

Introduction

Atrial septal defects (ASDs) are the second most common congenital heart lesion in adults representing around 30-33% of CHD diagnosed in adults (following bicuspid aortic valves). They represent approximately 7-10% of all cardiac anomalies (**Kazmouz et al,2013**). These defects are often undetected due to the lack of prominent clinical symptoms initially. If untreated, an ASD can eventually result in right ventricular (RV) heart failure, pulmonary hypertension, atrial arrhythmias, or paradoxical embolization and ischemic cerebral events (**Rigatelli et al., 2005**).

Before the advancement of interventional procedure, ASDs have only been closed surgically through midline sternotomy. More recently, these Procedures have been accomplished with minimally invasive surgical Techniques as well as a percutaneous transcatheter technique. The latter Technique has been increasing in popularity; especially in last two decades; for the avoidance of surgery associated risks (**Wilson et al., 2008**).

ASD repair with the transcatheter technique has been shown to have a high success rate but unfortunately, anatomy

and the size of the defect often limit their use (***Wilson et al., 2008***).

Different imaging modalities as conventional fluoroscopy and different echocardiographic techniques including transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), intracardiac echocardiography (ICE) and real time three-dimensional transesophageal echocardiography (3D TEE) are being used by many centers as an integral part of the procedure (**Silvestry et al. 2009**). And this raised the questioning for reliability of each of those tools.

Aim of the work

This study aims to evaluate the additional value of transesophageal echocardiography over the conventional methods (including transthoracic echocardiography and conventional fluoroscopy) in decision making and procedural guidance in percutaneous trans-catheter closure of secundum atrial septal defect.

Literature Review

Embryology and anatomy of IAS

Embryogenesis of the IAS:

Throughout cardiac embryogenesis an avenue for inter atrial blood flow is maintained, despite the development of two separate septal structures (*Allen et al., 2008*).

The septum primum, which is the first septum to develop, is an incomplete thin walled partition in which the anteroinferior free edge is above the atrioventricular canal and becomes lined by tissue derived from the superior and inferior endocardial cushions. Before the resultant inter atrial opening (ostium primum) becomes sealed by endocardial cushion tissue, new fenestrations develop form along the anterosuperior aspect of the septum primum. These fenestrations coalesce to form a large second interatrial communication (ostium secundum) maintaining inter atrial blood flow (*Anderson et al. 1987*).

At this time, to the right of the first septum, an anterosuperior infolding of the atrial roof occurs and forms a second septal structure (septum secundum) which expands posteroinferiorly as a thick-walled muscular ridge to form an incomplete partition that overlies the ostium secundum. The

channel for inter atrial blood flow through the ostium secundum is known as the foramen ovale (*Anderson et al. 1987*).

Concurrently with atrial septation, the left horn of the sinous venosus forms the coronary sinus, and the right sinus horn becomes a part of the right atrium. Infolding at the sinoatrial junction forms the right and left venous valves, whereas the right venous valve is maintained and forms the rudimentary valves of the inferior vena cava and the coronary sinus, the left venous valve becomes fused to the superior, posterior, and the inferior margins of the fossa ovalis (*Porter, 2000*).

Anatomy of the Interatrial Septum:

