CLINIQUIZ

Congenital heart defects

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Case Scenario: 1
A full term born, 6 month old child presented in outpatient department with chief complaints of respiratory distress, poor weight gain and recurrent chest infections since birth. On general examination, PR was 152/min, BP-80/48 mm Hg. Cardiac examination revealed an ejection systolic murmur at left lower sternal border, S2 split with accentuation of pulmonary component and apical diastolic rumble. ECG showed features of left atrial enlargement and left ventricular hypertrophy. Chest x-ray showed cardiomegaly and increased pulmonary vascular marking. Echocardiography revealed 12 mm ventricular septal defect with significant left to right shunt, aortic cusp prolapse and mild aortic insufficiency.

Q 1: What is the most appropriate line of treatment in this patient?
   a. Medical Management
   b. Surgical Treatment
   c. Percutaneous Device closure
   d. All of the above

Q 2: What is the cause of apical rumble in this patient?
   a. Increased flow through mitral valve
   b. Left ventricular dysfunction
   c. Increased pulmonary flow and pulmonary venous return
   d. None of the above

Q 3: VSD closure can be safely undertaken in all of the following conditions except
   a. Pulmonary vascular resistance less than 6 Wood units/m²
   b. Qp/Qs >1.5:1
   c. Evidence of pulmonary reactivity with pulmonary vasodilators
   d. Eisenmenger syndrome

Q 4: All of the following statements regarding VSD patients are true except
   a. Non-restrictive VSD leads to pulmonary hypertension and left ventricular dysfunction
   b. Paradoxical embolization by clot or air is a great risk in patients with intracardiac shunts.
   c. High FiO2 and low PaCO2 is optimal for VSD patients in the intraoperative period.
   d. Complete Heart block is a devastating postoperative complication of VSD closure.

Case Scenario: 2
A 3 month old child was brought to emergency department with history of severe bluish discoloration while crying followed by collapse. The mother gave a past history of such episodes with spontaneous recovery since birth. On physical examination, patient showed central cyanosis, PR was 155/min, respiratory rate was 50/min, BP-66/38 mmHg, SPO2- 55%. On auscultation, systolic ejection murmur was found at left upper sternal border. Lab investigation were within normal limits except elevated hematocrit. ECG showed right axis deviation and right ventricular hypertrophy. Chest X-ray demonstrated boot-shaped heart and pulmonary oligemia. Echocardiography revealed right ventricular outflow obstruction with RVH, non-restrictive ventricular septal defect with right to left shunt and infundibular pulmonary stenosis.

Q 5: The diagnosis of this patient is
   a. Transposition of great arteries
   b. Truncus arteriosus
   c. Tetralogy of Fallot
   d. Total anomalous pulmonary venous connection

Q 6: This patient should be immediately treated with all of the following except
   a. Intravenous Beta-blocker
   b. Intravenous Sodium bicarbonate
   c. Intravenous Phenylephrine
   d. Intravenous Digoxin

Q 7: Ejection systolic murmur is due to
   a. Right ventricular outflow obstruction
   b. Non-restrictive VSD
   c. Pulmonary stenosis
   d. Aortopulmonary collateral vessel

Q 8: Perioperative management of this patient include all of the following except
   a. Volume loading is useful
   b. Maintain or increase SVR relative to PVR
   c. Ketamine and Pancuronium must always be used for induction.
   d. Postoperative analgesic requirement should be adequately met.

Q 9: Which of the following statement is correct regarding this cyanotic congenital heart defect?
   a. Diagnostic catheterization is needed to delineate
coronary and pulmonary artery anatomy.

b. Blalock-Taussig Shunt is a common palliative surgery done for this heart defect.

c. High risk of cerebral and renal thrombosis due to polycythemia

d. All of the above

Q 10: Which of the following statement is incorrect regarding this defect?
a. Down’s syndrome is most commonly associated with this defect.
b. Reoperation is necessary in 10-15% of patients after reparative surgery in long term followup.
c. Junctional ectopic tachycardia is common in early postoperative period.
d. Risk factors for sudden death after surgical repair includes RV dilation and QRS >180msec.

