Review Article

Echocardiographic Evaluation of Atrial Septal Defects and Clinical Applications; A Focused Review

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Abstract:

Atrial septal defect (ASD) is a commonly encountered congenital lesion in paediatric and adult populations and accounts for 8-10 % of all congenital heart disease. Echocardiography is the most sensitive and specific imaging tool to diagnose this condition and also to determine management guidelines. Various modalities of echocardiography imaging can assess atrial septal defects completely with associated changes in cardiac chambers, haemodynamic status, relationship with neighboring structures, suitability for percutaneous closure or need for surgical intervention. Echocardiography can guide percutaneous device closure procedure as well.

Keyword: *Atrial septal defects, Echocardiography, device closure*

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Introduction

Atrial septal defects (ASD) is a septal communication which accounts for 8-10 % of all congenital heart defects and causes shunting of blood between the systemic and pulmonary circulations[1]. If the defects are significant and remain untreated, patient might have exercise intolerance, supra ventricular arrhythmias, right ventricular dysfunction and pulmonary arterial hypertension with Eisenmenger change[2]. The incidence of ASD is 1 in 5000 live births. It accounts for 30-40% of clinically important intracardial shunts in adults[3]. Patient foramen ovale (PFO) is also a common defect in atrial septum of adults and it accounts for 20-25%. There is debate whether PFO should be recognized as atrial septal defect as no septal tissue is missing but clinical syndrome associated with PFO, ASD are extremely variable and represent a health burden for community & involves specialty like pediatrics, internal medicine, neurology and cardiac & Neurosurgery etc.

So interatrial septum needs proper evaluation following a systematic approach. Echocardiography is the conventional and best method to analyze atrial septum and its abnormalities. 3 Transthoracic (TTE) Transesophageal (TEE) and Intracardiac (ICE) ultrasound are used and 2-dimentional(2D), 3-dimentional (3D), Doppler (color and spectra), transcranial Doppler types of imaging are utilized to delineate anatomy of the defect, flow direction and velocities[3].

The addition of 3-D imaging and TEE based description of anatomy of septum contributed to add more information's about atrial septum[4,5].

Development of atrial septum (IAS):

Atrial septum is composed of three separate components :1. Septum primum 2. Septum secundum 3. Atrioventricular canal septum (Endocardial cushion septum). Sinus venosus is not a component of atrial septum but defect in this results in atrial communication. Fig-1 showed development of atrial septum[1].

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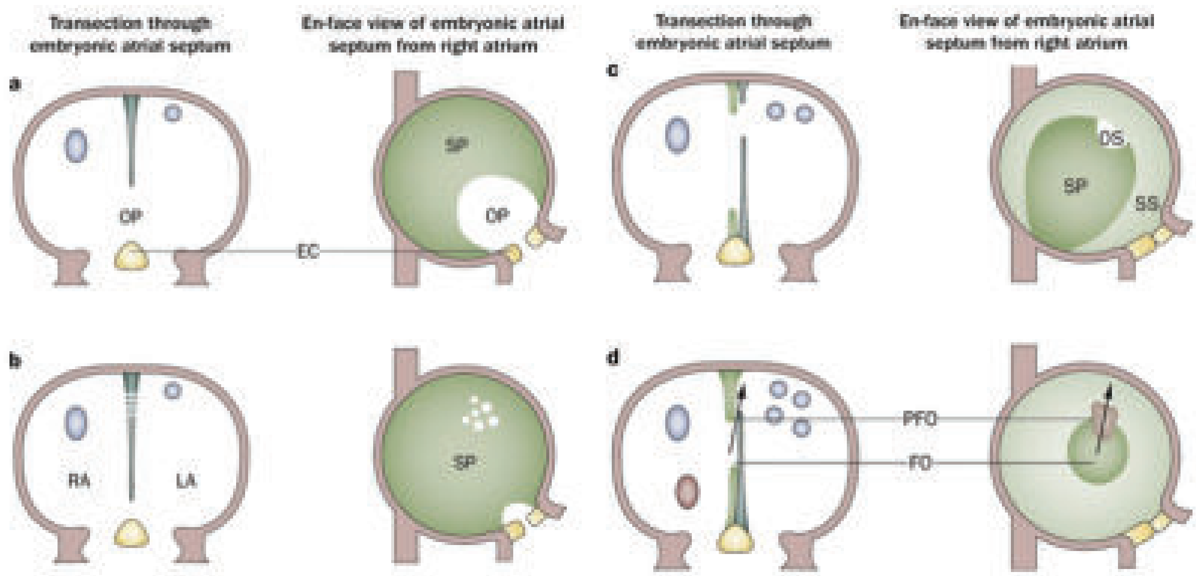