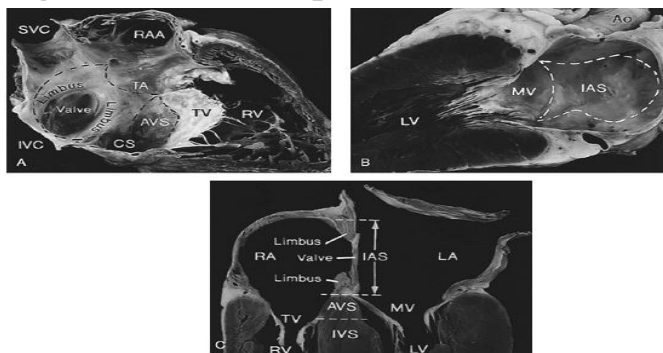


Figure (1): Atrial septal anatomy. A: Two-chamber view, right-sided. B: Two-chamber view, left-sided. The interatrial septum (IAS, outlined by dotted lines) is relatively small and is associated primarily with the limbus and valve of the fossa ovalis. Anterosuperiorly, the aortic root indents the right atrial free wall as the torus aorticus (TA). C: Four-chamber view. The IAS lies between the right and left atria (RA, LA), whereas the atrioventricular septum (AVS) lies between the RA and left ventricle (LV) (*Allen et al., 2008*).

The inter-atrial septum occupies an area on the right atrial medial wall, in its center lies the fossa ovalis bordered by a curved ridge of atrial myocardium called the limbus. The prominence and circumferential extent of the limbus varies in shape it may be circular, oval, or shaped like the letter C or an inverted U. The average area of the atrial septum was 8.9 cm² and that of the fossa ovalis 2.4 cm² as measured in adult autopsies by Sweeney and rosenquist (*Sweeney et al., 1979*).

Schwinger et al. (1990) used transesophageal echocardiography (TEE) to study the thickness of the atrial septum. They found that the latter was thickest peripherally (i.e. near its attachment to the atrial free wall) and was most thin at the fossa ovalis. In 82% of their 119 patients, the atrial septum on either side of the fossa ovalis was of approximately uniform thickness which varied enormously, from 2 to 20 mm. Atrial septal thickness at the fossa ovalis was only 1.8 ± 0.7 mm (mean).

The septum has quite different characteristics on its right and left sides. *Anderson et al. (1987)* described the anatomy of the interior of the right atrium, when the right atrium is opened through a wide incision; a large expanse of the atrial “septum” is seen between the orifices of the superior and inferior caval

veins and the attachment of the septal leaflet of the tricuspid valve. Only a small part of this area separates the cavities of the two atrial chambers, namely the floor of the fossa ovalis and its muscular margins (the limbus fossae ovalis).

The superior limbus, often called the septum secundum is made up in its larger part of an infolding of atrial wall between the base of the superior vena caval vein and the insertion of the right pulmonary veins to the left atrium. This is the area known to the surgeons as Waterston's groove; a substantial cleavage plane extends down to the margin of the fossa which can be opened by external dissection. The internal aspect of this groove is the prominent muscle band which separates the fossa from the orifice of the superior caval vein. The part of the limbus immediately adjacent to the fossa is a true septal structure (**Beerman et al., 1987**).

More anteriorly the margin of the limbus continues as a septal structure, but its larger part is the anterior atrial wall overlying the aortic root. Postero-inferiorly the fossa ovalis becomes directly continuous with the wall of the inferior caval vein. Antero-inferiorly the morphology becomes more complicated, since the fossa is separated from the orifice of the coronary sinus by the sinus septum. Through the sinus septum

runs the tendon of todaro, which is the continuation of the commissure of the Eustachian and Thebesian valves (**Beerman et al., 1987**).

The muscular structure between the mouths of the inferior caval vien and the coronary sinus is not septal, but is rather the free wall of the right atruim. The part of the atrial wall between the tendon of todaro and the septal leaflet of the tricuspid valve is an atrioventricular rather than inter atrial septal structure. This is because the tricuspid valve is attached to the septum more apically than the mitral valve, so the septum between them separates the right atruim from the left ventricle. Thus, only the thin translucent floor of the fossa ovalis (septum primum) together with the margins of the limbus immediately adjacent to the fossa is a true inter atrial septal structure (**Beerman et al., 1987**).

When viewed from the left atrial aspect, the morphology is far less complex. The floor of the fossa is smooth but in its anterosuperior margin it is roughened and wrinkled. Only a small part of this anterior region is septum, the rest being the area where the septum primum is fused with the anterior atrial wall (**Beerman et al., 1987**).

