PEDIATRIC BOARD CERTIFICATION AND RECERTIFICATION MULTIPLE CHOICE QUESTIONS AND ANSWERS

Volume 4

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PEDIATRIC BOARD

CERTIFICATION AND RECERTIFICATION

MULTIPLE CHOICE QUESTIONS AND ANSWERS

Volume 4

AN EXCELLENT GUIDE

AN ESSENTIAL TOOL FOR BOARD CERTIFICATION

by

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DEDICATION

Dedicated to all the readers

INTRODUCTION

This is volume 4 of the first edition of Pediatric Board Certification and Recertification Multiple Choice Questions and Answers. A total of 1005 questions and answers are presented here.

This book will help candidates to prepare for the examination. You are well aware that reading only multiple choice questions and answers is not enough to pass the Board Examination.

First, try to answer these questions by yourself; then evaluate your knowledge. If you have answered more than 85% correctly, you will probably do well in the examination. I have written this book after repeated requests from candidates and residents for a practice book that replicates the exam format.

My sincere thanks go to Josh Furman for his continued support, Preeti Agarwal and Atul Kumar (atul_kumar@apyl.com) for their first-rate desktop publishing expertise. Dr. Azfar Shabbir, M.D., Dr. Marion Tamesis, M.D., Dr. Mahmood Benham, M.D., Dr. Roland Tayaba, M.D., Dr. Maria Macapagal, M.D., Nivedita Kanjilal, Anirban Kanjilal for their support.

Good luck to all of you.

D. Kanjilal, M.D.

1. A routine physical examination in a 2-year-old boy reveals rooting reflex. The most likely diagnosis is:
   a) Normal
   b) Cleft lip
   c) Cleft palate
   d) Visual defects
   e) CNS abnormalities

2. A child begins to develop sense of individuality. She is aware of strangers. She is frightened by separation from her parents. The most likely age of this girl is:
   a) 6 months
   b) 8 months
   c) 10 months
   d) 12 months
   e) 15 months

3. A 12- month-old girl is toilet trained. The long-term outcome of this girl is:
   a) Questionable
   b) Good
   c) Very good
   d) Average
   e) Excellent

4. A 3-year-old boy can read. The long-term outcome of this boy is:
   a) Average
   b) Good
   c) Very Good
   d) Excellent
   e) Questionable

5. The first visible sign of puberty in girls is:
   a) Onset of menstruation
   b) Axillary hairs
   c) Pubic hairs
   d) Appearance of breast buds
   e) Enlargement of clitoris

6. In boys, testicular enlargement begins as early as:
   a) 8 years of age
   b) 8½ years of age
   c) 9 years of age
   d) 9½ years of age
   e) 10 years of age

7. The development of underarm odor and faint genital hair is most likely due to the following hormone:
   a) FSH
   b) LH
   c) GnRH
   d) Testosterone
   e) Dehydroepiandosterone sulfate (DHEAS)

8. A 12-month-old boy appears with failure to gain weight for the last 3 months. The boy has a history of repeated regurgitation of food after eating. Mother denies nausea, abdominal pain, dysuria, diarrhea, and bloody stools. The boy was completely asymptomatic prior to this episode. He was born by NSVD with Apgar scores of 9 and 9 at 1 and 5 minutes respectively. He was discharged home at 2 days of age. The mother is living alone with the child for the last 3 months. The most likely diagnosis is:
   a) Malrotation
   b) Volvulus
   c) Food allergy
   d) Rumination disorder
   e) Intussuception

9. A child is diagnosed to have restrictive pericardial effusion. The important clinical signs are the following:
   a) Tachycardia, hypotension, and decreased oxygen saturation
   b) Bradycardia, hypotension, and decreased perfusion
   c) Tachycardia, hypertension, and swelling of face
   d) Arrythmia, normal BP, and venous engorgement of neck
   e) Fever, tachycardia, and hypertension

10. The most common unintentional cause of death in children with epilepsy is:
   a) Drowning
   b) Motor vehicle accidents
   c) Medication overdose
   d) Head injury
   e) Pedestrian injury

11. The most recognized toxicity of halothane inhalation anesthetic agent is:
   a) Nephritis
   b) Pancreatitis
   c) Pneumonitis
   d) Myocarditis
   e) Hepatitis

12. A 26-week-gestational age preterm infant is placed on a mechanical ventilator. The infant is less responsive to stimulation. Head sonogram reveals a grade 2 intraventricular hemorrhage. The parents want to remove the baby from a ventilator and let him die. What should the doctor do in this situation?
   a) The physician should remove the infant from a mechanical ventilator.
   b) The physician should get a court order.
   c) The parents should get a court order.
   d) The physician should avoid the parents and continue patient's care.
   e) The physician should not remove the infant from a mechanical ventilator and arrange for a meeting with the parents and social worker.

13. All of the following methods can be used to prevent injuries from burn **except**:
   a) Crawling beneath smoke if indoors
   b) Practising escape procedures
   c) Wrapping with blanket
   d) Running if clothes catches fire
   e) Using materials for education

14. All of the following methods can be used to prevent fires **except**:
   a) Use flame retardant-treated clothing
   b) Stop cigarette smoking inside the house
   c) Use fire-matches-lighter
   d) Install smoke detectors
   e) Control of hot water thermostat (maximum temperature 128° F)

15. A physician wants to measure the clinical assessment of pain in a 5-year-old girl. All of the following pain measurement tools can be used **except**:
   a) Visual Analog Scale (VAS)
   b) Hormonal-metabolic measures
   c) Autonomic measures
   d) Faces scale
   e) Color and other analog scales

16. A physician wants to measure the clinical assessment of pain in a 3-year-old boy. All of the following pain measurement tools can be used **except**:
   a) Autonomic measures
   b) Behavioral scales
   c) Hormonal-metabolic measures

d) Wong-Baker faces scale
e) Combined behavior-physiological scales

17. A conscious child is on a mechanical ventilator. A physician wants to measure the clinical assessment of pain. The following pain measurement tool is useful in patients who are on a mechanical ventilator:
a) Behavioral scales
b) Hormonal-metabolic measures
c) Autonomic measures
d) Oucher faces scale
e) Behavioral-physiological measures

18. A child has congnitive limitations. All of the following pain measurements tools can be used **except**:
a) Autonomic measures
b) Visual Analog Scales
c) Hormonal-metabolic measures
d) Behavioral scales
e) Behavioral-physiological scales

19. A 3-month-old boy appears with hypoglycemic seizures. A physical examination reveals short stature, hepatomegaly, protruberant abdomen, and doll-like facies. The laboratory findings reveal hypoglycemia, lactic acidosis, hyperuricemia, and hyperlipidemia. The most likely enzyme deficiency is:
a) Glucose-6-phosphatase
b) Acid maltase
c) Galatose-1-phosphate uridyltransferase
d) Myophosphorylase
e) Fructokinase

20. A 4-month-old girl appears with hepatomegaly, massive cardiomegaly, pulmonary atelactasis with a secondary infection. ECG reveals prominent P waves, short P-R intervals, massive QRS voltages, biventricular hypertrophies, and intraventricular conduction delays. The most likely diagnosis is:
a) Endocardial cushion defect
b) Pompe disease
c) Myocarditis
d) Pericarditis
e) Pneumonia causing cardiac failure

21. The mode of inheritance in patients with Pompe disease (glycogenosis II) is:
a) Autosomal recessive
b) Autosomal dominant
c) X-linked recessive
d) X-linked dominant
e) Multifactorial

22. The enzyme deficiency in patients with Pompe disease is:
a) Pyruvate dehydrogenase
b) Glucose-6-phosphate translocase
c) Liver phosphorylase
d) Glycogen synthetase
e) Acid maltase

23. The preferred diagnostic study in patients with Pompe disease is:
a) Renal biopsy
b) Muscle biopsy
c) Serum CK level
d) Acid maltase levels in WBCs
e) Serum acid maltase level

24. An infant is born with erythroblatosis fetalis. The infant may have the following finding:
    a) High insulin levels in serum
    b) Low insulin levels in serum
    c) High glucagon levels in serum
    d) Hyperglycemia
    e) High TSH levels in serum

25. Beckwith-Wiedemann syndrome has all of the following features **except**:
    a) Exomphalos
    b) Gigantism
    c) Visceromegaly
    d) Lateral earlobe fissure
    e) Macrocephaly

26. The patients with Beckwith-Wiedemann syndrome may develop all of the following tumors **except**:
    a) Retinoblastoma
    b) Wilms' tumor
    c) Adrenal carcinoma
    d) Hepatoblastoma
    e) Rhabdomyosarcoma

27. Beckwith-Wiedemann syndrome is due to the mutation in the following chromosome:
    a) Chromosome 7
    b) Chromosome 11
    c) Chromosome 13
    d) Chromosome 15
    e) Chromosome 17

28. A full term male infant appears jaundice during the first 24 hours of life due to 'AO' incompatibility and Coombs positive results. His serum bilirubin level is 35 mg/dL. A double-volume exchange transfusion is performed. The most likely morbidity is:
    a) Seizures
    b) Spastic diplegia
    c) Hydrocephalus
    d) Spastic quadriplegia
    e) Choreoathetotic cerebral palsy

Match the following perinatal and neonatal illnesses and morbidities (29-33):

29. Sensorineural hearing loss
30. Blindness
31. Paralysis-paresis
32. Spastic diplegia
33. Communicating hydrocephalus

    a) Intraventricular hemorrhage
    b) Periventricular leukomalacia
    c) Gentamicin
    d) Birth trauma
    e) Retinopathy of prematurity

34. All of the following conditions can cause poor speech **except**:
    a) Hearing deficit
    b) Immaturity
    c) Hyperoxia
    d) Chronic illness
    e) Prolonged endotracheal intubation

35. All of the following conditions can cause microcephaly **except**:
    a) Asymmetric SGA
    b) CMV
    c) Congenital rubella
    d) Congenital toxoplasmosis
    e) Hypoxic-ischemic encephalopathy

36. All of the following conditions can cause encephalopathy **except**:
    a) Herpes simplex virus
    b) Congenital toxoplasmosis
    c) Congenital rubella
    d) IVH
    e) Human immunodeficiency virus

37. An adolescent white girl appears for a routine annual physical examination. She is sexually active. All of the following tests are indicated **except**:
    a) Hematocrit
    b) Screening for sickle cell anemia
    c) Papanicolaou smears
    d) Testing for tuberculosis
    e) Routine urine analysis

38. A 16-year-old girl has anorexia nervosa. However, she is not very depressed. All of the following therapies are indicated in this patient **except**:
    a) Nutritional rehabilitation
    b) Individual psychotherapy
    c) Family psychotherapy
    d) Antidepressant drug
    e) Behavioral modification therapy

39. All of the following are clinical manifestations in female patients with anorexia nervosa (AN) **except**:
    a) Postural hypotension
    b) Malnutrition
    c) Menstrual irregularities
    d) Cardiac arrythmias
    e) Tachycardia

40. In patients with anorexia nervosa, daily weight gain should be about:
    a) 200-400 grams
    b) 400-600 grams
    c) 600-800 grams
    d) 800-1,000 grams
    e) 1,000-1,200 grams

41. All of the following are complications of rapid rehydration in patients with anorexia nervosa **except**:
    a) Hyperphosphatemia
    b) Congestive heart failure
    c) Pulmonary edema
    d) Tachypnea
    e) Death

42. A patient with anorexia nervosa abused ipecac for induced vomiting. The patient may develop the following complication due to the abuse of ipecac:
    a) Mitral valve stenosis
    b) Aortic stenosis
    c) Endocarditis
    d) Myocarditis
    e) Mitral valve prolapse

43. The following chromosome is involved in patients with CATCH 22 syndrome:
    a) Chromosome 12
    b) Chromosome 15
    c) Chromosome 21
    d) Chromosome 22
    e) Chromosome X

44. All of the following features are present in patients with CATCH 22 syndrome **except**:
    a) Cardiac
    b) Abnormal facies
    c) Thymic hypoplasia
    d) Hypocalcemia
    e) Cleft lip

45. A routine CBC result in a healthy newborn reveals the total lymphocyte count is 1950/mm^3. The most likely diagnosis is:
    a) Normal
    b) Congenital CMV infection
    c) Congenital herpes infection
    d) Parvovirus infection
    e) Severe combined immunodeficiency (SCID)

46. A 5-month-old boy appears with respiratory distress for the last 24 hours. He had two episodes of pneumonia and four episodes of otitis media and six episodes of diarrhea. He is delayed for his growth and development. A physical examination reveals bilateral rales and right otitis media. All of the following findings are true for this patient **except**:
    a) Total CBC lymphocyte counts are 1600/mm^3.
    b) Chest x-ray reveals bilateral infiltrates.
    c) Blood culture is positive for Streptococcus pneumoniae.
    d) Serum immunoglobulin levels are elevated.
    e) Conductive hearing loss in the right ear.

47. All of the following are features of CHILD syndrome **except**:
    a) Congenital hemidysplasia
    b) Limb defects
    c) Ichthyosiform erythroderma
    d) Facial exfoliative erythematous dermatitis
    e) Unilateral hypomelia ipsilateral to the dermatosis

48. The mode of inheritance in patients with CHILD syndrome is:
    a) Autosomal recessive
    b) X-linked recessive
    c) Autosomal dominant
    d) X-linked dominant
    e) Multifactorial

Match the following diseases with enzyme defects (49-53):

49. Niemann-Pick type I
50. Gaucher disease
51. Metachromatic leukodystrophy
52. GM1 gangliosidosis
53. GM2 gangliosidosis

    a) Arylsulfatase A
    b) Beta-galactosidase
    c) Sphingomyelinase
    d) Beta-hexosaminidases A and B
    e) Glucocerebrosidase

Match the following porphyrias with different clinical manifestations (54-58):

54. Acute intermittent porphyria
55. Congenital erythropoietic porphyria
56. Porphyria cutanea tarda
57. Hepatoerythropoietic porphyria
58. Erythropoietic protoporphyria

a) Erythema, edema, itching, burning sensation, and scarring vesicles
b) Abdominal pain, diarrhea, vomiting, constipation, ileus, dysuria, muscular hypotonia, sensory neuropathy, seizure, and respiratory failure
c) Pink to brown staining urine, bullae, crusts, scar formations, hypopigmentation, hyperpigmentation, sclerodermoid lesions, alopecia, hypertrichosis, splenomegaly, hemolytic anemia, and erythrodontia
d) Bullae on the sun-exposed areas of the skin, particularly the dorsa of the hands, skin fragility, crusts, scar formations, sclerodermoid lesions, hyperpigmentation, hypopigmentation, and hypertrichosis
e) Pink urine, bullae, crusts, scar formation, sclerodermoid lesions, hypopigmentation, hyperpigmentation, and hypertrichosis

59. A child appears with burning sensations on the face and distal part of the hands within an hour exposure to the Sun. He also complains of occasional upper abdominal pain. A physical examination reveals normal skin and mild tenderness in the right upper quadrant. Abdominal sonogram reveals gallstones. The next step in management:
a) Psychiatric consultation
b) Beta-carotene
c) Upper GI series
d) Duodenoscopy
e) Observation and follow-up after 1 month

60. The brain metabolism mostly uses the following substrate:
a) Galactose
b) Fructose
c) Fatty acids
d) Amino acids
e) Glucose

61. The major, long-term outcome of severe, prolonged hypoglycemia in infants is:
a) Growth retardation
b) Microcephaly
c) Cerebral atrophy
d) Mental retardation
e) Periventricular leukomalacia

62. The glucose level normally reaches nadir in newborn infants:
a) 15-30 minutes after birth
b) 30-60 minutes after birth
c) 1-3 hours after birth
d) 3-5 hours after birth
e) 5-8 hours after birth

63. A poorly controlled diabetic mother with vascular disease delivered the newborn at 38-week-gestational age. A physical examination most likely reveals:
a) Macrosomia
b) Macrocephaly
c) Hepatomegaly
d) Large for gestational age
e) Small for gestational age

64. A 15-year-old girl appears for a routine physical examination. A 35-year-old male physician is going to examine her. She came with her mother. The following condition should be maintained during a physical examination:
a) The room should have very bright light.
b) The pelvic examination should be performed very gently.

c) The patient should lie down during breasts examination.
d) The physician should be very friendly during a physical examination.
e) A chaperone should be present during a physical examination.

Match the following drugs and major metabolites (65-69):

65. Heroin
66. Cocaine
67. Alcohol
68. Cannabinoids
69. Methaqualone

a) Acetaldehyde
b) Morphine
c) Benzoyl ecgonine
d) Carboxy-and hydroxymetabolites
e) Hydroxylated metabolites

Match the following clinical manifestations and drugs (70-74):

70. Euphoria, pinpoint pupils, hypothermia, respiratory depression, and pulmonary edema
71. Hypertension, hyperpyrexia, seizures, psychotic ideation, sudden violence, and ventricular irritability
72. Dizziness, dilated pupils, fever, flushing, tachycardia, and sensation of synesthesia or "seeing" smells and "hearing" colors
73. Headaches, syncope, lightheadedness, severe hypotension, cutaneous flushing followed by vasoconstriction, and tachycardia
74. Euphoria, jaw clenching, blurred vision, teeth grinding, anxiety, panic attacks, psychosis, hyperthermia after vigorous dancing at a "rave".

a) Lysergic acid diethylamide
b) Amphetamines
c) Methylenedioxymethamphetamine
d) Opiates
e) Volatile substance

Match the following disorders with functional deficiencies (75-79):

75. Wiskott-Aldrich syndrome
76. Ataxia-telangiectasia
77. Cartilage-hair hypoplasia
78. Major histocompatibility complex class II antigen deficiency
79. Lymphoproliferative syndrome

a) Low immunoglobulins, CD4 deficiency, and lack of T-cell responses to antigens
b) Thrombocytopenia, T-cell deficiency, and poor antipolysaccharide antibody production
c) Combined B- and T-cell defects of varying severity
d) Selective IgA deficiency and T-cell deficiency
e) Poor T-cell responses, autoimmunity, impaired apoptosis (programmed cell death), and increased bcl-2

80. A child has partial DiGeorge syndrome. All of the following features are present **except**:
a) Mild hypoplasia of thymus
b) Otitis media
c) P. carinii pneumonia
d) Normal growth
e) Mild hypoplasia of parathyroid glands

81. All of the following are complications of chronic systemic use of corticosteroids **except**:
a) Growth suppression
b) Hyperglycemia
c) Delayed sexual maturation

d) Diabetes mellitus
e) Hyperkalemia

82. The most common hypersensitivity reaction of eyes is:
a) Contact allergy
b) Atopic keratoconjunctivitis
c) Giant papillary conjunctivitis
d) Allergic conjunctivitis
e) Vernal keratoconjunctivitis

83. The major causes of death in patients with SLE include all of the following **except**:
a) Nephritis
b) Myocardial infarction
c) CNS disease
d) Intestinal bleeding
e) Infection

84. Myocardial infarction in patients with SLE may be due to:
a) Coronary artery abnormalities
b) Endocardial thickening
c) Pericardiac thickening
d) Aortic root thickening
e) Chronic use of corticosteroids

85. The most common inflammatory myopathy in pediatric age group is:
a) Juvenile dermatomyositis
b) SLE
c) Scleroderma
d) Sjogren syndrome
e) Behcet disease

86. A 9-year-old girls appears with fatigue, irritability, low-grade fever, arthralgia, abdominal pain, weight loss, and rash. A physical examination reveals erythema over the malar areas and bridge of the nose. She also has violaceous (heliotropic) discoloration of the upper eyelids and mild facial edema. All of the following laboratory findings are present in this patient **except**:
a) Coombs-positive anemia
b) Presence of antinuclear antibody
c) Absence of antibodies to DNA
d) Negative rheumatoid factor
e) Elevated serum creatine kinase

87. All of the clinical manifestations are more frequently present in patients with juvenile dermatomyositis **except**:
a) Gottron papules
b) Heliotrope eyelids
c) Gower sign
d) Retinitis
e) Avascular nailfold capillaries, dropout of capillary end-loops, and terminal bush formation.

88. The following laboratory finding is absent in patients with juvenile dermatomyositis:
a) Elevated ESR (erythrocyte sedimentation rate)
b) Elevated lactic acid dehydrogenase
c) Negative antibodies to SSA
d) Elevated CD4: CD8 ratio
e) Elevated levels of von Willebrand factor antigen

89. The therapy in patients with juvenile dermatomyositis includes all of the following **except**:
a) Bed rest
b) Refer to pediatric rheumatologist
c) Corticosteroids

d) Intravenous immunoglobulins
e) Sunscreen

90. A child appears with febrile episodes. The fever is characterized by febrile periods and normal temperature between febrile illnesses. The most likely the child has:
a) Remittent fever
b) Relapsing fever
c) Biphasic fever
d) Periodic fever
e) Intermittent fever

Match the diseases with different types of fever (91-95):

91. Hypothalamic disorder
92. Malaria causes by Plasmodium vivax
93. Poliomyelitis
94. Cyclic neutropenia
95. Malaria caused by Plasmodium malariae

a) Biphasic fever
b) Central fever
c) Periodic fever
d) Quartan fever
e) Tertian fever

Match the diseases with different clinical manifestations (96-100):

96. E. coli bacteremia
97. Drug fever
98. Dengue fever
99. Apthous stomatitis
100. Borrelia recurrentis infections

a) Relative bradycardia
b) Relative tachycardia
c) Biphasic fever
d) Relapsing fever
e) Periodic fever

101. All of the following are primary signs and symptoms of sepsis **except**:
a) Hypotension
b) Tachypnea
c) Shaking chills
d) Fever
e) Hypothermia

102. The definitive diagnostic test in patients with septic shock is:
a) Leukocytosis
b) Leukocytopenia
c) Prolonged PTT
d) High CSF cell counts
e) Positive blood cultures

103. A 2-year-old boy is admitted in pediatric intensive care unit with septic shock. He appears in the ER with tachypnea, fever, and lethargy. A physical examination reveals purpura fulminans, cyanosis, nasal flaring, and intercostal retractions. Vital signs reveal hypotension, tachycardia, hyperthermia, and tachypnea. The boy received 100% oxygen by mask immediately. All the blood work, urine, and CSF specimens are obtained. The chest x-ray is performed. The therapy for septic shock includes all of the following **except**:
a) A third-generation cephalosporin (e.g., ceftriaxone or cefotaxime) and aminoglycoside should be given.

b) Intravenous fluid therapy (e.g., normal saline, Ringer's solution) 20 mL/kg should be given immediately.
c) Continue to give 100% oxygen by face mask.
d) The patients with DIC should receive fresh frozen plasma, cryoprecipitates, and platelets.
e) Inotropic agents (e.g., dopamine, epinephrine) can be used in persistent hypotension.

104. The ominous laboratory finding of overwhelming sepsis is:
a) Elevated neutrophil counts
b) Increased band counts
c) Vacuolation of neutrophils
d) Toxic granulations
e) Neutropenia

105. The laboratory findings of sepsis include all of the following **except**:
a) Metabolic alkalosis
b) Thrombocytopenia
c) Anemia
d) Hypoxia
e) Hypercarbia

106. All of the following are common complications of group A Streptococcal pharyngitis when the treatment is inadequate or not provided **except**:
a) Peritonsillar abscess
b) Retropharyngeal abscess
c) Cervical lymphadenitis
d) Otitis media
e) Pneumonia

Match the following diseases that occurs due to complications of group A Streptococcal infections (107-108):

107. Acute glomerulonephritis
108. Acute rheumatic fever

a) Group A Streptococcal skin infection
b) Group A Streptococcal upper respiratory tract infection
c) All of the above
d) None of the above
e) Unknown

109. A child has poststreptococcal reactive arthritis. All of the following statements are true about this condition **except**:
a) Usually involves the smaller joints.
b) The latent period between the group A Streptococcal pharyngitis and subsequent reactive arthritis is usually shorter (i.e., less than 10 days).
c) Usually does not respond dramatically to therapy with aspirin or other NSAIDs.
d) All patients have serologic evidence of group A Streptococcal infection.
e) About 50% of patients have positive throat culture for group A Streptococcal infection.

110. A child appears for a routine physical examination. He was diagnosed with poststreptococcal reactive arthritis about 1 month ago. The child should be watched for the following complication:
a) Cholecystitis
b) Pancreatitis
c) Hepatitis
d) Encephalitis
e) Carditis

111. The Ekiri syndrome occurs due to the complication of the following infection:
a) E. coli
b) Pseudomonas aeruginosa
c) Staphylococcus aureus

d) Shigella
e) Salmonella

112. The most common complication of Shigella infection is:
a) Hypoglycemia
b) Protein loosing enteropathy
c) Sepsis
d) Dehydration
e) Inappropriate secretion of ADH

113. All of the following clinical manifestations are due to the complications of Shigella dysenteriae infections **except**:
a) Hemolytic-uremic syndrome
b) Rectal prolapse
c) Toxic megacolon
d) Blood-tinged vaginal discharge
e) Inappropriate secretion of ADH

114. A child has hemolytic-uremic syndrome (HUS). This syndrome can be produced by all of the following organisms **except**:
a) E. coli 0157 : H7
b) E. coli 0111 : NM
c) E. coli 026 : H11
d) Shigella sonnei
e) Shigella dysenteriae serotype 1

115. Bacteremia is more common in the following serotype of Shigella:
a) S. flexneri
b) S. boydii
c) S. sonnei
d) S. dysenteriae serotype 1
e) S. dysenteriae serotype 2

116. The American Academy of Pediatrics recommends the following supplementation for children between 6 months and 2 years of age who are hospitalized for measles or complications and for children older than 6 months of age with measles and immunodeficiency:
a) Iron
b) Folic acid
c) Vitamin A
d) Vitamin D
e) Calcium

117. All of the following clinical manifestations are due to the complications of measles **except**:
a) Otitis media
b) Pneumonia
c) Guillain-Barre syndrome
d) Cerebral bleeding
e) Encephalomyelitis

118. Hecht giant cell pneumonia occurs due to the following organism:
a) Rubella
b) Mumps
c) Herpes simplex virus
d) Varicella zoster virus
e) Measles

119. The best approach to prevent firearm injuries in children and adolescents is:
a) Counselling families and adolescents
b) Gun should be kept in a locked container
c) Gun should have a trigger lock.

d) Remove the firearm from the environment of children and adolescents.
e) Reading educational materials about the guns obtained from the American Academy of Pediatrics.

120. Most homicides due to firearm injuries are:
a) Acts of robbery
b) Acts of friendly fire
c) Acts of rage
d) Acts of routine practice
e) Acts of routine hunting

121. Most of the body calcium is present in bones in the form of:
a) Carbonate
b) Bicarbonate
c) Gluconate
d) Oxalate
e) Hydroxyapatite

122. The ionized or free calcium in serum is:
a) Slightly less than 10% of the total serum calcium
b) Slightly less than 25% of the total serum calcium
c) Slightly less than 50% of the total serum calcium
d) Slightly less than 60% of the total serum calcium
e) Slightly less than 70% of the total serum calcium

123. All of the following statements are true about serum calcium levels in different clinical conditions **except**:
a) A low total serum calcium concentration may be normal in patients with significant hypoalbuminemia.
b) In metabolic acidosis, less calcium is bound to albumin, more free or ionized calcium is present in serum.
c) In metabolic alkalosis, less calcium is bound to albumin, more free or ionized calcium is present in serum.
d) Each 1 gram/dL of albumin in serum binds with 0.8 mg/dL of calcium.
e) Serum ionized calcium concentration should be obtained in patients who are very ill, have low serum albumin levels, or clinical suspicion of abnormal ionized calcium level.

Match the following clinical manifestations with vitamin deficiencies and toxicities (124-128):

124. Vitamin A toxicity
125. Vitamin A deficiency
126. Vitamin B_1 deficiency
127. Vitamin B2 deficiency
128. Foliacin deficiency

a) Beriberi
b) Increased intracranial pressure
c) Ariboflavinosis
d) Megaloblastic anemia
e) Nyctalopia

129. A 10-day-old boy appears with hard nodules with overlying erythematous skin over the cheeks, thighs, buttocks, and back for the last 2 days. The boy was born by forcep delivery with Apgar scores were 3 and 6 at 1 and 5 minutes respectively. He is drinking infant formulas 2 ounces every 4 hours. His birth weight was 9 lbs. The laboratory finding should reveal:
a) Hypocalcemia
b) Hyponatremia
c) Hyperkalemia
d) Hypercalcemia
e) Pseudomonas aeruginosa bacteremia

130. A 2-year-old girl appears for brief developmental assessment. All of the following tests can be performed **except**:
a) Early screening inventory
b) Denver-II
c) Early language milestone scale (ELM)

d) Clinical adaptive test (CAT)
e) Age and stages questionnaires (ASQ)

131. A 7-year-old boy needs developmental assessment. The following test is appropriate for this age:
a) Parents evaluation of developmental status
b) Denver-II
c) Early screening inventory
d) Clinical linguistic and auditory milestones scale (CLAMS)
e) Early language milestone scale (ELM)

132. A 15-month-boy girl appears with history of breath holding for the last 1 month. The preferred method of therapy is:
a) Hug the child
b) Give whatever she demands
c) Ignore the behavior
d) Give toys immediately
e) Mouth-to-mouth resuscitation

133. A 17-month old boy appears in the ER with history of first episode of generalized seizure that lasted 30 seconds. The boy had breath holding spell followed by loss of consciousness and seizure activity. The risk of later developing seizure disorder is about:
a) 0%
b) 25%
c) 50%
d) 66%
e) 75%

134. A 2-year-old boy is lying frequently to his parents to handle conflict and reduce anxiety. His parents are very sensitive and supportive to him. The preferred initial method to manage the situation is:
a) Parents should not allow him to watch TV until he stops lying.
b) Parents should confront the child to give clear message that lying is not acceptable.
c) Parents should ignore the lying completely.
d) Parents should take it seriously if the lying is about serious matter only
e) Parents should use time-out and confined him in his room for about 2 hours.

135. A 4-year-old boy has high level of aggressive behavior. The most likely outcome of this boy in adolescence:
a) He will remain aggressive, especially without family-focused intervention
b) He will become normal adolescent without any intervention.
c) He will most likely become murderer.
d) He will most likely end up in jail.
e) He will most likely use illegal drugs.

136. All of the following behaviors may be developmentally appropriate **except**:
a) Lying
b) Stealing
c) Temper tantrums
d) Breath holding
e) Truancy

137. A 2-year-old boy appears with history of many episodes of temper tantrums for the last 2 months. His mother usually gets angry and frustrated. The preferred method to manage temper tantrums is:
a) The mother should turn away briefly from the child during the episode of temper tantrum.
b) The mother should stop the temper tantrum immediately by force.
c) The mother should lock the child in his bedroom for 2 hours.
d) The mother should not allow him to watch TV for 4 hours after each episode of temper tantrum.
e) The mother should not give food for at least 6 hours after each temper tantrum episode.

138. A 19-month-old girl appears with pain, pseudoparalysis, and edema of the legs. She has digestive disturbances, loss of appetite, and irritability. A physical examination reveals frogleg position with the hips and knees semiflexed with outward rotation of the feet. The x-ray of the lower end of femur and the upper end of tibia reveals zone of rarefaction under the white line at the metaphysis. The lateral parts of the zone of rarefaction appear as triangular defect. The most likely diagnosis is:

a) Nutritional rickets
b) Hypervitaminosis D
c) Child abuse with fracture
d) Scurvy
e) Vitamin D deficiency rickets

139. The most frequent trisomy in humans is:
a) Trisomy 8
b) Trisomy 13
c) Trisomy 16
d) Trisomy 18
e) Trisomy 21

140. A newborn appear with the midline cleft lip and cleft palate, microcephaly, hypotelorism, microopthalmos, bulbous nose, and midline scalp defect. The most likely diagnosis is:
a) Trisomy 18
b) Trisomy 13
c) Trisomy 9
d) Trisomy 16
e) Trisomy 21

141. All of the following syndromes have chromosomal breakage **except**:
a) Klinefelter syndrome
b) Fanconi anemia
c) Bloom syndrome
d) Werner syndrome
e) Nijmegen syndrome

142. The most common heritable form of mental retardation in males is:
a) Down syndrome
b) Williams syndrome
c) Klinefelter syndrome
d) Turner syndrome
e) Fragile X syndrome

Match the following syndromes with the clinical manifestations (143-147):

143. Williams syndrome
144. Prader-Willi syndrome
145. Alagille syndrome
146. Rubinstein-Taybi syndrome
147. Angelman syndrome

a) Bile duct paucity with cholestasis
b) Uncontrollable bouts of laughter and seizures
c) Hypotonia, hypogonadism, and obesity
d) Supravalvular aortic stenosis
e) Broad thumbs and toes

Match the following syndromes with the clinical manifestations (148-152):

148. WAGR syndrome
149. Smith-Magenis syndrome
150. Alagille syndrome
151. Miller-Dieker syndrome
152. Velocardiofacial-DiGeorge syndrome

a) Brachycephaly
b) Aniridia

c) Conotruncal cardiac defects
d) Pulmonary artery stenosis
e) Microcephaly

153. A 2-day-old male newborn infant appears with vomiting, tachypnea, lethargy, and convulsion. He was born by NSVD with Apgar scores of 9 and 9 at 1 and 5 minutes respectively. A physical examination reveals mild hepatosplenomegaly. A sepsis work up is performed. The boy is receiving ampicillin, gentamicin, and phenobarbital. The laboratory studies reveal normal arterial blood gas and serum electrolytes. The serum ammonia level is 1,050 μM. All of the following enzyme deficiencies can produce hyperammonemia **except**:
a) Carbamyl phosphate synthetase
b) Glycine cleavage enzyme
c) Ornithine transcarbamylase
d) Arginosuccinate synthetase
e) N-Acetylglutamate synthetase

154. The therapy for hyperammonemia in infants includes all of the following **except**:
a) High protein therapy intravenously.
b) Give adequate calories, fluid, and electrolytes intravenously.
c) Sodium benzoate
d) Sodium phenylacetate
e) Arginine hydrochloride

155. A 3-day-old preterm female infant appears sudden onset of respiratory distress. Her condition worsens despite antibiotics (ampicillin and gentamicin) and ventilator therapies. The blood tests reveal elevated citrulline, alanine, glutamine, and ammonia (3,500 μM) levels. All of the following statements are true about this condition **except**:
a) The cause is carbamyl phosphate synthetase deficiency.
b) The therapy for hyperammonemia should be started as soon as possible.
c) The patient recovers without sequelae.
d) Hyperammonemia does not recur with normal protein diet.
e) The therapy for hyperammonemia should be continued vigorously.

156. A male child appears with night blindness, icthyosis, and peripheral neuropathy. He has Refsum disease. The preferred therapy is:
a) Phytanic acid-restricted diet
b) Carboxylic acid-restricted diet
c) Pristanic acid-restricted diet
d) Citric acid-restricted diet
e) Bile acid-enriched diet

157. The enzyme deficiency in patients with classic Refsum disease is:
a) Acyl-CoA oxidase
b) 2-Methyl acyl CoA oxidase
c) Phytanoyl-CoA
d) D-bifunctional protein
e) Very long chain fatty acids

158. At birth, the eyes of all newborn infants should be protected against gonococcal infection by instilling:
a) 0.5% erythromycin ointment
b) 1% erythromycin ointment
c) 0.5% silver nitrate drops
d) 2.5% povidone-iodine solution
e) 1% silver nitrate drops

159. Apgar evaluations of newborn infants include all of the following **except**:
a) Color
b) Heart rate
c) Muscle strength
d) Respiratory effort
e) Response to catheter in nostril tested after oropharynx is clear.

160. A full term female newborn is delivered by NSVD. Her heart rate 110 per minute. She is pale and blue. She is limp. She has no respiratory effort. She has no response to catheter in nostril (tested after oropharynx is clear). Apgar score is:
   a) 1
   b) 2
   c) 3
   d) 4
   e) 5

161. All of the following conditions can give false-positive findings in Apgar evaluations **except**:
   a) Magnesium sulfate
   b) Immaturity
   c) Maternal acidosis
   d) Acute cerebral injury
   e) Morphine

162. All of the following conditions reduce the effects of maternal drugs on fetus **except**:
   a) Drugs composed large molecules
   b) Drugs that are strong acids
   c) Drugs that are plasma bound
   d) Poorly absorbed from maternal GI tract
   e) Poorly absorbed from newborns GI tract

163. Breast feedings are contraindicated in the USA in all of the following conditions **except**:
   a) HIV
   b) HTLV-1
   c) HTLV-2
   d) CMV (preterm infants)
   e) CMV (term infants)

164. All of the following organisms are present in the breast milk and therefore, cause infections to newborn infants **except**:
   a) CMV
   b) HTLV-1
   c) HTLV-2
   d) HIV
   e) Mycobacterium tuberculosis

165. All of the following drugs are contraindicated in breast feeding **except**:
   a) Aldomet
   b) Nicotine
   c) Cocaine
   d) Heroin
   e) Cimetidine

166. The following category drugs are contraindicated during pregnancy:
   a) Category A
   b) Category B
   c) Category C
   d) Category D
   e) Category X

Match the following drugs and their teratogenic effects (167-171):

167. Phenytoin
168. Warfarin
169. Propylthiouracil
170. Accutane
171. Lithium

a) Hypothyroidism
b) Hypoplastic nasal structures
c) Neuroblastoma
d) Facial-ear defects
e) Ebstein's anomaly

Match the followings drugs and their mechanism of teratogenic effects (172-173):

172. Warfarin
173. Phenytoin

a) Prevents the carboxylation of gamma-carboxyglutamic acid
b) Deficiency of epoxide hydrolase

Match the following fetal conditions and fetal therapies (174-178):

174. 21-hydroxylase deficiency
175. Bartter syndrome
176. Isoimmune thrombocytopenia
177. Supraventricular tachycardia
178. Methylmalonic acidemia

a) Indomethacin to the mother
b) Intravenous immunoglobulin to the mother
c) Flecainide to the mother
d) Vitamin B_{12}
e) Dexamethasone to the mother

Match the following drugs and their side effects (179-183):

179. Sulfisoxazole
180. Furosemide
181. Erythromycin
182. Indomethacin
183. Fentanyl

a) Nephrocalcinosis
b) Kernicterus
c) Intestinal perforation
d) Seizures
e) Pyloric stenosis

Match the following congenital anomalies and clinical manifestations (184-188):

184. Choanal atresia
185. Potter syndrome
186. VATER syndrome
187. Pierre Robin syndrome
188. Diaphragmatic hernia

a) Aspiration pneumonia
b) Regurgitation of milk through the nose
c) Scaphoid abdomen
d) Unable to pass urine
e) Baby is blue when stops crying and becomes pink when starts crying.

189. The most common site of spinal cord injury in newborns with cephalic presentation is:
a) C1
b) C2

c) C3
d) C4
e) C7

190. The most common sites of spinal cord injuries in newborns with breech presentation are:
a) C1-C2
b) C3-C4
c) C5-C6
d) C7-T1
e) T2-T3

191. A newborn appears with asymmetric facial features. The right eye cannot be closed, right side of the forehead is smooth, right nasolabial fold is absent, and right corner of the mouth drops. He was born by forcep-assisted vaginal delivery. The most likely diagnosis is:
a) Absence of depressor anguli oris muscle
b) Nuclear agenesis of the facial nerve
c) Left facial nerve palsy
d) Right sixth cranial nerve palsy
e) Right peripheral facial nerve palsy

Match the intrauterine infections with the following findings (192-196):

192. CMV
193. Toxoplasmosis
194. Herpes simplex virus
195. Rubella
196. Syphilis

a) Cataracts
b) Ganciclovir therapy may be effective
c) Life-threatening illness
d) Organism is not resistant to penicillin therapy.
e) Transmitted through cat feces or undercooked meat or raw vegetables

197. A 4-day-old full term infant appears with poor feeding, lethargy, pallor, jaundice, tachypnea, and tachycardia. She was born by vaginal delivery with breech presentation. Her Apgar scores were 2 and 8 at 1 and 5 minutes respectively. She is A(+), Coombs (-ve). Her mother is O(+). A physical examination reveals bluish discoloration and palpable mass in right upper quadrant. The most likely diagnosis is:
a) Hemolytic jaundice (AO set up)
b) Congenital spherocytosis
c) G6PD deficiency
d) Ruptured duodenum
e) Subcapsular hematoma of the liver

198. A 29-week-gestational age preterm infant appears with cessation of breathing for longer than 20 seconds, cyanosis, and sinus bradycardia. The initial therapy includes all of the following **except**:
a) Bag and mask ventilation
b) Oxygen
c) Aminophylline
d) Gentle cutaneous stimulation
e) Antireflux medication

199. A 10-day-old male infant appears with kernicterus. All of the following clinical manifestations are present **except**:
a) Hypotonia
b) Stupor
c) Poor suck
d) Seizures
e) Opisthotonos

200. A 2-week-old female infant appears with jaundice. She was born in another institution. She is A(+)ve and Coombs(-)ve. Her mother is O(+)ve. The differential diagnosis includes all of the following **except**:
   a) Biliary atresia
   b) Neonatal hepatitis
   c) Galactosemia
   d) Hypothyroidism
   e) Hemolytic jaundice due to 'AO' incompatibility

201. According to American Academy of Pediatrics all of the following conditions are preventable causes of kernicterus **except**:
   a) Lack of parenteral concern regarding the presence of jaundice.
   b) Early discharge (less than 48 hours) was done but no early follow-up (within 48 hours of discharge) was arranged.
   c) Failure to obtain serum bilirubin levels in newborns who appear jaundice during the first 24 hours.
   d) Lack of clinical experience to recognize the jaundice.
   e) Lack of concern regarding the presence of jaundice in newborn infants.

