

COMMON CARDIAC CONDITIONS

Introduction

The incidence of congenital heart disease is 6-8:1000. It can exist in isolation or be associated with chromosomal abnormalities or syndromes.

Congenital heart disease can be classified as follows:

1. 'Simple' left to right shunt: increased pulmonary blood flow
 - Patent Ductus Arteriosus (PDA)
 - Atrial Septal Defect (ASD)
 - Ventricular Septal Defect (VSD)
 - Atrioventricular Septal Defect (AVSD)
2. 'Simple' right to left shunt: results in cyanosis
 - Tetralogy of Fallot (TOF)
 - Pulmonary atresia
 - Tricuspid atresia
 - Ebstein's anomaly
3. Complex shunts: mixing of pulmonary and systemic blood flow with cyanosis
 - Transposition of Great Arteries
 - Total anomalous pulmonary venous drainage (TAPVD)
 - Hypoplastic left heart syndrome (HLHS)
 - Double outlet right ventricle (DORV)
 - Truncus arteriosus
4. Obstructive lesions
 - Coarctation of Aorta
 - Interrupted or hypoplastic aortic arch
 - Aortic Stenosis
 - Mitral Stenosis
 - Tricuspid Stenosis

5. Regurgitant lesions

Surgery for congenital heart disease can be

- a. Corrective (eg. PDA ligation/ ASD closure/ VSD closure) or
- b. Palliative (eg. Pulmonary artery (PA) banding/ Blalock- Taussig (BT) shunts/ cavo-pulmonary shunts)

Surgery can be either

- a. Open (on cardiopulmonary bypass (CPB) or
- b. Closed (non-CPB)
 - ligation of PDA (thoracotomy)
 - Repair of coarctation of aorta (thoracotomy)
 - PA banding (thoracotomy or sternotomy)
 - BT shunt (thoracotomy or sternotomy)

Considerations for anaesthetic plan include:

1. The cardiac lesion:
 - the predominant one on the basis of pathophysiology
 - myocardial reserve /functional capacity (feeding, sweating, grunting, recurrent chest infections, failure to thrive, exercise tolerance, Hb, SpO₂)/ rhythm/ complications (neurological, heart failure, tet spells)
 - nature of shunt /obstruction
 - impact on pulmonary and systemic pulmonary vascular resistance (PVR/SVR)
2. Planned surgery
3. Management of CPB
4. Other patient factors
 - age
 - associated lesions /syndromes
 - respiratory tract infection
 - vascular access
 - airway and dental status
 - medications
 - previous surgery

Investigations and results

- FBC
- U/E/Cr
- PT/PTT
- CXR
- ECG
- 2D echo report /cardiac catheterisation report

Pathophysiology

The pathophysiology depends on the nature and size of the cardiac lesion. Where more than one cardiac lesion exist, usually one lesion will predominate.

Other factors that can impact on the overall behaviour of the cardiac lesion include:

1. Transition circulation:
 - This in the first few days to weeks of life.
 - The pulmonary circulation is very reactive and potential reopening of the ductus arteriosus can occur during this period. The circulation is extremely sensitive to hypoxia, hypercarbia, acidosis and prostaglandins at this stage. Shunting or reversion to fetal circulation can occur.
2. Presence of a duct-dependent circulation. This could be either:
 - duct-dependent systemic circulation (eg critical aortic stenosis, hypoplastic left heart syndrome, coarctation of the aorta). It can present with collapse or cardiac failure.
 - duct-dependent pulmonary circulation will present with increasing cyanosis unresponsive to increasing oxygen concentrations. The duct must be kept open until further surgical management is possible by avoiding high FiO_2 , allowing moderate hypercarbia and administering PGE1 via an infusion. Cardiac output may have to be supported with fluids and inotropes.
3. Presence of non-restrictive/ balanced shunts
 - Balancing the systemic and pulmonary circulation is important to avoid cyanosis, cardiac failure or systemic hypotension.

Determine if the following are present:

1. Cyanosis

This results in chronic hypoxia, compensatory polycythemia with increased risk of thromboembolic phenomena, coagulopathy and metabolic acidosis. The severity is indicated by the baseline saturations in room air and polycythemia. These children benefit from hydration in the perioperative period and it is useful to order an intravenous drip for the duration in which they are kept nil by mouth. In selected cases, preoperative oxygen therapy, correction of acidosis and inotropic support may be required.

2. Reduced pulmonary blood flow

Oxygen therapy, hyperventilation and avoiding a decrease in the systemic vascular resistance aid in reducing pulmonary vascular resistance and promoting blood flow to the lungs. Selective agents to reduce PVR eg nitric oxide may be beneficial especially in those infants with right ventricular hypertrophy.

3. Increased pulmonary blood flow and cardiac failure /duct- dependent systemic circulation /high pressure or volume shunts.

These children benefit from moderate hypercarbia, room air or avoiding high FiO_2 and inotropes in some instances. Marked falls in SVR should be avoided.