Chapter 5
Cardiology

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Questions

1. Paroxysmal supraventricular tachycardia:
   1. Can cause low output congestive heart failure
   2. Can be treated with IV adenosine
   3. Can be prevented with PO digoxin
   4. Should be initially treated with verapamil
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

2. The ECG of a newborn or infant:
   1. Will show increased right ventricular forces
   2. Will show prominence R wave in V1
   3. Will show inverted T waves in lead V1
   4. Will show right bundle branch block pattern until 5 years of age
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

3. The most common cardiac lesions, exclusive of PDAs in premies, are (choose 2):
   A. ASD
   B. Tetralogy of Fallot (TOF)
   C. Hypoplastic left heart syndrome
   D. VSD
   E. Coarctation of the aorta (CoA)

4. A relatively large VSD:
   1. Will often present with dyspnea, feeding difficulty, and poor growth
   2. Presents at 1–3 months of age as pulmonary vascular resistance (PVR) decreases
   3. Will cause a Qp:Qs >2:1
   4. Will present with a harsh holosystolic murmur and a loud P2
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

Answers

1. A. 1, 2, 3

SVT is a tachydysrhythmia that originates from above the bifurcation of the bundle of His. The onset is usually paroxysmal, hence paroxysmal supraventricular tachycardia (PSVT). In children, the rate is generally >230 beats/min, and in infants the rate can exceed 300 beats/min. The QRS is narrow, differing little from the QRS seen during normal sinus rhythm. Predisposing factors for the development of PSVT are the preexcitation syndromes such as Wolf-Parkinson-White and congenital heart disease and sympathomimetic medications such as atropine or glycopyrrolate.

2. A. 1, 2, 3

At birth, the RV and LV walls are of approximately equal thickness, thus the infant has relative RV hypertrophy with prominence of right and anterior forces.

3. A. ASD and D. VSD

Atrial septal defects (ASDs) come in four varieties: patent foramen ovale (PFO), secundum (at the fossa ovalis) ASDs, coronary sinus defects (absence of wall separating the coronary sinus and LA), and sinus venosus defects (immediately below the SVC opening). Sinus venosus defects are associated with partial anomalous pulmonary venous return.

4. E. All of the above

VSD is the most common congenital cardiac malformation in children with an incidence of approximately 2–3:1,000 live births. VSDs are often not apparent in the newborn because the relatively high pulmonary vascular resistance limits the right to left flow through the defect. Congestive heart failure (CHF) becomes clinically apparent as the infant grows and PVR decreases. The severity of the shunt is characterized by the Qp:Qs ratio (pulmonary to systemic blood flow). With a Qp:Qs >2, signs and symptoms of CHF are seen. CHF in infants and newborns presents with poor feeding, diaphoresis with feeding, effortless tachypnea, lethargy, and FTT. The typical murmur of a VSD is holosystolic, harsh, and best heard along the left sternal border.

5. Regarding ventricular septal defects (VSD's):
   1. They occur with an overall incidence of 3–4/1000 live births.
   2. They undergo spontaneous closure in approximately 25 % of cases.
   3. Complications of repair are rare and include acquired complete heart block.
   4. Most VSD's occur in association with other congenital anomalies.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

6. Regarding ASD's:
   1. They can be divided into primum, secundum, and sinus venosus based on location and etiology.
   2. Secundum ASDs are the most common type.
   3. Secundum ASDs often are asymptomatic in childhood.
   4. There may be associated partial anomalous venous return with an ASD.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

7. Which of the following organisms are associated with sepsis occurring after cardiac surgery?
   A. *Haemophilus influenzae* type b
   B. *Neisseria gonorrhoeae*
   C. *Staphylococci*
   D. Enteric gram-negative rods

8. The so-called innocent murmur of childhood:
   1. May be heard in up to 30 % of children at some point in their lives
   2. Is best heard in a localized area along the left lower sternal border
   3. Is short, vibratory ejection-type murmur
   4. Is generally heard in children between the ages of 3 and 7 years
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

5. A. 1, 2, 3

VSDs are classified by location in the septum. Membranous or perimembranous defects are the most common. These are located below the aortic valve and adjacent to the tricuspid valve. Other types of VSD include AV canal type, subpulmonary, conoventricular, and muscular VSDs. Many VSDs undergo spontaneous closure with larger defects less likely to do so. Currently, repair is undertaken in infancy. The risk of heart block in the postoperative period is related to the size and location of the patch used to repair the defect.

