1. Three years ago, a 15-year-old boy was treated for Ewing sarcoma. For the past month, he has complained of worsening fatigue, exercise intolerance, loss of appetite, persistent dry cough, and edema of the lower extremities. The total dose of adriamycin he received was 575 mg/m²; he also received irradiation to the thorax.

The MOST likely diagnosis is:

A. Anemia
B. Congestive heart failure
C. Pericarditis
D. Reactive airway disease
E. Supraventricular tachycardia

2. An irritable 8-year-old is seen with respiratory distress, tachypnea, tachycardia, and a temperature of 102.2°F (39°C). On physical examination, the patient has distended neck veins, hepatomegaly, a quiet precordium, distant heart sounds, and a pulsus paradoxus of >10 mmHg. These findings are suggestive of which of the following diagnoses?

A. Rheumatic heart disease
B. Myocarditis
C. Lyme syndrome
D. Pericarditis
E. Congestive heart failure
3. While performing ultrasonography on a 31-week fetus, an obstetrician notes that the fetal heart rate ranges from 62 to 66 beats/min. Fetal growth appears normal and no structural cardiac anomalies are identified. Fetal echocardiography reveals that the fetal atria appear to be contracting at 140 beats/min, with a ventricular rate of 65 beats/min.

Of the following, the NEXT step in the management of this infant is to:

A. Administer beta-agonist drug therapy to the mother
B. Assess the cardiac status of the infant following labor and delivery
C. Counsel the parents that intrauterine fetal death is likely
D. Perform amniocentesis to confirm lung maturity and if mature, perform immediate cesarean section
E. Repeat the fetal echocardiogram and fetal ultrasound in 1 week

4. An 18-month-old boy has had persistently high fever for 7 days. You suspect Kawasaki disease (mucocutaneous lymph node syndrome).

Of the following, physical examination is LEAST likely to reveal:

A. Cervical lymphadenopathy
B. Edema of the hands and feet
C. Polymorphous rash
D. Purulent conjunctivitis
E. Strawberry tongue
5. A 1-year-old child is brought to the emergency department because his parents thought his heart was “pounding” as they were putting him to bed. ECG reveals a heart rate of 300 beats/min that spontaneously converts to sinus rhythm 100 bpm. The parents estimate the tachycardia lasted 20 minutes; the child was asymptomatic throughout.

Of the following, the MOST appropriate management of this child is:
A. Administration of a beta blocker
B. Administration of digoxin
C. Administration of procainamide
D. Administration of verapamil
E. Observation without drug therapy

6. A 3-month-old boy has respiratory distress and lethargy. Findings include tachypnea, hepatomegaly, heart rate 300 beats/min, and a narrow QRS complex consistent with supraventricular tachycardia.

Of the following, the MOST appropriate treatment for the patient at this time is:
A. Electrical cardioversion
B. Intravenous verapamil
C. Oral digoxin
D. Oral verapamil
E. Parenteral digoxin

7. A 30-year-old woman who has active systemic lupus erythematosus is pregnant.

Of the following, the abnormality of cardiac conduction that is MOST likely to occur in her infant is:
A. Atrial flutter
B. First-degree heart block
C. Supraventricular tachycardia
D. Third-degree heart block
E. Ventricular tachycardia
A 5-year-old girl is very excited following a ride on a roller coaster. In the midst of her excitement she suddenly loses consciousness and falls to the ground. Paramedics on the scene document ventricular tachycardia. Family history reveals a maternal uncle who died suddenly at 16 years of age.

8. Following treatment of the ventricular tachycardia, an ECG MOST likely will demonstrate:
   A. Corrected QT interval of 0.52 sec
   B. P wave axis of –30 degrees
   C. PR interval of 0.81 seconds
   D. QRS axis of –15 degrees
   E. QRS interval of 0.12 seconds

9. In addition to a prolonged QT interval, an individual who has Jervell and Lange-Nielsen syndrome is MOST likely to have:
   A. Alopecia universalis
   B. Cranial bruits
   C. Hepatosplenomegaly
   D. Rotary nystagmus
   E. Sensorineural hearing loss
10. The sport you are most likely to suggest that patients who have LQTS avoid is:

A. Bicycling
B. Bowling
C. Ice skating
D. Swimming
E. Tennis

11. A 16 year old athlete has a syncopal episode immediately following a high school basketball game. ECG reveals a QTc of 0.52 sec.

Which of the following family members would you recommend must have a screening ECG?

A. All first-degree relatives
B. Brothers and male first cousins
C. Sisters and female cousins
D. Father and both grandfathers
E. Mother and both grandmothers

12. A 14-year-old girl falls during a race. She is unconscious, cyanotic and has no pulse, but spontaneously revives within seconds. Both patient and family histories are benign. Results of physical exam, chest x-ray, echocardiography, EEG, and exercise ECG during a treadmill stress test are normal.

The most appropriate NEXT step in management is to

A. Order a 30-day ECG event recorder
B. Perform cardiac catheterization studies
C. Perform 24-hour ambulatory ECG monitoring
D. Perform tilt-table testing
E. Reassure the family that cardiac etiologies have been excluded
13. Which one of the following statements regarding coronary artery aneurysms due to Kawasaki syndrome is TRUE?

