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Atrial Septal Defect – A Review

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1. Introduction

Defects in the atrial septum cause left to right shunt because the left atrial pressure is higher than that in the right atrium. This causes volume overloading of the right ventricle. While this is generally well tolerated in infancy and childhood, development of exercise intolerance and arrhythmias in later childhood and adolescence, and the risk for development of pulmonary vascular obstructive disease in adulthood make these defects important. There are four major types of atrial septal defects (ASDs) and these include ostium secundum, ostium primum, sinus venosus and coronary sinus defects. The clinical features are essentially similar and I will present detailed discussion of ostium secundum and primum ASDs followed by brief presentation of the other two defects.

Persistent patency of the foramen ovale in nearly one third of normal population makes the patent foramen ovale (PFO) a normal variant, although these become important in the presence of other structural abnormalities of the heart and when they become the seat of right to left shunt causing paradoxical embolism resulting in stroke/transient ischemic attacks or other problems, such as migraine, Caisson's disease and platypnea-orthodexia syndrome. The issues related these types of PFOs will be briefed at the conclusion of this chapter.

2. Secundum atrial septal defect

Atrial septal defects constitute 8% to 13% of all congenital heart defects (CHDs). Pathologically, there is deficiency of the septal tissue in the region of fossa ovalis. These may be small to large. Most of the time, these are single defects, although, occasionally multiple defects and fenestrated defects can also be seen. Because of left-to-right shunting across the defects, the right atrium and right ventricle are dilated and somewhat hypertrophied. Similarly, main and branch pulmonary arteries are also dilated. Pulmonary vascular obstructive changes are not usually seen until adulthood.

Mitral valve abnormalities, including mitral valve prolapse and mitral insufficiency may be seen in some patients. It is not clear whether these abnormalities are to due to right ventricular volume overloading or intrinsic abnormality of the mitral valve. Pulmonary valvar pressure gradients are seen frequently and are thought to be related to increased flow and/or differences in expression of kinetic and potential energies in the right ventricle and pulmonary artery (Rao et al 1973); however, true pulmonary stenosis is present in only 5% of ASD patients. Persistent left superior vena cava may be present in 10% patients.

2.1 Clinical features

The clinical features are essentially similar in all types of ASDs mentioned in the Introduction section.

2.1.1 Symptoms

Isolated ASD patients are usually asymptomatic and are most often detected at the time of preschool physical examination. Sometimes these defects are detected when echocardiographic studies are performed for some unrelated reason. A few patients do present with symptoms of heart failure in infancy, although this is uncommon.

2.1.2 Physical examination

The right ventricular and right ventricular outflow tract impulses are increased and hyperdynamic. No thrills are usually felt. The second heart sound is widely split and fixed (splitting does not vary with respiration) and is the most characteristic sign of ASD. Ejection systolic clicks are rare with ASDs. The ejection systolic murmur of ASD is soft and is of grade I-II/VI intensity and rarely, if ever, louder. The murmur is secondary to increased blood flow across the pulmonary valve and is heard best at the left upper sternal border. A grade I-II/VI mid-diastolic flow rumble is heard (with the bell of the stethoscope) best at the left lower sternal border. This is due to large volume flow across the tricuspid valve. There is no audible murmur because of flow across the ASD.

2.2 Noninvasive evaluation

2.2.1 Chest x-ray

Chest film usually reveals mild to moderate cardiomegaly, prominent main pulmonary artery segment and increased pulmonary vascular markings.

Fig. 1. Chest x-ray in posterior-anterior view demonstrating mild cardiomegaly, increased pulmonary vascular markings and a slightly prominent main pulmonary artery segment as seen in patients with atrial septal defect.