ANSWERS:
Ans. 1(b): Patient with defect size > 6.5mm almost always require surgical closure. Failure to thrive with signs of left ventricular failure (i.e. fatigue with feeding) and significant left to right shunt leading to recurrent chest infections are also indications for surgical closure. Medical therapy in moderate to large defects is usually started to improve symptoms before surgery. Percutaneous device closure is not done in cases of associated anomalies such as aortic valve prolapse and insufficiency which commonly occur in supracristal VSD, due to long term risk of endocarditis.

Ans. 2(a): Apical rumble reflects increased flow through mitral valve due to left to right shunting.

Ans. 3(d): Eisenmenger syndrome is an end result of chronic, large left to right shunt. It is a consequence of the pathologic changes caused by chronically increased pulmonary artery pressure leading to increased pulmonary vascular resistance (PVR) so that the shunt becomes bidirectional or reversed. Correction of underlying cardiac disorder is contraindicated once eisenmenger syndrome is established.

Ans. 4(c): Anesthetic management of a patient with ventricular septal defect is tailored to minimize the excessive reduction in PVR and increase in systemic vascular resistance, in order to reduce the magnitude of shunt. Oxygen and low Paco2, both are potent pulmonary vasodilator and therefore, not optimal for defects with high pulmonary flow.

Ans. 5(c): Tetralogy of Fallot is a cyanotic congenital heart disease characterized by ventricular septal defect with right to left shunt, right ventricular hypertrophy, infundibular pulmonary stenosis and overriding to aorta. Chest X-rays shows classic boot-shaped heart or “coeur en sabot” appearance.

Ans. 6(d): Patient is having an episode of hypercyanotic “Spell” characterized by paroxysmal cyanosis and hyperpnea which can be initiated by crying, feeding or defecation. These events causes increase in oxygen demand and hypoxemia which results in decrease in SVR. These episodes usually resolve spontaneously but can be progressive and fatal in rare cases. Episodes can be terminated by intravenous beta-blocker. Inj. Phenytoine increases SVR, while Inj. Sodium bicarbonate corrects peripheral metabolic acidosis with return to normal SVR.

Ans. 7(a): The murmur is due to RVOT obstruction and length and volume of the murmur vary inversely with the degree of obstruction to antegrade pulmonary blood flow. In patients with TOF, VSD is non-restrictive and does not produce a murmur. Aortopulmonary collateral vessels produces continuous murmur best heard at the back.

Ans. 8(c): Patients with history of hypercyanotic spells are particularly vulnerable during induction and emergence of anesthesia. Endogenous catecholamines induces infundibular muscle spasm and can precipitate spell. There is a increased need of preoperative sedation and postoperative analgesia to minimize catecholamine release. Although, anesthetic agents which increase sympathetic discharge such as ketamine and pancuronium are often used for induction of cyanotic patients, should be used with caution in patients with frequent episodes of hypercyanotic spells.

Ans. 9(d): Diagnostic catheterization in TOF is required in cases of associated lesion like anomalous origin of left anterior descending artery from right coronary artery and to delineate pulmonary blood supply and presence of collateral vessels. Blalock-Taussig shunt is palliative procedure for intensely cyanotic TOF patients and involves direct anastomosis of subclavian artery with a branch of ipsilateral pulmonary artery. It helps in increasing pulmonary artery size and improved haemodynamics following complete repair. Chronic hypoxia leads to polycythemia in TOF patients which can cause renal, cerebral and pulmonary thrombosis.

Ans. 10(a): Tetralogy of fallot is most commonly associated with Digeorge Syndrome and chromosome 22q11 deletion. Down’s syndrome is most commonly associated with atrioventricular canal defect.

REFERENCES