1. The most common site of pulmonary atelectasis in children is:
   a) Right upper lobe
   b) Right middle lobe
   c) Right lower lobe
   d) Left upper lobe
   e) Left lower lobe

2. A routine newborn physical examination of a newborn reveals a grade 2/6, short, harsh systolic murmur at the apex. The newborn
   is completely asymptomatic. The most likely diagnosis is:
   a) Atrial septal defect
   b) Ventricular septal defect
   c) Mitral stenosis
   d) Mitral regurgitation
   e) Patent ductus arteriosus

3. All of the following conditions can present with spherocytosis in a peripheral blood smear except:
   a) ABO incompatibility
   b) Thermal injury
   c) Wilson disease
   d) Autoimmune hemolytic anemia
   e) Pneumococcal sepsis

4. A 7-day-old newborn girl appears for a routine physical check-up. Her mother noticed a grapelike mass protruding through the
   child’s vagina. The girl is asymptomatic. The most likely diagnosis is:
   a) Vaginal prolapse
   b) Rectovaginal fistula
   c) Urethral prolapse
   d) Uterine prolapse
   e) Sarcoma botryoides

5. A VCUG study in a male newborn reveals massive bilateral ureteral dilatations due to a severe vesicoureteral reflux. The urinary
   bladder is very distended. There is no urethral obstruction. There is absence of neuropathic dysfunction. The newborn is receiving
   prophylactic antibiotics. The preferred therapy is:
   a) Resection of posterior urethral valve
   b) Dilatation of urethra
   c) Removal of prostate
   d) Permanent vesicostomy
   e) Reimplantation of ureters into an urinary bladder

6. The most important hormone responsible for the onset and progression of puberty is:
   a) Estrogen
   b) ACTH
   c) TSH
   d) Progesterone
   e) GnRH

7. A 2½ years boy appears with intermittent loose stools for the past 1 month. Stools typically occur during the day and not overnight.
   The boy is otherwise healthy. The growth and development are normal. The most likely diagnosis is:
   a) Chronic enteritis
   b) Rotavirus enteritis
   c) Salmonella enteritis
   d) Food allergy
   e) Toddler’s diarrhea

8. The best diagnostic antibody titer in patients with a streptococcal skin infection is:
   a) Antistreptolysin O titer is elevated.
   b) Serum C3 level is decreased.
   c) Serum C3 level is increased
33. The most common presentation in patients with congenital hypothyroidism is:
   a) Umbilical hernia
   b) Constipation
   c) Jaundice
   d) Poor feeding
   e) Asymptomatic

34. The most newborn screening programs in USA measure the following parameter to rule out congenital hypothyroidism is:
   a) $T_3$
   b) $T_4$
   c) TSH
   d) TRH
   e) Free-$T_4$

35. The most common cause in newborns with 46, XX intersex and ambiguous genitalia is:
   a) Maternal use of testosterone during pregnancy.
   b) Aromatase deficiency
   c) Glucocorticoid receptor gene mutation
   d) Maternal ovarian androblastomas
   e) Congenital adrenal hyperplasias

36. The most common site of the vanishing testes in patients with XY gonadal agenesis syndrome is:
   a) Abdomen
   b) Scrotum
   c) Superficial inguinal ring
   d) Deep inguinal ring
   e) Inguinal canal

37. The cause of undescended testes in patients with a generalized myopathy is:
   a) Decreased dehydrotestosterone level
   b) Decreased testosterone level
   c) Weakness of the gubernaculum
   d) Absence of the Y chromosome
   e) Weakness of the Y chromosome

38. The most frequent cause of toxic neuropathies in children is:
   a) Lead poisoning
   b) Organophosphates
   c) Vincristine
   d) Cisplatin
   e) Idiopathic

39. The most important risk factor in patients with a retinopathy of prematurity is:
   a) Hyperoxia
   b) Apnea
   c) Infection
   d) Hypoxia
   e) Prematurity

40. The first retinal sign in patients with a hypertensive retinopathy is:
   a) Edema
   b) Cotton-wool spots
   c) Flame-shaped hemorrhages
   d) Papilledema
   e) Generalized constrictions of retinal arterioles.

41. The most common organism in neonates with an acute otitis media is:
   a) S. aureus
   b) Streptococcus pneumoniae
161. All of the following statements are true about extrathoracic obstructive disease except:
   a) Duration of inspiration is reduced.
   b) Respiratory rate is decreased.
   c) Duration of expiration is unchanged.
   d) Physical examination reveals a inspiratory stridor.
   e) Chest x-ray findings are normal.

162. The following statement is not true about the intrathoracic obstructive lung disease except:
   a) Duration of inspiration is prolonged.
   b) Respiratory rate is either normal or increased.
   c) Duration of expiration is prolonged.
   d) A physical examination reveals expiratory wheezes.
   e) A chest x-ray reveals an increased lung volume.

163. The most common presentation in patients with a tracheal foreign body is:
   a) Wheezing
   b) Stridor
   c) Positive chest x-ray
   d) Choking and aspiration
   e) Positive soft tissue x-ray of the neck

164. A 2-month-old infant appears with a brassy cough. The most likely diagnosis is:
   a) Subglottic hemangioma
   b) Bronchiolitis
   c) URI
   d) Chlamydia infection
   e) Vascular ring

165. A child appears with a staccato cough. The most likely diagnosis is:
   a) Habit cough
   b) Subglottic hemangioma
   c) Laryngomalacia
   d) Croup
   e) Chlamydia pneumonitis

166. A 10-year-boy appears with a history of cough only during the daytime for the last 2 months. However, he sleeps well at night without coughing. The boy is coughing during the physical examination. A physical examination reveals a normal throat and clear breath sounds in both lungs. The most likely diagnosis is:
   a) Chlamydia infection
   b) Bronchial asthma
   c) Sinusitis
   d) Habit cough
   e) Carrier of group A Streptococcus

167. A congenital subglo ttic hemangioma is associated with the following anomaly:
   a) Renal
   b) CNS
   c) Liver
   d) Spleen
   e) Cutaneous

168. The most common complication of patients with a supracrystal VSD is:
   a) Right ventricular failure
   b) Left ventricular failure
   c) Pulmonic stenosis
   d) Aortic insufficiency
   e) Arrythmias
d) Psychotherapy

e) Reassurance

209. The side effect of imipramine is:
   a) Arrhythmias
   b) Hypertension
   c) Excessive salivation
   d) UTI
   e) Alertness

210. The hours per night sleep necessary for adolescents are:
   a) 6 hours
   b) 7 hours
   c) 8 hours
   d) 9 hours
   e) 10 hours

211. Munchausen syndrome by proxy (MSBP) may include all of the following except:
   a) A mother is very concerned about a child’s condition.
   b) A mother fabricates a medical history.
   c) A mother may have a background in health care.
   d) A mother may alter laboratory samples.
   e) A child may be exposed to physical abuse, toxin, medication, or infectious agents.

