Ventricular septal defects (VSDs) are the commonest group of congenital heart lesions, accounting for \( \approx \)30% of congenital heart defects in liveborn infants.\(^4\) Defects in the ventricular septum are classified according to their position when the ventricular septum is viewed from the right ventricle and these may vary in their location, size, and number.\(^2,3\) VSDs can occur as isolated lesions or as part of more complex disease, for example, transposition of the great arteries or double outlet right ventricle, where the location and size of such defects assume great importance in planning the optimal mode of surgical repair.\(^4\) In most cases, surgical closure remains the mainstay treatment for VSDs, but selected lesions may be amenable to catheter closure.\(^5,6\)

Cross-sectional echocardiography is the most commonly used noninvasive diagnostic method to delineate the position of a VSD before surgery.\(^7\) Multiple sonographic cuts and cross-sectional sweeps of the region of interest are used to build a virtual reconstruction of the three-dimensional (3D) anatomy in the mind of the echocardiographer. However, for the surgeon who deals solely with 3D images there can remain an element of uncertainty when called to operate on the patient. The number, size, location of VSDs, and their relationship to adjacent structures are central for selecting whether the defects might be best closed via the tricuspid valve, via the semilunar valves, or via a ventriculotomy or whether these might not be readily approachable and a palliative pulmonary artery banding is desirable.\(^8,9\)

In this review, we present key 3D echocardiographic projections and examples of different types of VSDs where the images have a direct effect on the approach taken to VSD closure. Comparison of 3D echocardiography (3DE) with other 3D imaging modalities is also described. Images are presented in an intuitive anatomic format, which is consistent with other cardiac imaging modalities, such as MRI and angiography.\(^10\)

Echocardiographic Methods

Echocardiographic images were obtained either by transthoracic or transesophageal echocardiography using the Philips IE33 ultrasound system (Philips Medical Systems, Andover, MA). Probes used for acquisition included the X7-2, X5-1, X3-1, and 7-2t ultrasound probes. Each probe houses a matrix array system, which permits either live 3D echocardiographic imaging or acquisition of a volume of ultrasound data \( \geq 1 \) to \( \geq 6 \) cardiac cycles. Postprocessing was performed using Qlab V8-9 software either on cart or on a Philips Xcelera workstation. This software permits recropping of data in any desired plane in either a 3D rendered mode or multiplanar reformatted mode as is highlighted in Movie I in the Data Supplement.

For transthoracic studies in young children, a subcostal approach was most commonly preferred. The rationale for this approach is that the ultrasound beam is approximately orthogonal to the ventricular septum and that the area of interest is in the middle of the sonographic field. Anticlockwise rotation of the probe to permit a cross-sectional reference view most akin to that used to visualize the right ventricular outflow tract has been most effective in our experience. Adjustment of the lateral width and depth of image on cart permits inclusion of relevant landmarks, such as the tricuspid valve, outflow tracts, and the diaphragmatic surface of the heart. Particular care is necessary to include the true apex of the heart where defects in this region are suspected. If the patient is cooperative, then breathhold and lack of patient motion normally permit acquisition of a good quality full-volume acquisition. For patients who are ventilated, this is briefly suspended during acquisition. If the patient is moving during the examination, live 3D echocardiographic modes are preferred to avoid the potential for stitch artifact. In older children and in adults, the subcostal view can be challenging and the apical four-chamber view or parasternal long- and short-axis views are commonly used. The apical view remains limited because the ultrasound beam is parallel to the ventricular septum, but in most cases muscular VSDs can be well visualized from the right and left ventricular perspective. In contrast, membranous defects are poorly seen. Parasternal long- and short-axis views are limited by the near-field location of the VSD so that these are at the top of pyramid of the 3D data set, meaning that there is less potential to include adjacent landmarks. A further consideration for images acquired in parasternal projections is that...
orientation of the image into an anatomic projection is less intuitive than a subcostal approach, particularly for the less experienced operator.