A. Aneurysmal rupture and hemorrhage is the most common mechanism of sudden death.
B. Coronary artery aneurysms typically appear 3 months after the onset of Kawasaki syndrome.
C. Risk of coronary artery aneurysm is decreased by intravenous gamma globulin therapy.
D. Serial echocardiographic evaluation at 3 month intervals over a 2 year period is necessary for all patients.
E. There is no correlation between the duration of fever and the development of giant coronary aneurysm.

14. For treatment of Kawasaki syndrome, the BEST time to administer gamma globulin is:

A. After the congestive heart failure subsides.
B. After the fever subsides.
C. After treatment with corticosteroids is initiated.
D. As soon as the diagnosis is made.
E. When cutaneous manifestations of vasculitis start to resolve.

15. The most important factor in planning follow-up care for a child who has Kawasaki syndrome is:

A. Age of the patient.
B. Duration of the febrile illness.
C. Presence or absence of arthritis.
D. Presence or absence of coronary artery abnormalities.
E. Severity of the original liver disease.
16. A systolic ejection murmur is detected during the routine sports preparticipation examination of a highly competitive 16-year-old high school athlete. ECG demonstrates a sinus bradycardia (50 bpm) and borderline left ventricular hypertrophy. Echocardiography reveals mildly enlarged left and right ventricles with a ventricular shortening fraction of 40% (normal > 28%).

Of the following, the MOST likely explanation for these findings is:

A. Autonomic dysfunction
B. Cardiovascular response to athletic training
C. Early evidence of dilated cardiomyopathy
D. Early evidence of hypertrophic cardiomyopathy
E. Myocarditis

17. A previously healthy 8-year-old boy cannot walk today because of right ankle pain. His left knee was sore and swollen 3 days ago. Findings include: temperature, 39°C; blood pressure, 102/64 mm Hg; pulse, 112 beats/min; grade 3/6 holosystolic murmur at the apex radiating to the left axilla; and swelling, warmth, and tenderness of the right ankle.

Of the following, the laboratory test that is MOST likely to be associated with these findings is:

A. Blood culture growing *Streptococcus viridans*
B. Elevated antistreptolysin O (ASO) titer
C. Positive serum antinuclear antibody (ANA) titer
D. Positive serum rheumatoid factor
E. Urinalysis showing microscopic hematuria

18. Following an uncomplicated delivery, a 3.7 kg term infant develops cyanosis in the first hour of life. Findings at 3 hours of age include: cyanosis, heart rate 140 bpm, respiratory rate 45 breaths/min, no heart murmurs, pulse oximetry in room air, 70% in the right hand, 85% in the foot; in 100% FIO₂ via head hood O₂ sat increases to 90% in the foot; Normal chest x-ray.

These findings are MOST consistent with:

A. Primary pulmonary hypertension of the newborn
B. Pulmonary valve atresia
C. Transient tachypnea of the newborn
D. Transposition of the great arteries
E. Truncus arteriosus
19. A three-week-old infant presents with tachypnea, tachycardia, hepatomegaly, and poor feeding for 4 days. 

_The LEAST likely diagnosis is:_

A. Ventricular septal defect 
B. Tetralogy of Fallot 
C. Coarctation of the aorta 
D. Aortic stenosis 
E. Congestive cardiomyopathy

20. A 5-month-old girl is being evaluated for her first episode of wheezing. She feeds slowly and sweats during feedings. Findings include: weight, 5th percentile; length, 50th percentile; soft systolic ejection murmurs at the left upper sternal border and in the left posterior thorax; liver edge, 3cm below the right costal margin; and diffuse expiratory wheezing.

_The most appropriate INITIAL step is:_

A. Administration of nebulized albuterol and observation for clinical response. 
B. Chest radiography to exclude foreign body. 
C. Echocardiography. 
D. Palpation of pulses and measurement of blood pressures in all 4 extremities. 
E. Sweat chloride testing for cystic fibrosis.

21. A 1-day-old term infant has dysmorphic features that include a low nasal bridge, flat occiput, clinodactyly of the fifth digits, wide spacing between the first and second toes, and hypotonia. He is cyanotic, and results of lung and abdominal examinations are normal. There is no heart murmur and pulses are normal.

_The likelihood that this infant will have clinically significant congenital heart disease is CLOSEST to:_

A. less than 20% 
B. 20% to 35% 
C. 40% to 55% 
D. 60% to 70% 
E. 80% to 95%
22. A term infant is cyanotic and requires intubation.
Findings include: heart rate, 175 bpm; blood pressure, 60/30 mm Hg; increased right ventricular activity; single S₂; short systolic murmur; and equal arm and leg pulses; chest x-ray: normal heart size and pulmonary congestion. Arterial blood gases (right radial artery in 100% FIO₂): pH 7.31; pO₂ 43 Torr; pCO₂ 48 Torr.

