perfumes, vanilla) instead of volatile substances (ammonia), which irritate the nasal mucosa and stimulate the trigeminal nerve.

14.(C). Visual acuity has been estimated to be 20/200 in newborns and 20/20 in 6-month-old infants.

15.(D). Facial muscles are assessed by observing the face during rest, crying, and blinking. Weakness of all unilateral muscles of the face, including the forehead, eye, and mouth, indicates a lesion of the ipsilateral peripheral facial nerve (Bell palsy). Because the upper third of the face receives bilateral cortical innervations, if the weakness affects only the lower face and mouth, a contralateral lesion of upper motor neuron in the brain (tumor, stroke, abscess) must be considered.

16.(C). Power is graded as follows:

- 5 Normal
- 4 Weak but able to provide resistance
- 3 Able to move against gravity but not against resistance
- 2 Unable to move against gravity
- 1 Minimal movement
- 0 Complete paralysis

17.(B). Gower sign.

18.(C). In many lower motor neuron conditions (neuropathies, SMA), muscle bulk is diminished or atrophic. Boys with Duchenne muscular dystrophy have pseudohypertrophy of their calves.

19.(C). By 6 years, a child is able to walk on toes, walk on heels, and tandem walk (heel to toe). Cerebellar dysfunction results in a broad-based, unsteady gait accompanied by difficulty in executing turns. Corticospinal tract dysfunction produces a stiff, scissoring gait and toe walking. Arm swing is decreased, and the affected arm is flexed across the body. Extrapyramidal dysfunction produces a slow, stiff, shuffling gait with dystonic postures. A waddling gait occurs with hip weakness due to lower motor neuron or neuromuscular disorders.

20.(D). Reflexes scale:
- 0 Absent
- 1 Trace
- 2 Normal
- 3 Exaggerated reflex, with spread to contiguous areas (tapping a patellar reflex and observing a bilaterally brisk quadriceps response)
- 4 Clonus (self-limited or sustained)

21. (B). Increased lymphocytes and decreased CSF glucose seen in: mycobacterial infection (tuberculosis), fungal infection, carcinomatous meningitis and sarcoidosis.

22. (E). Bloody CSF seen in subarachnoid hemorrhage, subdural hemorrhage, intraparenchymal hemorrhage, hemorrhagic meningoencephalitis (group B streptococci, HSV), CNS trauma, vascular malformation, coagulopathy and traumatic lumbar puncture. Other distracters are causes of increased CSF protein.

23. (B). Fixed slow wave foci (1 to 3 Hz) delta rhythms suggest an underlying structural abnormality (brain tumor, abscess, and stroke). Bilateral disturbances of brain activity (increased ICP, metabolic encephalopathy) must be suspected when there is diffuse slow wave activity (delta frequency). Spikes, polyspikes, and spike-and-wave abnormalities, either in a localized region (focal) or distributed bihemispherically (generalized), indicate an underlying seizure tendency.

24. (A). Abnormal muscle responses to repetitive nerve stimulation are seen with neuromuscular junction disorders, such as myasthenia gravis and botulism. The amplitude and duration of the muscle compound action potentials are decreased in primary diseases of muscle. NCVs assess the action potential transmission along peripheral nerves. NCVs are slowed in demyelinating neuropathies (Guillain-Barré syndrome). The amplitude of the signal is diminished in axonal neuropathies.

25. (A). When the headache has a sudden, severe onset, emergent computed tomography (CT) can quickly evaluate for intracranial bleeding. If the CT is negative, a lumbar puncture should be performed, with measurement of opening pressure and evaluation for red and white blood cells, protein, glucose, or xanthochromia.

26. (C). Chronic progressive headache: most ominous of the temporal patterns—implies a gradually increasing frequency and severity of headache. The pathologic correlate is increasing ICP. Causes of this pattern include pseudotumor cerebri, brain tumor, hydrocephalus, chronic meningitis, brain abscess, and subdural collections.

27. (B).

28. (E). Papilledema or focal neurological deficits such as sixth cranial nerve palsy may be seen on examination.

29. (B). In these cases, brain magnetic resonance imaging, with and without gadolinium contrast, is the study of choice, providing the highest sensitivity for detecting posterior fossa lesions and other, more subtle abnormalities.
30. (B). Triptans are contraindicated for patients with focal neurological deficits associated with their migraines or signs consistent with basilar migraine (syncope) because of the risk of stroke. Other distracters are preventive agents use to reduce both attack frequency and severity.

31. (E).

32. (A). Seizure is a transient occurrence of signs or symptoms resulting from abnormal excessive or synchronous neuronal activity in the brain while epilepsy is defined as recurrent, unprovoked seizures. Epileptic seizures are generally classified as focal (or partial), arising from one region of the cortex, or generalized seizures, which arise from both hemispheres, simultaneously.

33. (A). Benign childhood epilepsy with centrotemporal spikes, also known as benign Rolandic epilepsy, is among the most common epilepsy syndromes and usually begins between ages 5 and 10 years.

34. (D). Atypical absence; they are associated with slower EEG discharges (2 Hz) and other seizure types. Acquired epileptic aphasia; the EEG is highly epileptiform in sleep, the peak area of abnormality often being in the dominant perisylvian region (language areas). Rolandic epilepsy: centrotemporal spikes. West syndrome is the triad of infantile spasms, developmental regression, and a dramatically abnormal EEG pattern (hypsarrhythmia). Hypsarrhythmia consists of chaotic high-voltage slow waves, spikes, and polyspikes.

35. (E). The diagnosis is absence seizure, a subset of patients also has generalized tonic clonic seizures, for these children, valproic acid is the first choice as it can prevent both absence and convulsive seizures.

36. (A). First-line treatment options for infantile spasms include adrenocorticotropic hormone, high-dose oral corticosteroids, and vigabatrin. For infants with underlying tuberous sclerosis, vigabatrin is considered the treatment of choice.

37. (B).

38. (E). Either IV phenytoin or fosphenytoin is effective, but cardiac monitoring is required to evaluate for arrhythmia. If seizures persist, a loading dose of phenobarbital or valproic acid is appropriate. Alternatively, continuous infusions, such as midazolam or pentobarbital, can be employed as third-line agents. If this approach is ineffective, preparations for general anesthesia are undertaken.


40. (D). Disorder of anterior horn cell causing weakness in infants and children include spinal muscular atrophy and poliomyelitis. Other distracters are result of peripheral nerve lesions.

41. (B). Proximal muscle weakness seen in:
- muscular dystrophy
- Duchenne / Becker
- limb-girdle
- dermatomyositis
- polymyositis
- Kugelberg-Welander disease (spinal muscular atrophy type 2).
- Other distracters are causes of distal limb weakness.

42. (A). Progressive degeneration of anterior horn cells is the key manifestation of spinal muscular atrophy (SMA), a genetic disease that may begin in intrauterine life or any time thereafter. Infants with SMA type 1 present in early infancy with severe hypotonia, generalized weakness, and facial involvement. Infants have normal cognitive, social, and language skills and sensation. Fasciculations (quivering of the lateral aspect of the tongue) are best identified by inspecting the mouth when the child is asleep. Deep tendon reflexes are absent.