Fig.-1: *Atrial septum development.*

Atrium developed as a common cavity first. At about 28 days of gestation, septum primum originated for the aortic root and continue to grow towards endocardial cushions. The gap between these two is known “Ostium primum”. Later infoldings of atrial roof formed septum secundum in the right side of septum primum[6] Ostium primum normally closed by fusion of septum and superior / inferior endocardial cushion with the help of mesenchymal cells.

By two months’ septum primum and secundum fuses, leaving foramen ovale as only communication between two atrium[6,7,8]. Fossa ovalis is the flap of PFO formed by the septum secundum, septum premium and the AV canal septum. Sinus venosus part of septum is an adjacent structure that separates the right pulmonary veins from superior vena cava (SVC) and posterior right atrium.

Coronary sinus septum is a wall of tissue that separates coronary sinus from left atrium. To understand ASD’s, knowledge about septal development is very important and various defects are classified according to location in the inter atrial septum during development.

Atrial Septal abnormalities:

1. **Patent Foramen Ovale (PFO):**
   It is not a true defect but a potential space between septum primum and secundum (fig-2) PFO remain functionary closed as long as LA Pressure is high. PFO may be tunnel like if septum primum form a flap valve. PFO may be even circular, elliptical in shape and stretching of septum secundum due to atrial dilation lead to patency of foramen ovale. PFO is must in fetal life to maintain circulation of oxygenated blood from placenta to vital organs like heart, brain etc. After birth PFO close but 20-25% population has PFO up to adulthood. [9-11]

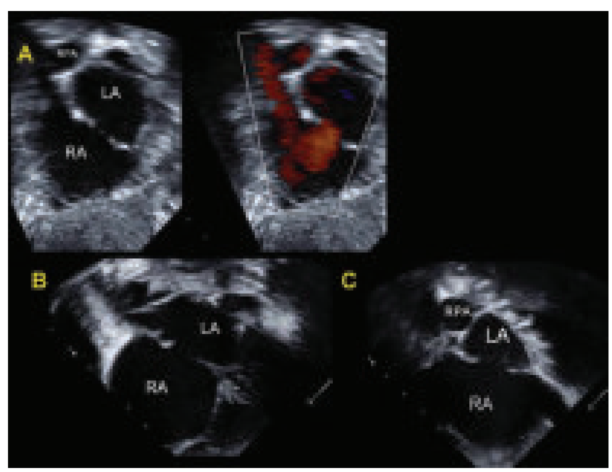


Fig.-2: *Ostium secundum ASD in TTE.*

PFO with left to right shunt occurs when atrial haemodynamics results in shunting of blood through potential communication and demonstrated by Doppler imaging. “Stretched PFO” is the defect when

PFO stretched open by atrial haemodynamics thus causing a defect. Direction of shunt depends on differences in the right and left atrial pressure.[12-14.]

Right to left shunt through PFO is of clinical significance and may cause stroke.

2. **Secundum type on Fossa ovalis Atrial Septal Defects:**

   Secundum ASD is the most common type of ASD results for true deficiency of septum primum tissue. This defect is bordered superiorly and posteriorly by septum secundum, anteriorly by AV canal septum and inferiorly by septum secundum, Primum and left venous valve of inferior vena cava. These defects may be round or elliptical. Crossing of persistent strand of septum primum results in formation of fenestrated ASD's with multiple holes. Absence of superior limbic band of septum secundum results in a rare form of ASD which is located high in the septum not same as sinus venosus ASD which is often associated with anomalous pulmonary venous drainage. Secundum ASD can enlarge over time with age and cardiac growth.