For patients who are large enough to accommodate the 3D transesophageal echocardiography probe (typically >25 kg), this produces extremely high-resolution images of the ventricular septum particularly the membranous septum and the muscular septum close to the AV valves. In these authors’ experience angles of 0°, 40° to 60°, and 120° are all good to visualize the ventricular septum. The exact probe position, rotational angle, and flexion are adjusted on an individualized basis, mainly to achieve an imaging projection, which is not parallel to the ventricular septum itself. In cases of doubly committed defects and those complicated by aortic cusp prolapse, 120° has the benefit that both the defect and the cusp prolapse are visualized optimally in the same projection. Transgastric views are helpful in some cases because the insonation is orthogonal to the ventricular septum, but these projections are particularly prone to whole heart motion through the cardiac cycle. Suspension of ventilation is used briefly during acquisition where appropriate.

The use of 3D color flow Doppler may assist in sizing of defects and localizing true flow through a VSD. The limitations with 3D color flow are that the temporal resolution is significantly reduced when compared with 3D without color. Furthermore, the area of interest, which can be interrogated, is relatively small if unacceptably low frame rates are to be avoided. Motion artifact remains a significant issue when volumes are obtained during several cardiac cycles. In keeping with color flow Doppler on 2DE, the optimal color scale has to be adjusted although color gain can be adjusted during post-processing using the 3D technique. Our approach to color flow Doppler assessment of VSDs involves the use of cross plane imaging, which uses the capacity of the 3D matrix probe to project 2 user-defined orthogonal planes simultaneously. In our current clinical practice, we have not used 3DE to calculate flow or pulmonary:systemic flow ratio. Accurate determination of these parameters is an ongoing area of research in our department but does not currently extend to clinical studies.11

Orientation of 3D Echocardiographic Images

The means of presentation of 3D echocardiographic images has been the subject of some debate, and recent standards have been published for adult cardiology.12 The approach that has been adopted at our center and others is to present images in an anatomic format, particularly for views that are unique to 3DE such as en face visualization of septal structures and AV valves.10 This approach is both intuitive and consistent with other imaging modalities, such as MRI and computed tomography. Anatomic presentation is readily achieved in post-processing by appropriate rotation of rendered images. This approach is described in detail elsewhere.10

Normal Ventricular Septum-Identification of Landmarks

Three-dimensional echocardiography permits the display of the ventricular septum as a true reflection of actual anatomy rather than depiction of user-defined cross-sectional images.13 This provides a unique opportunity for accurate localization of a VSD with regards to landmarks (Figure 1; Movie II in the Data Supplement).

Visualization of the ventricular septum by 3DE is achieved by cropping away the free wall of the right ventricle.14,15 This en face view of the ventricular septum demonstrates the tricuspid valve and chordal support, the trabeculated right ventricular septal surface, landmarks such as the septomarginal trabeculation (septal band) and the right ventricular outflow tract up to the pulmonary valve. The diaphragmatic surface assists in delineating the inferior border of the heart.

When acquiring images of the ventricular septum in 3D, the gain setting of the ultrasound system is set slightly above that for 2D studies to avoid the potential creation of artifactual defects in the ventricular septum. Rotation of the image during postprocessing to visualize the septum from the left ventricle assists in ensuring that defects are real. This is assisted by rapid view of the multipplanar reformatted images to detect any areas of acoustic shadowing which affect the rendered image, as well as correlation with cross-sectional images.

Insights From 3DE Useful for Perimembranous VSD Closure

Perimembranous VSDs are the commonest type of VSD.16 The defects vary in size and are localized immediately adjacent to

