

Ventricular Septal Defect

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A ventricular septal defect (VSD) is a communication between both ventricles. There are different types of VSDs based on anatomy (Figure 11-1):

- **Perimembranous.** Underneath the tricuspid valve and with continuity between the tricuspid and aortic valves. The septal leaflet of the tricuspid valve forms one of the rims of the defect.
- **Muscular.** Completely surrounded by muscular tissue. It can be subclassified into outlet, inlet, mid-muscular, and apical, depending on the area of the muscular septum where it is mainly located.
- **Inlet.** Analogous to the VSD seen in patients with AV septal defects. Both the right and left AV valves are in continuity and form the posterior border of the defect. It may be associated with a mitral cleft.
- **Doubly committed juxta-arterial (DCJA).** Also called supracristal or subpulmonary VSD. The defect is bordered by both arterial valves (aortic and pulmonary), which are in continuity. There is a lack of infundibular septum and the pulmonary valve lies at the same level as the aortic valve. The defect can have perimembranous extension, in which case there is continuity between the aortic and tricuspid valves.

Pathophysiology and Clinical Presentation

The magnitude and direction of shunting depends on the size of the defect and the relationship between the resistance of the systemic and pulmonary circulations. Classically, since SVR is higher than PVR, shunting occurs from left to right (Figure 11-2). In situations in which the defect is very small (pressure-restrictive), shunting will be relatively small and the RV and PA pressures will remain normal (i.e., much lower than the LV and aortic pressures). In a large VSD, there is significant left-to-right shunting and the LV and RV pressures are equalized (non-restrictive VSD).

In newborns, given the transitional circulation after birth, there is a gradual decrease in PVR that translates into progressive left-to-right shunting. Once the PVR has found its lowest point during the first 6 weeks of life, symptoms of CHF become evident. In newborns and infants, these symptoms include tachypnea and diaphoresis with feeds, failure to thrive, and tachycardia.

If the defect is large and left untreated, the patient will eventually develop pulmonary vascular changes that will translate into a high PVR with eventual reversal of shunting and development of cyanosis. The development of irreversible pulmonary vascular disease (Eisenmenger syndrome) is variable but may happen early, especially in patients with genetic syndromes such as trisomy 21.

The physical exam findings relate to the amount of shunting, the size of the defect, the relative pulmonary and systemic vascular resistances, and the presence of secondary abnormalities such as MR (from significant left-heart dilation) or AI (from prolapse of the aortic valve into the defect). The diagnosis of a VSD is usually made prenatally during echocardiographic screening or after birth when a murmur is detected. Children

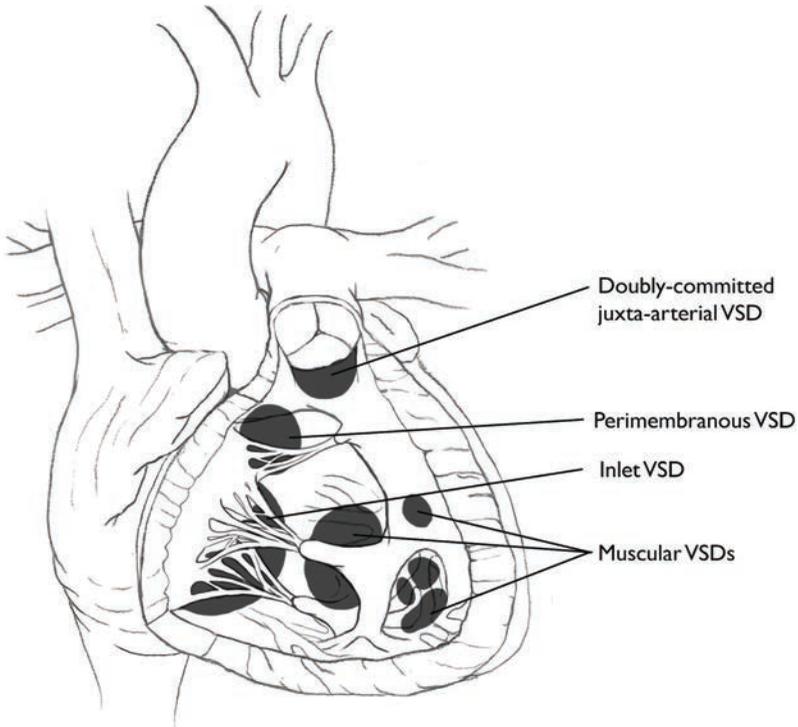


Figure 11-1. Anatomic variants of VSD based on location.