3. **Ostium primum Atrial Septal Defect:**

   This defect is characterized by absence of AV canal part of the septum in association with a common AV valve annulus and two different orifices. AV valve tissue attached to crest of ventricular septum and no shunt seen at ventricular level. Septum primum ASD is also known as incomplete or partial AV canal defect.

4. **Sinus Venosus Defects:**

   Sinus venosus ASD is not true ASD and occurs due to partial or complete absence of sinus venosus septum between superior vena cava (SVC) and right upper pulmonary vein (SVC type) or between middle and lower pulmonary vein and right atrium (IVC type).

   SVC type defect is most common and associated with anomalous drainage of right pulmonary vein.

5. **Coronary sinus defects:**

   This is a rare type of ASD where roof of coronary sinus is absent and also known as unroofed coronary sinus. Left atrial blood drained to right through coronary sinus. Association of this defect with persistent left SVC is known as "Raghib syndrome"

6. **Common Atrium:**

   All components of atrial septum as e.g., septum primum , secundum and AV canal septum may be absent result in common atrium.

7. **Atrial septal Aneurysm (ASA):**

   It is a redundancy or secular deformity of atrial septum associated with increased mobility of the septal tissue. Excursion of septal tissue more than 1 mm or LA or RA side is defined as aneurysm. ASA is associated with PFO, stroke and embolic events.

8. **Eustachian valve and Chiari network:**

   This is a remnant of the valve of inferior vena cava (IVC) that directs IVC flow to left atrium through fossa ovalis in fetal life. Chiari network is the remnant of right valve of sinus venosus and have filamentous appearance inside RA. Large and prominent eustachian valve with PFO contribute to paradoxical embolism.

Echocardiographic evaluation of interatrial septum:

Most important imaging modality to analyze interatrial septum (IAS) is Transthoracic echocardiography (TTE). In small children, image quality is good and permit full diagnostic study, selection of patient for percutaneous closure and even guidance of the device closure procedure [15,16,17.] In adult TTE is helpful for initial diagnosis of ASD, PFO but TEE is required for comprehensive evaluation of atrial septum. In adults TEE can identify margin and rims of the ASD clearly and also assess the proximity of the surrounding structures like aorta, pulmonary vein, AV valves, vena cava, coronary sinus etc. 3D TEE, offered real time anatomical details during device closure procedure.[18,19]

Intracardiac Echocardiography or ICE has been used to guide percutaneous ASD/PFO device closure procedure. Transcranial Doppler and contrast echocardiography with agitated saline has role in assessing shunt in PFO / doubt full ASD's and is not used for preliminary diagnostic purpose.[20,21]Three dimensional imaging of interatrial septum.

IAS is a dynamic complex structure & does not exist in a true flat plane. Moreover, both ASD and PFO exists in heterogeneous size, shape and configurations. 3D imaging provides clear view of IAS and allows Enface viewing of ASD and surrounding fossa and change of the morphology with cardiac cycle. It also delineates the relationship of the ASD with surrounding structures [22-25.] Table-I: showed components to be evaluated by echocardiography.[26] Table-II Showed strategy for overall evaluation of interatrial septum[1]

Transthoracic Echocardiography (TTE):

Atrial septum can be fully evaluated by using TTE guide (Table III). Size and shape of the detect, shunt direction, rim assessment, relationship of ASD to surrounding structure can be assessed thoroughly in children and even adults. TEE guide may be required in some cases with poor echo window. Fig 2 showed examples of ostium secundum ASD in TTE.

Table-I

Components to be evaluated by echocardiography.[26]

Ser	Name
1	Location of defect in the septum and type of ASD
2	Detection and qualification of shape and size of the ASD.
3	Measurement of rims surrounding ASD and adjacent structures.
4	Direction of shunting.
5	Change & remodeling in size and function of the chambers.
6	Qualification of pulmonary artery pressure.
7	Estimation of pulmonary / Systemic flow ratio
8	Examination of right heart.
9	Presence of fenestration
10	Dynamic nature of ASD / Measurement of area and maximum/ minimum diameter in end systole and diastole.
11	Stop flow diameters of ASD during balloon sizing.

