15

Pulmonary Atresia with Intact Ventricular Septum

15.1 Morphology & Pathophysiology

The spectrum of this anomaly ranges from, a *normal-sized tripartite right ventricle* (infundibular, trabecular, and inlet portions) and an *imperforate pulmonary valve* with a simple commissural fusion, to a severely *hypoplastic or diminutive right ventricle* with atretic infundibulum, atresia of the pulmonary valve, and *coronary artery - myocardial sinusoid connections* (see Figure 15.1).

The prevalence of coronary artery-myocardial sinusoid connections are inversely related to size of the tricuspid valve, RV (right ventricular) cavity size, and the degree of tricuspid incompetence.

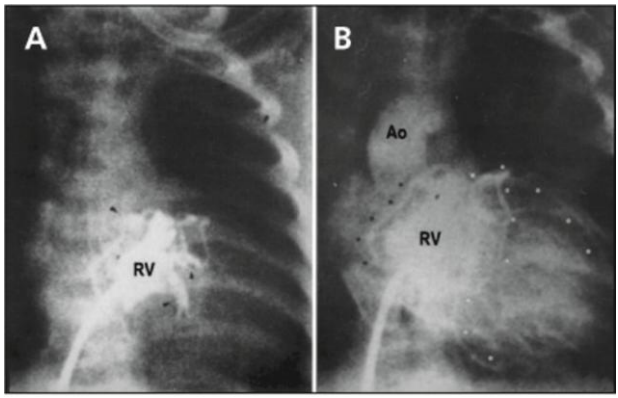

Figure 15.1 *Antero-posterior angiocardiogram of a hyponastic right ventricle (RV) in pulmonary atresia with intact ventricular septum: A. Note the presence of coronary artery - RV cavity connections (black arrowheads) originating from the inlet and infundibular portions of the RV. The right and left coronary artery trunks are beginning to be visualized. Note also the absence of a trabecular portion of the RV. B. After opacification of the coronary trunks, the right coronary artery (black dots) and the left anterior descending and circumflex branches (white dots) of the left coronary artery are filled with a contrast. Ascending aorta is also visualized. The annulus of the tricuspid valve is small showing mild valve regurgitation. Reproduced from: Santos MA, Azevedo VMP. Angiographic morphologic characteristics in pulmonary atresia with intact ventricular septum. Arquivos Brasilerios de Cardiologia. São Paulo. 2004; Vol 82.*

The tricuspid valve is often abnormal with severe annular hypoplasia and stenosis or severe regurgitation. *A patent foramen ovale or secundum atrial septal defect* allows an obligatory right-to-left shunt. The patent ductus arteriosus provides pulmonary blood flow.

Majority of the infants with PAIVS (pulmonary atresia with intact ventricular septum) become cyanotic a few hours after birth with a closure of the ductus arteriosus. Some of the patients with PAIVS (with tricuspid regurgitation) may manifest congestive cardiac failure early in life after right ventricular outflow-tract reconstruction and a systemic-pulmonary artery shunt or left-to-right ductal-dependent pulmonary blood flow.

If the coronary artery - right ventricular myocardial sinusoid connections are present, patient is a risk for myocardial ischemia due to coronary steal in the diastole or admixture of coronary arterial and right ventricular venous blood. If coronary artery stenoses also do exist, the myocardial perfusion is dependent on the right ventricular systolic events (RV pressure dependent), and patients are at a higher risk for myocardial ischemia, ventricular arrhythmias, and sudden death.

15.2 Principles of Surgical Management

15.2.1 Prostaglandin E_1 Infusion

Initial treatment consists of maintaining the ductal patency with a continuous intravenous infusion.

15.2.2 Metabolic Acidosis

Judicious administration of fluids and sodium bicarbonate are needed for correction.

15.2.3 Mechanical Ventilation

Persistent acidosis requires respiratory support.

15.2.4 Surgical or Catheter Intervention

Treatment algorithms for PAIVS depend on the size and morphology of both the right ventricle and the tricuspid valve as well as the presence of coronary artery - right ventricular myocardial sinusoid connections, as discussed below.

1. Absent ventriculocoronary connections and mild right ventricular hypoplasia and TV (tricuspid valve) hypoplasia (Z values 0 to 2):

   Transannular patch or surgical valvotomy or / transcatheter valvotomy with or without stenting of the patent ductus arteriosus.

   Growth of the right ventricle and tricuspid valve allows biventricular correction in the future.

2. Absent ventriculocoronary connections and moderate to severe right ventricular and tricuspid valve hypoplasia (Z= -2 to -3).

   Transannular patch or surgical valvotomy with a systemic-to-pulmonary artery shunt or a tanscatheter valvotomy with stenting of the patent ductus arteriosus is a procedure of choice.

   If the growth of the RV and tricuspid valve is not likely or optimal, Fontan repair is indicated in the future.

3. Presence of significant ventriculocoronary connections and proximal coronary artery stenoses (right ventricular dependent coronary circulation) and moderate to severe right ventricular and tricuspid valve hypoplasia (Z= -3 to less):

   The systemic-to-pulmonary artery shunt alone or stenting of the patent ductus arteriosus is performed. This is followed by a bidirectional

cavopulmoanry anastomosis at 6 months of age and Fontan repair at 2 to 3 years of age.

At Fontan operation, a modification of a technique to perfuse the RV with oxygenated blood and recruitment of coronary sinus in the atrium (in Fontan circuit), would improve myocardial oxygen supply.

4. Presence of mild forms of right ventricular dependent ventriculocoronary connections *(i.e., coronary-myocardial sinusoid connections without significant or absent coronary stenoses*).

The initial palliative procedure in this category depends on the z value of the tricuspid valve:

Tricuspid Z value= 0 to -2: RVOT procedure.

Tricuspid Z value= -2 to -3: RVOT procedure + shunt.

Tricuspid Z value= -3 to less: Shunt alone.

15.3 Postoperative Management

Systemic pulmonary artery shunt: (see Blalock-Taussig shunt, Chapter 21).

Bidirectional cavopulmonary anastomosis: (see bidirectional Glenn or hemi-Fontan in univentricular heart, Chapter 17).

Fontan operation: (see Fontan circulation in univentricular heart, chapter 17, tricuspid atresia, chapter 16).