Of the following, the MOST likely diagnosis is:

A. Hyaline membrane disease  
B. Hypoplastic left heart  
C. Intrauterine constriction of the ductus arteriosus  
D. Tetralogy of Fallot  
E. Total anomalous pulmonary venous connection

23. The most common cause of syncope in young children is:

A. Autonomic dysfunction associated with prolonged standing  
B. Breath-holding spells  
C. Cataplexy  
D. Panic attack associated with hyperventilation  
E. Prolonged QT syndrome

24. Regarding infective endocarditis:

A. Clinical diagnosis is made utilizing the Duke Criteria  
B. Echocardiography is the definitive diagnostic modality  
C. Atrial septal defect is a predisposing heart condition  
D. Most cases are “culture-negative”  
E. Systolic ejection murmur indicates valvar regurgitation
25. Which of the following organisms is least likely to cause infective endocarditis:

A. Staphylococcus epidermidis
B. Streptococcus viridans
C. Haemophilus influenzae
D. Staphylococcus aureus
E. Streptococcus Faecalis (Enterococcus)

26. Which of the following signs is most likely to be present in children with infective endocarditis?

A. Osler nodes
B. Fever
C. Systolic murmur
D. Splenomegaly
E. Splinter hemorrhages

27. A 13 year old girl loses consciousness while standing in line at a roller coaster. She regains consciousness spontaneously within one minute.

The most likely cause for her syncopal event is:

A. Temporal lobe epilepsy
B. Long QT syndrome
C. Neurocardiogenic syncope
D. Hypertrophic cardiomyopathy
E. Shy-Drager syndrome
28) The physical finding most suggestive of hypertrophic cardiomyopathy in an adolescent athlete is:

A. Bilateral ptosis
B. Single second heart sound
C. Diminished pulse in the right arm
D. Continuous murmur heard in the left back
E. Systolic murmur accentuated by standing

29) Which of the following findings may be normal in a newborn?

A. Widely split, fixed second heart sound
B. Precordial thrill
C. Systolic ejection murmur
D. Lower O₂ saturation in the right arm than in the right leg
E. Systolic ejection click

30. Two weeks after undergoing surgical closure of an atrial septal defect, a 2 year old boy presents with several days of irritability and anorexia. On physical exam, he is pale and mildly tachypneic. His pulses are weak and irregular. The auscultated heart rate is regular at 160 bpm.

The intervention MOST likely to be effective is:

A. Oral ibuprofen 10 mg/kg q 6 hrs.
B. Intravenous adenosine
C. Pericardiocentesis
D. Synchronized direct current cardioversion
E. Transfusion of packed red blood cells, 10 ml/kg
31. During a sports pre-participation evaluation, you obtain the information that an older sister has a seizure disorder and a younger brother has a history of syncope.

Before approving this patient for sports you MUST perform which study?

A. Echocardiogram  
B. Electrocardiogram  
C. Electroencephalogram  
D. Genetic testing for ion channel abnormalities  
E. Cardiac catheterization

32. At a routine one month evaluation you hear a 3/6 harsh high pitched holosystolic murmur at the mid left sternal border.

The MOST likely diagnosis is:

A. Ventricular septal defect  
B. Atrial septal defect  
C. Aortic stenosis  
D. Pulmonary stenosis  
E. Tetralogy of Fallot

33. A 6 day old infant is brought to the Emergency Department with a two day history of poor feeding and lethargy. Pregnancy, labor and delivery were uncomplicated. He was discharged at 48 hours of age. On physical exam, the HR is 190 bpm, RR is 80/min and BP is 50/30. He is pale and mottled with cool extremities and weak pulses.

The MOST likely diagnosis is:

A. Ventricular septal defect  
B. Atrial septal defect  
C. Hypoplastic left heart syndrome  
D. Double inlet left ventricle  
E. Double aortic arch
34. A 19 year old college student with a history of a small VSD presents to the Emergency Department with a 4 hour history of sharp right sided chest pain. The pain is worse with inspiration. He reports that over the past month he has had night sweats and has lost 10 lbs.

The MOST appropriate initial response is:

A. Transesophageal echocardiography
B. Administration of intravenous Ceftriaxone
C. Transthoracic echocardiography
D. Tuberculin skin test
E. Blood cultures

35. Two weeks after surgical repair of an atrial septal defect, a 2 year old boy presents with anorexia and irritability. On physical examination, he is pale and his pulses are difficult to palpate. His heart rate is 160 bpm but his radial pulse is difficult to palpate during inspiration.

Of the following the intervention most likely to be beneficial is

A. Intravenous adenosine
B. Thoracentesis
C. Pericardiocentesis
D. Intravenous normal saline bolus
E. D/C cardioversion

36. Regarding the proposed use of pulse oximetry screening for critical congenital heart disease, the AAP recommends:

A. Screening should be performed within 12 hours of age
B. Screening should be performed by the family
C. Oxygen saturation should be obtained in both hands and one foot
D. Pulse oximetry reading with a ≤5% absolute difference between the upper and lower extremity is considered a pass
E. In the event of a positive screening result, CCHD needs to be excluded with a diagnostic echocardiogram.
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