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Pulmonary Atresia with Ventricular Septal Defect

11.1 Morphology

In this anomaly, the right ventricular outflow tract is underdeveloped with atresia of the pulmonary valve and presence of a large VSD (ventricular septal defect) with overriding of the aorta.

If a large patent ductus arteriosus is associated with this anomaly, the pulmonary arteries are usually normal in size and distribute blood to all segments of the lungs. This subtype of pulmonary atresia with VSD is managed as a severe form of tetralogy of Fallot.

If there is no patent ductus arteriosus or only a small ductus is associated with this anomaly, pulmonary artery blood supply is provided partially or entirely by major aorto-pulmonary collateral arteries (MAPCAs).

11.2 Operative Principles

The operative approach to the patient with pulmonary atresia and VSD should be tailored to the morphology of the defect, and total repair of the lesion is an ultimate goal. A singe stage total repair may be performed in early infancy if the pulmonary artery anatomy would allow, or correction of the defect is performed in stages.

Total repair is accomplished by:

1) Constructing central pulmonary arteries from the MAPCAs.
2) Establishing continuity between the RV and reconstructed pulmonary artery confluence.
3) VSD closure.

The total repair should proceed as early as the patient's pulmonary anatomy will allow.

11.3 Postoperative Management

The postoperative course following repair of pulmonary atresia with VSD and major aorto-pulmonary collateral arteries (MAPCA) may vary and depends upon individual anatomy and surgical course. The hospital stay may average one to two weeks.

11.3.1 Hemodynamic Management

Maintain adequate prograde pulmonary artery blood flow and prevent RV (right ventricular) dysfunction by preventing rises in pulmonary vascular resistance (PVR) postoperatively, especially after a single stage total repair in early infancy.

Arterial oxygen saturation and intracardiac pressures should be normal following surgery.

11.3.2 Prevention of Pulmonary Artery Hypertension or Rises in PVR

The pulmonary artery hypertensive episodes are either prevented from occurring or treated during the postoperative period by institution of either one or more of the following measures:

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

11.3.3 Invasive Monitors

Arterial, CVP (central venous pressure), and LA (left atrial) catheters.

An oximetric catheter is utilized to monitor cardiac output.

11.3.4 Vasoactive Drug Infusions

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

11.3.5 Postoperative Bleeding

Excessive bleeding can occur if pulmonary artery angioplasty is performed extensively.

11.3.6 Atrioventricular Conduction Abnormalities

Occasionally, occur as VSD closure requires manipulation near the atrioventricular conduction tissue. Temporary atrioventricular pacing should be available at the bed side.