2.2.2 Electrocardiogram

The ECG shows mild right ventricular hypertrophy; the so-called diastolic volume overload pattern with rsR' pattern in the right chest leads.
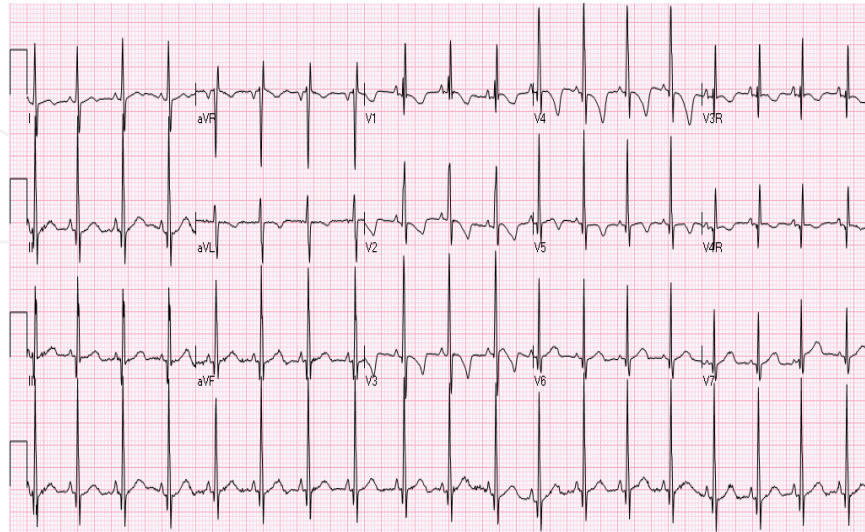

Fig. 2. An electrocardiogram demonstrating rsR' pattern in right chest leads, the so called diastolic overloading pattern, indicative mild right ventricular hypertrophy, seen in patients with atrial septal defects.

2.2.3 Echocardiogram

Echocardiographic studies reveal enlarged right ventricle with paradoxical septal motion, particularly well-demonstrable on M-mode echocardiograms in patients with moderate to large ASDs. Dilatation of the right ventricle may not be present in small defects. By two-dimensional echocardiogram, the defect can be clearly visualized (Figure 3 left panel).
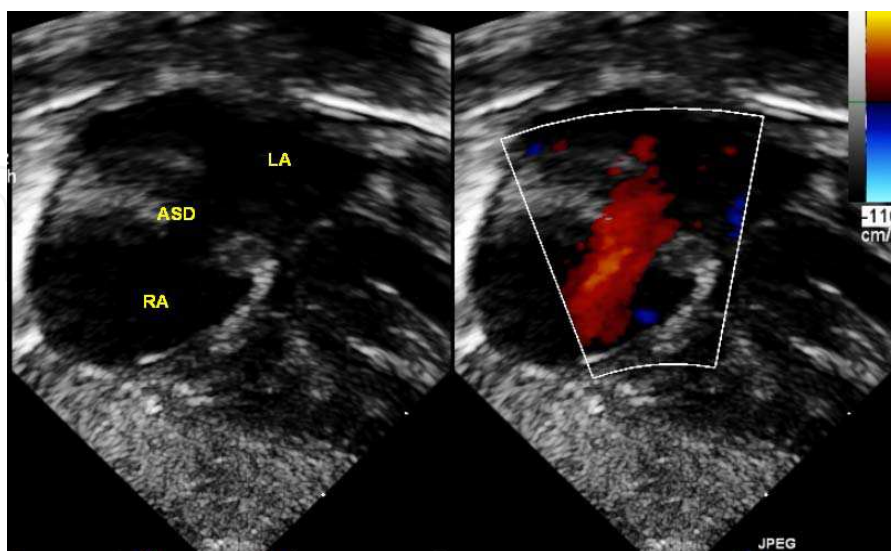

Fig. 3. Two dimensional subcostal echocardiographic views of the atrial septum demonstrating secundum atrial septal defect (ASD) in the mid septum (left panel) and color Doppler with left to right shunt (right panel). LA, left atrium; RA, right atrium.