212. The most common clinical manifestation in neonates with early onset group B Streptococcus (GBS) infections is:
   a) Pneumonia
   b) Meningitis
   c) Sepsis
   d) UTI
   e) Asymptomatic bacteremia

213. The most common clinical manifestation in infants with late onset group B Streptococcus (GBS) infections is:
   a) Meningitis
   b) Bacteremia
   c) Pneumonia
   d) Osteomyelitis
   e) UTI

214. The most common type of group B Streptococcus (GBS) responsible for early or late-onset meningitis is:
   a) Ia
   b) Ib
   c) II
   d) III
   e) V

215. All of the following serotypes are most commonly associated with neonatal GBS diseases except:
   a) Ia
   b) Ib
   c) II
   d) III
   e) IV

216. The most common sexually transmitted disease noted in sexually abused children is:
   a) Chlamydia
   b) Syphilis
   c) Gonorrhea
   d) HIV
   e) Herpes simplex virus
d) Pseudomonas infections  
e) Coccidioidomycosis infections

225. Medical personnel are more prone to acquire the following herpes infection:
   a) Herpetic whitlow  
   b) Herpes gladiatorum  
   c) Genital herpes  
   d) Eczema herpeticum  
   e) Acute herpetic gingivostomatitis

226. Newborns acquire a herpes infection from their mothers. The most common time to acquire an herpes infection is:
   a) First trimester  
   b) Second trimester  
   c) Third trimester  
   d) During delivery  
   e) During breast-feeding

227. Negri body is a characteristic of the following disease:
   a) Herpes zoster infections  
   b) Herpes simplex virus infections  
   c) Rabies  
   d) CMV infections  
   e) Toxoplasmosis

228. A child is receiving an active immunization for a postexposure prophylaxis for rabies. The following therapy may suppress the response of an active immunization is:
   a) Antibiotic therapy  
   b) Antiviral therapy  
   c) Antifungal therapy  
   d) Antimalarial therapy  
   e) Antiepileptic therapy

229. The most common fungal infection in HIV-infected children is:
   a) Fungal genital infections  
   b) Oral candidiasis  
   c) Fungal scalp infections  
   d) Fungal bacteremia  
   e) Fungal UTI

230. The most common cause of bacterial pneumonia in HIV-infected children is:
   a) H. influenzae  
   b) Moraxella catarrhalis  
   c) P. aeruginosa  
   d) E. coli  
   e) Streptococcus pneumoniae

231. The most common MRI finding in HIV-infected children with neurologic symptoms is:
   a) Cerebellar atrophy  
   b) Cerebral atrophy  
   c) Leukomalacia  
   d) Periventricular calcifications  
   e) Diffuse calcifications

232. AIDS enteropathy is most likely due to the following reason:
   a) Salmonella infections  
   b) Cryptosporidium infections  
   c) CMV infections  
   d) Candida infections  
   e) Direct HIV-infections
242. A child can hold a cup by:
   a) 6-7 months of age
   b) 7-8 months of age
   c) 8-9 months of age
   d) 9-10 months of age
   e) 11-12 months of age

243. Children can feed themselves by the end of:
   a) 18 months of age
   b) 24 months of age
   c) 30 months of age
   d) 36 months of age
   e) 48 months of age

244. Moderately active 4-6 years old children need:
   a) 1,200 Kcal/day
   b) 1,400 Kcal/day
   c) 1,600 Kcal/day
   d) 2,000 Kcal/day
   e) 2,400 Kcal/day

245. Moderately active 6-10 years old children need:
   a) 2,000 Kcal/day
   b) 2,200 Kcal/day
   c) 2,400 Kcal/day
   d) 2,600 Kcal/day
   e) 2,800 Kcal/day

246. Active teenage boys need:
   a) 2,400 Kcal/day
   b) 2,600 Kcal/day
   c) 2,800 Kcal/day
   d) 3,000 Kcal/day
   e) 3,200 Kcal/day

247. A child appears with nausea, diarrhea, polyuria, weight loss, and nocturia. The most likely diagnosis is:
   a) Vit A toxicity
   b) Vitamin A deficiency
   c) Vit D toxicity
   d) Vit K deficiency
   e) Niacin toxicity

248. A child appears with alopecia, drying skin, slow growth, hepatosplenomegaly, increased intracranial pressure, painful and swollen long bones. The most likely diagnosis is:
   a) Vitamin B₁₂ toxicity
   b) Vitamin B₆ toxicity
   c) Niacin deficiency
   d) Vitamin E toxicity
   e) Vitamin A toxicity

249. Breast-fed infants should receive iron supplementation by:
   a) 1-2 months of age
   b) 2-4 months of age
   c) 4-6 months of age
   d) 6-9 months of age
   e) 9-12 months of age
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b) Ceftriaxone
c) Ibuprofen
d) High calcium diet
e) Bone marrow transplantation

330. The most common organism causing recurrent skin and pulmonary infections in patients with a hyper IgE syndrome is:
   a) Pseudomonas aeruginosa
   b) S. aureus
   c) S. epidermidis
   d) E. coli
   e) S. pneumoniae

331. The most common organism causing skin, mucous membranes, and respiratory tract infections in patients with a Chediak-Higashi syndrome is:
   a) P. aeruginosa
   b) S. epidermidis
   c) E. coli
   d) S. aureus
   e) Candida albicans

332. The most common organism causing recurrent lymphadenitis, hepatic abscesses, and osteomyelitis at multiple sites in patients with a chronic granulomatous disease is:
   a) S. aureus
   b) Serratia marcescens
   c) Burkholderia cepacia
   d) Candida albicans
   e) Salmonella

333. The following condition appears with a pancreatic insufficiency with fatty replacement, anemia, thrombocytopenia, and metaphyseal dysostosis:
   a) Kostman syndrome
   b) Chediak-Higashi syndrome
   c) Hyper IgE syndrome
   d) Cartilage-hair hypoplasia
   e) Shwachman-Diamond syndrome

334. The best method for detection of allergen-specific IgE is:
   a) Serum IgE levels
   b) Blood eosinophilia
   c) Eosinophils in nasal smear
   d) Total serum immunoglobulin levels
   e) Epicutaneous skin tests

335. A good asthma management means that almost all children with asthma can experience all of the following except:
   a) Sleep well without disturbance due to a chronic cough
   b) Attend school regularly
   c) Participate fully in sports activities
   d) Avoid severe exacerbations of asthma
   e) Little or no adverse effects from asthma medications

336. The most common cause of childhood emergency department visits, hospitalizations, and missed school days is:
   a) Gastroenteritis
   b) Pneumonias
   c) Asthma
   d) Child abuse
   e) Motor vehicle accidents

337. The following statement is not true about patients with an asthma:
   a) All young children who experienced wheezing, a majority of them will develop asthma in later childhood.
354. A child is recovered from a Kawasaki disease after IVIG and aspirin therapies. The following vaccination should be delayed:
   a) IPV
   b) MMR
   c) DTaP
   d) Pneumococcal
   e) Hepatitis B vaccine

355. Churg-Strauss syndrome includes all of the following except:
   a) Destructive upper airway disease
   b) Chronic sinusitis
   c) History of asthma
   d) Blood eosinophilia
   e) Eosinophilic cutaneous vasculitis

356. The most common site of intussusception in patients with a Henoch-Schonlein purpura is:
   a) Ileocolic
   b) Colocolic
   c) Jejunoileal
   d) Duodenjejunal
   e) Ileoleal

357. The definitive diagnostic study in patients with a Henoch-Schonlein purpura is:
   a) Elevated serum IgA levels
   b) Elevated serum IgM levels
   c) Positive antcardiolipin antibodies
   d) Positive antiphospholipid antibodies
   e) Biopsy of skin lesions

358. A pulse rate can be low in the presence of fever in all of the following conditions except:
   a) Drug fever
   b) Typhoid
   c) Brucellosis
   d) Leptospirosis
   e) S. aureus

359. A preterm infant is receiving total Parenteral nutrition and intralipid through a central line. Suddenly, the infant develops apnea, bradycardia, and mild abdominal distension. CBC and blood culture are performed. CBC reveals a normal WBC count (12,000) and low platelet count (75,000). A peripheral blood culture is positive for Candida albicans. The next step in management is:
   a) Amphotericin
   b) Fluocytosine
   c) Remove the central line only
   d) Repeat the blood culture
   e) Amphotericin and removal of the central line

360. A preterm infant’s peripheral blood culture is positive for S. epidermidis. She is NPO and receiving TPN and intralipid through a central line. She is in a stable condition. The next step in management is:
   a) Nafcillin and remove the central line
   b) Vancomycin and remove the central line
   c) Remove the central line and give ampicillin
   d) Repeat the blood culture and keep the central line
   e) Vancomycin, repeat the blood culture, and keep the central line