with significant volume overload will present with respiratory distress, tachypnea, and in many cases with emesis (usually from hepatomegaly-associated compression of the stomach).

Usually, there is a hyperdynamic LV impulse and a harsh holosystolic high-frequency murmur at the left lower sternal border. The murmur is well heard in the back. When PA pressures are normal, the second heart sound (S_2) will be split with a normal pulmonary component (P_2). When PA pressures are elevated, the S_2 will be narrowly split or single, with a loud P_2 . The frequency of the murmur depends on the pressure drop between the LV and RV. A low-frequency diastolic rumble (absence of silence during diastole) heard at the left lower sternal border usually means that there is at least 2:1 shunting from left to right. At times, an S_3 gallop may be heard at the apex. A high-frequency end-diastolic murmur will be a sign that there is concomitant AI.

Diagnosis

- **ECG (Figure 11-3).** There can be LV and sometimes RV hypertrophy and occasional atrial enlargement.
- **CXR (Figure 11-4).** Helpful in the initial evaluation and follow-up of children with VSDs, particularly while titrating diuretics or when trying to establish the nature of

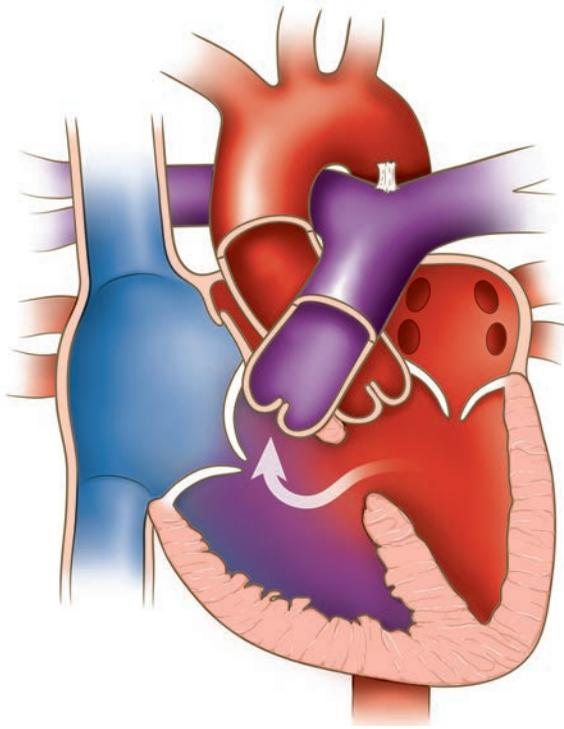


Figure 11-2. Pathophysiology of VSDs with left-to-right shunting.

their respiratory distress. It tends to show cardiomegaly and increased pulmonary vascular markings due to left-to-right shunting.

- **Echocardiogram (Figure 11-5).** Mainstay of diagnosis. It is important to obtain complete sweeps to demonstrate the location and size of the defect, measure chamber dilatation (atrial and/or ventricular), profile MR, and interrogate the aortic valve for potential prolapse. It is also important to obtain additional imaging to demonstrate the Doppler velocity of the defect to indirectly estimate the size of the defect and the PVR. In older children, with long standing defects, the presence of a double-chamber RV should be investigated.
- **Cardiac catheterization.** Rarely needed. However, in patients who present late, it may be indicated to assess PVR for suitability of VSD closure and/or the need for pulmonary vasodilator therapy.