Figure 1. A, Diagrammatic representation of the ventricular septum as seen from the right ventricle. The free wall of the right ventricle is removed so that the en face view of the ventricular septum can be obtained and landmarks can be identified. Image obtained using Heartworks simulator, Inventive Medical, London, United Kingdom. B, Three-dimensional echocardiographic view of the ventricular septum as seen from the right ventricle. ASL indicates anterosuperior leaflet of tricuspid valve; IL, inferior leaflet of tricuspid valve; IVC, inferior vena cava; MB, moderator band; PA, pulmonary artery; PV, pulmonary valve; RA, right atrium; SL, septal leaflet of tricuspid valve; SMT, septomarginal trabeculation; SVC, superior vena cava; TV, tricuspid valve; and VIF, ventriculo-infundibular fold.
the tricuspid valve (Figure 2A; Movie III in the Data Supplement). Extensions to the inlet (Figure 2B; Movie IV in the Data Supplement) or outlet part of the septum can also be seen and formation of aneurysmal tissue along the right ventricular side can often develop and decrease the amount of shunting from the left to the right side of the heart or be associated with spontaneous closure of the defect.

Most perimembranous defects are closed through the tricuspid valve using a right atrial approach that enables the surgeon, in most cases, to visualize the defect when lifting the septal leaflet of the tricuspid valve.\textsuperscript{17–20} However, residual defects and compromise of the tricuspid valve function have been described in the postoperative period; complications that potentially can be avoided if further information about the size and extension of the defect or tricuspid valve attachments in relation to the defect could be provided in the presurgical planning.\textsuperscript{20}

Using 3DE, the depth of field of view permits the tricuspid valve chords and the defect to be seen in a single projection, which cannot be achieved using cross-sectional imaging. In addition, during postprocessing, the septal leaflet of the tricuspid valve can be cropped away to permit direct visualization and accurate measurement of the size and shape of the defect (Figure 2A). The position of the tricuspid valve and chordal attachments in relation to the defect can be visualized in relation to the VSD (Figure 2B).

Ventricular Component of AVSDs

Atrioventricular septal defects (AVSDs) are characterized by a common AV junction, which results in unwedging and anterior displacement of the aortic valve.\textsuperscript{21} The superior and inferior bridging leaflets cross the ventricular septum, and there is a variable size and position of the atrial and ventricular components of the defect.\textsuperscript{22} Two-dimensional echocardiography has been the primary imaging modality used in the diagnostic evaluation of the AVSDs,\textsuperscript{23,24} however, this technique cannot project an en face view of the right ventricular aspect of the ventricular septum to delineate the extent of the ventricular component of the defect and valvar attachments.\textsuperscript{25,26}

A complete AVSD can be seen from the right ventricle after cropping of the right free wall to permit an en face view of the ventricular and atrial septums (Movie V in the Data Supplement). The size and shape of the atrial and ventricular components of the defect can be visualized. The extent of the defect can be comprehensively delineated and attachments of the AV valve leaflets and its chordae to the crest of the interventricular septum can be assessed accurately.\textsuperscript{27} Complementary projections from the atrial and ventricular aspects can provide further information on the valvar apparatus. Complex AV valve and left ventricular outflow abnormalities associated with the defect can also be identified using 3DE and these may assist surgical planning.\textsuperscript{13,28,29}

Muscular VSDs

Muscular VSDs can be found in different areas of the muscular part of the interventricular septum and can have different shapes, which are far from regular. Preoperative guidance of the location, size, number, and immediate relations of muscular VSDs can be particularly helpful to surgeons, considering that some of these defects can be obscured by heavy trabeculations and missed during surgical repair, particularly when the heart is arrested.\textsuperscript{15} Figure 3A (Movie VI in the Data Supplement) demonstrates an inferiorly located muscular VSD, close to the tricuspid valve but immediately adjacent to the diaphragmatic border of the heart. Forewarning of the location of the defect allows surgeons to locate and close the defect rapidly. Figure 3B (Movie VII in the Data Supplement) shows both a large muscular inlet and a smaller apical VSD from a patient with transposition of the great arteries. The major defect has a vertical slit-like shape, where the major axis of the defect has a superoinferior orientation. If assessed solely in a conventional 4-chamber projection, the defect might be considered small but the enface 3D projection illustrates accurately the shape and size of the defect.\textsuperscript{9} In this case, the additional muscular apical...
defect could be assessed accurately and in this case was not considered sufficiently large to merit closure. This could also be applied to multiple muscular VSDs (swiss-cheese defects), which pose particular problems in interpretation when assessed only by multiple 2D sonographic planes.