361. All of the following conditions can occur due to a colonization of VA (ventriculoatrial) shunt except:
   a) Shunt nephritis
   b) Pulmonary hypertension
   c) Septic pulmonary emboli
492. A child appears with a history of tibial bone pain in the night and the pain is relieved by aspirin. The most likely diagnosis is:
   a) Osteosarcoma
   b) Ewing sarcoma
   c) Osteoid osteoma
   d) Osteochondroma
   e) Enchondroma

493. A child appears with a pain in the lone bone that is not relieved by aspirin. The pain awakens the child at night. The most likely diagnosis is:
   a) Fibroma
   b) Aneurysmal bone cyst
   c) Osteochondroma
   d) Unicameral bone cyst
   e) Osteosarcoma

494. A routine screening urinalysis should be obtained initially at:
   a) 1 year of age
   b) 2 years of age
   c) 3 years of age
   d) 4 years of age
   e) 5 years of age

495. A 14-year-old girl noticed blood in the urine. Her menstrual periods are irregular. She is constipated. The routine urinalysis reveals blood. She denies suprapubic tenderness and flank pain. The most likely diagnosis is:
   a) Asymptomatic UTI
   b) Hemorrhoidal bleeding
   c) Glomerular disease
   d) Ureteral bleeding
   e) Urine contaminated with menstrual bleeding

496. A 10-year-old healthy boy appears for a routine physical examination. The urinalysis reveals more than 100,000 RBCs per 12 hours period. The next step in management is:
   a) Urine culture
   b) Renal ultrasonography
   c) Intravenous pyelography
   d) Voiding cystourethrogram
   e) Reassurance

497. A fair skinned and blond child appears with polyuria, polydipsia, growth failure, rickets, and photophobia. This condition is usually associated with all of the following except:
   a) Hyperthyroidism
   b) Retinopathy
   c) Impaired visual acuity
   d) Delayed sexual maturation
   e) Hepatosplenomegaly

498. The preferred diagnostic study in patients with a cystinosis is:
   a) RBC cystine content
   b) WBC cystine content
   c) Plasma cystine level
   d) Urine cystine content
   e) Presence of cystine crystals in the cornea

499. The following statement is not true about immunizations in patients with a chronic renal failure (CRF):
   a) CRF patients respond to immunizations optimally.
   b) All children with CRF should receive a yearly influenza vaccine.
   c) Live virus vaccines (MMR, Varicella) should not be given during immunosuppressive therapies.
   d) Live virus vaccines should be given before a renal transplantation.
   e) All children with CRF should receive all standard immunizations like healthy children
c) Chronic renal disease
d) Allergic reaction
e) Chronic liver disease

532. A 13-year-old rapidly growing boy has an elevated alkaline phosphatase level. The rest of the LFTs are normal. The most likely diagnosis is:
   a) Obstructive liver disease
   b) Acute hepatitis A
   c) Acute hepatitis B
   d) Hepatoblastoma
   e) Normal

533. An ultrasonographic examination of an infant with Biliary atresia reveals all of the following except:
   a) Small gallbladder
   b) ‘Triangular cord sign’
   c) Absent gallbladder
   d) Increased echogenicity of the liver
   e) Nonvisualization of the common bile duct

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

535. An ophthalmologic examination of a child with a Wilson disease reveals:
   a) Brush field spots in the iris
   b) Cataracts
   c) Glaucoma
   d) Retinitis pigmentosa
   e) Kayser-Fleischer rings in the cornea

536. In patients with a Wilson disease, the abnormal gene is located on the:
   a) Chromosome 1
   b) Chromosome 7
   c) Chromosome 13
   d) Chromosome 18
   e) Chromosome 21

537. The predominant clinical manifestation in younger patients with a Wilson disease is:
   a) Hemolysis
   b) Neurologic manifestations
   c) Behavioral changes
   d) Hepatic manifestations
   e) Kayser-Fleischer rings in the cornea

538. All of the following hepatic conditions can occur in patients with a Wilson disease except:
   a) Asymptomatic hepatomegaly
   b) Subacute hepatitis
   c) Chronic hepatitis
   d) Hepatoblastoma
   e) Fulminant hepatic failure

539. Kayser-Fleische rings in the cornea are always present in patients with Wilson disease and have the following symptoms:
   a) Hepatic
   b) Behavioral
   c) Hematologic
547. The most common presentations in patients with a primary ciliary dyskinesia are:
   a) Situs inversus, otitis media, and chronic sinusitis
   b) Chronic sinusitis, otitis media, and wheezing
   c) Bronchiectasis, chronic sinusitis, and asthma
   d) Asthma, pneumonia, and bronchiectasis
   e) Productive cough, sinusitis and otitis media

548. The mode of inheritance in patients with a primary ciliary dyskinesia is:
   a) Autosomal recessive
   b) Autosomal dominant
   c) X-linked recessive
   d) X-linked dominant
   e) Unknown

549. The gold standard to make the diagnosis of primary ciliary dyskinesia is:
   a) CT scan of paranasal sinuses
   b) Pulmonary function tests
   c) CT scan of lungs
   d) Ultrasonography of sinuses
   e) Nasal biopsy

550. A pulmonary function test in older children with a primary ciliary dyskinesia reveals:
   a) Normal
   b) Restrictive lung disease
   c) Obstructive lung disease
   d) Both restrictive and obstructive lung diseases with an equal frequency
   e) More restrictive than an obstructive lung disease

551. The cardiac output in a normal healthy newborn is about:
   a) 75 mL/kg/minute
   b) 150 mL/kg/min
   c) 200 mL/kg/minute
   d) 300 mL/kg/min
   e) 350 mL/kg/minute

552. The foramen ovale is functionally closed in a normal healthy newborn by:
   a) 12 hours of life
   b) 24 hours of life
   c) 7 days of life
   d) 1 month of life
   e) 3 months of life

553. The ductus arteriosus is functionally closed in a normal healthy newborn by
   a) 0-5 hours of life
   b) 5-10 hours of life
   c) 10-15 hours of life
   d) 15-20 hours of life
   e) 20-25 hours of life

554. The ductus arteriosus closes when the PO\textsubscript{2} of the blood at the ductus is about:
   a) 45 mm Hg
   b) 50 mm Hg
   c) 55 mm Hg
   d) 60 mm Hg
   e) 65 mm Hg

555. The most common cardiac anomaly in patients with an autosomal dominant polycystic kidney disease is:
   a) ASD
   b) VSD
c) Mitral valve prolapse
d) Peripheral pulmonic stenosis
e) Tricuspid atresia

556. Facio-auriculo-vertebral spectrum (FAVS) is associated with the following cardiac anomaly:
   a) ASD
   b) Tetralogy of Fallot
   c) Tricuspid stenosis
   d) Truncus arteriosus
   e) Transposition of great arteries

557. Cat-eye syndrome has the following chromosomal abnormality:
   a) Trisomy 9
   b) Trisomy 21
   c) Trisomy 21
   d) Trisomy 13
   e) Trisomy 5

558. The most common cause of death in pediatric patients with a heart transplantation is:
   a) CMV infection
   b) Candida albicans infection
   c) Protozoal infection
   d) Toxoplasma infection
   e) S. aureus infection

559. All of the following suggestions are useful as a prevention of high blood pressure in pediatric population except:
   a) Control obesity
   b) Reduce dietary sodium intake
   c) Physical exercise
   d) Reduce serum cholesterol levels
   e) Consume a small amount of red wine

560. A 9-month-old child is treated with an iron for iron deficiency anemia. The iron therapy has failed to improve the child's condition. Some of the factors responsible for the failure include:
   a) Wrong diagnosis
   b) The child does not receive iron therapy
   c) Iron is given in a form that is poorly absorbed.
   d) Unrecognized intestinal blood loss
   e) Unrecognized 'ABO' hemolytic disease

561. A child is diagnosed to have a hereditary spherocytosis. He has a positive family history. A physical examination reveals splenomegaly. The peripheral smear reveals spherocytes. The presence of spherocytes in the blood can be confirmed by:
   a) Positive Coombs test
   b) RBC membrane protein analysis
   c) Presence of anti-RBC antibodies
   d) Osmotic fragility test
   e) Measurement of RBC sizes