Table-II

Evaluation of interatrial septum [1].

Ser	Patient Population	For establishing diagnosis	For guidance of device closure	Post-operative follow up
1	Paediatric patient <35kg	TTE	TTE/TEE/ICE	TTE
2	Paediatric patient>35-40kg	TTE,TEE,3D TEE	TEE,3D TEE,ICE	TTE
3	Adult patient	TTE, TEE,3D TEE	TEE,3D TEE,ICE	TTE

Table -III

TTE views for assessment of atrial septal anatomy.

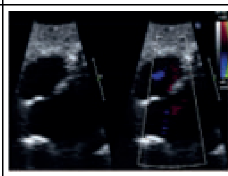
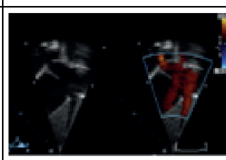
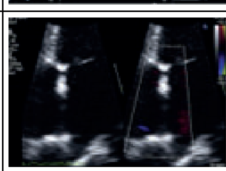
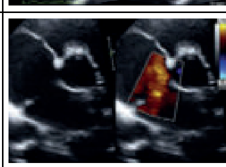
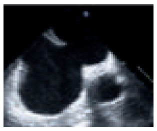
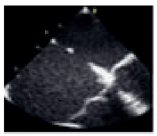
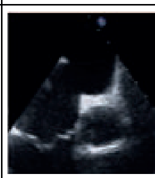
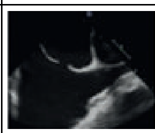
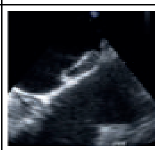
View	Example	Septal anatomy	Procedural assessment
Subxiphoid long –axis (frontal) or left anterior oblique (45^0)		Right pulmonary vein ASD rim, atrial septal defect diameter, and atrial septum length.	Position of device with regard to right pulmonary veins and assessment for residual leak.
Subxiphoid short-axis (sagittal)		SVC and IVC rim and atrial septal defect diameter.	Position of device with regard to SVC and IVC and assessment for residual leak.
Apical four-chamber		Rim of defect to AV valves , assessment of RV dilation RV pressure estimation from tricuspid regurgitation jet	Position of device with regard to AV valves.
Parasternal short-axis		Aortic and posterior atrial wall rim, atrial septal defect diameter, assessment of RV dilation.	Device relationship to aortic valve, assessment for impingement on aorta or straddle and relationship of device to posterior wall

Table-IV
Views for assessment of ASD by TEE.

View	Example	Atrial Septal anatomy	Procedural assessment	Suggested multiplane angles	Esophageal position
Basal transverse		SVC, superior aortic ,RUPV	Device relationship in atrial roof	0^0,15^0,30^0,45^0	Mid to upper esophagus
Four-chamber		Posterior and AV rims, maximal ASD diameter	Device relationship to AV valves	0^0,15^0,30^0	Mid-esophagus
Short-axis		Posterior and aortic rims, maximal ASD diameter	Device relationship to AoV and posterior atrial wall	30^0,45^0,60^0,75^0	Mid- to upper esophagus
Bicaval		IVC and SVC rims, maximal ASD diameter	Device relationship to RA roof/dome	90^0,105^0,120^0	Mid-to upper esophagus and deep transgastric
Long -axis		Dome/roof of LA	Device relationship to LA dome/ roof	120^0,135^0,150^0	Mid to upper esophagus

a) **Apical four chamber view:** In this view, ASD measurement may be overestimated as ultrasound beams is parallel to septum. This view can assess haemodynamic consequence of ASD like RA, RV dilation, Tricuspid valve regurgitation velocity etc.

b) **Subxiphoid four chamber view:** This is the best view for assessing atrial septum. The septum run perpendicular to ultrasound beams and provide highest resolution to measure ASD in long axis. Sinus venosus defects are difficult to visualize as vena cavae are not viewed longitudinally.