Medical Management

Loop diuretics are effective at mitigating congestion symptoms and allowing for better feeding in patients with VSDs. Furosemide 1-2 mg/kg/dose scheduled up to every 6 hours tends to be enough to manage moderate-to-large defects. Chlorothiazide 5-10 mg/kg/dose given twice a day can be a helpful adjuvant in cases of significant volume loading or when there is some degree of diuretic resistance. Spironolactone in doses of 0.5-1 mg/

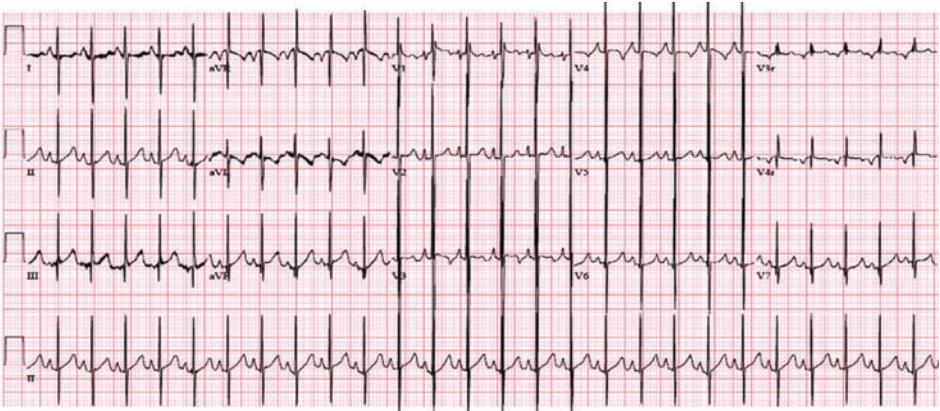


Figure 11-3. ECG on a patient with VSD demonstrating right atrial enlargement and biventricular hypertrophy.

kg/dose given twice a day can help with potassium sparing and mitigate loop diuretic resistance. Afterload reduction with angiotensin-converting enzyme (ACE) inhibitors can also be helpful by decreasing SVR and potentially reducing left-to-right shunting.

Large perimembranous VSDs can be initially managed with diuretics and ACE inhibitors. If there is no aortic valve prolapse on echocardiography and the child is gaining weight, medical management can be continued. If the S_2 becomes single and there is rapid improvement of CHF signs with weight gain and no change on the size of the defect on echocardiography, one should suspect increased PVR. In these cases, early surgical intervention should be undertaken, or if later in life, a cardiac catheterization should be considered to evaluate pulmonary vascular reactivity.

Muscular VSDs tend to close spontaneously by 2 years of age. As such, conservative management is usually indicated, unless the defects are large and the patient fails medical management. Patients with multiple apical muscular VSDs (“Swiss-cheese” septum) can have significant shunting and placement of a PA band may be necessary to control CHF symptoms. It is thought that Swiss-cheese VSDs may be a part of the LV non-compaction spectrum and if LV function decreases, cardiomyopathy should be suspected.

Indications / Timing for Intervention

The vast majority of VSDs (80%) will close spontaneously. However, some defects should be repaired in order to prevent long-term complications such as AI, endocarditis, and development of pulmonary vascular disease. In asymptomatic patients requiring intervention, repair is usually delayed until later in infancy.

Indications for intervention include:

- **Failure of medical management.** Patients with large VSDs and symptoms resistant to medical management are usually repaired in infancy.
- **Left-heart dilation.** VSDs that have a large left-to-right shunt as manifested by



Figure 11-4. CXR on an 8-week-old patient with a large and symptomatic VSD, in addition to prematurity-associated lung disease. The CXR shows cardiomegaly with increased pulmonary vascular markings.

left-heart dilation may benefit from intervention if they have not closed spontaneously after infancy.

- **DCJA defects.** DCJA defects are associated with aortic valve prolapse and virtually all patients will develop AI before childhood or adolescence. In addition, these defects tend to not close spontaneously.
- **Aortic valve prolapse.** With or without AI.

The likelihood of spontaneous closure decreases with age and is <10% in VSDs that are patent beyond 2-3 years of age. True inlet VSDs rarely close spontaneously; strong consideration should be made for early intervention.