6. E. All of the above

Isolated secundum ASDs, which represent 80 % of all ASDs, generally do not present in infancy. Children with this defect generally are in sinus rhythm. Primum ASDs are often associated with a cleft mitral valve. Sinus venosus and coronary sinus ASDs are actually defects in the embryologic sinus venosus. The murmur noted in children with relatively large ASDs is a pulmonary flow murmur with associated fixed splitting of S2. Patients with unrepaired ASDs often do relatively well into their twenties when progressive cyanosis and dyspnea develop.

7. C. Staphylococci

8. E. All of the above

The history is unremarkable in these children since they have no cardiac disease. Murmurs are more frequently heard in children during febrile episodes. The cause of the murmur is unknown. Some speculate that it is heard only in childhood because the relatively thin chest wall of young children transmits extra-cardiac sounds more easily.

9. Congestive heart failure in childhood may present:
   1. Chronic cough
   2. As failure to thrive
   3. With respiratory distress during feedings
   4. With the child's complaint that it is difficult to keep up with peers during play
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

10. Tetralogy of Fallot:
    1. Is the most common cyanotic congenital cardiac lesion presenting after 2 weeks of age
    2. Includes pulmonary stenosis, VSD, overriding aorta, and RVH
    3. May have infundibular and valvar pulmonary stenosis
    4. Is inherited as an autosomal dominant
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

11. The so-called tet spells:
    1. Occur in infants with unrepaired tetralogy of Fallot
    2. Are the result of increased tone in the infundibulum of the RV outflow tract
    3. Result in intense cyanosis and diminution of the systolic ejection murmur as pulmonary blood flow dramatically decreases
    4. Can be treated with IV fluid administration and/or increasing SVR
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

12. The most common congenital cardiac defect presenting with cyanosis in the newborn is:
    A. Transposition of the great arteries (TGA)
    B. Patent ductus arteriosus (PDA)
    C. Hypoplastic left heart syndrome (HLHS)
    D. Tetralogy of Fallot (TOF)
    E. Truncus arteriosCus

9. E. All of the above

CHF presents in infancy with tachycardia, tachypnea, poor feeding, and failure to thrive. In older children, decreased exercise tolerance is noted, as in adults. Recurrent respiratory infections are common, although rales are not heard until later in the course of CHF.

10. A. 1, 2, 3

TOF is found in approximately 6–10 % of infants with cyanotic congenital heart disease. TOF has no known inheritance pattern, but it is found in association with a number of syndromes such as Goldenhar (oculo-auriculo-vertebral hypoplasia), VACTERL (vertebral anomalies, esophageal atresia with tracheoesophageal fistula, radial dysplasia, renal anomalies, imperforate anus, cardiac defects), CHARGE association (choanal atresia, heart defects, deafness, genital hypoplasia in males, coloboma), and Klippel-Feil syndrome (short neck, limited neck motion, low occipital hairline).

11. E. All of the above

With repair of TOF now routinely performed in the neonate and infant, "tet spells" are rare.

12. A. Transposition of the great arteries (TGA)

Without mixing of the two parallel circulations, the newborn with TGA cannot survive.

The presentation of TGA is affected by the presence of other anomalies such as a VSD, left ventricular outflow tract obstruction (LVOTO), or sub-pulmonic stenosis. Newborns with intact ventricular septa rely on the presence of a PFO or PDA to mix oxygenated and deoxygenated blood.

13. A newborn with isolated TGA may present with:
    1. Cyanosis
    2. Tachypnea without dyspnea or respiratory distress
    3. Normal peripheral pulses
    4. An ECG showing right ventricular hypertrophy, indistinguishable from that of a newborn with a normal heart
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

14. Which of the following may be done to newborns with TGA to improve arterial SpO_2?
    1. A Rashkind-Miller procedure
    2. Administration of milrinone
    3. Prostaglandin E1 administration
    4. Dilation of the ductus arteriosus in the catherization lab
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

15. Regarding the ductus arteriosus:
    1. More than 90 % of fetal RV output passes through the ductus.
    2. If it remains open postnatally, it has flow through it both during systole and diastole.
    3. It closes functionally during the first day of life in most term infants.
    4. Spontaneous closure of a persistently open PDA is unlikely after the age of 6 months.
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

16. Endocardial cushions contribute to which of the following cardiac structures?
    1. The lower part of the atrial septum
    2. The part of the ventricular septum where the AV valves insert
    3. Tissue that forms part of the mitral and tricuspid valves
    4. Part of the intraventricular conduction system
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

13. E. All of the above

In newborns with TGA and IVS (intact ventricular septum), there often is no murmur present. A CXR may be normal, but approximately 30 % shows an "egg on a string" pattern of the cardiothymic shadow.

