4

Atrioventricular Septal Defects

4.1 Morphology

Failure in development and fusion of the embryonic endocardial cushions gives rise to the spectrum of lesions ranging from a partial to complete atrioventricular septal defect (AVSD) or AV (atrioventricular) canal defect. The cardiac anatomy of the partial and the complete AV canal defects is the same but distinct from normal.

The atrioventricular valve annulus and valves are displaced apically, so the distance from mitral valve annulus to ventricular apex is shorter, and the left ventricular outflow tract is displaced anteriorly producing a characteristic "gooseneck" deformity" seen on the angiogram.

Though the inlet ventricular septum is deficient (scooped out septum) in both the partial and complete AV canal, there *is no interventricular communication* in the partial AVSD.

4.1.1 AV Conduction Tissue

The AV node and His bundle are displaced inferiorly and posteriorly to lie between the inferior margin of the primum atrial septal defect and the tricuspid valve annulus, medial to coronary sinus within the crest of the ventricular septum.

The AV valve has four or five leaflets (superior, inferior, right lateral, and left lateral) at two ventricular orifices with varying degrees of bridging across the ventricular septum by the superior and the inferior leaflets. The extent of malformation of the AV valve distinguishes the partial AVSD from the complete AVSD. The partial AVSD is characterized by a crescent shaped primum atrial septal defect and two AV valves. The central fusion of the superior and the inferior bridging leaflets of the AV valves with underlying crest of the ventricular septum prevents interventricular communication.

4.1.2 The AV Valve

The right superior, right inferior, and right lateral leaflets form the right AV valve. The left superior, left inferior, and left lateral leaflets form the left AV valve. The cleft, which is a slit or a triangular, divides the left superior and the left inferior leaflet and correspond to a cleft in the septal leaflet of the mitral valve, producing various degrees of mitral insufficiency. In most, a cleft may also be seen in the septal leaflet of tricuspid valve. (see Figure 4.1)

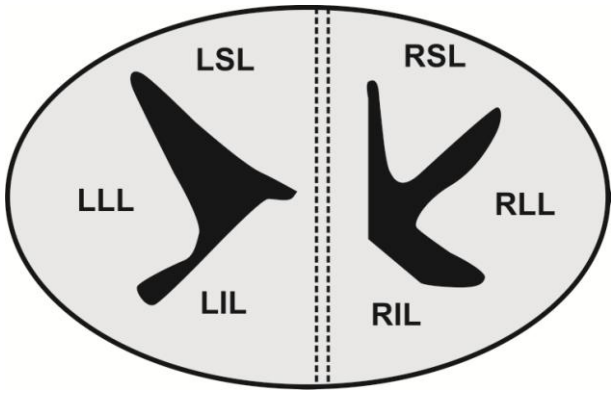

Figure 4.1 *Diagram depicting components of atrioventricular valve in the partial atrioventricular canal septal defect. Note the central fusion of the right and left superior and the inferior leaflets, to distinct the AV valve into the tricuspid and mitral components. The dashed lines show fusion of the bridging leaflet tissue to the underlying crest of the ventricular septum to prevent inter-ventricular communication. RSL= right superior leaflet, RIL=right inferior leaflet, RLL= right lateral leaflet, LSL=left superior leaflet, LIL= left inferior leaflet, LLL= left lateral leaflet.*

4.1.3 The Complete Atrioventricular Septal Defect (AVSD)

In this lesion, in addition to the presence of crescent shaped primum atrial septal defect, the AV valve orifice is not partitioned into tricuspid and mitral orifice (producing so called common atrioventricular valve and a common AV valve annulus) due to failure in fusion of superior and inferior common leaflets centrally. The superior and inferior leaflets also do not fuse with the underlying crest of the ventricular septum producing interventricular communication (see

Figures 4.2, 4.3 & 4.4). The complete AVSD is further classified by morphology of the superior bridging leaflet.

(I) Rastelli Type A

The anterior or superior leaflet has a division into the mitral and tricuspid components. The chordae attach the margins of this medial portion of this divided leaflet to the crest of the ventricular septum. The lateral chordae tendineae from the lateral portions of the leaflet attach to the anterior papillary muscles in each ventricle (see Figure 4.2).

