Chapter

62 Ventricular Septal Defects

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A 23-month-old female presents for annual physical examination. Her current vital signs are: blood pressure 78/22 mmHg; heart rate 97/min; respiratory rate 26/min; SpO_2 100% on room air. On auscultation, there is a holosystolic murmur at the left sternal border with a split S2. Upon further questioning, the mother reports that she has "fast breathing" when eating with occasional sweating at meal times.

Chest X-ray reveals prominent pulmonary vascular markings and mild enlargement of the right-sided cardiac silhouette. The patient is sent for transthoracic echocardiography revealing a large conoventricular type ventricular septal defect.

DIAGNOSIS

What Is the Incidence for VSDs?

The incidence of ventricular septal defects (VSDs) is approximately one to two per 1,000 live births making it one of the most common congenital heart defects. Isolated VSDs account for nearly 20–30% of all congenital heart defects. VSDs are also present in a wide range of other cardiac defects, e.g., atrioventricular (AV) canal defects and pulmonary atresia with VSD. Approximately 25% of isolated VSDs are in association with another anomaly either genetic or morphologic (trisomy 21, VATER syndrome, VACTERL).

What Is the Most Likely Age for Presentation in Patients with VSDs?

As opposed to atrial septal defects, VSDs can relay symptoms more readily in the neonatal period. VSDs may occur as isolated defects or as part of a larger continuum of congenital structural defects. Smaller VSDs can often be diagnosed early in life by an astute clinician auscultating a murmur prompting further evaluation. VSDs in associated with larger congenital heart defects, e.g., complete AV canal defects, are often diagnosed in the early neonatal period.

Identify the Four Main Types of VSDs

VSDs are classified based on the anatomic location of the defect (Figure 62.1). There are two main types of basic nomenclature for VSDs, and this is somewhat institution dependent. The main types will be listed here with the lesser-used classification in parentheses. The main types of ventricular septal defects are: conventricular (perimembranous), conotruncal (outlet), inlet (AV canal type), and muscular (trabecular).

Briefly Describe the Three Main Segments of the Ventricular Septum

The ventricular septum has three primary components:

- The ***inlet portion*** of the septum extends from beneath the septal leaflet of the tricuspid valve to the tricuspid valves, papillary muscle attachment.
- The ***trabecular or muscular portion*** extends from the chordal attachment of the tricuspid valve to the apex of the ventricles and cephalad toward the conal septum.
- The ***outlet portion*** is the infundibular septum extending to the pulmonary and aortic annuli.

What Is the Natural History of Untreated VSDs in the Neonatal Period?

Approximately 50–70% of VSDs identified in the neonatal period will close spontaneously or regress in size by 6–12 months, specifically the conventricular and muscular type VSDs.

VSDs of the conotruncal (outlet) and inlet (AV canal type) generally do not close spontaneously and require intervention.

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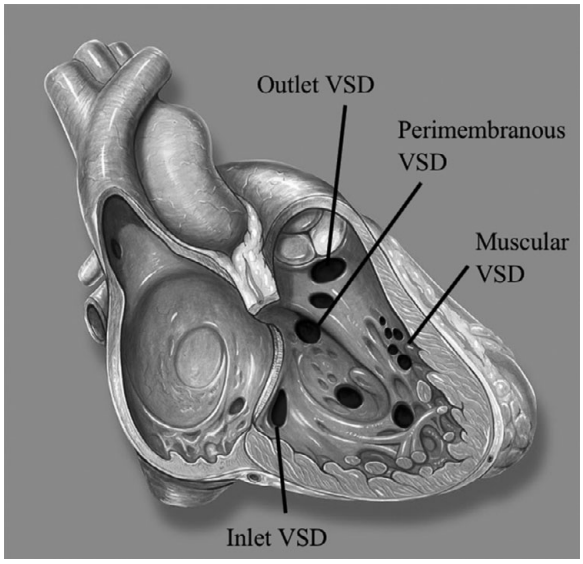

Figure 62.1 Types of ventricular septal defects. Image credit: Patrick J. Lynch, medical illustrator, and C. Carl Jaffe, MD, cardiologist. Reproduced here under CC BY 2.5 license https://creativecommons.org/licenses/by/2.5/

Describe the Typical Presentation of Children with Large Unrepaired VSDs

With a large and unrestrictive VSD, there is often significant left-to-right shunting. As the pulmonary vascular resistance (PVR) falls in the first days to weeks of life, left-to-right shunting increases. If the VSD is moderate and the PVR is high, it can be missed during early auscultation. The shunt results in excessive pulmonary blood flow (overcirculation) and these neonates can present with tachypnea and dyspnea especially with feeding.