Morphological variants of interatrial communications

The accurate classification of atrial septal defects is important, not because the location of the defect alters hemodynamics, but rather because of differences in the incidence of associated anomalies and differences in techniques of repair (either surgical or transcatheter). It's obviously important for the surgeon in planning repair to be aware of the anomalously connected pulmonary viens which accompany a sinus venosus defect or of the atrioventricular valve abnormalities which go with an ostuim primum defect (**Latson, 2002**).

Patent foramen ovale:

In the fetus, the interatrial septum consists mainly of the septum secundum, which is a non-mobile, flat structure deficient in its center. Immediately to the left of this is the septum primum, a mobile flap that allows blood to flow from right to left. All through intrauterine life blood flows through this valve-like foramen ovale from right to the left atrium, but at birth the left atrial preasure rises above of the right atrium, and the flap like septum primum closes the foramen ovale and forms the floor of the fossa ovalis (**Beerman et al., 1987**).

Fusion of the septum primum with septum secundum occurs at a variable time after birth in the majority of individuals, thus sealing off the two atria from each other permanently. In a minority of otherwise normal persons, such fusion does not occur, though the foramen ovale remains functionally closed. In other words, an oblique, slit-like potential communication persists between the right and the left atrium. In a series of 965 autopsies from the Mayo clinic (*Hegan et al., 1984*), 27% had a valve–patent foramen ovale, the prevalence being higher (34%) in children and young adults than in elderly (20%).

Right to left blood flow through a patent foramen ovale may be demonstrable on color flow Doppler. *Chenzbraun et al. (1993)* found the best TEE plane for this purpose to be a vertical plane passing through the fossa ovalis as well as both venae cavae.

Ten milliliters of agitated saline solution is injected into an arm vein, resulting in dense right atrial opacification by micro bubbles. A right-to-left shunt is judged to be present if micro bubbles are found in the left atrium within three cardiac cycles after complete right atrial opacification. Five or more

micro bubbles must be visualized others say more than 3 micro bubbles (*Lynch et al., 1984*).

Right –to-left shunting through a patent foramen ovale can be enhanced or provoked by the Valsalva maneuver or coughing, both of which cause abrupt transient elevation of right –sided heart pressure. Between 10% and 18% of apparently healthy normal persons have a patent foramen ovale by contrast TEE (*Lynch et al., 1984*).

Aneurysm of the atrial septum:

Since the floor of the fossa ovalis is developed from the septum primum of the embryo, aneurysms of this region are sometimes referred to as septum primum aneurysms. The floor of the fossa ovalis is commonly taut and flush with the limbos of the fossa and adjacent atrial septum or the floor of the fossa maybe mildly depressed with the limbus a conspicuous ridge or shoulder. In yet other cases, the thin fossa floor is somewhat lax or floppy on pressure but cannot be pushed far into either atrium; such mild redundancy is considered within the range of normal variance and should not be called an IAS aneurysm. The latter term should be reserved for cases with severe redundancy such that the aneurysmal bulge protrudes far into the atrial cavity (*Hanely et al., 1985*).

Quantitative measurements of IAS aneurysms were reported by **Silver et al. (1978)** in their 16 autopsy cases. The diameter of the base of the aneurysm varied from 1.5 to 2.5 cm, and the aneurysm projected 1.1 to 2.4 cm into the atrium (usually the right). The Mayo Clinic's echocardiographic criteria for diagnosis of IAS aneurysm were 1.5-cm protrusion beyond the plane of the atrial septum and a base diameter of at least 1.5 cm (**Massani et al., 2001**).