562. A patient with a paroxysmal nocturnal hemoglobinurea can be treated with all of the following except:
   a) Prednisone
   b) Fluoxymesterone
   c) Erythropoietin
   d) Splenectomy
   e) Bone marrow transplantation

563. A pregnant mother has a normal platelet count. She is carrying a fetus with alloimmune thrombocytopenia. All of the following therapies are indicated except:
   a) Monitoring of fetal platelet counts by PUBS (percutaneous umbilical blood sampling)
   b) Give IVIG to the mother beginning in the second trimester.
657. A severe form of familial hyperinsulinemic hypoglycemia is most likely due to the defect of the:
   a) Islet alpha cell $K_{ATP}$ channel
   b) Islet beta cell $K_{ATP}$ channel
   c) Islet gamma cell $K_{ATP}$ channel
   d) Islet delta cell $K_{ATP}$ channel
   e) Islet beta cell non-$K_{ATP}$ channel

658. A 17-year-old boy for a routine physical examination. He is asymptomatic but experienced occasional headaches. His recorded blood pressure is above the 95th percentile for his age. The most likely diagnosis is:
   a) Aortic coarctation
   b) Renal artery stenosis
   c) Pheochromocytoma
   d) Hyperthyroidism
   e) Suspected hypertension

659. A 16-year-old boy appears for a routine physical examination. He is asymptomatic. His recorded blood pressures are between 90th and 95th percentiles. He is obese. In addition to weight loss, the next step in management is:
   a) A follow-up examination after 1 month
   b) A follow-up examination after 3 months
   c) A follow-up examination after 6 months
   d) Furosemide
   e) Aldomet

660. A 13-year-old boy appears for a routine physical examination. He is asymptomatic. His recorded blood pressure is below the 90th percentile. The next step in management is:
   a) Echocardiography
   b) Renal ultrasonography
   c) Follow-up after 1 year
   d) Low-salt diet
   e) Blood pressure should be measured on three separate occasions

661. A 17-year-old girl appears at 2 AM in ER with a violent behavior and psychotic ideation. The girl was asymptomatic prior to this episode. A physical examination reveals hypotension, hyperpyrexia, slowing of cardiac conduction and ventricular irritabilities. The most likely diagnosis is:
   a) Cocaine intoxication
   b) Opiates overdose
   c) Amphetamine overdose
   d) Marijuana overdose
   e) Lysergic acid diethylamide intoxication

662. A 16-year-old girl appears with a white, floccular vaginal discharge and itching for the last 7 days. The pH of the discharge is less than 4.5. The discharge has no WBCs, clue cells, and trichomonads; absence of mycelia. The most likely diagnosis is:
   a) Bacterial vaginosis
   b) Trichomonas infection
   c) Candida infection
   d) Herpes simplex virus infection
   e) Normal vaginal discharge

663. A 17-year-old girl appears with a gray, yellow homogenous vaginal discharge for the last 10 days. The pH of discharge is more than 4.5. The discharge contains clue cells, few white cells, but absence of lactobacilli. The most likely diagnosis is:
   a) Trichomonas infection
   b) Candida infection
   c) Herpes simplex virus infection
   d) Foreign body
   e) Bacterial vaginosis

664. The preferred screening test for patients with complement defects is:
   a) $C_3$
   b) $C_4$
688. The most common extraintestinal manifestation in patients with a bacillary dysentery is:
   a) Cardiac
   b) Hepatic
   c) Pancreatic
   d) Neurologic
   e) Renal

689. The most common cause of neurologic manifestations in patients with a bacillary dysentery is:
   a) Shiga toxin
   b) Hypercalcemia
   c) Hyponatremia
   d) Increased ADH secretion
   e) Unknown

690. The preferred therapy in children with intrathoracic tuberculosis (pulmonary TB and/or hilar lymphadenopathy) is:
   a) A regimen of INH and rifampin for 9 months.
   b) A regimen of INH and ethambutol for 9 months.
   c) A regimen of INH and rifampin for 6 months plus first 2 months of treatment with pyrazinamide.
   d) A regimen of rifampin and pyrazinamide for 6 months.
   e) A regimen of INH and pyrazinamide for 6 months.

691. The preferred therapy in HIV-seropositive children with a drug-susceptible tuberculosis is:
   a) A regimen of INH, rifampin (RIF), pyrazinamide (PZA) for the first 2 months followed by INH and PZA for a total duration of at least 9 months.
   b) A regimen of INH, RIF, PZA for the first 2 months followed by RIF and PZA for a total duration of at least 6 months.
   c) A regimen of INH and RIF for the first 2 months followed by INH and PZA for a total duration of at least 12 months.
   d) A regimen of INH and PZA for the first 3 months followed by INH and RIF for a total duration of at least 9 months.
   e) A regimen of INH, RIF and PZA for the first 2 months followed by INH and RIF for a total duration of at least 9 months.

692. All of the following statements are true about drug-resistant tuberculosis except:
   a) The most drug resistance in children is primary.
   b) The major cause of secondary drug resistance is poor compliance with the medication.
   c) A noncompliance with one drug is more important cause of secondary resistance than failure to take all medications.
   d) The major cause of primary drug resistance is an inadequate treatment regimen prescribed by the doctor.
   e) A successful therapy in a drug-resistant tuberculosis should include at least two bactericidal drugs.

693. The preferred therapy in children with INH-resistant tuberculosis is:
   a) A regimen of RIF, PZA, and ethambutol (EMB) for 9 months.
   b) A regimen of RIF and PZA for the first 3 months followed by RIF, PZA, and EMB for at least 9 months.
   c) A regimen of PZA and EMB for the first 2 months followed by PZA, EMB, and RIF for at least 6 months.
   d) A regimen of RIF, PZA, and EMB for 6 months.
   e) A regimen of streptomycin for the first 1 month followed by RIF, PZA, and EMB for at least 9 months.

694. A child appears with mild fever, cough, and sore throat for the last 7 days. A physical examination reveals pharyngeal redness, bilateral rales, and mild wheezing. His mother denies history of asthma. He was healthy prior to this episode. The chest x-ray reveals bilateral diffuse infiltrates and mild pleural effusions. The complete blood count is normal without a left shift. The most likely diagnosis is:
   a) Newly diagnosed asthma
   b) Streptococcus pneumonia
   c) Staphylococcus aureus pneumonia
   d) HIV-infected pneumonia
   e) Chlamydia pneumonia

695. The most common site of infection in newborns with untreated maternal genital chlamydia infections is:
   a) Rectum
   b) Lungs
   c) Nasopharynx
   d) Oropharynx
   e) Eyes
727. A child appears in the ER with a shock, decreased urinary output, and altered mental status. A physical examination reveals tachycardia, tachypnea, cool extremities, dry mucous membranes, poor skin turgor, and hypotension. The most likely diagnosis is:
   a) Hypovolemic shock
   b) Obstructive shock
   c) Distributive shock
   d) Cardiogenic shock
   e) Late septic shock

728. The most common complication occurs in patients during induction of anesthesia is:
   a) Vomiting
   b) Bronchospasms
   c) Laryngospasms
   d) Aspiration pneumonia
   e) Aspiration of gastric contents

729. The most common postoperative complication after anesthesia is:
   a) Apnea
   b) Hypothermia
   c) Malignant hyperthermia
   d) Atelactasis
   e) Nausea and vomiting

730. All of the following are clinical features of neurologic malignant syndrome except:
   a) Muscle rigidity
   b) High fever
   c) Hypercarbia
   d) Metabolic alkalosis
   e) Hemodynamic collapse

731. A 7-year-old boy has appendicitis. He is going for an appendectomy. His mother died during anesthesia that was introduced for hysterectomy. The boy developed high fevers, metabolic acidosis, hypercarbia, muscle rigidity, and cardiovascular collapse, during anesthesia. The management includes all of the following except:
   a) Discontinue inhalation anesthesia
   b) Give succinylcholine
   c) Sodium bicarbonate
   d) Sodium dantrolene
   e) Mechanical ventilation

732. All of the following clinical features are diagnostic of neurofibromatosis 1 except:
   a) Optic glioma and iris hamartomas
   b) Freckling in the axillary regions and the child’s father has neurofibromatosis 1.
   c) Six cafe au lait macules more than 5 mm in greatest diameter in a prepubertal child and sphenoid dysplasia.
   d) Six cafe au lait macules more than 5 mm in greatest diameter in a postpubertal adolescent and his/her mother has neurofibromatosis 1.
   e) Six cafe au lait macules more than 5 mm in greatest diameter in a postpubertal adolescent and his/her mother has neurofibromatosis 1.