Often, these children have sweating during feeding and are generally small for their age due to excessive work of breathing. The excessive overcirculation results in congestive heart failure that can be severe. For small patients, medical management with diuretics and feeding through an indwelling nasogastric tube to reduce the work of breathing can allow for the child to grow, making the surgical repair technically easier to perform.

What Is the Meaning of the Qp:Qs Ratio in Patients with VSDs?

The Qp:Qs ratio is the ratio of the pulmonary blood flow to systemic blood flow (Figure 62.2). Normally this ratio equals one with the entire preload to the right ventricle (RV) eventually becoming the preload to the left ventricle (LV). With large mixing lesions, it is important to determine the % of blood that is recirculating.

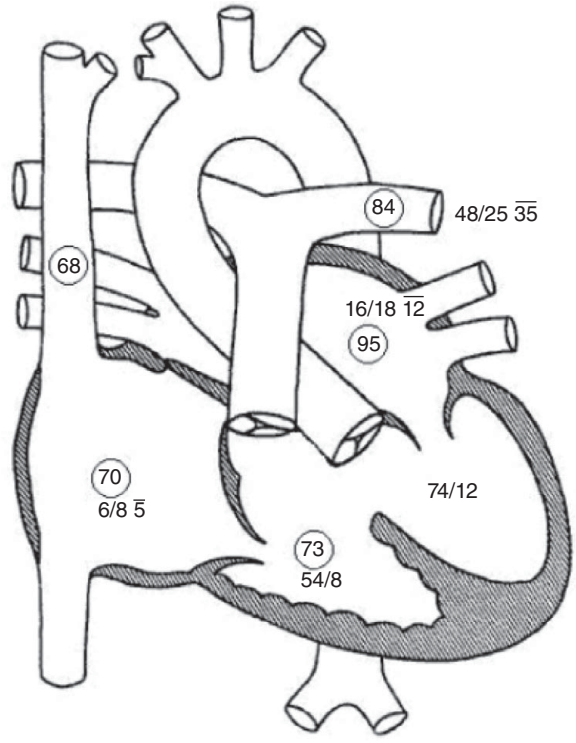

Figure 62.2 Diagram of cardiac catheterization findings for a patient with perimembranous VSD

A simplified formula for determining the Qp:Qs can calculated using only oxygen saturations.

$$\text{Qp : Qs} = (\text{Aortic sat\%} - \text{Right atrial sat\%})/(\text{Pulmonary venous sat\%} - \text{Pulmonary artery sat\%}).$$

Small VSDs will have a Qp:Qs ratio of <1.5:1
Medium VSDs have a large Qp:Qs of 2–3:1
Large VSDs can have Qp:Qs ratios exceeding 3:1 or greater.

For Figure 62.2, What Is the Calculated Qp:Qs Ratio When the Saturation in the Aorta Is 100%?

From the formula,

$$\text{Qp : Qs} = (\text{Aortic sat\%} - \text{Right atrial sat\%})/(\text{Pulmonary venous sat\%} - \text{Pulmonary artery sat\%})$$

In this case, the formula becomes Qp:Qs = (100–70)/(95–84) giving a Qp/Qs of 2.7.

Notice that from the right atrium to the RV, the saturation increases from 70 to 73. This is known as a step-up. Normally, the saturation from the right

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atrium to ventricle should be identical. The fact that there is an increase, or step-up, signifies the presence of a mixing lesion.

What Is the Consequence of Leaving a Large VSD Unrepaired?

Aside from the issues with feeding and poor weight gain, significant overcirculation leads to pulmonary vascular occlusive disease. The muscularization of the pulmonary vasculature develops over time from persistent elevations in blood volume and pressure within this normally low-pressure system. Over time, this leads to pulmonary hypertension, right ventricular hypertrophy and even flow reversal across the VSD in severe cases (right-to-left shunting).

List the Indications for Closing a VSD in the Neonatal Period

- Large VSDs
- Symptomatic VSDs
- VSDs with associated aortic insufficiency
- Medium-sized VSDs with failure to thrive
- Conotruncal (outlet), inlet (AV canal type)
- Residual VSDs >3 mm and those with associated Pulmonary artery pressure elevation
- Smaller VSDs when the patient is already undergoing cardiac surgery

Smaller lesions that are less symptomatic may be managed by diuretics while allowing the child to grow with the hopes of spontaneous closure. Delay until the child is two to three years of age provides a larger patient that is surgically more appealing and avoids cardiopulmonary bypass in the neonatal period.

My Patient Is Three Years Old at the Time of Diagnosis. Why Are They Doing a Cardiac Catheterization?

For patients older than one to two years when a large VSD is discovered, a cardiac catheterization is recommended to evaluate if the lesion is amenable to closure. Patients with high pulmonary vascular resistance and those with desaturation during physical activity should not have their VSD closed immediately. If the patient's PVR is elevated from prolonged overcirculation, the Qp:Qs may be decreased indicating that right-sided (RV) pressure is elevated. Desaturation during activity signifies reversal of flow across the VSD (right heart pop-off) and closure of the VSD can result in right heart failure.

