3

Ventricular Septal Defect

3.1 Morphology

There are four anatomic types of ventricular septal defects (VSD) based upon their location on the ventricular septum as described below:

The inlet portion of the septum is the area between the annulus of the tricuspid valve and the attachments of the tricuspid valve to the right ventricular wall and the septum. *The inlet VSD* is posterior and inferior to perimembranous defects. Inlet defects are uncommon.

The trabecular area of the septum extends from the inlet area out to the apex and includes the coarse trabeculae of the right ventricle. *Muscular septal defects* occur in the trabeculated muscular septum and are often multiple and account for 5 to 20 percent of VSDs. These include *marginal defects* along the right ventricular free wall-septal junction, *central defects*, and *apical defects*. Multiple muscular defects have resulted in the pseudonym "Swiss cheese" septum. Apical defects are frequently difficult to visualize on the right ventricular side because they can be multiple and covered over by trabeculations.

The outlet area of the septum or infundibular septum extends up to the pulmonary valve from the trabecular area. *The conal or sub-arterial septal defect*, also referred to as a supracristal, infundibular, sub-pulmonary or doubly committed defect, occurs in this area of the septum and accounts for 5 to 7 percent of all defects, but the incidence of this defect is approximately 30 percent in Far Eastern countries.

The membranous septum is the area of the septum between the trabecular, inlet, and outlet areas, and is usually partially covered over by the septal leaflet of the tricuspid valve. *Perimembranous VSD* or infracristal defects occur in this area of the septum and account for approximately 80 percent of VSDs. These defects are also termed as conoventricular or malaligned type of VSDs due to

mal-alignment of the conal septum with the trabeculated septum during development of the interventricular septum and bi- partition of the ventricle. These defects account for approximately 80 percent of VSDs. The papillary muscle of the conus is within the perimembranous area. (see Figure 3.1.)

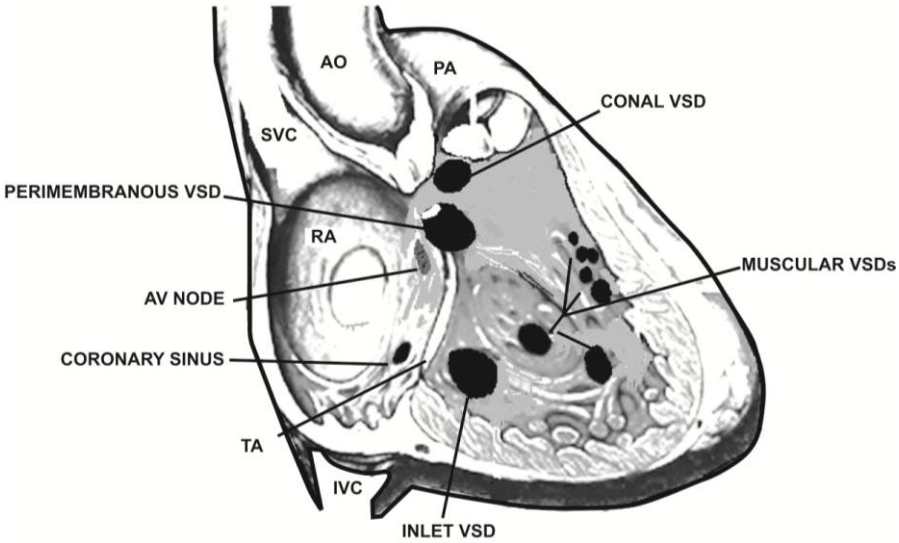

Figure 3.1 *Diagram depicting four anatomic types of ventricular septal defects in the inter-ventricular septum as viewed from the right ventricular side. AO= ascending aorta, AV node= atrioventricular node, IVC= inferior vena cava, PA= pulmonary artery, RA= right atrium, SVC= superior vena cava, TA= septal margin of the tricuspid annulus after removal of the septal leaflet and other leaflets of the tricuspid valve. VSD= ventricular septal defect.*

VSD and conduction bundle: The bundle of His lies in the subendocardial position on the left ventricular side of the ventricular septum as it courses along the posterior-inferior margin of perimembranous defect. It is the only defect in which heart block is a potential surgical complication. The His bundle passes anterior and superiorly to the inlet defects, and is usually remote from the margins of outlet and muscular defects.

3.2 Pathophysiology

The physiology of this lesion is determined by the variables such as resistance to flow across the VSD, and is related to 1) the size of the defect and 2) the relative resistance of systemic and pulmonary circulations. A small VSD with high resistance to flow (as in restrictive VSD) permits only a small left-to-right shunt. In contrast, with a large defect that is equal in diameter to the aortic annulus, there is equalization of the left and right ventricular peak systolic pressures, and both the ventricles function as a single pumping chamber with two outlets. The degree and direction of shunting is determined by relative resistance of systemic and pulmonary circulations. The magnitude and direction of shunting varies also during various phases of cardiac cycle and mainly dependent on pressure gradients in the ventricular cavity.

3.2.1 Left to Right Shunting

a) It occurs in *early phase of systole* as the left ventricular pressure rises faster than the right ventricular pressure and the left ventricle contraction occurs before the right ventricle.

b) It occurs normally *in mid and late systole* as the pressure in the ventricles in the mid and late systole is determined by resistance for the right (pulmonary vascular resistance) and left ventricular ejection (systemic vascular resistance).

c) *It* occurs during *mid to late diastole* due to a slightly higher left ventricular diastolic pressure.

3.2.2 Right to Left Shunting

It occurs in *early diastole as* the LV (left ventricle) relaxes more quickly than the RV (right ventricle). The high pressure in the RV cavity results in a transient pressure gradient favoring the right to left shunt. The blood that is shunted

flows back into the right ventricle during mid to late diastole. As a result, there is no net right to left shunt.

