

Three-dimensional echocardiographic assessment of atrial septal defects

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ABSTRACT

Echocardiography provides a useful tool in the diagnosis of many congenital heart diseases, including atrial septal defects, and aids in further delineating treatment options. Although two-dimensional echocardiography has been the standard of care in this regard, technological advancements have made three-dimensional echocardiography possible, and the images obtained in this new imaging modality are able to accurately portray the morphology, location, dimensions, and dynamic changes of defects and many other heart structures during the cardiac cycle.

Key words: Atrial septal defects, Congenital heart disease, Echocardiography, Transthoracic echocardiography, Transesophageal echocardiography, Three-dimensional echocardiography, Two-dimensional echocardiography

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Congenital heart disease (CHD) represents a common and broad group of disorders affecting not only newborns, but adults as well. Particularly in the pediatric population, complications of critical CHD such as shock, cyanosis, and pulmonary edema increase dramatically when there is a delay in diagnosis and subsequent medical and/or surgical treatment. Two-dimensional echocardiography (2DE) has been the gold standard in diagnoses due to its low side-effect profile when compared with magnetic resonance imaging and computed tomography (CT). However, the advent of three-dimensional echocardiography (3DE) provides better visualization of the more subtle intricacies of cardiac anatomy that may have been more difficult to discern on 2D imaging, thus increasing its power to detect cardiac anomalies.

Atrial septal defects (ASDs) account for approximately 13% of congenital heart disorders, with a prevalence ranging from 1.6 to 1.8 of 1000 live births.^[1] There are five major defects, which include the secundum

and primum types, sinus venosus and coronary sinus defects, and patent foramen ovale (PFO). However, there is debate as to whether a PFO should be included in this category because PFOs do not have missing septal tissue. In addition, coronary sinus defects (unroofed coronary sinus) are not considered true septal defects as they represent a communication between the roof of the coronary sinus and the adjacent left atrium. Regardless, these defects often go undiagnosed at birth if asymptomatic, and often can be found incidentally on routine imaging. Understanding their type, size, location, and presence or absence of other congenital defects is paramount in determining the appropriate therapy.

Secundum defects are the most common type of ASDs and are more prevalent in females as compared to males. They usually affect the middle portion of the atrial septum within the fossa ovalis, a remnant of the foramen ovale, and result from either excessive absorption of the septum primum or from arrested growth of the septum secundum. 2DE is adequate in detecting these defects, but the images obtained

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can lack the detail necessary in accurately measuring their size, shape, and location, specifically with respect to the Swiss cheese pattern or multiple hole pattern, in which these limitations become more pronounced. 3D transthoracic echocardiography (TTE) performed via the apical, para-apical, right parasternal and subcostal views provide good visualization of the defect in the majority of patients [Figure 1]. Furthermore, 3D TTE has been shown to better approximate the location and dimensions of ASDs as well as the surrounding anatomy and rim size when compared to 2D images.^[2,3] In one study, Morgan *et al.* compared results obtained from 2D transesophageal echocardiography (TEE) to 3D TTE. Though the differences in precisely measuring the defect's diameter, area, and circumference was not statistically significant when comparing the two imaging modalities, it was clinically significant, in that 3D TTE was just as accurate in its ability to recognize appropriate candidates for percutaneous closure of ASDs, thus diminishing the need for the more invasive TEE procedure and circumventing its major complications such as GI bleeding, esophageal hematoma formation, and perforation.^[4]

The majority of ASDs, including the secundum type, have classically been repaired via median sternotomy, though recent advancements have made percutaneous closure the current treatment of choice. This decision is based on the parameters of the defect, including its size, shape, and location with respect to the surrounding tissue.^[2,5]

Percutaneous transthoracic repair can be done if the rim size is more than 5 mm. Thus, it is important to accurately obtain a comprehensive image of the

defect in order to determine its eligibility for the less invasive procedure. The ability of 3D TTE to estimate measurements correctly not only helps when planning the transcatheter approach to the procedure,^[2,6] but also provides important data needed to select the appropriate size of the occluder device which is imperative to avoid procedure related complications, including breakdown of the device, persistence of the shunt, device embolization, and even perforation of the heart.^[7] Moreover, 3D TTE has also been shown to help in visualizing the above-mentioned complications, allowing for immediate detection and reversal of the complication, which can be life-threatening.^[8] For example, color Doppler 3D TTE can uncover residual shunt after implantation of an ASD occluder^[9] and can help in evaluating the effectiveness of other percutaneous closure devices used for ASDs and PFOs.