List the Types of VSDs from Most to Least Frequent

- Conventricular (perimembranous) approximately 70–80% of VSDs.
- Muscular (trabecular) approximately 10–20% of VSDs.
- Conotruncal (outlet) approximately 5% of VSDs.
- Inlet (AV canal type) approximately 5% of VSDs.

Describe the Defect Associated with a Conventricular (Perimembranous) VSD

These VSDs are defects between the conal portion of the septum and extend toward the tricuspid valve annulus. The aortic valve can be visualized through the defect and occasionally, the noncoronary leaflet of the aortic valve may prolapse through the defect leading to aortic insufficiency.

Describe the Defect Associated with a Conotruncal (Outlet) VSD

These VSDs are in the conotruncal region of the septum immediately beneath the pulmonary valve. Occasionally, these VSDs are referred to as subpulmonary or infundibular VSDs due to their location. In this defect, the tissue separating the aortic and pulmonary valves is deficient and can lead to aortic insufficiency due to prolapse of the aortic valve leaflet into the defect.

Describe the Defect Associated with an Inlet (AV Canal Type) VSD

These VSDs are situated at the level of the tricuspid valve with the septal leaflets and annulus forming their border. The conduction tissue is located near the posterior border of the defect at the point of division into the right bundle branch. Occasionally, there may be an associated cleft of the anterior mitral valve leaflet.

Describe the Defect Associated with a Muscular VSD

These defects are located at any place within the muscular septum. They can be a single defect or multiple

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defects. These defects can have numerous openings on the RV side of the septum but usually one opening on the LV side of the septum. Generally, muscular VSDs are not notable for their proximity to the conduction system unless there is an associated conventricular VSD or the muscular VSD is located near the tricuspid valve.

What Is the Pathophysiology of a VSD?

A VSD serves as a left-to-right shunt. The degree of shunting is determined by the size of the defect, relative diastolic compliance of the ventricles, presence of pulmonary valve stenosis and pulmonary vascular resistance.

What Are the Major Hemodynamic Consequences of an Uncorrected VSD?

With a VSD, the right ventricle is subjected to chronic volume overload. This results in ventricular hypertrophy and dilation resulting in RV dysfunction and atrial arrhythmia. The pulmonary vasculature also undergoes changes from the chronic volume overload resulting in muscularization of the pulmonary vascular bed and elevated pulmonary vascular resistance. Over time, elevated pulmonary vascular resistance results in RV pressure elevations and hypertrophy. In severe cases, this can result in reversal of flow (right-to-left shunting) across the VSD, a condition known as Eisenmenger syndrome.

Describe the Appearance of a Chest X-Ray in a Patient with a Large VSD

Generally, these patients will have right ventricular hypertrophy and enlargement of the cardiac silhouette. There may be prominence of pulmonary vasculature due to pulmonary overcirculation.

What Is the Characteristic Murmur in Patients with a VSD?

With the increase in left-to-right flow, the right ventricle experiences an excessive volume overload and takes longer to eject its stroke volume. This leads to delayed closure of Pulmonary valve and thus a split S2 may result. A holosystolic systolic murmur best heard at the left sternal border may be observed.

Describe the Two Main Invasive Approaches to Closure of VSDs

Common approaches for closure of VSDs include surgical closure or catheter-based device closure.

Some muscular type VSDs may be amenable to a catheter-based closure using a device similar to ones used for secundum ASDs. More commonly, surgical intervention is done using cardiopulmonary bypass most commonly with bicaval cannulation to optimize visualization. The VSD is approached by right atriotomy and generally through the tricuspid valve. These defects are closed with a Dacron patch, homograft patch, occasionally with a PTFE-based Gore-Tex patch or simply sutured closed with or without a pledget.

What Are the Major Anesthetic Considerations for Surgical Repair of VSDs?

Generally, patients with VSDs have pulmonary overcirculation. These patients may be managed with diuretics if significant pulmonary congestion is present. Mask induction is usually performed with a premedicant if needed.

Most often, the repair of a routine VSD is uneventful. The patient should be prepared for extubation either in the operating room or shortly afterwards in the intensive care unit. Repair of conotruncal and conventricular VSDs can affect the aortic valve due to its close proximity to the sutures required for the repair. Repair of inlet VSDs can affect the conduction system and means of artificial pacing should be readily available. A transesophageal echocardiograph (TEE) is often performed at the termination of cardiopulmonary bypass and should specifically evaluate for residual VSDs and damage to the anterior mitral valve leaflet, aortic non and right coronary leaflets and the septal leaflet, of the tricuspid valve.

Suggested Reading

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