Catheter-Based Intervention

Device closure can be performed in patients with muscular VSDs, either as primary therapy (Figure 11-6) or as an adjunct to surgical closure, as some muscular VSDs may not be able to be closed surgically due to the difficulty in distinguishing them from trabeculations on the RV side. The procedure frequently involves the creation of an arteriovenous loop to facilitate delivery of the VSD device from the venous side. Retrograde deployment of VSD devices can also be performed in some instances without the creation of an arteriovenous loop. Transesophageal and transthoracic echocardiography provide vital imaging guidance during the procedure.

In some very small patients, or when a concomitant operation is being performed

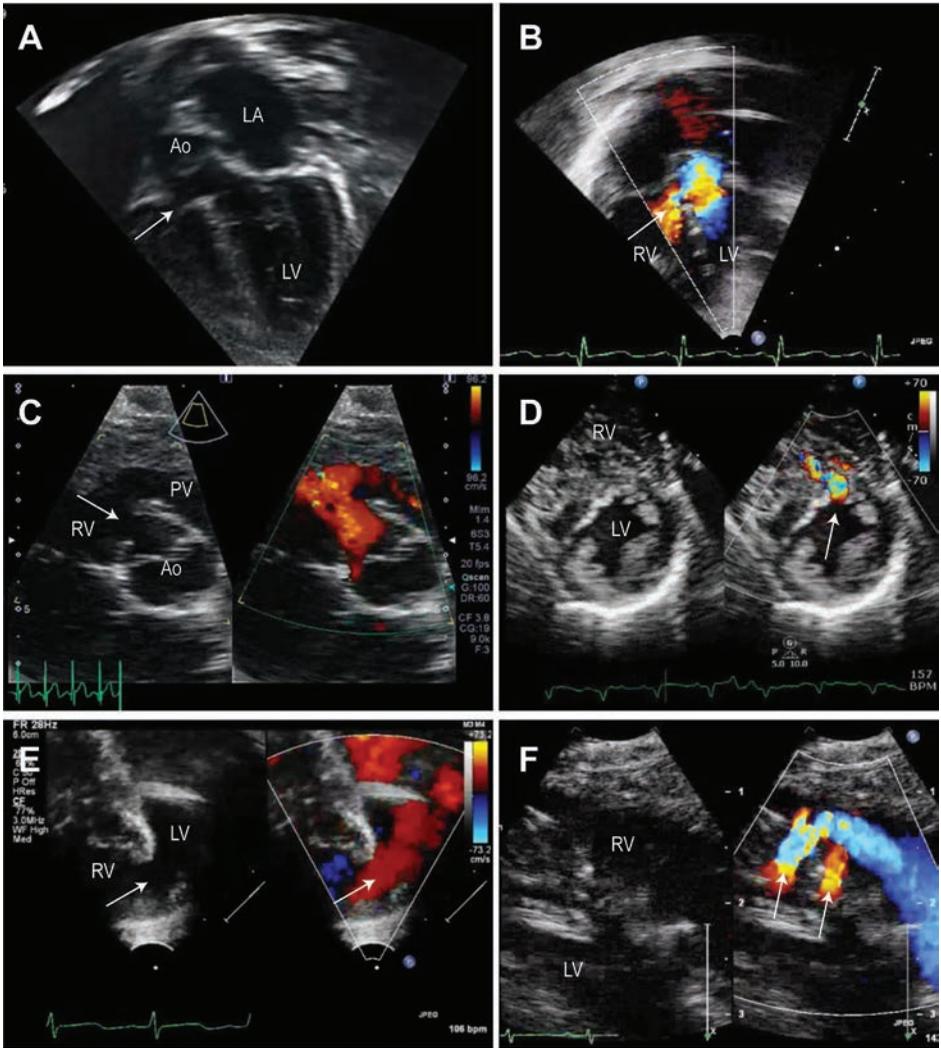


Figure 11-5. Echocardiography for diagnosis of VSD. A) 5-chamber view of a perimembranous VSD (arrow). B) 5-chamber view with color Doppler of a perimembranous VSD (arrow). C) Parasternal short-axis color-compare view of a large perimembranous VSD (arrow). D) Short-axis color-compare view of a mid-muscular VSD (arrow). E) 4-chamber color-compare view of a large apical muscular VSD (arrow). F) Parasternal long-axis color-compare view of several mid-muscular VSDs (arrows). Ao: aorta, PV: pulmonary valve. Images courtesy of Dr. Josh Kailin, www.pedecho.org.