43. (E). Other distracters are causes of primary microcephaly.

44. (E). Guillain-Barré syndrome (acute inflammatory demyelinating poly radiculoneuropathy) is a post infectious autoimmune peripheral neuropathy that can occur about 10 days after a respiratory or gastrointestinal infection. Dysfunction of autonomic nerves can lead to blood pressure changes, tachycardia and other arrhythmias, urinary retention or incontinence, or stool retention.

45. (A). Manifests with ataxia, partial ophthalmoplegia, and areflexia.

46. (B). Plasma exchange and immunosuppressive drugs are alternatives when IVIG treatment is unsuccessful or in rapidly progressive disease. Physical, occupational, and speech therapies are mainstays of treatment.

47. (A). The diagnosis is clinical, although EMG or nerve biopsy may confirm the diagnosis.

48. (E).

49. (C). Hereditary Motor Sensory Neuropathy HMSN (also called Charcot-Marie-Tooth Disease [CMT]) is a group of progressive peripheral nerve diseases.

50. (E). Rapid fatigue of muscles distinguishes myasthenia from other neuromuscular disorders, with progressive worsening over the day or with repetitive activity.

51. (D). Myotonia is a disorder of muscle relaxation after contraction. Patients grasp onto an object and have difficulty releasing their grasp, peeling their fingers away slowly.

52. (A).

- Lissencephaly results in a smooth brain without sulcation (agyria).
- Pachygyria, the gyri are few in number and too broad.
- Polymicrogyria, the gyri are too many and too small.
Gray matter heterotopias are abnormal islands within the central white matter of neurons that have never completed the migratory process.

53. (B).

54. (B). Other distracters are causes of macrocrania (increased skull thickness).

55. (A). Prader-Willi syndrome and trisomy 21 are the most common genetic causes of neonatal hypotonia.

56. (E). Infants with benign congenital hypotonia typically exhibit the condition at 6 to 12 months old with delayed gross motor skills. They are unable to sit, creep, or crawl, but have good verbal, social, and manipulative skills. Strength appears normal, and the infants can kick arms and legs briskly and bring their toes to their mouths.

57. (D). Pediatric strokes may be due to ischemia (arterial ischemic stroke, cerebral sinovenous thrombosis) or hemorrhage. The most common causes of HS are vascular malformations, head trauma, and vasculitis. Other distracters are causes of arterial ischemic stroke.

58. (E). The most common causes of acute ataxia in childhood are post infectious acute cerebellar ataxia and drug intoxications.

59. (A). Neuroblastoma located in the adrenal medulla or anywhere along the paraspinal sympathetic chain in the thorax or abdomen is associated with degeneration of Purkinje cells and the development of severe ataxia, dysmetria, irritability, myoclonus, and opsoclonus.

60. (C). Dystonia are caused by
- Inherited primary dystonias
- Acute dystonic reaction
- Tardive dyskinesia
- Cerebral palsy
- Metabolic disorders (Wilson disease).

Other distracters are causes of athetosis.

61. (D).

62. (C). Most tic disorders in children are transient and not intrusive into the child’s life, but may be a source of parental anxiety.

63. (C). Consciousness represents awareness of self and environment (place and time). Lethargic patients have difficulty maintaining an aroused state. Patients who are obtunded have decreased arousal but are responsive to stimuli. Stupor is a state of responsiveness to pain but not to other stimuli. Coma is a state of unresponsive unconsciousness and is caused by dysfunction of the cerebral hemispheres (bilaterally), the brainstem or both simultaneously.

64. (D). Therefore, airway, breathing and circulation are addressed first. Vital signs, including pulse oximetry, must be assessed. A glucose level should also be checked.
immediately because hypoglycemia is a rapidly treatable cause of altered mental status.

65.(C).

66.(B). While acute subdural hemorrhage onset occur over hours.

67.(D). Subarachnoid cause stiff neck, worst headache of life, or late hydrocephalus.

68.(C). Other distracters are causes of hydrocephalus result from obstruction of CSF pathways.

69.(A). Idiopathic intracranial hypertension (pseudotumor cerebri) is a cause of increased ICP with normal brain imaging. Other distracters are mechanisms of increased ICP in bacterial meningitis.

70.(A). Any patient with a degenerative neurologic condition of unknown cause should have leukocytes or skin fibroblasts harvested for measurement of a standard battery of lysosomal, peroxisomal, and mitochondrial enzymes. Neuroimaging usually with brain magnetic resonance imaging (MRI), is also typically warranted.

71.(B). Neurologic symptoms characteristically begin in the early teenage years with dysarthria, dysphasia, drooling, fixed smile, tremor, dystonia, and emotional lability. MRI shows abnormalities of the basal ganglia. Treatment is with a copper-chelating agent, such as oral penicillamine.

72.(D). Tuberous sclerosis brain lesions (cortical tubers, subependymal nodules and hydrocephalus). Other distracters are causes of intracranial and spinal tumors in neurofibromatosis type 2.

73.(C). Sturge-Weber syndrome is sporadic (not inherited) and characterized by abnormal blood vessels (angiomas) of the leptomeninges overlying the cerebral cortex in association with an ipsilateral facial port-wine stain involving the ophthalmic division of the trigeminal nerve (forehead and upper eyelid) and, often, glaucoma. The port-wine stain, also known as nevus flammeus.

74.(A). Seizures are the most common associated neurologic abnormality in patients with Sturge-Weber syndrome, occurring in 75% of patients, and develop because of ischemic injury to the brain.

75.(B). Within a family, an anencephalic birth may be followed by the birth of a child affected with a lumbosacral myelomeningocele. The inheritance of neural tube defects is polygenic.
1. A primary lesion in dermatology is defined as the basic lesion that arise de novo. Of the following, the statement that is correctly defines primary lesion is
   A. macule is an elevated, solid lesion > 1 cm in diameter
   B. patch is an elevated, palpable lesion < 1 cm in diameter
   C. papule is a flat, non palpable lesion > 1 cm in diameter
   D. plaque is an elevated lesion > 1 cm in diameter, with a flat and broad surface
   E. nodule is a fluid-filled (usually clear or straw-colored) epidermal lesion < 1 cm in diameter

2. Secondary skin lesions may be seen in the absence of a primary lesion. Which of the following is considered as secondary rather than primary skin lesion?
   A. pustule
   B. milia
   C. comedo
   D. telangiectasia
   E. scale

3. The face is the most common site of involvement by acne. Which of the following is another common site of acne development?
   A. back
   B. upper extremity
   C. abdomen
   D. lower extremity
   E. groin

4. A medical student is asking you about the type of skin lesions that develops in adolescents with acne. You state that development of acne has different stages. Of the following, the primary event in all acne lesions is development of
   A. papules
   B. pustules
   C. micro-comedones
   D. cysts and nodules
   E. scars
5. The mainstays of treatment of acne are topical keratolytic agents and topical antibiotics.
Of the following, the topical forms that are commonly and effectively used in the treatment of acne is
   A. gels
   B. foams
   C. creams
   D. ointments
   E. lotions