733. The following syndrome is due to deletion of paternally derived chromosome 15:
   a) Angelman syndrome
   b) Down syndrome
   c) Prader-Willi syndrome
   d) Turner syndrome
   e) Noonan syndrome

734. The following syndrome is due to deletion of maternally derived chromosome 15:
   a) William syndrome
   b) Rubinstein-Taybi syndrome
   c) Angelman syndrome
   d) Klinefelter syndrome
   e) Velocardiofacial syndrome
d) Fire ants
e) Penicillins

759. An adolescent boy appears with a pain in the flower back. His lower spine remains straight when he bends forward. The x-ray of both sacroiliac joint reveals a severe sclerosis, erosion of joint margins, and apparent widening of the joint space. The most likely diagnosis is:
   a) Reactive arthritis
   b) Post traumatic arthritis
   c) Ankylosing spondylitis
   d) Reactive arthritis
   e) Psoriatic arthritis

760. A 7-year-old appears with URI syndrome and pain in the right hip joint for the last 5 days. He denies history of a trauma. A physical examination reveals a tenderness in the right hip joint. CBC and ESR values are normal. X-ray of the right hip reveals a widening of the joint space. The next step in management is:
   a) Orthopedic consultation
   b) Hip spica casting
   c) Azithromycin
   d) Ampicillin
   e) Cefuroxime

761. A child appears with rashes and multiple joints arthritis that resembles serum sickness. The girl is not immunized properly. The most likely diagnosis is:
   a) Rubella-associated arthropathy
   b) Poststreptococcal arthritis
   c) Shigella-associated arthritis
   d) Salmonella-associated arthritis
   e) Arthritis-dermatitis syndrome

762. A child developed arthralgias of the knees and hands after the rubella vaccination. Postvaccination arthritis due to rubella usually occurs:
   a) 1-3 days after vaccination
   b) 4-9 days after vaccination
   c) 10-28 days after vaccination
   d) 29-60 days after vaccination
   e) 61-90 days after vaccination

763. All of the following statements are true in patients with a rubella-associated arthropathy except:
   a) More common after natural rubella infections than after rubella vaccinations.
   b) Arthralgias commonly occurs in knees and hands.
   c) Common in preadolescent children
   d) Arthralgias usually begin within 7 days of onset of the rash due to a natural infections.
   e) Typically occurs in young women.

764. A 14-year-old girl is diagnosed with SLE (systemic lupus erythematosus). She is receiving corticosteroids therapy. All of the following are side effects of corticosteroids therapy except:
   a) Gastritis
   b) Cataracts
   c) Hypertension
   d) Osteoporosis
   e) Cushingoid features

765. The most specific diagnostic laboratory test in patients with a SLE is:
   a) Positive LE cell preparation
   b) Elevated ANA titers
   c) Decreased CH50
   d) Anti-DNA antibody to native DNA in abnormal titer
   e) Anti-Sm-presence of antibody to Sm nuclear antigen
782. A child appears with a photophobia, dry cough, coryza, conjunctivitis, and mild fever for the last 2 days. A physical examination reveals a transverse line of conjunctival inflammation and the line is sharply demarcated along the eyelid margin. The most likely diagnosis is:
   a) Rubella
   b) Glaucoma
   c) Bacteria conjunctivitis
   d) Measles
   e) Chlamydia conjunctivitis

783. A Koplik spot is a pathognomonic sign of measles. Koplik spots usually appear as follows:
   a) Grayish white dots appear opposite the lower molars.
   b) Bluish white dots appear opposite the upper molars.
   c) Pinkish white dots appear opposite the lower molars.
   d) Grayish yellow dots appear opposite the lower molars.
   e) Yellowish white dots appear opposite the lower molars.

784. Koplik spots consist of the following:
   a) Polymorphonuclear leukocytes and pus cells
   b) Lymphocytes and pus cells
   c) Eosinophils and pus cells
   d) Serous exudates and endothelial cells
   e) Mucous exudates and lymphocytes

785. The preferred diagnostic study in patients with a herpes simplex virus encephalitis beyond the neonatal period is:
   a) MRI of the brain
   b) CT scan of the brain
   c) PCR analysis of CSF
   d) Brain biopsy
   e) Analysis of CSF cells

786. The patients with a sarcoidosis usually develop the following condition:
   a) Hypocalciuria
   b) Hypercalcemia
   c) Hypercalciuria
   d) Hyperkalemia
   e) Hypocalcemia

787. The most common cause of neonatal hyperparathyroidism is:
   a) Maternal hypercalcemia
   b) Maternal hyperparathyroidism
   c) Parathyroid adenoma
   d) Pituitary tumor
   e) Absence of functional calcium-sensing receptors in the parathyroid glands

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

789. A 2-year-old girl has a history of persistent thumb sucking. Her mother is worried. The next step in management:
   a) Put a bandage in the thumb
   b) Put a gloves in the hand
   c) Use a noxious agent
   d) Don’t give food until she stops thumb sucking
   e) Reassurance
c) 70% of cases are resolved by 10 months of age.
d) 80% of cases are resolved by 11 months of age.
e) 90% of cases are resolved by 12 months of age.

805. The most common congenital laryngeal anomaly in infants is:
   a) Laryngomalacia
   b) Subglottic stenosis
   c) Subglottic hemangioma
   d) Laryngeal web
   e) Vocal cord paralysis

806. A full term newborn appears with low pitched, inspiratory stridor at birth. The stridor worsens with crying, agitation, and feeding. The newborn is NPO and receiving intravenous fluids. He was born by an elective cesarean section with Apgar scores are 8 and 9 at 1 and 5 minutes respectively. The most likely diagnosis is:
   a) Vocal cord paralysis
   b) Laryngeal web
   c) Laryngeal atresia
   d) Laryngeal nodule
   e) Laryngomalacia

807. The preferred diagnostic study in infants with laryngomalacia is:
   a) AP x-ray of the neck
   b) Lateral x-ray of the neck
   c) Flexible laryngoscopy
   d) Barium swallow of the esophagus
   e) Flexible bronchoscopy

808. A 2-day-old newborn male appears with cyanosis and tachypnea for the past 2 hours. He was born by NSVD with Apgar scores are 8 and 9 at 1 and 5 minutes respectively. The physical examination reveals a grade 2/6 systolic murmur at the left sternal border. The arterial PO$_2$ is 80 in hyperoxia test. The oxygen saturation values are as follows: RA 58%, RV 58%, LA 100%, LV 94%, pulmonary artery 58%, and ascending aorta 58%, the volume of blood passes 1.0 L/min, different structures are as follows: RA 3 L/min/m$^2$, RV 3 L/min/m$^2$, LA 2 L/min/m$^2$, LV 2 L/min/m$^2$, pulmonary artery 2 L/min/m$^2$, and ascending aorta 3 L/min/m$^2$. The newborn is receiving PGE1 infusions. The preferred surgical therapy is:
   a) Switch operation (Jatene)
   b) Modified Blalock-Taussig shunt
   c) Starnes procedure
   d) Norwood operation
   e) Fontan procedure