202. A 16-hour-old male newborn appears with jaundice. His serum total bilirubin level is 18 mg/dL. His blood type is B(+)ve and Coombs positive. His mother's blood type is O(+)ve. The double phototherapy is started. Double volume exchange transfusion should be initiated when the serum indirect bilirubin level is:
   a) 19 mg/dL
   b) 20 mg/dL
   c) 22 mg/dL
   d) 24 mg/dL
   e) 25 mg/dL

203. Double volume exchange transfusions are performed in patients with severe hyperbilirubinemia secondary to hemolytic disease. The composition of blood used in this procedure should be about:
   a) 15% packed-RBCs and 85% plasma
   b) 25% packed-RBCs and 75% plasma
   c) 33% packed-RBCs and 67% plasma
   d) 50% packed-RBCs and 50% plasma
   e) 66% packed-RBCs and 34% plasma

204. During phototherapy, photo-isomerization predominantly converts bilirubin:
   a) From 4Z, 15E to 4Z, 15Z
   b) From 4Z, 15Z to 4Z, 15E
   c) From 4E, 15Z to 4Z, 15Z
   d) From 4E, 15E to 4Z, 15E
   e) From 4E, 15Z to 4E, 15Z

205. All of the following are common complications of phototherapy **except**:
   a) Loose stools
   b) Overheating
   c) Erythematous maculopapular rash
   d) Eye injury
   e) Dehydration

206. All of the following conditions can cause hemolytic disease of the newborn (or erythroblastosis fetalis) **except**:
   a) D antigens of the Rh group
   b) ABO incompatibility
   c) C antigens of the RBCs
   d) E antigens of RBCs
   e) Anti-Lewis antibodies

207. The majority of the patients with ataxia-telangiectasia have the following clinical manifestation besides cerebellar ataxia and oculocutaneous telangiectasias:
   a) Adenocarcinomas
   b) Recurrent sinopulmonary infections
   c) Varicella

d) Otitis media
e) Lymphoreticular type malignancy

208. The most common type of malignancy in patients with ataxia-telangiectasia is:
a) Cerebellar tumor
b) Pituitary tumor
c) Hemangiosarcomas
d) Neurofibrosarcomas
e) Lymphoreticular type

209. The mode of inheritance in patients with Wiskott-Aldrich syndrome is:
a) Autosomal recessive
b) Autosomal dominant
c) X-linked recessive
d) X-linked dominant
e) Multifactorial

210. The laboratory findings in patients with Wiskott-Aldrich syndrome includes all of the following **except**:
a) Elevated serum level of IgM
b) Elevated serum level of IgA
c) Elevated serum level of IgE
d) Normal or slightly low IgG level
e) Moderately reduced percentages of T cells

211. The most common associated finding in patients with chronic asthma and/or pneumonia is:
a) Mental retardation
b) Cerebral palsy
c) Allergy
d) Tracheoesophageal fistula (H-type)
e) Gastroesophageal reflux

212. The most common cause of bronchiectasis in the developed countries is:
a) Cystic fibrosis
b) Ciliary dyskinesia
c) TB
d) Measles
e) Right middle lobe syndrome

213. The preferred diagnostic study in patients with bronchiectasis is:
a) Chest x-ray (AP view)
b) Bronchoscopy
c) Chest x-ray (lateral view)
d) Bronchography
e) CT scan

214. The most common combination of pathogens causing pneumonia is:
a) RSV and Mycoplasma pneumoniae
b) RSV and Chlamydia trachomatis
c) Streptococcus pneumonia and S. aureus
d) Streptococcus pneumonia and C. trachomatis
e) Streptococcus pneumonia and RSV

215. The most common causes of empyema are:
a) S. aureus and S. pneumoniae
b) S. aureus and Pseudomonas aeruginosa
c) S. pneumoniae and RSV
d) S. pneumoniae and Mycoplasma pneumoniae
e) S. pneumoniae and Chlamydia trachomatis

216. The top five most common clinical manifestations in patients with cystic fibrosis are the following **except**:
   a) Acute or persistent respiratory symptoms
   b) Failure to thrive (or malnutrition)
   c) Abnormal stools
   d) Meconium ileus
   e) Nasal polyps

217. The parents noticed salty taste when they kiss the child. The most likely diagnosis is:
   a) Facial eczema
   b) Acrodermatitis enteropathica
   c) SLE
   d) Cystic fibrosis
   e) Ectodermal dysplasia

218. The diagnostic criteria in patients with cystic fibrosis (CF) include all of the following **except**:
   a) Presence of typical clinical manifestations and two elevated sweat chloride concentrations (60 mEq/L or more) performed on separate days
   b) A history of cystic fibrosis in a sibling and identification of two CF mutations
   c) A positive newborn screening test and two elevated sweat chloride concentrations performed on separate days
   d) A history of CF in a sibling and an abnormal nasal potential difference measurement
   e) A history of CF in a sibling and positive newborn screening test

219. The following statement is **not** true in patients with cystic fibrosis:
   a) Sexual development is often delayed.
   b) Sexual function is usually impaired.
   c) Majority of males are azoospermic.
   d) Female fertility rate is reduced.
   e) Females may have cervicitis and accumulation of tenacious mucus in the cervical canal.

220. All of the following complications are due to use of aminoglycosides in patients with cystic fibrosis **except**:
   a) Hypomagnesemia
   b) Hyperuricemia
   c) Hearing loss
   d) Vestibular dysfunction
   e) Renal tubular dysfunction

221. A child has cystic fibrosis. The PFTs (pulmonary function tests) are not performed until:
   a) 1-2 years of age
   b) 3-4 years of age
   c) 5-6 years of age
   d) 7-8 years of age
   e) 9-10 years of age

222. The following statement is not true about pulmonary function tests in patients with cystic fibrosis:
   a) Restrictive lung changes occur early in the disease process.
   b) Obstructive lung changes typically present by 5-6 years of age.
   c) Small airway obstruction is manifested early by decrease in mid-maximal flow rate
   d) Residual volume and functional residual capacity are increased early in the course of the disease.
   e) Presence of an obstructive lung disease and modest response to bronchodilator are diagnostic of cystic fibrosis.

223. The presence of the following organisms on culture of lower airways or sputum is diagnostic of cystic fibrosis in children:
   a) Streptococcus pneumoniae
   b) Mucoid forms of Pseudomonas
   c) Mycoplasma pneumoniae
   d) RSV
   e) Chlamydia trachomatis

Match the following therapies and their complications in patients with cystic fibrosis (224-228):

224. Spironolactone
225. Corticosteroids
226. Ibuprofen
227. Semisynthetic penicillins
228. Iodine-containing expectorants

a) Interstitial nephritis
b) Gynecomastia
c) Goiter
d) Gastrointestinal bleeding
e) Hyperglycemia

Match the following syndromes and different cardiac defects (229-233):

229. Asplenia syndrome
230. Polysplenia syndrome
231. Mulibery nanism
232. Fragile X syndrome
233. PHACE syndrome

a) Pericardial thickening
b) Mitral valve prolapse
c) VSD
d) Increased pulmonary blood flow
e) Complex cyanotic heart lesions with decreased pulmonary blood flow

Match the following syndromes and chromosomal defects (234-238):

234. Williams syndrome
235. Alagille syndrome
236. Noonan syndrome
237. Marfan syndrome
238. Char syndrome

a) Chromosome 6
b) Chromosome 7
c) Chromosome 12
d) Chromosome 15
e) Chromosome 20

239. A 3-day-old male newborn appears with heart murmur. Oxygen saturation values are the following: right atrium (72%), right ventricle (83%), left atrium (100%), left ventricle (100%), pulmonary artery (87%), and aorta (100%). Blood flow volume (L/minute/m^2) through the heart and blood vessels is the following: right atrium (3), right ventricle (5), left atrium (6), left ventricle (6), pulmonary artery (5), and aorta (4). The most likely diagnosis is:
a) Tetralogy of Fallot
b) VSD
c) Pulmonary stenosis
d) Aortic stenosis
e) Transposition of great arteries

240. A child had surgery for right ventricular outflow tract obstruction. The expected postoperative complication is:
a) Pulmonic stenosis
b) Tricuspid regurgitation
c) Pulmonary valvular insufficiency
d) Tricuspid stenosis
e) Rupture interventricular septum

241. A child has congenital absence of the pulmonary valve. The most common associated anomaly is:
   a) ASD
   b) VSD
   c) Aortic stenosis
   d) Pulmonic stenosis
   e) Congenital absence of tricuspid valve

Match the different cardiac conditions with different types of murmurs (242-246):

242. Coarctation of the aorta
243. Mitral valve prolapse
244. Pulmonary valvular insufficiency
245. Congenital mitral insufficiency
246. Congenital mitral stenosis

   a) A low-pitched decrescendo diastolic murmur at the upper and midleft sternal border
   b) A short systolic murmur is noted along the left sternal border at the 3rd and 4th intercostal spaces; murmur is transmitted to the left infrascapular area.
   c) Apical murmur is late systolic and associated with a click.
   d) A high-pitched ("cooing dove"), apical holosystolic murmur
   e) A rumbling apical diastolic murmur is followed by loud 1st sound; 2nd sound is loud and split.

247. A child has sickle cell disease with hereditary persistence of fetal hemoglobin. The following complication may occur:
   a) Pulmonary hypertension
   b) Gross hematuria
   c) Avascular necrosis of the hips
   d) Pituitary infarction
   e) Intracranial hemorrhage

248. The most common organism responsible for post-splenectomy sepsis is:
   a) Haemophilus influenzae
   b) Neisseria meningitides
   c) Plasmodium falciparum
   d) Babesia microti
   e) Streptococcus pneumoniae

249. All of the following conditions can cause functional hyposplenism **except**:
   a) Congenital polysplenia
   b) Premature infants
   c) Malaria
   d) Severe hemolytic anemias
   e) Irradiation to the left upper quadrant

250. Splenic hypofunction is characterized by:
   a) Platelet inclusions in smear
   b) Thrombocytopenia
   c) Thrombocytosis
   d) Large size platelets
   e) RBC inclusions in peripheral blood smears

251. All of the following preoperative, intraoperative, and postoperative management can reduce the risk of infection in post-splenectomy patients **except**:
   a) It is better to postpone the operation until 4 years of age.
   b) Pneumococcal, meningococcal, and H. influenzae vaccines should be given before splenectomy.
   c) Laparascopic splenectomy has reduced the morbidity and hospitalization time after surgery.
   d) After surgical splenosis (spreading small pieces of spleen throughout the abdomen) or spontaneous splenosis, the splenic tissue that regrows usually has inadequate function.
   e) Oral prophylactic penicillin therapy should be provided for at least 2 years after splenectomy or 6 years of age.

Match the following diseases with the tumors (252-256):

252. Peutz-Jeghers syndrome
253. Neurofibromatosis 2
254. Tuberous sclerosis
255. Rendu-Osler-Weber syndrome
256. von Hippel-Lindau disease

a) Angioma
b) Gastrointestinal cancer
c) Myocardial rhabdomyoma
d) Hemangioblastoma of the cerebellum
e) Bilateral acoustic neuromas

Match the following syndromes with the tumors (257-261):

257. Wermer syndrome
258. Sipple syndrome
259. Bloom syndrome
260. Klinefelter syndrome
261. Noonan syndrome

a) Parathyroid adenoma
b) Medullary carcinoma of the thyroid
c) Breast cancer
d) Schwannoma
e) Leukemia

262. A male child has vesicoureteral reflux. He has a female sibling who is asymptomatic. The incidence of reflux in siblings of children with reflux is:
a) 10%
b) 15%
c) 25%
d) 35%
e) 25%

263. The incidence of renal scarring in asymptomatic siblings with vesicoureteral reflux is:
a) 1%
b) 6%
c) 12%
d) 15%
e) 25%

264. A 20-year-old mother appears with of vesicoureteral reflux. She delivered a newborn male infant. The incidence of reflux in an offspring when mother appears with a history of reflux is:
a) 10%
b) 15%
c) 25%
d) 35%
e) 50%

265. The most common obstructive lesion in urinary tract is:
a) Prune-belly syndrome
b) Posterior urethral valve
c) Urethral stricture
d) Ureterovesicle junction obstruction
e) Ureteropelvic junction obstruction

266. The most common cause of UPJ obstruction is:
   a) Intrinsic
   b) Abnormal rein veins causing obstruction
   c) Abnormal renal artery causing obstruction
   d) Lymph nodes causing pressure on the ureter
   e) Mesenteric band causing pressure on the ureter

267. All of the following are usual presentations in patients with UPJ obstruction **except**:
   a) Hypertension
   b) Prenatal maternal ultrasonographic findings reveal fetal UPJ obstruction
   c) A palpable renal mass during the routine physical examination.
   d) Pain in the flank or abdomen
   e) A febrile urinary tract infection

268. A girl has ureterocele. The most common associated finding is:
   a) Single ureter
   b) Ureteral duplication
   c) Small urinary bladder
   d) Single kidney
   e) Hypertrophied urinary bladder

269. A boy has ureterocele. The most common associated finding is:
   a) Horseshoe kidney
   b) Single ureter
   c) Multicystic kidney
   d) Posterior urethral valve
   e) Thickened urinary bladder

270. The time to repair in children with epispadias is:
   a) 1-5 months of age
   b) 6-12 months of age
   c) 13-18 months of age
   d) 19-24 months of age
   e) 25-36 months of age

271. The most common tumor occurs in undescended testes is:
   a) Embryonal carcinoma
   b) Choriocarcinoma
   c) Yolk sac carcinoma
   d) Sertoli-Leydig cell tumor
   e) Seminoma

272. Hepatocellular carcinoma can occur with all of the following conditions **except**:
   a) Hepatitis B infection
   b) Biliary cirrhosis
   c) Hepatitis A infection
   d) Glycogen storage disease
   e) Alpha 1-antitrypsin deficiency

273. The most common site of metastasis in patients with hepatoblastoma is:
   a) CNS
   b) Lungs
   c) Spleen
   d) Bones
   e) Small intestine

274. A full term newborn has left undescended testis. The mother is worried. The physician should tell the mother:
   a) The testis will descend by 1 month of age.
   b) The testis will descend by 2 months of age.

c) The testis will descend by 3 months of age.
d) If the testis does not descend by 5 months of age, it will remain undescended.
e) If the testis does not descend by 6 months of age, it will remain undescended.

275. A newborn has bilateral undescended testes. The physician should suspect the following diagnosis:
a) Seminoma of the testes
b) Testicular torsion in utero
c) Prader-Willi syndrome
d) Klinefelter syndrome
e) Virilized female with congenital adrenal hyperplasia

276. The orchiopexy is performed for an undescended testis between:
a) 1-3 months of age
b) 3-6 months of age
c) 6-9 months of age
d) 9-15 months of age
e) 15-24 months of age

277. The most common associated finding in patients with an undescended testis is:
a) Malignancy
b) Teratoma
c) Testicular torsion
d) Direct inguinal hernia
e) Indirect inguinal hernia

278. A 5-year-old boy appears for a routine physical examination. His right testis is not in the scrotum. The physician is able to bring the testis down into the scrotum. He had normal physical examination at birth. The most likely diagnosis is:
a) Retractile testis
b) Inguinal hernia
c) Testicular torsion
d) Cryptorchid testis
e) 'Acquired' or ascending undescended testis

Match the following different conditions of the testis and underlying mechanisms (279-282):

279. Undescended testis
280. Retractile testis
281. 'Acquired' or ascending undescended testis
282. Vanishing testis

a) Incomplete disappearance of processus vaginalis
b) Brisk cremasteric reflex
c) Failure of hormonal and/or mechanical factors
d) Testicular torsion in utero

283. A 12-year-old boy appears with sudden onset of pain in the scrotum for the last 2 hours. He denies history of trauma but he was playing soccer. A physical examination reveals swollen and tender right testis. The cremasteric reflex is absent. The preferred therapy is:
a) Prompt surgical exploration, detorsion of the testis, and scrotal orchiopexy
b) Surgical exploration, detorsion of the testis, and orchiopexy after 24 hours
c) Cold compress locally and observation for 24 hours
d) Hot compress locally and analgesic therapy
e) Bed rest; NSAIDs for 5 days

284. A 9-year-old boy appears with painless paratesticular mass on the right side. The mass is described as 'bag of worms'. The following investigation should be performed:
a) Testicular scan
b) Urine culture
c) Doppler ultrasonography

d) Ultrasonography of the testis
e) Ultrasonography of the abdomen

285. A female patient appears with intermittent sharp right lower abdominal pain for the last 24 hours. The pain radiates to the right extremity. She has no fever. She denies history of trauma. She has irregular menstrual cycles for the last 1 year. The preferred therapy is:
a) Appendectomy
b) Aspiration of right hip joint
c) Cefuroxime
d) Laxative therapy
e) Laparascopic detorsion of the right ovary

286. The triad of problems in female athletes are:
a) Amenorrhea, disordered eating, and using steroids
b) Dysmenorrhea, anorexia, and hyperthyroidism
c) Dysmenorrhea, anorexia nervosa, and hypothyroidism
d) Amenorrhea, disordered eating, and ovarian failure
e) Amenorrhea, disordered eating, and osteoporosis

287. The most common associated anomaly in patients with Mayer-Rokitansky-Kuster-Hauser syndrome is:
a) Renal
b) Liver
c) Intestinal
d) CNS
e) Cardiac

288. All of the following are manifestations of Mayer-Rokitansky-Kuster-Hauser syndrome **except**:
a) Primary amenorrhea
b) Normal vagina
c) Normal ovaries
d) Attenuated fallopian tubes
e) Duplication anomaly of the uterus

289. The mode of inheritance in patients with congenital absence of vaginal (Mayer-Rokitansky-Kuster-Hauser syndrome) is:
a) Autosomal recessive
b) Autosomal dominant
c) X-linked recessive
d) X-linked dominant
e) Multifactorial/polygenic

290. Wolfram syndrome includes all of the following manifestations **except**:
a) Diabetes mellitus
b) Nephrogenic diabetes insipidus
c) Optic atrophy
d) Deafness
e) Central diabetes insipidus

Match the following diseases with the different parameters (291-293):

291. Syndrome of inappropriate antidiuretic hormone secretion
292. Central diabetes insipidus
293. Cerebral salt wasting

a) Low (serum sodium, intravascular volume, vasopressin levels); high (urine output); very high (urine sodium); normal or high (serum uric acid levels)
b) Low (serum sodium, serum uric acid); high (urine sodium, vasopressin level); normal or high (intravascular volume); normal or low (urine output)
c) Low (urine sodium, intravascular volume, vasopressin level); high (serum sodium, urine output, serum uric acid level)

294. All of the following conditions cause fetal overgrowth **except**:
   a) Sotos syndrome
   b) Weaver syndrome
   c) Maternal diabetes mellitus
   d) SHOX excess syndrome
   e) Beckwith-Wiedemann syndrome

295. All of the following conditions cause childhood tall stature **except**:
   a) Fragile X syndrome
   b) Marfan syndrome
   c) Klinefelter syndrome
   d) Testicular feminization syndrome
   e) McCune-Albright syndrome

296. A 12-year-old boy has tall stature. His predicted adult height is 77 inches or 196 cm. His both parents are tall. He does not like his tall stature. The preferred therapy is:
   a) Testosterone enanthate
   b) Ethinyl estradiol
   c) Anabolic steroid
   d) Estrogen and progesterone
   e) Reassurance

297. A 10-year-old girl has tall stature. Her predicted adult height is 70 inches or 178 cm. Her both parents are tall. She does not like her tall stature. The preferred therapy is:
   a) Ethinyl estradiol
   b) Estrogen and progesterone
   c) Growth hormone inhibiting factor
   d) Anabolic steroid
   e) Reassurance

298. All of the following are features of Sotos syndrome (cerebral gigantism) **except**:
   a) Large hands and feet
   b) Large, dolichocephalic head
   c) Abnormal EEG findings
   d) Elevated GH and IGF-I levels
   e) Clumsiness and awkward gait

299. The most common type of thyroid cancer is:
   a) Medullary carcinoma
   b) Follicular carcinoma
   c) Papillary carcinoma
   d) Undifferentiated carcinoma
   e) Mixed papillary-follicular carcinoma

300. All of the following clinical manifestations are present in infants and younger children with multiple endocrine neoplasia, type II B **except**:
   a) Neuromas
   b) Poor sucking
   c) Diarrhea
   d) Constipation
   e) Failure to thrive

301. A child appears with history of muscle cramps and pains for the last 1 month. He develops numbness, tingling, and stiffness of the hands and feet for the last 7 days. The most likely the child has the following finding:
   a) Elevated serum calcium level
   b) Low serum phosphorus level
   c) Elevated serum alkaline phosphatase level
   d) A short QT interval on the ECG
   e) A positive Chvostek or Trousseau sign

302. All of the following findings are complications of long-standing hypocalcemia **except**:
   a) Mental deficiency
   b) Cataracts
   c) Mucocutaneous candidiasis
   d) Dry and scaly skin
   e) Premature eruption of teeth

303. A child has hypoparathyroidism. However, the serum calcium level is normal. The most likely the child is also suffering from:
   a) Hyperparathyroidism
   b) Hyperthyroidism
   c) Hyperpituitarism
   d) Pseudohypoparathyroidism
   e) Addison disease

304. A child appears for a routine physical examination. He is asymptomatic. His serum calcium level is elevated, PTH level is normal, serum magnesium level is high normal, but the rate of calcium to creatinine clearance is decreased. He has two younger siblings. They are both asymptomatic. The preferred management is:
   a) Subtotal parathyroidectomy
   b) Total parathyroidectomy
   c) Remove malignant rhabdoid tumors of the kidney
   d) Remove rhabdomyosarcoma of the extremity
   e) Detection of a serum calcium level in siblings

305. The preferred therapy in patients with hypercalcemia secondary to sarcoidosis is:
   a) Furosemide
   b) Intravenous hydration
   c) High phosphate diet
   d) Vitamin D therapy
   e) Prednisone

306. All of the following conditions can cause hypercalcemia **except**:
   a) Prolong immobilization
   b) Hypophosphatasia
   c) Williams syndrome
   d) Albright hereditary osteodystrophy
   e) Jansen-type metaphyseal chondrodysplasia

307. A child has history of adrenal insufficiency. She receives oral cortisol therapy. She is in the operating room for appendectomy. She should receive the following therapy:
   a) Reduce the hydrocortisone during surgery
   b) Reduce the hydrocortisone dose before and during surgery
   c) Slightly increase the hydrocortisone dose during surgery
   d) Give fludrocortisone during surgery
   e) Give high intravenous dose of hydrocortisone

308. All of the following drugs can cause adrenal insufficiency **except**:
   a) Rifampin
   b) Ketoconazole
   c) Phenytoin
   d) Phenobarbital
   e) INH

309. The following serotype of group A beta-hemolytic streptococci can cause **epidemics of nephritis after throat infections** is:
   a) Serotype 1
   b) Serotype 6
   c) Serotype 12
   d) Serotype 49
   e) Serotype 55

310. The following serotype of group A beta-hemolytic streptococci (Streptococcus pyogenes) can cause **epidemics of nephritis after skin infections** is:
   a) Serotype 24
   b) Serotype 49
   c) Serotype 55
   d) Serotype 57
   e) Serotype 59

311. The most common complication of acute poststreptococcal glomerulonephritis is:
   a) Hypertension
   b) Seizures
   c) Cardiac failure
   d) Hyperkalemia
   e) Hyperphosphatemia

312. A 15-year-old girl has **comedonal acne**. The therapy **not** indicated is:
   a) Oral doxycycline
   b) Topical tretinoin
   c) Topical salicylic acid
   d) Topical azelaic acid
   e) Topical adapalene

313. A 16-year-old boy has **mild papulopustular acne**. The therapy **not** indicated is:
   a) Oral isotretinoin
   b) Topical benzoyl peroxide
   c) Oral doxycycline plus topical retinoid
   d) Topical benzoyl peroxide gel with clindamycin
   e) Topical benzoyl peroxide gel with erythromycin

314. A 17-year-old girl has severe papulopustular and nodular acne. She used benzoyl peroxide gel, oral doxycycline plus topical retinoid. The acne has not improved. The preferred therapy is:
   a) Oral isotretinoin
   b) Oral minocycline
   c) Oral minocycline plus topical retinoid
   d) Continue benzoyl peroxide gel
   e) Benzoyl peroxide gel with clindamycin

315. A child has congenital scoliosis. The following investigation should be performed in all children with congenital scoliosis:
   a) Echocardiogram
   b) Renal ultrasonography
   c) Head CT scan
   d) Pulmonary function tests
   e) Liver ultrasonography

316. The most common renal anomaly in patients with congenital scoliosis is:
   a) Unilateral renal agenesis
   b) Horseshoe kidney
   c) Pelvic kidney
   d) Polycystic kidney
   e) Duplication of the ureter

317. The most common associated anomaly in patients with congenital scoliosis is:
   a) Spinal dysraphism
   b) Unilateral renal agenesis
   c) Mitral valve prolapse
   d) Intestinal obstruction
   e) Biliary atresia

318. The most common type of spinal dysraphism in patients with congenital scoliosis is:
    a) Meningomyelocele
    b) Spina bifida occulta
    c) Intradural Lipoma
    d) Spinal cord cyst
    e) Spinal cord tethers

319. A 5-year-old girl complains of back pain for the last 7 days. She denies history of trauma. The most common cause of back pain in preadolescent children is:
    a) Organic
    b) Psychological
    c) Mechanical
    d) Abnormal posture
    e) Unknown

320. The preferred initial diagnostic procedure for patients with back pains is:
    a) CT scan
    b) MRI
    c) Bone scan
    d) ESR
    e) Plain x-ray of the spine

321. The most consistent presenting feature in patients with Marfan syndrome is:
    a) Positive Steinberg sign
    b) Positive wrist sign
    c) Echocardiography reveals aortic root dilatation
    d) Dislocated lens
    e) Tall stature with diminished upper segment/lower segment ratio

322. The patients with Marfan syndrome should be followed by all of the following pediatric subspecialists **except**:
    a) Cardiologist
    b) Opthalmologist
    c) Orthopedic surgeon
    d) Nephrologist
    e) Physical therapist

323. A pregnant mother with Marfan syndrome is receiving beta-adrenergic blocking agent. All of the following complications can occur in the newborn **except**:
    a) Hypothermia
    b) Hypoglycemia
    c) Bradycardia
    d) Hypotension
    e) Hypogonadism

324. The following statement is **not** true about genetic counseling of Marfan syndrome:
    a) Marfan syndrome is autosomal dominant.
    b) About 15-30% of cases are the first Marfan syndrome in their families.
    c) Fathers of sporadic Marfan syndrome cases are usually not older than fathers in the general population.
    d) There is minimal risks to the future offspring of normal parents.
    e) Each offspring of Marfan syndrome patients has 50% chance of inheriting the chromosome 15 with mutant FBN1 allele.

325. The **hematopoietic syndrome** (or clear hematologic depression) occurs after acute whole body irradiation from X-ray or Gamma rays above:
    a) 50 rads (0.5 Gy)
    b) 100 rads (1.0 Gy)
    c) 200 rads (2.0 Gy)
    d) 400 rads (4.0 Gy)
    e) 800 rads (8.0 Gy)

326. The **gastrointestinal syndrome** occurs after acute whole body irradiation from X-rays or Gamma rays above:
   a) 100 rads (1.0 Gy)
   b) 200 rads (2.0 Gy)
   c) 400 rads (4.0 Gy)
   d) 800 rads (8.0 Gy)
   e) 3,000 rads (30.0 Gy)

327. The symptoms (e.g., nausea and vomiting) begin in **some patients (about 10% of cases)** after acute whole body irradiation from X-rays or Gamma rays above:
   a) 5 rads (0.05 Gy)
   b) 10 rads (0.10 Gy)
   c) 15 rads (0.15 Gy)
   d) 50 rads (0.5 Gy)
   e) 100 rads (1.0 Gy)

328. A 12-year-old boy appears with persistent vomiting for the last 12 hours. He has history of recurrent episodes of abdominal pain with occasional episodes of vomiting and diarrhea for the last 9 years. He was seen by gastroenterologist several times. He visited emergency room on several occassions. The diagnosis of viral gastritis/enteritis/gastroenteritis were made in different visits. The parents denied history food allergy. He did not eat foods from restaurant recently. He is doing very well in school. His height and weight measurement are at the 10th percentile. He had no bowel surgery in the past. His birth history was unremarkable. He has no family history of a chronic intestinal disease. The physical examination reveals a soft but mildly tender mid-abdomin and epigastric regions. His bowel pattern is normal. The most likely diagnosis is:
   a) Psychologic vomiting
   b) Intussuception
   c) Hirschspring disease
   d) Recurrent appendicitis
   e) Malrotation with intermittent volvulus

329. The diagnosis of malrotation can be made by all of the following findings **except**:
   a) The abdominal plain film reveals double-bubble sign.
   b) The barium enema reveals malposition of the cecum.
   c) The upper gastrointestinal series reveals malposition of the ligament of Treitz.
   d) The ultrasonography reveals superior mesenteric vein located to the left of the superior mesenteric artery.
   e) The ultrasonography reveals superior mesenteric vein located to the right of the superior mesenteric artery.

330. The Jejunoileal atresia is most likely due to:
   a) Intrauterine nerve injury
   b) Intrauterine muscular injury
   c) Intrauterine vascular accidents
   d) Failure to recanalize the lumen
   e) Intrauterine epithelial injury

331. The duodenal atresia occurs most likely due to:
   a) Failure to recanalize the lumen in utero
   b) Intrauterine epithelial injury
   c) Intrauterine muscular injury
   d) Intrauterine mesenteric injury
   e) Intrauterine vascular accident

332. All of the following congenital anomalies are associated with duodenal atresia **except**:
   a) Down syndrome
   b) Malrotation
   c) Esophageal atresia
   d) Congenital heart disease
   e) Jejunal atresia

Match the following diseases with the different findings (333-337):

333. Malrotation
334. Pyloric stenosis
335. Duodenal atresia
336. Jejunal atresia
337. Hirschsprung disease

a) Double-bubble sign
b) Ladd band
c) Hypochloremic metabolic alkalosis
d) Absence of ganglion cell
e) Triple-bubble sign

338. The most common cause of lower intestinal obstruction in neonates is:
a) Imperforate anus
b) Hypoplastic left colon
c) Hypoplastic right colon
d) Hirschsprung disease
e) Hypoplastic transverse colon

339. Hirschsprung disease is associated with all of the following conditions **except**:
a) Down syndrome
b) Waardenburg syndrome
c) Cartilage-hair hypoplasia
d) Turner syndrome
e) Smith-Lemli-Opitz syndrome

340. In older patients with Hirschsprung disease, the Currarino triad may be present. **The Currarino triad** includes all of the following **except**:
a) Ectopic anus, hypoplastic sacral bones, and presacral anterior meningocele
b) Rectal stenosis, poor segmentation of sacral bones, and presacral teratoma
c) Ectopic anus, poor segmentation of sacral bones, and presacral cyst
d) Rectal stenosis, hypoplastic sacral bones, and presacral anterior meningocele
e) Ectopic anus, hypoplastic sacral bones, and posterior meningomyelocele

341. All of the following features can distinguish Hirschsprung disease from functional (or acquired) constipation **except**:
a) Anal tone
b) Age of onset
c) Enterocolitis
d) Malnutrition
e) Rectal biopsy findings

342. All of the following laboratory findings are characteristics of Hirschsprung disease **except**:
a) Rectal biopsy reveals absence of ganglion cells.
b) Rectal biopsy reveals decreased acetylcholinesterase staining.
c) Barium enema reveals a transition zone and delayed evacuation (more than 24 hours).
d) Anorectal manometry reveals absence of internal sphincter relaxation with rectal distension.
e) Anorectal manometry reveals a paradoxical increase in internal sphincter pressure with rectal distension.

343. A 4-year-old girl appears with abdominal pain, nausea, vomiting, and low grade fever for the last 6 hours. The abdominal pain started before the nausea, vomiting, and fever. She denies history of trauma. The physical examination reveals flat abdomen, periumbilical tenderness, and normal bowel sounds. The most likely diagnosis is:
a) Acute gastritis
b) Acute gastroenteritis
c) Hemolytic-uremic syndrome
d) Food allergy
e) Acute appendicitis

344. All of the following laboratory findings suggest acute appendicitis **except**:
   a) Presence of 31 WBCs per high-power field in urine.
   b) Leukocytosis
   c) Presence of 20 RBCs per high-power field in urine
   d) A shift in differential diagnosis of WBCs
   e) Mild proteinuria

345. A male newborn infant is diagnosed to have rectovesical fistula. The most likely associated anomaly is:
   a) Cardiac
   b) CNS
   c) Hepatic
   d) Musculoskeletal
   e) Urologic

346. A simple anoplasty is performed in the newborn with perianal fistula. The colostomy is not required. The following procedure is indicated 2 weeks after surgery:
   a) Barium enema
   b) Upper GI follow through
   c) Colostomy
   d) Anal dilatations
   e) Rectal manometry

347. All of the following therapies are not indicated in patients with anal fissure **except**:
   a) Stretching of the anus
   b) Excision of the fissure
   c) Anal dilatations
   d) Stool softener
   e) 'Internal' anal sphincterotomy

348. The most common cause of rectal prolapse is:
   a) Chronic constipation
   b) Cystic fibrosis
   c) Pertussis
   d) Diarrhea
   e) Idiopathic

349. A 22-month-old male child appears with low-grade fever, perianal redness, and painful defecation for the last 2 days. He was completely asymptomatic prior to this episode. A physical examination reveals perianal cellulitis and rectal tenderness. The diagnosis of perianal abscess is made. The preferred therapy is:
   a) Incision and drainage
   b) Excision and drainage
   c) Vancomycin
   d) Gentamicin
   e) No therapy is indicated

350. A mother is pregnant. She has one sibling with congenital adrenal hyperplasia (CAH). The earliest preferred method to diagnose the fetus with CAH is:
   a) Amniocentesis
   b) Elevated 17-hydroxyprogesterone levels in the amniotic fluid.
   c) Elevated 17-hydroxyprogesterone levels in the fetal blood.
   d) Elevated ACTH levels in the fetal blood.
   e) DNA analysis of specimens obtained by chorionic villus sampling

351. The most important reason to make early prenatal diagnosis is:
   a) To relieve parental anxiety
   b) To monitor the fetal growth by fetal ultrasonography
   c) To look for an ambiguous genitalia by fetal ultrasonography
   d) To facilitate appropriate prenatal therapy of the affected male fetus
   e) To facilitate appropriate prenatal therapy of the affected female fetus

352. The preferred prenatal therapy for the fetus with congenital adrenal hyperplasia (CAH) is:
   a) Administration of dexamethasone directly into the fetal umbilical venous blood in utero.
   b) Administration of dexamethasone directly into the fetal umbilical arterial blood in utero.
   c) Administration of dexamethasone directly into the amniotic fluid in utero.
   d) Administration of dexamethasone to the mother (20 micrograms/kg prepregnancy maternal weight daily in two or three divided doses) of an affected fetus.
   e) Administration of dexamethasone to the mother (40 micrograms/kg pregnancy maternal weight daily in two or three divided doses) of an affected fetus.

353. The goal of early prenatal therapy in fetus with congenital adrenal hyperplasia is:
   a) To prevent the virilization of the external genitalia of the male fetus.
   b) To prevent the virilization of the external genitalia of the female fetus.
   c) To prevent the electrolyte imbalance of the male fetus.
   d) To prevent the electrolyte imbalance of the female fetus.
   e) To prevent the adrenal atrophy of the female fetus

354. The dexamethasone therapy to the mother of an affected fetus with CAH should be started to prevent the virilization of the external genitalia of the female fetus by:
   a) 6 weeks of age
   b) 8 weeks of age
   c) 10 weeks of age
   d) 12 weeks of age
   e) 15 weeks of age

355. The specific side effect on fetus when mother was treated with dexamethasone is:
   a) Adrenal crisis
   b) VSD
   c) Hyperglycemia
   d) Cleft lip
   e) Unknown

356. All of the following statements are true about the newborn screening in infants to diagnose congenital adrenal hyperplasia (CAH) **except**:
   a) The nonclassic form of CAH is detected reliably.
   b) The screening test analyze 17-hydroxyprogesterone levels in dried blood.
   c) The initial abnormal results should be confirmed by repeat serum 17-hydroxyprogesterone levels and electrolytes about 2 weeks of age.
   d) This test has high sensitivity and low specificity.
   e) This test has more problems in premature infants.

357. The preferred therapy in children with pituitary Cushing disease is:
   a) Transphenoidal pituitary microsurgery
   b) Pituitary irradiation
   c) Cyproheptadine
   d) Bilateral adrenalectomy
   e) Unilateral adrenalectomy

358. A 10-month-old girl appears with URI symptoms. A physical examination reveals runny nose, rounded face, prominent cheeks, flushed appearance, and mildly injected pharynx. The most likely underlying diagnosis is:
   a) Acute bacterial pharyngitis
   b) Pulmonic stenosis
   c) Acute tonsillitis
   d) Cushing syndrome
   e) Infant of a diabetic mother

359. All of the following are clinical manifestations in older children with Cushing syndrome **except**:
   a) Obesity of the face and trunk
   b) Short stature
   c) Hypertension

d) Osteoporosis
e) Advanced pubertal development

360. All of the following laboratory findings can make the diagnosis of Cushing syndrome **except**:
a) Cortisol levels in blood are elevated at midnight and 8 AM.
b) Nighttime salivary cortisol levels are decreased.
c) In a 24-hour-urine sample, an increased ratio of micrograms of cortisol excreted per gram of creatinine.
d) An abnormal glucose tolerance test despite elevated levels of insulin.
e) A single-dose dexamethasone suppression test (25-30 micrograms/kg, given at 11 PM) fails to decrease the plasma cortisol level of less than 5 micrograms/dL at 8 AM in the next morning.

361. The following serum electrolyte is abnormal in patients with Cushing syndrome, especially tumors with ectopic ACTH secretion:
a) Sodium
b) Potassium
c) Chloride
d) Bicarbonate
e) BUN and creatinine

Match all of the following findings (362-366):

362. Tall stature
363. Short stature
364. Elevated urinary excretion of cortisol
365. Elevated nighttime salivary levels of cortisol
366. Cortisol secretion is suppressed by oral administration of low doses of dexamethasone.

a) Simple obesity
b) Cushing syndrome
c) Both

Match all of the following findings (367-371):

367. Two-step dexamethasone suppression test (30 and 120 micrograms/kg/day in four divided doses, on consecutive days) does not suppress cortisol levels.
368. Two-step dexamethasone suppression test reveals that only larger dose suppresses cortisol levels.
369. ACTH concentrations are suppressed.
370. Intravenous corticotropin-releasing hormone (CRH) increases ACTH and cortisol levels.
371. Intravenous CRH does not increase ACTH and cortisol levels.

a) Pituitary Cushing syndrome (ACTH-dependent)
b) ACTH-independent Cushing syndrome

372. The preferred test to distinguish adolescents with secondary hypogonadism from adolescents with a delayed puberty is:
a) The gonadotropin response to stimulation with GnRH.
b) The gonadotropin response to stimulation with more potent analogue of GnRH
c) The use of GnRH analogue or the GnRH test after 36 hours of 'priming' of the hypothalamic-pituitary-gonadal axis with administration of pulsatile GnRH.
d) A single, 8 AM testosterone level is more than 0.7 mmol/L (20 mg/dL).
e) A single, 8 AM testosterone level is less than 0.7 mmol/L (20 mg/dL).

373. An adolescent male is diagnosed with secondary hypogonadotropic hypogonadism. He was treated with testosterone enanthate ester. The testes remain small. He also received hCG therapy. After 12 months of therapy, sufficient growth of the tests has not occurred. The following therapy may be added at this point:
a) Growth hormone
b) ACTH
c) Dehydrotestosterone
d) Prolactin
e) Human menopausal gonadotropin

374. All of the following manifestations are noted in patients with diabetes mellitus with infection **except**:
   a) Hypoglycemia
   b) Hyperglycemia
   c) Ketonuria
   d) Ketosis
   e) Overhydration

375. A child with diabetes mellitus becomes sick. She receives glargine and NPH insulin regularly. She should call her physician or nurse in all of the following conditions **except**:
   a) If the blood glucose remains elevated after two extra doses of insulin.
   b) If the blood glucose remains elevated after three extra doses of insulin.
   c) Signs of dehydration
   d) If the blood glucose less than 70 mg/dL and the child cannot take oral glucose.
   e) If the child has moderate to large ketones in each urine specimen despite extra doses of insulin and vomiting.

376. A child with diabetes mellitus developed large ketonuria and persistent vomiting. The nurse told him to go to the ER. The ER physician should perform all of the following tests **except**:
   a) Serum glucose
   b) Blood pH
   c) Serum electrolytes
   d) Total CO_2
   e) Serum insulin levels

377. A child with diabetes mellitus is going to have an elective surgery. She is asymptomatic. Her laboratory values are normal. Her insulin doses are adjusted appropriately. The following fluid therapy recommended during surgery is:
   a) 5% glucose, 0.45% saline solution with 10 mEq/L of potassium acetate is given at 1.0 time maintenance rate.
   b) 0% glucose, 0.9% saline solution with 20 mEq/L of potassium acetate is given at 1.5 times maintenance rate.
   c) 10% glucose, 0.9% saline solution with 40 mEq/L of potassium acetate is given at 1.5 times maintenance rate.
   d) 5% glucose, 0.3% saline solution with 20 mEq/L of potassium acetate is given at 1.5 times maintenance rate.
   e) 5% glucose, 0.45% saline solution with 20 mEq/L of potassium acetate is given at 1.5 times maintenance rate.

378. A child has diabetes mellitus and developed acute appendicitis. She received her insulin doses prior to surgery. During surgery, her blood glucose level is 180 mg/dL. the preferred therapy is:
   a) No insulin infusion is indicated.
   b) 0.03 units/kg/hour insulin infusion is indicated.
   c) 0.06 units/kg/hour insulin infusion is indicated.
   d) 0.08 units/kg/hour insulin infusion is indicated.
   e) 0.10 units/kg/hour insulin infusion is indicated.

379. All of the following statements are true regarding management of children with diabetes mellitus during perioperative period of surgery **except**:
   a) If glargine is used as the basal insulin, one half of the dose is given in the evening before surgery.
   b) If NPH or lente insulin is used, one half of the morning dose is given before surgery.
   c) An elective surgery should be performed early in the day.
   d) A full oral intake is expected shortly after surgery when the duration of surgery is less than 1 hour.
   e) The child should not be discharged until oral intake is tolerated and blood glucose levels are stable.