The type of ASD, ostium secundum (Figure 3) versus ostium primum (Figure 4) can also be delineated by the echocardiographic study.
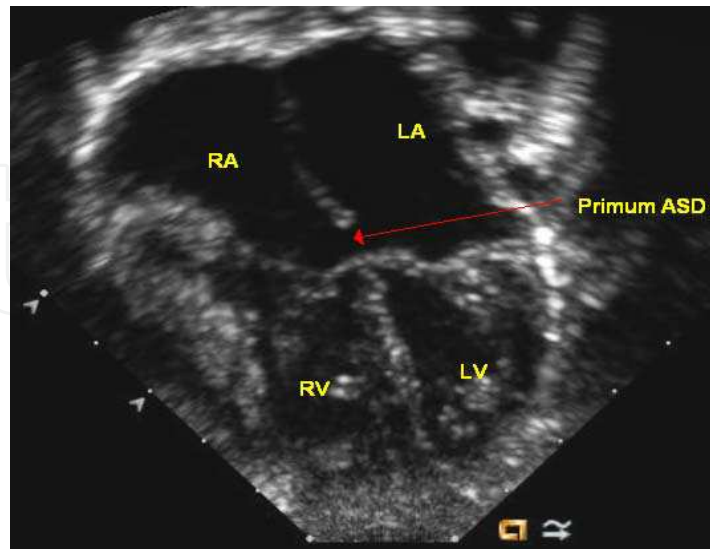

Fig. 4. Four chambered view of the heart demonstrates ostium primum atrial septal defect (ASD), arrow. Note absence of any atrial septal tissue superior to the crest of the ventricular septum. The right atrium (RA) and right ventricle (RV) are enlarged. LA, left atrium; LV. left ventricle.

Apical and precordial views may show "septal drop-outs" without an ASD because of thinness of the septum in the region of fossa ovalis. Therefore, subcostal views should be scrutinized for evidence of ASD. In addition, demonstration of flow across the defect with pulsed Doppler and color Doppler (Figure 3, right panel) echocardiography is necessary to avoid false positive studies. In adolescents and adults transesophageal echo (TEE) is needed to make definitive diagnosis of ASD. (Figures 5 and 6)
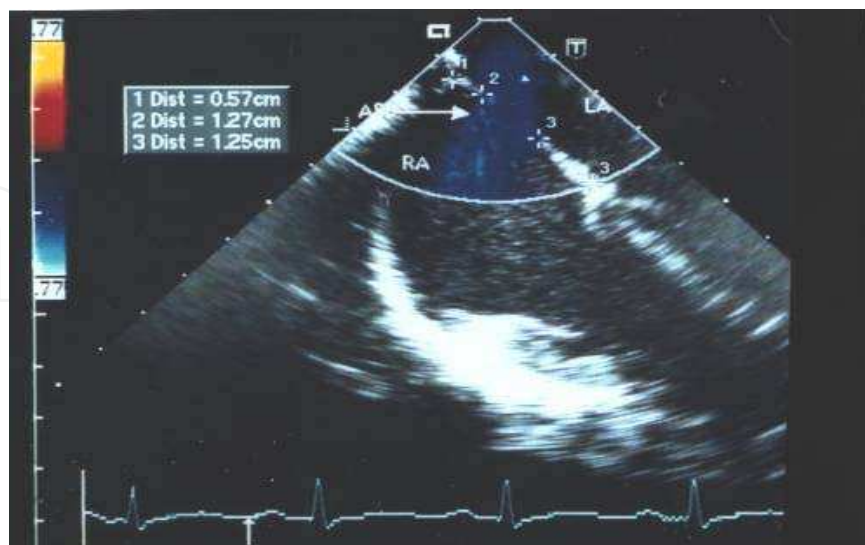