The American Society of Echocardiography currently recommends 2D TEE during percutaneous closure and repair of ASDs, however, this is contingent on the observer's ability to mentally recreate these images in 3D space, which can be difficult. 3D TEE allows the observer to evade this problem and can offer a clearer view of the defect, leading to more accurate measurements, increased repair rates, and better identification of patients at higher risk of complications, prompting closer follow up.

Specifically, 3D TTE allows the user to measure defect size, rim size, left and right atrial occluder disc dimensions, and the distance between the left atrium and aorta which can help recognize appropriate candidates with secundum type ASDs for percutaneous closure.^[8]

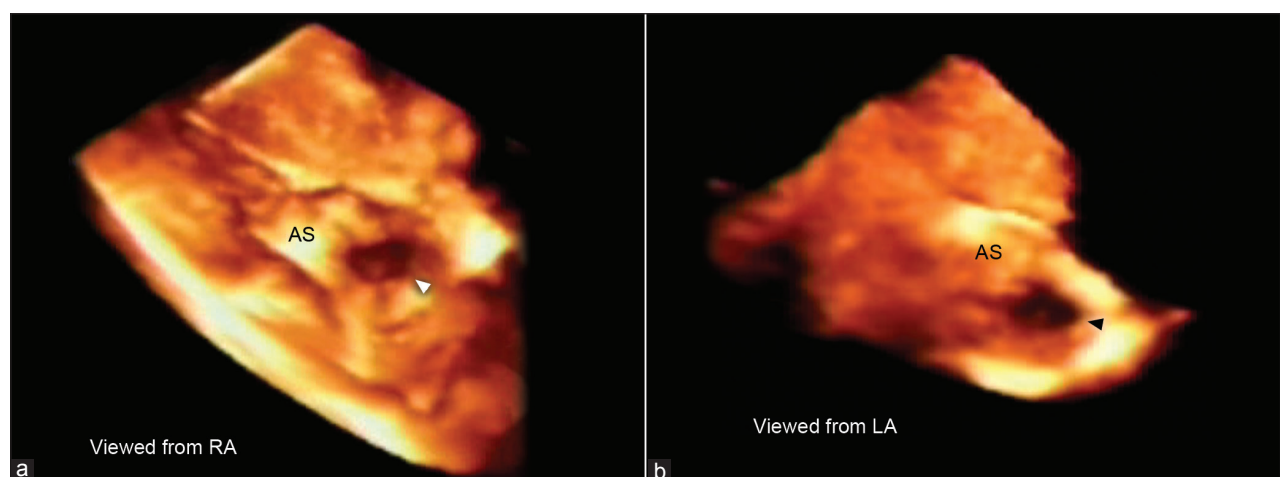


Figure 1: (a and b) Live/real time three-dimensional transthoracic echocardiographic (3D TTE) assessment of atrial septal defect (ASD). Arrowhead points to a large secundum ASD visualized from both right atrial (RA) (a) and left atrial (LA) (b) aspects. Note the large rim of tissue surrounding the defect (AS: Atrial septum). Reproduced with permission from Mehmood F, Vengala S, Nanda NC, et al. Usefulness of live three-dimensional transthoracic echocardiography in the characterization of ASD in adults. *Echocardiography J* 2004;21:707-13

In another example, Wei *et al.*, described a patient with a Swiss cheese type secundum ASD who refused surgery and hence transcatheter repair was attempted under both 2D TEE and 3D TEE guidance. Two closure devices were successfully placed without any complications, but attempted placement of a third device to close a residual defect resulted in device embolization to the left atrium and right iliac artery.^[10] The embolized device was pushed back transcatheter into the proximal descending aorta under 3D TEE monitoring and retrieved during subsequent surgery to close the defects [Figure 2]. The risk of complications from percutaneous closure of ASDs increases linearly with the complexity of the ASD, but 3D TEE is useful in their monitoring and aids in their management.^[8] Similarly, 3D TEE can also be useful in assessing encroachment of the closure device on surrounding structures such as the aorta [Figure 3].

