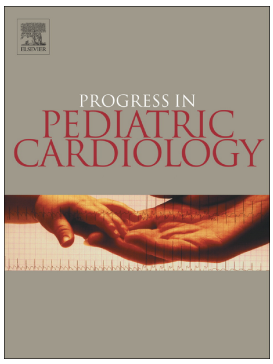

Echocardiographic assessment of ventricular septal defects

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Title: Echocardiographic Assessment of Ventricular Septal Defects.

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Abstract: A ventricular septal defect (VSD) is one of the most common congenital cardiac lesions. Echocardiography is the primary mode for evaluation of ventricular septal defects. Echocardiographic evaluations of ventricular septal defects are easily reproducible, can be done in a timely fashion and do not require a complex set up. They give detailed information needed for the diagnosis, treatment and long term follow up. Although diagnosis can now be made prenatally, a more detailed evaluation with confirmation of the diagnosis is not made until after birth with transthoracic echocardiography. With a combination of 2D, color Doppler and spectral Doppler, the VSD location, size and hemodynamic components can be established. Additionally, sequelae from the VSD and other associated congenital

cardiac lesions can also be identified. A review of the standard components of a thorough transthoracic echocardiogram to evaluate the different types of ventricular septal defects will be discussed in this article.

Keywords: Ventricular, septal, defect, shunt, echocardiography, congenital.

Introduction: Ventricular septal defects (VSDs) are among the most common congenital cardiac lesions, comprising up to 40% of all congenital heart defects (1). They can occur in isolation but more commonly are associated with other complex heart defects. The reported incidence of an isolated ventricular septal defect in live births varies significantly. It is estimated that in the United States about 1 in every 240 live births have a ventricular septal defect (2). It is important to note that its frequency is dependent on age at examination and diagnostic technique, as some defects will close spontaneously during the first year of life. It is also important to note that most of those are asymptomatic. Overall, utilizing echocardiography, VSDs have a prevalence of up to 3.94 per 1000 individuals (3). Echocardiography remains as the mainstay for the diagnosis and follow up of VSDs, providing a comprehensive evaluation of the anatomy and associated physiology.

Anatomy and nomenclature of VSDs: There are several nomenclatures (Table 1) that are currently being used in classifying VSDs (4) (5) (6). In this article, the unified classification for VSDs by the International Society for Nomenclature of Paediatric and Congenital Heart Disease will be utilized (5). This has been accepted by the World Health Organization into the 11th Iteration of the International Classification of Diseases and as follows:

1. Perimembranous central VSD
2. Inlet VSD without a common AV junction
    a. Without AV septal malalignment
    b. With AV septal malalignment
    c. Inlet muscular
3. Trabecular muscular VSD

   a. Mid septal
   b. Apical
   c. Postero-inferior
   d. Antero-superior
   e. Multiple (Swiss cheese)
4. Outlet VSD

It is essential to have a thorough understanding of the anatomy of the interventricular septum when evaluating VSDs (Figure 1).

The interventricular septal anatomy viewed from the right ventricular aspect does not correspond to the left ventricular aspect. VSDs which traverse more than one portion of the septum are named to reflect the location of the majority of the lesion followed by its extension into the adjacent segment of the interventricular septum.

It is important to visualize all segments of the interventricular septum, keeping in mind that it is a curved structure which cannot be fully studied in a single plane or from a single window.

The right ventricular aspect of the interventricular septum can be divided into four main regions: the inlet, trabecular, membranous and outlet (Figure 1). The inlet septum lies infero-posteriorly and extends from the tricuspid valve septal leaflet hinge point, down to the distal tricuspid valve septal attachments. The trabecular septum is a coarse muscular structure which extends inferior to the inlet septum. The outlet septum lies superior to the trabecular septum, with the septal band separating the two. The aortic valve is located posterior and to the right of the outlet septum. The membranous septum is a thin fibrous structure with two components, the interventricular and atrioventricular, separated by the septal leaflet of the tricuspid valve. It is surrounded superiorly by the outlet septum, inferiorly by the trabecular septum, and posteriorly by the inlet septum.