Fig. 5. Selected two-dimensional and color flow frame from a transesophageal echocardiographic (TEE) study (in adult patient) of the atrial septum shows an atrial septal defect (arrow) with left to right shunt (blue flow). Measurements of septal margins (1 Dist and 3 Dist) and of the defect (2 Dist) are shown in the insert. LA, left atrium; RA, right atrium.
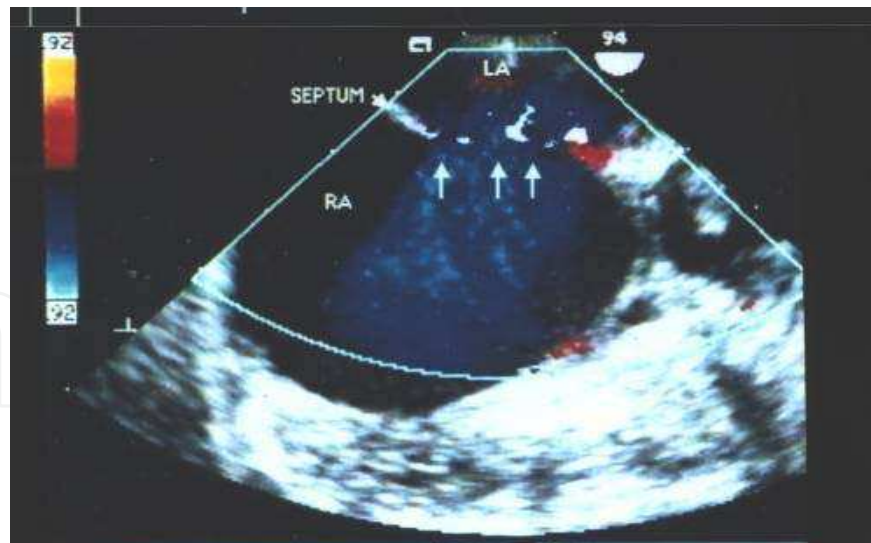
Fig. 6. Selected two-dimensional and color flow frame from a transesophageal echocardiographic (TEE) study (in another adult patient) of the atrial septum shows multi-fenestrated atrial septal defect (arrows) with left to right shunt (blue flow). LA, left atrium; RA, right atrium.

2.2.4 Other imaging studies

Other imaging studies such as three-dimensional echo, MRI and CT can and do demonstrate the defect, but are not necessary for routine cases.

2.3 Catheterization and angiography

Clinical and echocardiographic features are sufficiently characteristic so that cardiac catheterization is not necessary for the diagnosis. However, cardiac catheterization is an integral part of transcatheter occlusion of the ASD.

When catheterization is performed, one will observe step-up in oxygen saturation at the right atrial level. The right ventricular or pulmonary arterial saturations may be better to estimate the degree of shunting because of improved mixing in these distal sites. The pulmonary venous, left atrial, left ventricular and aortic saturations are within normal range. In large defects, the pressures in both atria are equal while in small defects, an inter-atrial pressure difference is noted. The right ventricular and pulmonary arterial pressures are usually normal during childhood. Calculated pulmonary-to-systemic flow ratio (Qp:Qs) is used to quantify the degree of shunting and a Qp:Qs in excess of 1.5:1 is considered an indication for closure of ASD.

Selective cineangiography in the right upper pulmonary vein at its junction with the left atrium in a left axial oblique view will reveal location and the size of the ASD. When anomalous pulmonary venous connection is suspected, selective left or right pulmonary arterial angiography should be performed and the levophase of angiogram should be scrutinized for anomalous pulmonary venous connections.

To avoid missing a diagnosis of partial anomalous pulmonary venous return, we usually perform a number of routine maneuvers and these include (i) measurement of oxygen

saturations from both right and left innominate veins at the time of superior vena caval sampling, (ii) left innominate vein cineangiogram in posterior-anterior view with diluted contrast material, (iii) probe for all the four pulmonary veins from the left atrium and (iv) as mentioned before, obtain cineangiography from the right upper pulmonary vein at its junction with the left atrium in a left axial oblique (30^0 LAO and 30^0 cranial) view.

2.4 Management

The management of ASD patients is largely dependent of the age at presentation, presence of symptoms, particularly those of congestive heart failure and the size of the defect (and magnitude of the shunt).

2.4.1 Medical management

As mentioned earlier, congestive heart failure is rare with ASDs, although occasionally, failure symptoms may be present in infancy. In these infants anti-congestive measures (diuretics and digoxin) should be instituted. If they do not improve, surgical and more recently trans-catheter intervention to close the defects are considered.

Small ASDs, not requiring closure may be followed at infrequent intervals. SBE prophylaxis and activity restriction are not generally recommended for ASD patients.

