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

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.\textsuperscript{[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.\textsuperscript{[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.\textsuperscript{[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,\textsuperscript{[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.\textsuperscript{[6]} 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.\textsuperscript{[8]}

For example, color Doppler 3D TTE can uncover residual shunt after implantation of an ASD occluder\textsuperscript{[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 TEE 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.\textsuperscript{[6]}

![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](image-url)
In another example, Wei et al., described a patient with a Swiss cheese type secundum ASD who refused surgery and hence transcutaneous 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. The embolized device was pushed back transcutaneously 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. 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 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 [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, 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.

When measuring the size and dimensions of AVSDs, and surrounding cardiac structures, 3D TTE has proven to be far more exact than 2D TTE. For example, 3D TTE can provide a clearer picture of the characteristic five leaflets in complete defects [Figure 5], and can better identify the superior bridging leaflet and its attachment used to categorize AVSDs into the modified Rastelli types. 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 function.
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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