The outlet septum, also known as the conal septum, is located between the two limbs of the septal band on the right ventricular side. When the outlet septum is muscular, the resulting VSD is a malalignment type. When the outlet septum is fibrous or absent then the VSD is a doubly committed outlet VSD (6).

A malalignment of the outlet septum with the muscular septum can create a VSD that may involve the membranous septum which is then referred to as a malalignment VSD (Image 1). The defect created by the malalignment can be either from an anterior or posterior malalignment of the outlet septum. In the case of an anterior malalignment, the resulting VSD is that seen in Tetralogy of Fallot, which may result in narrowing of the right ventricular outflow tract and overriding of the aorta. This is a result of the anterior deviation of the outlet septum. In posterior malalignment of the outlet septum, the VSD created is usually associated with interrupted aortic arch and narrowing of the left ventricular outflow tract (7).

Echocardiography:

There are several key components of a VSD evaluation with echocardiography. These include:

- Exact location and margins

- Size and number of VSDs

- Relationship to neighboring structures.

- Hemodynamic/physiologic assessment of shunt including flow assessment by color and spectral Doppler evaluation.

- Evaluation of associated lesions.

The evaluation can be accomplished via a systematic approach. It is crucial to identify the location of the VSD and its relationship to other cardiac structures, such as the valves and valve apparatuses. The ventricular septum should be thoroughly inspected from several windows and in multiple planes. It requires both 2-dimensional and color sweeps to ensure that it is evaluated in its entirety. The sweeps need to cover the septum from apex to base; and from anterior to posterior. The ventricular septum

presents a challenging configuration with a curved shape which requires imaging in several windows and planes to adequately cover all its segments. Ideally, the septum is visualized with the ultrasound beam perpendicular to it, and with the beam parallel to the flow across the defect.

Advancing technology has added significantly to the information obtained by echocardiography. In the majority of cases, a transthoracic echocardiogram is sufficient for obtaining an accurate diagnosis using 2 dimensional imaging (2D), M-mode, color Doppler, and spectral Doppler (continuous and pulsed wave) (8).

Transthoracic Imaging:

The four main types of VSDs are perimembranous, inlet, muscular and subarterial (Figure 2).

1. Perimembranous VSDs: These constitute the most common type of VSD, about 80%. They are located in the membranous region of the IVS, at the intersection of the trabecular, inlet or outlet regions. In the parasternal short axis views at the level of the aortic valve, these lesions are located between 9 and 11 o'clock, with anterior extension showing between 11 and 12 o'clock. (Image 2a) In the parasternal long axis view they can be visualized just below the aortic valve. In the apical five chamber view, they are again seen below the aortic valve (Image 2b). In the subcostal coronal and sagittal views, these defects can be well seen using similar landmarks. Perimembranous defects are also adjacent to the tricuspid valve, beneath the septal leaflet, which can be seen in the parasternal short axis views of the left ventricular outflow tract. The tricuspid valve plays an important role in the closure of this type of VSD through apposition of its septal leaflet with the VSD. Sometimes this apposition creates an aneurysm of the interventricular septum which can be visualized in the parasternal long and short axis views, as well as subcostal and apical views. Associated lesions like a double chambered right ventricle (Image d), left ventricle-to-right atrial shunt, or a sub-aortic ridge may be seen (9) (29). Due to high velocities

from the VSD in proximity to the aortic valve, the right or non-coronary leaflets may prolapse into this type of VSD (Venturi effect) resulting in aortic regurgitation (Image 2c and 2e) (30).