2.4.2 Indications for closure

Despite lack of symptoms at presentation, closure of moderate to large ASDs is recommended so as to 1) prevent development of pulmonary vascular obstructive disease later in life, 2) reduce chances for supra-ventricular arrhythmias and 3) prevent development of symptoms during adolescence and adulthood. Elective closure around age 4 to 5 years is recommended. Closure during infancy is not undertaken unless the infant is symptomatic. Right ventricular volume overloading by echocardiogram and a Qp:Qs >1.5 (if the child had cardiac catheterization) are indications for closure.

2.4.3 Surgical management

Following the introduction of cardiopulmonary bypass techniques for open heart surgery and the description of surgical closure of ASD by Gibbon, Lillehei and Kirklin in 1950s, it rapidly became a standard treatment for atrial defects. The conventional treatment of choice of moderate and large defects until recently is surgical correction. Under general anesthesia, a median sternotomy or a right submammary incision is made, the aorta and vena cavae are cannulated and the patient placed on cardiopulmonary bypass. Right atriatomy is made and the defect exposed and closed either by approximating the defect margins with suture material or by using a pericardial patch, depending upon the size of the defect.

While surgical closure of ostium secundum ASDs is safe and effective with low (<1%) mortality, the morbidity associated with sternotomy/thoracotomy, cardiopulmonary bypass and potential for postoperative complications cannot be avoided. Other disadvantages of surgical therapy are the expense associated with surgical correction, residual surgical scar and psychological trauma to the patients and/or the parents. Because of these reasons

several trans-catheter methods have been developed (Chopra and Rao, 2000; Rao, 2003) which will be reviewed in the next section.

At the present time, surgical repair is largely reserved for defects with poor septal rims in which the interventional cardiologist deems that defect is difficult to close with trans-catheter methodology or was unsuccessful in closing the defect. Also, if intra-cardiac repair of other defects is contemplated, surgical closure of ASD could be performed at the same time.

2.4.4 Trans-catheter closure

As alluded to above, a large number of devices have been developed over the last three and one-half decades. Some of the devices have been discontinued and others modified and redesigned (Rao, 1998; Rao, 2000; Rao, 2003b). Clinical trials have been undertaken with a large number of devices as reviewed elsewhere (Rao, 2000; Rao, 2003b) and feasibility, safety and effectiveness of these devices in occluding the ASD have been demonstrated.

Clinical trials have been undertaken in a large number of patients with Bard clamshell septal occluder and buttoned device and feasibility and effectiveness of these devices in occluding the ASD have been demonstrated. Fractures of one or more arms of the clamshell device with occasional embolization, has prompted the investigators and the US Food and Drug Administration (FDA) to withdraw the device from clinical trials. The buttoned device has undergone clinical trials and, immediate and short-term follow-up results are encouraging (Rao et al 1992, Rao et al 1994, Rao et al 2000, Rao and Sideris 2001). However, pre-market-approval (PMA) application was not made and consequently it is not approved by the FDA and is not available for general clinical use. Subsequently, a large number of other devices (Das Angel-Wing, ASDOS, Amplatzer, CardioSeal, HELEX and others) have been introduced and clinical trials began (Chopra and Rao 2000). At the present time however, Amplatzer Septal Occluder and HELEX are the only two devices that are approved for general clinical use by the FDA. The experience with Amplatzer for most defects has been encouraging. HELEX device is only useful in small to medium-sized defects. A number of other devices are in clinical trials either in the US or in other countries with local, national or regional IRB supervision. These devices, to the best of my knowledge, are CardioSeal/StarFlex devices, transcatheter patch, pfm ASD-R device, bio-absorbable NMT devices (Bio-STAR and Bio-TREK), Occlutech Flex device, Cardia devices (INTRASEPT, ATRIASEPT I/II-ASD and ULTRASEPT), Solysafe Septal Occluder, Heart R Septal Occluder (manufactured in China) and others. The Amplatzer Septal Occluder is rapidly becoming the device of choice because of ease with which the device can be implanted, retrieved and repositioned plus the comfort that the device is FDA approved.