Sinus venosus ASDs (SVASD) are characterized by malposition of the insertion of the superior or inferior vena cava straddling the atrial septum^[11] and these defects account for approximately 5-10% of ASDs. Although both 2D and 3D echocardiography have been shown to be effective in detecting SVASDs, the latter has proven to be advantageous. For example, 3D TEE reconstructed images can correctly visualize the defect in relation to the superior vena cava (SVC) and the anomalously draining right superior pulmonary vein, whereas 2D imaging has been inferior in this regard^[12] [Figure 4]. In addition, 3D TEEs can calculate the area of SVASDs due to the extra dimensions obtained from the 3D rendered image.

Atrioventricular septal defects (AVSD) result from incomplete fusion of the superior and inferior endocardial cushions during fetal development, which can result in aberrant development of the atrioventricular septum and valves,^[13] leading to both atrial and ventricular septal defects. AVSDs can be classified as complete, intermediate, and partial. Complete AVSDs can be further categorized based on the superior bridging leaflet and its attachments to the crest of the ventricular septum and right ventricle via the Rastelli system.^[14]

When measuring the size and dimensions of AVSDs, and surrounding cardiac structures, 3D TTE has proven to be far more exact than 2D TTE.^[13] For example, 3D TTE can provide a clearer picture of the characteristic five leaflets in complete defects^[14] [Figure 5], and can

better identify the superior bridging leaflet and its attachment used to categorize ASDs into the modified Rastelli types.^[13,14] Left atrioventricular valve (LAVV) regurgitation is a common sequela of AVSD repair, and quick identification is key. Live, real time 3D TTE is better than 2D imaging in evaluating LAVV regurgitation after repair due to its superiority in visualizing valve