2. Inlet VSDs without a common atrioventricular valve junction: These defects are located below the medial papillary muscle, postero-inferior limb of the septal band and antero septal commissure of the tricuspid valve. It is further subdivided into: a. without atrioventricular septal malalignment and without a common atrioventricular junction; b. with atrioventricular septal malalignment and without a common atrioventricular junction; c. inlet muscular VSD (5) (10) (11). These defects are best viewed from the apical four chamber view but can also be seen in the parasternal short axis views. Any atrioventricular valve straddling and/or override must be identified and evaluated. When associated with an atrioventricular septal defect, a cleft is often seen in the mitral valve anterior leaflet (12). The tricuspid valve can also contribute to the closing of an inlet VSD.
3. Muscular VSDs: These can be located in any region of the trabecular septum and are surrounded by muscular borders. They can be located in the anterior muscular (anterior to the septal band), mid-muscular (posterior to the septal band), posterior (below the septal leaflet of the tricuspid valve) or in the apical trabecular regions. Muscular VSDs distal to the moderator band are classified as apical muscular VSDs (Image 3c) (10). Figure 2 demonstrates these landmarks. The exact location of a muscular ventricular septal defect is determined after imaging from several views, as it can be challenging to sort out the subtypes of muscular VSDs. Adequate sweeps in the parasternal, subcostal and apical views are needed to evaluate the corresponding segments of the interventricular septum. A parasternal short axis sweep towards the apex of the heart can demonstrate anterior muscular VSDs between 11 and 12 o'clock (Image 3a), mid-muscular VSDs between 10 and 12 o'clock and posterior muscular VSDs between 7 and 10 o'clock (Image 3b). Muscular VSDs often exist as multiple lesions in one area resulting in a "Swiss cheese" appearance of the interventricular septum (20).

4. Outlet VSDs: These are among the least common types of VSDs, constituting about 5-10% of VSDs. These are more commonly seen in the Asian population. These defects can be present with or without malalignment between the outlet septum and the apical part of the muscular septum. Furthermore, these defects can be further subdivided into: a. outlet perimembranous defects, b. outlet muscular defects and c. doubly committed juxtaarterial with a muscular or fibrous posteroinferior rim (5).

   In the parasternal short axis view at the level of the aortic valve, these defects are seen between 12 and 2 o'clock (Image 4). These defects can also be demonstrated in the parasternal long axis views with slight counterclockwise rotation of the transducer. They are immediately adjacent to the aortic and pulmonary valves resulting in fibrous continuity of the two valves (13) (14) (10). A very common complication of a subarterial defect is distortion and often prolapse of the unsupported right aortic leaflet into the defect (25). This has been reported in up to 70% of patients with subarterial VSDs. The leaflet prolapse may range from mild to severe; with or without aortic valve regurgitation. The prolapse may sometimes be associated with tethering of the right aortic leaflet to the VSD (11). It is generally recommended that subarterial VSDs be surgically repaired to avoid the development of this complication over time (19).

Size

The size of a VSD should be evaluated in several views as it may have different dimensions in different planes (oval defects). This is also due to the complex configuration of the interventricular septum. The two-dimensional (2D) defect size is generally determined by comparison to the aortic annulus, with small lesions measuring less than 25% of the aortic diameter, moderate defects 25-75% and large defects being larger than 75% of the aortic annulus (31). It is important to keep in mind that the size of a VSD may vary throughout the cardiac cycle. Some defects may also have multiple openings on the RV side, making a simple diameter measurement impossible. This is where the shunt evaluation adds to the information obtained regarding the effective size and hemodynamic significance of the lesion.

Hemodynamic assessment

This is a very important component in the evaluation of a VSD; it provides important information that helps predict the clinical course of a VSD. It additionally facilitates management decisions (7).

The left to right shunt from a VSD (seen with color and spectral Doppler) will result in increased pulmonary blood flow producing left atrial and left ventricular dilation. In addition to M mode, left atrial and ventricular volumes can be measured using Simpson's method or 3D imaging. This is particularly important as M mode measurement may not be accurate in the presence of a flattened interventricular septum, which can be seen with large VSDs. Significant left heart dilatation may also result in mitral annular dilation with mitral regurgitation.

Although with its limitations in the pediatric echocardiogram setting, conventionally, the amount of shunting is measured using the ratio of pulmonary-to-systemic blood flow (Qp/Qs) that can be calculated (Qp/Qs) as follows:

Qp/Qs=RVOT VTI x RVOT area/ LVOT VTI x LVOT area.

(*RVOT*, Right Ventricular Outflow Tract. *LVOT*, Left Ventricular Outflow Tract. *VTI*, Velocity Time Integral).

It is important to know that variations in these measurements can occur with minor differences in the measurements of the outflow tract diameters which can be a source of error in the final calculations (12)(22).