2.4.4.1 Amplatzer septal occluder

Amplatzer septal occluder is a double disk device constructed with 0.004" to 0.007" Nitinol (nickle-titanium compound) wire with shape memory. A 4 mm wide waist connects the left and right atrial disks and stents the ASD. The left atrial disk is slightly larger than the right. Dacron polyester patches are sewn into each disk. Multiple sizes are available from the manufacturer (AGA); the device size is expressed as the size of waist of the device. The device can be withdrawn into a delivery sheath and can be implanted across the defect and if necessary pulled back into the sheath and repositioned.

2.4.4.1.1 Method of device implantation

The procedure involves percutaneous right heart catheterization to confirm the clinical and echocardiographic diagnosis with particular attention to exclude partial anomalous pulmonary venous return. A left atrial cineangiogram in a left axial oblique view (30^0 LAO and 30^0 Cranial) with the catheter positioned in the right upper pulmonary vein at its junction with the left atrium is then performed. This is followed by transesophageal (TEE) or intracardiac (ICE) echocardiography to measure the size of the ASD, to visualize entry of all pulmonary veins into the left atrium and to examine the atrial septal rims. Static balloon sizing of the ASD using NuMed PTS or AGA Amplatzer sizing balloons is performed routinely by some cardiologists. During balloon occlusion, color Doppler evaluation of the atrial septum to rule out additional atrial defects should be carried out. However, I do not routinely perform balloon sizing, but rely on the TEE sizing; I utilize the thick margins of the defect to measure the size of the ASD, leaving out the flail margins, a method similar to that suggested by Carcagnì and Presbitero (2004).

An Amplatzer Septal Occluder that is 1 to 2 mm larger than the diameter of the ASD is selected for implantation. The size of delivery sheath accommodating the selected device is then be positioned in the left upper pulmonary vein, taking appropriate precautions to avoid inadvertent air entry into the system. The selected device is screwed onto the delivery cable; the device is loosened by unscrewing by one turn and drawn into the loader sheath under saline. The device is deposited into the delivery sheath while flushing the loader sheath continuously with saline or a similar flushing solution. This is to prevent inadvertent air entry into the system. The device is advanced within the sheath under fluoroscopic guidance until it reaches the tip of the delivery sheath in the left upper pulmonary vein. It is important not to rotate the delivery cable to prevent inadvertent unscrewing of the device. The entire system is withdrawn until the tip of the sheath slips into the free left atrium and the device advanced, thus releasing the left atrial disk. Under echocardiographic guidance, the entire system is withdrawn such that the left atrial disk is flush against the left atrial side of the atrial septum occluding the ASD. Then, while the device cable is held steady, the delivery sheath is withdrawn releasing the waist of the device within the atrial septal defect, followed by further withdrawal of the sheath deploying the right atrial disk in the right atrium. The position of the device is verified by echocardiography and residual shunt looked for. If the device position is satisfactory, the device cable is moved back and forth (so called Minnesota Wiggle). The position of the device is again verified by TEE (or ICE). If the device position is unsatisfactory, the device can be withdrawn into the sheath and redeployed. Then the device cable is rotated counterclockwise, releasing the device. A repeat TEE to ensure good position of the device is undertaken. Right atrial cineangiography through the delivery sheath is performed by some cardiologists prior to withdrawal of the delivery sheath out of body.

Arterial line to monitor the systemic pressures throughout the procedure, administration of heparin (100 units/kg) and monitoring the ACT to keep it above 200 seconds, and administration of Ancef or a similar antibiotic are routine parts of the procedure. Aspirin 5 mg/kg as a single daily dose for six months is usually recommended. Clopidogrel (Plavix) is used in adult patients.

2.4.4.1.2 Complex defects

Large defects, small septal rims, multiple defects and septal aneurysms pose additional problems and appropriate adjustments in the technique (Nagm and Rao 2004) should be undertaken to ensure success of the device implantation.

2.4.4.1.3 Results

Both immediate and mid-term follow-up results of Amplatzer Septal Occluder appear excellent with immediate complete closure rates varying from 62% to 96% which improved to 83% to 99% at six to 12 month follow-up (Hamdan et al 2003). We undertook closure of 80 ostium secundum defects with this device; there was a small residual shunt in two patients at the conclusion of the procedure. This shunt disappeared at one and six month follow-up visits respectively. No residual shunts were observed during a mean follow-up of 24 months.