380. The Mauriac syndrome includes all of the following **except**:
   a) Moon face
   b) Enlarged liver
   c) Proximal muscle wasting
   d) Protuberant abdomen
   e) Over-insulinization

Match all of the following cranial nerve involvement and clinical manifestations (381-385):

381. Hypoglossal nerve (12th cranial nerve)
382. Vagus nerve (10th cranial nerve)
383. Glossopharyngeal nerve (9th cranial nerve)
384. Trigemial nerve (5th cranial nerve)
385. Accessory nerve (11th cranial nerve)

a) Paralysis of the sternomastoid and trapezius muscles
b) Absence of corneal reflex
c) Absence of gap response of the posterior pharyngeal wall
d) Presence of wasting, weakness, and fasciculations of the tongue
e) Asymmetry of the soft palate and hoarseness

Match all of the following cranial nerve involvement and clinical manifestations (386-390):

386. Oculomotor nerve (3rd cranial nerve)
387. Trochlear nerve (4th cranial nerve)
388. Abducens nerve (6th cranial nerve)
389. Trigeminal nerve (5th cranial nerve)
390. Vagus nerve (10th cranial nerve)

a) Nasal regurgitation of fluids, pooling of secretions, and immobile, low-lying soft palate
b) Absence of jaw jerk
c) Ptosis, dilatation of the pupil, displacement of the eye outward and downward, and impairment of adduction and elevation
d) Medial deviation of the eye and inability to abduct beyond the midline
e) Upward and outward deviation of the eye

391. All of the following statements are true about the average rate of head growth in healthy premature infants **except**:
a) 0.25 cm during the 1st week of life
b) 0.50 cm during the 2nd week of life
c) 0.75 cm during the 3rd week of life
d) 1.0 cm during the 4th week of life
e) 1.0 cm per week from the 5th week until the 40th week of development

392. The head circumference of full term infants at birth is about:
a) 32-33 cm
b) 33-34 cm
c) 34-35 cm
d) 35-36 cm
e) 36-37 cm

393. The head circumference of 6-month-old children who were born full term is about:
a) 40 cm
b) 44 cm
c) 47 cm
d) 50 cm
e) 52 cm

394. The head circumference of 12-month-old children who were born full term is about:
a) 45 cm
b) 46 cm
c) 47 cm
d) 48 cm
e) 49 cm

395. The most common cause of anosmia is:
a) Congenital
b) Hydrocephalus

c) Meningitis
d) Fracture of nasal bones
e) URI

396. All of the following conditions can produce loud cranial bruits **except**:
a) Hydrocephalus
b) Purulent meningitis
c) Intracranial tumor
d) Subdural effusions
e) A normal 2-year-old child

397. All of the following conditions can produce soft cranial bruits **except**:
a) Children with febrile illness
b) Murmurs arising from the heart
c) Severe anemia
d) Children younger than 4 years of age
e) Arteriovenous malformation of vein of Galen

398. A routine full-term newborn physical examination reveals bilateral retinal hemorrhages. He was born by NSVD with Apgar scores 9 and 9 at 1 and 5 minutes respectively. The rest of the physical examination is unremarkable. The next step in management is:
a) Opthalmology consultation
b) Head CT scan
c) Head ultrasonography
d) MRI of the eye
e) Reassurance

399. A routine full-term newborn physical examination reveals the size of the anterior fontanel is about 2 by 2 cm. The next step in management is:
a) Head ultrasonography
b) Head CT scan
c) Skull x-ray
d) Neurology consultation
e) Reassurance

400. The average time of closure of anterior fontanel is:
a) 9 months
b) 12 months
c) 15 months
d) 18 months
e) 24 months

401. The average time of closure of posterior fontanel is after the first:
a) 3-5 weeks of life
b) 6-8 weeks of life
c) 9-12 weeks of life
d) 13-15 weeks of life
e) 16-18 weeks of life

402. A 9-month-old girl appears for a routine physical examination. A physical examination reveals that the anterior fontanel is closed. The next step in management is:
a) Skull x-ray
b) Head CT scan
c) Head MRI
d) Neurosurgical consultation
e) Reassurance

Match the following clinical manifestations and different diagnosis (403-405):

403. Hydrocephalus
404. Microcephaly
405. Chronic Subdural hematomas

a) A full-term newborn with head circumference of 25 cm
b) Box-shaped skull
c) Broad forehead

406. Horner syndrome includes all of the following features **except**:
a) Miosis
b) Dilated pupil
c) Ptosis
d) Enopthalmos
e) Ipsilateral anhidrosis of the face

407. All of the following are contraindications for performing a lumber tap **except**:
a) Platelet count is 55,000
b) Brain tumor
c) Meningitis with flexion of upper extremities and extension of lower extremities
d) Skin infection at the site of lumber tap
e) Spinal cord tumor

408. The most common cause of elevated opening pressure in CSF is:
a) Bacterial meningitis
b) Viral meningitis
c) Tuberculous meningitis
d) Fungal meningitis
e) Crying constantly

409. The following problems have increased significantly due to the obesity epidemic:
a) Type I diabetes
b) Type II diabetes
c) Hypertension
d) Malignancies
e) Hypercholesterolemia

410. The most common complication of acute hypophosphatemia is:
a) Rhabdomyolysis
b) Acute respiratory failure
c) Slow weaning from the ventilator
d) Proximal muscle weakness
e) Proximal muscle atrophy

411. All of the following are manifestations of severe hypophosphatemia **except**:
a) Cardiac dysfunctions
b) Tremor
c) Seizures
d) Hemolysis
e) Leukocytosis

412. The appropriate metabolic compensation in patients with chronic respiratory alkalosis is:
a) Increased renal alkali excretion
b) Increased renal acid excretion
c) Decreased renal acid excretion
d) Decreased renal alkali excretion
e) Increased renal chloride excretion

413. All of the following statements are true about metabolic acidosis **except**:
   a) The normal respiratory response to metabolic acidosis is compensatory hyperventilation.
   b) When the serum pH is less than 7.20, there are impaired cardiac contractilities and increased risk of cardiac arrythmias.
   c) Acidemia causes potassium to move from the extracellular space to the intracellular space.
   d) Many patients with metabolic acidosis have low serum potassium level due to excessive body losses of potassium.
   e) Acidemia causes pulmonary vasoconstrictions, especially in newborn infants.

414. All of the following therapies are appropriate in patients with Batter syndrome and Gitelman syndrome **except**:
   a) Oral potassium supplementation is indicated in both the syndromes.
   b) Potassium-sparing diuretics are used in both the syndromes.
   c) Children with Gitelman syndrome often require magnesium supplementation.
   d) Children with Batter syndrome often need indomethacin.
   e) Children with Gitelman syndrome need calcium supplementation.

415. All of the following are characteristics of Bartter syndrome and Gitelman syndrome **except**:
   a) Hypokalemia
   b) Metabolic alkalosis
   c) Secondary hyperaldosteronism
   d) High blood pressure
   e) High urine chloride

416. The mode of inheritance in patients with Bartter syndrome or Gitelman syndrome is:
   a) Autosomal recessive
   b) Autosomal dominant
   c) X-linked recessive
   d) X-linked dominant
   e) Multifactorial

417. All of the following are clinical features in children with pseudohypoparathyroidism type 1A **except**:
   a) Short stature
   b) Round face
   c) Stocky build
   d) Brachydactyly
   e) Moderate mental retardation is noted in about 80% of cases.

418. The preferred diagnostic test in patients with pseudohypoparathyroidism is:
   a) Measurement of urinary cAMP after infusion of synthetic PTH
   b) Decreased production of PTH
   c) Decreased serum calcium levels
   d) Increased serum phosphorus levels
   e) Decreased serum 1, 25-dihydroxy vitamin D levels

419. A child appears with psychiatric symptoms such as confusion, disorientation, depression, psychosis, and hallucinations. The child is also hypotonic and lethargic. The laboratory test results most likely reveal:
   a) Low serum calcium levels
   b) High serum calcium levels
   c) Low serum magnesium levels
   d) High serum magnesium levels
   e) High serum phosphorus levels

420. The first line of therapy in patients with hypercalcemia is:
   a) Loop diuretic
   b) Calcitonin
   c) Glucocorticoids
   d) Biphosphonates
   e) Hydration with normal saline

421. The most common presentation in hospitalized patients with hypomagnesemia is:
   a) Asymptomatic
   b) Constipation
   c) Diarrhea
   d) Rickets
   e) Arrythmias

422. All of the following are manifestations of hypomagnesemia **except**:
   a) Tetany
   b) Tall T waves
   c) Positive Chvostek sign
   d) Positive Trousseau sign
   e) Seizures

Match the following syndromes with different conditions (423-424):

423. Gitelman syndrome
424. Bartter syndrome

   a) Hypocalciuria
   b) Hypercalciuria

425. The most common cause of transient hypomagnesemia in newborn is:
   a) Idiopathic
   b) IUGR
   c) Mother treated for preeclampsia
   d) Post exchange transfusions
   e) Infant of diabetic mother

426. All of the following medications can cause respiratory alkalosis **except**:
   a) Salicylate intoxication
   b) Theophylline
   c) Estrogen
   d) Progesterone
   e) Exogenous catecholamines

Match the all of the following conditions that cause metabolic alkalosis and their mechanism of actions (427-429):

427. Salicylates poisoning
428. Sepsis
429. Pain

   a) Cytokine mediated
   b) Psychogenic hyperventilation
   c) Directly stimulate the respiratory center.

430. The most common cause of all unintentional cause of death in children under 1 year of age is:
   a) Motor vehicle accident
   b) Suffocation
   c) Drowning
   d) Homicide
   e) Fire and burn

431. The most common cause of injury death in all children over 1 year of age is:
   a) Homicide
   b) Suicide
   c) Drowning
   d) Motor vehicle accidents
   e) Suffocation

432. All of the following are indications for hospitalization in burn patients **except**:
   a) Burn to hands, feet, genitals
   b) Inhalation injury regardless of the size of the burn
   c) High tension wire electrical burn
   d) Suspected child abuse or neglect
   e) Burns involve 9-10% of the body surface area

433. All of the following therapies are indicated in children with burns greater than 15% of body surface area **except**:
   a) Initially, oral fluid therapy is given before IV hydration.
   b) Lactated Ringer solution (10-20 mL/kg/hour) is given initially.
   c) Consultation with specialized burn unit is indicated.
   d) All inhalation injuries need IV access to control fluid intake.
   e) All high-tension and electrical injuries need IV access to provide forced alkaline diuresis to avoid myoglobinuric renal damage.

434. The pain management in patients with sickle cell anemia includes all of the following **except**:
   a) Generous dosing of opiods and NSAIDs at home are indicated.
   b) Treatment of breakthrough pain in a day-treatment program or clinic is preferred than in the ER or hospital admission.
   c) Self-care and maintenance of normal activities are indicated.
   d) Teaching of biobehavioral techniques and coping strategies before cycle develops involving positive experience with health care workers.
   e) The use of ketorolac may decrease the need for the total doses of opoids and avoid hospitalizations.

Match the following syndromes with chromosomes (435-436):

435. Prader-Willi syndrome
436. Angelman syndrome

   a) Maternally derived chromosome 15
   b) Paternally derived chromosome 15

Match the following inborn errors of amino acid metabolisms with abnormal urine odors (437-440):

437. Isovaleric acidemia
438. Maple syrup urine disease
439. Oasthouse urine disease
440. Trimethylaminuria

   a) Hops-like
   b) Rotting fish
   c) Maple syrup
   d) Sweaty feet, acrid

Match all of the skin manifestations in newborns with the diagnosis (441-444):

441. Mongolian spots
442. Lanugo
443. Erythema toxicum
444. Pustular melanosis

   a) Skin lesions appear at birth as vesiculopustular eruption on the neck, back, chin, palms, soles, and extremetries. Skin lesions contain neutrophils and last 2-3 days.
   b) Skin lesions appear 1-3 days after birth as small, white, occasionally vesicopustular papules on an erythematous base on the face, trunk, and extremities. Skin lesions contain eosinophils and last up to 1 week.
   c) Skin lesions appear at birth as slate blue, well demarcated areas of pigmentation on the buttocks and back. Skin lesions disappear within the first year.
   d) Fine, soft, immature hair on the scalp, brow, and face in premature infants.

Match all the clinical findings with different diagnosis (445-449):

445. Johanson-Blizzard syndrome
446. Spina bifida
447. Ehlers-Danlos syndrome
448. Amniotic band
449. Osteogenesis imperfecta

a) Excessive skin fragility
b) Face clefts
c) Aplasia cutis congenita
d) Wormian bones
e) Tufts of hair over the lumbosacral spine

450. A routine newborn physical examination reveals an enlarged scrotum. The transillumination is positive. The rest of the examination is unremarkable. The most likely diagnosis is:
a) Inguinal hernia
b) Femoral hernia
c) Hydrocele
d) Testicular torsion
e) Torsion of the spermatic cord

Match all the clinical findings with the different clinical conditions (451-453):

451. Sinus bradycardia in neonates
452. Sinus bradycardia after neonatal period
453. Supraventricular tachycardia in neonates

a) Heart rate is 231 beats/minute
b) Heart rate is less than 90 beats/minute.
c) Heart rate is less than 60 beats/minute.

454. A routine physical examination in a full-term newborn reveals heart rate 90 beats/minute during sleeping. The newborn is asymptomatic. The next step in management is:
a) Cardiology consultation
b) EKG
c) Sepsis work up
d) Chest x-ray
e) Reassurance

455. A routine physical examination of a newborn infant reveals heart rate 180 beats/minute during crying. The newborn is asymptomatic. The next step in management is:
a) EKG
b) Cardiology consultation
c) Chest x-ray
d) Sepsis work up
e) Reassurance

456. The most common cause of abdominal mass in newborn infants is:
a) Pancreatic cyst
b) Hydronephrosis
c) Adrenal hemorrhage
d) Hepatoblastoma
e) Intestinal duplication

457. All of the following conditions can cause renal vein thrombosis in infants **except**:
a) Dehydration
b) Asphyxia
c) Sepsis

d) Polycythemia
e) Increased level of antithrombin III

458. The Apgar evaluation in a newborn infant includes all of the following **except**:
a) Color
b) Respiratory effort
c) Heart rate
d) Reflex irritation
e) Eyes blinking

459. A newborn is intubated for respiratory distress. A physical examination reveals the decreased breath sounds on the right side of the chest wall. The most likely diagnosis is:
a) Left diaphragmatic hernia
b) Right pneumothorax
c) Left pneumothorax
d) Dextroxardia
e) Complete collapse of the right lung

460. A 14-day-old breast-fed infant appears with jaundice. His serum bilirubin level is 19 mg/dL. Mother is A(+)ve, baby is B(+)ve and Coombs (–)ve. The most likely diagnosis is:
a) 'ABO' incompatibility
b) G6PD deficiency
c) Rh incompatibility
d) Congenital spherocytosis
e) Breast-feeding jaundice

461. The serum glucose level is 20 mg/dL in a 1-hour-old newborn girl of a diabetic mother. She is asymptomatic. The next step in management is:
a) Intravenous D_{10} glucose water
b) Oral feeding with D_5 glucose water
c) Intravenous D_5 glucose water
d) Intravenous D_5 0.45 (half normal saline)
e) Reassurance

462. The serum glucose level is 50 mg/dL in 1-hour-old asymptomatic newborn boy of diabetic mother. The next step in management is:
a) Oral feeding with formula as soon as possible
b) Oral feeding with D_5 glucose water
c) Intravenous D_5 glucose water
d) Intravenous D_{10} glucose water
e) Oral feeding with breast milk after 3 hours

463. A child appears in the ER with aspiration of a button battery. The next step in management is:
a) Chest x-ray AP view
b) Chest x-ray lateral view
c) ENT consultation after the chest x-ray results
d) Observation for 24 hours
e) Removal of the battery as quickly as possible

464. A child appears in the ER with aspiration of a coin. The child is asymptomatic. The flat surface of the coin is visible in the anteroposterior view and the edge of the coin is visible in the lateral view. The next step in management is:
a) Removal of the coin immediately
b) Observe for up to 24 hours
c) Observe for up to 48 hours
d) MRI study of the neck and upper part of the thoracic cavity
e) Intravenous glucagon

465. A child appears in the ER with meat impactions in the esophagus. The child is asymptomatic. He has no problems in handling the secretions. The next step in management is:
a) Meat should be removed immediately
b) Chest x-ray AP-view
c) Chest x-ray lateral view
d) Observation for up to 12 hours
e) Observation for up to 48 hours

466. A symptomatic child appears in the ER with aspiration of a plastic material. The plain x-ray appears unremarkable. The next step in management is:
a) Remove the plastic material
b) Barium contrast study of the esophagus
c) MRI study of the esophagus
d) CT scan study of the upper chest areas
e) Observation for 24 hours

467. A child appears with right lower quadrant pain, followed by nausea, vomiting, and fever for the last 6 hours. A physical examination reveals tenderness at the McBurney point, rigidity over the rectus muscle, presence of rebound tenderness, and mild abdominal distension. The next step in management is:
a) Call pediatric surgeon
b) CT scan of the abdomen
c) Barium enema
d) Upper GI series
e) Ultrasonography of the abdomen

468. A child developed fever and right lower quadrant pain 24 hours after appendectomy. A physical examination reveals tenderness and redness in the right lower quadrant of the abdomen. The next step in management is:
a) CT scan of the abdomen
b) Ultrasonography of the abdomen
c) Needle aspiration of the affected area
d) Laparatomy
e) Barium enema

469. A full-term newborn appears with thick meconium stain. Her Apgar scores are 2 and 8 at 1 and 5 minutes respectively. The meconium is removed from the oropharynx. However, no meconium is visualized below the vocal cord. Most likely, this child will develop:
a) Meconium aspiration
b) Transient tachypnea of the newborn
c) Pneumothorax
d) Pneumomediastinum
e) No meconium aspiration

470. A full-term newborn male developed seizure activities secondary to perinatal asphyxia. A physical examination reveals absence of Moro reflex and tendon reflexes. There is presence of hypotonia, and extension of both upper and lower extremities. Most likely, the newborn will develop:
a) Normally
b) Cerebral palsy
c) Intraventricular hemorrhage
d) Subdural bleeding
e) Epidural bleeding

471. A child is receiving digoxin and furosemide for a long period of time. The most likely complication is:
a) Hyperkalemia
b) Hypokalemia
c) Metabolic acidosis
d) Hypercalcemia
e) Hypernatremia

472. All of the following are complications of furosemide therapy **except**:
   a) Ototoxicity
   b) Hypouricemia
   c) Hypercalciuria
   d) Ischemic hepatitis
   e) Photosensitivity

473. All of the following are complications of digoxin therapy **except**:
   a) Bradycardia
   b) Ototoxicity
   c) Diplopia
   d) Yellow or green vision
   e) Depression

474. The following electrolyte abnormalities enhances digoxin toxicity:
   a) Hyperkalemia and hypocalcemia
   b) Hypernatremia and hypocalcemia
   c) Hyponatremia and hypomagnesemia
   d) Hypomagnesemia and hypocalcemia
   e) Hypokalemia and hypercalcemia

Match all the fetal conditions and appropriate therapies (475-479):

475. Parvovirus infection and hydrops
476. HIV-AIDS (mother)
477. Prevention of neural tube defects
478. Meconium-stained amniotic fluid
479. Lung immaturity

   a) Zidovudine (AZT) and protease inhibitors
   b) Folate
   c) Intrauterine P-RBCs transfusion
   d) Betamethasone
   e) Amnioinfusion

480. A routine newborn physical examination reveals petechiae of the skin of the head and neck. The newborn is delivered by NSVD and remains asymptomatic. The next step in management is:
   a) Head ultrasonography
   b) Sepsis work up
   c) Rule out isoimmune thrombocytopenia
   d) Reassurance
   e) Intravenous immunoglobulin therapy

481. A skull x-ray of newborn infant reveals linear fracture. She is asymptomatic. The next step in management is:
   a) Neurology consultation
   b) Head ultrasonography
   c) Head MRI
   d) Elevation of the skull bones
   e) Reassurance

482. A skull x-ray of the newborn infant reveals severely depressed fracture. He is asymptomatic. The next step in management is:
   a) Reassurance
   b) Neurology consultation
   c) Head MRI
   d) Head ultrasonography
   e) Elevation of the depressed skull bones

483. A routine newborn physical examination reveals the Ping-Pong ball sensation over the right parietal bone. The most likely diagnosis is:
   a) Linear fracture of the skull
   b) Depressed fracture of the skull
   c) Craniosynostosis
   d) Intraventricular hemorrhage
   e) Subarachnoid hemorrhage

484. The use of narcan in newborns is contraindicated in the following condition:
   a) Mother received morphine before delivery
   b) A severely depressed newborn at birth
   c) Mother is addicted to opiates
   d) A full-term newborn who is receiving gentamicin therapy.
   e) A newborn who is receiving artificial ventilation.

485. All of the following are clinical manifestations of a newborn whose mother received narcotics before delivery **except**:
   a) Cyanosis
   b) Weak cry
   c) Slow to breathe
   d) Hypertonic
   e) Hypotonic

486. A mother received morphine shortly before the delivery of her newborn infant. The infant appears cyanotic, hypotonic, lethargic, and apneic. The therapy includes all of the following **except**:
   a) Oxygen
   b) Tactile stimulation
   c) Bag and mask ventilation
   d) Narcan
   e) CNS stimulant drugs

487. In multifactorial inheritance, the risk of recurrence for the next child of unaffected parents with one affected child is:
   a) Between 1-2%
   b) Between 3-5%
   c) Between 6-8%
   d) Between 9-12%
   e) 25%

488. When a mother is balanced 21/21 translocation carrier, the risk of recurrence of Down syndrome in children is:
   a) 10%
   b) 25%
   c) 50%
   d) 66%
   e) 100%

489. The congenital deformity most commonly occurs in the following system:
   a) CNS
   b) Cardiac
   c) Pulmonary
   d) Gastrointestinal
   e) Musculoskeletal

490. The most common extrinsic cause of a fetal deformity is:
   a) Small uterus
   b) Bifid uterus
   c) Breech presentation
   d) Fetal crowding
   e) Small maternal pelvis

491. The external fetal deformity most commonly occurs during:
   a) Early half of 1st trimester
   b) Late half of 1st trimester
   c) Early half of 2nd trimester
   d) Late half of 2nd trimester
   e) 3rd trimester

492. The risk of recurrence in talipes equinovarus is:
   a) 1-2%
   b) 3-5%
   c) 6-9%
   d) 10-15%
   e) 16-25%

493. All of the following are features of Costello syndrome **except**:
   a) Mental retardation
   b) Microcephaly
   c) Cerebral atrophy
   d) Cardiomyopathy
   e) Papillomas

494. All of the following are features of Kabuki syndrome **except**:
   a) Prominent fingertip pads
   b) Cardiac anomaly
   c) Short palpebral fissures
   d) Mental retardation
   e) Protuberant ears

495. The current cutoff for duration of rupture membrane that increases the risk of neonatal infections is longer than:
   a) 6 hours
   b) 12 hours
   c) 18 hours
   d) 20 hours
   e) 24 hours

496. All of the following conditions are indications of intrapartum penicillin prophylaxis **except**:
   a) GBS bacteriuria during current pregnancy
   b) Previous infant with an invasive GBS disease
   c) Positive GBS screening culture during current pregnancy
   d) Maternal GBS status is unknown and rupture membrane is more than 18 hours.
   e) Previous pregnancy with a positive GBS screening culture

497. The most commonly reported illicit drug ever used by adolescents is:
   a) Marijuana
   b) Cocaine
   c) Ecstasy
   d) Heroin
   e) Amphetamine

498. All of the following are clinical manifestations of alcohol ingestion **except**:
   a) CNS stimulations
   b) Euphoria
   c) Impaired short-term memory
   d) Epigastric pain
   e) Guiac-positive stools

499. An adolescent is considered a problem drinker. All of the following statements indicate problem drinker **except**:
   a) He was drunk 5 times in the past year.
   b) He has problems with school authorities.

c) He has driven after drinking.
d) He has been criticized by dates for drinking habits.
e) He has problem with his friends

500. A 16-year-old boy appears in the ER at 4 AM. He has alcoholic aroma in his breath. He is severely depressed, disoriented, lethargic, and unresponsive. His serum alcohol level is 65 mg/dL. The pupils are normal in sizes and react to light normally. The funduses appear normal. The most likely cause of his depression is:
a) Alcohol overdose syndrome
b) Psychiatric illness
c) Cerebral hemorrhage
d) Brain tumor
e) Ingestion of other drugs along with alcohol

501. The usual mechanism of death from the alcohol overdose syndrome is:
a) Cardiac failure
b) Cerebral hemorrhage
c) Acute GI bleeding
d) Respiratory depression
e) Encephalopathy

502. The preferred therapy in patients with blood alcohol level 450 mg/dL is:
a) Charcoal through OG tube
b) Ipecac
c) Charcoal enema
d) Dialysis
e) Phenobarbital

503. The hallucinogenic properties of marijuana is due to:
a) Benzoyl ecgonine
b) Acetaldehyde
c) Dihydrocannabinol
d) Trihydrocannabinol
e) Tetrahydrocannabinol

504. An "amotivational" syndrome occurs most likely due to the following drug:
a) Toluene intoxication
b) Cocaine use
c) Amphetamine use
d) Phencyclidine use
e) Chronic marijuana use

505. A 17-year-old girl appears in the ER with euphoria, impairment of short-term memory, distortion of time perception, visual hallucination and recall of frightening hallucinations. The most likely diagnosis is:
a) Metamphetamine use
b) Cocaine use
c) Marijuana use
d) Opiate overdose syndrome
e) Lysergic acid diethylamide use

506. All of the following are features of marijuana intoxication **except**:
a) Hypothermia
b) Tachycardia
c) Systolic hypertension
d) Diastolic hypertension
e) Increased intraocular pressure

Match the following drug interactions with marijuana and their effects (507-509):

507. Diazepam
508. Cocaine
509. Propranolol

a) Antagonistic effect
b) Potentiate sedation
c) Potentiate stimulation

Match all the diseases with clinical manifestations (510-514):

510. Wiskott-Aldrich syndrome
511. Ataxia-telangiectasia
512. Cartilage hair hypoplasia
513. Omenn syndrome
514. Hyper IgE syndrome

a) Recurrent staphylococcal abscesses, persistent pneumatoceles, and coarse facial features.
b) Chronic sinopulmonary disease and a high incidence of cancer
c) Prolonged bleeding from circumcision sites, bloody diarrheas, atopic dermatitis, and S. pneumoniae infections
d) Fatal varicella infections, redundant skin, hyperextensible joints of the hands and feet but an inability to extend the elbow completely.
e) Intractable diarrhea, exfoliative erythroderma, hepatosplenomegaly, and lymphadenopathy

515. The most common cause of eosinophilia in the USA is:
a) Hyper IgE syndrome
b) Inflammatory bowel disease
c) Trichinosis
d) Eosinophilic leukemia
e) Allergy

516. The most common cause of hypereosinophilic syndrome is:
a) Idiopathic
b) Malignancy
c) Allergy
d) Immunodeficiency disease
e) Helminth infections

517. The most serious and life-threatening complication of hypereosinophilic syndrome is:
a) Brain involvement
b) Cardiac involvement
c) Pulmonary involvement
d) Hepatic involvement
e) Splenic involvement

518. The preferred initial therapy in patients with hypereosinophilic syndrome is:
a) Hydroxyurea
b) Alpha-interferon
c) Corticosteroids
d) Methotrexate
e) Partial exchange transfusion

519. All of the following are diagnostic criteria of hypereosinophilic syndrome **except**:
a) Eosinophilia greater than 1,500 cells/micro L persisting more than 1 year.
b) Eosinophilia greater than 1,500 cells/micro L persisting more than 6 months.
c) Lack of another diagnosis to explain the eosinophilia
d) Signs of organ involvements
e) Symptoms of organ involvements

520. The curative therapy in patients with chronic granulomatous disease is:
   a) Interferon-gamma
   b) Amphotericin B
   c) Prednisone
   d) Trimethoprim-sulfamethoxazole
   e) Bone marrow transplantation

521. The curative therapy in patients with Chediak-Higashi syndrome is:
   a) High-dose ascorbic acid
   b) High-dose vitamin A
   c) Interferon-gamma
   d) Interferon-alpha
   e) Bone marrow transplantation

Match all the different diseases with the laboratory tests (522-526):

522. Chediak-Higashi syndrome
523. Leukocyte adhesion deficiency-1 (LAD-1)
524. Leukocyte adhesion deficiency-2 (LAD-2)
525. Myeloperoxidase deficiency
526. Chronic granulomatous disease

   a) Histochemical analysis reveals deficiency of neutrophils and monocytes enzyme.
   b) Dihydrorhodamine 123 fluorescence (DHR) test detects oxidant productions.
   c) Wright-stained blood films reveal large inclusions in all nucleated blood cells.
   d) Lack of sialyl-Lewis X on the neutrophils
   e) Flow cytometric measurements of surface CD1 1b in stimulated and unstimulated neutrophils using monoclonal antibodies directed against CD1 1b.

527. All of the following are physical examination findings in patients with allergic rhinitis **except**:
   a) Continuous open-mouth breathing
   b) Dark circles under eyes
   c) Longitudinal nasal crease
   d) Dental malocclusions
   e) Abnormal facial developments

528. A 10-year-old girl appears with nasal congestion, sneezing, itching, clear rhinorrhea, conjunctival irritation, loss of sense of smell, and difficulty in sleeping. A nasal examination reveals all of the following **except**:
   a) Bluish mucous membranes
   b) Purulent nasal secretions
   c) Swollen turbinates
   d) Edematous mucous membranes
   e) Mildly erythematous mucous membranes

529. All of the following are effective prevention strategies in patients with allergic rhinitis **except**:
   a) For animal allergens in the home, the only effective measure is removal of the pet.
   b) HEPA filters reduce the airborne mold spores.
   c) Sealing the mattresses, pillows, and covers with allergen-proof encasing
   d) Bed linen and blankets should be washed every week in hot water (125°F)
   e) Air conditioning lowers the pollen exposure.

530. The majority of patients with allergic rhinitis have:
   a) Otitis media
   b) Tonsillopharyngitis
   c) Asthma
   d) Obstructive sleep apnea
   e) Secondary bacterial infections

Match all of the following medications and their effects on glucocorticoids metabolism and clearance (531-534):

531. Phenytoin
532. Rifampin
533. Ketoconazole
534. Erythromycin

a) Delays glucocorticoid clearance
b) Enhances glucocorticoid clearance
c) Increases glucocorticoid metabolisms
d) Delays glucocorticoid metabolisms

535. The following statements is true about long-term inhaled glucocorticoids therapy and its effect on growth:
a) Long-term inhaled glucocorticoid therapy has no effect on adult attained height.
b) Long-term inhaled glucocorticoid therapy decreases the adult height by 2.5 cm.
c) Long-term inhaled glucocorticoid therapy increases the adult height b 2.5 cm.
d) Long-term inhaled glucocorticoid therapy decreases the adult height by 5.0 cm.
e) Long-term inhaled glucocorticoid therapy increases the adult height by 5.0 cm.

536. The most likely cause of oral thrush due to inhaled glucocorticoids is:
a) Generalized immunosuppression
b) Local immunosuppression
c) Uncleaned oral cavity
d) Direct irritation of the oral mucous membranes
e) Predisposing oral ulcers

537. All of the following are adverse effects of frequently administered beta-agonist therapy in patients with severe asthma **except**:
a) Irritability
b) Tachycardia
c) Tremor
d) Hypokalemia
e) Hyponatremia

538. The following monitoring is indicated when patients with severe asthma those who are receiving beta-agonist therapy frequently:
a) Urine output
b) Blood pressure
c) Temperature
d) EKG
e) Pulse oximetry

539. The important side effect of intravenous magnesium sulfate therapy in patients with asthma is:
a) Hypotension
b) Hypertension
c) Constipation
d) Sedation
e) Respiratory stimulation

540. All of the following therapies are indicated in hospitalized patients with an acute severe asthma attack **except**:
a) Oxygen
b) Inhaled beta-agonist
c) Systemic glucocorticoid
d) Fluids at or slightly below maintenance
e) Fluids above maintenance

541. A severely asthmatic patient is receiving oxygen, inhaled beta-agonist, and systemic glucocorticoid therapies. The child is intubated because of respiratory failure. All of the following therapies are useful in this patient **except**:
a) Intravenous beta-agonist
b) Intravenous theophylline
c) Heliox

d) Intramuscular magnesium sulfate
e) Intravenous magnesium sulfate

542. All of the following therapies are given in patients with acute asthma attacks in the ER **except**:
a) Oxygen
b) Systemic glucocorticoids
c) Subcutaneous epinephrine may be given
d) Inhaled beta-agonist
e) Heliox

543. All of the following measures can reduce the development of asthma **except**:
a) Influenza vaccine
b) Healthy diet
c) Breast feeding more than 4 months
d) Active life style
e) Avoid environmental tobacco smoke beginning prenatally

544. All of the following are adverse effects of chronic systemic glucocorticoid therapy **except**:
a) Dermal thinning
b) Cataracts
c) Glaucoma
d) Increased skin fragility
e) Increased IgG levels

545. The following statements is not true about beta-lactum hypersensitivity reactions:
a) Patients with late-onset morbilliform rashes with amoxicillin require skin testing before penicillin therapy.
b) If a patient needs penicillin and has past history of pencillin allergy, skin testing should be performed to determine the presence of penicillin-specific IgE with both the major and minor determinants of penicillin.
c) If a patient has history of penicillin allergy and needs cephalosporin, skin testing should be performed.
d) If a patient has history of IgE-mediated reaction to cephalosporin and needs penicillin, skin testing should be performed.
e) Aztreonam (monobactam) can be used safely in most penicillin-allergic patients.

546. In addition to high spiking temperature (104°F or higher) that is remittent and unresponsive to antibiotics therapy, all of the following clinical manifestations can make the diagnosis of acute febrile phase of Kawasaki disease **except**:
a) Polymorphous, vesicular rash primarily over the trunk
b) Bilateral bulbar conjunctival injection, generally nonpurulent
c) Strawberry tongue, injected oropharynx, injected and/or dry fissured lips
d) Edema and/or erythema of the hands or feet
e) Cervical lymphadenopathy, 1.5 cm or more, usually unilateral

547. All of the following are clinical manifestations of subacute phase of Kawasaki disease **except**:
a) Periungual desquamation of the fingers and toes
b) Thrombocytosis
c) Development of coronary aneurysms
d) Conjunctival injection
e) Normal ESR (erythrocyte sedimentation rate)

548. The following laboratory finding is usually **not** present in patients with Kawasaki disease during the first week of illness:
a) Thrombocytosis
b) Elevated ESR
c) Normal or elevated WBC counts
d) Normocytic anemia
e) Elevated neutrophils and bands count

549. A child is diagnosed to have Kawasaki disease. The 1st echocardiogram is performed on the 5th day of illness when the diagnosis is made. The echocardiogram result is normal. The next step in management is :
a) No more echocardiogram is needed.
b) The 2nd echocardiogram should be performed after 2-3 weeks of illness.
c) The 2nd echocardiogram should be performed after 4-6 weeks of illness.

d) The 2nd echocardiogram should be performed after 6-8 weeks of illness.
e) The 2nd echocardiogram should be performed after 1 year of illness.

550. A child has Kawasaki disease and coronary artery aneurysms. The next step in management is:
a) Repeat echocardiogram after 1 week
b) Coronary angiography
c) Thallium scan
d) Follow-up chest x-ray daily
e) Consult with a pediatric cardiologist

551. A child appears with fever, weight loss, myalgias, arthralgia, chronic sinusitis, hemoptysis, hematuria, and proteinuria. The most likely diagnosis is:
a) Tuberculosis
b) Wegener granulomatosis
c) DIC
d) HIV-infection
e) Churg-Strauss syndrome

552. The most common organism responsible for toxic shock syndrome is:
a) S. aureus
b) S. epidermidis
c) Group A Streptococcus
d) Group B Streptococcus
e) Pseudomonas aeruginosa

553. A child appears with sudden onset of high fever (more than 38.8°C), vomiting, diarrhea, sore throat, and followed by diffuse erythematous macular rash (sunburn-like or scarlatiniform). A physical examination reveals hypotension, altered level of consciousness, and strawberry tongue. The most likely diagnosis is:
a) Toxic shock syndrome
b) Kawasaki disease
c) Group A Streptococcus infection
d) Pseudomonas infection
e) AIDS

554. The most common risk factor in women with toxic shock syndrome is:
a) Menstruation and tampon use
b) Multiple sexual partners
c) HIV-infection
d) Candida infection
e) Ectopic pregnancy

555. The preferred initial antibiotic therapy in patients with toxic shock syndrome is:
a) Gentamicin
b) Ampicillin
c) Nafcillin
d) Ceftriaxone
e) Clindamycin

Match all food with food proteins (556-559):

556. Peanut
557. Egg
558. Cow's milk
559. Fish

a) Paralbumin
b) Casein
c) Ovomucoid
d) Vicilin

560. All of the following recommendations are useful in prevention of food allergies **except**:
   a) Avoid seafood until 3 years of age
   b) Avoid egg until 18-24 months of age
   c) Avoid cow's milk until 9 months of age
   d) Promotion of breast feeding with maternal exclusion of nuts
   e) Promotion of breast feeding with maternal exclusion of peanuts

561. The following side effect can occur due to the **prolonged use** of acetaminophen:
   a) Hepatic injury
   b) Renal injury
   c) GI injury
   d) Pancreatic injury
   e) CNS injury

562. The **massive overdose** of acetaminophen can produce:
   a) Renal injury
   b) Hepatic injury
   c) CNS injury
   d) GI injury
   e) Pancreatic injury

563. All of the following drugs can cause neutropenia **except**:
   a) Penicillins
   b) Phenothiazines
   c) Sulfonamides
   d) Anticonvulsants
   e) Gentamicin

564. The following finding is **not** present in patients with Kostmann syndrome:
   a) Monocytosis
   b) Eosinophilia
   c) Neutrophilia
   d) Anemia
   e) Recurrent, severe pyogenic infections

565. The following finding is **not** present in patients with Shwachman-Diamond syndrome:
   a) Constipation
   b) Failure to thrive
   c) Delayed puberty
   d) Neutropenia
   e) Metaphyseal chondrodysplasia

566. The following finding is **absent** in patients with cartilage-hair hypoplasia:
   a) Short stature
   b) Short limbs
   c) Very fine, thin hair
   d) Neutropenia
   e) Microcytic anemia

567. The most common organism isolated in patients with neutropenia is:
   a) Group A Streptococcus
   b) Staphylococcus epidermidis
   c) Group B Streptococcus
   d) Streptococcus pyogenes
   e) Staphylococcus aureus

568. Leukoencephalopathy is a clinical syndrome characterized by all of the following **except**:
   a) Ataxia
   b) Slurred speech

c) Seizures
d) Dysphagia
e) Decorticate posturing

569. The most common cause of fatal motor vehicle crashes between 16-19 years of age is:
a) Speeding
b) Driver error
c) Alcohol
d) Three passengers
e) Less frequently use of seat belts

570. The leading causes of morbidity and mortality in adolescents are:
a) Automobile and motorcycle accidents
b) Firearm and drowning
c) Fall and suffocation
d) Poisoning and drowning
e) Drowning and suffocation

571. Obesity is correlated with all of the following diseases **except**:
a) Hypercholesterolemia
b) Diabetes insipidus
c) Gallbladder disease
d) Hypertension
e) Arthritis

572. The body mass index (BMI) of a child is 86th percentile. The most likely interpretation is:
a) Underweight
b) Normal weight
c) Overweight
d) Moderately obese
e) Severely obese

573. The following chemical changes may be associated with suicide:
a) Low levels of serotonin 5-HT (hydroxytryptamine)
b) High levels of testosterone
c) High levels of dehydrotestosterone
d) Low levels of prolactin
e) Low levels of hypothalamic releasing hormone

574. All of the following are risk factors of suicide **except**:
a) Substance abuse
b) Gay youth
c) Bisexual youth
d) Heterosexual youth
e) Parent-child conflict

575. The cause of chronic fatigue syndrome is:
a) Unknown
b) Influenza
c) Anemia
d) Undiagnosed UTI
e) Infectious mononucleosis

576. The most common life-threatening emergency of the gastrointestinal tract in newborns is:
a) Malrotation
b) Volvulus
c) Stress ulcer
d) Hirschsprung disease
e) Necrotizing enterocolitis

577. The most common cause of necrotizing enterocolitis is:
   a) Multifactorial
   b) Aggressive feeding
   c) E. coli infection
   d) Thickened formula feeding
   e) Hypoxic damage to intestine

578. The first signs of NEC are:
   a) Gastric retention with abdominal distension
   b) Guiac-positive stools with vomiting
   c) Portal venous gas with abdominal distension
   d) Pneumatosis intestinalis with Guiac-positive stools
   e) Vomiting and thickening of bowel wall

579. The preferred diagnostic study in patients with NEC is:
   a) Upper GI series
   b) Barium enema
   c) Abdominal CT scan
   d) Plain abdominal x-ray
   e) Abdominal MRI

580. The radiological sign that indicates a severe disease in patients with NEC is:
   a) Paralytic ileus
   b) Edema of bowel wall
   c) Portal vein gas
   d) Air-fluid levels
   e) Pneumatosis intestinalis

581. A 6-day-old preterm infant develops gastric retension, abdominal x-ray reveals pneumatosis intestinalis. The management includes all of the following **except**:
   a) NPO and intravenous fluids
   b) Orogastric decompression
   c) Blood culture
   d) Coagulation profile
   e) Keep umbilical catheters for ABC, CBC, and SMA-6 monitoring

582. After the initial diagnosis of NEC, the preferred diagnostic study to detect perforation is:
   a) Anteroposterior abdominal x-ray
   b) Abdominal CT scan
   c) Abdominal MRI
   d) Abdominal paracentesis
   e) Cross-table lateral abdominal x-ray

583. The indication for surgery in patients with NEC is:
   a) Neutropenia
   b) Perforation
   c) Thrombocytopenia
   d) Gross bloody stools
   e) Metabolic acidosis

584. The cause of an idiopathic focal intestinal perforation in newborns is:
   a) Steroids
   b) Aminophylline
   c) Gentamicin
   d) Hypoxia
   e) Vancomycin

585. The clinical features of acute form of kernicterus during phase **1 (1st 1-2 days)** are all of the following **except**:
   a) Poor sucking
   b) Seizures
   c) Stupor
   d) Hypotonia
   e) Hypertonia

586. The clinical features of chronic form of kernicterus **during the first year** are all of the following **except**:
   a) Hypotonia
   b) Delayed motor skills
   c) Seizures
   d) Obligatory tonic neck reflexes
   e) Active deep tendon reflexes

587. The clinical features of chronic form of kernicterus **after first year** are all of the following **except**:
   a) Choreoathetosis
   b) Ballismus
   c) Upward gaze
   d) Tremor
   e) Mixed hearing loss

588. All of the following therapies are effective in hyperbilirubinemia **except**:
   a) Phototherapy
   b) Tin (Sn)-protoporphyrin
   c) Intravenous immunoglobulin
   d) Exchange transfusion with PRBCs
   e) Exchange transfusion with whole blood

589. A 7-day-old boy develops a dark grayish brown discoloration of the skin while undergoing phototherapy. The following statement is most appropriate about this infant:
   a) The boy has only indirect hyperbilirubinemia.
   b) The boy has only direct hyperbilirubinemia.
   c) The boy has both direct and indirect hyperbilirubinemias.
   d) The discoloration is due to intensive phototherapy and dehydration.
   e) The color usually disappears within 24 hours after discontinuation of phototherapy.