In the presence of left to right shunt, pulmonary blood flow is increased well above the systemic blood flow with increased flow in the PA, left atrium, with elevations of pulmonary artery, left atrial, and pulmonary venous pressures and consequent increase in interstitial fluid of the lungs.

The work load of both the right and left ventricles is increased, and as the interstitial fluid in the lungs increases in the presence of non-complaint lungs (seen often in young infants), the work of breathing is increased. In the presence of reduced systemic blood flow, increased work of breathing, and increased work load of both the ventricles, the energy expenditure is greatly increased resulting in severe growth failure in infants.

Pulmonary artery hypertension leads to the development of pulmonary vascular disease with elevated pulmonary vascular resistance resulting in reversal of shunt (i.e., right-to-left shunt), decreased pulmonary blood flow, ejection of unoxygenated blood out of the aorta, and variable degrees of cyanosis (Eisenmenger syndrome).

3.3 Surgical Treatment

A large VSD with severe or intractable congestive cardiac failure should be closed in all young infants of < 3 months of age. The rare exception is a minimally symptomatic and a large VSD may be anticipated to close spontaneously or would decrease in size.

Even in the presence of moderate elevations of PVR (pulmonary vascular resistance) (8 Wood units), any large VSD in infants 6 months of age or less should be promptly closed. A large VSD in infants and children > 6 months of age with normal or low PVR < 5 should be closed. If the PVR is moderately

elevated (5 to 10 units & Qp/Qs 1.5 - 1.8 at rest) closure may be advised though some of the patients would not benefit from the operation due to progressive pulmonary vascular disease. The latter patient subsets are identified preoperatively (Qp/Qs falls 1.0 or < 1.0 with a fall in O_2 saturation on moderate exercise) and the operation is denied. Operation is contraindicated if PVR is > 10 units (Qp/Qs < 1.5 at rest), as closure prevents obligatory R to L shunting during exercise, and the life expectancy is same whether the defect is closed or left open.

A moderate sized VSD with minimal symptoms and only with pulmonary plethora (Qp/Qs 3.0, moderate PAH (40 to 50 mm Hg), PVR < 5), operation is indicated if not spontaneously closed or reduced after an observation period of 5 years.

In asymptomatic or minimally symptomatic children with restrictive and small VSD, the surgery is not indicated as spontaneous closure is very likely. The operative closure is done for infundibular VSD, even small, which persists in children beyond 5 years of age or associated with diastolic murmur in the aortic area.

3.4 Operative Technique

The procedure is performed under CPB with moderate hypothermia (28-34 ℃) using the standard aortic and bicaval venous cannulation and antegrade cardioplegic arrest. The left ventricle is decompressed by venting through the right superior pulmonary vein. Alternately, the procedure may be performed under deep hypothermia and circulatory arrest or a low flow CPB.

3.4.1 Right Atrial Approach (Transatrial Approach)

Most cono-ventricular or perimembranous VSDs may be closed working through a vertical right atriotomy. The surgeon inspects and repairs the VSD looking across the tricuspid valve and into the right ventricle. To aid in visualization of the defect, detachment of the septal leaflet of the tricuspid valve may be required. The defect is closed with adequately sized PTFE (Impra) or Dacron patch using a running polypropylene suture or horizontal pledgeted mattress sutures. While suturing, careful attention is necessary to not to injure the conduction bundle which lies on the left ventricular side of the interventricular septum, from the level of the insertion of the papillary muscle of the conus to the AV node in the vicinity of tricuspid annulus, traversing at the posteroinferior margin of the ventricular septal defect. Care is also advised to avoid injury to the aortic valve and it's annulus with sutures.

3.4.2 Transpulmonary Approach

The conal or supracristal infundibular VSDs may be approached through an incision in the main pulmonary artery, working across the pulmonary valve.

3.4.3 Transaortic Approach

If significant aortic valvular incompetence accompanies the conal or perimembranous defects due to aortic valve leaflet prolapse, the aortic valve repair (valvuloplasty) is performed. The elongated free edge of the distorted leaflet is shortened, and the leaflet is re-suspended against the aortic wall with sutures.

3.4.4 Transventricular Approach

Most of the muscular VSDs may be approached through an incision in the ventricular apex. The defects are closed using a synthetic patch, such as Dacron or

polytetrafluoroethylene, sewn to the rightward aspect of the VSD with a running, nonabsorbable, and monofilament suture. Care should be taken to avoid placing deep sutures in the area of conduction tissue.

3.5 Postoperative Management

The postoperative course following an isolated VSD closure is usually uneventful. The hospital stay averages 5 to 8 days.

3.5.1 Hemodynamic Management

If surgical intervention is delayed, pulmonary artery hypertension co-exists and requires management.

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

Elevated left atrial pressure as compared to the right atrial pressure in the presence of a low cardiac output may suggest a persistent left to right shunt due to a residual VSD.

Persistent of low cardiac output state requires evaluation by doppler echocardiogram or repeat cardiac catheterization and surgical re-intervention.

3.5.2 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.

3.5.3 Invasive Pressure Monitors

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

PA catheter if preoperative pulmonary artery hypertension exists.

3.5.4 Vasoactive Drug Infusions

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

3.5.5 Postoperative Bleeding

Excessive bleeding is rare.

3.5.6 AV Conduction Abnormalities

Edema around the VSD suture lines* may result in transient AV block.

Temporary pacing (atrioventricular pacing) should be readily available at the bed side (see Section I Chapter 4).

* Conduction bundle lies in the subendocarium of the left ventricular surface of the septum and courses on the infero-posterior margin of the perimembranous VSD.