809. A newborn appears with cyanosis and respiratory distress within the 1st hour of life. She was born by NSVD with Apgar score of 7 and 8 at 1 and 5 minutes respectively. The physical examination reveals a soft systolic ejection murmur at the midleft sternal border and single, loud 2nd heart sound. The arterial PO$_2$ is 60 in hyperoxia test. The oxygen saturation values are as follows: both vena cava 58%, RA 73%, RV 73%, LA 90%, LV 90%, pulmonary artery 90%, right and left pulmonary arteries 80%, pulmonary veins 100%, and ascending aorta 73%. The newborn is placed on a mechanical ventilator and PGE$_1$ infusions have started. The preferred initial surgical therapy is:
   a) Norwood operation
   b) Glenn operation
   c) Fontan procedure
   d) Aortic valvuloplasty
   e) Rashkind procedure

810. The pulmonary arteriovenous fistulas are most commonly present is:
   a) Williams syndrome
   b) Down syndrome
   c) Turner syndrome
   d) Prader-Willi syndrome
   e) Osler-Weber-Rendu syndrome
819. The rate of urine production at term is about:
   a) 40 mL/hour
   b) 45 mL/hour
   c) 51 mL/hour
   d) 60 mL/hour
   e) 70 mL/hour

820. The glomerular filtration rate at term is about:
   a) 15 mL/min/1.73 m²
   b) 20 mL/min/1.73 m²
   c) 25 mL/min/1.73 m²
   d) 30 mL/min/1.73 m²
   e) 35 mL/min/1.73 m²

821. The glomerular filtration rate triples since birth in full term infants by:
   a) 1 month of age
   b) 3 months of age
   c) 6 months of age
   d) 9 months of age
   e) 12 months of age

822. All of the following are manifestations of septo-optic dysplasia except:
   a) Nystagmus
   b) Visual impairments
   c) Optic nerve dysplasia
   d) Large anterior pituitary gland
   e) Incomplete development of septum pellucidum

823. In girls, McCune-Albright syndrome appears with all of the following clinical and laboratory features except:
   a) Early vaginal bleeding
   b) Precocious puberty
   c) Decreased level of estrogen
   d) Decreased levels of LH
   e) Does not respond to GnRH stimulation

824. In boys, McCune-Albright syndrome, all of the following statements are true except:
   a) Precious puberty is more common.
   b) Testicular enlargements are fairly symmetric.
   c) Phallic enlargement occurs after testicular enlargement.
   d) During puberty, the response to GnRH becomes pubertal.
   e) 

825. The preferred therapy in patients with McCune-Albright syndrome whose puberty has shifted from a pseudo (gonadotropin-independent) to true (gonadotropin-dependent) mechanism is:
   a) Estrogen
   b) Progesterone
   c) FSH
   d) Short-acting analogs of GnRH
   e) Long-acting analogs of GnRH

826. A 3-week-old girl appears with sudden onset of tetany and seizure activities. She was born by NSVD with Apgar score of 8 and 9 at 1 and 5 minutes respectively. She is receiving breast milk since birth. She was completely asymptomatic prior to this episode. The serum calcium level is 5 mg/dL and ionized calcium level is markedly reduced. The serum magnesium level is 1.2 mg/dL. The next step in management is:
   a) Intravenous calcium
   b) Calcium gluconate
   c) PTH
   d) Magnesium
   e) 1, 25-dihydroxycholecalciferol
Match the following growth findings and different conditions (899-903):

899. A child remains short until puberty, then increases height, and bone age is less than chronological age.
900. A child remains short for rest of life. Both parents are short. Bone age is the same as chronological age.
901. A child suddenly stops growing and then, started to grow again after medication.
902. A child suddenly stops growing and the, started to grow after surgery.
903. A child suddenly stops growing, has electrolyte imbalance, and then, started to grow after medication.

   a) Congenital adrenal hyperplasia
   b) Cranioopharyngioma
   c) Constitutional short stature
   d) Hypothyroidism
   e) Genetic short stature

Match the following arterial blood gases and different conditions (904-908):

904. pH 7.21, PCO₂ 62, PO₂ 90, bicarbonate 34
905. pH 7.58, PCO₂ 17, PO₂ 95, bicarbonate 11
906. pH 7.56, PCO₂ 36, PO₂ 88, bicarbonate 37
907. pH 7.19, PCO₂ 18, PO₂ 49, bicarbonate 12
908. pH 7.36, PCO₂ 61, PO₂ 92, bicarbonate 35

   a) Respiratory alkalosis
   b) Respiratory acidosis
   c) Metabolic alkalosis
   d) Metabolic acidosis
   e) Compensated respiratory acidosis

Match the following visual pathway lesions and findings (909-913):

909. Unilateral vision loss
910. Bitemporal hemianopsia
911. Left homonymous hemianopsia
912. Right homonymous hemianopsia
913. Partial homonymous hemianopsia

   a) Left optic tract
   b) Lateral geniculate body
   c) Optic chiasma
   d) Right optic tract
   e) Optic nerve

Match the following hematologic findings and different conditions (914-918):

914. Hemoglobin A 95%, hemoglobin A2 3%, hemoglobin F less than 1%, hemoglobin S 0%, normal hemoglobin, and normal MCV
915. Hemoglobin A 0%, hemoglobin A2 normal, hemoglobin F 8%, hemoglobin S 92%, hemoglobin 7, and normal MCV
916. Hemoglobin A 55%, hemoglobin A2 normal, hemoglobin F normal, hemoglobin S 45%, normal hemoglobin, and normal MCV
917. Hemoglobin A 0%, hemoglobin A2 0%, hemoglobin F 1%, hemoglobin S 50%, hemoglobin C 50%, and hemoglobin 9.
918. Hemoglobin A 28%, hemoglobin A2 normal, increased ratio A2/A, hemoglobin F 72%, hemoglobin S 0%, hemoglobin 4, and MCV 45.

   a) Sickle cell disease
   b) Hemoglobin SC disease
   c) Normal
   d) Thalassemia major
   e) Sickle cell trait
Questions

Match the characteristic of sinusitis and site of pain/headache (955-959):

955. Frontal
956. Sphenoidal
957. Anterior ethmoidal
958. Posterior ethmoidal
959. Maxillary

a) Suboccipital area
b) Temporal area and eyes
c) Trigeminal area particularly mastoid
d) Over maxilla and teeth
e) Above and medial to eyes

Match the glucose levels and insulin doses in type 1 diabetes mellitus (960-964):

960. Increased glucose level in morning
961. Increased glucose level at noon
962. Increased glucose level in evening
963. Increased glucose level in night
964. Increased glucose level in night

a) Decrease this P.M. dose of regular insulin
b) Increase last P.M. dose of NPH insulin
c) Increase this A.M. dose of regular insulin
d) Increase this A.M. dose of NPH insulin
e) Increase this P.M. dose of regular insulin

Match different types of cough and diseases (965-969):

965. Tracheitis
966. Chlamydia pneumonia
967. Laryngeal obstruction
968. Habit cough
969. Allergic reaction

a) Cough with stridor
b) Disappears with sleep
c) Brassy
d) Nocturnal cough
e) Staccato

Match different characteristics of cough and diseases (970-974):