2.4.4.2 HELEX device

HELEX device is constructed with a single stand super-elastic, Nitinol wire frame with ultrathin poly-tetra-fluro-ethelene (ePTFE) covering the entire length of the wire; the device can be loaded into a 9-F delivery sheath. The delivery system has three components, a delivery catheter, control catheter and a mandrel. When deployed, it forms two interconnected round disks, designed to be placed on either side of the atrial septum. The device is available in 15 thru' 35 mm diameter sizes in 5 mm increments.

2.4.4.2.1 Method of device implantation

The procedures of catheterization and defect sizing are similar those described in Amplatzer device section. The method of implantation is detailed elsewhere (Latson et al 2003). In brief, the delivery catheter (Green) is placed in the left atrium over a guide wire and the wire removed. Push-pinch-pull method is used to form the left atrial disk and the disk pulled back gently to engage the left side of the atrial septum, under fluoroscopic and/or TEE or ICE guidance. Then the delivery (Green) catheter is withdrawn over the control (Gray) catheter until the mandrel (Tan) engages the hub. Then the green catheter is held study while the gray catheter is advanced to deliver the right atrial disk on the right atrial side of the septum, again using the "push-pinch-pull" technique. Once the device position is verified by echocardiography (TEE or ICE), the device is locked and then released. Intra and post procedural management is similar to that described in the Amplatzer device section.

2.4.4.2.2 Results

Results of the multicenter trial (Jones et al 2007) suggest successful implantation in 87% patients with low incidence of residual leaks (2.6% at one year follow-up) and modest incidence (8%) of wire frame fractures. It is generally considered to be a good device for occlusion of small to medium-sized ASDs.

2.5 Prognosis

The prognosis following surgical or transcatheter closure of ASDs is excellent, provided that they do not have pulmonary hypertension or atrial tachycardia. Actuarial survival rate following surgery were 97%, 90%, 83% and 74% at 5, 10, 20 and 30 years respectively (Murphy et al 1990) and were slightly worse than that of control (normal) population (99%,

98%, 94% & 85%). However, if surgical correction is performed prior to 25 years of age, the actuarial survival rates are similar to normal population. Similar favorable results can be expected if the defect is closed by trans-catheter methodology prior to 25 years of age.

3. Ostium primum ASDs

Ostium primum ASDs belong to the group of defects called atrio-ventricular septal defects (AVSDs) and are thought to be caused by defective embryonic development of embryonic endocardial cushions. There is persistence of the embryonic ostium primum, located in the posterior portion of the lower part of the atrial septum, usually large in size. A cleft in the anterior leaflet of the mitral valve is present, causing mitral insufficiency of varying degree. Depending upon the direction of mitral insufficiency jet, there may be a left ventricular-to-right atrial shunt as well. A cleft in the septal leaflet of the tricuspid valve may be present in some patients. These defects are formerly known as partial endocardial cushion defects. These defects are also called partial AVSDs; this is in contradistinction to complete AVSDs in which atrial and ventricular septal defects and clefts in the mitral and tricuspid valves with common atrio-ventricular valve are present. There may be associated ostium secundum ASD, patent foramen ovale or a persistent left superior vena cava draining into the coronary sinus.

The left ventricular outflow tract is long and narrow and sometimes the abnormal attachments of the atrio-ventricular valve tissue may cause left ventricular outflow tract obstruction.

Dilatation of the right heart structures is similar to that described for ostium secundum atrial septal defects. In the presence moderate to severe mitral insufficiency left ventricular dilatation may also be present.

3.1 Clinical features

The clinical features are essentially similar to that described for ostium secundum ASDs; however in the presence of significant mitral insufficiency symptoms of heart failure may be present.

3.1.1 Symptoms

Isolated ostium primum ASD patients are usually asymptomatic and are most often detected at the time of preschool physical examination. However, murmurs associated with mitral insufficiency of ostium primum defects may also result in early detection of these defects. A few patients do present with symptoms of heart failure in infancy or childhood especially in the presence of significant mitral insufficiency.