- Right ventricular systolic pressures (RVSP): Right ventricular function may be affected as the pulmonary artery pressures increase in untreated VSDs

With increasing right ventricular systolic pressure, septal flattening in systole can be seen. RVSP is usually obtained by using the Bernoulli equation as follows: measurement of the pressure gradient across the TV (TR jet peak gradient) plus the right atrial pressure:

RVSP= 4V TR^2 + Right atrial pressure.

Right ventricular systolic pressure can also be obtained by Doppler interrogation of a VSD when the angle of Doppler interrogation is appropriate (13) (18).

The velocity across a VSD can be obtained by spectral Doppler (Image 5). Ideally, the Doppler pattern across the VSD peaks in mid systole, to avoid over estimation of the VSD gradient. However, it is also important to note that VSD gradient may be underestimated in some cases, based on the angle and location of spectral Doppler sampling.

Furthermore, by measuring the pressure gradient across the VSD using the Bernoulli equation (4V VSD^2), the right ventricular systolic pressures can be calculated by subtracting the VSD pressure gradient from the systolic blood pressure (SBP): RVSP= SBP- (4V VSD^2). It is important to note that a simultaneous measurement of the systemic blood pressure should be obtained for an accurate value. Untreated hemodynamically significant VSDs result in severe increase in right ventricular systolic pressure resulting in a right to left shunt across the VSD (Eisenmenger syndrome).

It may also be challenging to discern accurate VSD gradients in the setting of a double chambered right ventricle as it may be confounded by the obstruction in the right ventricular outflow tract.

Perioperative imaging:

It is crucial that the exact location, size and number of VSDs is clarified prior to any surgical repair. The surgical approach to the VSD is planned based on the VSD location. It is usually helpful to show the relationships and distances from adjacent structures. Any preoperative valvar regurgitation should be evaluated and included in the planning of the repair. A distorted aortic valve leaflet, for example, may need to be addressed and repaired, in addition to the VSD (13)(14)(27).

Transesophageal echocardiography (TEE) is used in the operating room prior to surgery, to confirm the diagnosis and to obtain any possible additional information. During surgery a transesophageal approach can be used to provide information about the repair prior to chest closure (21).

Post operatively, with the surgical approach in mind, the imaging must provide detailed information regarding any residual lesions (additional VSDs or patch leaks) (26), adjacent valvar regurgitation or new valvar regurgitation. The tricuspid valve, for example, may be damaged and may develop regurgitation post operatively, mostly after repair of a perimembranous or malalignment VSD. The aortic valve may be affected following repair of a subarterial or perimembranous VSD (7). This can either be in the form of aortic leaflet distortion or perforation of the leaflet with a suture during repair. In some cases, additional VSDs are newly visualized postoperatively after surgical closure of a large VSD, with the smaller VSDs being obscured preoperatively. This is due to the systemic right ventricular pressures produced by the larger VSD. It is not uncommon to see residual VSDs after repair of a muscular VSD due to the coarse trabeculations in that region of the septum. It is important to evaluate gradients across the left ventricular outflow tract after closure of a VSD as the VSD may mask the gradient across the aortic valve. Small residual lesions are, however, well tolerated and mostly resolve over time (14).

An intramural VSD is a unique type of VSD which can present in the post-operative period. This is usually a defect seen after repair of a conotruncal lesion and can be challenging from an imaging perspective. It is located anterior to the VSD patch between the great artery where the patch is attached, and the right ventricular trabeculations (Figure 3) (28). It is particularly difficult to visualize intraoperatively especially as the channels through the muscular trabeculations usually dilate over time, resulting in a significant left to right shunt which requires re-operation. This is best demonstrated on subcostal views on transthoracic echocardiogram and deep transgastric on transesophageal echocardiogram.

Additionally, it is important to assess ventricular function after surgical repair. And in some cases, a follow up study may demonstrate a VSD patch dehiscence.

The goals in perioperative evaluation of a VSD can be summarized in Table 2.

Transcatheter Device Closure:

In the catheterization laboratory, echocardiography is used to describe the location, size, rims and relationships to adjacent structures. This information is key in determining whether the VSD is amenable to device closure and if so, assists with choosing the appropriate device to be used. In addition, during the procedure, echocardiography assists in determining the location and appropriateness of the device closure (17). Key points to determine after device deployment are the presence or absence of a residual shunt, any impingement of the device on adjacent structures, ventricular function and the presence of any pericardial effusion.