c) **Subxiphoid sagittal view: This** view is good for imaging atrial septum along its superior inferior axis. Dimension measured in this view can be compared with subxiphoid four chamber view to determine the shape (circular/ oval) of the defect, SVC and IVC rim can be measured here and is an excellent view to assess sinus venosus ASD. This view is obtained by 90^0 clockwise rotation of frontal view.

d) **Left anterior oblique view:** This view is obtained by 45^0 counter clockwise rotation of the frontal view. This view allows imaging of length of atrial septum and good to identify ASD primum, right sided pulmonary veins, coronary sinus dilation.

e) **Parasternal short axis view:** This view allows visualization of base of the heart anterior to aortic root with anteroposterior orientation of the defect and aortic rim can be measured nicely.

Transesophageal Echocardiography (TEE) guideline for inter atrial septum:

Multiple and sequential views are required to evaluate IAS, size and shape of the ASD's, its rim and relationship with surrounding structure.

American society of echocardiography recommend to start form a standard view and then stepwise increment of transducer angle by 15^0 or sweeping the beam through area of interest is good to image IAS. The color Doppler scale should be 35-40 cm/sec to visualize low velocity flow across PFO or ASD. Continuous and pulsed Doppler can measure the velocity and direction of shunt[27]. Fig-3 showed large osteum secundum ASD in TEE [1]. Five basal views are important to interrogate IAS and its surrounding structures (Table -IV).

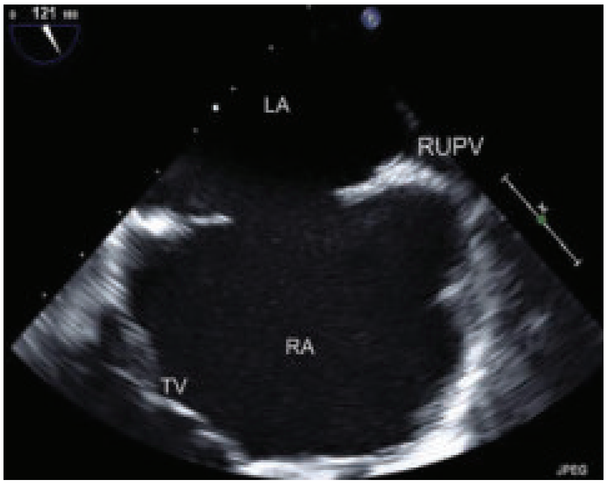


Fig.-3: *Ostium secundum ASD in TEE*

Upper esophageal short axis view: This view is obtained at upper esophagus with step wise sweeping at 15^0, 30^0 and 45^0. Superior aspect of atrial septum, ASD secundum, roof of RA, LA, surrounding vessels (SVC, aorta) entry of right pulmonary vein is visualized.

Mid esophageal short axis view

Mid oesophageal aortic valve short axis view is obtained at mid oesophageal level sweeping at 45^O, 60^0, 75º Progression of angles allow visualization of AoV in short axis view to modified bicaval tricuspid valve view. This view capture anterior and posterior plane of atrial septum and anteroposterior diameter can be measured.

Mid Oesophageal four chamber view:

This view is obtained by multiple angles of 0^0 at mid oesophageal level with stepwise increment to 15o and 300. It can evaluate atrial Atrioventricular septum (Septum primum) and relationship of ASD if any to AV valve. .

Mid esophageal bicaval view: This view is obtained by sweeping with multiplane angles of 90^0, 105^0 and 120^0. It can image superior and inferior plane of IAS and surrounding SVC, IVC and right pulmonary veins. This view is good to visualize sinus venosus ASD (SVC & IVC type) and anomalous pulmonary veins.

Mid esophageal long axis view: This view is obtained by sweeping at 120^0,135^0 and 150^0 and good to evaluate roof of LA when a device is implanted.