590. All of the following clinical manifestations are indications of packed red blood cells transfusion **except**:
   a) Asymptomatic infant with hemoglobin 7.1, hematocrit 21, and absolute reticulocyte count 100,000 cells/μL
   b) Infant required moderate mechanical ventilation (mean airway pressure 9 cm H_2O and Fio_2 more than 40%), hemoglobin 10.9, and hematocrit 34
   c) Infant required minimal mechanical ventilation (Fio_2 35%), hemoglobin 9.7, and hematocrit 29
   d) Infant required supplemental oxygen (35%), heart rate 182 per minute, hemoglobin 7.9, and hematocrit 24
   e) Infant required nasal CPAP (continuous airway positive pressure, Fio_2 38%), respiratory rate 82 per minute, hemoglobin 7.8, and hematocrit 24

591. The following drugs can cause gynecomastia **except**:
   a) Digoxin
   b) Furosemide
   c) Thiazides
   d) Cimetidine
   e) Omeprazole

592. The most common organism in CSF shunt infections is:
   a) Staphylococcus aureus
   b) Staphylococcus epidermidis
   c) E. coli
   d) Streptococcus pneumoniae
   e) Neisseria meningitidis

593. The most common organism is **distal** infections of VP shunt is:
   a) Gram-negative organisms
   b) S. aureus
   c) S. epidermidis
   d) S. pneumoniae
   e) S. haemolyticus

594. The most common clinical manifestation in patient with a **distal** shunt infection is:
   a) Abdominal symptoms
   b) Neurologic symptoms
   c) Cardiac symptoms
   d) Pulmonary symptoms
   e) Endocrine abnormalities

595. The most common organism in wound infections of the VP shunt is:
   a) S. aureus
   b) S. epidermidis
   c) E. coli
   d) S. saprophyticus
   e) S. pneumoniae

596. All of the following precautions can prevent infections due to urethral catheter **except**:
   a) Duration of use should be minimized
   b) Prophylactic antibiotics should be used.
   c) Casual use of catheter should be avoided.
   d) Antibiotic-impregnated urinary catheters reduces the rate of infection.
   e) Silver-impregnated urinary catheters lowers the rate of infection.

597. The most common organism in patients with a long-term implanted orthopedic prosthesis is:
   a) Pseudomonas aeruginosa
   b) Candida albicans
   c) S. epidermidis
   d) E. coli
   e) Streptococcus pneumoniae

598. The most common organism in children with osteomyelitis is:
   a) Pseudomonas aeruginosa
   b) S. epidermidis
   c) S. aureus
   d) S. pneumoniae
   e) E. coli

599. The most common organism in children with suppurative arthritis is:
   a) S. epidermidis
   b) E. coli
   c) Pseudomonas aeruginosa
   d) S. aureus
   e) Streptococcus pneumoniae

600. The specific laboratory test to diagnose toxic shock syndrome is:
   a) Positive urine culture
   b) Positive blood culture
   c) Positive CSF culture
   d) Positive wound culture
   e) No specific laboratory test

601. A physical examination of the newborn male reveals a "fish mouth" meatus of the distal urethra and normally developed foreskin. The most likely diagnosis is:
   a) Epispadius

b) Urethral stricture
c) Hypospadius
d) Meatal stenosis
e) Penile torsion

602. A 17-year-old boy recently immigrated to the USA. A physical examination reveals distal hypospadius. He is asymptomatic and is sexually active. The next step in management is:
a) Renal ultrasonography
b) Immediate surgical repair
c) VCUG
d) Reassurance
e) Surgical repair after 1 month

603. The preferred technique to repair a cleft lip is:
a) W-plasty
b) M-plasty
c) N-plasty
d) A-plasty
e) Z-plasty

Match all of the following diagnosis and clinical manifestations (604-608):

604. Phenytoin therapy
605. Vitamin D-dependent rickets
606. Neutrophil chemotactic deficiency
607. Immunosuppression
608. Kidney failure

a) Enamel hypoplasia
b) Juvenile periodontitis
c) Facial dysmorphology
d) Oral candidiasis
e) Gingival enlargement

609. The most common cause of premature loss of **primary teeth** is:
a) Vitamin D-dependent rickets
b) Nutritional rickets
c) Hypoparathyroidism
d) Hypothyroidism
e) Premature eruption of permanent teeth

Match the different conditions and teeth discoloration (610-612):

610. Porphyria
611. Neonatal hyperbilirubinemia
612. Tetracycline

a) Brown-yellow discoloration
b) Blue to black discoloration
c) Red-brown discoloration

613. All of the following may be complications of natal/neonatal teeth **except**:
a) Pain
b) Abrasion of nipple
c) Aspiration
d) Riga-Fede disease
e) Easy to feed solid food

614. Delayed eruption of the primary teeth may be due to all of the following conditions **except**:
   a) Hypothyroidism
   b) Hypopituitarism
   c) Cleidocranial dysplasia
   d) Progeria
   e) Turner syndrome

615. Pyloric atresia is associated with the following condition:
   a) Down syndrome
   b) Epidermolysis bullosa
   c) Duodenal atresia
   d) Congenital adrenal hyperplasia
   e) Infant of diabetic mother

616. A full term newborn is delivered by NSVD. Maternal history of polyhydramnios. Aspiration of infant's stomach reveals 20 mL of bile stained gastric fluid. The most likely diagnosis is:
   a) Normal condition
   b) H-type TE fistula
   c) Swallowed amniotic fluid
   d) High intestinal obstruction
   e) Low intestinal obstruction

617. A high intestinal obstruction is usually associated with the following condition:
   a) Metabolic acidosis
   b) Respiratory acidosis
   c) Hypochloremic metabolic alkalosis
   d) Hyperchloremic metabolic alkalosis
   e) Hypochloremic metabolic acidosis

618. A newborn is diagnosed with an intestinal obstruction. All of the following findings suggest bowel infarction **except**:
   a) Leukocytosis without thrombocytopenia
   b) Leukocytosis with thrombocytopenia
   c) Metabolic acidosis
   d) Hematochezia
   e) Leukopenia with thrombocytosis

619. The most common clinical manifestation in patients with portal hypertension is:
   a) Ascites
   b) Jaundice
   c) Palmar erythema
   d) Vascular telangiectasia
   e) Bleeding from esophageal varices

620. An infant is admitted for liver transplantation. Pretransplantation management includes all of the following **except**:
   a) Medium-chain triglycerides
   b) Nocturnal nasogastric tube feedings
   c) Intravenous nutrition, especially lipids
   d) Vitamin A, D, E, and K
   e) Caloric requirements up to 120 Kcal/kg/day

621. All of the following immunizations should be given in children prior to hepatic transplantation **except**:
   a) Hepatitis A
   b) Hepatitis B
   c) Varicella
   d) MMR
   e) Hepatitis C

622. Early complications after hepatic transplantation include all of the following **except**:
   a) Electrolyte imbalance
   b) Renal dysfunction
   c) Hypertension
   d) Fluid shifts
   e) Bacterial infections

623. The most ominous early problem after hepatic transplantation is:
   a) Hypertension
   b) Acute hypotension
   c) Ascites
   d) Renal failure
   e) Thrombosis of graft vessels

624. The most common infection after hepatic transplantation is:
   a) Bacterial
   b) CMV
   c) Adenovirus
   d) Candida albicans
   e) Pneumocystis carinii

Match the following organs and their predominance in hematopoiesis in different gestational ages (625-627):

625. Liver
626. Yolk sac
627. Bone marrow

   a) 2 to 6 weeks
   b) 6 to 24 weeks
   c) 24 to 42 weeks

Match the following drugs and their side effects (628-632):

628. Diazoxide
629. Hydrochlorthiazide
630. Enalaprilat
631. Labetalol
632. Prazocin

   a) First dose, orthostatic hypotension
   b) Transient hypotension
   c) Bronchospasm
   d) Hyperuricemia
   e) Hyperglycemia

633. The principal effect of vasopressin on the kidney is via the following receptor:
   a) V1 receptors
   b) V2 receptors
   c) V3 receptors
   d) V4 receptors
   e) V5 receptors

634. The human V2 receptor gene is located on the following chromosome:
   a) Short arm of the X chromosome
   b) Long arm of the X chromosome
   c) Short arm of the Y chromosome
   d) Long arm of the Y chromosome
   e) Chromosome 21

635. The most common cause of thyroid disease in children and adolescents is:
   a) Lymphocytic thyroiditis
   b) Endemic goiter
   c) Iodide goiter
   d) Subacute nonsuppurative thyroiditis
   e) Acute suppurative thyroiditis

636. The most common cause of acquired hypothyroidism is:
   a) Iodine deficiency
   b) Congenital goiter
   c) Iodide goiter
   d) Simple goiter
   e) Lymphocytic thyroiditis

637. The most common cause of sporadic goiter is:
   a) Lithium carbonate
   b) Lymphocytic thyroiditis
   c) Hyperthyroidism
   d) Amiodarone
   e) Thyroid cyst

638. A 14-year-old girl is diagnosed with Graves disease. A physical examination reveals apical systolic murmur. The most likely diagnosis is:
   a) Tricuspid regurgitation
   b) Tricuspid stenosis
   c) Mitral regurgitation
   d) Mitral stenosis
   e) Aortic stenosis

639. A 9-year-old boy is diagnosed with Graves disease. All of the following laboratory findings are noted in patients with Graves disease **except**:
   a) Elevated serum levels of thyroxine (T_4)
   b) Decreased serum levels of thyroid stimulating hormone (TSH)
   c) Elevated serum levels of triiodothyronine (T_3)
   d) Elevated serum levels of free T_3 and free T_4
   e) Absence of thyroid receptor-stimulating antibody (TRSAb)

640. The radiographic finding in very young children with Graves disease is:
   a) Delayed skeletal maturation
   b) Craniotabes
   c) Increased bone density
   d) Decreased bone density after therapy
   e) Advanced skeletal maturation

641. All of the following syndromes produce craniostenosis **except**:
   a) Dandy-Walker syndrome
   b) Carpenter syndrome
   c) Apert syndrome
   d) Kleeblattschadel syndrome
   e) Crouzon disease

Match the different causes of cataract and different manifestations of cataract (642-646):

642. Corticosteroids
643. Wilson disease
644. Infantile galactosemia
645. Persistent strands of the pupillary membrane
646. Persistent remnants of the primitive hyaloid vascular system

a) Sunflower cataract
b) Posterior Subcapsular cataract
c) Anterior capsular cataract
d) Zonular type cataract
e) Posterior capsular cataract

647. Opthalmologic examination of an infant reveals a cluster of tiny vacuoles in the distribution of the Y sutures of the lens. This type of cataract is most likely noted in:
a) Premature infants
b) Toxoplasmosis
c) Congenital rubella
d) CMV infection
e) Infant of diabetic mother

648. All of the following conditions can cause cataract **except**:
a) Child abuse
b) Trauma
c) Radiation
d) Atopic dermatitis
e) Hypercalcemia

649. All of the following conditions can cause congenital lens opacities **except**:
a) Parvovirus B19 infection
b) Hypoglycemia
c) Congenital syphilis
d) Infants of diabetic mothers
e) Infants of prediabetic mothers

650. The drug of first choice for uncomplicated acute otitis media is:
a) Amoxicillin
b) Amoxicillin-clavulanate
c) Cefuroxime axetil
d) Azithromycin
e) Trimethoprim-sulfamethoxazole

651. The drug of choice for acute otitis media in patients who are allergic to beta-lactams is:
a) Erythromycin
b) Azithromycin
c) Cefuroxime axetil
d) Ceftriaxone
e) Trimethoprim-sulfamethoxazole

652. A child is diagnosed to have acute otitis media. He did not respond to amoxicillin therapy. The drug of choice is:
a) Azithromycin
b) Erythromycin
c) Trimethoprim-sulfamethoxazole
d) Cefotaxime
e) Amoxicillin-clavulanate

653. The most common side effect of using amoxicillin-clavulanate, especially in infants and young children is:
   a) Nausea
   b) Vomiting
   c) Constipation
   d) Diarrhea
   e) Pyloric stenosis

654. The children do not like to ingest cefuroxime-axetil because:
   a) It has an unattractive color.
   b) It has a bad smell.
   c) It has a bad taste.
   d) It causes diarrhea.
   e) It causes nausea.

655. The second-line drugs for acute otitis media should be effective against all of the following organisms **except**:
   a) Beta-lactamase producing strains of H. influenzae
   b) M. catarrhalis
   c) Susceptible strains of S. pneumoniae
   d) Most nonsusceptible strains of S. pneumoniae
   e) Staphylococcus aureus

656. A child appears in ER with multiple vesiculobulous lesions on the face, trunk, and extremities for the last 24 hours. His mother stated that initially he had a flulike upper respiratory illness, burning sensation, edema, and erythema of the lips and buccal mucosa. The lesions also involve eyes and oral cavity. He refused to eat and drink. Mother denies history of drug intake. The lesions have a central necrosis. The management must include all of the following **except**:
   a) Opthalmologic consultation
   b) Prophylactic systemic antibiotics
   c) Topical anesthetics
   d) Mouthwashes and glycerin oral swabs
   e) Denuded skin lesions are cleansed with saline or Burrow solution compresses.

657. All of the following infections agents can cause Stevens-Johnson syndrome. The most convincingly identified infectious agent is:
   a) Herpes simplex virus
   b) Epstein-Barr virus
   c) Group A streptococci
   d) Enteroviruses
   e) Mycoplasma pneumoniae

658. All of the following drugs can produce Stevens-Johnson syndrome **except**:
   a) Captopril
   b) Etoposide
   c) Allopurinol
   d) Vancomycin
   e) Nonsteroidal anti-inflammatory agents

659. A child appears with multiple painful oral ulcers for the last 2 months. The most likely diagnosis is:
   a) Dermatitis herpetiformis
   b) Bullous pemphigoid
   c) Linear IgA dermatosis
   d) Pemphigus foliaceus
   e) Pemphigus vulgaris

660. Marcus Gunn phenomenon involves the following cranial nerves:
   a) Facial and abducens nerves
   b) Trigeminal and oculomotor nerves
   c) Facial and glossopharyngeal nerves
   d) Abducens and vagus nerves
   e) Optic and olfactory nerves

661. Mobius syndrome includes all of the following **except**:
   a) Mental retardation
   b) Bilateral facial weakness
   c) Abducens nerve paralysis
   d) Dull facies
   e) Hypoplasia of brainstem nuclei

662. All of the following syndromes can cause microcephaly **except**:
   a) Down syndrome
   b) Cri-du-chat syndrome
   c) Edward syndrome
   d) Turner syndrome
   e) Cornelia de Lange syndrome

663. The most common cause of obstructive or noncommunicating hydrocephalus is:
   a) Subarachnoid hemorrhage
   b) Vein of Galen malformation
   c) Tuberculous meningitis
   d) Pneumococcal meningitis
   e) Aqueductal stenosis

664. The most common cause of nonobstructive or communicating hydrocephalus is:
   a) Leukemic infiltrates
   b) Subdural hemorrhage
   c) Subarachnoid hemorrhage
   d) Pneumococcal meningitis
   e) Tuberculous meningitis

Match the following conditions with different findings (665-667):

665. Dandy-Walker malformation
666. Chiari malformation (type II)
667. Aqueductal stenosis

   a) Foreshortened occiput, cerebellar tonsils protruding downward into the cervical canal
   b) Prominent occiput, large posterior fossa cyst
   c) Large head, dilated lateral ventricles

668. All of the following feature are usually present in patients with Chiari malformation (type I) **except**:
   a) Hydrocephalus
   b) Urinary frequency
   c) Recurrent headache
   d) Neck pain
   e) Progressive lower extremity spasticity

669. A toddler appears with history of sudden onset of ataxia, refuses to walk, nausea, and vomiting. The child experienced a similar attack during the physical examination. The physician noted horizontal nystagmus. The attack lasted for about 1 minute. The child appears frightened and pale but is able to verbalize without lethargy, drowsiness, and loss of consciousness. The most likely diagnosis is:
   a) Simple syncope
   b) Breath-holding spells
   c) Prolonged QT syndrome
   d) Shuddering attacks
   e) Benign paroxysmal vertigo

670. A 2-year-old girl appears with history of a loss of consciousness, generalized cyanosis, clonic jerky movements, and opisthotonos. She had two episodes within 1 hour. The child was upset before the episode. She had a brief, shrill cry before the apneic episodes. The preferred therapy is:
   a) Diphenhydramine

b) Atropine
c) Propranolol
d) Phenytoin
e) Reassurance

671. A child wakes up shortly after the onset of sleep with coughing paroxysm. He is plethoric, frightened, and agitated, followed by loss of consciousness, clonic muscle contractions, vertical upward gaze, generalized flaccidity, and urinary incontinence. He recovered within seconds and consciousness is restored within a few minutes. Mother denies a prior episode. He is a known asthmatic. The most likely diagnosis is:
a) Seizure disorder
b) Cyanotic spells
c) Pallid spells
d) Simple syncope
e) Cough syncope

672. The most important causes of significant headache in children are the following **except**:
a) Refractive errors
b) Stress
c) Migraine
d) Psychologic factors
e) Increased intracranial pressure

673. The most prevalent type of migraine in children is:
a) Common migraine
b) Complicated migraine
c) Migraine variants
d) Classic migraine
e) Idiopathic

Match the different types of migraine and their clinical manifestations (674-677):

674. Common migraine
675. Classic migraine
676. Migraine variants
677. Complicated migraine

a) The child develops neurologic signs (e.g., diplopia, third-nerve palsy, or hemiplegia) during a headache that persist after termination of the headache.
b) The child develops cyclic vomiting (i.e., recurrent bouts of severe vomiting, sometimes monthly) or appears with confusion, disorientation, hyperactivity, unresponsiveness, vomiting, lethargy, and memory disturbances.
c) The child has history of throbbing or pounding headache either unilateral, bifrontal or temporal regions. The headache is associated with severe nausea and vomiting. The migraine is not associated with an aura. A strong family history, especially on the maternal side.
d) The child has history of an aura (e.g., blurred vison, scotoma, flashes of light) before the onset of headache. The child has distortions of body image, perioral paresthesia, and numbness of the hands and feet. After the aura, the child appears with clinical manifestations that is described in item (c).

678. The treatment for migraine may include all of the following medications **except**:
a) Acetaminophen
b) Ibuprofen
c) Dimenhydrinate
d) Chlorpromazine
e) Monosodium glutamate

679. The mode of inheritance of patients with tuberous sclerosis is:
a) Autosomal recessive
b) Autosomal dominant
c) X-linked recessive

d) X-linked dominant
e) Multifactorial

680. The hallmark of neurofibromatosis (NF-1) is:
a) Optic glioma
b) Sphenoid dysplasia
c) Iris Lisch nodules
d) Café-au-lait spots
e) A first-degree relative with NF-1

681. The most distinctive finding in neurofibromatosis (NF-2) is:
a) Bilateral acoustic neuromas
b) Posterior subcapsular cataracts
c) Skin neurofibromas
d) A sibling with NF-2
e) A parent with NF-2

682. The most common orthopedic manifestation of neurofibromatosis (NF-1) is:
a) Scoliosis
b) Lordosis
c) Kyphosis
d) Sphenoid dysplasia
e) Cortical thinning of long bones

683. The preferred therapy in patients with port-wine stain is:
a) Cryotherapy
b) Excision
c) Excision and skin grafting
d) Interferon-alpha
e) Flashlamp-pulsed laser therapy

684. A child has Sturge-Weber disease. He has normal mental development. His seizure activities are well-controlled with anticonvulsant medication. The preferred therapy for this patient is:
a) Hemispherectomy
b) Conservative
c) Frontal lobectomy
d) Parietal lobectomy
e) Temporal lobectomy

685. A 10-year-old boy with Down syndrome suddenly develops neck pain, head tilt, and gait abnormalities. A physical examination reveals weakness, hyperreflexia, and spasticity. The preferred therapy is:
a) Intravenous immunoglobulin
b) Intravenous ceftriaxone
c) Intravenous cefuroxime
d) Low-dose intravenous methylprednisolone
e) High-dose intravenous methylprednisolone

686. A lateral x-ray of the neck in a patient with Down syndrome reveals a distance of 4.0 mm between the odontoid process of the axis and the anterior arch of the atlas. The most likely diagnosis is:
a) The child has atlantoaxial instability.
b) The child has instability between C2 and C3 vertebrae.
c) The child has instability between C3 and C4 vertebrae.
d) The child has instability between C4 and C5 vertebrae.
e) The child has no atlantoaxial instability.

687. A routine physical examination of a newborn reveals lipoma in the midline of the lower back. The most likely diagnosis is:
a) Syringomyelia
b) Spinal cord tumor
c) Diastematomyelia

d) Tethered cord
e) Syringobulbia

688. A 16-year-old girl appears with low back pain and paresthesias of the lower extremities for the last 24 hours. She experienced a viral infection prior to this episode. She also has sphincter disturbances. A physical examination reveals weak and flaccid lower extremities; a sensory level in the midthoracic region; decreased pain, temperature, and light touch sensations; normal vibration sense and joint position. CSF study reveals moderate lymphocyte pleocytosis and a normal protein level. The most likely diagnosis is:
a) Spinal cord tumor
b) Guillain-Barre syndrome
c) Epidural abscess
d) Transverse myelitis
e) Devic disease

689. All of the following infections can cause transverse myelitis **except**:
a) Herpes
b) Epstein-Barr virus
c) Varicella
d) Chlamydia trachomatis
e) Mycoplasma pneumoniae

Match the different diseases with different opthalmologic examination findings (690-694):

690. Retinopathy of prematurity
691. Retinoblastoma
692. Retinitis pigmentosa
693. Tuberous sclerosis
694. Diabetic retinopathy

a) Retinal phakomas
b) Neovascularization and proliferation of fibrovascular tissue on the retina, extending into the vitreous.
c) Progressive retinal degeneration characterized by pigmentary changes, arteriolar attenuation, some degree of optic atrophy
d) White pupillary reflex and strabismus
e) Ridge formation, extraretinal vascular tissue

695. The preferred therapy in patients with infantile glaucoma is:
a) Furosemide
b) Acetazolamide
c) Topical steroid therapy
d) Systemic steroid therapy
e) Surgery

696. The classic triad of signs and symptoms in patients with infantile glaucoma include:
a) Corneal edema, photophobia, and visual impairment
b) Epiphora, photophobia, and visual impairment
c) Corneal enlargement, photophobia, and conjunctival injection
d) Epiphora, corneal edema, and photophobia
e) Epiphora, photophobia, and blepharospasm

697. The most common tumor of the optic nerve in children is:
a) Optic nerve glioma
b) Optic nerve Lipoma
c) Optic nerve neuromas
d) Optic nerve meningioma
e) Optic nerve blastoma

698. A child is diagnosed with juvenile-onset (insulin-dependent) diabetes mellitus and heterodegenrative optic atrophy. The most likely associated condition is:
a) Cardiac anomaly

b) Nephrotic syndrome
c) Hepatic abnormalities
d) Herpes infection
e) Sensorineural hearing loss

699. The most common benign tumor of the orbit is:
a) Lipoma
b) Fibroma
c) Neuroma
d) Osteoma
e) Hemangioma

700. The malignant tumors of the orbit include all of the following **except**:
a) Rhabdomyosarcoma
b) Lymphosarcoma
c) Adenocarcinoma
d) Extension of retinoblastoma
e) Metastatic neuroblastoma

701. The most common hip disorder in adolescent is:
a) Transient synovitis
b) Legg-Calve-Perthes disease
c) Septic arthritis
d) Osteomyelitis
e) Slipped capital femoral epiphysis

702. The most common form of scoliosis is:
a) Congenital
b) Traumatic
c) Bone tumor
d) Spinal cord tumor
e) Idiopathic

703. A routine physical examination of a patient reveals idiopathic scoliosis 50 degrees. The preferred therapy is:
a) Observation
b) Bracing
c) Physical exercise
d) Reevaluation after 6 months
e) Surgery

704. The school screening examination for scoliosis is:
a) Asymmetric shoulder height
b) Asymmetric waist line
c) Asymmetric elbow-to-flank distance
d) Vertebral rotation
e) Asymmetry of the posterior chest wall on forward bending

705. The preferred diagnostic study in patients with idiopathic scoliosis is:
a) MRI
b) CT scan
c) Supine posteroanterior x-ray
d) Prone posteroanterior x-ray
e) Standing posteroanterior x-ray

706. All of the following statements are true regarding treatment of idiopathic scoliosis **except**:
a) The progressive adolescent scoliosis are usually treated with bracing and surgical correction.
b) The chiropractic therapy can improve the outcome of scoliosis.
c) Most orthopedic surgeons recommend a trial of brace therapy for immature patients with curves less than 40 degrees.

d) Bracing does not correct curvature but can improve Cobb angle during active brace therapy.
e) Surgical therapy is indicated when curves greater than 45 degrees.

707. The preferred therapy in patients with clavicular fracture is:
a) Shoulder cast
b) Internal fixation
c) Open reduction
d) Arm and shoulder cast
e) Figure-of-eight clavicle strap

708. The most common type of elbow dislocation is:
a) Anterior dislocation
b) Posterior dislocation
c) Superior dislocation
d) Inferior dislocation
e) Lateral dislocation

709. The mechanism of supracondylar humeral fracture is:
a) Falling backward onto the outstretched arm with the elbow flexed
b) Falling forward onto the outstretched arm with the elbow flexed
c) Falling backward onto the outstretched arm with the elbow extended
d) Falling laterally onto the flexed arm with the elbow flexed
e) Falling laterally onto the extended arm with the elbow extended

710. The most common coexisting injury in patients with supracondylar humeral fracture is:
a) Median nerve injury
b) Radial nerve injury
c) Ulnar nerve injury
d) Brachial artery injury
e) Compartment syndrome

711. During the initial evaluation of the injured extremity, the first priorities are to maintain:
a) Nerve and skeletal stability
b) Nerve and muscle stability
c) Muscle and skeletal stability
d) Nerve and vascular stability
e) Vascular and skeletal stability

712. The most common site of epiphyseal injury is:
a) Proximal radius
b) Distal radius
c) Proximal tibia
d) Distal tibia
e) Distal femur

713. A patient has shoulder injury and complains of radiating pain down the arm. The most likely associated injury is at the site of:
a) Proximal humerous
b) Distal humerous
c) Clavicle
d) Scapula
e) Neck

714. A 16-year-old boy appears with a history that his shoulder had "popped out of place" while playing football. He was tackling another player only with his arm. The most likely diagnosis is:
a) Posterior dislocation of the shoulder
b) Anterior dislocation of the shoulder
c) Fracture of the lateral end of clavicle
d) Fracture of the proximal humeral bone
e) Acromioclavicular separation

715. A football player appears in ER with pain in the right shoulder. He collided with another player and the point of contact was the distal part of the right clavicle. A physical examination reveals tenderness at the right acromioclavicular joint and step-off between the distal clavicle and the acromion. The most likely diagnosis is:
   a) Fracture of the proximal humeral bone
   b) Fracture of the distal right clavicle
   c) Fracture of the acromion
   d) Anterior dislocation of the right shoulder
   e) Acromioclavicular separation of the right shoulder

716. All of the following statements are true about atlantoaxial instability (AAI) **except**:
   a) About 40% of patients younger than 21 years of age with Down syndrome have AAI.
   b) Symptomatic AAI is rare in patients with Down syndrome
   c) Asymptomatic AAI is not a risk factor for symptomatic AAI.
   d) Special Olympics require atlantoaxial x-ray in asymptomatic individuals with Down syndrome.
   e) All symptomatic patients (i.e., upper motor neuron or posterior column dysfunction) should be evaluated with radiographs.

717. All of the following are features of sports-related heat stroke **except**:
   a) Dry, hot skin
   b) Altered mental status
   c) Related to intense exertion
   d) Profuse sweating
   e) Body temperature is more than 104°F

718. The therapy for patients with heat stroke includes all of the following **except**:
   a) Removal of excess clothing
   b) Ice water bathing
   c) Airway
   d) Breathing
   e) Intravenous fluid should be given at a rate of 500 mL/m^2 in the first hour with dextrose and saline solution.

719. All of the following measures can prevent sports-related dehydration and heat illness **except**:
   a) Athletes should be well hydrated before participation in sports.
   b) Athletes should drink every 20 minutes during sports.
   c) Athletes should have free access to cold water.
   d) Salt pills should be used to avoid hyponatremia.
   e) Fluid (i.e., electrolytes and carbohydrate) should be reserved for sports lasting more than 1 hour.

720. Amenorrheic athletes have an increased risk of the following condition than their eumenorrheic athletic peers:
   a) Heart problems
   b) Uterine abnormalities
   c) Ovarian abnormalities
   d) Osteopetrosis
   e) Osteopenia

721. The treatment for hypothalamic amenorrhea in athletes includes all of the following **except**:
   a) Reduced exercise
   b) Increased energy intake
   c) Increased calcium intake
   d) Increased vitamin D intake
   e) Estrogen replacement therapy

722. The current recommended daily allowance for calcium in women with amenorrhea is:
   a) 1200 mg/day
   b) 1500 mg/day
   c) 1800 mg/day
   d) 2000 mg/day
   e) 2200 mg/day

723. The following skin infection is a contraindication to wrestling:
   a) Contact dermatitis
   b) Eczema
   c) Herpes simplex
   d) Superficial fungal infections
   e) Staphylococcal furunculosis

724. The most common musculoskeletal injury at the time of preseason examinations is:
   a) Neck injury
   b) Shoulder injury
   c) Elbow injury
   d) Wrist injury
   e) Knee injury

725. A 17-year-old boy has "tennis elbow". The boy has the following injury:
   a) Injury to the medial epicondyle
   b) Injury to the lateral epicondyle
   c) Injury to the anterior part of the elbow
   d) Injury to the posterior part of the elbow
   e) Injury to the elbow caused by tennis ball

Match the skeletal dysplasias and their associated anomalies (726-730):

726. Achondroplasia
727. Osteogenesis imperfecta
728. Robinow syndrome
729. Beemer-Langer syndrome
730. Saldino-Noonan

   a) Renal cysts
   b) Hydrocephalus
   c) Omphalocele
   d) Blue sclera
   e) Genital hypoplasia

731. A child appears with cerebellar ataxia, posterior column dysfunction, opthalmoplegia, loss of deep tendon reflexes, and chronic cholestasis. The most likely diagnosis is:
   a) Hepatic encephalopathy
   b) Vitamin A deficiency
   c) Vitamin D deficiency
   d) Vitamin E deficiency
   e) Vitamin K deficiency

732. A 10-year-old girl has vitamin E deficiency. The following laboratory investigation is suggestive of this diagnosis:
   a) The ratio of serum vitamin E to total serum protein 0.4.
   b) The ratio of serum vitamin E to total serum glucose is 1.0.
   c) The ratio of serum vitamin E to total serum lipid is 0.3.
   d) The ratio of serum vitamin E to vitamin D level is 0.9.
   e) The ratio of serum vitamin E to vitamin A level is 0.5.

733. A child appears with a chronic abdominal pain, vomiting, abdominal distension, diarrhea, steatorrhea, and megaloblastic anemia. She underwent intestinal operation a few months prior to this episode. The most likely diagnosis is:
   a) Disaccharidase deficiency
   b) Vitamin B_{12} deficiency
   c) Protein-losing enteropathy
   d) Fat malabsorption
   e) Stagnant loop syndrome

734. The preferred therapy for children with bacterial overgrowth syndrome is:
   a) Ciprofloxacin
   b) Tetracycline
   c) Gentamicin
   d) Colistin
   e) Metronidazole

735. The breast milk has all of the following components **except**:
   a) High levels of IgA
   b) High levels of nucleotides
   c) High levels of leukocytes
   d) High levels of vitamin K
   e) Epidermal growth factor

736. The following complication commonly occurs in infants receiving TPN who had terminal ileal resection:
   a) Renal stones
   b) Gallstones
   c) Venous thrombosis
   d) Vitamin deficiency
   e) Hyperoxaluria

737. The long-term complications of TPN include all of the following findings **except**:
   a) Thrombosis
   b) Hepatotoxicity
   c) S. epidermidis infections
   d) Gallstones
   e) Bleeding

738. A child developed short bowel syndrome after intestinal resection due to volvulus. Enteral feeding started very slowly along with TPN. The child was tolerating enteral feeding well. Suddenly, he develops bloody diarrhea. The most likely diagnosis is:
   a) Recurrence of volvulus
   b) Malrotation
   c) Nectrotizing enterocolitis
   d) Feeding colitis
   e) Large bowel obstruction

739. The celiac disease is due to the following food component:
   a) Lactose
   b) Galactose
   c) Triglycerides
   d) Sucrose
   e) Gluten

740. The most common symptom of celiac disease is:
   a) Failure to thrive
   b) Diarrhea
   c) Irritability
   d) Vomiting
   e) Anorexia

741. The most common sign of celiac disease is:
   a) Wasted muscles
   b) Body weight less than 25th percentile
   c) Abdominal distension
   d) Edema
   e) Height less than 25th percentile

742. The gold standard to make the diagnosis of celiac disease is:
   a) Antigliadin IgG antibodies in serum
   b) Antigliadin IgA antibodies in serum
   c) Antiendomysial antibodies in serum
   d) Antibodies to tissue transglutaminase (tTG)
   e) Small intestinal biopsy

743. The preferred therapy for patients with celiac disease is:
   a) One year gluten-free diet
   b) Three years gluten-free diet
   c) Six years gluten-free diet
   d) Lifelong, strict gluten-free diet
   e) Corticosteroids

744. A 10-month-old boy develops perianal abscess. He is otherwise asymptomatic. The preferred therapy is:
   a) Incision and drainage
   b) Amoxicillin-clavulanate
   c) Gentamicin
   d) Vancomycin
   e) Reassurance

745. A 3-year-old boy appears with perianal abscess. He has a predisposing condition. The therapy includes all of the following **except**:
   a) Excision and drainage
   b) Antibodies to cover gram-negative bacteria
   c) Antibodies to cover Staphylococcus aureus
   d) Antibodies to cover fecal anaerobic organism
   e) Observation only

746. A 3-year-old girl appears with prolapse of the rectum during defecation. This is the first episode. She has no pain. A physical examination reveals a bright red mass that is 6 cm long protruding through the anus. Neurological examination is normal. The therapy may include all of the following **except**:
   a) Surgical placement of a subcutaneous ring-like band in the perianal area.
   b) Reduction of protrusion with a gentle pressure with warm compress.
   c) Stool softeners in case of constipation
   d) Laxative in case of constipation
   e) Treatment for intestinal parasites when present

747. A 2-year-old boy appears with procidentia. He had meninomyelocele that was repaired after birth. He has a ventriculoperitoneal shunt. He has procidentia for the last 6 months. The preferred therapy is:
   a) Laparotomy and internal fixation of the rectum to the presacral fascia
   b) Sclerosing agent
   c) Stool softeners
   d) Laxative
   e) Manual reduction of protrusion

748. The most common cause of malabsorption in white American children is:
   a) Biliary atresia
   b) Jejunal atresia
   c) Duodenal atresia
   d) Lactose intolerance
   e) Cystic fibrosis

749. The following newborn screening test is used to make the diagnosis of cystic fibrosis:
   a) Elevated serum levels of chymotrypsinogen
   b) Decreased serum levels of chymotrypsinogen
   c) Elevated serum levels of trypsinogen
   d) Decreased serum levels of trypsinogen
   e) Elevated serum levels of triglyceride

750. The most widely used screening test for malabsorption is:
   a) A qualitative examination of the stools for microscopic fat globules.
   b) Measurement of stool trypsinogen levels
   c) Measurement of stool chymotrypsinogen levels
   d) Measurement of stool lipase levels
   e) Measurement of stool amylase levels

751. The most commonly used test to determine pancreatic function is:
   a) Measurement of stool trypsinogen levels
   b) Measurement of stool chymotrypsinogen levels
   c) Measurement of stool bicarbonate levels
   d) Breath tests to detect CO_2 in the expired air
   e) Collection and measurement of pancreatic juice after stimulation with secretion and cholecystokinin during endoscopy

752. The most common mechanism of arterial hypoxemia in pulmonary disease is:
   a) Ventilation-perfusion mismatch
   b) Ventilation failure
   c) Perfusion abnormalities
   d) Arterial obstruction
   e) Venous obstruction

753. All of the following statements are true in infants who are sleeping supine or on their side **except**:
   a) They develop less fever at 1 month of age.
   b) They develop less stuffy nose at 6 months of age.
   c) They have less trouble sleeping at 6 months of age.
   d) They have less outpatient visits for ear infection at 1 and 2 months for sleeping supine.
   e) They have less outpatient visits for ear infection at 3 months for sleeping on their side.

754. The preferred therapy in patients with antral choanal polyps is:
   a) Surgery
   b) Intranasal steroid sprays
   c) Systemic steroids
   d) Local decongestants
   e) Systemic decongestants

755. The preferred therapy in patients with nasal polyps is:
   a) Intranasal steroid sprays
   b) Systemic steroids
   c) Local decongestants
   d) Systemic decongestants
   e) Functional endoscopic sinus surgery

756. The most common organism for common cold is:
   a) Coronavirus
   b) Adenovirus
   c) Influenza virus
   d) Rhinovirus
   e) Respiratory syncytial virus

757. The most common complication of cold is:
   a) Sinusitis
   b) Asthma
   c) Bronchiolitis
   d) Croup
   e) Otitis media

758. The best method of prevention of common cold is:
   a) Influenza vaccine
   b) Vitamin C

c) Echinacea
d) Good handwashing
e) Chicken soup

Match all the different diseases and their clinical manifestations (759-763):

759. Pertussis
760. Allergic rhinitis
761. Streptococcal nasopharyngitis
762. Foreign body in nose
763. Sinusitis

a) Persistent or paroxysmal cough
b) Nasal discharge that excoriates the nares
c) Persistent rhinorrhea or cough for more than 10-14 days, headache, facial pain, or periorbital edema
d) Itching, sneezing, and nasal eosinophilia
e) Unilateral, foul-smelling discharge

764. Prolonged use of nasal topical adrenergic medications (e.g., xylometazoline, oxymetazoline, phenylephrine) can produce the following complication:
a) Nasal ulcers
b) Hypertension
c) Tachycardia
d) Irritability
e) Rhinitis medicamentosa

765. A young woman appears with recurrent pneumothorax during menstruation. The investigation of the following organ is indicated:
a) Uterus
b) Fallopian tubes
c) Ovary
d) Vagina
e) Diaphragm

766. The most common organism isolated in patients with pneumothorax, especially in infants is:
a) S. aureus
b) S. epidermidis
c) Streptococcus pneumoniae
d) Pseudomonas aeruginosa
e) Group B Streptococcus

767. The most common cause of chylothorax is:
a) Chest injury
b) Metastatic lung disease
c) Child abuse
d) Thrombosis of thoracic duct
e) Rupture of thoracic duct during cardiac surgery

768. The mode of inheritance in patients with achondroplasia is:
a) Autosomal recessive
b) Autosomal dominant
c) X-linked recessive
d) X-linked dominant
e) Multifactorial

769. The most of inheritance in patients with Jeune syndrome is:
a) Autosomal recessive
b) Autosomal dominant
c) X-linked recessive

d) X-linked dominant
e) Multifactorial

770. All of the following findings may be present in patients with achondroplasia **except**:
a) Hydrocephalus
b) Adenotonsillar hypertrophy
c) Dilated foramen magnum
d) Reduced vital capacity
e) Muscular upper airway obstruction

771. The preferred therapy for patients with re-stenosis of coarctation of the aorta after earlier surgery is:
a) Aortic stent placement
b) Repeat surgical procedure
c) Antihypertensive agent
d) Balloon angioplasty
e) Small dose of acetyl salicylic acid daily

772. All of the following clinical manifestations are present in newborns with critical pulmonic stenosis **except**:
a) Pulmonary edema
b) Peripheral edema
c) Hepatomegaly
d) Cyanosis
e) Dyspnea

773. All of the following clinical manifestations are present in newborns with critical aortic stenosis **except**:
a) Pulmonary edema
b) Poor perfusion
c) Hepatomegaly
d) Peripheral edema
e) Hypertension

774. The most common cardiac defect in patients with autosomal dominant Holt-Oram syndrome is:
a) PDA
b) ASD
c) VSD
d) Aortic stenosis
e) Pulmonic stenosis

775. The most common cardiac defect in patients with Alagille syndrome is:
a) VSD
b) ASD
c) Mitral regurgitation
d) Tricuspid regurgitation
e) Pulmonary stenosis

776. PHACE syndrome includes all of the following features **except**:
a) **P**osterior brain fossa anomalies
b) **H**emangiomas (facial)
c) **A**rterial anomalies
d) **C**ardiac anomalies and aortic dilatation
e) **E**ye anomalies

777. CATCH 22 syndrome can be associated with the following cardiac defect:
a) Tetralogy of Fallot
b) Transposition of great vessels
c) Total anomalous pulmonary venous return
d) Tricuspid atresia
e) Hypoplastic left heart syndrome

778. CATCH 22 syndrome includes all of the following findings **except**:
   a) **C**ardiac defects
   b) **A**bnormal facies
   c) **T**hymic hypoplasia
   d) **C**left lip
   e) **H**ypocalcemia

Match all different postoperative complications with clinical manifestations (779-783):

779. Phrenic nerve injury
780. Recurrent laryngeal nerve injury
781. Sympathetic nerve chain injury
782. Hyponatremia
783. Spinal artery ischemia

   a) Horner syndrome
   b) Seizures
   c) Diaphragmatic paralysis
   d) Vocal cord paralysis
   e) Paraplegia

Match all of the following complications after cardiac surgery with different manifestations (784-788):

784. Postcoarctectomy syndrome
785. Repair of TAPVR
786. ARDS postpump syndrome
787. SIADH
788. Serous postpericardiotomy syndrome

   a) Hyponatremia
   b) Hypertension
   c) Pericardial tamponade
   d) Pulmonary hypertension
   e) Possible release of vasoactive substances

789. All of the following statements are true in patients with Wolff-Parkinson-White syndrome **except**:
   a) WPW syndrome may be associated with Ebstein anomaly.
   b) WPW syndrome is absent in patients with a normal heart.
   c) WPW syndrome is usually seen when the patient does not have tachycardia.
   d) EKG reveals a short P-R interval and slow upstroke of QRS (delta wave).
   e) The anatomic re-entrant circuit are the AV node and an accessory preexcitation pathway consisting of a muscular bridge connecting atrium to the ventricle.

790. The synthetic drug desmopressin (DDAVP) is effective in the following type of von Willebrand disease:
   a) Type 1
   b) Type 3
   c) Type 2B
   d) Type 2M
   e) Platelet-type pseudo

791. The most common cause for acute onset of thrombocytopenia in otherwise well children is:
   a) Bacterial infection
   b) Alloimmune etiology
   c) Fungal infection
   d) Viral infection
   e) Idiopathic

792. A 2-year-old boy appears with sudden onset of generalized petechiae and purpura for the last 24 hours. The boy was fine before this episode. He experienced a viral infection 2 weeks ago. A physical examination reveals petechiae and purpura all over the

body. The laboratory test results reveal platelet count is 10,000 but platelet sizes are increased. The rest of CBC results are normal including differential counts. The bone marrow examination reveals increased numbers of megakaryocytes and the rest of the examination is normal. The preferred therapy is:

a) Antibiotics
b) Acyclovir
c) Amphotericin
d) Ribavirin
e) Intravenous immunoglobulin

793. The side effect of IVIG therapy is:

a) Thrombocytosis
b) Polycythemia
c) Leukocytosis
d) Leukopenia
e) Aseptic meningitis

794. The long-term side effects of corticosteroid therapy includes all of the following **except**:

a) Growth failure
b) Diabetes mellitus
c) Osteoporosis
d) Aseptic necrosis of bone
e) Lymphocytosis

Match the different disorders with cancer/tumor (795-799):

795. Chromosome 11p deletion
796. Chromosome 13q deletion
797. Dysplastic nevus syndrome
798. Xeroderma pigmentosa
799. Fanconi anemia

a) Squamous cell skin cancer
b) Retinoblastoma
c) Wilms tumor
d) Melanoma
e) Leukemia

Match all the following viruses with oncogenesis (800-803):

800. Epstein-Barr virus (EBV)
801. Hepatitis C virus
802. Human herpesvirus 8 (HHV 8)
803. Human papillomaviruses

a) Splenic lymphoma
b) B-cell lymphoproliferative disease
c) Kaposi sarcoma
d) Cervical cancer

804. A newborn with Down syndrome develops high leukocyte counts, blast cells in the peripheral blood, anemia, and thrombocytopenia. A physical examination reveals hepatosplenomegaly. The following statement is true about this patient:

a) The patient needs chemotherapy immediately.
b) The patient should receive chemotherapy after bone marrow examination.
c) A close follow-up is indicated because 70-80% will develop leukemia within 6 months of life.
d) A close follow-up is indicated because 40-60% will develop leukemia within 12 months of life.
e) A close follow-up is indicated because 20-30% will develop leukemia within the first few years of life.