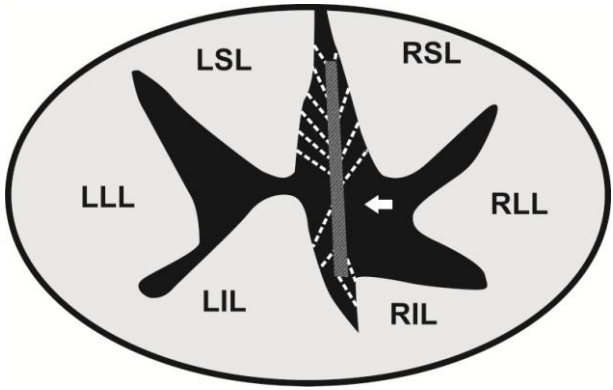

Figure 4.2 *Diagram depicting components of common atrioventricular valve in the complete atrioventricular canal septal defect. Note the failure of fusion of superior and inferior leaflet tissue to producet the common AV valve. Note also the complete division of the superior and inferior bridging leaflets into the right and left components with attachment of medial margins of the leaflets to the underlying crest of the ventricular septum (Rastelli type A valve). RSL= right superior leaflet, RIL=right inferior leaflet, RLL= right lateral leaflet, LSL=left superior leaflet, LIL= left inferior leaflet, LLL= left lateral leaflet. Arrow head showing the crest of the ventricular septum.*

(II) Rastelli Type B

The anterior or superior bridging leaflet has incomplete division into the mitral and tricuspid components. The chordae from the margins of the division have no direct insertion to the ventricular septum but rather insert on to an anomalous papillary muscle positioned in the right ventricle (see Figure 4.3).

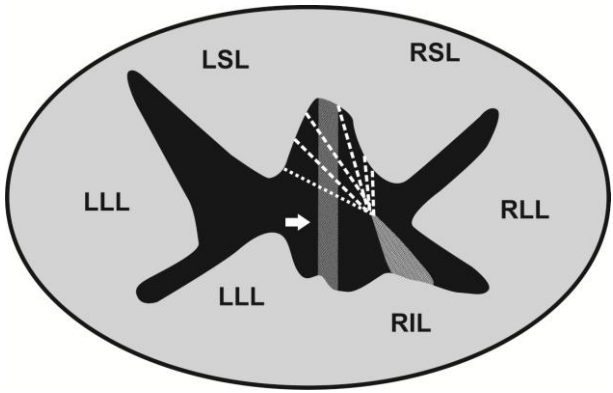

Figure 4.3 *Diagram depicting components of common atrioventricular valve in the complete atrioventricular canal septal defect. Note the failure of fusion of superior and inferior leaflet tissue to produce the common AV valve. Note the incomplete division of both the superior and inferior bridging leaflets into the right and left components with chordal attachment of medial margins of the leaflets to an anomalous papillary muscle in RV (Rastelli type B valve). RSL= right superior leaflet, RIL=right inferior leaflet, RLL= right lateral leaflet, LSL=left superior leaflet, LIL= left inferior leaflet, LLL= left lateral leaflet. Arrow head showing the crest of the ventricular septum.*

(III) Rastelli Type C

The anterior or superior bridging leaflet is not divided, larger, looks like a single leaflet and overhangs the septum. It is not attached to the ventricular septum and is referred to as being "free floating". The chordae attach the lateral margins of the leaflet to the papillary muscles of the right and left ventricular wall (see Figure 4.4).

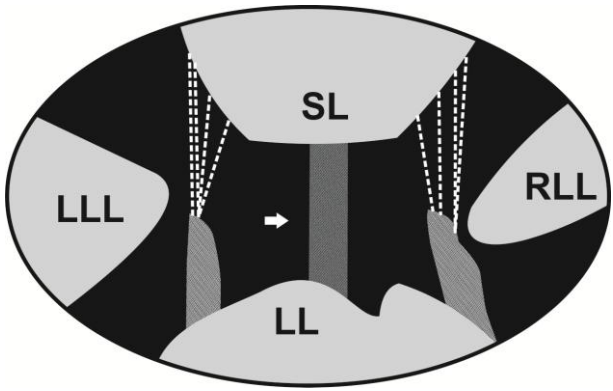

Figure 4.4 *Diagram depicting components of common atrioventricular valve in the complete atrioventricular canal septal defect. Note the failure of fusion of superior and inferior leaflet tissue to produce the common AV valve. Note the superior bridging leaflet is not divided (into the mitral or tricuspid components) and overhangs the septum. It is not attached to the ventricular septum. The chordae attach the lateral margins of the leaflet to the papillary muscles of the right and left ventricular wall (Rastelli type C valve). RSL= right superior leaflet, RIL=right inferior leaflet, RLL= right lateral leaflet, LSL=left superior leaflet, LIL= left inferior leaflet, LLL= left lateral leaflet. Arrow head showing the crest of the ventricular septum.*

4.1.4 The Intermediate AVSD

The morphology of this lesion refers to the combination of a partial AVSD with a small interventricular communication. The superior and inferior bridging leaflets fuse overlying the ventricular septum, so two distinct valvular components are observed.

4.2 Pathophysiology

The left to right shunt through the primum septal defect and severity of mitral insufficiency (MR) determine the physiologic consequences of partial AVSD. If MR is severe, cardiac failure and pulmonary hypertension develops early by 1 to 3 years of age. In the absence of mitral insufficiency, the physiology is similar to the secundum ASD.