Three-Dimensional Echocardiography (3D):

This type of imaging provides additional information which may be important in the pre-surgical assessment, intraoperatively, during device closure, or in the post-repair period. In the past decade 3D transducers have evolved with high 3D resolution, a small footprint and higher frequencies which allow for 3D imaging of small infants with higher heart rates (23). 3D imaging can be obtained via the transthoracic or transesophageal approach. 3D echocardiography can be especially helpful in visualization of VSDs with unusual shapes, hence obtaining accurate information regarding the location and the dimensions of the defect, which may be difficult to see en face by 2D echocardiography. The interventricular septum can be visualized from the right ventricular or left ventricular sides, after cropping away the free walls of the ventricles in post-processing. This can be done in the axial plane, using live 3D or ECG-gated full volume acquisition. With 3D imaging it is possible to visualize the interventricular septum in its entirety (12). Different imaging windows may be needed to optimally view different portions of the septum given its curved shape. Windows including apical, low parasternal, or subxiphoid can be used. 3D imaging is superior in visualizing the borders or rims of a VSD (12). This is crucial and particularly helpful in guiding the decision as to the size and type of VSD device to be used in the

catheterization laboratory. Accurate visualization and measurement of the rims of a VSD is important to avoid compromise and potential damage of the aortic or tricuspid valves (28). One limitation to 3D imaging is the "dropout" of segments of the interventricular septum, giving the false appearance of a defect.

Transesophageal echocardiography (TEE):

A transesophageal echocardiogram is predominantly utilized intraoperatively or during device closure of VSDs. It can help detect a residual VSD which may require going back on bypass to be repaired (14). Transesophageal echocardiogram is also used to evaluate new valve regurgitation and ventricular function after repair of a VSD (24). It is important to remember that an under-filled ventricle can appear to have poor function, thus appropriate filling of the ventricles is needed for proper interpretation of function by transesophageal echocardiogram (14).

In the catheterization laboratory TEE can provide information regarding device position, residual lesions, encroachment of the device on nearby structures or any flow abnormalities created by the device.

The septum can be viewed starting in the trans-gastric short axis view at the apex, with some anteflexion and left lateral deflection, with approximately 20 degrees of head rotation. From this position the probe can be gently withdrawn until the entire septum is visualized. A four chamber view can then be obtained through retroflexion of the probe. The anterior and posterior portions of the septum can then be visualized by ante or retro-flexion, to view the corresponding segments of the interventricular septum. A long axis view (usually 110 degrees) can then be obtained from a low esophageal view, scanning right to left to view the inlet then outlet portions (27). Limitations of transesophageal echocardiogram for VSD evaluation include the limited windows, restricting the possible angles for spectral Doppler interrogation.

In addition, it is important during the post-surgical period to rule out intracardiac or myocardial air. Air is usually seen anteriorly in the chamber, or alongside the chamber walls or interventricular septum.

Myocardial air is seen as a localized echogenicity in the myocardium accompanied by dysfunction of the affected myocardium.

SUMMARY

Ventricular septal defects (VSDs) are among the most common congenital heart defects. Echocardiography is currently the primary tool used in the evaluation of VSDs. Anatomic and hemodynamic assessment of VSDs using the different components of transthoracic and transesophageal echocardiogram (2 dimensional imaging, color Doppler, spectral Doppler and 3 dimensional imaging) play an important role in the diagnosis, management and long term follow up of patients with VSDs.

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Table 1.

International Society for Nomenclature of Paediatric and Congenital Heart Disease	Congenital Heart Surgery Nomenclature and Database Project (CHS)	Van Praagh Classification	Anderson Classification

Perimembranous central	Perimembranous	Conoventricular	Perimembranous,
Inlet VSD without a common AV junction	Inlet	"Atrioventricular Canal type defect"	Perimembranous, Inlet outlet
Trabecular Muscular	Muscular	Muscular	Muscular
Outlet	Subarterial	Conal septal defects	Juxta-arterial

Commonly used classifications of ventricular septal defects.[4, 5, 6]