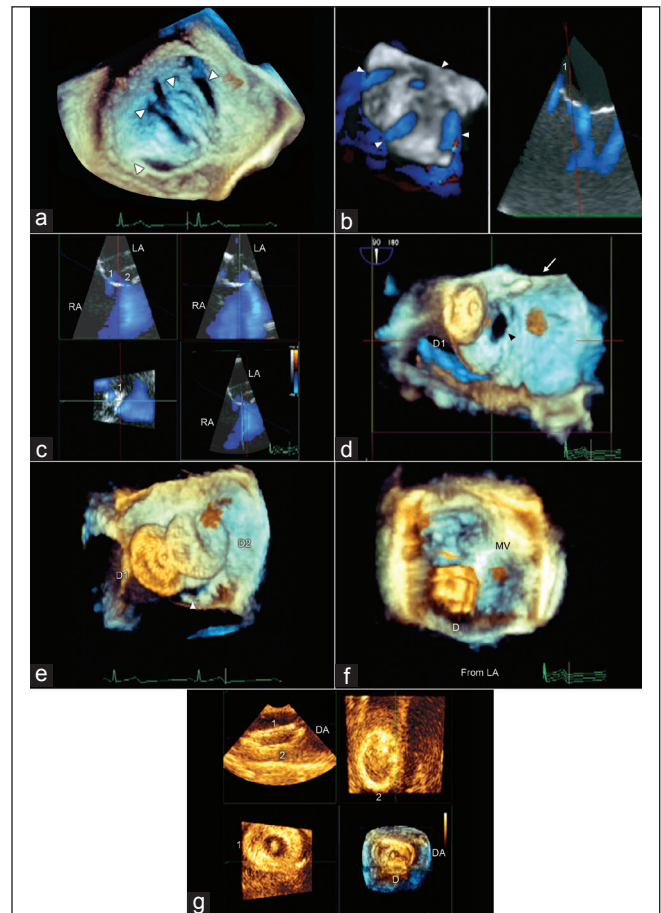


Figure 2: (a-g) Live/real time three-dimensional transesophageal echocardiographic assessment of device embolization during percutaneous atrial septal defect (ASD) closure. (a) The arrowheads point to multiple secundum ASD (ASD, "swiss cheese" appearance) viewed en face from the left atrium (LA). (b) Color Doppler assessment showing flow signals within the defects viewed en face (left panel). QLAB examination (right panel) showing four defects numbered 1, 2, 3, and 4. (c) QLAB examination demonstrating en face view of one of the defects (1) using color Doppler. In the upper left panel the cropping plane is positioned exactly parallel to the defect which resulted in en face viewing of the defect in the lower left panel. Subsequently the area was measured by planimetry. (d) Demonstrates the first ASD closure device (D1) in position (viewed from left atrium and anatomically correct). Arrowhead shows a large residual defect viewed en face. The arrow points to the device placement catheter. (e) The second ASD closure device (D2) in position, partially overlapping D1 (viewed from left atrium and anatomically correct). Arrowhead shows the presence of one of the two significant residual defects. (f) Embolization of one of the closure devices (d) to the LA. (g) The device (d) in the proximal descending thoracic aorta (DA) after percutaneous manipulation from the iliac artery. 1 and 2 denote the right and the left atrial sides of the device, which are viewed en face in the left lower and the right upper panels (MV: Mitral valve; RA: Right atrium). Reproduced with permission from Wei J, Hsiung MC, Tsai SK, et al. Atrial septal occluder device embolization to an iliac artery: A case highlighting the utility of three-dimensional transesophageal echocardiography during percutaneous closure. *Echocardiography* 2012;29:1128-31

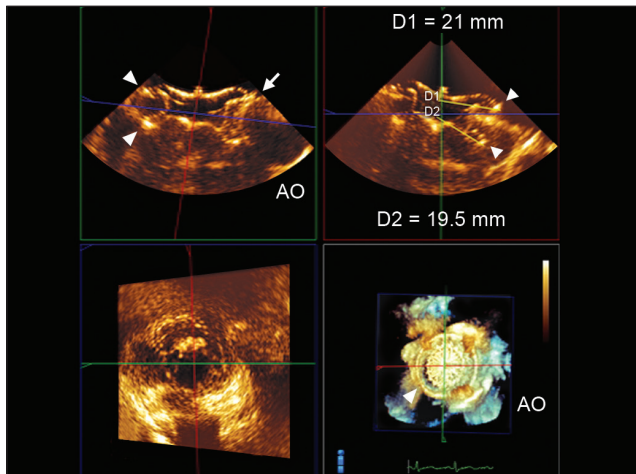


Figure 3: Live/real time three-dimensional transesophageal echocardiographic demonstration of encroachment of the aorta by the atrial septal defect (ASD) closure device. Multiplanar rendering mode: Upper panels. Top arrowheads point to the left atrial disc, lower arrowheads to the right atrial disc. D1 represents the measurement of the radius of the left atrial disc from the middle of the central marker band to the outer edge of the disc. D2 represents the measurement of the radius of the right atrial disc from the middle of the end screw to the outer edge of the disc. The arrow in the top right panel points to the area of contact of the left atrial disc with the aorta (AO). Lower panels. Arrowhead in the right lower panel points to the left atrial disc covering the ASD. Reproduced with permission from Bhaya M, Mutluer FO, Mahan III EF, et al. Incremental utility of live/real time three-dimensional transesophageal echocardiography in percutaneous closure of ASD. *Echocardiography* 2013;30:345-53