(e.g., PA band takedown), a hybrid approach can be used (direct “perventricular” VSD closure) in the OR or cardiac catheterization laboratory with a surgeon and an interventional cardiologist.

Patients with perimembranous VSDs are currently not suitable candidates for

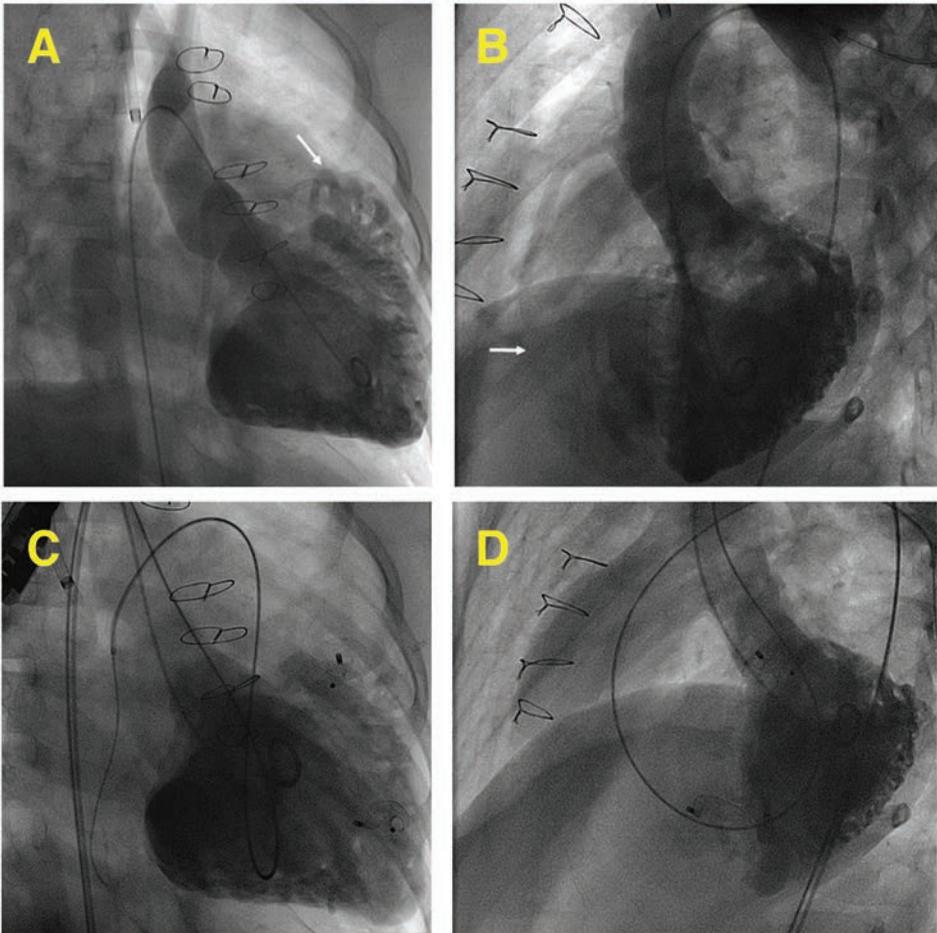


Figure 11-6. LV angiogram in a 6-year-old girl with multiple muscular VSDs. Left-to-right shunting via multiple defects in the anterior muscular septum (arrow) can be seen on a right anterior oblique/caudal projection (A). Significant shunt via apical/mid muscular VSDs to the body of the right ventricle (outlined by arrow) is also seen on the left anterior oblique/cranial projection (B). After placement of percutaneous devices in the anterior and apical muscular septum, a significant reduction in left to right shunt is seen in the corresponding post procedure angiograms (C and D).

catheter-based closure of their defects in general, due to the proximity of the defect to the AV node (with risk of heart block after device closure), and the tricuspid and aortic valves. Exceptions are patients with perimembranous defects and aneurysmal tricuspid valve tissue, which may allow a device to be placed within the defect(s), away from the AV node and surrounding valves.