6. Effective treatment of acne focuses on minimizing sebum production, hyperkeratosis, and bacterial proliferation.
Of the following, the MOST effective treatment that can be used as monotherapy for acne is
   A. topical salicylic acid
   B. topical retinoids
   C. topical antimicrobials
   D. topical steroids
   E. oral antibiotics

7. A 14-year-old female adolescent develops deep cystic acne not responding to topical regimen. You decide to give oral antibiotics for such condition in combination with topical regimen.
Of the following, the antibiotic that is LEAST likely effective is
   A. tetracycline
   B. erythromycin
   C. clindamycin
   D. doxycycline
   E. minocycline

8. A 16-year-old girl is having refractory severe nodulocystic acne involving the face with some disfigurement that prevents her from social activities. She tries many anti-acne regimen but with slight improvement.
Of the following, the BEST treatment in such scenario is
   A. topical keratolytic agents
   B. oral antibiotics
   C. oral isotretinoin
   D. combination of topical keratolytic agents and antimicrobials
E. combination of topical keratolytic agents, antimicrobials, and oral antibiotics

9. Atrophic, or hypertrophic scars may develop in patients with acne. Which of the following types of acne has the highest incidence of scar development?
   A. open (blackhead) comedonal acne
   B. closed (whitehead) comedonal acne
   C. papular acne
   D. cystic acne
   E. pustular acne

10. You are meeting parents of a 2-year-old boy with atopic dermatitis; you state that atopic dermatitis is a chronic inflammation that predisposes to the development of allergy. Of the following, this child is **MOST** frequently predisposed to development of
   A. allergic rhinitis
   B. asthma
   C. food allergy
   D. drug allergy
   E. insect bites anaphylaxis

11. You are discussing atopic dermatitis with medical students. You state that atopic dermatitis is a skin disease with a characteristic finding that may superimpose by secondary lesions. Of the following, the secondary lesion that may be encountered in the acute stage of atopic dermatitis is
   A. lichenification
   B. scarring
   C. hypopigmentation
   D. hyperpigmentation
   E. weeping

12. You are evaluating a 7-month-old male infant who recently diagnosed with atopic dermatitis. The location of the lesions you expect to see in this infant is the face and
   A. extensor surfaces of the extremities
   B. flexural surfaces of the antecubital and popliteal fossae
   C. wrists
   D. back
E. upper chest

13. A 3-year-old boy develops fever for the last 3 days. He has been diagnosed with atopic dermatitis since infancy and is well-controlled by topical steroids. Examination reveals presence of pustules and exudative lesions. You suspect secondary bacterial infections.
Of the following, the **MOST** common causative micro-organism is
A. staphylococcus aureus
B. staphylococcus epidermidis
C. streptococcus pyogenes
D. streptococcus pneumoniae
E. mycoplasma pneumoniae

14. Topical corticosteroids are the mainstay of anti-inflammatory therapy for atopic dermatitis. They are classified according to strength and potency from I to VII. Which of the following classes should be avoided in young children?
A. I
B. III
C. V
D. VII
E. no precaution for any class

15. Different potencies and forms of topical corticosteroids are available. Of the following, the form of the topical corticosteroids that is preferred in children with atopic dermatitis is
A. gel
B. lotion
C. solution
D. cream
E. ointment

16. A 5-year-old boy develops extensive atopic dermatitis involving both upper and lower extremities, back, and upper chest. Your decision is to start with potent topical corticosteroids. Of the following, the **MOST** serious complication that may occur with prolonged use of such topical corticosteroids is
A. acne
B. striae
C. skin atrophy
17. A 3-year-old girl has atopic dermatitis involving mainly the face; the mother has a concern regarding skin atrophy that may develop with the use of potent topical corticosteroids. Of the following, the **BEST** treatment for this girl is
   A. systemic corticosteroids
   B. systemic cyclosporine
   C. topical tacrolimus
   D. antihistamines
   E. ultraviolet light therapy

18. Atopic dermatitis is characterized by an increased tendency toward bacterial, viral, and fungal skin infections. Of the following, the **MOST** potentially serious infection in atopic dermatitis is
   A. impetigo
   B. molluscum contagiosum
   C. skin candidiasis
   D. cellulitis
   E. eczema herpeticum

19. You are meeting a pregnant mother who is going to deliver a new baby in near future; she states that she has two children with atopic dermatitis and she wants to prevent the development of this disease for her new baby. Of the following, the **BEST** advice you can offer to this lady is
   A. exclusive breastfeeding for the first 6 months of age
   B. artificial feeding with hydrolyzed casein-based formulas
   C. artificial feeding with soy-based formulas
   D. delaying the introduction of complementary foods beyond 4 to 6 months of age
   E. avoiding peanuts or other foods during pregnancy and while breastfeeding

20. A 4-month-old male infant is brought to your clinic by his mother complaining of napkin rash. The mother has been used different topical agents without any improvement; she has a concern that this rash is caused by fungal infection. Of the following, the character of the rash that’s typically caused by candida albicans napkin dermatitis is
   A. usually extensive and severe
B. affects the perianal region and the buttocks while sparing the protected groin folds  
C. primarily affecting intertriginous areas  
D. not responding to topical nystatin  
E. resolves spontaneously  

21. A 9-month-old male infant has a greasy, scaly, erythematous patches and plaques involving the scalp, face, and posterior auricular folds that have been developed since the age of 1 month. Of the following, the character of this eruption that differentiates it from infantile atopic dermatitis is  
   A. well-demarcated  
   B. thick  
   C. greasy and scaly  
   D. non-pruritic  
   E. involvement of the scalp and face  

22. Seborrheic dermatitis is a common, chronic inflammatory disease that has different clinical presentations at different ages. Of the following, the classic seborrheic dermatitis during adolescence is typically localized to the  
   A. scalp  
   B. axillae  
   C. groin  
   D. antecubital fossa  
   E. umbilicus  

23. A 1-month-old male infant develops a greasy, erythematous scale of the scalp. Examination reveals a thick greasy and waxy yellow-white patch involving the vertex of the scalp. Of the following, the BEST initial treatment for this infant is  
   A. ketoconazole shampooing  
   B. zinc pyrithione  
   C. local application of low-potency steroids  
   D. salicylic acid shampoo  
   E. olive oil messaging followed by shampooing  

24. Pityriasis Rosea is characterized by the appearance of a solitary oval patch followed by multiple red scaly macules. Although it may occur at any age, the peak incidence usually occurs during
A. neonatal period  
B. infancy  
C. early childhood  
D. late childhood  
E. adolescence

25. Psoriasis is a common papulosquamous condition that may occur at all ages and of various subtypes. Of the following, the **MOST** common variety of psoriasis is
   A. psoriasis vulgaris  
   B. guttate psoriasis  
   C. erythrodermic psoriasis  
   D. pustular psoriasis  
   E. inverse psoriasis

26. A 9-year-old girl develops plaque-type psoriasis involving the extensor surfaces of the elbow and knee joints, posterior occipital scalp, and lumbosacral region. Which of the following medications should be avoided for this girl?
   A. topical corticosteroids  
   B. oral corticosteroids  
   C. topical vitamin D analogs  
   D. tar preparations  
   E. phototherapy