Where there are multiple defects, assessment by Doppler technique is particularly limited because velocities across both defects will be relatively low (unrestrictive) and Doppler assessment of the smaller defect would only be helpful once the larger defect is closed. In this situation, surgical decision making is based on the judgment of size and potential significance once other defects have been closed.

**Associated Lesions**

There are many important associated abnormalities, which may complicate surgical management of VSDs. These include prolapse of one of the aortic cusps into the VSD and straddling of AV valves. Recognition of these associated lesions is helpful before surgical repair.

Aortic cusp prolapse can be a feature of either a perimembranous (Figures 4A; Movie VIII in the Data Supplement) or a doubly committed subarterial VSD (Figure 4B). The 3D rendered technique permits visualization of the prolapsed cusp from the right ventricular aspect, consistent with the surgical view of the anatomy at repair (Movie IX in the Data Supplement). Such views can illustrate the dynamic nature of the prolapse of the cusp, which changes through the cardiac cycle (Movie X).

In other cases, there can be straddling of the tricuspid or mitral valve through the VSD. This complicates surgical closure of the VSD. Preoperative delineation of the anatomy can assist with assessing the feasibility of VSD closure. This includes determining whether straddling chords are integral to the functioning of the straddling valve. Conventional 2D imaging does not always permit visualization of the straddling chords along their entire length from the point of origin on the AV valve to the point of insertion into the contralateral ventricle. Figure 5A (Movie XI in the Data Supplement) shows straddling of tricuspid valve chords into the medial papillary muscle of the mitral valve. The depth of field provided by rendered views permits the image to be rotated to provide all necessary preoperative information. By way of contrast, Figure 5B (Movie XII in the Data Supplement) illustrates straddling mitral valve chords into the free wall of the right ventricle.
The location and size of a VSD assume particular importance in cases where consideration is given to baffling of blood flow across the ventricular septum into the aorta. This can be particularly important in complex cases, such as transposition of great arteries with VSD and subpulmonary stenosis (Movie XIII in the Data Supplement). In these cases, the size and location of the VSD can be central to decision making as to whether outflow from the left ventricle can be baffled through the communication between the ventricles to the aorta. Similar considerations are necessary in the surgical decision making in double outlet right ventricle, which we have reported previously.34

In some clinical situations, there may be debate as to whether a VSD should be closed surgically or whether the defect may be amenable to catheter intervention. One of the considerations is whether there are sufficient rims around a VSD for an occlusion device to be placed effectively without causing incompetence of the tricuspid valve or aortic valve.35–37 Three-dimensional echocardiography facilitates accurate sizing of defects to assist in device selection and also permits visualization of the rims of the defect from both the right and the left ventricular aspects, which we have previously reported.35

**Application of Other 3D Imaging Modalities in the Assessment of VSDs**

VSDs in most cases can be identified adequately using cross-sectional echocardiography and color flow Doppler; however, in complicated cases, other 3D imaging modalities including cardiac MRI (CMR) or less often computed tomography can be of assistance. The latter is usually reserved for patients with known contraindication to CMR (ie, metallic implants) or when the ability to tolerate CMR is limited by claustrophobia (Table).

CMR has the advantage over echocardiography of not being limited by acoustic windows. The wider field of view allows for imaging of the whole heart and great vessels, which are helpful for surgical planning in complex repairs. In addition to anatomic data, it permits accurate assessment of ventricular volumes, function, and flow. CMR quantification of the ratio of pulmonary:systemic blood flow (Qp:Qs) is accurate and allows assessment of the hemodynamic significance of the VSD to aid clinical decision making.38 CMR-derived ventricular volumes are also helpful in the assessment of suitability for biventricular repair in cases with borderline ventricular dimensions.

