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Is Three-Dimensional Echocardiography Useful in Evaluation of Atrial Septal Defects?

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Of all birth defects, Congenital Heart Disease (CHD) remains the most prevalent. These malformations are largely multifactorial with both environmental and genetic components, but known chromosomal abnormalities and mutations of single genes account for less than 10% of all cardiac defects (1). They affect approximately 6 to 13 newborns per 1000 live births (2) and are made up of 5 major Atrial Septal Defects (ASDs), including primum and secundum type defects, sinus venosus and coronary sinus defects, and Patent Foramen Ovale (PFO). However, there is debate within the medical community regarding inclusion of PFOs and coronary sinus defects within the realm of CHD. PFOs do not have absent septal tissue, and coronary sinus defects, or unroofed coronary sinus, represent an abnormal communication between the superior portion of the coronary sinus and the neighboring left atrium. Regardless, these anomalies can go undetected at birth, particularly if asymptomatic. Though some defects may be innocuous at birth, the continued shunting of blood between the atria can lead to pulmonary hypertension, heart failure, and even death. Thus, finding an accurate and reliable means of diagnosis via echocardiography is necessary in establishing the optimal treatment and ultimately improving patient mortality.

Atrioventricular Septal Defects (AVSD) are a product of incomplete fusion of the superior and inferior endocardial cushions during fetal development, resulting in irregular development of the atrioventricular septum and accompanying valves (3), leading to both atrial and ventricular septal defects. Primum type ASDs are a subtype of AVSDs because their pathophysiology is related to anomalous fusion of the endocardial cushions with the interatrial septum. AVSDs can be classified as partial, intermediate, and complete.

The Rastelli system provides another way to organize AVSDs based on the superior bridging leaflet and its attachments to the crest of the ventricular septum and right ventricle (4). When measuring the parameters of AVSDs and other cardiac structures, 3D Transthoracic Echocardiography (3D TTE) has proven to be more accurate than 2D TTE (3). For example, TTE can provide a comprehensive picture of the characteristic five leaflets in complete defects (4) (Figure 1) and can easily recognize the superior bridging leaflet and its attachment used to categorize ASVDs to determine their modified Rastelli type (3, 4). Likewise, Left Atrioventricular Valve (LAVV)