970. Bronchitis
971. Cystic fibrosis
972. Reactive airways
973. Pertussis airway
974. Asthma

a) Most severe on awakening in morning
b) With vigorous exercise
c) Paroxysmal
d) Tight (wheezy)
e) Loose (discontinuous), productive
122. c) Subpial transection is indicated in LK syndrome when medical management fails.
123. e) ABR should be performed if a child failed OAEs testing. OAEs are absent if a hearing loss is more than 30-40 dB.
124. b) Paranasal sinusitis is the most common cause of orbital cellulitis in children. Answers (a), (c), (d), and (e) also can cause orbital cellulitis.
125. d) Tuberous sclerosis
126. b) Preceding 2-3 months
127. e) Thalassemia and other conditions have elevated fetal hemoglobin levels that falsely reduce the HbA1C values.
128. d) Sickle cell disease
129. a) Normal (less than 6%) HbA1C; a good control (6-8.5%); a fair control (9-10%); a poor control (11% and above).
130. d) Rhabdomyomas are present at the apex of left ventricle. Rhabdomyomas can cause arrhythmia and congestive cardiac failure.
131. c) Hamartomas or polycystic kidney disease can cause pain, hematuria, and in some cases, renal failure.
132. b) Angiomyolipomas can produce cystic or fibrous pulmonary changes resulting in spontaneous pneumothorax.
133. e) Subarachnoidal hemorrhages (traumatic or nontraumatic)
134. e) Purtscher retinopathy; Purtscher-like fundus is also noted in acute pancreatitis, SLE, and childbirth.
135. d) Elevated serum alkaline phosphatase activities are noted first; serum PTH levels is usually normal; answers (a), (b), (c), and (d) are also noted in familial hypophosphatemia. Increased urinary excretion of phosphate is due to a defect in renal tubular phosphate reabsorption probably secondary to failure of PHEX (phosphate-regulating gene with homologies to endopeptidases on the X chromosome) to cleave and decrease F6F23 action.
136. e) Bowing of lower extremities are smooth; answers (a), (b), (c), and (d) are noted in patients with calcium-deficient rickets.
Familial hypophosphatemia (a pure phosphate-deficient rickets) also manifests with waddling gait, cow-hoof, genu varum, genu valgum, and short stature.
137. c) Erythromycin. Tonsillectomy is curative therapy.
138. c) CT scan of an affected area or needle aspiration and culture can make the diagnosis of peritonsillar abscess.
139. e) A third-generation cephalosporin and ampicillin-sulbactum or clindamycin; surgical drainage is indicated in a patient with respiratory distress or failure to improve with antibiotic therapy.
140. e) Surgical drainage and a third-generation cephalosporin and clindamycin or ampicillin-sulbactum
141. d) ECG can reveal long Q-T syndrome.
142. d) Long Q-T syndrome; a heart-rate corrected Q-T interval of more than 0.47 second is highly indicative of long Q-T syndrome and more than 0.44 second is suggestive of long Q-T syndrome.
143. e) A patient with myxoma appears with fainting spells and have a positional character of murmur. Myxomas should be removed completely.
144. b) Left atrium (75% of cases); these smooth, pedunculated masses arise from an interatrial septum and protrude into the left atrium.
145. d) Overhydration; however, dehydration causes stress ulcers.
146. e) CT scan results are more sensitive and specific than that of ultrasonography.
147. a) Intravenous morphine should be used in patients with acute appendicitis to control the abdominal pain even before the diagnosis is confirmed. Morphine therapy does not delay the diagnosis.
148. a) PFT rarely makes a diagnosis. PFT can detect a functional impairment in patients with a normal physical examination but minor complaint. PFT is useful in determining responses of a bronchodilator in patients with an obstructive lung disease.
149. e) A deletion of a single phenylalanine residue at amino acid 508.
150. e) Unknown
151. b) Trisomy 18 (90% of cases); trisomy 21 (50% of cases); Turner syndrome (40% of cases)
152. e) Transposition of great vessels and left-sided obstructive lesions (e.g., aortic stenosis, hypoplastic left heart syndrome) are more common in boys than in girls.
153. d) Left upper lobe
154. a) Measure blood pressures in both lower extremities. In a normal person, systolic blood pressure in legs is 10-20 mm Hg higher than in arms. In patients with a coarctation of the aorta, blood pressures in lower extremities are lower than in upper extremities.
155. e) Breast bud is the first sign of puberty in girls.
269. c) 18 mm. The formula is:

\[
\text{Uncuffed ETT size (mm)} = 16 + \frac{\text{Age in years}}{4}
\]

\[
= 16 + \frac{8}{4} = 18 \text{ mm}
\]

270. d) All of the findings are characteristic of an early septic shock.

A patient with a **hypovolemic shock** has an increased SVR, decreased CO, WP, and CVP, normal or decreased MAP.

A patient with an **obstructive shock** has an increased SVR, WP, and CVP, decreased CO, and normal or decreased MAP.

A patient with **distributive shock** has an increased CO, decreased SVR, and normal or decreased MAP, WP, and CVP.

A patient with a **late septic shock** has an increased WP, decreased CO, SVR, and MAP, and normal or increased CVP.

A patient with a **systolic cardiogenic shock** has an increased SVR, WP, and CVP, decreased CO, and normal or decreased MAP.

A patient with a **diastolic cardiogenic shock** has an increased SVR, WP, and CVP, and normal CO and MAP.

A patient with an **early septic shock** has a normal, decreased, or increased MAP.

Please remember, WP, CVP, and pulmonary artery diastolic pressures are the same.

271. c) Cardiac output (CO) is increased in a distributive shock and early septic shock. Cardiac output is normal in a diastolic cardiogenic shock.

272. c) SVR is decreased in an early septic shock, late septic shock, and distributive shock.

273. a) CVP is decreased in an early septic shock and hypovolemic shock. CVP is normal or decreased in a distributive shock. CVP is increased or normal in a late septic shock.

274. a) Late septic shock. MAP is either normal or decreased in hypovolemic shock, cardiogenic (systolic, diastolic) shock, obstructive shock, and distributive shock. MAP is either normal, decreased, or increased in an early septic shock.

275. a) Infectious pneumonia and sepsis are the most common causes of ARDS. Other risk factors are shock, multiple transfusions, trauma, pulmonary contusion, near-drowning, donor oxygen reperfusion injury after lung transplantation, and acute pancreatitis.

276. b) 1-4 years of age (37% of cases); next group is 15-19 years of age (29% of cases)

277. b) 7-15 months of age (86% of cases)

278. d) Inadequate supervision by parents; they overestimated their child’s abilities or coordination.

279. a) Bathtub

280. e) Swimming pool

281. a) Freshwater

282. a) Most submersion events occur when there is a brief lack of supervision (i.e., less than 5 minutes); up to 80% of children are hospitalized for at least 24 hours.

283. d) An endotracheal tube can be maintained for months without tracheostomy. If necessary, a tracheostomy should be performed electively and delayed until burns at and near the site of tracheostomy have healed. Inhaled corticosteroid therapy may be useful. A patient with severe carbon monoxide poisoning needs hyperbaric oxygen therapy. A high-frequency ventilation is indicated when PO2 below 90% when on Fio2 between 90-100% and PEEP of at least 12.5 cm in a convention ventilator therapy.

284. e) Redness and blisters on the penis can occur. EMLA is a topical eutectic mixture of lidocaine and prilocaine.

285. a) FISH cannot be used as a screening tool. FISH is used to answer specific questions. FISH can identify specific DNA sequences in different recognizable diseases. FISH cannot provide information regarding the physical state of DNA or chromosome segment.

286. c) Phenotypically normal family members do not transmit the disease to their children. Autosomal dominant inheritance has a male-to-male (i.e., father to son) transmission that is absent in X-linked disorders because father gives only Y-chromosome to his son.

287. b) The patients with Klinefelter syndrome are relatively tall and have delayed secondary sexual development.

288. a) Autosomal recessive; the patients with this syndrome have a reduced number of melanosomes. These melanosomes are large in size (macromelanosomes).