27. Dermal melanosis is the most frequently encountered congenital pigmented lesion that gradually disappears during the first few years of life. Of the following, the site that is **MOST** likely to have persistence of these lesions is
   A. lumbosacral area  
   B. buttocks  
   C. flank  
   D. extremities  
   E. face

28. Malignant melanoma may develop in a pre-existing cutaneous melanocytic lesion. Which of the following skin lesion is **MOST** likely to develop malignant melanoma during childhood?
   A. dermal melanosis  
   B. congenital giant melanocytic nevi  
   C. Café au lait macules
D. acquired nevi
E. epidermal nevus

29. Cutaneous hemangiomas may cause functional compromise depending on the location and extent.
Of the following, the area involved by hemangioma that is **MOST** frequently pose a considerable risk and should be monitored carefully is

A. face
B. nasal tip
C. periorbital region
D. buttock
E. groin

30. You are evaluating a 2-month-old male infant with extensive beard hemangioma involving the chin and jaw; he has no hoarseness of voice or stridor.
Of the following, the **NEXT** step in the management of this infant is

A. laryngoscopy
B. ophthalmoscopy
C. MRI of the spinal cord
D. CT scan of the brain
E. echocardiography

31. A 3-year-old girl develops melena and bleeding per rectum. Examination reveals multiple cutaneous hemangiomas involving the face, upper back, and both extremities. Upper and lower endoscopies show multiple hemangiomas involving the stomach, small intestine, and sigmoid colon. Other lab investigations are normal.
Of the following, the **NEXT** step in the management of this girl is

A. oral propranolol
B. laser therapy
C. surgical resection of the largest size hemangioma
D. packed RBCs transfusion
E. α-interferon

32. You are discussing vascular malformations with medical students; you state that port-wine stains are malformations of the superficial capillaries of the skin. An important statement that should be included in your discussion is

A. they do not enlarge after birth
B. the spine is the most commonly involved site
C. they appear as purple or port-wine patches in infancy
D. they do not bleed
E. steroids is the most successful treatment modality

33. You are evaluating a 7-month-old male infant with a pink-red, sharply demarcated macule involving the left eyelid. He doesn`t have any other ocular manifestations. He has normal developmental milestone. There is no history of seizures. Examination including neurological system is unremarkable.
Of the following, the MOST important step in the management of this infant is
   A. frequent MRI of the brain
   B. prophylactic anticonvulsant treatment
   C. cosmetic surgery
   D. frequent ocular pressure monitoring
   E. frequent neurological examination

34. A 5-year-old boy develops a pruritic skin rash involving the lower extremities; he has a history of fever 1 week before that has been resolved spontaneously. Examination reveals many target lesions distributing symmetrically at knees, legs and feet; the outer layer of each lesion is red, the inner layer is purple, while the intermediate layer is white.
Of the following, the MOST likely organism that may cause these target lesions is
   A. herpes simplex virus
   B. mycoplasma pneumoniae
   C. staphylococcus aureus
   D. streptococcus pyogenes
   E. varicella-zoster virus

35. A 7-year-old girl develops red macules that coalesce into large patches distributing over more than half of the body especially the face and trunk; the lesions evolve rapidly into bullae. Her lips are swollen and red with erosion of the oral mucosa. She has also bilateral conjunctival injections and erosions. She has a history of fever, malaise, and upper respiratory symptoms ten days before.
Of the following, the MOST likely diagnosis is
   A. erythema multiforme (EM)
   B. Steven-Johnson syndrome (SJS)
   C. toxic epidermal necrolysis (TEN)
   D. Kawasaki disease
   E. staphylococcal scalded skin syndrome
36. Scabies is caused by the mite Sarcopes scabiei. The clinical presentation is variable depending on the duration of infestation, age, and immune status of the patient.

Of the following, the hallmark of the disease in children is
A. severe itching that is frequently worse than the eruption would suggest
B. diffuse eczematous eruption
C. presence of linear papules
D. presence of pustules in the axillae, umbilicus, and groin
E. nodular eruption in the instep of the foot, and web spaces of the fingers and toes

37. Eruption due to scabies is characterized by papules involving many sites of the body with sparing of the face and scalp. In which of the following age group these sites are usually involved?
A. adults
B. adolescents
C. older children
D. toddlers
E. infants

38. Pediculoses may infest human by direct contact or by indirect spread through contact with personal utensils and clothes. Which of the following manifestations indicate active infestation?
A. lymphadenopathy
B. crusting
C. excoriations
D. brown nits
E. white nits
1. (D). Macule is a flat, non palpable lesion < 1 cm in diameter; patch is similar to macule, but >1 cm in diameter; papule is an elevated, solid lesion < 1 cm in diameter; nodule is similar to papule, but >1 cm in diameter with a rounded surface in contrast to a plaque; vesicle is a fluid-filled (usually clear or straw-colored) epidermal lesion < 1 cm in diameter.

2. (E). Secondary skin lesions are the residue, or result, of the effects of the primary skin lesion. They may be created by scratching or secondary infection. Scale results from abnormal keratinization; it may be fine or sheet-like.

3. (A). The back and upper chest are other common sites of acne development.

4. (C). Rupture of a comedo into adjacent dermis and proliferation of Propionibacterium acnes induce an inflammatory response and development of inflammatory papules and pustules. Larger, skin-colored or red cysts and nodules represent deeper plugging and cystic acne. Increased and persistent inflammation, especially with rupture of a deep cyst, increases the risk of scarring.

5. (A). Gels and solutions are commonly used because acne skin is generally greasier and these agents tend to be drying, but they have the tendency to be irritating and may not be as well tolerated. Creams and lotions are better tolerated but may not be as effective.

6. (B). The topical retinoids (tretinoin, adapalene, tazarotene) are based on the vitamin A molecule. They decrease keratin and sebum production and have some anti-inflammatory and antibacterial activity; thus they can be the most effective when used as monotherapy.

7. (B). Erythromycin efficacy has decreased as P. acnes have become increasingly resistant to this antibiotic. Tetracyclines are the most effective antibiotics because of their significant anti-inflammatory activity.

8. (C). For recalcitrant or severe nodulocystic acne, oral isotretinoin may be instituted. Isotretinoin, an oral analog of vitamin A, normalizes follicular keratinization, reduces sebum production, and decreases 5α-dihydrotosterone formation and androgen receptor–binding capacity. It is the only medication that can permanently alter the course of acne and induce a durable remission. It is teratogenic and must not be used immediately before or during pregnancy.

9. (D). Cystic acne has the highest incidence of scarring because rupture of a deep cyst induces the greatest inflammation, though scarring may be caused by milder pustular or even comedonal acne.
10. (A). Asthma develops in up to half of children with atopic dermatitis, and allergic rhinitis even more frequently. Food allergies are commonly associated with atopic dermatitis.

11. (E). Lichenification is found in older lesions. Formation of fissures is common in both acute and chronic lesions. Temporary hypo- and hyperpigmentation can be seen after lesions resolve. Atopic dermatitis is not usually scarring unless secondary features become severe (e.g., infection or scratching).