14. B. 1, 3

Maneuvers that enhance mixing of the parallel circulations improve the situation in newborns with TGA. Prostaglandins keep the PDA open, and the Rashkind procedure involves creation of an atrial septostomy using a specially designed catheter.

15. E. All of the above

In full-term infants, persistent patent ductus arteriosus accounts for approximately 10 % of congenital heart disease. PDAs are much more common in the premature newborn (see question/answer 18 and 19 in newborn medicine). Commonly, the PDA is picked up when a murmur is heard in an asymptomatic child who is being examined for another reason. The typical murmur is continuous (machine-like) and heard best in the midclavicular line between the first and second interspace.

16. A. 1, 2, 3

AV canal defects, also called endocardial cushion defects, involve a primum ASD, defects in one or more of the AV valves, and also a defect in the ventricular septum.

17. Common atrioventricular canal defects (CAVC):
    1. Result in communication between all four cardiac chambers
    2. Have abnormal mitral and/or tricuspid valves
    3. Often present in a manner similar to large VSDs
    4. Predispose the child to the early development of pulmonary vascular obstructive disease (PVOD)
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

18. In total anomalous pulmonary venous return (TAPVR):
    1. It is often divided into several types based upon the site of pulmonary venous drainage.
    2. All the pulmonary veins drain into the systemic venous system, not the left atrium.
    3. There often is pulmonary venous obstruction.
    4. There is an ASD or PFO allowing a right to left shunt to compensate for the left to right shunt resulting from the TAPVR.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

19. Regarding the locations of the pulmonary venous drainage in TAPVR:
    1. Supracardiac, the most common, involves drainage into an anomalous vein which eventually empties into the SVC.
    2. With intracardiac TAPVR, venous drainage is directly into the RA or coronary sinus.
    3. With infracardiac TAPVR, a vein passes through the diaphragm.
    4. Mixed TAPVR, a combination of the other types, is the least common.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

17. E. All of the above

AV canal defects have variable anatomy. The atrial and ventricular septa and AV valves are affected in many different ways. Down syndrome is frequently associated with CAVC defects. Two-dimensional echo demonstrates absence of the atrial septum, while pulsed Doppler echo demonstrates the presence of AV valve regurgitation.

18. E. All of the above

TAPVR is classified into four types. In decreasing order of frequency, they are supracardiac, cardiac, infracardiac, and mixed. Obstruction of the anomalous venous drainage leading to pulmonary congestion may occur at any point along the anomalous venous pathway. Obstruction almost always occurs in the infracardiac type.

19. E. All of the above

The presence and degree of venous obstruction and the degree of intra-atrial mixing determine the severity of clinical symptoms. Infants with obstruction in the anomalous venous connections develop cyanosis and respiratory distress early in life. Infants without obstruction and a nonrestrictive inter-atrial communication may have only minimal symptoms during the first year of life.

20. Coarctation of the aorta (CoA):
    1. Is a congenital narrowing of the aorta near the insertion of the ductus arteriosus
    2. Is commonly associated with VSD or hypoplastic left heart syndrome (HLHS)
    3. May cause reduced lower body perfusion
    4. Produces aortic insufficiency
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

21. Newborns and infants presenting with coarctation of the aorta:
    1. Generally have more severe coarctation
    2. May have metabolic acidosis as a result of poor lower body perfusion
    3. May present with signs of LV failure
    4. Have an extensive network of collaterals
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

22. Children with coarctation of the aorta:
    1. Have had time to develop collateral flow through intercostal and other arteries
    2. Usually require cardiopulmonary bypass for surgical repair
    3. May have systemic hypertension
    4. Are often managed medically until adulthood
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

23. The hypoplastic left heart syndrome can include:
    1. Hypoplasia of the LV and RA
    2. Mitral atresia
    3. Coronary, carotid, and subclavian flow via retrograde filling of a small ascending aorta from the ductus
    4. RV hypertrophy
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

20. A. 1, 2, 3

The aortic narrowing seen in CoA is located in the descending thoracic aorta just across from the insertion of the ductus arteriosus. This anomaly has a 2:1 male predominance. The major problem associated with CoA is increased LV afterload. For CoA to become clinically significant, the aortic diameter must be decreased by at least 50 %. Pulses and measured blood pressure in the lower extremities are diminished compared to the upper extremities.

21. E. All of the above

Newborns with CoA may appear well initially, but cardiac failure and respiratory distress quickly develop as the ductus closes. Prostaglandin administration may improve the situation as the dilated ductus allows improved lower extremity and renal perfusion.