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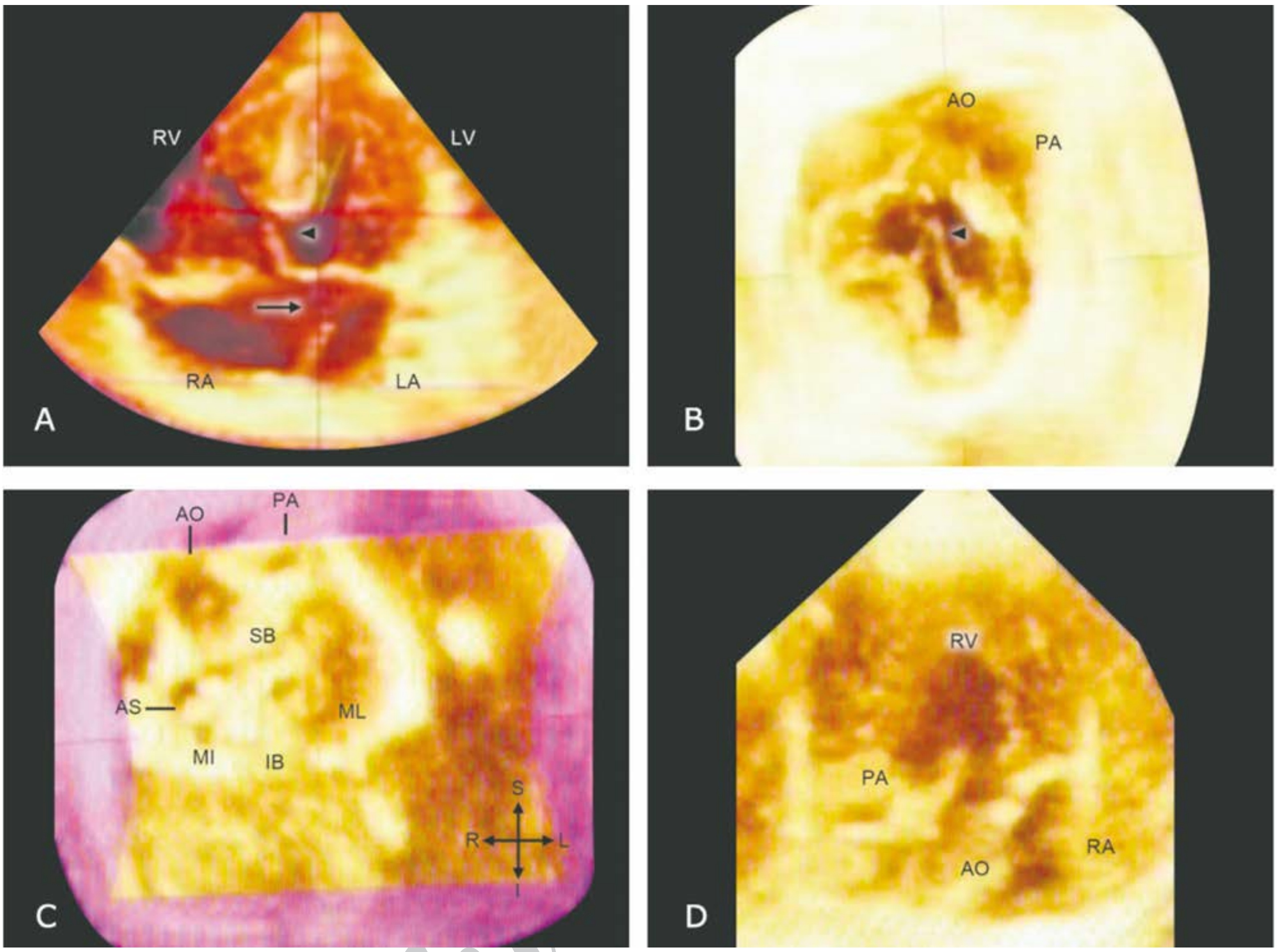
Figure 1 A-D. Live/Real Time Three-Dimensional Transthoracic Echocardiography (3D TTE) in Atrioventricular Septal Defects (AVSDs). (A) Complete AVSD. Arrowhead Shows Attachment of the Common Atrio-Ventricular Valve (CAV) to the Crest of the Ventricular Septum (Rastelli Type A). Arrow Points to Atrial 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) Leaf Let 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 Tridging 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.

regurgitation is a common complication of AVSD repair, and rapid identification is important. Live, real time 3D TTE is better than 2D TTE when evaluating LAVV regurgitation after repair due to its superiority in determining its specific location, as well as quantitatively estimating its severity through area measurements of the regurgitation vena contracta, the most narrow central flow region of a jet that occurs at or downstream to the orifice of a regurgitant valve. 3DE provides an en face view of the vena contracta, which is essentially the hole or defect through which regurgitation occurs, thereby facilitating precise assessment of its shape and area. Because 2DE images lack the extra dimensions, they cannot view the vena contracta en face and hence cannot evaluate its area which is essential for reliable calculation of the regurgitant volume (5).

Secundum defects remain the most common type of ASDs and are twice as prevalent in females compared to males. They generally involve the middle portion of the atrial septum and result from either arrested growth of the septum secundum or accelerated absorption of the septum primum. 2DE can detect these defects with some accuracy, but the obtained images lack the detail necessary to measure their true size, shape, and location. Specifically, this becomes apparent when evaluating the Swiss cheese or multiple hole pattern ASD. 3D TTE performed via the apical, para-apical, right parasternal, and subcostal views provide excellent en face visualization of the defect in the majority of patients, facilitating reliable evaluation of defect dimensions and area (Figure 2). It has been previously shown that 2DE may underestimate defect dimensions because each 2DE plane represents only a thin slice through the heart. Unless this slice happens to be exactly parallel to the long axis of the defect, its maximum dimension will be underestimated, which has been shown to occur in a significant number of patients (6). In addition, 2DE cannot reliably measure the area or circumference of the defect because it lacks en face viewing abilities. Furthermore, the rim size, in relation to the surrounding structures such as the aorta, superior and inferior vena cavae, tricuspid and mitral valves, can be accurately measured because of en face views provided