Gradient echo CMR sequences allow for real-time cine imaging of the heart structures in a single predetermined slice. The commonly used sequence is a balanced steady-state free precession because it is relatively fast and provides good contrast between the myocardium and the blood pool. Images are usually limited to 20 to 40 phases per cardiac cycle. The final image displayed is a composite of multiple images from several cardiac cycles and is thus an average of heart motion. Cine imaging can be used to visualize multiple VSDs (Movie XIV in the Data Supplement). Although these images provide...
dynamic visualization of the cardiac structures including the AV valves, they are limited to 2D images. The selection of the imaging plane and the number of slices through the heart is user dependent, and any inadvertent foreshortening may result in the underestimation of the true dimensions of the VSD. Unlike 3DE, there is no facility for manipulation of the planes of the 2D cine data set on a postprocessing platform.

Three-dimensional CMR whole heart imaging permits the acquisition of large volume data sets, which include all the margins of the VSD and the surrounding structures. Importantly, it allows postacquisition interrogation of the data set in using the multiplanar reformatting tool or rendered views similar to 3DE. The main modality for this is dual-phase 3D steady-state free precession imaging, which is ECG-gated and respiratory navigated. ECG gating allows the images to be obtained during the systolic and diastolic rest periods of the cardiac cycle. The motion artifact is reduced by means of a respiratory navigator whereby images are only obtained within a defined window of diaphragmatic position in expiration. The size and boundaries of the VSD can be assessed in both phases of the cardiac cycle to obtain the minimal and maximal dimensions (Figure 6A and 6B). Again, the data set obtained is a composite over multiple cardiac cycles. The disadvantage of this mode of imaging is the lack of any information on the relationship of finer structures, such as the AV valves in relation to the VSD because of the static nature of the images with limited spatial resolution.

Contrast-enhanced 3D CMR angiography using gadolinium-based contrast agents can help define the position and size of the VSD because the contrast fills the blood pool within the VSD. This is helpful in defining the size and the position of the VSD. Importantly, it also allows visualization of the path of complex ventricular baffles during septation in double outlet right ventricle (Figure 7). However, this is a non–ECG-gated static image. In common with 3D steady-state free precession images, there is poor visualization of the relationship of the VSD to the AV valves and their attachments, which are better seen on 3D echocardiographic images.

Echocardiography remains the main imaging modality in the assessment of VSDs with a limited number of reports in the literature on VSD evaluation by CMR. CMR remains expensive and has a requirement for sedation or anesthesia in younger patients. Furthermore, it is contraindicated in patients with metallic implants. In our unit, CMR is used in selected cases where additional information is needed to complement data from 2DE and 3DE. This is in keeping with a general consensus that CMR is only useful in complex cases.
Figure 7. Volume rendered three-dimensional contrast-enhanced angiographic image demonstrating the position of the ventricular septal defect (VSD). The ventricular septum is marked with an asterisk (*) separates the 2 ventricles and is void of contrast. LV indicates left ventricle; and RV, right ventricle.

Challenges of 3DE Relevant to VSD Closure

There are several reports of the value of using 3DE as a complementary or alternative imaging modality to cross-sectional imaging before closure of a VSD.15,35,44,45 Chen et al16 compared real-time 3DE and 2DE with surgical findings in 38 patients. They demonstrated that real-time 3DE produced novel views of VSDs and improved quantification of the size of the defect. Similarly, Cheng et al47 and Mehmood et al48 in 38 patients and 10 patients, respectively, demonstrated that sizing of VSDs using 3DE was more highly correlated with surgical findings than the diameter measured by 2DE and accurately defined VSD location, size, and surrounding anatomy in all patients. In complex lesions, De Castro et al49 reported that the use of real-time 3DE provided incremental information to 2DE in a variety of complex congenital heart lesions which affected therapeutic decision making in 82 patients. These reports support the capability of 3DE to provide complementary information to 2DE and provision of a clear perspective to surgeons so that planning of the surgical approach is facilitated. However, these results should be interpreted with caution considering that 2DE continues to have superior spatial and temporal resolution to 3D.50