Certain aspects of the pathology of IAS aneurysm may have clinical implications: (1) fine fibrin-like tags have been noted on the aneurysm's convexity, causing a rough or ragged appearance of the latter, (2) rarely, small annular thrombi have been seen in the groove formed between the aneurysm and the limbic ridge, (3) multiple small perforations on the order of 1 or 2mm often occur at the apex of the IAS aneurysm, presumably the result of progressive thinning and eventual fenestrations (such tiny holes are not recognizable by conventional echocardiography but have been detected by TEE), and (4) valve-patent patent foramen ovale and (5) mitral valve prolapse are more common in patients with IAS aneurysms than in the general population (**Burston et al., 1990**).

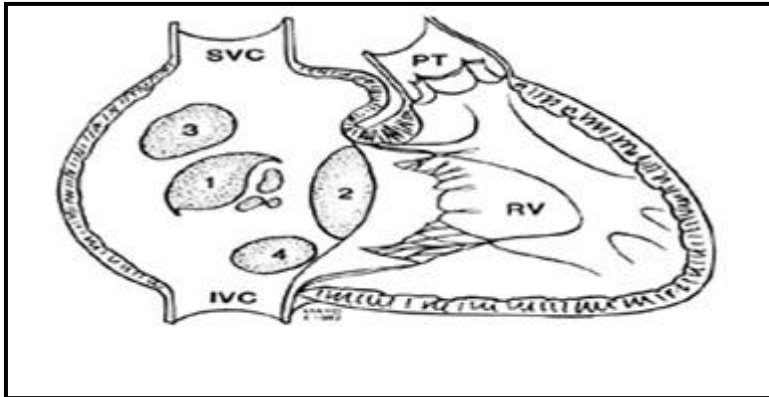


Figure (2): Schematic diagram showing the location of atrial septal defects, numbered in decreasing order of frequency: 1, secundum; 2, primum; 3, sinus venosus; 4, coronary sinus (CS) type. (IVC), inferior vena cava; (IVS), interventricular septum; (MV), mitral valve; (PT), pulmonary trunk; (RAA), right atrial appendage; (RV), right ventricle; (SVC), superior vena cava; (TV), tricuspid valve (*Allen et al., 2008*).

Ostium secundum ASD:

This is by far the most common variety, accounting for about two-thirds of all ASDs. They are restricted to the fossa ovalis region, though variable in size within the area enclosed by the ridge or the limbus of the fossa ovalis. An ASD can permit flow from the left to the right atrium or in a reversed direction and is a different hemodynamic situation from a valve-patent foramen ovale, which allows only right to left shunting if right atrial pressure exceeds left atrial pressure (*Anderson et al., 2010*).

A secundum ASD can result either from excessive resorption of the septum primum or from deficient growth of

the septum secundum. Some believe that gross right atrial dilatation, due to right –sided heart failure in a patient with patent foramen ovale, can stretch the latter so as to convert it into a true ASD in the sense that a hiatus is formed in part of the fossa ovalis floor (*Anderson et al., 2010*).

10 morphological variants have been defined for this defect, the most common type being the defect with deficient aortic (anterosuperior) rim (42.1%). The other variants included central defects (24.2%), deficient IVC (inferoposterior) rim (12.1%), perforated aneurysm of the septum (7.9%), multiple defects (7.3%), combined deficiency of mitral (anteroinferior) and aortic rims (4.1%), deficient SVC (posterosuperior) rim (1%), and deficient coronary sinus rim (1%) (*podnar et al.,2001*).

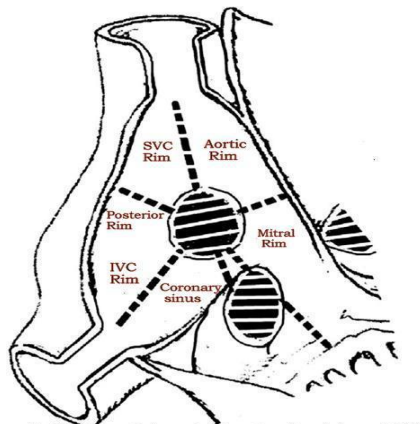


Figure (3): Septal rims surrounding secundum ASD (*Butera et. al, 2008*).