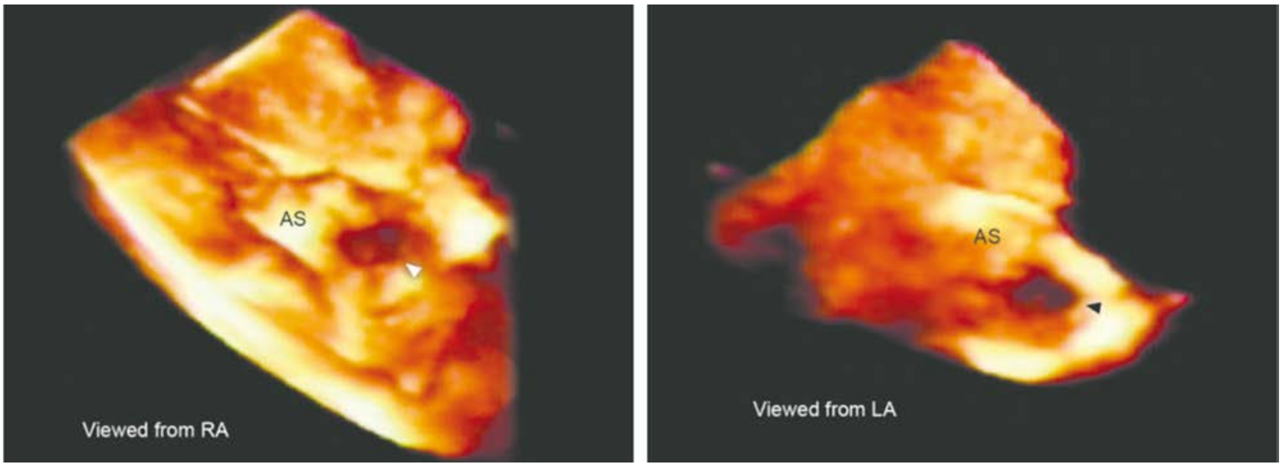


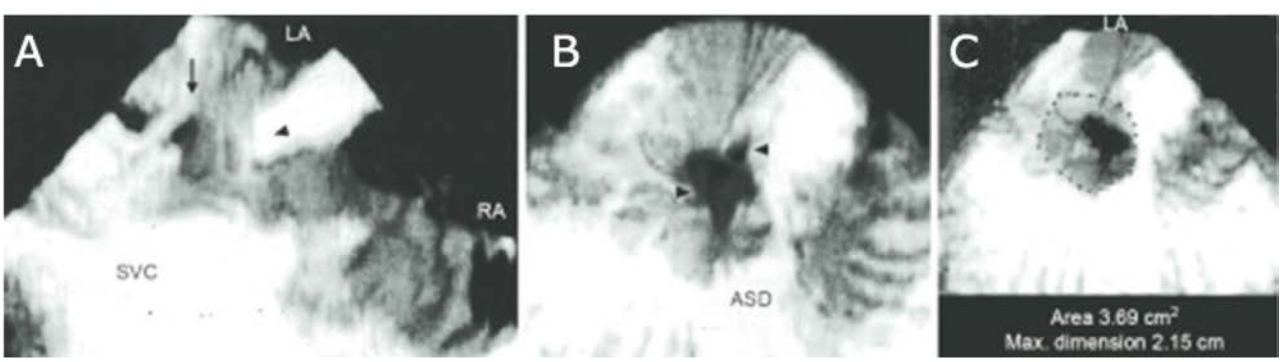
Figure 2 A and B. Live/Real Time Three-Dimensional Transthoracic Echocardiographic (3D TTE) Assessment of Atrial Septal Defect. Arrowhead Points to a Large Secundum Atrial Septal Defect Visualized from Both Right (RA, A) and Left Atrial (LA, B) Aspects. Note the large rim of Tissue Surrounding the Defect. (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 atrial septal defects in adults. Echocardiography J. 2004; 21: 707 – 713.