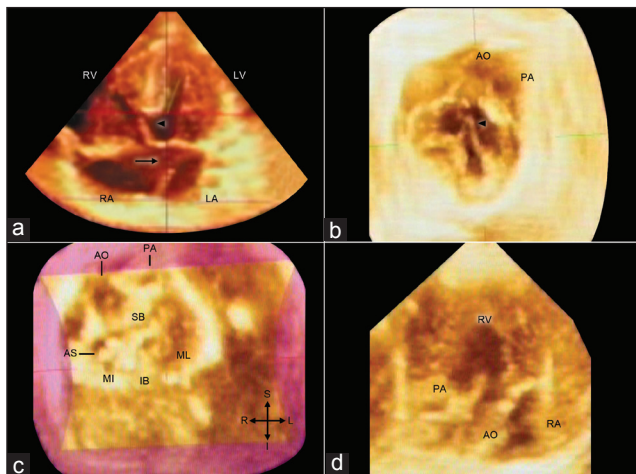


Figure 5 : (a-d) Live/real time three-dimensional transthoracic echocardiography (3D TTE) in atrioventricular septal defects (AVSDs). (a) Complete AVSD. Arrowhead shows attachment of the common atrioventricular valve (CAV) to the crest of the ventricular septum (Rastelli type A). Arrow points to atrial component of the defect; (b) Complete AVSD. Arrowhead points to an anomalous papillary muscle projecting into the left ventricular outflow tract causing subaortic obstruction; (c and d) Intermediate AVSD; (c) En face view of CAV shows superior bridging (SB) leaflet crossing over into the RV; (d) Both the AO and the PA are seen arising from the RV consistent with double outlet right ventricle (AS: Anterosuperior leaflet; IB: Inferior bridging leaflet; L: Liver; LA: Left atrium; LV: Left ventricle; MI: Mural inferior leaflet; ML: Mural lateral leaflet; PA: Pulmonary artery; PV: Pulmonary valve; RA: Right atrium; RV: Right ventricle; RAV: Right atrioventricular valve; RVO: Right ventricular outflow tract; SB: Superior bridging leaflet). Reproduced with permission from Singh A, Romp RL, Nanda NC, et al. Usefulness of live/real time three-dimensional transthoracic echocardiography in the assessment of atrioventricular septal defects. *Echocardiography* 2006;23:598-606

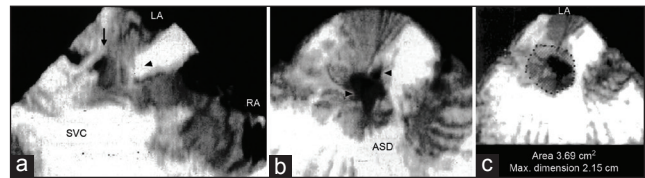


Figure 4: (a-c) Multiplane three-dimensional transesophageal echocardiographic reconstruction of sinus venosus ASD. (a) The arrowhead points to the large defect in the superior portion of the atrial septum. The arrow shows the right superior pulmonary vein entering the SVC-atrial junction at the site of the defect. (b and c) Orthogonal views demonstrating the size of the defect (ASD), which measured 3.69 cm² in area. The maximal dimension of the defect was 2.15 cm, which corresponded to the diameter of 2 cm measured at surgery. The top arrowhead in B points to the right superior pulmonary vein, and the bottom arrowhead points to the defect. (ASD: Atrial septal defect; SVC: Superior vena cava; LA: Left atrium; RA: Right atrium). Reproduced with permission from Nanda NC, Ansingkar K, Espinal M, et al. Transesophageal three-dimensional echo assessment of sinus venosus ASD. *Echocardiography* 1999;16:835-7

anatomy, morphology, function, size, shape, and location.^[15]

In conclusion, echocardiography has been pivotal as a tool for diagnosis and treatment planning for all forms of CHD, including ASDs. While 2DE is still recommended by current guidelines as the gold standard, it is clear that 3DE can perform all of the functions of 2DE and more. As the technology continues to improve, 3DE use will lead to better detection of anomalies, reduction in repair complications rates, and ultimately improved patient care.

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