289. a) MSUD (Maple Syrup Urine Disease)

290. d) Beta-hexosaminidase A isozyme
d) Smallpox; this boy has a characteristic presentation of smallpox. The synchronous nature of the rash with a centrifugal (away from center of a body) distribution can distinguish smallpox from chickenpox that rashes are nonsynchronous and centripetal (toward center of a body). The survivors of smallpox develop disfiguring and de-pigmented scars over the affected areas. Patients who are exposed to lewisite (blistering agent) experienced immediate pain, cutaneous vesicles, and injuries to eyes and airways.

e) Nonvenomous snakes have a rounded head and rounded pupil. Therefore, reassurance and local wound care are indicated because they have no fangs. Venomous snakes have a triangular-shaped head, elliptical pupil, and fangs (e.g., pit vipers, or Crotalidae). These are the most common poisonous snakes in USA.

e) Applying ice to the site of injury or using excision and suction is contraindicated because it causes more tissue damage.

e) Paralysis and respiratory failure are the most common complications. Antivenin is only effective if given before to the onset of symptoms but it cannot reverse the symptoms once started. The patient may remain asymptomatic for a few hours.

b) VSD (about 50% of cases)

e) Congenital absence of the pulmonary valve produces a syndrome that manifests with signs of an upper airway obstruction. Marked aneurysmal dilatation of the main and right or left pulmonary artery resulting in compression of the bronchi that causes wheezing and recurrent pneumonia. Cyanosis may be absent, mild, or moderate. This syndrome may be associated with TOF.

b) William syndrome has a deletion of elastin gene on chromosome 7q.

e) Subvalvular aortic stenosis may become apparent after a successful repair of other cardiac defects (e.g., VSD, PDA, or coarctation of aorta).

a) Konno procedure (i.e., obstruction of a left ventricular outflow tract can be relieved by borrowing space anteriorly from a right ventricular outflow tract).

d) Pulmonary atelactasis can occur in asthma, pneumonia, and trauma. Massive collapse of the lungs most commonly occurs after a surgery. The most common site of atelactasis is the right upper lobe.

c) Nausea is absent in hemoptysis.

d) Frothy blood is absent in hematemesis.

c) Right upper lobe abscesses. This is a rare chest x-ray finding of going asleep.

c) Lung CT scan is better than a chest x-ray.

a) A secondary lung abscess is more common on the left side.

d) Lung function is not present in all cardiac conditions are single ventricle, pulmonic stenosis, and pulmonary atresia.

e) Blalock-Taussig shunt or total repair is performed in patients with a tetralogy of Fallot. This patient has a high right ventricular pressure 40/6 mm Hg (normal 25/3 mm Hg), low pulmonary artery pressure 5 mm Hg (normal 25/10 mm Hg), reduced left ventricular saturation 80% (normal 94-100%), and reduced aortic saturation 80% (normal 94-100%).

b) Arterial switch (Jatene) operation is the preferred procedure for transposition of great vessels. However, Rashkind balloon atrial septostomy is the initial procedure of choice. This patient has a high right ventricular pressure 100/8 mm Hg (normal 25/3 mm/Hg), high pulmonary artery pressure 40/2 mm Hg (normal 25/10 mm Hg), decreased left ventricular pressure 40/2 mm Hg (normal 100/8 mm Hg).

a) Repair of VSD is indicated. This patient has an increased right ventricular pressure 35/5mm Hg (normal 25/3mm Hg), increased right ventricular saturation 80% (normal 74%), and increased pulmonary artery saturation 80% (normal 74%).

e) Correct hyponatremic dehydration.

d) Correct hypernatremia over 48 hours.

c) The patient should be admitted because the home-made formulas result in salt poisoning to this child.

a) Give excessive fluids, then DDAVP in a patient with diabetes insipidus.

d) Furosemide, hydrochlorothiazide, and bumetanide cause hypokalemia. Furosemide also causes metabolic alkalosis.

e) Nifedipine causes facial flushing and tachycardia.

a) Enalapril and diazoxide cause hypotension; enalaprilat causes transient hypotension; prazocin causes orthostatic hypotension.

a) Clonidine causes sedation, constipation, rebound withdrawal, and hypertension.

b) Vitamin B₁₂ deficiency can occur in breast-fed infants whose mothers are vegans or themselves have pernicious anemia.

a) Macrocytic anemia is present; Pearson’s syndrome is a unique variant of congenital sideroblastic anemias that are microcytic.
455. c) 1.0; 
Reticulocyte index = reticulocyte% x observed hematocrit x \( \frac{1}{\text{normal hematocrit}} \) 
(\( \mu \) is a maturation factor related to the severity of the anemia)

456. d) 3-4; a maximum of 6-8 corresponds to a maximal marrow response.

457. d) Hereditary spherocytosis is diagnosed by a positive family history, large number of spherocytes in peripheral blood smear, reticulocytosis, and splenomegaly.

458. e) Reassurance; the average heart rate in newborns ranges from 120 to 140 beats per minute and may increase up to 170+ beats per minute during crying.

459. e) Reassurance; the heart rate can drop to 70-90 beats per minute during sleep. When the baby is awake heart rate goes above 100-120 per minute.

460. e) Reassurance; the heart rate can normally drop up to 40 beats per minute in athletic adolescents.

461. e) A 16-years-old athlete’s heart rate up to 40 beats per minute is normal.

462. d) Differential cyanosis is noted in coarctation of aorta, interrupted aortic arch, and persistent pulmonary hypertension (PPHN).

463. a) Transposition of great arteries

464. e) Prominent venous plexus in lips

465. e) Prominent venous plexus on the forehead.

466. d) Unwrapped and cold extremities (i.e., acrocyanosis)

467. a) S. epidermidis is common in the presence of an indwelling central venous catheter or prosthetic valve. For a native valve, the common organisms are viridans group streptococci (e.g., S. mutans, S. sanguis, S. mitis), S. aureus, and Group D Streptococcus (enterococcus) (S. bovis, S. faecalis).

468. a) S. aureus

469. a) Viridans group streptococci (e.g., S. mutans, S. sanguis, S. mitis)

470. a) Group D streptococcus (enterococcus) (e.g., S. bovis, S. faecalis)

471. e) Group D streptococcus (enterococcus) (e.g., S. bovis, S. faecalis)

472. b) Pseudomonas aeruginosa or Serratia marcesces

473. c) Staphylococcus epidermidis

474. d) Poor dental hygiene

475. e) Repair of a simple ASD or closure of PDA almost eliminates the risk of developing endocarditis. A surgical correction of a congenital heart disease can reduce but does not eliminate the risk of endocarditis except the two conditions mentioned above.

476. d) Tricuspid regurgitation (TR) is not a preoperative condition and does not produce infective endocarditis. TR is common in newborns with a perinatal asphyxia secondary to transient papillary muscles dysfunction. TR is a self-limiting condition. However, mitral valve prolapse, bicuspid aortic valves, PDA, ASD, and other anatomical cardiac defects, especially involving the left side of the heart can develop infective endocarditis.

477. d) The patients with congenital hypoplastic anemia (Diamond-Blackfan anemia) may appear clinically with short stature, craniofacial abnormalities, or defects in upper extremities (e.g., triphalangeal thumbs). Answer (c) should be ruled out by a PCR study. Answer (e) patients usually manifest after 6 months of age.

478. b) Fetal hemoglobin levels are elevated; folic acid and vitamin B\(_12\) levels are normal; thrombocytosis or thrombocytopenia is noted; bone marrow chromosome studies are normal unlike in Fanconi anemia.

479. b) Corticosteroid; the patients who do not respond to corticosteroid therapy, transfusions at intervals of 1-2 months are indicated to maintain a normal growth and development, and a chelating agent (e.g., deferoxamine, deferiprone) is used to avoid iron overload; stem cell transplantation is indicated if above therapies fail.

480. d) Glionomas are common between 2-5 years of age. Medulloblastomas and primitive neuroectodermal tumors (PNET) are common in less than 1 year of age.

481. e) Non-Hodgkin lymphomas, gliomas, and acute lymphoid leukemias are more common between 2-5 years of age. Testicular and ovarian cancers are more common during puberty.

482. d) Osteosarcomas; a therapeutic radiation to the brain causes brain cancers. Ionizing radiation therapy can also cause ALL (acute lymphoid leukemia).

483. a) Non-Hodgkin lymphomas (2-5 years of age) and Hodgkin lymphomas (6-18 years of age) are associated with E-B virus infections.