by 3DE. This helps not only in assessing the suitability for percutaneous repair but also in selecting the proper device size for closure. Thus, 3DE supplements 2DE by enhancing the confidence level of assessment of various parameters needed for evaluation of a patient with an ASD referred for surgical repair versus percutaneous closure (7, 8). In a study conducted by Morgan and associates, 2D TEE and 3D TTE imaging results were compared based on several parameters to ascertain a clear picture of the defect. Although the differences in precisely measuring the defect's diameter, area, and circumference were not statistically significant in their study, they were clinically significant. Because 3D TTE was shown in their study to be as accurate as 2D TEE in recognizing appropriate candidates for percutaneous closure of ASDs, it can potentially reduce the need for the more invasive TEE procedure and thus avoid major but rare complications, such as esophageal bleeding and perforation (9).

Next, Sinus Venosus ASDs (SVASD) are characterized by improper positioning of the insertion of the superior, or rarely inferior vena cava straddling the atrial septum. These defects account for approximately 5 - 10% of ASDs (10). While 2D and 3D echocardiography can both identify SVASDs with respectable accuracy, the latter has proven to be superior in many aspects. For example, 3D TEE images can correctly visualize the defect in relation to the Superior Vena Cava (SVC) and the anomalously draining right superior pulmonary vein, a significant benefit over 2D imaging (11) (Figure 3). Additionally, 3D TEEs have the added advantage of calculating the area of the defect with the extra dimensions obtained, while both can size the SVASD accurately compared to surgical measurements.

Although some do not deem PFOs to be true ASDs, they will be considered here because of their clinical significance and high incidence reportedly detected in 20 - 25% of the general population who underwent TEE (12). These defects arise due to incomplete fusion of the septum primum with the septum secundum, potentially creating a right to left shunt if the right atrial pressure is higher than in the left atrium. An accurate and specific diagnosis can be made if a saline bubble study during the Valsalva or other maneuver to increase the right atrial pressure shows bulging of the interatrial septum into the left atrium with bubbles crossing the PFO from the right atrium into the left atrium. Not surprisingly, TEE is the preferred imaging modality over TTE, but both can miss the diagnosis.

Figure 3A-C. Multiplane Three-Eimensional Transesophageal Echocardiographic Reconstruction of Sinus Venosus Atrial Septal Defect. (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 cm 2 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 atrial septal defect. Echocardiography. 1999; 16: 835 – 837.

Good contrast effect is necessary and complete filling of the right atrial cavity with microbubbles as well as bulging of the atrial septum into the left atrium produced by maneuvers deemed to increase the right atrial pressure. These parameters should be met before a PFO can be excluded with certainty. 3D TEE provides incremental value by comprehensively assessing the PFO and its surroundings and may aid in distinguishing cardiac from intrapulmonary shunts (6).

Moreover, 3DE can accurately detect complications related to ASD repair, often times at exceptional rates compared to 2DE. Historically, ASD repair has been done via median sternotomy though advancements in technology have made percutaneous closure the current standard of care in many patients. Percutaneous intravenous repair can be done if the rim size is greater than 5 mm. Accordingly, it is important to choose a diagnostic imaging modality that will give the clinician the most representative picture of the defect so that complications can be avoided. The ability of 3D TTE to approximate measurements correctly not only helps when planning the transcatheter approach to repair (7, 13), but also, as mentioned previously, provides important information used to select the appropriately sized occluder device. Poorly fitting devices can lead to major complications, such as device dislodgement and embolization, cardiac perforation, and large residual shunts (14). 3D TTE can also aid in detecting these complications, allowing for immediate recognition and reversal of the problem, thereby improving recovery rates (15). For example, color Doppler 3D TTE can not only expose but also assess the severity of a residual shunt after ASD occluder implantation (16).

In conclusion, echocardiography has become a fundamental diagnostic tool for assessing various forms of CHD, including ASDs. Not only can it provide positional data, but it can also identify valuable information to help delineate the optimal treatment. While 2DE is still recommended by the American Society of Echocardiography as the current gold standard for diagnosis, it is clear that 3DE can perform all the functions of 2DE and more. 3DE allows the observer to avoid the complicated task of recreating 2D images mentally in space, leading to better repair rates and decreased complications. As the technology continues to advance and improve and more clinicians become familiar with 3DE, management and treatment will continue to improve in patients with ASDs.

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