12. (A). Characteristic locations vary with the age of the patient. Infantile atopic dermatitis typically affects the face and extensor surfaces of the extremities and is often generalized. Childhood lesions predominate in flexural surfaces (antecubital and popliteal fossae), wrists, ankles, hands, and feet. The adult phase occurs after puberty and manifests in the flexural areas including the neck, as well as predominant involvement on the face, dorsa of the hands, fingers and toes, and the upper arms and back.

13. (A). Secondary bacterial infection, most commonly with staphylococcus aureus or less commonly with streptococcus pyogenes, is frequently present.

14. (A). Class I is the highest potency and class VII is the lowest potency. Class I and II steroids are typically avoided in young children or in areas of thinner skin or enhanced penetration.

15. (E). In general, ointments are preferred because of their increased efficacy, occlusive nature, and tolerability. Creams, lotions, sprays, solutions, and gels can be particularly irritating when applied to atopic skin and should generally be avoided on areas of open skin.

16. (D). Systemic side effects of adrenal suppression or Cushing syndrome can result with application of a potent topical corticosteroid to large surface areas or occluded areas at risk of enhanced penetration.

17. (C). Topical calcineurin inhibitors (topical immune modulators) such as topical tacrolimus and pimecrolimus selectively inhibit T-cell proliferation by inhibiting calcineurin and subsequent interleukin 2 production. They are particularly useful for face or genital lesions as there is no potential for skin atrophy. Antihistamines are useful adjunctive therapy; they have only mild effect on pruritus but can improve the sleeplessness due to scratching during the night. Systemic corticosteroids, cyclosporine, and ultraviolet light therapy can be used for cases of severe disease.

18. (E). Eczema herpeticum (Kaposi’s varicelliform eruption) is one of the potentially serious infectious complications in atopic dermatitis. After herpes simplex virus (HSV) infection, an eruption of multiple, pruritic, vesiculopustular lesions occurs in a disseminated pattern, both within plaques of atopic dermatitis and on normal-appearing skin.
19. (A). There is some evidence suggests that breastfeeding for at least 4 months prevents or delays the occurrence of atopic dermatitis in early childhood.
20. (C).
21. (D). The eruption of seborrheic dermatitis is usually asymptomatic, which helps differentiate it from infantile atopic dermatitis, which is pruritic.
22. (A). The mild form is commonly known as dandruff, a fine, white, dry scaling of the scalp with minor itching.
23. (E). For infants with cradle cap, oil (such as mineral oil or olive oil) may be gently massaged into the scalp and left on for a few minutes before gently brushing out the scale and shampooing. Daily shampooing with ketoconazole, zinc pyrithione, selenium sulfide, or salicylic acid shampoos can treat scalp scale. Seborrheic dermatitis with inflamed lesions responds rapidly to treatment with low-potency steroids two times daily.
24. (E).
25. (A). The most common variety is plaque-type psoriasis (psoriasis vulgaris), which can be localized or generalized. Guttate (numerous small plaques diffusely distributed on the torso), erythrodermic (covering large body surface areas), inverse (moist red patches affecting body folds), and pustular forms may occur.
26. (B). The foundation of therapy is topical corticosteroids. Treatment of psoriasis with oral corticosteroids can induce pustular psoriasis and should be avoided. Topical vitamin D analogs, salicylic acid, and tar preparations are useful adjuvants to topical corticosteroids. Phototherapy with ultraviolet B (UVB) light can be useful as secondary therapy in older children.
27. (E). Although most of these lesions are found in the lumbosacral area (Mongolian spot), they also occur at other sites such as the buttocks, flank, extremities, or, rarely, the face. Aberrant lesions in unusual sites are more likely to persist.
28. (B). Malignant melanoma develops in approximately 2% to 10% of patients with giant congenital melanocytic nevi during childhood. Signs of malignant transformation include irregular pigmentation, rapid growth, bleeding, and a change in configuration or borders.
29. (C). Periorbital hemangiomas pose considerable risk to vision. Amblyopia can result from the hemangioma causing obstruction of the visual axis or pressure on the globe, resulting in astigmatism. If there is any concern, the patient should have urgent evaluation by an ophthalmologist. Treatment may be indicated to prevent blindness.
30. (A). Symptomatic airway hemangiomas develop in more than 50% of infants with extensive facial hemangiomas on the chin and jaw (beard distribution); any infant with a beard hemangioma should be referred for laryngoscopy.
31. (A). Most hemangiomas do not necessitate medical intervention and involute spontaneously; however, if complications arise and treatment is warranted, oral propranolol is the mainstay of therapy.

32. (A). Port-wine stains do not enlarge after birth; any apparent increase in size is caused by growth of the child. Facial lesions are the most common. They are pink-red, sharply demarcated macules and patches in infancy; with time, they darken to a purple or port-wine color. Vascular blebs may form within the lesions and become symptomatic or bleed. The most successful treatment modality in use is the pulsed dye laser, which can result in 80% to 90% improvement. Treatment is more effective if undertaken in infancy.

33. (D). Glaucoma can occur in association with port-wine stains located on the eyelid, even in the absence of Sturge-Weber syndrome, and these patients need lifelong monitoring of ocular pressures.

34. (A). Most erythema multiforme (EM) cases in children are precipitated by herpes simplex virus infection, though the infection may no longer be apparent by the time EM develops. Mycoplasma pneumoniae and other infectious organisms may also trigger EM.

35. (C). SJS is defined as epidermal detachment of less than 10% of the body surface area, whereas SJS/TEN overlap has 10% to 30% and TEN has greater than 30% body surface area involvement. Patients with Kawasaki disease have conjunctival injection and hyperemia of the mucous membranes but necrosis of the lesions does not occur; blistering, erosions, and severe crusting are not observed. The mucosal changes of staphylococcal scalded skin syndrome are minor, and frank erosions are not present. The blistering of the skin is more superficial and favors intertriginous regions.

36. (A). Most children exhibit an eczematous eruption composed of red, excoriated papules and nodules. The classic linear papule or burrow is often difficult to find. Distribution is the most diagnostic finding; the papules are found in the axillae, umbilicus, groin, penis, instep of the foot, and web spaces of the fingers and toes.

37. (E). Infants infested with scabies have diffuse erythema, scaling, and pinpoint papules. Pustules, vesicles, and nodules are much more common in infants and may be more diffusely distributed. The face and scalp usually are spared in adults and older children, but these areas are usually involved in infants.