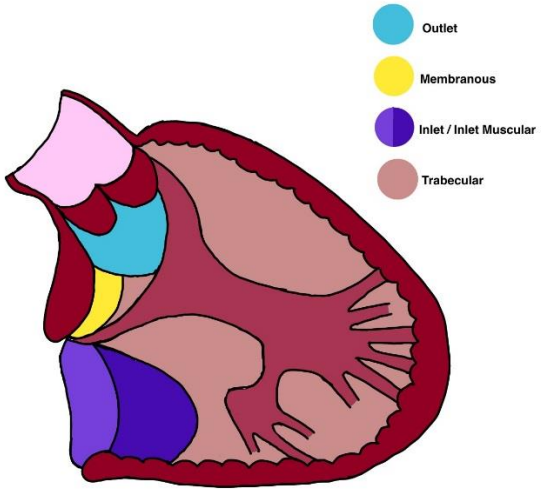

Figure 1. Different regions of the interventricular septum as viewed from the right ventricle. (*Original illustration © by Reena Turpin)*

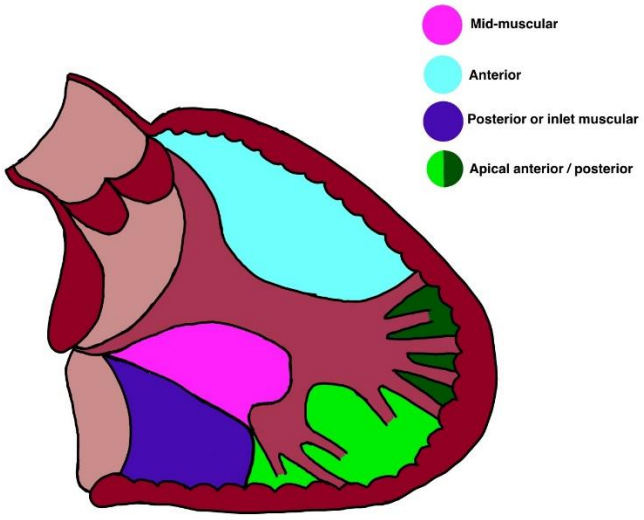

Figure 2. Muscular ventricular septal defects as viewed from the right ventricle. (*Original illustration © by Reena Turpin*).

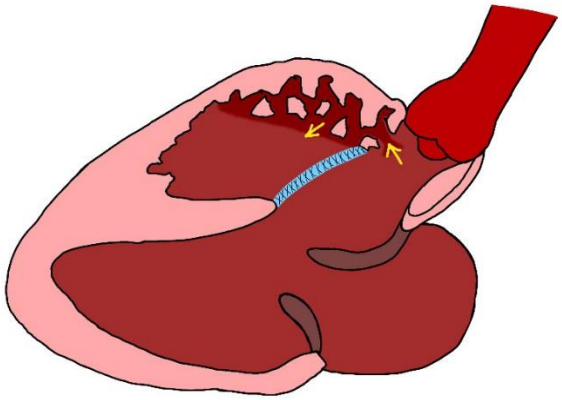

Figure 3. Intramural ventricular septal defect (arrows) after surgical patch repair. (*Original illustration © by Reena Turpin.)*

Table 2

Preoperative assessment	Post-operative assessment
Location, Type, Size and Number of VSDs	Residual or additional VSD
Adjacent valvar regurgitation or stenosis	Adjacent valvar regurgitation or stenosis

Associated lesions	Associated lesions
Left atrial size; Left ventricular size and function	Left ventricular size and function
Estimated right ventricular pressure	Estimated right ventricular pressure
Left outflow tract obstruction	Left outflow tract obstruction
Right outflow tract obstruction	Right outflow tract obstruction
	Pericardial effusion

Perioperative VSD evaluation checklist

Declarations of interest
None.

Highlights:

- Ventricular septal defect (VSD) is the most common congenital cardiac lesion.
- Echocardiographic evaluations of ventricular septal defects are easily reproducible, can be done in a timely fashion, do not require a complex set up and give detailed information needed for the diagnosis, treatment and long term follow up.
- Transthoracic, transesophageal and 3D echocardiogram with 2D, color and spectral Doppler can accurately determine the VSD's location, size, its hemodynamic components, sequelae and other associated congenital cardiac lesions.