Match all of the following diseases with brain tumors (805-809):

805. Cowden syndrome
806. Neurofibromatosis 1
807. Neurofibromatosis 2
808. Tuberous sclerosis
809. von Hippel-Lindau disease

a) Hemangioblastoma
b) Vestibular schwannomas
c) Optic pathway gliomas
d) Subependymal giant cell astrocytomas
e) Dysplastic gangliocytoma of the cerebellum (Lhermitte-Duclos disease)

810. A child appears with ataxia and "dancing eyes and dancing feet". The most likely diagnosis is:
a) Pheochromocytoma
b) Neuroblastoma
c) Medulloblastoma
d) Pituitary tumor
e) Cerebellar astrocytoma

811. The most common site of neuroblastoma is:
a) Brain
b) Neck
c) Chest
d) Abdomen
e) Pelvis

812. The most common sites of metastasis in patients with neuroblastoma are all of the following **except**:
a) Lungs
b) Long bones
c) Skull
d) Bone marrow
e) Liver

813. The following stage refers to children younger than 1 year of age with neuroblastoma is:
a) Stage 1
b) Stage 2
c) Stage 3
d) Stage 4
e) Stage 4S

814. A child appears with abdominal discomfort, proptosis and ecchymoses of both eyes. The preferred diagnostic study is:
a) CT scan of abdomen
b) CT scan of eyes
c) MRI of abdomen
d) Biopsy of tumor tissue
e) Decreased homovanillic acid (HVA) and vanillylmandelic acid (VMA) in the urine

815. A bone marrow examination reveals rosettes of cells surrounding an inner mass of fibrillary material. The most likely diagnosis is:
a) Neuroblastoma
b) Pheochromocytoma
c) Ganglioneuroma
d) Osteosarcoma
e) Chronic myelogenous leukemia

816. A 6-year-old girl appears with lethargy, malaise, abdominal pain, fever, oliguria, and edema for the last 48 hours. She experienced pharyngitis 10 days prior to this episode. A physical examination reveals hypertension, edema, and mild abdominal tenderness. Urinalysis reveals RBCs, RBC cast, polymorphonuclear leukocytes, and proteinuria. The serum C3 level is reduced. All of the following statements are true about this condition **except**:
a) Complete recovery occurs in more than 95% of cases.
b) The risk of recurrence is extremely high.
c) A 10-day course of oral penicillin therapy is recommended to limit the spread of the specific strain of the organism.
d) Antibiotic therapy does not affect the natural history of the disease.
e) The diagnosis must be questioned in patients with chronic renal dysfunction.

817. A 10-year-old boy is diagnosed to have acute poststreptococcal glomerulonephritis. He has acute nephritis syndrome, evidence of recent streptococcal infection, and a low serum C3 level. The boy does not need renal biopsy. However, renal biopsy is indicated in all of the following conditions **except**:
a) Acute renal failure
b) Nephrotic syndrome
c) Absence of evidence for streptococcal infection
d) Abnormal complement levels
e) Persistence of hematuria, proteinuria, diminished renal function and/or a low serum C3 level persist more than 2 months after onset.

818. A 3-year-old girl appears with sudden onset of pallor, weakness, lethargy, irritability, and oliguria for the last 24 hours. She experienced fever, vomiting, diarrhea, and abdominal pain 7 days prior to this episode. A physical examination reveals petechiae, edema, dehydration, marked irritability, and hepatosplenomegaly. The laboratory findings include all of the following results **except**:
a) Microangiopathic hemolytic anemia
b) Positive Coombs test
c) Thrombocytopenia
d) Acute renal failure
e) Leukocytosis

819. The therapy for patients with hemolytic-uremic syndrome includes all of the following **except**:
a) Antithrombotic agent
b) Antihypertensive agent
c) Aggressive nutrition
d) Meticulous management of fluid and electrolytes
e) Early peritoneal dialysis

820. Antibiotic therapy should be avoided in patients with enteritis secondary to E. coli 0157 : H7 because the patient can develop the following complication:
a) Pyloric stenosis
b) Bloody stools
c) NEC
d) Ulcerative colitis
e) Hemolytic-uremic syndrome

821. The following therapy is contraindicated in patients with hemolytic-uremic syndrome caused by Streptococcus pneumoniae:
a) Peritoneal dialysis
b) Plasmaphresis
c) Nutrition
d) Antihypertensive agent
e) Fluid and electrolytes

822. All of the following medications can cause hemorrhagic cystitis **except**:
a) Penicillins
b) Busulfan
c) Thiotepa
d) Cyclophosphamide
e) Acyclovir

823. All of the following conditions can cause transient proteinuria **except**:
   a) Exercise
   b) Dehydration
   c) Congestive heart failure
   d) Seizure
   e) Temperature greater than 100.4°F

824. The most common cause of persistent proteinuria in school-aged children and adolescents is:
   a) Glomerular proteinuria
   b) Tubular proteinuria
   c) Nephrotic syndrome
   d) Acute glomerulonephritis
   e) Orthostatic proteinuria

825. A 9-year-old girl appears with persistent proteinuria that is detected in a routine urinalysis. She is asymptomatic. The preferred method to make the diagnosis is:
   a) Renal scan
   b) VCUG
   c) Renal ultrasonography
   d) Urine culture
   e) Collection of urine in both resting (supine) and active (upright) position, especially a split 24-hour urine collection.

826. The most common cause of nephrotic syndrome in children is:
   a) Membranous nephropathy
   b) Minimal change disease
   c) Focal segmental glomerulosclerosis
   d) Mesangial proliferation
   e) Membranoproliferative glomerulonephritis

827. A 4-year-old boy appears with mild edema around the eyes and lower extremities for the last 24 hours. He also complains of anorexia, diarrhea, and abdominal pain. He has idiopathic nephrotic syndrome. All of the following laboratory findings are suggestive of this disease **except**:
   a) Urinalysis reveals 3+ or 4+ proteinuria
   b) Spot urine protein to creatinine ratio is 1.2
   c) Serum creatinine level is normal.
   d) Serum albumin level is less than 2.5 g/dL.
   e) Serum C3 and C4 levels are normal.

828. All of the following are clinical features of patients with Bartter syndrome **except**:
   a) Triangular facies
   b) Drooping mouth
   c) Small ears
   d) Signs of dehydration
   e) Large eyes with strabismus

829. All of the following are laboratory findings in patients with Bartter syndrome **except**:
   a) Severe hypokalemia
   b) Metabolic alkalosis
   c) Hypercalcuria
   d) Abnormal renal functions
   e) Elevated serum renin, aldosterone, and prostaglandin E levels

830. The therapy in patients with Bartter syndrome includes all of the following **except**:
   a) High dose of potassium
   b) Indomethacin
   c) Adequate hydration
   d) Adequate nutrition
   e) Sodium bicarbonate

831. The mode of inheritance in patients with Bartter syndrome is:
   a) Autosomal recessive
   b) Autosomal dominant
   c) X-linked recessive
   d) X-linked dominant
   e) Multifactorial

832. The most common organism responsible for urinary tract infections (UTIs) is:
   a) Adenovirus
   b) E. coli
   c) Klebsiella
   d) Proteus
   e) Staphylococcus saprophyticus

833. A 16-year-old girl appears with urgency, dysuria, frequency, bladder and pelvic pain. The pain is relieved after voiding. The urine culture is negative. The preferred therapy is:
   a) Ampicillin
   b) Cefotaxime
   c) Gentamicin
   d) Ceftriaxone
   e) Bladder hydrodistension and laser ablation

834. The following statement is **not** true in patients with acute hemorrhagic cystitis:
   a) Rarely caused by E. coli
   b) Adenovirus types 11 and 21 can cause cystitis
   c) Adenovirus cystitis is more common in males than females.
   d) Adenovirus cystitis is a self-limiting condition.
   e) Hematuria lasts about 4 days in patients with adenovirus cystitis.

835. All of the following statements are true about collection of urine to make the diagnosis of UTI **except**:
   a) Urine samples can be kept at room temperature up to 2 hours before plating.
   b) A midstream urine sample is good in toilet-trained children.
   c) In uncircumcised males, prepuce should be retracted before collection of urine samples.
   d) A catheterized specimen is more reliable to diagnose UTI.
   e) In infants, a sterile collection bag specimen can give falsely positive UTI.

836. All of the following statements suggest UTI **except**:
   a) A midstream urine sample reveals greater than 100,000 colonies of E. coli in a symptomatic boy.
   b) A midstream urine sample reveals greater than 10,000 colonies of E. coli in a symptomatic child.
   c) A sterile collection bag urine reveals greater than 100,000 colonies of Klebsiella in an asymptomatic infant.
   d) A catheterized specimen reveals greater than 100,000 colonies of E. coli.
   e) A suprapubic urine sample reveals greater than 10,000 colonies of Klebsiella in a symptomatic child.

837. The side effect of fluroquinolones in children is:
   a) Hepatic damage
   b) Gastric ulcer
   c) Duodenal ulcer
   d) Pancreatitis
   e) Cartilage damage

838. The preferred therapy for patients with E. coli cystitis is:
   a) Nitrofurantoin
   b) Amoxicillin
   c) Pencillin
   d) Azithromycin
   e) Trimethoprim-sulfamethoxazole

839. The preferred therapy for patients with Klebsiella-Enterobacter cystitis is:
   a) Azithromycin
   b) Ampicillin
   c) Penicillin
   d) Erythromycin
   e) Nitrofurantoin

840. The preferred oral therapy for patients with E. coli pyelonephritis is:
   a) Cefixime
   b) Ampicillin
   c) Amoxicillin
   d) Azithromycin
   e) Erythromycin

841. The preferred oral therapy for patients with Pseudomonas pyelonephritis is:
   a) Ciprofloxacin
   b) Nitrofurantoin
   c) Ampicillin
   d) Azithromycin
   e) Erythromycin

842. The preferred intravenous therapy for patients with acute pyelonephritis is:
   a) Ampicillin
   b) Gentamicin
   c) Cefotaxime
   d) Ceftriaxone
   e) Ceftazidime

843. The following medication should **not** be used in children with a febrile UTI:
   a) Ampicillin
   b) Cefixime
   c) Nitrofurantoin
   d) Gentamicin
   e) Trimethoprim-sulfamethoxazole

844. The preferred prophylactic therapy against recurrent UTIs is:
   a) Amoxicillin
   b) Cefalexin
   c) Ampicillin
   d) Erythromycin
   e) Trimethoprim-sulfamethoxazole

845. A child is being treated for UTI. The follow-up urine culture should be performed:
   a) 48 hours after onset of antibiotic therapy
   b) 72 hours after onset of antibiotic therapy
   c) Immediately after the termination of antibiotic therapy
   d) 1 week after the termination of antibiotic therapy
   e) 1 month after the termination of antibiotic therapy

846. A child was treated for UTI. A long-term follow-up urine cultures should be performed regardless of predisposing anatomic abnormalities. The follow-up should be as follows:
   a) Periodically for 6 months, if the child is asymptomatic.
   b) Periodically for 1-2 years, even when the child is asymptomatic.
   c) Periodically for 3-5 years, even when the child is asymptomatic.
   d) Periodically for 6-8 years, even when the child is asymptomatic.
   e) Periodically for 3 months, if the child is asymptomatic.

847. The preferred nonsurgical therapy for boys with persistent physiologic or pathologic phimosis is:
   a) Estrogen cream locally
   b) Progesterone cream locally
   c) Testosterone cream locally
   d) Corticosteroid therapy orally
   e) Corticosteroid cream locally

848. A child appears in ER with pain and swelling of the glans penis. He has phimosis and his father forcefully retracts the foreskin. The preferred therapy is:
   a) Estrogen cream locally
   b) Corticosteroid cream locally
   c) Corticosteroid therapy orally
   d) Lubrication of the foreskin and glans, and then compressing the glans and push the phimotic ring distal to the coronal sulcus.
   e) Emergency circumcision

849. The following disease can cause galactorrhea:
   a) Hypothyroidism
   b) Hyperthyroidism
   c) Hypopituitarism
   d) Hypoparathyroidism
   e) Hyperparathyroidism

Match different diagnosis with different types of nipple discharge (850-853):

850. Papilloma
851. Mastitis
852. Galactorrhea
853. Duct ectasia

   a) Purulent
   b) Watery
   c) Bloody
   d) Milky

854. All of the following conditions can cause serous/serosanguineous nipple discharge **except**:
   a) Cancer
   b) Duct ectasia
   c) Fibrocystic changes
   d) Galactorrhea
   e) Intraductal papilloma

855. All of the following features may be present in infants with congenital hypopituitarism **except**:
   a) Apnea
   b) Cyanosis
   c) Hyperglycemia
   d) Seizures
   e) Elevation of both conjugated and unconjugated bilirubins

856. A newborn has normal size and weight at birth. A physical examination reveals round head, short and broad face, prominent frontal bone, small, depressed, and saddle-shaped nose, bulging eyes, underdeveloped chin and mandible, short neck, small hands and feet, and high-pitched cry. The most likely diagnosis is:
   a) Congenital syphilis
   b) Down syndrome
   c) Congenital hypopituitarism
   d) Congenital hypothyroidism
   e) Silver Russell syndrome

857. A child has growth hormone deficiency type 1. This condition can be due to large deletions in growth hormone gene cluster on the following chromosome:
   a) Chromosome 1
   b) Chromosome 7
   c) Chromosome 9
   d) Chromosome 15
   e) Chromosome 17

858. The definitive diagnosis of growth hormone (GH) deficiency can be made by the following test results:
   a) CT scan reveals destroyed sella.
   b) MRI reveals enlarged, ballooning, and calcification of sella.
   c) X-ray skull reveals destroyed sella.
   d) X-ray reveals markedly delayed skeletal maturation.
   e) Peak levels of growth hormone are less than 10 μg/L after two provocative tests.

Match the different causes of hyponatremia with intravascular volume and urine sodium (859-862):

859. Pseudohyponatremia
860. SIADH
861. Cerebral salt wasting
862. Systemic dehydration

   a) Low intravascular volume and low urine sodium
   b) Low intravascular volume and very high urine sodium
   c) High intravascular volume and high urine sodium
   d) Normal intravascular volume and normal urine sodium

863. The most common cause of Addison disease is:
   a) Tuberculosis
   b) Hemorrhage
   c) Autoimmune
   d) AIDS
   e) Waterhouse-Friderichsen syndrome

864. The most infectious cause of Addison disease is:
   a) Streptococcus pneumoniae
   b) Meningococcus infection
   c) Staphylococcus aureus
   d) HIV infection
   e) Tuberculosis

865. A newborn infant appears with lethargy, poor feeding, and vomiting. Sepsis work up is performed. Ampicillin and gentamicin are given. He was delivered by forcep delivery with Apgar scores were 2 and 8 at 1 and 5 minutes respectively. CBC results are unremarkable except mildly low hematocrit. The SMA 6 results reveal hypoglycemia, hyponatremia, and hyperkalemia. Blood culture has no growth. Hyperpigmentation of skin is absent. The most likely diagnosis is:
   a) Group B Streptococcus sepsis
   b) Severe perinatal asphyxia
   c) Meconium aspiration syndrome
   d) Phenylketonuria
   e) Adrenal insufficiency

866. In older children with Addison disease, all of the following manifestations are noted early in the course of the disease **except**:
   a) Orthostatic hypotension
   b) Hyponatremia
   c) Hyperkalemia
   d) Hypoglycemia
   e) Ketosis

867. The most definitive test to diagnose primary adrenal insufficiency is:
   a) Elevated serum ACTH levels
   b) Decreased serum aldosterone levels
   c) Presence of hyponatremia, hyperkalemia, hypoglycemia, and ketosis
   d) Decreased serum levels of cortisol before the administration of ACTH but significantly increased levels of cortisol after the administration of ACTH
   e) Decreased serum levels of cortisol before and after the administration of ACTH

868. A child has acute adrenal insufficiency. All of the following statements are true about therapy **except**:
   a) Intravenous administration of 5% glucose with 0.9% saline solution should be given to correct hypovolemia, hyponatremia, and hyperkalemia.
   b) Hydrocortisone sodium succinate should be given intravenously every 6-hour intervals for the first 24 hours.
   c) Fludrocortisone should be given to achieve mineralocorticoid effects.
   d) Severe hyperkalemia is treated with intravenous calcium and/or sodium bicarbonate, intrarectal potassium-binding resin (Kayexalate), or intravenous infusion of glucose and insulin.
   e) Therapy should be started immediately and vigorously.

869. Chronic overdose of fludrocortisone may cause all of the following manifestations **except**:
   a) Tachycardia
   b) Palpitations
   c) Hypertension
   d) Obesity
   e) Occasionally hypokalemia

870. A newborn male appears with abnormally small testes and penis. The most likely diagnosis is:
   a) Primary hypogonadism
   b) Secondary hypogonadism
   c) Down syndrome
   d) Fragile X-syndrome
   e) Pseudohypogonadism

871. A 12-year-old boy appears for a routine physical examination. He has high pitched voice. A physical examination reveals absence of facial, pubic, and axillary hair. He has small penis, scrotum, and testes. He has long extremities. He has fat accumulations in hips and buttocks. The distance from the symphysis pubis to the soles of the feet is much longer than that from the symphysis pubis to the vertex. The following laboratory finding can make the diagnosis:
   a) Low levels of serum FSH
   b) Low levels of serum LH
   c) Low levels of both serum FSH and LH
   d) High levels of both serum FSH and LH
   e) A random plasma testosterone level that is low.

872. The patients with Klinefelter syndrome have increased risk of all of the following conditions **except**:
   a) Breast cancer
   b) Pulmonary disease
   c) Varicose veins
   d) Testicular cancer
   e) Mediastinal germ cell tumors

873. The most common testicular lesions in patients with Klinefelter syndrome are:
   a) Spermatogenic arrest and Sertoli cell predominance
   b) Spermatogenic arrest and absence of Sertoli cells
   c) Spermatogenic arrest and Leydig cell predominance
   d) Spermatogenic arrest and absence of Leydig cells
   e) Normal spermatogenesis and absence of Leydig and Sertoli cells

874. The preferred therapy in patients with Klinefelter syndrome is:
   a) FSH
   b) LH
   c) Estrogen

d) Testosterone
e) Human chorionic gonadotropin

875. An adolescent has diabetes mellitus. He takes insulin regularly. He is going to participate in long distance running competition. The following recommendation appropriate for this boy is:
a) Increased insulin doses by 10%
b) Increased insulin doses by 15%
c) Increased insulin doses by 50%
d) Decreased insulin doses by 15%
e) Decreased insulin doses by 50%

876. The insulin dose may be adjusted during a regular exercise in patients with diabetes mellitus. The usual recommendation is:
a) Decreased insulin doses by 10-15%
b) Decreased insulin doses by 25%
c) Decreased insulin doses by 50%
d) Increased insulin doses by 10-15%
e) Increased insulin doses by 50%

877. All of the following viruses can cause prolonged pancytopenia **except**:
a) Parvovirus B19
b) Herpesviruses
c) CMV
d) Hepatitis viruses
e) Epstein-Barr virus

878. The most common hereditary bleeding disorder is:
a) Hemophilia A
b) Hemophilia B
c) Hemophilia C
d) Factor V deficiency
e) von Willebrand disease

879. A child appears with an acute abdominal pain and petechiae. A physical examination reveals diffuse abdominal tenderness and involuntary guarding. The abdominal x-ray reveals free air. The diagnosis is ruptured acute appendicitis with DIC (disseminated intravascular coagulation). The most specific laboratory finding for activation of coagulation and fibrinolysis is:
a) Fibrin degradation products (FDPs)
b) D-dimers
c) Profound thrombocytopenia
d) Blood smear reveals fragmented, burr, and helmet-shaped RBCs (schistocytes)
e) Prolonged PT and PTT

880. All of the following therapies are indicated in patients with disseminated intravascular coagulation (DIC) **except**:
a) Platelet transfusions for thrombocytopenia
b) Cryoprecipitate therapy for hyperfibrinogenemia
c) Fresh frozen plasma (FFP) is used for replacement of other coagulation factors and natural inhibitors.
d) Activated protein C (APC)
e) Heparin is occasionally effective in patients with DIC.

881. Heparin therapy is **not** indicated in all of the following conditions **except**:
a) DIC associated with purpura fulminans and acute promyelocytic leukemia.
b) Septic shock
c) Massive head injury
d) Heat stroke
e) Snake envenomation

882. A severe preterm infant has bronchopulmonary dysplasia. The infant is not responding to therapy and experiences frequent exacerbations without clear triggers. The most likely associated finding is:
a) Cyanotic heart disease
b) VSD

c) Sinusitis
d) RSV infection
e) Gastroesophageal reflux

883. A severely affected patient with bronchopulmonary dysplasia is usually benefited from a multidisciplinary team of caregivers. They include all of the following specialists **except**:
a) Audiologist
b) Pediatrician
c) Pulmonologist
d) Nutritionist
e) Developmental specialist

884. A 12-year-old girl appears with sudden loss of large amount of hair, often with combing, brushing, and washing of hair. She had appendectomy 2 months prior to this episode. She is doing well in school. A physical examination reveals absence of scalp inflammation. The hair follicles are intact. The most likely diagnosis is:
a) Toxic alopecia
b) Telogen effluvium
c) Trichotillomania
d) Traction alopecia
e) Seborrheic dermatitis

Match all different diseases with nail findings (885-889):

885. Rubinstein-Taybi syndrome
886. Nail-patella syndrome
887. Bronchiectasis
888. Plummer-Vinson syndrome
889. Arsenic poisoning

a) Small nails
b) Yellow nails
c) Leukonychia
d) Large nails
e) Koilonychia

Match all different diseases with findings of tongue and mucous membranes (890-894):

890. Fusobacterium nucleatum infection
891. Pseudomonas aeruginosa infection
892. AIDS
893. Chronic smoking
894. Vitamin A deficiency

a) Vincent gingivitis
b) Oral hairy leukoplakia
c) Scrotal tongue
d) Black hairy tongue
e) Noma neonatorum

895. A child appears with multiple painful ulcerations on the buccal, lingual, sublingual, and gingival mucosa. The lesions are well demarcated, necrotic ulcers with a gray fibrinous exudates and an erythematous halo. All of the following conditions can produce this manifestation **except**:
a) Herpes simplex virus
b) Vitamin B_{12} deficiency
c) Folate deficiency
d) Trauma
e) Emotional stress

896. The most common organism in patients with blistering distal dactylitis is:
   a) Staphylococcus epidermidis
   b) Pseudomonas aeruginosa
   c) E. coli
   d) Klebsiella
   e) Group A beta-hemolytic Streptococcus

897. The most common organism in patients with sycosis barbae is:
   a) S. aureus
   b) S. epidermidis
   c) Pseudomonas aeruginosa
   d) Proteus
   e) Group A beta-hemolytic Streptococci

898. The most common organism in patients with hot tub folliculitis is:
   a) Pseudomonas aeruginosa
   b) Proteus
   c) S. aureus
   d) E. coli
   e) Klebsiella

899. The most common organism in patients with ecthyma gangrenosa is:
   a) Candida
   b) E. coli
   c) Serratia marcescens
   d) S. aureus
   e) Pseudomonas aeruginosa

900. The most common organism in patients with erythrasma is:
   a) Actinomycetes
   b) Dermatophilus
   c) S. aureus
   d) E. coli
   e) Corynebacterium minutissimum

901. The most common organism in patients with infection secondary to total parenteral infection (TPN) is:
   a) E. coli
   b) S. aureus
   c) S. epidermidis
   d) Group B Streptococcus
   e) Pseudomonas aeruginosa

902. The water intake in term infants on day 1 usually begins at:
   a) 50-60 mL/kg/day
   b) 60-70 mL/kg/day
   c) 70-80 mL/kg/day
   d) 80-90 mL/kg/day
   e) 90-100 mL/kg/day

903. The water intake in term infants by days 2-3 is usually:
   a) 80-90 mL/kg/day
   b) 90-100 mL/kg/day
   c) 100-120 mL/kg/day
   d) 120-130 mL/kg/day
   e) 130-140 mL/kg/day

904. A low birthweight (LBW) infant is receiving greater than 100 kcal/kg/day. The infant is expected to gain weight about:
   a) 10 g/kg/day
   b) 15 g/kg/day

c) 20 g/kg/day
d) 25 g/kg/day
e) 30 g/kg/day

905. A bottle or breast feeding in premature infants usually begins at:
a) 28 wk gestational age
b) 30 wk gestational age
c) 32 wk gestational age
d) 34 wk gestational age
e) 36 wk gestational age

906. The supplementation of following vitamin can reduce bronchopulmonary dysplasia (BPD) in very low birthweight infants:
a) Vitamin A
b) Vitamin B_2
c) Vitamin B_{12}
d) Vitamin C
e) Vitamin D

907. Breast milk has all of the following advantages **except**:
a) Reduces risk of infants obesity
b) Improves neurodevelopmental outcome
c) Protects from a wide range of infections
d) Reduces risk of sudden infant death syndrome
e) Reduces risk of necrotizing enterocolitis in preterm infants

908. All of the following infants need an eye examination to screen for ROP (retinopathy of prematurity) **except**:
a) A 1250 g infant who did not require oxygen therapy.
b) A 900 g infant who required oxygen therapy for 15 days.
c) A 1490 g infant who did not required oxygen therapy.
d) A 1990 g infant who had unstable clinical course and required oxygen therapy.
e) A 2010 g infant who had unstable clinical course and required oxygen therapy.

909. The most common pattern of idiopathic apnea of prematurity is:
a) Obstructive apnea
b) Central apnea
c) Mixed apnea, obstructive apnea usually occurs before central apnea.
d) Mixed apnea, obstructive apnea usually occurs after central apnea.
e) Mixed apnea, obstructive apnea and central apnea occur simultaneously.

910. Umbilical arterial catheterization can produce all of the following complications **except**:
a) Cardiac perforation
b) Embolization
c) Thrombosis
d) Spasm
e) Perforation

911. Umbilical venous catheterization can produce all of the following complications **except**:
a) Thrombosis
b) Cardiac perforation
c) Pericardial tamponade
d) Portal hypertension
e) Gangrene of lower extremities

912. A preterm infant develops blanching in the right lower extremity after the insertion of a umbilical arterial line. The blanching persisted. The umbilical catheter is removed immediately. Persistent spasm after removal of the catheter may be relieved by:
a) Warming the affected leg
b) Warming the opposite leg
c) Cooling the affected leg

d) Topical nitroglycerin applied over the femoral artery
e) Topical nitroglycerin applied over the dorsalis pedis artery

913. A full term infant is delivered by cesarean section due to fetal distress and meconium in the amniotic fluid. The Apgar scores are 2, 3, and 5 at 1, 5, and 10 minutes respectively. The meconium was suctioned through the endotracheal tube. The infant is placed on a mechanical ventilator. The chest x-ray appears normal. His arterial PO_2 is 50 in 100% oxygen. Heart sounds are normal. The most likely diagnosis is:
a) Meconium aspiration
b) Truncus arteriosus
c) Transposition of great arteries
d) Total anomalous pulmonary venous return
e) Persistent pulmonary hypertension

914. The characteristic chest x-ray findings in patients with meconium aspiration are all of the following **except**:
a) Patchy infiltrates in both lungs
b) Coarse streaking of both lung fields
c) Flattening of the diaphragm
d) Increased anteroposterior diameter
e) Presence of air leak in both lungs

915. All of the following are complications of ECMO (extracorporeal membrane oxygenation) **except**:
a) Systemic hypotension
b) Edema
c) Bleeding
d) Seizures
e) Thromboembolism

916. A full term healthy newborn infant appears with oral thrush. She has no candidal diaper rash. Her mother is healthy. The next step in management is:
a) HIV testing of the newborn
b) HIV testing of the mother
c) Culture of the oral lesions
d) Reassurance
e) HIV testing of the mother and the newborn

Match all the different diseases with skin lesions (917-921):

917. CMV infection
918. Candida albicans infection
919. Listeria monocytogenes infection
920. Herpes simplex virus infection
921. Pseudomonas infection

a) Satellite pustules
b) 'Blueberry-muffin' rash
c) Vesicular rash
d) Ecthyma gangrenosum
e) Salmon-pink pustules

922. All of the following clinical findings are associated with violent behavior in adolescents **except**:
a) Mental retardation
b) Learning disabilities
c) Anxiety
d) Severe motor disorders
e) Attention-deficit/hyperactivity disorder

923. CRAFFT mnemonic tool that is used for evaluation by interview and questionnaire to adolescents with alcohol or drug abuse includes all of the following **except**:
   a) 'C' stands for **C**ar driving under influence of alcohol or drugs
   b) 'R' stands for **R**elaxation after using alcohol or drugs
   c) 'A' stands for **A**lone while using alcohol or drugs
   d) 'F' stands for **F**ighting after using alcohol or drugs
   e) 'T' stands for **T**rouble while using alcohol or drugs

924. A 16-year-old girl appears with ulcerative lesions in the genitalia. She is sexually active. She has genital ulcer syndromes. All of the following organisms can produce genital ulcer syndromes **except**:
   a) Treponema pallidum
   b) Haemophilus ducreyi
   c) Herpes simplex virus type 1
   d) Herpes simplex virus type 2
   e) Group B Streptococci

925. A 17-year-old girl appear with pelvic inflammatory disease. All of the following should be considered as part of the initial evaluation **except**:
   a) GYN consultation
   b) Pelvic examination
   c) Endocervical culture for gonorrhoeae
   d) Endocervical culture for Chlamydia or enzyme immunoassay (EIA)
   e) Complete blood counts with differential

926. Common variable immunodeficiency (CVID) can be produced by all of the following drugs **except**:
   a) Phenytoin
   b) Phenobarbital
   c) Gold
   d) Sulfasalazine
   e) D-penicillamine

927. Selective IgA deficiency is noted in patients treated with all of the following drugs **except**:
   a) D-penicillamine
   b) Gold
   c) Phenytoin
   d) Pyrimethamine
   e) Sulfasalazine

928. A child appears with short, pudgy hands, redundant skin, hyperextensible joints of the hands and feet but unable to extend the elbow completely. The child also has fine, sparse, light hair, and eyebrows. The most likely diagnosis is:
   a) Ataxia-telangiectasia
   b) Omenn syndrome
   c) Cartilage hair hypoplasia
   d) Hyper IgE syndrome
   e) Wiskott-Aldrich syndrome

929. Ataxia telangiectasia mutation (ATM) is maped on the long arm of chromosome:
   a) X-chromosome
   b) Y-chromosome
   c) Chromosome 7
   d) Chromosome 11
   e) Chromosome 15

930. All of the following are features of Wiskott-Aldrich syndrome **except**:
   a) Prolonged bleeding after circumcision
   b) Atopic dermatitis
   c) Recurrent infections
   d) Bloody diarrhea
   e) Initially, thrombocytopenia is due to antiplatelet antibodies.

931. The frequent major complication in patients with juvenile dermatomyositis is:
a) Bowel perforation
b) Partial lipodystrophy
c) Calcinosis
d) Aspiration pneumonia
e) Depression

932. The most common cause of nonthrombocytopenic purpura in children is:
a) Polyarteritis nodosa
b) Kawasaki disease
c) Meningococcemia
d) Listeria infection
e) Henoch-Schonlein purpura

933. Hypersensitivity angiitis occurs most likely due to:
a) Sulfonamides
b) Ampicillin
c) Pseudomonas infection
d) Listeria infection
e) Kawasaki disease

934. A child appears with episodes of intense pain, erythema, and heat in the distal portion of the lower extremities. He noticed that heat exposure triggers clinical manifestations. The most likely diagnosis is:
a) Erythromelalgia
b) Reflex sympathetic dystrophy
c) Allergic reaction
d) Leukocytoclastic vasculitis
e) Takayasu arteritis

935. The most common cause of community-acquired bacterial pneumonia is:
a) Haemophilus influenzae type b
b) Streptococcus pneumoniae
c) Staphylococcus aureus
d) Pseudomonas aeruginosa
e) E. coli

936. The most common cause of community-acquired otitis media is:
a) Pseudomonas aeruginosa
b) E. coli
c) Haemophilus influenzae type b
d) Staphylococcus aureus
e) Streptococcus pneumoniae

937. A child appears with upper respiratory tract infection and a characteristic rash. The rash appeared within 24-48 hours after the onset of symptoms. The rash began around the neck and subsequently involved trunk and extremities. The rash is diffuse, finely papular, and erythematous. The bright red discoloration of the skin, which blanches on pressure. The skin feels rough and has goose-pimple appearance. The rash is more prominent along the creases of the elbows, axillae, and groins. She also has pharyngitis. The most likely diagnosis is:
a) Kawasaki disease
b) Rubella
c) Roseola
d) Toxic shock syndrome
e) Scarlet fever

938. The most common cause of bacterial pharyngitis is:
a) Group C Streptococcus
b) Group G Streptococcus
c) Group A Streptococcus

d) Corynebacterium diphtheriae
e) Arcanobacterium haemolyticum

939. All of the following clinical manifestations suggest Streptococcal toxic shock syndrome **except**:
a) Hypotension, renal impairment, and coagulopathy
b) Hypotension, hepatic involvement, and generalized erythematous maculopapular rash
c) Hypotension, adult respiratory distress syndrome, and soft tissue necrosis
d) Hypotension, splenic involvement, and coagulopathy
e) Hypotension, soft tissue necrosis, and hepatic involvement

940. The definitive diagnosis of Streptococcal toxic shock syndrome is made by the following findings:
a) Clinical criteria and detection of group A Streptococcus from a normally sterile site
b) Clinical criteria and detection of group A Streptococcus from a normally unsterile site
c) Clinical criteria and detection of group B Streptococcus from a normally sterile site
d) Clinical criteria and detection of group D Streptococcus from a normally unsterile site
e) Clinical criteria and detection of group C Streptococcus from a normally sterile site

941. The most common clinical manifestations in patients with disseminated gonococcal infection (DGI) include all of the following **except**:
a) Carditis
b) Asymmetric arthralgia
c) Petechial acral skin lesions
d) Tenosynovitis
e) Suppurative arthritis

942. The most common site of positive culture in women with DGI (disseminated gonococcal infection) is:
a) Urethra
b) Cervix
c) Vagina
d) Pharynx
e) Rectum

943. The most common site of positive culture in men with DGI (disseminated gonococcal infection) is:
a) Rectum
b) Urethra
c) Pharynx
d) Anus
e) Eyes

944. Haemophilus influenzae requires the following factors for growth:
a) Factors I and II
b) Factors III and IX
c) Factors IV and VIII
d) Factors V and X
e) Factors II and IV

945. The most common clinical manifestation of tularemia in children is:
a) Lymphadenopathy
b) Fever
c) Ulcer
d) Pharyngitis
e) Myalgia

946. The most common clinical syndrome of tularemia in children is:
a) Ulceroglandular
b) Glandular
c) Pneumonia
d) Oropharyngeal
e) Oculoglandular

947. The preferred therapy in patients with tularemia is:
   a) Chloramphenicol
   b) Gentamicin
   c) Cefotaxime
   d) Ceftriaxone
   e) Quinolones

948. The preferred diagnostic test in patients with tularemia is:
   a) Blood culture
   b) Wound culture
   c) Gram stain
   d) Urine culture
   e) Serum agglutination test

949. All of the following statements are true about prevention of herpes infections **except**:
   a) Wresters with herpes simplex virus (HSV) cutaneous lesions should not participate in practice and competition until all lesions are healed.
   b) All children with active herpes lesions should not go to daycare or school activities even when the lesions can be covered.
   c) Children with immunodeficiencies or chronic skin diseases should not be cared by individuals with herpetic whitlow or active uncovered fever blisters.
   d) Administration of acyclovir before a known trigger factor (e.g., intense sunlight) usually prevents recurrences.
   e) Wrestling mats should be cleaned with a bleach solution at least daily.

950. A fetus is infected with varicella at 6-12 of gestation. The most likely affected part of the body is:
   a) Eyes
   b) Brain
   c) Limbs
   d) Heart
   e) Kidneys

951. A fetus is infected with varicella at 16-20 wk of gestation. The most likely affected parts of the body are:
   a) Limbs and liver
   b) Eyes and brain
   c) Heart and lungs
   d) Liver and spleen
   e) Kidneys and pancreas

952. A child appears in ER with paroxysms of fever alternating with periods of fatigue but otherwise he is relatively well. He experiences high fever, rigors, sweats, myalgias, abdominal pain, vomiting, diarrhea, pallor, and jaundice during febrile paroxysms. He migrated in USA recently. However, he received malaria chemoprophylaxis prior to his journey. The most likely diagnosis is:
   a) Typhoid fever
   b) Pneumococcal bacteremia
   c) Pneumococcal meningitis
   d) Malaria
   e) Viral gastroenteritis

953. A child pretends to drink from cup. The age of this child is:
   a) 9 months
   b) 12 months
   c) 15 months
   d) 18 months
   e) 24 months

954. A child speaks 4-6 words. The age of this child is:
   a) 12 months
   b) 15 months
   c) 18 months
   d) 24 months
   e) 30 months

955. A child scribbles. The average age of attainment is:
   a) 11 months
   b) 12 months
   c) 13 months
   d) 14 months
   e) 15 months

956. A child can put hands together in midline. The age of this child is:
   a) 2 months
   b) 3 months
   c) 4 months
   d) 5 months
   e) 6 months

957. A child bangs two cubes. The average age of attainment is:
   a) 6 months
   b) 7 months
   c) 8 months
   d) 9 months
   e) 10 months

958. A child speaks two-word sentences (e.g., "Mommy milk"). The age of this child is:
   a) 12 months
   b) 15 months
   c) 19 months
   d) 24 months
   e) 30 months

959. A child builds tower of six cubes. The age of this child is:
   a) 15 months
   b) 18 months
   c) 22 months
   d) 30 months
   e) 36 months

960. A child stares at own hand. The age of this child is:
   a) 2 months
   b) 4 months
   c) 6 months
   d) 8 months
   e) 10 months

961. A child uses stick to reach a toy. The average age of attainment is:
   a) 15 months
   b) 17 months
   c) 20 months
   d) 24 months
   e) 30 months

962. A child's asymmetric tonic neck reflex and palmer grasp are absent. The age of this child is:
   a) 4 months
   b) 6 months
   c) 8 months
   d) 10 months
   e) 12 months

Match the different age groups and growth in head circumference (963-967):

963. 0-3 months
964. 3-6 months
965. 6-12 months
966. 1-3 years
967. 4-6 years

a) 2 cm/month
b) 1 cm/month
c) 0.5 cm/month
d) 0.25 cm/month
e) 1 cm/year

Match the different age groups and average monthly weight gain (968-972):

968. 0-3 months
969. 3-6 months
970. 6-9 months
971. 9-12 months
972. 1-3 years

a) 2 lb
b) 1.25 lb
c) 1 lb
d) 13 oz
e) 8 oz

973. About 70-80% of children sleep through the night at:
a) 3 months of age
b) 6 months of age
c) 9 months of age
d) 12 months of age
e) 15 months of age

Match the different age groups and average duration of sleep (974-978):

974. 0-3 months
975. 3-12 months
976. 1-3 years
977. 3-6 years
978. 6-12 years

a) 16-20 hours per day
b) 14-15 hours per day
c) 12-14 hours per day
d) 11-12 hours per day
e) 10 hours per day

Match the different drugs and their side effects (979-983):

979. Clonidine
980. Desipramine
981. Carbamezepine
982. Valproic acid
983. Lamotrigine

a) Sudden death in high doses
b) Sudden deaths when used with stimulants
c) Hepatic failure occurs in the first few months of treatment
d) Risk of Stevens-Johnson syndrome
e) Fever, sore throat, drowsiness, blurred vision, neuromuscular disturbance, and leukopenia

984. A child's IQ test result is 60. The most likely the child has:
a) Mild mental retardation
b) Moderate mental retardation
c) Severe mental retardation
d) Profound mental retardation
e) Normal mental status

985. All of the following statements are true about pain and symptom management **except**:
a) Oral therapy is preferred.
b) Use topical anesthetics when possible
c) Use anesthetic blocks for regional pain
d) Cognitive, physical, and behavioral therapies are always used.
e) Prescribe PRN medication for constant pain

Match the use of different adjuvant medications with different clinical pain manifestations (986-990):

986. Stimulants
987. Steroids
988. Neuroleptics
989. Sedatives and hypnotics
990. Antidepressants and anticonvulsants

a) Agitation and nausea
b) Resistant pain
c) Muscle spasm or anxiety
d) Opioid-induced somnolence
e) Neuropathic pain

991. The following statement is not true about opioid use guidelines:
a) Strong medications should be saved for last use.
b) Always give constipation therapy when starting opioids.
c) Begin with weak opioids (e.g., codeine) for mild pain and replace with strong opioids (e.g., morphine) for strong or unresponsive pain.
d) Begin with short-acting opioids at regular intervals then switch to long-acting only when dose requirements have established.
e) When oral/rectal routes are no longer usuable, then subcutaneous infusions are used.