38. (D). Brown nits located on the proximal hair shaft suggest active infestation. White nits located on the hair shaft 4 cm or farther from the scalp indicate previous infestation. Because nonviable nits can remain stuck in the hair for weeks to months after an infestation has resolved, many children with nits do not have active lice infestation.
1. All the following matching are true EXCEPT
   A. pes cavus = high medial arch of the foot
   B. pes planus = flat foot
   C. subluxation = displacement of bones at a joint
   D. abduction = movement away from midline
   E. adduction = movement toward midline

2. Toe walking is a common complaint in early walkers. A physician should evaluate any child who still toe walks after the age of
   A. 2 years
   B. 3 years
   C. 4 years
   D. 5 years
   E. 6 years

3. Common fracture patterns that should increase the index of suspicion of child abuse include all the following EXCEPT
   A. multiple fractures in different stages of radiographic healing
   B. metaphyseal corner fractures
   C. fractures too severe for the history
   D. toddler’s Fracture
   E. fractures in nonambulatory infants

4. One of the following is a feature of developmental dysplasia of the hip (DDH)
   A. right hip is affected three times as often as the left hip
   B. male infants are at higher risk (9:1)
   C. positive family history in 70% of cases
   D. breech presentation is a risk factor
   E. firstborn child is usually not affected

5. Which of the following conditions should receive a careful examination of the hips to rule out developmental dysplasia of the hip (DDH)?
   A. congenital muscular torticollis
   B. Klippel-Feil syndrome
C. scoliosis
D. spina bifida occulta
E. tethered cord

6. In developmental dysplasia of the hip (DDH), the hip may develop muscular contractures, preventing positive Ortolani tests after the age of
   A. 2 months
   B. 4 months
   C. 6 months
   D. 8 months
   E. 10 months

7. Ultrasound is used for initial evaluation of infants with DDH. To avoid confusion with physiologic laxity, ultrasound should be obtained after the age of
   A. 2 weeks
   B. 4 weeks
   C. 6 weeks
   D. 8 weeks
   E. 10 weeks

8. Bilateral developmental dysplasia of the hip (DDH) presents a diagnostic dilemma.
   Of the following, the **MOST** useful test in this situation is
   A. Ortolani test
   B. Klisic test
   C. Barlow test
   D. Galeazzi sign
   E. decreased hip abduction

9. The **MOST** important and severe complication of DDH is
   A. pressure ulcers
   B. redislocation of the femoral head
   C. subluxation of the femoral head
   D. residual acetabular dysplasia
   E. iatrogenic avascular necrosis of the femoral head

10. A 6-year-old child presented with acute onset of pain in the groin and anterior thigh, he gives history of an upper respiratory tract infection in the preceding 10
days. The patient is afebrile, walks with a painful limp, and has normal white blood cell count, C-reactive protein, and erythrocyte sedimentation rate. Of the following, the MOST likely diagnosis is

A. septic arthritis
B. osteomyelitis
C. myositis
D. transient synovitis
E. Legg-Calve-Perthes disease

11. All the following are risk factors for slipped capital femoral epiphysis (SCFE) EXCEPT

A. obesity
B. trisomy 21
C. hyperthyroidism
D. pituitary tumor
E. growth hormone deficiency

12. The MOST common cause of in-toeing in children 2 years or older is

A. internal femoral torsion
B. internal tibial torsion
C. metatarsus adductus
D. talipes equinovarus (clubfoot)
E. developmental dysplasia

13. The MOST common cause of in-toeing in a child younger than 2 years old is

A. internal femoral torsion
B. internal tibial torsion
C. metatarsus adductus
D. talipes equinovarus (clubfoot)
E. developmental dysplasia

14. It is important to inform families of children with in-toeing due to femoral anteversion and internal tibial torsion that it can take until

A. 3 to 4 years of age for correction
B. 5 to 6 years of age for correction
C. 7 to 8 years of age for correction
D. 9 to 10 years of age for correction
E. 11 to 12 years of age for correction
15. The **MOST** common cause of out-toeing is
   A. external femoral torsion
   B. external tibial torsion
   C. calcaneovalgus feet
   D. hypermobile pes planus (flatfoot)
   E. slipped capital femoral epiphysis

16. Infants are born with maximum genu varum. The lower extremity straightens out around the age of
   A. 12 months
   B. 18 months
   C. 24 months
   D. 30 months
   E. 36 months

17. A 20-month-old child presented with symmetrical genu varum. His height is on the 10\textsuperscript{th} percentile, weight on 25\textsuperscript{th} percentile with no dysmorphic features. His calcium, phosphorus, alkaline phosphatase were normal for his age with no abnormal radiological findings.
Of the following, the **MOST** likely diagnosis is
   A. physiologic bowlegs
   B. Blount disease
   C. physiologic knock-knees
   D. skeletal dysplasia
   E. active rickets

18. Leg-length discrepancy (LLD) is common and may be due to differences in the femur, tibia, or both bones. The **MOST** accurate method with reduced-radiation to measure (LLD) is
   A. clinical measurements using bony landmarks (anterior superior iliac spine to medial malleolus)
   B. teloradiograph
   C. orthoradiograph
   D. computed tomography (CT) scanogram
   E. EOS/slot scanning
19. Treatment of Leg-length discrepancy (LLD) is usually required if it is greater than
   A. 2 cm
   B. 3 cm
   C. 4 cm
   D. 5 cm
   E. 6 cm

20. A 13-year-old boy presented with pain, tenderness, and local swelling over the tibial tubercle mostly during and after activity for the last 3 months. Of the following, the MOST likely cause is
   A. osteochondritis dissecans
   B. Osgood-Schlatter disease
   C. patellofemoral pain syndrome
   D. idiopathic anterior knee pain
   E. recurrent patellar subluxation

21. A 16-year-old female athlete presented with anterior knee pain that worsens with activity, going up and down stairs and soreness after sitting in one position for an extended time with no associated swelling. The patient also complains of a grinding sensation under the kneecap. Of the following, the MOST likely cause is
   A. osteochondritis dissecans
   B. Osgood-Schlatter disease
   C. patellofemoral pain syndrome
   D. idiopathic anterior knee pain
   E. recurrent patellar subluxation

22. In infants, radiographs and advanced imaging are rarely necessary for assessment because their tarsals have incomplete ossification. The navicular ossifies in boys at the age of about
   A. 2 years
   B. 3 years
   C. 4 years
   D. 5 years
   E. 6 years
23. Metatarsus adductus is the most common foot disorder in infants. One of the following is a feature of this disorder
   A. It is caused by in utero positioning
   B. It is rarely bilateral
   C. It is more common in boys
   D. It is less common in first-born children
   E. Developmental dysplasia of the hip is a common association

24. A 9-month-old infant presented with adducted forefoot, normal midfoot and hindfoot, and a convex lateral border of his foot, while his ankle dorsiflexion and plantar flexion are normal.
Of the following, the **BEST** action at this time is
   A. evaluation by a pediatric orthopedist
   B. serial casting
   C. serial bracing
   D. reassurance
   E. surgery

25. Newborn and toddler flatfoot is the result of ligamentous laxity and fat in the medial longitudinal arch. This is called developmental flatfoot and usually improves by the age of
   A. 2 years
   B. 4 years
   C. 6 years
   D. 8 years
   E. 10 years

26. A young athlete develops heel pain with activity that decreases with rest; he is limping with no swelling. He has pain to palpation of the posterior calcaneus and tight heel cords. Infection and tumor were excluded by radiographs
Of the following, the **MOST** likely diagnosis is
   A. Kohler disease
   B. Sever disease
   C. Freiberg disease
   D. tarsal coalition
   E. skewfoot
27. Polydactyly (extra toes) is usually found on the initial newborn physical examination. It may be associated with the following malformation syndrome
   A. Apert syndrome
   B. de Lange syndrome
   C. Holt-Oram syndrome
   D. Fetal hydantoin syndrome
   E. Rubinstein-Taybi syndrome