The magnitude of blood flow through the AVSD and the amount of atrioventricular valve regurgitation determines the physiology in complete AV canal. As pulmonary vascular resistance decreases normally over first 4 to 6 weeks after birth, most infants have significant left-to-right shunt through both the atrial and ventricular septal defects with increased PBF, development of CHF, pulmonary artery hypertension, and early onset of pulmonary vascular disease. Infants with clinically significant atrioventricular valve regurgitation may also have signs of CHF and growth failure.

Some infants with minimal AV valve regurgitation and elevated PVR may remain asymptomatic until second or third decade when they develop increasing cyanosis from advanced pulmonary vascular disease.

4.3 Treatment

4.3.1 Medical Treatment

The main stay of this therapy is combination of diuretics, digoxin, and ACE (angiotensin converting-enzyme) inhibitors. Medical treatment for children with CHF in complete AV canal defect is given in preparation for surgical RX though its effectiveness has been questioned. Medical treatment, however, should not be pursued for more than a few weeks before a definitive repair, as results of surgical repair in young infants (2-3 months age) have been generally fair.

4.3.2 Surgical Treatment

(I) Partial AVSD

Operation is performed usually between 1 to 4 years. If the patients present with severe CHF or pulmonary hypertension, the surgery is undertaken < 2 years. A defect without MR (mitral regurgitation) is managed like the secundum ASD and is electively repaired before school going age (i.e., 5 years).

(II) Complete AVSD

Operation should be performed by 1 year or usually by 6 months of age. Earlier operation is indicated in patients with severe CHF. PA banding would not prevent the hemodynamic consequences of mitral regurgitation and CHF, so a complete and definitive repair of common AV canal is currently preferred even at an early age.

(III) Probable Indications for Pulmonary Artery Banding

1. A rare case of refractory CHF in a low-birth-weight infant may be palliated with the placement of PA band.
2. In complete AVSD with additional muscular ventricular septal defects (VSDs), the PA band may alleviate CHF for 6-12 months during which time the VSDs may spontaneously close and thus simplify the eventual complete repair.

4.4 Operative Technique

4.4.1 Principles of Repair of Partial AVSD

1. Repair the cleft of mitral valve to correct mitral insufficiency and avoid overcorrection.
2. Closure of the primum atrial septal defect with a patch.
3. Prevention of heart block and it is accomplished by
   - Placing series of interrupted sutures superficially and to the left of the postero-inferior rim of the primum septal defect in the mitral annulus in the area of the coronary sinus or

- Placing sutures to the right of the conduction bundle and the coronary sinus, thus avoiding the bundle in the sutures but recruiting the coronary sinus in the left atrium.

4.4.2 Principles of Repair of Complete AVSD

1. Construction of a competent mitral valve by approximation of left superior and the left inferior leaflets.

2. A prosthetic patch is attached to the crest of the ventricular septum, placing the sutures to the right side of the septum in the region of coronary sinus, to avoid heart block.

3. Attachment of both the superior and inferior leaflets of the mitral and tricuspid valve to a patch at the level where they would normally attach to the intact ventricular septum.

4. Closure of the primum atrial septal defect with a patch.

4.5 Postoperative Management

The postoperative course following the repair of AV septal defect is variable and the hospital stay averages 1 week to 12 days.

4.5.1 Hemodynamic Management

Delayed surgical intervention is associated with moderate pulmonary hypertension which requires management.

Closely monitor the right, the left atrial, and the pulmonary artery (PA) pressures.

Elevated left atrial pressure or right atrial pressures may suggest the AV valve regurgitation.

Decrease in cardiac output as a consequence of moderate or severe pulmonary hypertension, and/ or supraventricular arrhythmias requires prompt management.

A loud systolic murmur at the apex with decrease in cardiac output suggests significant valve regurgitation of the left AV valve and mandates evaluation by doppler echocardiogram.

4.5.2 Management of Pulmonary Artery Hypertension

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 PA pressure.

4.5.3 Invasive Pressure Monitors

Arterial, central venous, and LA (left atrial) line.

PA catheter if preoperative pulmonary artery hypertension exists.

4.5.4 Vasoactive Drug Infusions

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

4.5.5 Postoperative Bleeding

Excessive bleeding is rare.

4.5.6 AV Conduction Abnormalities

Complete heart block may be due to a faulty technique. The suture line at the ventricular septal crest near the coronary sinus and tricuspid annulus may injure the conduction tissue and may result in transient or permanent AV block.

Temporary pacing (atrioventricular pacing) should be readily available at the bed side. The permanent pacemaker is indicated at discharge if the AV block persists (see Section I Chapter 4).