992. The plasma level of leptin is directly related to the following component:
a) Fat mass
b) Size of pituitary
c) Size of hypothalamus
d) Muscle mass
e) Bone mass

993. A 4-day-old newborn infant's serum phosphorus level is 8 mg/dL. The serum electrolytes and calcium levels are normal. The next step in management is:
   a) Decrease phosphorus intake
   b) Discontinue phosphorus intake
   c) Decrease calcium intake
   d) Decrease magnesium intake
   e) Reassurance

994. A child has decreased urine output after surgery. He has normal blood pressure and heart rate. He gained weight slightly. He has elevated urinary excretion of sodium. The next step in management is:
   a) Increased the fluid intake by giving a bolus.
   b) Increased the maintenance fluid by intravenous route.
   c) Restrict the fluid intake
   d) Resume oral hydration immediately
   e) Give furosemide

995. A child develops oliguria after surgery. She also develops tachycardia, hypotension, and decreased urinary excretion of sodium. The next step in management is:
   a) Give isotonic solution immediately through intravenous route.
   b) Start oral hydration immediately
   c) Start 10% glucose infusion immediately
   d) Start 0.45% saline solution intravenously
   e) Restrict the fluid immediately

Match all the different drugs and their effects (996-1000):

996. Amrinone
997. Dobutamine
998. Phenylephrine
999. Norepinephrine
1000. Epinephrine

   a) Can produce sudden hypertension
   b) Phosphodiesterase inhibitor
   c) Strengthens cardiac contractions
   d) Produces short-run rise in blood pressure
   e) May reduce renal perfusion

Match all different diseases and sites of injury resulting in respiratory failure (1001-1005):

1001. Werdnig-Hoffman disease
1002. Cystic fibrosis
1003. Prune-belly syndrome
1004. Guillain-Barre syndrome
1005. Croup

   a) Chest wall deformity
   b) Neuromuscular
   c) Peripheral airway obstruction
   d) Central airway obstruction
   e) Spinal cord

ANSWERS

No.		**Correct Answer**
1.	e)	CNS abnormalities; a 1-month-old normal child has rooting reflex that facilitates sucking.
2.	b)	8 months of age
3.	a)	Questionable
4.	e)	Questionable. The long-term benefits of early accomplishments are questionable.
5.	d)	Enlargement of breast buds (between 8 to 13 years of age)
6.	d)	9½ years of age
7.	e)	DHEAS may appear as early as 6 years of age.
8.	d)	Rumination disorder is characterized by weight loss or failure to gain weight because of repeated regurgitation of food without nausea or other GI problems.
9.	a)	Tachycardia, hypotension, and decreased oxygen saturation
10.	a)	Drowning most commonly occurs in bathtubs or swimming pools; most children are older than 5 years of age.
11.	e)	Hepatitis (e.g., elevated liver enzymes, jaundice, or acute hepatic necrosis) rarely occurs in children.
12.	e)	The physician should not remove the infant from a mechanical ventilator but arrange for meeting with the parents and social worker. The grade 2 IVH is a benign condition and does not cause brain injury. A 26-week-gestational age preterm infant can survive without a major problem.
13.	d)	Running causes more fire; rolling is indicated if clothes catches fire.
14.	e)	Control of hot water thermostat (maximum water temperature 120°F)
15.	a)	VAS (Visual Analog Scale) is used in patients who are 8 years of age and older. Answers (d) and (e) are used in patients who are 4 years of age and older. Answers (b), (c) and behavioral or combined behavioral-physiologic scales can be used in all age groups. VAS is a vertical 10-cm line, patient marks on line between "no pain" and "worst pain imaginable" (or sad face).
16.	d)	Faces scales (e.g., Wong-Baker, Oucher, Bieri, McGrath scales) are used in patients who are 4 years of age and older. **In faces scale**, patients compare their pain to line drawings of faces or photos of children.
17.	c)	Autonomic measures (e.g., heart rate, BP, heart rate spectral analysis) are used in all ages especially those who are receiving a mechanical ventilation. Behavioral scales are facial expressions and limb movements. Physiological scales are heart rate and BP. Hormonal-metabolic measures are plasma or salivary samples of hormones (e.g., cortisol, epinephrine).
18.	b)	VAS (Visual Analog Scales), color and other analog scales **cannot** be used in children with cognitive limitations.
19.	a)	Glucose-6-phosphatase deficiency or absence is noted in patients with type Ia glycogen storage disease (Von Gierke)
20.	b)	Pompe disease (or type II glycogen storage disease)
21.	a)	Autosomal recessive
22.	e)	Acid maltase (acid alpha-glucosidase)
23.	b)	Muscle biopsy or liver biopsy specimen reveals decreased quantity of acid maltase activity.
24.	a)	Hyperinsulinemia and large for gestational age (LGA) may be present in patients with erythroblastosis fetalis like IDM (infant of diabetic mother).
25.	e)	Microcephaly, macroglossia, and islet cell hyperplasia are also present in Beckwith-Wiedemann syndrome.
26.	a)	Retinoblastoma
27.	b)	Chromosome 11p
28.	e)	Bilirubin encephalopathy (kernicterus) can cause choreoathetotic cerebral palsy and sensorineural hearing loss.
29.	c)	Gentamicin, furosemide, high serum bilirubin, and hypoxia with or without hyperventilation
30.	e)	ROP
31.	d)	Birth trauma (e.g., brachial plexus, phrenic nerve, and spinal cord injuries)
32.	b)	PVL and hypoxic-ischemic encephalopathy
33.	a)	IVH and meningitis
34.	c)	Hypoxia can cause poor speech.
35.	a)	Asymmetric SGA (small for gestational age) patients have normal head circumference unlike symmetric SGA.

36. d) IVH; CMV infections can cause encephalopathy.
37. b) Screening for sickle cell anemia is not indicated in white adolescent females. Sickle cell disease is common in African-American children. HIV-testing is indicated in high-risk adolescents. Spirometry is useful in adolescents who smoke routinely. Sexually active adolescents should be screened for sexually transmitted diseases.
38. d) Antidepressant medications are useful in depressed patients with anorexia nervosa.
39. e) Bradycardia with pulse rate as low as 20 beats per minute can occur in patients with AN. The mortality occurs in 10% of cases and is due to electrolyte imbalance, cardiac arrythmias, or CHF (congestive heart failure) in the recovery phase.
40. a) A daily weight gain should be limited to 200-400 grams.
41. a) A "refeeding syndrome" includes hypophosphatemia that is due to insulin-induced shift of phosphate from extracellular to intracellular space.
42. d) A prolonged use of ipecac can cause myocarditis.
43. d) Chromosome 22q
44. e) Cleft palate is included in CATCH 22 syndrome (not the cleft lip)
45. e) The patients with severe combined immunodeficiency (SCID) have lymphopenia (less than 2,000 per mm^3) at birth.
46. d) Serum immunoglobulin levels are decreased or absent in patients with severe combined immunodeficiency (SCID) disease and no antibodies are formed after immunizations.
47. d) The patients with CHILD syndrome appears with unilateral exfoliative erythematous dermatitis (psoriasis-like) **with facial sparing**, airway cartilage calcification, epiphyseal calcification, and punctuate vertebrae.
48. d) X-linked dominant
49. c) Sphingomyelinase
50. e) Glucocerebrosidase
51. a) Arylsulfatase A
52. b) Beta-galactosidase
53. d) Beta-hexosaminidases A and B
54. b) **Neurologic: Abdominal pain**, vomiting, diarrhea, constipation, ileus, dysuria, muscular hypotonia, sensory neuropathy, seizure, and respiratory failure
55. c) **Pink to brown staining urine**, bullae, crusts, scar formations, hypo-and hyperpigmentation, alopecia, sclerodermoid lesions, hypertrichosis, splenomegaly, hemolytic anemia, and erythrodontia
56. d) **Photosensitivity: Bullae on the sun-exposed areas of the skin** (e.g., dorsa of the hands), skin fragility, crusts, scar formations, sclerodermoid lesions, hyperpigmentation, hypopigmentation, and hypertrichosis
57. e) **Photosensitivity: Pink urine, bullae**, crust, scar formations, sclerodermoid lesions, hypopigmentation, hyperpigmentation, and hypertrichosis
58. a) **Photosensitive: Erythema, edema, itching, burning sensation**, and scarring vesicles
59. b) This boy has erythropoietic protoporphyria (EPP). Some patients do not have cutaneous clinical manifestations (e.g., erythema, edema, petechiae, vesicles, and crusting). Gallstones occur early and fairly common. Hepatic diseases are uncommon. Anemia is absent. The therapy includes beta-carotene, topical sun-screen agents, and avoidance of the Sun.
60. e) Glucose
61. d) Mental retardation and/or seizure activities can occur.
62. c) 1-3 hours after birth
63. e) SGA (small for gestational age)
64. e) A chaperone should be present whenever adolescent female patients are examined by male physicians. However, a chaperone's presence is not yet indicated whenever adolescent male patients are examined by female physicians.
65. b) Morphine
66. c) Benzoyl ecgonine
67. a) Acetaldehyde
68. d) Carboxy-and hydroxymetabolites
69. e) Hydroxylated metabolites
70. d) Opiates
71. b) Amphetamines

72. c) LSD (lysergic acid diethylamide)
73. e) Volatile nitrites (e.g., amyl nitrite, butyl nitrite, and related compounds used in room deodorizers), airplane glue (toluene), and gasoline.
74. c) Methylenedioxymethamphetamine
75. b) Thrombocytopenia, T-cell deficiency, and poor antipolysaccharide antibody production
76. d) Selective IgA deficiency and T-cell deficiency
77. c) Combined B- and T-cell defects of varying severity (e.g., defective antibody-mediated immunity, combined immunodeficiency, and severe combined immunodeficiency)
78. a) Low immunoglobins, CD4 deficiency, and lack of T-cell responses to antigens
79. e) Poor T-cell responses, autoimmunity, impaired apoptosis, and increased bcl-2
80. c) P. carinii, fungi, and viruses attack the children with complete DiGeorge syndrome. Patients with partial DiGeorge syndrome experienced only mild infections.
81. e) Hypokalemia, hyperlipidemia, weight gain, and Cushingoid habitus can occur.
82. d) Allergic conjunctivitis
83. d) Intestinal bleeding does not cause death in patients with SLE. However, pulmonary hemorrhage can cause death in patients with SLE.
84. e) Chronic use of corticosteroids
85. a) Juvenile dermatomyositis
86. a) Coombs-negative anemia is present in juvenile dermatomyositis.
87. d) Retinitis is a less frequent finding. **Gottron papules** (skin over the metacarpal and proximal interphalangeal joints are hypertrophic and reddish pink), **Gowers sign** (a child cannot rise unassisted from the floor without climbing up the body). Mechanic's hands, with thickened skin, skin overgrowth, and range losses are due to refractory, inflammatory myopathy that is more common in adults.
88. a) **ESR is normal**. The laboratory findings in patients with juvenile dermatomyositis are the following: **elevated** serum levels of muscle-derived enzymes (e.g., creatine kinase, aldolase, lactic acid dehydrogenase, serum glutamic-oxaloacetic transaminase); **presence of ANA (nonspecific**, 60% of cases); **negative antibodies** to SSA, SSB, Sm, RNP, and DNA; **negative rheumatoid factor**; elevated percentage of peripheral blood CD19B cells but decreased lymphocyte counts and CD8 cells; presence of Coombs-negative anemia.
89. a) Bed rest should be avoided because weight bearing improves bone density. A sunscreen (free of para-aminobenzoic acid) provides maximum protection against ultraviolet rays A and B throughout the year.
90. b) **Relapsing fever** (febrile periods that are separated by intervals of normal temperature); **remittent fever** (persistent and varies by more than 0.5°C/24 hours); **biphasic fever** (a single illness with two distinct periods of fever over 1 or more weeks); **periodic fever** (fever syndrome with regular periodicity); **intermittent fever** (a period of normal temperature on most days).
91. b) Central fever
92. e) Tertian fever (fever occurs on the 1st and 3rd days)
93. a) Biphasic fever
94. c) Periodic fever (fever syndromes with a regular periodicity)
95. d) Quartan fever (fever occurs on the 1st and 4th days)
96. b) **Relative tachycardia** (i.e., pulse rate is elevated out of proportion to the body temperature) is noted in infectious diseases with release of toxin or noninfectious diseases.
97. a) **Relative bradycardia** (i.e., temperature-pulse dissociation, pulse rate remains low in the presence of fever) is noted in drug fever, typhoid fever, brucellosis, leptospirosis, and conduction defect (e.g., Lyme disease, acute rheumatic fever, infective endocarditis, viral myocarditis).
98. c) **Biphasic fever** is noted dengue fever, leptospirosis, yellow fever, and the African hemorrhagic fevers (e.g., Ebola, Lassa fever, Marburg).
99. e) Periodic fever is noted in apthous stomatitis, pharyngitis, adenopathy, and cyclic neutropenia.
100. d) Relapsing fever is due to Borrelia recurrentis, Q fever, typhoid, TB, Dengue fever, Lyme disease, malaria, Epstein-Barr virus, acute rhematic fever, and many other causes.
101. a) Hypotension, cyanosis, oliguria, anuria, jaundice, direct hyperbilirubinemia, purpura fulminans, and signs of cardiac failure are the **secondary manifestations of sepsis.**

Other primary manifestations include tachycardia, skin lesions (e.g., petechiae, diffuse erythema, ecchymoses, ecthyma gangrenosum), and altered mental status (e.g., agitation, confusion, excitation, anxiety, lethargy, obtundation, coma).

102. e) Positive blood cultures; answers (a), (b), (c), (d), positive Gram stain, positive leukocytes in urine, thrombocytopenia, prolonged PT, low fibrinogen levels, D-dimer, elevated LFTs, metabolic acidosis, and hypocalcemia are noted in septic shock.
103. c) A prophylactic intubation and mechanical ventilation are indicated in infants and children with respiratory distress because they are susceptible to develop respiratory failure.
104. e) Neutropenia; answers (a), (b), (c), (d), and Dohle bodies suggest bacterial infections.
105. a) Metabolic alkalosis does not occur in sepsis. Other laboratory findings of sepsis are prolonged PT, prolonged PTT, reduced serum fibrinogen levels, elevated fibrin split products, leukocytosis, leukopenia, elevated band counts, toxic granulations, and vacuolation of neutrophils.
106. e) Pneumonia occurs rarely. Other complications are sinusitis, mastoiditis, acute rheumatic fever, and acute poststreptococcal glomerulonephritis.
107. c) All of the above; acute glomerulonephritis can occur after group A Streptococcal skin or upper respiratory tract infection.
108. b) Acute rheumatic fever can occur only after group A Streptococcal upper respiration tract infection.
109. a) Poststreptococcal reactive arthritis usually involves the larger joints; a small number of patients develop valvular heart disease.
110. e) Carditis; some physicians give secondary penicillin prophylaxis up to 1 year from the onset of infection.
111. d) Shigella, Ekiri syndrome or "lethal toxic encephalopathy" is a rare syndrome of severe toxicity, severe hyperpyrexia, headaches, brain edema, and convulsion without sepsis or significant dehydration. The mechanism is unclear.
112. d) Dehydration; answers (a), (b), (c), and (e)r are also complications of Shigella infection.
113. d) Blood-tinged vaginal discharge (vaginitis) is rare and usually associated with Shigella flexneri infections.
114. d) Shigella sonnei organism does not cause HUS because it does not produce Shiga toxin. All organisms in answers (a), (b), (c), and (e) produce Shiga toxin. HUS is caused by Shiga toxin-mediated endothelial damage. Shiga toxin is also produced by ShET-1 by Shigella flexneri 2a.
115. d) Shigella dysenteriae type 1
116. c) Vitamin A deficiency (hyporetinemia) can occur in children with measles (up to 72% of cases).
117. d) Cerebral bleeding is absent; **rare** complication of measles are answer (c), hemiplegia, cerebral thrombophlebitis, and retrobulbar neuritis.
118. e) Measles can causes Hecht giant cell pneumonia (interstitial pneumonia).
119. d) Remove the firearm from the environment of children and adolescents
120. c) Acts of rage between relatives or acquaintaces
121. e) Hydroxyapatite
122. c) Slightly less than 50% of the total serum calcium
123. c) In metabolic alkalosis, more calcium is bound to albumin, less free or ionized calcium is present in serum resulting in hypocalcemic symptoms and signs (e.g., sodium bicarbonate infusion in metabolic acidosis).
124. b) Increased intracranial pressure, slow growth, alopecia, drying and cracking of skin, hepatosplenomegaly, painfull swelling of long bones, fragile bone, and carotenemia
125. e) Nyctalopia, photophobia, xeropthalmia, conjunctivitis, retarded growth, faulty epiphyseal bone formation, defective tooth enamel, impaired resistance to infection, and skin keratinization.
126. a) Beriberi, constipation, insomnia, tachycardia, cardiac failure, edema, polyneuritis, aphonia, and elevated pyruvic acid level in blood
127. c) Ariboflavinosis, photophobia, burning and itching eyes, cheilosis, corneal vascularization, and poor growth
128. d) Megaloblastic anemia, glossitis, pharyngeal ulcers, and impaired immunity
129. d) Hypercalcemia; the boy has subcutaneous fat necrosis; there is history of birth asphyxia or trauma; hypercalcemia is probably due to excessive 1, 25-dihydroxyvitamin D that is probably produced by the macrophages infiltrating the areas of fat necrosis.
130. a) Early screening inventory test is performed in children between 3-6 years of age. Answers (b) is performed between 0-6 years of age. Answers (c) and (d) are performed between 0-3 years of age. Answer (e) is performed between 4-48 months of age.

131. a) PEDS (parents evaluation of developmental status) is performed between 0-8 years of age. PEDS is a well-documented psychometric and can be done by parents.
132. c) Ignore the behavior; breath holding behavior can occur during the first years of life. Infants and toddlers do breath holding spell to control the parents and their caregivers.
133. a) 0%
134. b) Initially, parents should confront the child to give clear message that lying is not acceptable. If the situation does not improve, professional help is indicated.
135. a) A boy (3-6 years of age) with high level of aggressive behavior remains aggressive, especially without family-focused intervention.
136. e) Truancy (disorganization within the home or developing personality problem) and run-away behavior are never developmentally normal.
137. a) The mother should turn away briefly from the child during the episode of temper tantrum and oppsitionalism. After the child is calm, the mother should tell the child that the reasons for frustrations are understandable but the opposional behavior is not acceptable.
138. d) Scurvy or vitamin C deficiency; the x-ray findings are characteristic of scurvy.
139. e) Trisomy 21
140. b) Trisomy 13
141. a) Klinefelter syndrome (47, XXY)
142. e) Fragile X syndrome
143. d) Supravalvular aortic stenosis
144. c) Hypotonia, hypogonadism, and obesity
145. a) Bile duct paucity with cholestasis
146. e) Broad thumbs and toes
147. b) Uncontrollable bouts of laughter, seizures, and jerky movements
148. b) Aniridia; WAGR syndrome represents Wilms tumor, aniridia, male genital hypoplasia, and mental retardation.
149. a) Brachycephaly, midfacial hypoplasia, cleft palate, short stature, behavioral problems, and mental retardation
150. d) Pulmonary artery stenosis, butterfly vertebrae
151. e) Microcephaly, hypoplastic male external genitalia, seizures, growth retardation, and severe mental retardation
152. c) Conotruncal cardiac defects, hypoplasia or agenesis of the thymus and parathyroid glands, hypoplasia of auricle and external auditory canal
153. b) Glycine cleavage enzyme deficiency produces hyperglycinemia.
154. a) **Minimal amount of protein** is used as a mixture of essential amino acids (0.25 g/kg/day) during the first 24 hours of therapy. Infants usually become symptomatic within a few days of protein feeding. Answers (b), (c), (d), and (e) are therapies in patients with hyperammonemia. A peritoneal dialysis or hemodialysis is indicated if above therapies fail.
155. a) The cause is unknown. Urea cycle enzyme activities are normal. The newborn girl has severe transient hyperammonemia.
156. a) Phytanic acid-restricted diet; early institution of phytanic acid-restricted diet can reverse peripheral neuropathy and prevent the progression of the eye and CNS clinical manifestations. In classic Refsum disease, phytanic acid is increased and pristanic acid is decreased.
157. c) Phytanoyl-CoA deficiency is localized to the peroxisome.
158. e) 1% silver nitrate; answers (a), (d), or 1% tetracycline opthalmic ointment is an alternative therapy.
159. c) Muscle tone (**not** muscle strength)
160. b) Apgar score is 2 (heart rate is over 100/minute). Infants with Apgar scores 0-3 need immediate resuscitation.
161. c) False-negative (presence of acidosis but normal Apgar) results are due to maternal acidosis, some full-term infants, and high fetal catecholamine levels. False-positive (absence of fetal acidosis or hypoxia but low Apgar) results are due to spinal cord trauma, CNS anomalies, airway obstructions, pneumonia, sepsis, myopathy, neuropathy, lung anomalies (hypoplasia, diaphragmatic hernia), and hemorrhage resulting in hypovolemia.
162. b) Drugs that are weak acids have less effects on neonates.
163. e) CMV (term infant) is allowed to take breast feeding. Breast feeding is allowed in hepatitis B virus infections **after the infant receives hepatitis B immunoglobulin and hepatitis B vaccine** (**not** before the immunizations). HTLV is human T-cell leukemia virus types 1 and 3. Breast feedings are also contraindicated in active TB, breast abscesses, herpetic vesicles on the breasts only, and herpes zoster vesicles on the breasts only. Herpes simplex and herpes zoster lesions in areas other

than breasts can be covered and breast feeding is allowed. Rubella (wild type) present in breast milk, rarely causes newborn infections, but the breast feeding is not contraindicated.

164. e) Mycobacterium tuberculosis is present in breast milk. However, breast feeding is contraindicated due to aerosol spreads during maternal active TB (**not** due to transmission through the breast milk). Hepatitis (A, B, and C) viruses are present in breast milk but do not cause infections through the breast milk. However, hepatitis C virus transmission is theoretically possible.
165. a) Aldomet is probably safe but used with caution.
166. e) Category X drugs have more risks than benefits.
167. c) Neuroblastoma, bleeding (vitamin K deficiency), IUGR, and other anomalies occur in phenytoin therapy.
168. b) Hypoplastic nasal structures, fetal bleeding, and death occur in warfarin therapy.
169. a) Hypothyroidism also occurs due to maternal iodides intake and propylthiouracil.
170. d) Facial-ear defects and heart disease occur in accutane therapy.
171. e) Ebstein's anomaly and macrosomia occur in lithium therapy.
172. a) Warfarin prevents the carboxylation of gamma-carboxyglutamic acid that is a component of osteocalcin and other vitamin K-dependent bone proteins. Warfarin is vitamin K antagonist.
173. b) Phenytoin causes deficiency of epoxide hydroxylase resulting in accumulation of a metabolite that causes teratogenic effects.
174. e) Dexamethasone; it is also used in congenital heart block.
175. a) Indomethacin may cause nephrocalcinosis and postnatal sodium losses
176. b) Maternal intravenous immunoglobulin and umbilical vein platelet transfusions
177. c) Flecainide is the drug of choice. If the fetus has hydrops, administration of Flecainide through umbilical vein is indicated in utero. Other useful drugs are digoxin, procainamide, amiodarone, and quinidine.
178. d) Vitamin B_{12} may be effective in methylmalonic acidemia.
179. b) Kernicterus
180. a) Nephrocalcinosis, biliary stones, deafness, low (sodium, chloride, and K)
181. e) Pyloric stenosis is caused by erythromycin and prostaglandins.
182. c) Intestinal perforation, oliguria, and hyponatremia
183. d) Seizures, withdrawal, and chest wall rigidity
184. e) Baby is blue when stops crying and becomes pink when starts crying.
185. d) Anuria, oligohydramnios, pulmonary hypoplasia, and pneumothorax
186. a) Aspiration pneumonia is due to esophageal atresia and TE fistula.
187. b) Regurgitation of milk is due to cleft palate.
188. c) Scaphoid abdomen and respiratory distress
189. d) C4 (4th cervical vertebra level) is injured in NSVD.
190. d) C7-T1 (lower cervical and upper thoracic levels) are injured in breech delivery.
191. e) Right peripheral facial nerve palsy due to the pressure effect from forcep delivery. The forehead wrinkles on the affected side with central paralysis that involves only lower two thirds of the ipsilateral side of the face.
192. a) Ganciclovir therapy is being evaluated.
193. e) Transmitted through cat feces or undercooked meat
194. c) Life-threatening illness
195. a) Cataracts
196. d) Penicillin is the drug of choice.
197. e) Liver laceration during breech delivery causes subcapsular hematoma of the liver. The diagnosis is made by ultrasonographic findings.
198. e) Antireflux therapy is not indicated in preterm infants with apnea because the role of GE reflux in apnea of prematurity is controversial. Mechanical ventilation is indicated when nasal CPAP (continuous positive airway pressure, 3-5 cm H_2O) failed to improve the clinical conditions (i.e., apnea).
199. a) Hypotonia is noted in **phase 1** (1st 1-2 days) (e.g., poor sucking, stupor, hypotonia, and seizures). **Phase 2** (middle of 1st week) patients appear with hypertonia of extensor muscles, opisthotonos, retrocollis, and fever. **Phase 3** (after 1st week) patients appear with hypertonia.

200. e) Hemolytic jaundice due to 'ABO' incompatibility manifests during the first 24-36 hours of life.

201. a) Lack of **physicians** concern regarding the presence of jaundice, delay in measuring serum bilirubin levels despite marked jaundice, delay in starting phototherapy, and failure to respond to parenteral concern regarding jaundice, lethargy, and poor feeding are also the preventable causes of jaundice.

202. b) 20 mg/dL or higher; patients with hemolysis, double volume exchange transfusions should be initiated when the serum indirect bilirubin level is 20 mg/dL or higher.

203. c) Whole blood (33% packed-RBCs and 67% plasma) is used for double volume exchange transfusions; answers (e) is used for simple transfusions in newborns with anemia.

204. b) From 4 Z, 15 Z-bilirubin to 4 Z, 15 E-bilirubin

205. d) Eye injury and nasal occlusion from the bandages are not common complications. Other common complications are chilling from exposure of the infant, purpuric rash associated with transient porphyrinemia, and bronze baby syndrome. Phototherapy should not be used in patients with porphyria.

206. e) Anti-Lewis antibodies do **not** cause hemolytic disease of the newborn. Other rare causes are RBC antigens (e.g., Kell, Duffy, Lutheran, Diego, Kidd, M, S, P, MNS, Xg, C^W, C^X, and D^U).

207. b) Recurrent sinopulmonary infections (80% of cases)

208. e) Lymphoreticular type of malignancy

209. c) X-linked recessive

210. a) Low serum level of IgM

211. e) Gastroesophageal reflux (over 50% of cases)

212. a) Cystic fibrosis

213. e) Thin-section high-resolution CT scanning has replaced bronchography as the gold standard. CT scan typically reveals **cylindrical** ("tram lines," "signet ring appearance"), **varicose** (bronchi with "beaded contour"), **cystic** (cysts appear in "strings and clusters"), or mixed types.

214. e) Streptococcus pneumoniae with either RSV or Mycoplasma pneumoniae

215. a) Staphylococcus aureus and Streptococcus pneumoniae

216. e) Nasal polyps or sinus diseases occur less frequently than rectal prolapse, electrolytes or acid-base abnormalities. Answers (a), (b), (c), and (d) are in the order of decreasing frequencies (i.e., answer (a) is the most common presentation). Hepatobiliary diseases occur **less** frequently than nasal polyps.

217. d) Cystic fibrosis

218. e) Diagnostic criteria for cystic fibrosis are the following:

Presence of typical clinical manifestations (respiratory, GI, or GU) **or** history of CF in a sibling or positive newborn screening test

PLUS

Laboratory evidence for CFTR (cystic fibrosis transmembrane regulator) dysfunction: Two elevated sweat chloride concentrations performed on separate days **or** identification of two CF mutations **or** an abnormal nasal potential difference measurement

219. b) Sexual function is usually unimpaired. Sexual development is often delayed by an average of 2 years. Females may experience secondary amenorrhea. Pregnancy is well tolerated in CF women with good pulmonary functions.

220. b) Hyperuricemia and colonic stricture can occur due to the use of very large doses of pancreatic extracts.

221. c) 5-6 years of age; by this time most patients have obstructive pulmonary disease. A decrease in **mid-maximal flow rate** is an early finding that suggests small airway obstructions.

222. a) Restrictive lung changes occur **late** in the disease process (i.e., decreased total lung capacity and vital capacity) and are due to extensive lung injuries and fibrosis.

223. b) Mucoid forms of pseudomonas; the presence of Staphylococcus aureus, Pseudomonas aeruginosa, or Burkholderia cepacia organism suggests CF.

224. b) Gynecomastia

225. e) Hyperglycemia (systemic); growth retardation (systemic, inhaled)

226. d) GI bleeding

227. a) Interstitial nephritis

228. c) Goiter

229. e) Complex cyanotic heart disease with decreased pulmonary blood flow
230. d) Acyanotic lesions with increased pulmonary blood flow
231. a) Pericardial thickening and constrictive pericarditis
232. b) Mitral valve prolapse and aortic root dilatation
233. c) VSD
234. b) Chromosome 7; supravalvular aortic stenosis and hypercalcemia
235. e) Chromosome 20; peripheral pulmonic stenosis and bile duct hypoplasia
236. c) Chromosome 12; pulmonic stenosis
237. d) Chromosome 15; aortic root dilatation
238. a) Chromosome 6; craniofacial, limb defects, and PDA.
239. b) VSD
240. c) Pulmonary valvular insufficiency also occurs due to severe pulmonary hypertension.
241. b) VSD
242. b) A short systolic murmur is noted along the left sternal border at the 3rd and 4th intercostal spaces; murmur is transmitted to the left infrascapular area.
243. c) Apical murmur is late systolic and may be associated with a click.
244. a) A low-pitched decrescendo diastolic murmur is noted at the upper and midleft sternal border
245. d) A high-pitched ("cooing dove"), apical holosystolic murmur; moderate to severe insufficiency causes low-pitched, mid-diastolic rumbling murmur.
246. e) A rumbling apical diastolic murmur is followed by loud 1st sound; 2nd sound is loud and split.
247. c) Avascular necrosis of the hips has been reported.
248. e) Streptococcus pneumoniae (over 60% of cases); answers (a), (b), (c), and (d) also cause infections in post-splenectomy patients.
249. a) The children with congenital polysplenia have normal functions of spleen.
250. e) RBC inclusions in peripheral blood smears, 'pits' on interference microscopy, and decreased uptake of technetium on spleen scan indicate splenic hypofunctions.
251. a) It is better to postpone the operation **until 5 years of age**. Vaccine is less effective in children less than 2 years of age and in immune-suppressed patients. Oral penicillin V is given (125 mg twice daily in less than 5 years of age and 250 mg twice daily for children 5 years or older).
252. b) Gastrointestinal cancer and ovarian tumors
253. e) Bilateral acoustic neuromas and meningiomas
254. c) Myocardial rhabdomyoma and fibroangiomatous nevi
255. a) Angioma
256. d) Hemangioblastoma of the cerebellum and retina, renal cancer, and pheochromocytoma
257. a) Parathyroid adenoma, pancreatic islet cell tumor, and pituitary adenoma carcinoid are noted in Wermer syndrome **(multiple endocrine neoplasia, type I)**.
258. b) Medullary carcinoma of the thyroid, hyperparathyroidism, and pheochromocytoma are noted in Sipple syndrome **(multiple endocrine neoplasia, type II). Multiple endocrine neoplasia type III** has Marfan habitus, mucosal neuromas, pheochromocytomas, medullary carcinoma of thyroid, and neuropathy.
259. e) Leukemia and lymphoma
260. c) Breast cancer and extragonadal germ cell tumors
261. d) Schwannoma and myelodysplasia
262. d) 35%; the incidence of reflux does **not** depend on the grade of reflux or sex of the index child.
263. c) 12%
264. e) 50%
265. e) UPJ obstruction
266. a) Intrinsic
267. a) Hypertension is **not** an usual presentation in patients with UPJ obstruction. Some children appears with hematuria after minimal trauma in UPJ obstruction.

268. b) In girls, ureteral duplication is almost always associated with ureterocele.
269. b) In boys, single ureter is oftenly present with ureterocele. Ureterocele is a cystic dilatation of the terminal ureter and is obstructive lesion because of pinpoint ureteral orifice.
270. b) 6-12 months of age
271. e) Seminoma (65% of cases)
272. c) Hepatitis A infection does **not** cause hepatocellular carcinoma. Hepatocellular carcinoma can occur with hepatitis C infection and chronic form of hereditary tyrosinemia.
273. b) Lungs and regional lymph nodes
274. e) If the testis does **not** descend by 6 months of age, it will remain undescended. The majority of the undescended testes descend spontaneously during the first 3 months of life.
275. e) Virilized female with congenital adrenal hyperplasia
276. d) 9-15 months of age
277. e) Indirect inguinal hernia
278. e) 'Acquired' or ascending undescended testis on the right side. It occurs between 4-10 years of age. He has descended testes at birth. The testis does not remain in the scrotum.
279. c) Failure of hormonal and/or mechanical factors cause an undescended testis.
280. b) A brisk cremasteric reflex causes retractile testis.
281. a) Incomplete disappearance of processus vaginalis causes 'acquired' or ascending undescended testis because spermatic cord does not grow as rapidly as the child.
282. d) Vanishing testis (or atrophy) is due to the testicular torsion in utero.
283. a) The boy has right **testicular torsion**. This is the most common cause of testicular pain in boys **12 years of age and older**. However, torsion of the appendix testis is the most common between **2 and 11 years of age**. Answer (e) is the therapy for the **torsion of the appendix testis**. Testicular torsion is surgical emergency. Therefore, prompt surgical exploration, detorsion of the testis, and scrotal orchiopexy are the treatment of choice.
284. e) Ultrasonography of the abdomen should be performed to rule out abdominal or retroperitoneal mass in **boys younger than 10 years of age with varicocele or presence of varicocele on the right side**.
285. e) Laparascopic detorsion of the right ovary is performed after the diagnosis of ovarian torsion is made by color Doppler flow studies. The oophoropexy (placation) of the contralateral adenexa may be indicated. Ovarian torsion is associated with ovary cyst or tumor.
286. e) Amenorrhea, disordered eating, and osteoporosis
287. a) Renal (i.e., unilateral absence of kidney or ureter, 27% of cases); next is skeletal, especially vertebral.
288. b) This syndrome represents the congenital absence of vagina; the patients have normal vulva, phenotype, and karyotype (46, XX).
289. e) Multifactorial/polygenic
290. b) Nephrogenic diabetes insipidus is not part of Wolfram syndrome.
291. b) Low (serum sodium, serum uric acid); high (urine sodium, vasopressin level); normal or high (intravascular volume); normal or low (urine output).
292. c) Low (urine sodium, intravascular volume, vasopressin level); high (serum sodium, urine output, serum uric acid level).
293. a) Low (serum sodium, intravascular volume, vasopressin levels); high (urine output); very high (urine sodium); normal or high (serum uric acid levels).
294. d) SHOX excess syndrome causes childhood tall stature. Answers (a), (b), and (e) also cause childhood tall stature.
295. d) Testicular feminization syndrome causes tall stature in adulthood.
296. e) Reassurance; answer (a) is used for boys with tall stature (i.e., predicted adult height is more than 3 SD above the mean that is **78 inches or 198 cm**); answer (b) is used in girls with tall stature.
297. e) Reassurance; answer (a) is used for girls with tall stature (i.e., predicted adult height is more than 3 SD above the mean that is **71 inches or 180 cm**). Estrogen mediates epiphyseal fusion in both girls and boys. The medication therapy is also indicated if both boys and girls with tall stature have **significant psychosocial problems**.
298. d) GH, IGF-I levels, and results of other endocrine studies are normal. Sotos syndrome is not endocrine disorder. The children have accelerated growth rate for the first 4-5 years of life and then return to normal rate. Most patients have some degree of mental retardation. Osseous maturation is compatible with the patient's height. They have difficulties in sports activities due to clumsiness and awkward gait. Puberty usually occurs in normal time but may occur slightly early.

299. c) Papillary carcinoma (80% of cases)
300. a) Neuromas and endocrine symptoms appear later in childhood. MEN, type II B includes mucosa neuroma syndrome, medullary carcinoma, and pheochromocytoma.
301. e) A positive Chvostek or Trousseau sign is present in patients with hypocalcemia secondary to hypoparathyroidism. They usually have low serum calcium (total and ionized), elevated serum phosphorus level, normal or low serum alkaline phosphatase, usually low 1, 25 $[OH]_2 D_3$, normal serum magnesium level, low serum PTH level, prolonged QT interval on the ECG, and calcification in basal ganglia (x-ray or CT scan).
302. e) Delayed and irregular teeth eruption, irregular enamel formations, and may be unusually soft teeth.
303. e) When hypoparathyroidism occurs with Addison disease, the serum calcium level may be normal. Hypocalcemia reveals after effective therapy of the adrenal insufficiency.
304. e) Detection of serum calcium levels in siblings is important to avoid unnecessary subtotal parathyroidectomy in patients with familial hypocalciuric hypercalcemia (familial benign hypercalcemia).
305. e) Oral prednisone (2 mg/kg/day) therapy corrects hypercalcemia and reduces serum level of 1, 25 $[OH]_2 D_3$ to normal.
306. d) Albright hereditary osteodystrophy (or pseudohypoparathyroidism)
307. e) A major surgery under general anesthesia in patients with adrenal insufficiency needs high intravenous dose of hydrocortisone like acute adrenal crisis. The dose of hydrocortisone should be increased 2-3 times during minor surgery or infection.
308. e) INH does **not** cause adrenal insufficiency. Mitotane can cause adrenal insufficiency. Mitotane is used in the treatment of adrenal carcinoma and refractory Cushing syndrome.
309. c) Serotype 12
310. b) Serotype 49
311. a) Hypertension (60% of cases); answers (b), (c), (d), (e), hypocalcemia, acidosis, and uremia are also potential complications of acute poststreptococcal glomerulonephritis.
312. a) Oral doxycycline is not used in mild papulopustular acne. Topical tazarotene, or nodular acne. Topical tazarotene can also used in comedonal acne. All topical applications are used.
313. a) Oral isotretinoin (1 mg/kg/day) is **not** used in mild papulopustular acne but is used in **severe** papulopustular acne.
314. a) Oral isotretinoin (1 mg/kg/day)
315. b) Renal ultrasonography should be performed to rule out genitourinary anomalies (20% of cases).
316. a) Unilateral renal agenesis
317. a) Spinal dysraphism (40% of cases)
318. b) Spinal bifida occulta; answer (a) is the most severe form of spinal dysraphism.
319. a) Organic (e.g., occult fracture, spondylolysis, scoliosis, kyphosis, tumor, and infection)
320. e) Plain x-ray of the spine (posteroanterior and lateral standing x-ray of the spine with right and left oblique views of the affected area)
321. e) Tall stature with diminished upper segment/lower segment ratio or an increased span to height ratio (i.e., more than 1.05) is the most consistent presenting feature in patients with Marfan syndrome. The **Steinberg sign** is that the thumb can be adducted across the palm. The wrist sign is that the thumb can overlap the fifth (or little) finger when encircling the wrist.
322. d) Nephrologist. However, neurologist can follow up the patient for neuromuscular hypotonia. Yearly evaluations are indicated for cardiac valvular disease, scoliosis, joint dislocation, and opthalmologic problems. Physical therapy may improve strength and hypotonia.
323. e) Hypogonadism
324. c) Fathers of sporadic Marfan syndrome cases are **usually 7 to 10 years older** than fathers in the general population. FBN1 is the fibrillin-1 locus resides within the long arm of chromosome 15.
325. c) 200 rads (2.0 Gy); the most obvious finding is lymphocytic depression due to bone marrow impairments.
326. d) 800 rads (8.0 Gy); the GI syndrome manifests with nausea, vomiting, diarrhea, then latent 1 week asymptomatic period followed by recurrence of GI symptoms, electrolyte imbalance, sepsis, and death.
327. e) 100 rads (1.0 Gy); **most patients** have symptoms of nausea and vomiting after 200 rads (2.0 Gy).
328. e) Malrotation with intermittent volvulus
329. e) A superior mesenteric vein located to the right of the superior mesenteric artery is a normal finding.
330. c) Intrauterine vascular accidents resulting in ischemic necrosis of the sterile bowel and resorption of the affected segment.

331. a) Failure to recanalize the lumen after the solid phase of intestinal development during the 4th and 5th weeks of gestation resulting in duodenal atresia.

332. e) Jejunal atresia; other associated congenital anomalies are renal anorectal anomalies.

333. b) Ladd band (or abnormal peritoneal folds) is connecting the cecum with the posterior abdominal wall and causing pressure on the 2nd part of duodenum from outside.

334. c) Hypochloremic metabolic alkalosis

335. a) A "double-bubble sign" (i.e., one bubble for the stomach and one bubble for the duodenum) is noted on plain abdominal x-ray.

336. e) A "triple-bubble sign" (i.e., one bubble for the stomach, one bubble for the duodenum, and one bubble for the jejunum) is noted on plain abdominal x-ray.

337. d) Absence of ganglion cells and large number of hypertrophied nerve bundles that stain positively with acetylcholinesterase are noted in submucosa that are obtained by rectal suction biopsy.

338. d) Hirschsprung disease (or congenital aganglionic megalcolon)

339. d) Turner syndrome. Hirschsprung disease is also associated with congenital hypoventilation ("Ondine curse") syndromes, urogenital anomalies, and cardiovascular anomalies.

340. e) The **Currarino triad** includes anorectal anomalies (e.g., ectopic anus, rectal stenosis), sacral bone anomalies (e.g., hypoplasia, poor segmentation), and presacral masses (e.g., anterior meningocele, teratoma, cyst).

341. a) Anal tone is normal in both conditions. Hirschsprung disease usually appears at birth but the functional constipation appears after 2 years of age. Answers (c) and (d) are possible in Hirschsprung disease but are absent in functional constipation. Answer (e) is normal in functional constipation and is abnormal in Hirschsprung disease.

342. b) In Hirschsprung disease, rectal biopsy reveals **increased** acetylcholinesterase staining. In normal children, rectal distension results in reflex decline in internal sphincter pressure.

343. e) Acute appendicitis. **The onset of an abdominal pain before the vomiting and fever suggests acute appendicitis**. Infectious enteritis **begins with vomiting** followed by the cramping abdominal pain due to hyperperistolsis.

344. a) Presence of more than 30 WBCs per high-power field in urine suggests urinary tract infections. Some children with acute appendicitis may have normal CBC counts and differential counts. Up to 30 WBCs and 20 RBCs per high-power field in urine have been noted in suppurative appendicitis. Presence of bacteria in the urine suggests UTI. Presence of **significant** proteinuria or cast indicates renal problems.

345. e) Urologic anomaly is also common in newborns with persistent cloaca.

346. d) Anal dilatations are started 2 weeks after the surgery and gradually increased to achieve the size of normal anus.

347. d) A stool softener is indicated to make stools soft, but to avoid both hard stools and diarrhea.

348. e) Idiopathic; answers (a), (b), (c), (d), intestinal parasites, malnutrition, meningocele, ulcerative colitis, and Ehlers-Danlos syndrome can cause rectal prolapse.

349. e) **No therapy is indicated in infants less than 2 years of age** without any predisposing disease because the condition is self-limited. Antibiotics are not useful. Occasionally, abscess is drained under local anesthesia. However, older children (more than 2 years of age) with a predisposing disease require combination of antibiotics that covers gram-negative organisms, anerobes and S. aureus. In addition to that, incision/excision and drainage in cases of sepsis and spreading cellulitis.