22. B. 1, 3

Children with isolated CoA often have no specific complaints. With a careful history, the child may report leg cramps. The coarctation may be discovered during an evaluation of systemic hypertension. The ECG may show no changes or LVH by voltage criteria may be seen. The pathognomonic CXR finding of rib notching, due to rib erosion by the enlarged collateral vessels, is rarely seen in children younger than 5–6 years of age.

23. A. 1, 2, 3

HLHS is seen in 3–4:10,000 live births. There is a spectrum of anomalies in this left-sided obstructive lesion. The LV and ascending aorta are underdeveloped. The mitral valve is often involved, exhibiting stenosis, hypoplasia, or atresia. The RV provides both pulmonary and systemic flow in HLHS. There is a L to R shunting of pulmonary venous return at the atrial level and a R to L shunting of RV output at the PDA, with the ascending aorta and its vessels (carotids, subclavian, and coronaries) perfused retrograde via flow from the PDA.

24. What happens to newborns with HLHS when the PDA closes?
    1. There is improved systemic blood pressure.
    2. There is reduced coronary flow.
    3. There is increased systemic flow.
    4. There is decreased systemic flow.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

25. In HLHS, systemic flow is affected by:
    1. SVR
    2. PaO_2
    3. PVR
    4. pH
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

26. Which of the following CHD diagnoses is matched with the past or present appropriate surgical procedure?
    1. TOF-BT shunt
    2. TGA switch
    3. HLHS Stage I – Glenn-Fontan
    4. VSD-PA band
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

24. C. 2, 4

The PDA allows the RV output to flow to the pulmonary circuit and the systemic circuit and retrograde into the ascending aorta. Once the diagnosis of HLHS is made, prostaglandin should be infused to keep the PDA open and the newborn monitored for the development of metabolic acidosis.

25. B. 1, 3

The ratio of systemic to pulmonary vascular resistances is crucial in determining flows into these vascular beds. Excessive pulmonary flow will lead to underperfusion of the body. Hyperventilation and oxygen administration may increase $Sp0_2$ but lead to systemic hypoperfusion and the development of metabolic acidosis. Chromosomal abnormalities have been reported in 11 % of infants with HLHS, and autopsies have revealed neurologic abnormalities in 29 % of these patients.

26. E. All of the above

Palliation of TOF is no longer done as a routine, but if done, the goal is to achieve an increase in pulmonary blood flow. A Blalock-Taussig shunt (B-T shunt) diverts subclavian artery flow to the pulmonary circulation, either using a Gortex graft or by an end-to-side anastomosis of the subclavian artery to the PA.

Transposition of the great arteries (TGA) is treated surgically with a so-called switch operation in which the PA and aorta are moved to the appropriate ventricular outflow tract. It is very important to know the coronary arterial anatomy beforehand. The coronaries are removed from the aortic root along with a small area of surrounding tissue and moved to the newly "switched" aorta.

Hypoplastic left heart syndrome accounts for 1 % of all CHD. There is a small LV, mitral valve, aortic valve, and aortic arch. A stage I procedure is done in the newborn period. Systemic flow is carried by the PDA, and coronary flow is retrograde in the small aortic arch. The stage I procedure involves creation of a neoaorta from the hypoplastic aortic arch, main PA, and homograft. A large ASD is created and pulmonary blood flow is via a modified (graft material) B-T shunt.

Ventricular septal defects often become clinically apparent in the third month of life, when PVR decreases substantially and the higher left ventricular pressures divert more and more blood to the lower pressure right ventricle, leading to CHF. The degree of shunt is characterized by the ratio of systemic to pulmonary flow (Qp:Qs).

27. The Fontan operation, also called total cavopulmonary connection:
    1. Directs systemic venous return to the PA
    2. Is the surgical procedure for many patients with single ventricle physiology
    3. Is generally performed at 1–2 years of age
    4. Is generally preceded by a bi-directional Glenn procedure

    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

A 2:1 shunt or greater is associated with CHF. Currently, most VSDs are repaired primarily in the OR or with a device in the cardiac catheterization lab. A PA band was used previously as a temporary means to increase resistance to pulmonary flow, thus decreasing the shunt.

27. E. All of the above

The Fontan procedure involves a so-called passive flow of systemic venous return into the pulmonary circuit. This requires a transpulmonary gradient of 3–8 mmHg. This can be achieved if the CVP (PA) pressure is kept at 12–15 mmHg with an LVEDP of 5–10 mmHg. In addition the cardiac rhythm must be kept in the sinus, and ventricular performance must often be supported pharmacologically.