3D TEE imaging of interatrial septum:

A through 3D examination begins with a real time narrow angled acquisition of images from standard views. To obtain higher resolution, 3D wide angled acquisition is performed. American Society of Echocardiography (ASE) recommends narrow angled, zoomed and wide angled acquisition of 3D data's sequentially is several important views.[28]

3D display: When IAS is viewed from LA, atrial septum should be oriented with the right upper pulmonary vein at the 1-0 clock position. The qualitative parameters obtained from 3D TEE imaging are type of ASD location in atrial septal shape, orientation etc. Quantitative analysis by 3D include maximum length, width and area measured in atrial diastole. ASD dimension measured in end systole is necessary to determine change with cardiac cycle.

Intracardiac Echocardiography imaging (ICE) Protocol:

Radial and phased array ICE is useful for comprehensive assessment of the atrial septum, septal defects, rims etc. ICE offer a radial rotational or phased area imaging plane that is manipulated by insertion and withdraw of catheter. In case of axial phased array, manipulation is controlled by steering with adjustable tension, so that catheter can be held in up to four directions (anterior, posterior, left and right) Insertion and withdrawal of probe images IAS superiorly and inferiorly. Axial rotation allows visualization in multiple planes.

Role of echocardiography in percutaneous device closure of secundum ASD:

Secundum ASD's the commonest type of ASD and are amenable for device closure if anatomy is favorable. Echocardiography is the most important imaging guide to select patient, guide whole intervention procedure and also to check complication like device embolization, cardiac perforation, tamponade and device erosion[29].

Table V showed indications and contraindications of ASD and PFO closures 1.

By virtue of its ease of use from patients bed side, lack of radiation and portability, it has taken a key role in interventional procedure in catheterization laboratory[30].

ASD secundum is surrounded by six rims and rim length of 5mm or more is considered as favorable for transcatheter closure. Less than 5mm is considered as deficient rim.

Deficient aortic rim is a potential risk for erosion [31,32].

ASD rims can be named as follows:

1. Aortic rim, the antero superior rim between ASD and aortic valve annulus and aortic root.
2. **AV valve rim:** The anteroinferior rim between ASD and the AV valves.
3. **SVC rim:** Posterosuperior rims between ASD and SVC.
4. **IVC rim:** Posteroinferior rim between ASD and IVC.
5. **Posterior rim:** Between ASD and posterior anterior walls.
6. Right upper pulmonary vein rim (RUPV rim) between the ASD and the RUPV.

TEE can evaluate all six rims in upper oesophageal short axis, mid esophageal short axis, four chamber and bicaval views. TTE can provide adequate information in paediatric patients. Mid esophageal 4 chamber view is (0^0-15^0) good for identifying anteroinferior rim and posterosuperior rim. Mid esophageal AV short axis view (30^0-45^0) is good for anterior and posterior rims. Mid esophageal bicaval view (110^0-130^0) is good to visualizing superior & inferior rims. During deployment of device, most important views are four chamber view and short axis view. [33,34]

Device size selected should be 2mm greater than stop flow diameter or largest diameter measured by TTE or TEE. LA disc is deployed first inside body of LA and remote from pulmonary veins and LA appendage. Waist of the device is partially deployed in LA with continuous pull towards septum with an aim to stent the ASD. Afterwards continuous traction is maintained towards RA and RA disc released. Delivery cable is advanced towards the septum to bring both disc closer. Follow up evaluation can be performed with TTE at 1,6,12 months of procedure and yearly thereafter for 3 years or as per institutional protocol. RV size normalize usually by one month but long standing RV dilation take time even might not normalize completely.

Conclusion:

Interatrial septum is a complex structure and associated with abnormalities varied from septal defects at different locations to atrial septal aneurysm, presence of remnant of eustachian valve, Chiari network etc. All information like type, size, shape, rims surrounding the defect, degree of shunting through defects changes in size and function of cardiac chambers, pulmonary artery pressure, Eisenmenger change can be assessed with the help of TTE, TEE commonly and ICE and 3D imaging during device implantation in some centers. In future more refinement in all modalities including 3D imaging, fusion imaging of Echocardiography with cardiac computed tomography, fluoroscopy and more procedural refinement of device implantation is under consideration. All will lead to more successful device implantation rate in coming days even for large secundum ASDs.

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