28. Scoliosis is alterations in normal spinal alignment that occur in the anteroposterior plane.
Of the following, the myopathic disease which cause scoliosis is
   A. spinocerebellar degeneration
   B. Friedreich ataxia
   C. Charcot-Marie-Tooth disease
   D. arthrogryposis
   E. syringomyelia

29. Idiopathic scoliosis is characterized by all the following EXCEPT
   A. It is the most common form of scoliosis
   B. It occurs in children with neurological defect
   C. Approximately 20% of patients have a positive family history
   D. The incidence is slightly higher in girls than boys
   E. The condition is more likely to progress and require treatment in females

30. Initial treatment for scoliosis is likely observation and repeat radiographs to assess for progression. The risk factors for curve progression include all the following EXCEPT
   A. gender
   B. curve location
   C. curve magnitude
   D. patient age
   E. skeletally maturity

31. More than 60% of patients with congenital scoliosis have other associated abnormalities.
Of the following, the MOST common one is
   A. renal anomalies
   B. congenital heart disease
   C. tethered cord
D. syringomyelia 
E. diastematomyelia

32. Silent radiolucent lesion with thin sclerotic border discovered incidentally on x-ray is a/an
A. nonossifying fibroma 
B. eosinophilic granuloma 
C. Brodie abscess 
D. unicameral bone cyst 
E. aneurysmal bone cyst

33. Congenital muscular torticollis is associated with
A. developmental dysplasia of the hip 
B. cataract 
C. microcephaly 
D. congenital heart disease 
E. spinal deformity

34. Red flags for childhood back pain include all the following EXCEPT
A. increasing pain 
B. fever 
C. bladder dysfunction 
D. school age 
E. painful left thoracic spinal curvature

35. The MOST common location of spondylolysis is
A. T9 
B. T11 
C. L1 
D. L3 
E. L5

36. The MOST common location of spondylolisthesis is
A. L2 on L3 
B. L3 on L4 
C. L4 on L5 
D. L5 on S1 
E. S1 on S2
37. An adolescent athlete complains of an insidious onset of low back pain persisting over 6 weeks. The pain tends to worsen with activity and with extension of the back and improves with rest, the pain radiated to the buttocks. Of the following, the **MOST** likely cause is
   A. diskitis
   B. ankylosing spondylitis
   C. vertebral osteomyelitis
   D. spondylolisthesis
   E. conversion disorder

38. A 4-year-old child presented with back pain, abdominal pain, irritability, and refusal to walk or sit. The child holds his spine in a straight position with loss of lumbar lordosis. The white blood cell count is normal, but the ESR and CRP are high. Of the following, the **MOST** likely cause is
   A. diskitis
   B. ankylosing spondylitis
   C. vertebral osteomyelitis
   D. spondylolisthesis
   E. spondylolysis

39. A 16-year-old athlete male presented with burning pain, weakness, and numbness of right upper extremity after lateral flexion of the neck away from the involved upper extremity, the symptoms resolve after 10 minutes. Of the following, the **MOST** likely cause is
   A. brachial plexopathy
   B. proximal humeral epiphysiolysis
   C. overuse injury
   D. glenohumeral dislocation
   E. Sprengel deformity

40. Radial head subluxation (nursemaid’s elbow) is characterized by all the following **EXCEPT**
   A. The subluxation is usually caused by a quick pull on the extended elbow
   B. The child usually holds the hand in a pronated position
   C. Radiographs are usually necessary
   D. There is a high rate of recurrence for this injury
   E. The problem generally resolves with maturation
41. A 10-year-old child presented with elbow pain, decreased range of motion, and tenderness to palpation over the capitellum. Radiographs reveal fragmentation of the capitellum. Of the following, the **MOST** likely diagnosis is

A. medial humeral epicondyle apophysitis
B. Panner disease
C. osteochondritis dissecans of the capitellum
D. nursemaid’s elbow
E. Little Leaguer’s elbow
1. (C). Subluxation is incomplete loss of contact between two joint surfaces.
2. (B). Toe walking is a common complaint in early walkers. A physician should evaluate any child older than 3 years of age who still toe walks. Although this is most likely habit, a neuromuscular disorder (cerebral palsy, tethered cord), Achilles tendon contracture (heel cord tightness), or a leg-length discrepancy should be considered.
3. (D). Toddler’s fracture is an oblique fracture of the distal tibia without a fibula fracture. There is often no significant trauma. Patients are usually 1 to 3 years old, but can be as old as 6 and present with limping and pain with weight bearing. There may be minimal swelling and pain. Initial radiographs do not always show the fracture; if symptoms persist, a repeat x-ray in 7 to 10 days may be helpful.
4. (D). Physiologic risk factors for DDH include a generalized ligamentous laxity, perhaps from maternal hormones that are associated with pelvic ligament relaxation (estrogen and relaxin). Female infants are at higher risk (9:1); family history is positive in 20% of all patients with DDH. Other risk factors include breech presentation, firstborn child (60%), oligohydramnios, and postnatal infant positioning.
5. (A). Congenital muscular torticollis, metatarsus adductus, and clubfoot are associated with DDH. An infant with any of these three conditions should receive a careful examination of the hips.
6. (A). After 2 months of age, the hip may develop muscular contractures, preventing positive Ortolani tests.
7. (C). Ultrasound should be obtained after 6 weeks of age to avoid confusion with physiologic laxity. Because the femoral head begins to ossify at 4 to 6 months of age, plain radiographs can be misleading until patients are older.
8. (B). Bilateral fixed dislocations present a diagnostic dilemma because of the symmetry on exam. The Klisic test is useful in this situation; it is done by placing the third finger over the greater trochanter and the index finger over the anterior superior iliac spine, then drawing an imaginary line between the two. The line should point to the umbilicus in a normal child. However, in a dislocated hip, the greater trochanter is elevated, which causes the line to project lower (between the umbilicus and pubis).
9. (E).
10. (D). Patients with transient synovitis are often afebrile, walk with a painful limp, and have normal to minimally elevated white blood cell count, C-reactive protein, and erythrocyte sedimentation rate compared with bacterial diseases of the hip.

11. (C). Hypothyroid.

12. (A). Internal femoral torsion or femoral anteversion is the most common cause of in-toeing in children 2 years or older.

13. (B). Internal tibial torsion is the most common cause of in-toeing in a child younger than 2 years old. When it is the result of in utero positioning, it may be associated with metatarsus adductus.

14. (C). It can take until 7 to 8 years of age for correction, so it is important to inform families of the appropriate timeline. Braces (Denis Browne splint) do not improve these conditions.

15. (B). External tibial torsion is the most common cause of out-toeing and may be associated with a calcaneovalgus foot. This is often related to in utero positioning. It may improve over time, but because the tibia rotates externally with age, external tibial torsion can worsen. It may be an etiologic factor for patellofemoral syndrome, especially when combined with femoral anteversion. Treatment is usually observation and reassurance, but patients with dysfunction and cosmetic concerns may benefit from surgical intervention.