350. e) DNA analysis of specimens obtained by chorionic villus sampling is performed in the first trimester between **9-12 weeks of gestation**. Genetic amniocentesis is usually performed at approximately **15 weeks of gestation**.

351. e) To facilitate appropriate prenatal therapy of the female fetus.

352. d) Administration of dexamethasone to the mother (20 micrograms/kg prepregnancy maternal weight daily in two or three divided doses) of affected fetus is indicated. Dexamethasone readily crosses the placenta and suppresses secretion of steroids by the fetal adrenals, including secretion of adrenal androgens.

353. b) To prevent the virilization of external genitalia of the female fetus.

354. a) 6 weeks of age. Then, choronic villus biopsy is performed to determine the sex and genotype of the fetus. **Please remember, therapy is continued only if the fetus is affected female with CAH**. Some centers can make early diagnosis by DNA analysis of fetal cells from maternal plasma for sex determination and CYP 21 gene analysis (for frequently occurring mutations).

355. e) Unknown

356. a) The nonclassic form of CAH is **not** detected reliably. The adrenal insufficiency does **not** occur in nonclassic form of 21-hydroxylase deficiency. Genotying for CYP21 can improve specificity.

357. a) Transphenoidal pituitary microsurgery is the treatment of choice. Answer (b) or reoperation is performed in patients with relapse. Answers (c) is centrally acting serotonin antagonist that blocks ACTH release and is rarely used in children. Answer (d) is needed in patients with pituitary adenoma that does not respond to therapy or ACTH is secreted by ectopic metastatic tumor. Answer (e) is needed in patients with benign cortical adenoma.

358. d) Cushing syndrome is characterized by moon facies (i.e., rounded face, prominent cheeks, and flushed appearance).

359. e) Pubertal development is delayed; other clinical manifestations may include secondary amenorrhea after menarche, purplish striae on the abdomen, hips, and thighs, pathologic fracture, emotional disturbances, weakness, and headaches.

360. b) Nighttime salivary cortisol levels are **elevated** and may be used as screening test in obese children. **Please remember**, cortisol levels in blood are normally elevated at 8 AM and reduces to less than 50% by midnight **except in infants and young children in whom diurnal variations may be absent**.

361. b) Hypokalemia can occur; the levels of serum electrolytes are normal except hypokalemia.

362. a) Tall stature is noted in simple obesity.

363. b) Short stature is present in Cushing syndrome.

364. c) An elevated urinary excretion of cortisol level is always present in Cushing syndrome and is often present in simple obesity.

365. b) An elevated nighttime salivary level of cortisol is noted in Cushing syndrome.

366. a) Cortisol secretion is suppressed by oral administration of low doses of dexamethasone in simple obesity.

367. b) ACTH-independent Cushing syndrome

368. a) Pituitary Cushing syndrome (e.g., pituitary adenoma)

369. b) ACTH concentrations are suppressed in patient with ACTH-independent Cushing syndrome or a cortisol-secreting tumor (e.g., adrenal tumors).

ACTH concentrations are **very high** in patients with **ectopic ACTH-secreting tumors** and may be **normal** in patients with **ACTH-secreting pituitary adenomas**.

370. a) ACTH-dependent Cushing syndrome (e.g., pituitary tumors)

371. b) ACTH-independent Cushing syndrome (e.g., adrenal tumors)

372. d) A single, 8 AM testosterone level more than 0.7 mmol/L (20 mg/dL) in boys indicates that all of them will achieve testicular volume greater than 4 mL within the next 15 months. Answer (c) may distinguish these two conditions. Boys with answer (e), only 25% developed puberty by 15 years of age.

373. e) Human menopausal gonadotropin may be added; recombinantly produced gonadotropins stimulates growth of the testes and spermatogenesis.

374. e) Overhydration does not occur. Children younger than 3 years of age usually develop hypoglycemia. Children older than 3 years of age usually develop hyperglycemia but overall outcome is **unpredictable**. Children often show signs of dehydration due to vomiting secondary to ketosis, poor oral intake, and osmotic diuresis secondary to hyperglycemia. **Please remember**, infections are not more common in diabetic children than in nondiabetic children.

375. a) She should call her physician or nurse if the blood glucose remains elevated after **three** extra doses of insulin. A child with diabetes should go to the ER when there is persistent vomiting, moderate to large ketonuria, and blood glucose levels are more than 250/dL despite extra insulin doses.

376. e) Serum insulin levels should **not** be performed. The physician should perform general examination to evaluate hydration and to determine the presence of ketoacidosis. A child with diabetes mellitus should go to the ER with persistent vomiting and blood glucose level is less than 50-60 mg/dL, especially with ketonemia.

377. e) 5% glucose, 0.45% saline solution with 20 mEq/L of potassium acetate is given at 1.5 times maintenance rate.

378. b) 0.03 units/kg/hour insulin infusion is indicated when the blood glucose levels are 121-200 mg/dL. Answer (a) is correct when the blood glucose levels are less than 120 mg/dL. Answer (c) is correct when the blood glucose levels are 200-300 mg/dL. Answer (d) is correct when the blood glucose levels are 300-400 mg/dL. Answer (e) is correct when the blood glucose levels are 400 and above.

379. a) If glargine is used as the basal insulin, **full dose** is given in the evening before surgery. The intravenous insulin therapy is continued after surgery until the child takes the full oral feedings. Then, IV insulin is switched to subcutaneous insulin.

380. e) The Mauriac syndrome is due to **under-insulinization**. This syndrome is rare in patients with diabetes because of the availability of longer-acting insulins.

381. d) Malfunction of the hypoglossal nerve produces wasting, weakness, and fasciculations of the tongue.

382. e) Unilateral injury of the vagus nerve produces hoarse voice and weakness of the ipsilateral soft palate.

383. c) A glossopharyngeal nerve injury results in absence of gag reflex of the posterior pharyngeal wall.

384. b) Malfunction of the sensory fibres of trigeminal nerve results in absence of corneal reflex. In corneal reflex, afferent fibres are the branches of 5th cranial nerve and efferent fibres are the branches of 7th cranial nerve (facial nerve).
385. a) An injury to the accessory nerve causes paralysis of the sternomastoid and trapezius muscles.
386. c) A complete paralysis of the 3rd cranial nerve causes ptosis, dilatation of the pupil, displacement of the eye outward and downward, and impairment of adduction and elevation.
387. e) A paralysis of the 4th cranial nerve produces upward and outward deviation of the eye.
388. d) A paralysis of the 6th cranial nerve produces medial deviation of the eye and inability to abduct beyond the midline.
389. b) A paralysis of the motor function of the 5th cranial nerve causes absence of jaw jerk.
390. a) A bilateral paralysis of the 10th cranial nerve produces nasal regurgitation of fluids, pooling of secretions, and an immobile low-lying soft palate.
391. a) Average head growth is 0.50 cm during the 1st week of life.
392. c) 34-35 cm
393. b) 44 cm
394. c) 47 cm
395. e) URI
396. e) A normal 2-year-old child can produce **soft symmetric bruits**. Answers (a), (b), (c), and (d) produce increased intracranial pressure.
397. e) Arteriovenous malformations of the vein of Galen or middle cerebral artery produce **loud cranial bruits**. A loud or localized bruit needs further investigation.
398. e) Reassurance; retinal hemorrhages disappear spontaneously by 1-2 weeks of age, occur in 30-40% of all full-term newborn infants, more common in NSVD than in cesarean section, and are not associated with birth injuries or neurologic complications.
399. e) Reassurance; the size of anterior fontanel is normal.
400. d) 18 months of age
401. b) After the first 6-8 weeks of life
402. e) Reassurance; anterior fontanel may close normally as early as 9 months of age.
403. c) A broad forehead indicated hydrocephalus
404. a) The normal head circumference of a full-term newborn is about 34-35 cm.
405. b) A box-shaped skull or square shaped skull indicates chronic subdural hematomas.
406. b) Dilated pupil is **not** part of Horner syndrome.
407. a) Lumber tap is contraindicated when the platelet count is less than 20,000 because it can cause spontaneous bleeding in the subarachnoid or subdural space. Rarely, critical illness is also contraindication of LP.
408. e) A crying and struggling patient has increased opening pressure in CSF. The normal opening pressure of CSF in recumbent and relaxed position is about **100 mm of fluid** and in flexed lateral decubitus position is **60-180 mm of fluid**.
409. b) Type II diabetes
410. a) Rhabdomyolysis; answers (b) and (c) are noted in ICU patients with hypophosphatemia; answers (d) and (e) are noted in patients with chronic hypophosphatemia.
411. e) Severe hypophosphatemia can cause dysfunction of WBCs and platelets, ataxia, paresthesias, and coma. Long-term phosphorus deficiency causes rickets in younger children and osteomalacia in older children.
412. c) Decreased renal acid excretion results in decreased serum bicarbonate concentration. If the respiratory alkalosis resolved quickly, patients develop metabolic acidosis that resolves over 1-2 days by increased acid excretion by the kidneys.
413. c) Acidemia causes potassium to move from the intracellular space to the extracellular space.
414. e) Children with Gitelman syndrome do not need calcium supplementation. Bartter syndrome is usually associated with hypercalciuria and often with nephrocalcinosis. Gitelman syndrome has high urinary magnesium losses and low urinary calcium losses.
415. d) Both the syndromes have normal blood pressure.
416. a) Autosomal recessive
417. e) Moderate mental retardation is noted in about 50% of cases. Children with pseudohypoparathyroidism type 1A have the phenotype features of Albright hereditary osteodystrophy. The presence of brachydactyly is due to shortened metacarpals and metatarsals.

418. a) Measurement of urinary cAMP after infusion of synthetic PTH is the diagnostic test for **pseudohypoparathyroidism in which there is no increase in urinary cAMP**, as normally occurs due to G protein-mediated activation of adenyl cyclase after PTH binds to its receptors. In pseudohypoparathyroidism there is normal production of PTH but lack of response at the target organs. Answers (c), (d), and (e) are the findings in patients with pseudohypoparathyroidism.

419. b) **High serum calcium levels** produce psychiatric symptoms, ventricular arrythmias, hypertension, decreased QT intervals, polyuria, nephrocalcinosis, renal failure, hypernatremia, and pancreatitis. **However, pancreatitis causes hypocalcemia**. Infants with hypercalcemia appear with poor feeding, vomiting, and failure to thrive.

420. e) **Hydration with normal saline** is followed by the use of **loop diuretic** that enhances calcium excretion. **Glucocorticoids** (e.g., prednisone) decreases intestinal absorption of calcium by blocking the action of 1, 25-dihydroxyvitamin D and especially effective in hypercalcemia secondary to vitamin D intoxication and subcutaneous fat necrosis. **Calcitonin** reduces calcium by inhibiting bone resorption. **Biphosphonates** inhibits bone resorption through their effects on osteoclasts. **Hemodialysis** is useful in patients with hypercalcemia and renal failure. Chronic hypercalcemia is treated with **diet with low calcium and vitamin D**.

421. a) Asymptomatic

422. b) Hypomagnesemia produces flattened T waves and prolonged ST segments. Answers (a), (c), (d), and (e) are the dominant clinical manifestations of **hypomagnesemia secondary to hypocalcemia**. Arrythmias usually occur in patients with an underlying heart disease.

423. a) Gitelman syndrome produces hypocalciuria.

424. b) Bartter syndrome produces hypercalciuria and secondary nephrocalcinosis. Both Gitelman and Bartter syndromes produce hypokalemic metabolic alkalosis and hypomagnesemia secondary to renal magnesium losses.

425. e) Infant of diabetic mother; answers (a), (b), and (d) can cause transient hypomagnesemia in newborns; answer (c) causes hypermagnesemia due to maternal magnesium sulfate therapy.

426. c) Estrogen does not cause respiratory alkalosis. However, caffeine causes respiratory alkalosis.

427. c) Salicylates directly stimulate the respiratory center and often concurrently produce metabolic acidosis.

428. a) Sepsis causes respiratory alkalosis probably due to cytokine release.

429. b) Pain, stress, panic, or anxiety causes psychogenic hyperventilation.

430. b) Suffocation (little over 50% of cases); 2nd most common is MVA (about 22%)

431. d) MVA is the most common cause of injury death in children and adolescents.

432. e) Burns grater than 10-15% of the body surface area and inadequate home situation are also indications of hospitalization.

433. a) Children with burns greater than 15% body surface area should **not** receive oral fluid therapy due to ileus. **A nasogastric tube** should be placed to avoid aspiration. **A Foley catheter** should be inserted to measure urine output in all children who need intravenous (IV) hydration. All wounds should be wrapped with **sterile towels**. If hypoxia or carbon monoxide poisoning is suspected, **100% oxygen** should be given. **An adequate airway** should be maintained.

434. d) Teaching of biobehavioral technique and coping strategies before cycle develops involving **negative** experience with health care workers. In severe cases, intravenous opoids and PCA (patient-controlled analgesia) are used. Amitrypline enhances sleep and effectiveness of opioids. Acupuncture may be considered for pain.

435. b) In **P**rader-Willi syndrome, the deletion is always of the paternally derived chromosome 15 (**P** and **P**) when there is deletion.

436. a) In Angelman syndrome, the deletion is always of the **maternally derived** chromosome 15 when there is deletion.

437. d) Sweaty feet, acrid odor in isovaleric acidemia

438. c) Maple syrup odor in MSUD

439. a) Hops-like odor in Oasthouse urine disease

440. b) Rotting fish odor in trimethylaminuria

441. c) Mongolian spots appear at birth as slate blue, well demarcated areas of pigmentation on the buttocks and back.

442. d) Lanugo appears at birth as fine, soft, immature hair on the scalp, brow, and face in premature infants.

443. b) Erythema toxicum appear 1-3 days after birth as small, white, occasionally vesicopustular papules on an erythematous base on the face, trunk, and extremities.

444. a) Benign pustular melanosis appears at birth as vesicopustular eruption on the neck, back, chin, palms, soles, and extremities.

445. c) Aplasia cutis congenita also occurs in trisomy 13, chromosome 4 deletion, or autosomal dominant inheritance.

446. e) Tufts of hair over the lumbosacral spine also noted in sinus tract or tumor (e.g., lipoma, teratoma).

447. a) Excessive skin fragility and extensibility with joint hypermobility are also noted in Marfan syndrome, disorders of collagen synthesis, or congenital contractural arachnodactyly.
448. b) Amnioniotic bands can also cause disruption of the skin; amputation, ring constriction, or syndactyly of the extremities; abdominal or thoracic wall defects.
449. d) Wormian bone formations are also noted cleidocranial dysostosis, lacunar skull, and sometimes, Down syndrome.
450. c) Hydrocele. Transillumination is negative in hernia.
451. b) In neonates with sinus bradycardia, heart rate is less than 90 beats/minute.
452. c) After neonatal period, heart rate less than 60 beats/minute represents sinus bradycardia
453. a) In neonates with SVT, heart rate is usually more than 230 beats/minute with abnormal P-wave axis. In other age groups with SVT, heart rate is above 180 and up to 300 beats/minute.
454. e) Reassurance
455. e) Reassurance
456. b) Renal (e.g., hydronephrosis, multicystic-dysplastic kidneys)
457. e) Renal vein thrombosis can occur in patients with antithrombin III deficiency, protein C deficiency, and in infants of diabetic mothers.
458. e) Eyes blinking. Apgar scores also include muscle tone. The reflex irritation (e.g., grimace, cough, or sneeze) is response to catheter in nostrils after the clearance of oropharynx.
459. b) Right pneumothorax. Answer (e) is wrong because the selective intubation into the left main bronchus is very difficult because of the anatomical position. Other answers are easy to understand.
460. e) Breast-feeding jaundice occurs after the 7th day of life, peaks during the 2nd-3rd weeks of life, and bilirubin reaches as high as 10-30 mg/dL.
461. a) Intravenous D_{10} glucose water.
462. a) Oral feeding with formula or breast milk should be started **as soon as possible** and continued every 3 hour intervals in infants of diabetic mother if they are clinically well and normoglycemic.
463. e) **Button batteries should be removed immediately** because they may cause esophageal mucosal injury within 1 hour of aspiration and involve all layers within 4 hours of aspiration.
464. b) **Asymptomatic patients with coin or blunt object lodged in the esophagus can be observed for up to 24 hours** in anticipation of passage of coin into stomach. Answer (a) is appropriate for symptomatic patients.
465. d) In asymptomatic patients, meat impactions in the esophagus can be observed up to 12 hours. Intravenous glucagon can be used in patients without prior esophageal surgery to fascilitate the passage of distal esophageal food boluses by decreasing the lower esophageal sphincter pressure.
466. a) In symptomatic patients, negative plain films indicate urgent endoscopic removal of the radiolucent foreign bodies (e.g., plastic, glass, wood, aluminum, and bones).
467. b) Early CT scan improves the diagnostic accuracy. CT scan is more sensitive and specific than ultrasonography. Abdominal distension indicates either perforation or obstruction. Once the diagnosis is established, the treatment is surgical appendectomy.
468. a) The CT scan of the abdomen is preferred in the diagnosis, localization, and percutaneous drainage of the abscess after appendectomy.
469. a) The newborn most likely had meconium aspiration because the child was depressed and required resuscitation at birth.
470. b) Cerebral palsy because the newborn has stage 3 hypoxic-ischemic encephalopathy. The stage 1 patients have good prognosis. The stage 2 patients have variable prognosis. The seizures are present in stage 2 and 3.
471. b) Hypokalemia. Chronic administration of furosemide also causes hyponatremia, and 'contraction alkalosis'.
472. b) Furosemide causes hyperuricemia; it also causes renal stones, GI intolerance, dehydration, hypokalemia, hyponatremia, and 'contraction alkalosis'.
473. b) Digoxin does not causes ototoxicity. Digoxin also causes vomiting, diarrhea, arrythmias, lethargy, vertigo, and photophobia.
474. e) Hypokalemia and hypercalcemia enhances digoxin toxicity.
475. c) Intrauterine P-RBCs transfusion
476. a) Zidovudine (AZT) and protease inhibitors
477. b) Folate and vitamins
478. e) Amnioinfusion
479. d) Betamethasone

480. d) Reassurance; petechiae occurs due to sudden increase of intrathoracic pressure as a result of normal delivery.
481. e) Reassurance. Linear fractures are the most common type of skull fractures. Patients are asymptomatic and require no treatment.
482. e) Elevation of the severely depressed skull bones are indicated to avoid cortical injuries.
483. b) Depressed fracture of the skull (right parietal bone)
484. c) When mother is addicted to opiates, narcan should not be used because it precipitates acute neonatal withdrawal with severe seizure activities. Answers (a) and (b) are indications of narcan therapy. Artificial ventilation should continue before and during the administration of narcan.
485. d) Hypertonicity is absent. When the respiration begins, it is extremely slow. These clinical manifestations can occur in a newborn whose mother received heavy doses of morphine, meperidine (Demerol), barbiturates, fentanyl, or tranquilizers shortly before delivery or from anesthesia during the 2nd stage of labor.
486. e) CNS stimulant drugs are contraindicated because they are harmful and ineffective.
487. b) Between 3-5%
488. e) 100%
489. e) Musculoskeletal
490. d) Fetal crowding is usually due to reduced volume of amniotic fluid. In breech presentation, the incidence of deformities is increased 10 times.
491. e) During 3rd trimester, fetuses have rapid growth.
492. b) 3-5%; the risk of recurrence in congenital hip dislocation is also 3-5%.
493. b) Microcephaly is absent. **Costello syndrome** also includes macrocephaly, loose skin, hyperkeratotic palms and soles, coarse face, papillomas (e.g., perioral, nasal, anal), and postnatal growth deficiency.
494. c) Kabuki syndrome appears with long palpebral fissures, preauricular pits, and postnatal growth deficiency.
495. c) Longer than 18 hours. Previous answer was longer than 24 hours.
496. e) Previous pregnancy with positive GBS screening culture but the culture should be negative in current pregnancy. Intrapartum prophylaxis (IP) is also **not** indicated in planned cesarean section in the absence of labor or rupture membrane regardless of maternal GBS culture results/or negative vaginal or rectal GBS screening culture during current pregnancy regardless of intrapartum risk factors. IP is indicated when maternal GBS status is unknown and maternal fever 100.4°F or delivery less than 37 weeks of gestational age.
497. a) Marijuana
498. a) Alcohol is CNS depressants. Alcohol also causes talkativeness, grogginess, increases the pain threshold, diuresis (inhibiting pituitary ADH), vasodilatation, and hypothermia (centrally mediated).
499. a) He should be drunk **six or more times** in the past year or if he had problems with the police that indicates problem drinker.
500. e) Ingestion of other drugs along with alcohol. The legal definition of alcohol intoxication means blood ethanol level is 80 or 100 mg/dL. The adolescent is at risk of death when blood ethanol level is greater than 200 mg/dL.
501. d) Respiratory depression
502. d) Dialysis should be considered when the blood alcohol level is higher than 400 mg/dL.
503. e) Tetrahydrocannabinol (THC) fraction of the resin causes hallucination.
504. e) Chronic marijuana users lose interest in age-appropriate behavior.
505. c) Acute marijuana use. Hallucinations and distortions of body image occur rarely but usually occur during stress or with fever.
506. e) Marijuana decreases intraocular pressure up to 5 hours in normal persons and patients with glaucoma. Tachycardia is noted within 20 minutes after smoking marijuana and is followed 30 minutes later by transient hypertension that disappears by 3 hours.
507. b) Diazepam and alcohol potentiate sedation.
508. c) Cocaine and amphetamine potentiate stimulation.
509. a) Propranolol and phenytoin have antagonistic effects.
510. c) Prolonged bleeding from circumcision sites, bloody diarrheas, atopic dermatitis, and S. pneumoniae infections.
511. b) Chronic sinopulmonary disease and high incidence of cancer
512. d) Fatal varicella infections, redundant skin, hyperextensible joins of the hands and feet but an inability to extend the elbow completely, short and pudgy hands.

513. e) Intractable diarrhea, exfoliative erythroderma, hepatosplenomegaly, and lymphadenopathy.
514. a) Recurrent staphylococcal abscesses (e.g., skin, joints, lungs), persistent pneumatoceles, coarse facial features, and hemihypertrophy
515. e) Allergy; acute allergic reaction may increase absolute eosinophil counts more than 20,000 cells/microL and chronic allergy reaction may rarely increase absolute eosinophil counts more than 2,000 cells/microL.
516. a) Idiopathic
517. b) Cardiac disease due to endomyocardial thrombosis and fibrosis
518. c) Corticosteroids; answer (a) or (b) is used when corticosteroids therapy fails.
519. a) Eosinophilia greater than 1,500 cells/microL persisting more than 1 year is an incorrect answer.
520. e) Bone marrow transplantation. Answer (a) is used before bone marrow transplantation. Answer (b) is used for aspergillus infections. Answer (c) is used for granulomas. Answer (d) is used for prophylaxis of infections.
521. e) Bone marrow transplantation. Answer (a) therapy improves the clinical conditions of some children when they are stable.
522. c) Large inclusions in all nucleated blood cells that is seen on Wright-stained blood films and are accentuated by peroxidase stain.
523. e) LAD-1 is diagnosed by flow cytometric measurements of surface CD1 1b in stimulated and unstimulated neutrophils using monoclonal antibodies.
524. d) LAD-2 is diagnosed by lack of sialyl-Lewis X on the neutrophils.
525. a) Myeloperoxidase (MPO) deficiency is diagnosed by deficiency of neutrophils and monocytes MPO.
526. b) For screening test of chronic granulomatous disease, DHR test is replacing nitroblue tetrazolium (NBT) dye test. DHR detects oxidant production by increasing fluorescence when oxidized by H_2O_2.
527. c) Allergic rhinitis has transverse nasal crease due to allergic salute (i.e., an upward rubbing of the nose with an open pump or extended fingers). Answer (a) is known as **allergic gape**. Answer (b) is known is **allergic shiners**.
528. b) This girl has allergic rhinitis. A thick, purulent nasal secretion indicates an infection.
529. d) Bed linen and blankets should be washed every week in hot water (**more than 130°F**). Answer (c) is the most effective strategy for the reduction of mite allergen. The avoidance and removal of allergens are indicated.
530. c) About 60% of patients with allergic rhinitis have asthma and even those who do **not** manifest with bronchial hyperresponsiveness. Chronic sinusitis is a common complication of allergic rhinitis.
531. c) Anticonvulsants (e.g., phenytoin, phenobarbital, carbamezepine) increase the metabolism of glucocorticoids.
532. b) Rifampin enhances glucocorticoid clearance.
533. d) Ketoconazole and oral contraceptives delay glucocorticoid metabolism.
534. a) Erythromycin and clarithromycin delay glucocorticoid clearance.
535. a) Long-term inhaled glucocorticoid therapy has no effect on adult attained height or on bone mineral density. However, this therapy decreases the height by 1.1 cm in the first year of therapy indicating that the growth suppression is transient phenomenon.
536. b) Local immunosuppression
537. e) Hyponatremia is absent.
538. e) Pulse oximetry is indicated because frequent beta-agonist therapy can produce ventilation-perfusion mismatch and hypoxia. Hypoxic patients should receive oxygen therapy.
539. a) Hypotension. Blood pressure should be monitored every 10-15 minutes during the infusion and for up to 90 minutes after the infusion.
540. e) Fluid above maintenance is a wrong answer because asthma increases ADH secretions.
541. d) IM magnesium sulfate should not be given.
542. e) Heliox is used in hospitalized patients. Inhaled beta-agonist should be given every 20 minutes for 1 hour, if needed, systemic (oral or IV) glucocorticoid is given. Inhaled ipratropium may be added if the 1st dose response of beta-agonist is inadequate.
543. a) Immunizations neither decrease nor increase the development of asthma. All childhood immunizations should be given to all asthmatic children including varicella and annual influenza vaccines. An early life microbial exposures may be associated with reduced allergen sensitization, atopic dermatitis, asthma, and bronchial hyperresponsiveness.
544. e) Glucocorticoids decrease IgG levels.

545. a) Patients with late-onset morbilliform rashes with amoxicillin are not at risk for IgE-mediated reactions to penicillin and do not require skin testing before penicillin therapy. Carbapenems (e.g., imipenem, meropenem) have high degree of cross reactivity with penicillin and are another class of beta-lactam antibiotics. Only aztreonam can be used safely.

546. a) **Nonvesicular**, polymorphous rash primarily over the trunk is noted in patients with a Kawasaki disease. Answers (b), (c), (d), and (e) are the manifestations of **acute febrile phase**, which usually last 1-2 weeks, of Kawasaki disease.

547. e) ESR returns to **normal** in convalescent phase that is 6-8 weeks after the onset of illness. Anorexia, irritability, and conjunctival injection may persist in **subacute phase when there is highest risk of sudden death**.

548. a) The platelet count is normal in the 1st week of illness, then rapidly increases by 2nd-3rd weeks of illness. Elevated C-reactive protein and other acute phase reactants are noted in acute phase of the disease. Antinuclear antibody and rheumatoid factor are absent. Sterile pyuria, CSF pleocytosis, and mild elevations of hepatic transaminases may be present.

549. b) **First echocardiogram** should be performed **at the time of diagnosis**. If the result is normal, **the 2nd echocardiogram** should be performed **after 2-3 weeks** of illness. If both the results are normal, the **3rd echocardiogram** should be performed **by 6-8 weeks** after the onset of illness. If the result is normal, there is no need to repeat the test again.

550. e) Consult with pediatric cardiologist

551. b) Wegener granulomatosis should be suspected in a child with chronic sever sinusitis, pulmonary granulomas, and nephritis.

552. a) S. aureus produces **TSST-1 toxin** that is the most common cause of toxic shock syndrome. However, TSST-1-negative strains have been isolated from patients with TSS (especially nonmenstrual) i.e., **enterotoxin** has role in producing TSS.

553. a) Toxic shock syndrome

554. a) Menstruation and tampon use/or abscess

555. c) Nafcillin or first-generation cephalosporin is the drug of choice. Clindamycin is added in unresponsive cases. Fluid therapy should be very aggressive. Inotropic agent is used for hypotension. Corticosteroids and intravenous immunoglobulin are used in severe cases. Drainage of the vagina is indicated in tampon-induced menstrual TSS.

556. d) Vicilin and conglutin are proteins of peanuts.

557. c) Ovomucoid is the protein of eggs.

558. b) Casein and beta-lactoglobulin are proteins of cow's milk.

559. a) Paralbumin is the protein of fish.

560. c) Avoid cow's milk until 1 year of age

561. b) Renal injury

562. b) Hepatic injury or failure

563. e) Gentamicin does not cause neutropenia.

564. c) Severe congenital **neutropenia** or Kostman syndrome (or disease)

565. a) Diarrhea usually occurs.

566. e) The patients with cartilage-hair hypoplasia appears with **macrocytic** anemia, hyperextensible fingers, abnormalities of spine, and increased rates of cancer.

567. e) Staphylococcus aureus and gram-negative bacteria are the most common organisms.

568. e) The patients with leukoencephaly manifest with **decerebrate** posturing. MRI or head CT scan reveals multifocal areas of white matter degeneration with necrosis. Leukoencephalopathy occurs exclusively in patients who received cranial irradiation or intrathecal chemotherapy.

569. b) Driver error (75-80% of cases); alcohol is a factor in approximately 33% of cases.

570. a) Automobile and motorcycle accidents

571. b) Diabetes insipidus. Obesity is correlated with diabetes mellitus and is a major risk factor for cardiovascular disease in adulthood.

572. c) A child is considered overweight when the BMI is more than 85th percentile and is considered obese when the BMI is more than 95th percentile.

573. a) Low or below average levels of serotonin 5-HT (hydroxytryptamine) or the metabolite 5-HIAA (hydroxyl-indoleacetic acid) may be associated with suicide due to poor impulse control.

574. d) Heterosexual youth is not a risk factor of suicide. Other community risk factors of suicide are local suicide and violent exposure rates, availability of firearms, suicide prevention programs, and supportive social networks.

575. a) Unknown

576. e) NEC

577. a) Multifactorial but the exact cause of NEC is unclear.
578. a) Gastric retention with abdominal distension
579. d) Plain abdominal x-ray reveals pneumatosis intestinalis that is a diagnostic finding in patients with NEC.
580. c) Portal vein gas; answers (a), (b), and (e) are also noted in patients with NEC. Answer (d) is noted in patients with intestinal obstruction.
581. e) Umbilical catheters should be removed and peripheral intravenous access should be maintained.
582. e) A cross-table lateral abdominal x-ray reveals free air between abdominal wall and abdominal contents in patients with a perforation.
583. b) Perforation. Other indications for surgery are failure to respond to medical management, palpable mass, erythema of abdominal wall and a single, fixed bowel loop. Preferably, surgery should be performed after necrosis but before perforation and peritonitis.
584. a) Early use of steroids and indomethacin can cause of idiopathic focal intestinal perforation. These patients are less sick than those with NEC.
585. e) Hypertonia is noted during the phase 2 (middle of 1st week) and phase 3 (after the 1st week).
586. c) Seizures are absent in chronic form of kernicterus.
587. e) Sensorineural hearing loss is noted after first year of chronic form of kernicterus.
588. d) Exchange transfusion with PRBCs (packed red blood cells) should not be performed because P-RBCs contain high hematocrit (66%). However, a whole blood has hematocrit 33%.
589. c) The bronze baby syndrome has both direct and indirect hyperbilirubinemias with significant elevation of direct-reacting bilirubin. The discoloration is due to photo-induced modification of porphyrins due to cholestatic jaundice. The discoloration may last for many months.
590. a) PRBCs transfusion is indicated in asymptomatic infants with hemoglobin 7 or less, hematocrit 20 or less, and absolute reticulocyte count less than 100,000 cells/μL **or** infants require moderate to high mechanical ventilation (MAP more than 8 cm H_2O, FiO_2 more than 40%), hemoglobin 11 or less, hematocrit 35 or less **or** infants require minimal respiratory support (low mechanical ventilation, endotracheal/NCPAP more than 6 cm H_2O and FiO_2 40% or more), hemoglobin 10 or less, hematocrit 30 or less **or** infants require supplemental oxygen or CPAP (40% oxygen or less with any one of the following condition (e.g., undergoing operation, heart rate more than 180, respiratory rate more than 80, persistent episodes of apnea and bradycardia requiring bag and mask ventilation, increased oxygen requirement) with hemoglobin 8 or less and hematocrit 25 or less.
591. b) Furosemide does not cause gynecomastia. However, spironolactone causes gynecomastia.
592. b) Staphylococcus epidermidis (more than 50% of cases), S. aureus (20% of cases), and gram-negative bacilli (15% of cases)
593. a) Gram-negative organisms; mixed infection is common.
594. a) Abdominal symptoms are due to peritonitis. The distal infection is most likely either due to perforation of the bowel at the time of VP shunt operation or movement of bacteria through the bowel wall.
595. a) S. aureus
596. b) Prophylactic antibiotics do not reduce the infection rate for long-term indwelling urethral catheters.
597. c) Coagulase-negative staphylococcus and S. aureus infections are equally divided.
598. c) Staphylococcus aureus
599. d) S. aureus
600. e) There is no specific laboratory test. Cultures from abscess or vagina usually reveal S. aureus but are not required to make the diagnosis.
601. c) Hypospadius. This is a megameatal variant of hypospadius and may not be diagnosed until after a circumcision is performed due to normally developed foreskin.
602. d) Reassurance. Boys with distal hypospadius and no functional abnormalities do not need surgical repair.
603. e) Z-plasty is usually performed by 3 months of age.
604. e) Phenytoin therapy can cause gingival enlargement. Cyclosporin also causes gingival hyperplasia.
605. a) Vitamin D-dependent rickets can cause enamel hypoplasia.
606. b) Neutrophil chemotactic deficiency can be associated with juvenile periodontitis.
607. d) Immunosuppression causes oral candidiasis.
608. c) Kidney failure causes dysmorphic facial features in newborns due to pressure effect from oligohydramnios.
609. e) Premature eruption of permanent teeth is the most common cause of premature loss of primary teeth.

610. c) Porphyria causes a red-brown discoloration.
611. b) Neonatal hyperbilirubinemia may produce blue to black discoloration of the primary teeth.
612. a) Tetracycline produces brown-yellow discoloration and hypoplasia of the enamel.
613. e) Children may refuse to feed. The tongue may become lacerated and rarely, the tip is amputated that is called Riga-Fede disease. Natal teeth are noted at birth. Neonatal teeth erupt in the first month of life.
614. e) Turner syndrome patients do not have delayed eruption of the primary teeth. Other causes of delayed eruption of the primary teeth are familial, rickets, incontinentia pigmenti, Albright osteodystrophy or multiple syndromes, especially Down syndrome.
615. b) Pyloric stenosis is associated with epidermolysis bullosa and is usually manifest in early infancy.
616. d) High intestinal obstruction. When there is a history of polyhydramnios, the infant's stomach should be aspirated immediately after birth. Aspiration of 15-20 mL or more gastric contents, especially if it is bile stained, is suggestive of a high intestinal obstruction (i.e., duodenum, proximal jejunum).
617. c) Hypochloremic metabolic alkalosis
618. e) Leukopenia with thrombocytosis does not occur.
619. e) Bleeding from esophageal varices
620. e) Caloric requirements up to 150 kcal/kg/day
621. e) Hepatitis C vaccination is not available.
622. e) Infections and organ rejections are the most common complications after the early phase.
623. e) Thrombosis of great vessels causes primary nonfunction of the graft and can lead to death unless the graft is replaced within 48 hours.
624. a) Bacterial infections are most common, followed by viral (e.g., CMV, adenovirus), fungal, and rarely, parasitic (e.g., Pneumocystis carinii) infections.
625. b) Liver (6 to 24 weeks)
626. a) Yolk sac (2 to 6 weeks)
627. c) Bone marrow (24 to 42 weeks, and thereafter)
628. a) Diazoxide causes hyperglycemia, hypotension, and tachycardia.
629. d) Hydrochlorthiazide causes hyperuricemia, hypercalcemia, and hypokalemia.
630. b) Enalaprilat causes transient hypotension.
631. c) Labetalol causes bronchospasm, orthostasis, and dizziness.
632. a) Prazocin causes first dose, orthostatic hypotension.
633. b) V2 receptors located primarily in the collecting tubule, thick ascending limb of the loop of Henle, and the periglomerular tubules of the kidney.
634. b) Long arm of the X chromosome (Xq28)
635. a) Lymphocytic thyroiditis (Hashimoto thyroiditis, autoimmune thyroiditis) is the most common cause of thyroid disease. Endemic goiter is due to dietary deficiency of iodine. Iodide goiter is due to administration of iodine containg cough medicine.
636. e) Lymphocytic thyroiditis is the most common cause of acquired hypothyroidism.
637. b) Lymphocytic thyroiditis
638. c) Mitral regurgitation
639. e) The presence of TRSAb establishes the diagnosis of Graves disease and disappearance of TRSAb indicates remission of the disease. However, the measurement of TRSAb is **not** necessary to make the diagnosis of Graves disease but is useful in equivocal cases.
640. e) Advanced skeletal maturation, craniostenosis, and decreased bone density; bone density returns to normal after therapy. **Craniostenosis** is a malformation of the skull caused by premature closure of the cranial sutures. **Craniosynostosis** is a premature fusion of the sutures of the skull.
641. a) **Dandy-Walker syndrome** is a cystic dilatation of the fourth ventricle in the posterior fossa due to a developmental failure of the roof of the fourth ventricle. About 90% of patients have hydrocephalus. **Carpenter syndrome** is known as acrocephalopolysyndactyly. **Apert syndrome** is known as acrocephalopolysyndactyly. **Kleeblattschadel syndrome** is known as cloverleaf skull. **Crouzon disease** is known as dysostosis craniofacialis.

642. b) Corticosteroids characteristically produce posterior subcapsular cataract.
643. a) Wilson disease causes sunflower cataract but is not commonly seen in children.
644. d) In classic infantile galactosemia (galactose-1-phosphate uridyl transferase deficiency), the cataract is typically of the zonular type. In galactokinase deficiency, cataract is zonular and is the sole clinical manifestation.
645. c) Anterior capsular cataract is associated with persistent strands of pupillary membrane or vascular sheath of the lens.
646. e) Posterior capsular cataract is associated with persistent remnants of the primitive hyaloid vascular system (the common Mittendorf dot).
647. a) Cataract of prematurity appears with a cluster of tiny vacuoles in the distribution of Y sutures of the lens.
648. e) Hypercalcemia does **not** cause cataract.
649. a) Parvovirus B19 infection causes hydrops and severe anemia but does **not** cause cataract.
650. a) Amoxicillin
651. b) Azithromycin is also used in children in whom palatability or convenience of administration are also important.
652. e) Amoxicillin clavulanate, cefuroxime-axetil, or intramuscular ceftriaxone is the second-line treatment that is indicated when patients have an unsatisfactory response to first-line treatment.
653. d) Diarrhea; yogurt use can reduce diarrhea in some cases.
654. c) Cefuroxime-axetil is not palatable.
655. e) S. aureus
656. b) Prophylactic systemic antibiotic is not indicated in patients with Stevens-Johnson syndrome. Management of this syndrome is supportive and symptomatic. Antibiotic therapy is used for a secondary bacterial infection. Most authorities prefer **not** to use corticosteroid therapy that enhance mortality due to sepsis. The drug should be discontinued in drug related Stevens-Johnson syndrome. Intravenous immunoglobulin may be used. In females, vaginal lesion should be observed closely to avoid vaginal stricture and fusion. Opthalmologic consultation is mandatory because corneal scars can lead to vision loss.
657. e) Mycoplasma pneumoniae
658. d) Vancomycin does **not** cause Stevens-Johnson syndrome. The most common offending drugs are sulfonamides, NSAIDs (e.g., ibuprofen, salicylates, butazones), and phenytoin.
659. e) Pemphigus vulgaris usually first appears as painful oral ulcers, which may be the only finding of the disease for weeks or months, followed by large, flaccid bullae emerge on nonerythematous skin on the face, trunk, groin, axialla, and pressure points. Nikolsky sign is present.
660. b) Marcus Gunn phenomenon (i.e., sucking jaw movements causing simultaneous eyelid blinking) is due an abnormal innervation of the trigeminal (5th) and oculomotor (3rd) cranial nerves.
661. a) Mobius syndrome patients have normal mental development.
662. d) Turner syndrome does not cause microcephaly. Rubinstein-Taybi, Smith-Lemli-Opitz, fetal alcohol, and fetal dydantoin syndromes also produce microcephaly.
663. e) Aqueductal stenosis
664. b) Subarachnoid hemorrhage
665. b) Dandy-Walker malformation appears with prominent occiput and CT/MRI study reveals large posterior fossa cyst.
666. a) Chiari malformation (type II) appears with foreshortened occiput and MRI/CT scan reveals cerebellar tonsil protruding downward into the cervical canal.
667. c) Aqueductal stenosis appears with large head and MRI/CT scan reveals dilated lateral ventricles.
668. a) Chiari malformation (type I) is usually **not** associated with hydrocephalus. Type I includes herniation of cerebellar tonsils into the cervical canal. Pathogenesis is unknown.
669. e) Benign paroxysmal vertigo is rare beyond 3 years of age. This is a typical presentation. Neurological examination is unremarkable except for the finding of abnormal vestibular function noted during ice water caloric testing. The preferred therapy is diphenhydramine.
670. e) The preferred therapy for cyanotic spells is reassurance and support of the parents. Both parents should be consistent and not to reinforce the similar behavior after the child recovers from the spell. Parents should put the child safely in bed. Parents should **not** play, hold, or cuddle the child until recovery is complete.
671. e) The child has couth syncope. This is a typical presentation. The child did not remember the attack except coughing. The therapy includes aggressive management to prevent bronchospasms.
672. a) The **less important** causes of headache in children are refractive errors, sinusitis, strabismus, and malocclusion of the teeth.

673. a) Common migraine (migraine without aura)

674. c) Common migraine is not associated with aura. In addition to the symptoms already mentioned, the child may experience paleness, photophobia, osmophobia (strong dislike to odors), phonophobia (dislike to sounds), and paresthesias of the hands and feet. A strong family history (90% of cases) is suggestive of common migraine.

675. d) Classic migraine has an aura precedes the onset of headache. Distortions of body image is a predominant clinical finding.

676. b) Migraine variants include cyclic vomiting, acute confusional states, and benign paroxysmal vertigo. Vomiting may lead to dehydration and electrolyte imbalance.

677. a) Complicated migraine presents with neurologic signs during and after the headache. Some patients have syndrome of alternating hemiplegia (i.e., one side to the other).

678. e) Monosodium glutamate is added in Chinese food and causes headache. Migraines can be prevented by avoiding certain stimuli (stress, fatigue, anxiety, bright lights, loud noise, nuts, chocolates, hot dogs, Chinese food, alcohol, and oral contraceptives).

679. b) Autosomal dominant

680. d) Café-au-lait spots are noted in 100% of NF-1 patients. However, they are less common in NF-2.

681. a) Bilateral acoustic neuromas are the most distinctive findings in patients with NF-2.