Although recent software advances in 3DE permit a higher frame rate than was previously possible, this can occur at the expense of spatial resolution. New 3D software developments permit the echocardiographer to specify the area of interest, which can avoid the need for acquisition during several cardiac cycles. This provides real-time 3D imaging of a user-defined field but with a significantly lower frame rate than cross-sectional imaging, particularly if color flow Doppler is used. Wider field of view with the best resolution continues to involve acquisition over multiple cardiac cycles with the potential for introduction of artifact to the 3D data set. The problem of acoustic windows is not overcome by 3DE, and 3D transesophageal echocardiography is of assistance only in older patients because pediatric-sized probes of this type are not currently available. Importantly, postprocessing has a learning curve and requires additional analysis time so that the technique is applied only where clinically relevant additional information is sought. Some of the key steps in the postprocessing 3D reconstruction of a perimembranous and an apical VSD are demonstrated in Movie XIV in the Data Supplement. Finally, acquisition of 3D echocardiographic probes and analysis software adds additional cost, which has to be factored during resource use analysis for every unit. However, recent advances have accommodated 3D probes, both for transthoracic echocardiography and transesophageal echocardiography, that can be used for both 2D and 3D, which reduces the overall cost.

Conclusions

The use of 3DE in the evaluation of a VSD can provide important complementary information to cross-sectional imaging before planning a surgical procedure. The defect can be visualized from the right and left ventricular aspects, and simulations of the surgeon’s view can be readily achieved. The whole circumference of the defect can be visualized for accurate localization and sizing. Furthermore, complex spatial relationships can be better understood. Three-dimensional echocardiography should be considered in the detailed presurgical echocardiographic evaluation VSDs in selected cases.

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Disclosures

None.

References


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Supplemental Material

Video Legends

Video 1. Step by step post processing to visualize a PM and an apical VSD in 3D reconstruction. Initially, the red plane is aligned along the septum to show the two VSDs (a PM and an apical) from the right ventricular aspect. Clicking on the edge of icrop box is important to change the direction of interrogation.

Video 2. Three-dimensional echocardiographic view of the ventricular septum as seen from the right ventricle.

Video 3. Three-dimensional reconstruction of perimembranous VSD as seen from the RV side. Surgeons use a right atrial (RA) approach through the tricuspid valve (TV) to reach the defect.

Video 4. The septal leaflet of TV and TV chords can cover the membranous area of the defect and create a misconception regarding the actual size of the defect. Asterisks demonstrate the rims of the perimembranous VSD.

Video 5. Three-dimensional TEE rendered image of an AVSD as seen from the right side of the ventricular septum. The right ventricular free wall has been cropped. The crescentic shaped atrial and ventricular components of the defect are easily visualized.

Video 6. Inferior muscular VSD projected from the RV aspect.
**Video 7.** Muscular VSDs from a patient with transposition of the great arteries. Inlet VSDs as seen from the right side of the heart in a rendered view.

**Video 8.** 3D TEE image of right coronary cusp prolapse in a patient with a perimembranous VSD. The depth of the cusp is clearly shown as well as the elongated nature of the prolapsing cusp.

**Video 9.** Multiplanar reformatted image of the prolapsing right coronary cusp. The red, green and blue cutplanes have been aligned through the right coronary cusp to permit visualization in both short axis and long axis planes.

**Video 10.** Doubly committed subarterial VSD from the right ventricular aspect. The defect is partially occluded by the right coronary cusp, which has prolapsed into the defect. The prolapse is dynamic and the appearance changes between ventricular systole and diastole.

**Video 11.** Straddling tricuspid valve (TV). The chords cross the ventricular septum and are attached to the medial papillary muscle of the mitral valve.

**Video 12.** Straddling MV chords pass through the VSD to the free wall of the right ventricle.

**Video 13.** Multiplanar reformatted image of transposition of the great arteries with VSD and pulmonary stenosis. The red, green and blue cutplanes have been aligned through the VSD and the relationship with the outflow tracts can be clearly defined.
Video 14. 2D cine four chamber image demonstrating multiple muscular VSDs.