16. (B). The majority of patients who present with knock-knees (genu varum) or bowlegs (genu valgum) are normal. Infants are born with maximum genu varum. The lower extremity straightens out around 18 months of age. Children typically progress to maximal genu valgum around 4 years. The legs are usually straight to a slight genu valgum in adulthood.

17. (A). Physiologic bowlegs are most common in children older than 18 months with symmetrical genu varum. This will generally improve as the child approaches 2 years of age.

18. (E). Computed tomography (CT) scanogram is the most accurate measure of LLD, but also has the highest radiation exposure. Technology such as EOS/slot scanning is an extremely accurate, reduced-radiation alternative to CT scan. The measured discrepancy is followed using Moseley and Green-Anderson graphs.

19. (A). LLD greater than 2 cm usually requires treatment. Shoe lifts can be used, but they will often cause psychosocial problems for the child and may make the shoes heavier and less stable. Surgical options include shortening of the longer extremity, lengthening of the shorter extremity, or a combination of the two procedures. Discrepancies less than 5 cm are treated by epiphysiodesis (surgical physeal closure) of the affected side, whereas discrepancies greater than 5 cm are treated by lengthening.
20. (B). Osgood-Schlatter disease is a common cause of knee pain at the insertion of the patellar tendon on the tibial tubercle. The stress from a contracting quadriceps muscle is transmitted through the developing tibial tubercle, which can cause a microfracture or partial avulsion fracture in the ossification center. It usually occurs after a growth spurt and is more common in boys. The age at onset is typically 11 years for girls and 13 to 14 years for boys. Patients will present with pain during and after activity as well as have tenderness and local swelling over the tibial tubercle. Radiographs may be necessary to rule out infection, tumor, or avulsion fracture.

21. (C). Idiopathic anterior knee pain is a common complaint in adolescents. It is particularly prevalent in adolescent female athletes. Previously, this was referred to as chondromalacia of the patella, but this term is incorrect as the joint surfaces of the patella are normal. It is now known as patellofemoral pain syndrome (PFPS).

22. (C). The navicular ossifies at about 3 years of age for girls and 4 years for boys.

23. (A). It is characterized by a convexity of the lateral foot and is caused by in utero positioning. It is bilateral in half of cases. Occurring equally in boys and girls, it is more common in first-born children because of the smaller primigravid uterus. Two percent of infants with metatarsus adductus have developmental dysplasia of the hip.

24. (D). True metatarsus adductus resolves spontaneously over 90% of the time without treatment, so reassurance is all that is needed. Metatarsus adductus that does not improve within 2 years needs evaluation by a pediatric orthopedist. Persistent cases may benefit from serial casting or bracing, and potentially surgery.

25. (C).

26. (B). The common presentation is a young athlete who develops heel pain with activity that decreases with rest. Swelling is rare, but limping may be associated with Sever disease. The child will have pain to palpation of the posterior calcaneus and often tight heel cords. Radiographs are rarely indicated, but with persistent pain they should be done to exclude infection or tumor.

27. (E). Polydactyly may be seen in
- Ellis-van Creveld syndrome
- Rubinstein-Taybi syndrome
- Carpenter syndrome
- Meckel-Gruber syndrome
- Polysyndactyly
- Trisomy 13
- Orofaciodigital syndrome

28. (D). Neuromuscular causes of scoliosis
Neuropathic diseases
- Upper motor neuron disease
- Cerebral palsy
- Spinocerebellar degeneration
- Friedreich ataxia
- Charcot-Marie-Tooth disease
- Syringomyelia
- Spinal cord tumor
- Spinal cord trauma
- Lower motor neuron disease
- Myelodysplasia
- Poliomyelitis
- Spinal muscular atrophy

Myopathic diseases
- Duchenne muscular dystrophy
- Arthrogryposis
- Other muscular dystrophies

29. (B). It occurs in healthy, neurologically normal children.
30. (E). The risk factors for curve progression include gender, curve location, and curve magnitude. Girls are five times more likely to progress than boys. Younger patients are more likely to progress than older patients. Typically, curves under 25° are observed. Progressive curves between 20° and 50° in a skeletally immature patient are treated with bracing. A radiograph in the orthotic is important to evaluate correction. Curves greater than 50° usually require surgical intervention.
31. (A). Renal anomalies occur in 20% of children with congenital scoliosis, with renal agenesis being the most common; 6% of children have a silent, obstructive uropathy suggesting the need for evaluation with ultrasonography. Congenital heart disease occurs in about 12% of patients. Spinal dysraphism (tethered cord, intradural lipoma, syringomyelia, diplomyelia, and diastematomyelia) occurs in approximately 20% of children with congenital scoliosis.
32. (A).
33. (A). Congenital muscular torticollis is associated with skull and facial asymmetry (plagiocephaly) and developmental dysplasia of the hip.
34. (D). Red flags for childhood back pain include persistent or increasing pain, systemic findings (e.g., fever, weight loss), neurologic deficits, bowel or bladder dysfunction, young age (under 4 is strongly associated with tumor), night waking, pain that restricts activity, and a painful left thoracic spinal curvature.
35. (E). Spondylolysis is a defect in the pars interarticularis. The most common location of spondylolysis is L5, followed by L4.
36. (D). Spondylolisthesis refers to bilateral defects with anterior slippage of the superior vertebra on the inferior vertebra. The lesions are not present at birth, but about 5% of children will have the lesion by 6 years of age. It is most common in adolescent athletes, especially those involved in sports that involve repetitive back extension. The most common location of spondylolisthesis is L5 on S1.

37. (D).

38. (A). Diskitis is an intervertebral disk space infection that does not cause associated vertebral osteomyelitis. The most common organism is Staphylococcus aureus. The infection can occur at any age but is more common in patients under 6 years of age.

39. (A). Brachial plexopathy is an athletic injury, commonly referred to as a stinger or burner. The symptoms are often likened to a dead arm. There is pain (often burning), weakness, and numbness in a single upper extremity. There are three mechanisms of injuries:

- Traction caused by lateral flexion of the neck away from the involved upper extremity
- Direct impact to the brachial plexus at Erb’s point
- Compression caused by neck extension and rotation toward the involved extremity

Symptoms are always unilateral and should resolve within 15 minutes. It is paramount to assess the cervical spine for serious injury. Bilateral symptoms, lower extremity symptoms, persistent symptoms, or recurrent injury are all signs of more serious disease and may need a more extensive workup.

40. (C). Usually, radiographs are not necessary unless reduction cannot be obtained or there is concern for fracture (swelling and bruising).

41. (B). Panner disease is an osteochondritis of the capitellum (lateral portion of distal humeral epiphysis) that occurs spontaneously in late childhood. Clinical features include elbow pain, decreased range of motion, and tenderness to palpation over the capitellum. Radiographs reveal fragmentation of the capitellum. Treatment is activity restriction and follow-up radiographs to demonstrate spontaneous reossification of the capitellum over several months. There is usually no need for further treatment or imaging studies. This is not to be confused with osteochondritis dissecans of the capitellum, which usually will occur in adolescents involved with throwing sports.
THANKS FOR ALLAH
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