682. a) Scoliosis; answers (d) and (e) are also noted in patients with NF-1.

683. e) Flashlamp-pulsed laser therapy

684. b) Conservative therapy is indicated in patients with Sturge-Weber disease who have normal or near-normal mental development and well-controlled seizure activities. Hemispherectomy or lobectomy may prevent the development of mental retardation in patients with recalcitrant seizures but the surgery should be performed during the 1st year of life.

685. e) High-dose intravenous methylprednisolone (30 mg/kg bolus followed by 5.4 mg/kg/hour for 24 hours) should be started STAT, even before the transport. If steroids are given within 3-8 hours after **injury of the spinal cord**, therapy should be continued for 48 hours. These are the signs and symptoms of spinal cord compression.

686. e) The child has no atlantoaxial instability. Atlantoaxial instability is defined as a distance **greater than 4.5 mm** between the odontoid process of the axis and the anterior arch of the atlas.

687. d) Tethered cord appears with a midline skin lesion (70% of cases) such as lipoma, tuft of hair, hyperpigmentation, dermal pit or cutaneous hemangioma. Plain x-ray usually reveals spina bifida in the lumbosacral region. In normal children, conus medullaris should be at the level of L1. In children with tethered cord, thickened ropelike filum terminale persists and pulls the conus at or below the L2 level.

688. d) Transverse myelitis. This is a typical manifestation of transverse myelitis. CSF protein may be slightly elevated. MRI is the preferred diagnostic study. About 60% of cases recover spontaneously over a period of weeks or months. Therapy includes bladder care and physiotherapy. Corticosteroids are **not** useful.

689. d) Chlamydia trachomatis infection does not cause transverse myelitis. Other infectious causes of transverse myelitis are Lyme disease, mumps, rubella, and influenza.

690. e) Retinopathy of prematurity

691. d) Retinoblastoma

692. c) Retinitis pigmentosa

693. a) Retinal phakomas (hamartomatous disorders) are noted in tuberous sclerosis.

694. b) Proliferative diabetic retinopathy

695. e) Surgery is the preferred therapy for infantile (or congenital) glaucoma that begins within the first 3 years of life.

696. e) The classic triad of infantile glaucoma includes epiphora (tearing), photophobia (sensitivity to light), and blepharospasm (eyelid squeezing). Only 30% of patients have signs and symptoms of classic triad. Other signs are visual impairment, conjunctival irritation, ocular enlargement, corneal enlargement, and corneal edema.

697. a) Optic nerve glioma (or juvenile pilocytic astrocytoma) is a benign and slowly progressive tumor.

698. e) Sensorineural hearing loss

699. e) Hemangiomas and dermoids are the most common benign tumors of the orbit.

700. c) Adenocarcinoma

701. e) Slipped capital femoral epiphysis

702. e) Idiopathic

703. e) Surgical therapy (i.e., correction of deformity with permanently placed internal fixation rods and posterior fusion of the involved vertebrae) is indicated when a curve is greater than 45 degrees.

704. e) Asymmetry of the posterior chest wall on forward bending **(the Adams test)**; answers (a), (b), (c), and (d) are also physical findings in patients with idiopathic scoliosis.

705. e) Standing posteroanterior and standing lateral radiograph of the entire spine should be obtained to determine the degree of curvature by measuring the angular relationships between the most tilted vertebrae at either end of the curve (the Cobb method). The CT scan and MRI are performed in patients with nonidiopathic scoliosis to determine the underlying cause.

706. b) The chiropractic therapy or exercise program does not improve the outcome of scoliosis. Transcutaneous electrical stimulation therapy is **not** effective. Curves greater than 45 or 50 degrees usually progress during adult life. Curves less than 30 degrees usually does not progress after complete skeletal maturation. Before menarche girls with curves between 20 and 30 degrees have a higher chance of progression than that of girls 2 years after menarche. Boys and girls usually have an equal risk of progression with same curves.

707. e) Figure-of-eight clavicle strap. The fracture usually heals in 3-6 weeks.

708. b) Posterior dislocation

709. c) Falling backward onto the outstretched arm with the elbow extended is the mechanism of supracondylar humeral fracture and elbow injuries.

710. d) Brachial artery; answers (a), (b), (c), and (e) can also occur in supracondylar humeral fracture.

711. e) Vascular and skeletal stability; orthopedic consultation should be performed immediately.

712. b) Distal radius

713. e) Neck injury

714. b) Anterior dislocation of the shoulder can also occur when a person falls onto an outstretched hand with a straight arm. There is a hollow region inferior to the acromion and bulge in the anterior portion of the shoulder.

715. e) Acromioclavicular separation of the right shoulder

716. a) About 15% of patients younger than 21 years of age with Down syndrome have AAI.

717. a) Dry, hot skin is absent in sports-related heat stroke but is present in '**classic**' heat stroke that is noted in chronically ill patients those who are exposed to summer heat waves.

718. e) Intravenous fluid (e.g., dextrose/saline solution) should be given at a rate of **800 mL/m2 in the first hour** in order to improve the intravascular volume.

719. d) Salt pills should not be used because they cause hypernatremia and delayed gastric emptying. Water (e.g., plain cold water) is adequate for exercise lasting less than 1 hour. Warm water should **not** be used because it causes delayed gastric emptying. Sports drinks usually contain 6-8% carbohydrate and can decrease nausea, bloating, and abdominal pain. Prepractice and postpractice weight can be useful to measure the fluid loss. Thirst is not an adequate indicator of dehydration because it appears at 2-3% dehydration that decreases atheletic performance.

720. e) Osteopenia and stress fracture can occur. A long-term persistence of osteopenia can lead to osteoporosis.

721. e) Estrogen replacement therapy, usually in the form of low dose oral contraceptive pills is recommended if amenorrhea persists for 6 months or longer. Bone mineral density should be measured to diagnose osteopenia. If amenorrhea persists more than 12 months, therapy with estrogen/progestin pills may be given regardless of the bone density findings. The American Academy of Pediatrics committee recommends not to give estrogen until 16 years of age. Estrogen therapy improves bone density and decreases adult height.

722. b) 1500 mg/day that is 300 mg/day more than the daily allowance for calcium in women with normal menstruation (i.e., 1200 mg/day).

723. c) Herpes simplex (herpes gladiatorum) and impetigo are contraindications to wrestling.

724. e) Knee injury

725. b) "Tennis elbow" or lateral epicondylitis is produced by repetitive contraction of the extensor muscles (e.g., extensoe carpi radialis brevis).

726. b) Hydrocephalus

727. d) Blue sclera

728. e) Genital hypoplasia and hypertelorism

729. c) Omphalocele

730. a) Renal cysts and intestinal atresia

731. d) Vitamin E deficiency; chronic cholestasis also causes vitamin A, D, and K deficiencies.

732. c) The normal ratio of serum vitamin E to total serum lipid is more than 0.6 in patients younger than 12 years and is more than 0.8 in patients 12 years and older. Vitamin E levels can be falsely elevated due to elevated blood lipid levels in patients with cholestasis.

733. e) Stagnant loop syndrome (blind loop syndrome, bacterial overgrowth syndrome) can occur after intestinal postsurgical adhesion. This syndrome causes both vitamin B_{12} deficiency (e.g., megaloblastic anemia) and disaccharidase deficiency (e.g., steatorrhea that is pale, foul-smelling, bulky stools).

734. e) Metronidazole is initially given for 2-4 weeks. If there is a relapse, metronidazole is used for 4-8 weeks. A surgery is indicated in patients with a partial small bowel obstruction. Answers (a) and (b) may be used in older teenagers. Answers (c) and (d) can be used for gram-negative bacterial infections.

735. d) Breast milk has low levels of vitamin K. Breast milk also contains long-chain fats, free amino acids, glutamine, and growth hormone.

736. b) Gallstones (60% of cases) but gallstones are not found in patients with intact ileum. All answers are also complications of short bowel syndrome.

737. e) Bleeding is absent. Answers (a), (b), (c), and (d) are long-term complications of short bowel syndrome that also include late vitamin B12 deficiency (i.e., may not appear until 1-2 years of age).

738. d) Feeding colitis; pathogenesis is unknown but it is a benign condition; usually respond to hypoallergenic diet or to sulfasalazine. In neonates, stricture after NEC can produce bloody stools.

739. e) Protein **gluten** is found in wheat, rye, and barley. The activity of gluten resides within **gliadin** fractions which contain amino acid sequences (**motifs**) that sensitize lamina propria lymphocytes.

740. a) Failure to thrive; answers (b), (c), (d), and (e) are common symptoms of celiac disease.

741. a) Wasted muscles; answers (b), (c), (d), and (e) are common signs of celiac disease.

742. e) Small intestinal biopsy and histologic confirmation (i.e., villus atrophy, crypt hyperplasia, and surface epithelium injury) is the gold standard. A rechallenge with gluten is recommended for **children younger than 2 years** when the diagnosis is in doubt because other conditions can lead to flattened mucosa. A rechallenge with gluten is **not** recommended for children 2 years of age and older.

743. d) Lifelong, strict gluten-free diet (i.e., all wheat, rye, and barley products should be eliminated). The child should receive vitamin and iron. Pancreatic enzyme is useful to gain weight in the first few months of therapy.

Corticosteroids are used in celiac crisis (i.e., weight loss, severe diarrhea, hypocalcemia, and hypoproteinemia).

744. e) Reassurance; children **younger than 2 years** of age without any predisposing condition usually do not require any treatment because this condition is self-limited. Conservative management (observation) is accepted in this age group with fistula because it disappears spontaneously. Rarely, incision and drainage under local anesthesia are indicated in patients who are very uncomfortable. Antibiotics are not useful in this age group.

745. e) Observation is **not** appropriate for children 2 years of age and older with a predisposing condition. They should be treated aggressively.

746. a) Surgical treatment is indicated if medical therapies fail. This is the preferred surgical procedure for patients with rectal prolapse.

747. a) Laparotomy and internal fixation of the rectum to the presacral fascia is the preferred therapy in patients with procidentia and meningomyelocele. **Rectal procidentia** means extrusion of all layers of the rectal wall. **Rectal prolapse** means extrusion of mucosal layer of the rectal wall.

748. e) Cystic fibrosis

749. c) Elevated serum levels of trypsinogen

750. a) A qualitative examination of the stool for microscopic fat globules. Random stool samples can have both false-positive and false-negative results. Therefore, a 72-hour collection of stools for quantitative analysis of fat content is preferable. Stools should be kept in a freezer. **Normal fat absorption is greater than 93% of intake**.

751. e) Direct stimulation of the pancreas with secretion and cholecystokinin, use a triple-lumen tube to collect pancreatic secretion, and measurement of bicarbonate levels and enzyme activities (e.g., trypsin, chymotripsin, lipase, amylase).

752. a) Ventilation-perfusion mismatch (or decreased) i.e., alveolar PO_2 is lower (e.g., 60) than normal (e.g., 100), therefore blood that passes through this unit of alveoli achieve lower (e.g., 89%) than normal (e.g., 95-100%) oxygen saturation. Supplemental oxygen therapy increases alveolar PO_2 and arterial PO_2.

753. d) Infants have less outpatient visits for ear infection at 3 and 6 months for those who are sleeping supine.

754. a) Surgery. Antral choanal polyps do not respond to medical therapy and are not associated with underlying disease process. Answers (b), (c), (d), and (e) are not effective therapies.

755. e) Functional endoscopic sinus surgery includes removal of polyps and other associated nasal disease. Answers (a) and (b) provide some shrinkage in nasal polyps. Answers (c) and (d) do not shrink nasal polyps but reduce mucosal edema.

756. d) Rhinovirus

757. e) Otitis media (5-30% of cases)
758. d) Good handwashing; in hospitals, protective face shields can prevent hand-to-eye and hand-to-nose contacts.
759. a) Pertussis causes persistent or paroxysmal cough.
760. d) Allergic rhinitis causes itching, sneezing, and nasal eosinophilia.
761. b) Streptococcal nasopharyngitis causes nasal discharge that excoriates the nares.
762. e) Foreign body in the nose causes unilateral, foul-smelling nasal discharge.
763. c) Sinusitis causes persistent rhinorrhea or cough for more than 10-14 days, headache, facial pain, or periorbital edema.
764. e) Rhinitis medicamentosa is a rebound effect that produces a sensation of nasal obstruction when the medication is discontinued.
765. e) Diaphragmatic defect results in passage of intraabdominal air into the pleural cavity. This girl has catamenial pneumothorax.
766. a) Staphylococcus aureus; Mycoplasma pneumoniae infection can cause pneumothorax.
767. e) Rupture of thoracic duct during cardiac surgery for complex congenital heart diseases. Answers (a), (b), (c), and (d) can cause chylothorax.
768. b) Autosomal dominant
769. a) Autosomal recessive. Jeune syndrome is called asphyxiating thoracic dystrophy (thoracic-pelvic-phalangeal dystrophy).
770. c) **Narrow** foramen magnum causes respiratory dysfunction in patients with achondroplasia. Achondroplasia may also manifest with kyphoscoliosis during infancy, decreased anteroposterior diameter of chest with anterior cupping of the ribs, small lungs but normal functions except reduced vital capacity.
771. d) Balloon angioplasty
772. a) Pulmonary circulation is decreased significantly in patients with critical PS. They appear with right-sided heart failure.
773. e) Patients with critical AS manifest with hypotension and subsequently, total circulatory collapse. They appear with both left-sided and right-sided cardiac failure.
774. b) Secundum ASD (atrial septal defect)
775. e) Pulmonary stenosis (e.g., pulmonary valve or branch pulmonary artery) is common in patients with arteriohepatic dysplasia (Alagille syndrome).
776. d) Cardiac anomalies and aortic **coarctation**
777. a) CATCH 22 syndrome can be associated with tetralogy of Fallot, pulmonary atresia with VSD (a severe form of TOF), or interrupted aortic arch.
778. d) Cleft **palate**
779. c) Phrenic nerve injury causes diaphragmatic paralysis.
780. d) Recurrent laryngeal nerve injury causes vocal cord paralysis.
781. a) Sympathetic nerve chain injury causes Horner syndrome.
782. b) Hyponatremia causes seizures that are also produced by hypoglycemia, CNS ischemia, and emboli.
783. e) Spinal artery ischemia after repair of coarctation causes paraplegia.
784. b) Postcoarctectomy syndrome causes hypertension and mesenteric arteritis.
785. d) Repair of TAPVR (total anomalous pulmonary venous return) can cause pulmonary hypertension that can occur trisomy 21 and Norwood 1st stage operation.
786. e) ARDS postpump syndrome can be due to possible release of vasoactive substances but the exact mechanism is unknown.
787. a) SIADH produces hyponatremia.
788. c) Serous postpericardiotomy syndrome produces pericardial tamponade.
789. b) WPW syndrome most often present in patients with a normal heart.
790. a) In type 1 von Willebrand disease, **DDAVP** induces the release of VWF (von Willebrand factor) from the endothelial cells. **VWF concentrates** are used in type 3, 2A, 2B, 2N, 2M and type 1. **Both DDAVP and VWF** concentrates are used in type 1, 2A, and 2N. **Platelets** are used in PT-VWD (platelet-type pseudo VWD) and BSS (Bernard Soulier syndrome) VWD.
791. e) Idiopathic (autoimmune) thrombocytopenia
792. e) IVIG (0.8-1 g/kg/day x 1-2 days) therapy increases the platelet count (over 20,000) in 95% of patients within 48 hours. Prednisone (1-4 mg/kg/day) is used for 2-3 weeks or until a rise in platelet count over 20,000. The platelet transfusion in ITP is contraindicated unless life-threatening bleeding is present. The patients may have normal size platelet in peripheral blood smear and have normal number of megakaryocytes in bone marrow.

793. e) IVIG therapy may cause headaches and vomiting (i.e., aseptic meningitis)
794. e) Prednisone therapy may cause lymphopenia and neutrophilia.
795. c) Chromosome 11p deletion with sporadic aniridia may cause Wilms tumor.
796. b) Chromosome 13q deletion may cause retinoblastoma and sarcoma.
797. d) Dysplastic nevus syndrome may cause melanoma.
798. a) Xeroderma pigmentosa may cause basal and squamous cell cancers.
799. e) Fanconi anemia may cause leukemia.
800. b) EBV can cause B-cell lymphoproliferative disease in immunocompromised individuals, especially those with AIDS.
801. a) Hepatitis C virus can cause splenic lymphoma and hepatocellular carcinoma. Hepatitis B virus can cause hepatocellular carcinoma.
802. c) HHV8 is associated with Kaposi sarcoma, primary effusion B-cell lymphoma, and plasma cell variant of Castleman disease, all of which occur primarily patients with AIDS.
803. d) HPV causes cervical cancer.
804. e) Neonates with Down syndrome can develop transient leukemia. They do not need chemotherapy. They may need temporary transfusions. A close follow-up is indicated because 20-30% will develop typical leukemia within the first few years of life.
805. e) Cowden syndrome can be associated with Dysplastic gangliocytoma of the cerebellum (Lhermitte-Duclos disease).
806. c) Neurofibromatosis 1 can be associated with optic pathway gliomas, astrocytomas, and malignant peripheral nerve sheath tumors.
807. b) Neurofibromatosis 1 can be associated with vestibular schwannomas, meningiomas, spinal cord ependymomas, and spinal cord astrocytomas.
808. d) Tuberous sclerosis can be associated with subependymal giant cell astrocytomas and cortical tubers.
809. a) von Hippel-Lindau can be associated with hemangioblastoma.
810. b) Neuroblastoma may appear as a paraneoplastic syndrome of autoimmune origin presenting as ataxia or opsomyoclonus ("dancing eyes and dancing feet").
811. d) Abdomen (e.g., adrenal gland or retroperitoneal sympathetic ganglia); a firm, nodular mass is palpable in the flank or midline. The patient complains of abdominal discomfort. The plain x-ray and CT scan reveals a mass with calcification and hemorrhage.
812. e) Lung metastases are **rare**. Other common sites of metastasis are skin and lymph nodes.
813. e) Neuroblastoma state 4S manifests with dissemination to liver, skin, or bone marrow without bone involvement and a primary tumor with stage 1 or 2.
814. d) Biopsy of tumor can make the pathologic diagnosis. The levels of HVA and VMA are **elevated** in urine (95% of cases). If the primary tumor cannot be located, the diagnosis can be made with a typical clinical manifestation, elevated HVA and VMA in urine, and presence of neuroblast in bone marrow.
815. a) Rosettes of cells surrounding an inner mass of fibrillary material are characteristic of neuroblastoma.
816. b) The risk of recurrence is extremely **rare** in patients with acute poststreptococcal glomerulonephritis. In addition to penicillin therapy, the patient should be treated with sodium restriction, diuretic (e.g., furosemide), and antihypertensive (e.g., calcium channel antagonists, vasodilators, or angiotensin-converting enzyme inhibitors) agent.
817. e) A normal serum complement (C 3) is an indication for renal biopsy to rule out other disease condition. The serum C 3 level is usually reduced in the acute phase of poststreptococcal glomerulonephritis and returns to normal 6-8 weeks after onset. Renal biopsy can rule out SLE (systemic lupus erythematosus), membranoproliferative glomerulonephritis, and acute exacerbation of chronic glomerulonephritis.
818. b) Coombs test is negative in hemolytic-uremic syndrome (HUS). Other laboratory findings in HUS are hemoglobin (5-9 g/dL), presence of burr cell, helmet cells, fragmented RBCs, reticulocytosis, usually normal PT and PTT, proteinuria, low-grade microscopic hematuria. Barium studies reveal colonic spasm and transient early filling defects. The patients may have abnormal PT and PTT those are caused by vitamin K deficiency than by DIC.
819. a) Antithrombotic therapy has no benefit in patients with HUS.
820. e) Hemolytic-uremic syndrome
821. b) Plasmaphresis or administration of FFP (fresh frozen plasma) may exacerbate HUS caused by Streptococcus pneumoniae.
822. e) Acyclovir does not cause hemorrhagic cystitis. Other causes of hemorrhagic cystitis are dyes, insecticides, amyloidosis, radiation, and viruses (e.g., influenza A, polyoma BK virus, adenovirus type 11 and 21). Cyclophosphamide induced

hemorrhagic cystitis can be prevented by administration of cyclophosphamide in the morning followed by aggressive oral hydration throughout the day and the use of MESNA disulfide that inactivates cyclophosphamide metabolites.

823. e) Temperature usually greater than 101°F (38.3°C); other causes of transient proteinuria are stress and cold exposure. The mechanism is unknown. Proteinuria does not exceed 2+ on the dipstick. No investigation is indicated.

824. e) Orthostatic (or postural) proteinuria (60% of cases)

825. e) In children with persistent asymptomatic proteinuria, orthostatic proteinuria should be ruled out first. They have proteins (up to 1 g/24 hours) in the active (upright) position and absence of proteins in resting (supine) position. The cause is unknown.

826. b) Most children (90% of cases) with nephrotic syndrome have idiopathic cause and remaining (10% of cases) are secondary. Answers (b), (c), and (d) are idiopathic. The most common idiopathic type is minimal change disease. Answers (a) and (e) are secondary causes of nephrotic syndrome.

827. b) Spot urine protein to creatinine ratio exceeds 2.0. Other laboratory findings of idiopathic nephrotic syndrome are elevated serum cholesterol and triglyceride levels, microscopic hematuria (20% of cases), increased urinary protein excretion (40 mg/m^2/hour), and sometimes elevated serum creatinine levels when intravascular volume is reduced. Renal biopsy is not usually required to make the diagnosis.

828. c) Bartter syndrome patients have large protruding ears.

829. d) Renal functions are **normal** in patients with Bartter syndrome. Blood pressure is usually **normal** in this syndrome **except** antenatal form with severe salt wasting resulting in hypotension and dehydration. Other features of Bartter syndrome are nephrocalcinosis, elevated urinary sodium and potassium levels, and rarely, hypomagnesemia. Hypomagnesemia is more common in Gitelman syndrome.

830. e) Sodium bicarbonate should **not** be given in patients with Bartter syndrome.

831. a) Autosomal recessive

832. b) E. coli, answers (a), (c), (d), and (e) also cause UTIs; answer (a) especially causes cystitis.

833. e) Bladder hydrodistension and laser ablation of ulcerated areas, but no therapy produces sustained relief. The girl has interstitial cystitis. The cause is **idiopathic**. Cystoscopy reveals mucosal ulcers with bladder hydrodistension.

834. a) Acute hemorrhagic cystitis is frequently caused by E. coli.

835. a) Prompt plating of urine samples are important. If the urine samples are kept at room temperature over 60 minutes, overgrowth of bacteria is common. Urine sample should be kept in refrigerator until it can be cultured.

836. c) Urine collected in a sterile collection bag is reliable if the culture is negative. However, it is not reliable if the culture is positive, especially in girls and uncircumcised boys. If the culture is positive, patient should be symptomatic, urinalysis should be positive, and there should be one organism with a colony count greater than 100,000 to diagnose a presumed UTI.

837. e) Fluoroquinolones cause cartilage damage in children.

838. e) Trimethoprim-sulfamethoxazole is the preferred therapy for E. coli cystitis.

839. e) Nitrofurantoin is effective against both Klebsiella and Enterobacter organisms.

840. a) Cefixime is the preferred oral therapy for pyelonephritis **except Pseudomonas**. Cefixime is oral third-generation cephalosporin.

841. a) Ciprofloxacin (fluoroquinolone) is effective against Pseudomonas and is used in patients older than 17 years.

842. d) Ceftriaxone (50-75 mg/kg/day, not to exceed 2 g) **or** ampicillin (100 mg/kg/day) with an aminoglycoside (e.g., gentamicin 3-5 mg/kg/day). Aminoglycosides are especially effective against Pseudomonas.

843. c) Nitrofuratoin should **not** be used in children with a febrile UTI because it does not achieve significant renal tissue levels.

844. e) Trimethoprim-sulfamethoxazole or nitrofurantoin is preferred prophylactic agent against recurrent UTIs (one third of normal dose is used). Answers (a) and (b) can be used but they can cause breakthrough UTIs because of bacterial resistance.

845. d) 1 week after the termination of antibiotic therapy to ensure that the urine is sterile.

846. b) A follow-up urine culture should be performed periodically for 1-2 years, even when the child is asymptomatic because of the tendency of UTI to recur.

847. e) Application of corticosteroid cream to the foreskin three times daily for 1 month releases the phimotic ring in 66% of cases. Circumcision is indicated when medical management fails. A forceful retraction of the prepuce should **not** be performed because it causes a cicatrical scar that prevents subsequent foreskin retraction.

848. d) Lubrication of the glans and foreskin, then compress the glans and simultaneously placing distal traction on the foreskin to try to push the phimotic ring distal to the coronal sulcus

849. a) Hypothyroidism in associated with elevated levels of thyroid-releasing hormone that also stimulates release of prolactin.

850. b) Papilloma causes watery discharge.

851. a) Mastitis causes purulent discharge.

852. d) Galactorrhea causes milky discharge.

853. c) Duct ectasia causes bloody discharge. It also produces multicolored/sticky or serous/serosanguineous discharge.

854. d) Galactorrhea

855. c) Congenital hypopituitarism also manifests with severe hypoglycemia, prolonged neonatal jaundice, and in boys, with microphallus.

856. c) Congenital hypopituitarism; this is a characteristic manifestation. **Silver-Russel syndrome** is characterized by small triangular facies, frontal bossing, sparse subcutaneous tissue, shortened and incurved 5th fingers, low birthweight for gestational age, short stature, and may appear with hemihypertrophy.

857. e) Chromosome 17q

858. e) Peak levels of GH are less than 10 μg/L after two provocative tests (i.e., administration of L-dopa, insulin, glucagon, arginine, or clonidine). Answers (a), (b), (c) and (d) are also noted in patients with GH deficiency.

859. d) **Pseudohyponatremia** manifests with normal intravascular volume and normal urine sodium same as **factitious hyponatremia**.

860. c) **SIADH** manifests with high intravenous volume and high urine sodium.

861. b) **Cerebral salt wasting** manifests with low intravascular volume and very high urine sodium.

862. a) **Systemic dehydration** manifests with low intravascular volume and low urine sodium same as **primary salt loss and decreased effective plasma volume**.

Patients with **primary polydipsia** manifest with normal or high intravascular volume and normal urine sodium.

Patients with **decreased free water clearance** manifest with normal or high intravascular volume and urine sodium.

863. c) The most common cause of Addison disease is autoimmune destruction of the glands. Answers (a) and (d) are less likely cause of Addison disease. Answers (b) and (d) can cause Addison disease.

864. b) Meningococcemia causes adrenal crisis that is called Waterhouse-Friderichsen syndrome.

865. e) Adrenal insufficiency in infants causes hypoglycemia, hyponatremia, and hyperkalemia. Hyperpigmentation takes weeks or months to develop. Ketosis is an inconsistent finding because infants produce ketones less effectively than older children. The infant has adrenal hemorrhage due to a forcep delivery resulting in adrenal insufficiency.

866. c) Hyperkalemia usually occurs later in the course of the disease in older children than in infants. Older children usually manifest with muscle weakness, vomiting, weight loss, and hypotension. Hyperpigmentation may or may not be present.

867. e) Primary adrenal insufficiency is diagnosed by decreased serum levels of cortisol before and after (30 to 60 minutes) the administration of ACTH. Answers (d) represents **secondary** adrenal insufficiency. Answers (a), (b), and (c) are also noted in patients with adrenal insufficiency. They also have increased renin and eosinophils in the blood. They also have increased excretion of sodium and chloride and decreased excretion of potassium in the urine.

868. c) Fludrocortisone (Florinef) is used for choronic replacement therapy if aldosterone deficiency is present. Measurements of plasma renin activity are useful in monitoring the adequacy of mineralocorticoid therapy (i.e., renin activity level should not be elevated after an adequate therapy).

869. d) Obesity, short stature, hyperglycemia, and osteoporosis are features of **chronic overdose of glucocorticoids**.

870. a) Primary hypogonadism (hypergonadotropic hypogonadism in the male) have both small testes and penis at birth. Secondary hypogonadism (hypogonadotropic hypogonadism in the male) have **normal testes at birth** and remain in the prepubertal state because lack of stimulation by gonadotropins.

871. d) The boy has primary hypogonadism. FSH and LH levels are highly elevated in boys after 11 years of age, reaching the castrate range. A random serum testosterone level is not useful in prepubertal boys because it is low. In normal children, serum testosterone levels are elevated significantly after administration of hCG but in children with primary hypogonadism, there is no rise or attenuated rise after administration of hCG.

872. d) Testicular cancer is not present. Answer (e) can produce precocious puberty in young boys due to increased production of hCG (human chorionic gonadotropin).

873. a) Spermatogenic arrest and Sertoli cell predominance. Most patients with Klinefelter syndrome have azoospermia and infertility.

874. d) Testosterone therapy increases prostate volume and prostate-specific antigen levels. Fertility can occur by an intracytoplasmic sperm injection after testicular sperm extraction.

875. e) Usual insulin doses should be reduced as much as 50% or more in prolonged exercise (e.g., long distance running).

876. a) Usual insulin doses should be reduced by 10-15% in regular exercise.

877. a) Parvovirus B19 typically produces **red cell aplasia**, but in patients with sickle cell disease or immunodeficiencies it can produce **transient** pancytopenia.

878. e) von Willebrand disease

879. b) The D-dimer assay is more specific and equally sensitive than the FDP test. Answers (a), (c), (d), (e), and decreased levels of factors (II, V, VIII, and fibrinogen) are also noted in patients with DIC.

880. b) Cryoprecipitate is used in patients with **hypofibrinogenemia**.

881. a) Heparin therapy is **occasionally** effective in patients with DIC associated with purpura fulminans and acute promyelocytic leukemia. Heparin therapy is not effective in answers (b), (c), (d), (e), and patients with incompatible blood transfusion reactions, unless there is vascular thrombosis.

882. e) Gastroesophageal reflux is common in patients with BPD (bronchopulmonary dysplasia). Medical therapies include promotility and antacid medications. If unresponsive, fundoplication with gastrostomy is performed.

883. a) Audiologist is not included in the team. However, speech therapist is a part of the team.

884. b) Telogen effluvium is loss of resting or telogen hairs due to premature conversion of growing or anagen hairs. Hair loss occurs 6 weeks to 3 months after the precipitating cause (e.g., surgery, fever, acute blood loss, blood donation, psychiatric stress; discontinuation of high-dose corticosteroids or oral contraceptives). Telogen bulbs are noted on shed hairs under microscopic examination. Patients need reassurance. Hairs will return to normal within 6 months.

885. d) Large nails are noted in Rubinstein-Taybi syndrome, hemihypertrophy, and paronychia congenita.

886. a) Small or absent nails are noted in nail-patella syndrome, ectodermal dysplasia, fetal hydantoin syndrome, acrodermatitis enteropathica, epidermolysis bullosa, and cartilage hair hypoplasia.

887. b) Yellow nail syndrome is associated with bronchiectasis, recurrent bronchitis, chylothorax, and focal edema of the limbs and face.

888. e) Koilonychia is associated with Plummer-Vinson syndrome, hemochromatosis, and hypochromic anemia. Koilonychia is flattening and concavity of the nail plate.

889. c) Leukonychia (or white opacity of the nail plate) is noted in aresenic poisoning (Mees lines), anemia, Hodgkin disease, tuberculosis, lichen planus, and Darier disease.

890. a) Fusobacterium nucleatum and Borrelia vincenti can produce Vincent gingivitis (acute necrotizing ulcerative gingivitis, trench mouth).

891. e) Pseudomonas aeruginosa infection can cause noma neonatorum that manifests with gangrenous lesions of the lips, mouth, nose, and anal regions.

892. b) AIDS can produce oral hairy leukoplakia but is rare in pediatric patients.

893. d) Black hairy tongue is produced by chronic smoking, tetracycline, bacterial overgrowth, and poor oral hygiene.

894. c) Scrotal (fissured) tongue is associated with vitamin A deficiency, trauma, infection, or malnutrition.

895. a) The child has aphthous stomatitis (canker sores) that is **not** produced by herpes simplex virus. Aphthous stomatitis is also produced by malabsorption (e.g., Crohn disease or celiac disease), decreased serum iron or ferrtin levels, food hypersensitivity, allergic or toxic drug reaction, and luteal phase (decreased progesterone) of menstruation.

896. e) Group A beta-hemolytic Streptococci is the most common organism. S. aureus also causes blistering distal dactylitis.

897. a) Staphylococcus aureus. Sycosis barbae is a recurrent, severe erythematous papules and pustules involving the chin, upper lip, and angle of the jaw.

898. a) Pseudomonas aeruginosa (predominantly serotype O-11)

899. e) Pseudomonas aeruginosa. Ecthyma gangrenosa is a necrotic ulcer cover with a gray-black eschar.

900. e) Corynebacterium minutissimum. Erythrasma is a benign, chronic, mildly pruritic superficial skin infections of the toe webs, groin, and axilla. Methylene blue or Gram stain reveals the filamentous coccobacillary forms. This condition is treated with topical erythromycin, clindamycin, miconazole or Whitfield ointment. Recurrence can be prevented by frequent use of antibacterial soap or 10-20% aluminum chloride in anhydrous ethyl alcohol.

901. c) Staphylococcus epidermidis is a coagulase-negative staphylococcus.

902. b) 60-70 mL/kg/day

903. c) 100-120 mL/kg/day

904. b) 15 g/kg/day

905. d) 34 wk gestational age. Premature infants less than 34 wk gestational age should receive gavage feeding or parenteral nutrition.

906. a) Vitamin A. Very low birthweight (VLBW) infants weigh 1,500 g or less at birth. Low birthweight (LBW) infants weight 2,500 g or less at birth.

907. a) Breast milk reduces risk of childhood/adolescent obesity.

908. e) All infants with a birthweight between 1500 and 2000 g with an unstable clinical course requiring oxygen and infants with a birthweight under 1500 g with or without unstable clinical course should have an eye examination to screen for ROP.

909. c) Mixed apnea; obstructive apnea usually occurs before central apnea.

910. a) Cardiac perforation is absent in patients with umbilical arterial catheterization. However, umbilical arterial catheterization can produce infection, accidental hemorrhage, decreased circulation in lower extremities resulting in gangrene, ischemic or chemical necrosis of abdominal organs.

911. e) Umbilical venous catheterization does **not** produce decreased circulation in lower extremities and gangrene. However, umbilical venous catheterization can produce infection and perforation. Answers (b) and (c) can occur when catheter is in right atrium. Answer (d) develops due to portal vein thrombosis, especially in the presence of omphalitis.

912. d) Topical nitroglycerin applied over the femoral artery or local infusion of tolazoline (Priscoline) within the artery, or **rarely**, warming the opposite leg is useful.

913. e) Persistent pulmonary hypertension is the diagnosis when the chest x-ray appears normal, absence of cardiac malformations, hypoxia is liable and out of proportion to the findings on chest x-ray.

914. e) Presence of air leak in both lungs may be the consequence of meconium aspiration.

915. a) ECMO can cause **systemic hypertension**. ECMO also causes air embolism, strokes, atelactasis, cholestatic jaundice, hemolysis, thrombocytopenia, neutropenia, and infectious complications of blood transfusions.

916. d) Reassurance because oral thrush in an otherwise healthy infant is a self-limited infection. However, treatment is indicated if there is associated candidal diaper rash.

917. b) 'Blueberry-muffin' rash is produced by CMV, rubella, and parvovirus. This rash represents dermal erythropoiesis.

918. a) Satellite pustules those affect the contiguous skin are a hallmark of localized candida infection.

919. e) The presence of salmon-pink pustules suggests Listeria monocytogenes infection.

920. c) The presence of vesicular rash suggests herpes simplex virus infection.

921. d) Ecthyma gangrenosum indicates Pseudomonas infection.

922. d) Severe motor disorders are not associated with violent behavior. Other conditions associated with violent behavior are moderately severe language disorders, mood disturbances, and personality disorders. They have increased vulnerability for substance abuse, sexual promiscuity, juvenile delinquency, incarceration, antisocial behavior, and adult criminal behavior.

923. d) 'F" stands for **F**orgot things while using alcohol or drugs. Other 'F' stands for **F**amily or **F**riends tell the adolescent to cut down on alcohol or drugs.

924. e) Group B Streptococci does not cause genital ulcer syndromes. The most common organism is herpes simplex virus type 2 in adolescents with genital ulcer syndromes.

925. a) GYN consultation is indicated immediately if pregnant or if needs ultrasonography. The initial evaluation also includes ESR or C-reactive protein, RPR, urine culture, urine dipstick, and urine beta-hCG (human chorionic gonadotropin). A surgical consultation is indicated in patients with suspected appendicitis or other surgical problems. Ultrasonography is indicated if there is a mass or difficult to perform an examination. Serum beta-hCG is done if urine beta-hCG is negative and suspected ectopic pregnancy.

926. b) Phenobarbital does not produce CVID.

927. d) Pyremethamine does not produce selective IgA deficiency. Answers (a), (b), (c), and (e) produce selective IgA deficiency and CVID.

928. c) Cartilage hair hypoplasia. Patients can develop fatal varicella infections, progressive vaccinia, and vaccine-associated poliomyelitis.

929. d) Chromosome 11

930. e) Thrombocytopenia is **not** initially due to antiplatelet antibodies.

931. d) Aspiration pneumonia occurs due to impairment of swallowing fluids. Answers (a), (b), (c), and (e) are also complications in patients with juvenile dermatomyositis. Progressive bowel infarction can lead to perforation and death.

932. e) Henoch-Schonlein purpura

933. a) Hypersensitivity angiitis is cutaneous vasculitis following exposure to drugs (e.g., sulfonamides). Patients usually appear with fever, myalgias, and arthralgia.

934. a) Erythromelalgia. Treatment includes avoidance of heat exposure and application of cold substance during attacks. Some children may respond to propranolol, carbamazepine, or sodium nitroprusside.
935. b) Streptococcus pneumoniae
936. e) Streptococcus pneumoniae
937. e) Scarlet fever is caused by group A Streptococcus.
938. c) Group A Streptococcus. Answers (a), (b), (d), and (e) also cause pharyngitis.
939. d) Splenic involvement is **not** a part of Streptococcal toxic shock syndrome. Hypotension plus two or more of the choices those are mentioned in answers (a), (b), (c), and (e) suggest Streptococcal toxic shock syndrome.
940. a) Clinical criteria and detection of group A Streptococcus from a normally sterile site.
941. a) Disseminated gonococcal infection **rarely** causes carditis, osteomyelitis, and meningitis. DGI also causes pustular acral skin lesions.
942. b) Cervix (80-90% of cases)
943. b) Urethra (50-60% of cases)
944. d) Factor V (phosphopyridine nucleotide) and factor X (hematin)
945. a) Lymphadenopathy (96% of cases). Answers (b), (c), (d), (e), vomiting, and hepatosplenomegaly are also clinical manifestations of tularemia in children.
946. a) Ulceroglandular (45% of cases). Answers (b), (c), (d), (e), and typhoidal are also clinical syndromes of tularemia.
947. b) Gentamicin or streptomycin
948. e) Serum agglutination test. A single titer of 1:160 or more in a patient with a typical history and physical findings can make the diagnosis. A fourfold increase in titer from two samples collected 2-3 wk apart is also diagnostic.
949. b) All children with active herpes lesions can go to daycare or school activities when the lesions can be covered.
950. c) Limbs
951. b) Eyes and brain
952. d) Malaria illness should be considered regardless of the use of chemoprophylaxis. Plasmodium falciparum infection should be suspected when children appear with clinical manifestations less than 1 month after return from an endemic area.
953. b) 12 months
954. b) 15 months
955. c) 13 months
956. b) 3 months
957. c) 8 months
958. c) 19 months
959. c) 22 months
960. b) 4 months
961. b) 17 months
962. a) 4 months
963. a) 2 cm/month
964. b) 1 cm/month
965. c) 0.5 cm/month
966. d) 0.25 cm/month
967. e) 1 cm/year
968. a) 2 lb
969. b) 1.25 lb
970. c) 1 lb
971. d) 13 oz
972. e) 8 oz
973. c) 9 months of age
974. a) 16-20 hours per day
975. b) 14-15 hours per day (13-14 hours at 6 months of age)

976. c) 12-14 hours per day
977. d) 11-12 hours per day
978. e) 10 hours per day. After 12 years of age, sleeping time 9 hours per day is ideal but actual sleeping time is 7 hours.
979. b) Clonidine can cause sudden death when used with stimulants. Clonidine also causes sedating bradycardia, hypotension, and withdrawal symptoms.
980. a) Desipramine can cause sudden death in doses more than 3.5 mg/kg. Desipramine can cause hypertension, orthostatic hypotension, arrythmias, and prolong PR or QRS intervals.
981. e) Carbamazepine can cause fever, sore throat, and leukopenia.
982. c) Valproic acid can cause hepatic failure usually in the first few months of treatment. Valproic acid can case birth defects in fetus, obesity, depression, and rashes.
983. d) Lamotrigine can cause Stevens-Johnson syndrome. A concurrent use with valproic acid doubles the lamotrigine blood levels.
984. a) Mild mental retardation (IQ 55-70), moderate (IQ 40-55), severe (IQ 20-40), and profound (IQ less than 20)
985. e) Prescrible regular (not PRN) medications for constant pain. Maintaining comfort is a priority.
986. d) Stimulants are used for opioid-induced somnolence.
987. b) Steroids are used for resistant pain.
988. a) Neuroleptics are used for agitation and nausea.
989. c) Sedatives and hypnotics are used for muscle spasms or anxiety.
990. e) Antidepressants and anticonvulsants are used for neuropathic pain.
991. a) Strong medications should not saved for last use.
992. a) Plasma levels of leptin correlate directly with fat mass. Human leptin is produced predominantly in adipose tissue. Leptin acts on hypothalamus. Low plasma levels of leptin and insulin during fasting and weight loss increase secretion of neuropeptide Y (NPY) that increases food intake and decreases energy expenditure.
993. e) Reassurance. Normal serum phosphorus levels are 4.8-8.2 mg/dL between 0-5 days of life.
994. c) Restrict the fluid intake because the child has increased ADH secretion after surgery.
995. a) Isotonic solution should be given immediately through intravenous route because of intravascular volume depletion and third spacing resulting in oliguria.
996. b) Amrinone is a phosphodiesterase inhibitor and slows cyclic AMP breakdown.
997. c) Dobutamine strengthens cardiac contraction, reduces afterload, and weak vasoconstrictor effects.
998. a) Phenylephrine can produce sudden hypertension.
999. d) Norepinephrine produces short-run rise in blood pressure (high systemic vascular resistance).
1000. e) Epinephrine may reduce renal perfusion.
1001. e) Werdnig-Hoffman disease affects spinal cord.
1002. c) Cystic fibrosis causes peripheral airway obstruction.
1003. a) Prune-belly syndrome may have chest wall deformities.
1004. b) Guillain-Barre syndrome involves neuromuscular system.
1005. d) Croup produces central airway obstruction.