Surgical Repair

Surgical repair of VSDs is performed using standard aorto-bicaval cannulation for CPB and mild-to-moderate hypothermia. Most perimembranous, inlet, and muscular defects are approached through a right atriotomy. If there is significant aneurysmal tricuspid valve tissue covering a perimembranous or inlet VSD, the tricuspid valve may be partially detached to allow full visualization of the defect. VSDs are usually repaired using glutaraldehyde-treated autologous pericardium and either interrupted pledgeted sutures or a combination of running and interrupted pledgeted sutures. In the case of perimembranous or inlet VSDs, the bundle of His travels on the posteroinferior margin of the defect, mainly towards the LV side. Care is taken to place sutures superficially and away from the rim of the VSD. Small defects may be closed primarily with interrupted double-pledgeted sutures.

DCJA defects are approached through a transverse pulmonary arteriotomy. Interrupted sutures with pericardial pledgets are placed through the pulmonary valve annulus and into the rim of the defect. A pericardial patch is then used to close the VSD. If there is no perimembranous extension, the conduction system is away from the VSD on these patients.

Apical defects and those muscular defects below the moderator band can be difficult to visualize through an atriotomy and alternative approaches should be considered (cath intervention vs. ventriculotomy). In some patients with multiple VSDs (i.e., Swiss-cheese septum), early closure is challenging and placement of a PA band may allow symptom control and growth. These patients can be brought back to the OR later in life for PA band takedown and VSD repair; some patients may require cath closure of the VSD at the same time.

Postoperative Management

Older patients are usually extubated in the OR. Neonates and small infants tend to be left intubated and are then extubated within the first few hours of arrival to the CICU. These patients have usually a very uncomplicated postoperative course where the most common event to manage is hypertension secondary to a hyperdynamic LV.

In patients with significant CHF symptoms preoperatively, optimization of preload, contractility, and afterload is key. These patients are commonly managed with a milrinone infusion to optimize cardiac output, and a tight fluid control, including diuretic treatment on postoperative day 1 to prevent and improve pulmonary congestion. Extubation within 24 hours of surgery is expected.

It is not unusual for the TEE to show mild-to-moderately depressed LV function after repair of a large VSD, usually due to volume unloading of the heart. This decreased function tends to have no clinical significance and is usually improved a few days after repair.

Patients tend to stay in the CICU 1-2 days. Patients that had significant overcirculation preoperatively tend to show dramatic improvements in oral intake, tachypnea, and weight gain immediately after surgery.

Complications

Potential complications after surgical closure of VSDs include:

- **Heart block.** Variable or complete heart block may complicate surgical VSD repair due to direct injury to the AV node or the bundle of His. If the patient remains in complete heart block 7-10 days after surgery, consideration should be made to placing a permanent pacemaker.
- **Arrhythmias.** Arrhythmias, such as JET, may develop in small children, likely from traction of the conduction system during repair.

TCH experience with VSD repairs (Scully et al. 2010)

Median number of repairs per year: 29 (22 – 43)

Median age at surgery: 10 months (20 days – 18 years)

Median ICU length of stay: 2 days (1 – 14 days)

Median hospital length of stay: 5 days (2 days – 6 months)

Perioperative mortality: 0.5%

Bleeding requiring reoperation: 1%

Perioperative complete AV block: 0

Suggested Reading

Scully BB, Morales DL, Zafar F, et al. Current expectations for surgical repair of isolated ventricular septal defects. *Ann Thorac Surg* 2010;89:544-9.