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Truncus Arteriosus

10.1 Morphology & Pathophysiology

During embryological differentiation of cono-truncus, arrest in septation fails to separate truncal artery into the aorta and pulmonary arteries. A single truncal vessel straddles over a large outlet ventricular septal defect. This single great vessel gives rise both the ascending aorta and pulmonary artery. Truncus is classified based on the origin of pulmonary arteries from the truncal artery:

Type I: The main pulmonary artery arises from the truncal artery.

Type II: The right and left branch pulmonary arteries arise separately but in close proximity.

Type III: The right and left branch pulmonary arteries may arise widely separately from the lateral aspects of the truncal artery.

Truncal valve: It may be abnormal. The valve has usually three cusps and severe valve incompetence is rare. It is important to evaluate the truncal valve function preoperatively since significant stenosis or insufficiency of the truncal valve will require intervention at the time of surgery.

The VSD in truncus arteriosus is large and non-restrictive and results in left to right shunting of blood with pulmonary overcirculation.

Congestive heart failure and pulmonary artery hypertension are common in truncus. Pulmonary vascular obstructive disease will occur early if no intervention is undertaken.

10.2 Surgical Treatment

Surgery for total repair is performed on cardiopulmonary bypass, aortic cross-clamping, and cardioplegic arrest. The pulmonary artery is separated from the truncal root, and the truncal vessel is closed directly or with a patch of

synthetic material or a cryopreserved homograft. Truncal valve repair, replacement, or valvotomy is performed as needed. An incision is made in the right ventricular outflow tract, and the ventricular septal defect is closed. A valved homograft conduit is implanted, connecting the right ventricle to the pulmonary arteries. Transesophageal echocardiography is often utilized to evaluate the adequacy of valve repair following weaning from cardiopulmonary bypass. The sternotomy is left open for a delayed closure. Pulmonary artery banding to prevent pulmonary overcirculation in infants with truncus is associated with a prohibitive mortality.

10.3 Postoperative Management

The postoperative course may be complex as the repair requires elaborate operative procedure. The hospital stay averages 8 to 12 days.

10.3.1 Hemodynamic Management

Hemodynamic instability can occur, especially in patients who have truncal valve dysfunction. Pulmonary artery hypertensive crises usually occur and require management. Systemic arterial oxygen saturation (SaO_2) should be normal. All intracardiac pressures should be normal.

10.3.2 Management of Pulmonary Artery Hypertensive Episodes

It is usually required to minimize pulmonary artery hypertensive episodes:

1. Sedation with infusions of a fentanyl.
2. Paralysis with neuromuscular blocking agent.
3. Oxygenation and a hyperventilation.
4. Inhaled nitric oxide: For reduction of pulmonary vascular resistance and pulmonary artery (PA) pressure.

10.3.3 Invasive Pressure Monitors

Arterial, central venous, and LA line. An oximetric catheter placed in the PA assesses cardiac output and pulmonary artery pressure.

10.3.4 Vasoactive Drug Infusions

Dopamine or dobutamine, milrinone, epinephrine, nitroprusside, and/or phenoxybenzamine (see Section I Chapters 4 & 16).

10.3.5 Postoperative Bleeding

Suture lines exposed to systemic arterial pressure and homograft suture lines under elevated pressure may predispose to early postoperative bleeding.

10.3.6 AV Conduction Abnormalities

They may be encountered due to manipulation in the area of the atrioventricular conduction tissue. Temporary pacing (atrioventricular pacing) should be available (see Section I Chapter 4).