3.1.2 Physical examination

The right ventricular and right ventricular outflow tract impulses are increased and hyperdynamic. No thrills are usually felt. The second heart sound is widely split and fixed (splitting does not vary with respiration) and is the most characteristic sign of ASD. Ejection systolic clicks are rare with ASDs. The ejection systolic murmur of ASD is soft and is of

grade I-II/VI intensity and rarely, if ever, louder. The murmur is secondary to increased blood flow across the pulmonary valve and is heard best at the left upper sternal border. A grade I-II/VI mid-diastolic flow rumble is heard (with the bell of the stethoscope) best at the left lower sternal border. This is due to large volume flow across the tricuspid valve. There is no audible murmur because of flow across the ASD. A holosystolic murmur of mitral insufficiency is heard best at the apex with radiation into the anterior and/or mid axillary line. A grade I-II/VI mid-diastolic flow rumble, heard best at the apex may be appreciated in the presence significant mitral insufficiency. Signs of heart failure may be present in cases with severe mitral insufficiency.

3.2 Noninvasive evaluation

3.2.1 Chest x-ray

Chest film usually reveals mild to moderate cardiomegaly, prominent main pulmonary artery segment and increased pulmonary vascular markings. In the presence of significant mitral insufficiency, the cardiomegaly may be more prominent.

3.2.2 Electrocardiogram

Prolongation of PR interval (first degree heart block) is commonly seen. Right atrial, left atrial or biatrial enlargement is seen nearly half of the patients. The ECG also shows mild right ventricular hypertrophy; the so-called diastolic volume overload pattern with rsR' pattern in the right chest leads. Left ventricular hypertrophy may be seen if there is significant mitral insufficiency. Characteristically, the mean frontal plane vector is oriented superiorly between -30^0 and -90^0, the so called left axis deviation and this is typical for endocardial cushion defects.
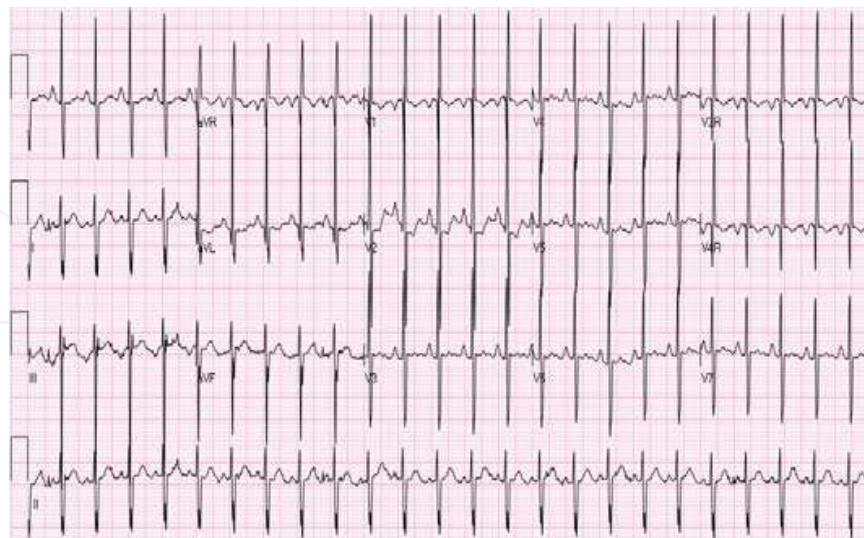

Fig. 7. An electrocardiogram of a child with ostium primum atrial septal defect demonstrating left axis deviation (-45^0 - deep S waves in leads II, III and AVF), right atrial enlargement (tall P waves in leads I and V2) and right ventricular hypertrophy (tall R waves in lead V2 and deep S waves in leads V5 and V6).

3.2.3 Echocardiogram

Echocardiographic studies reveal enlarged right ventricle with paradoxical septal motion, particularly well-demonstrable on M-mode echocardiograms in patients with moderate to large ASDs. By two-dimensional echocardiogram, the defect can be clearly visualized (Figure 4). The type of ASD, secundum versus primum can also be delineated by the echocardiographic study (Figure 3 & 4). Demonstration of flow across the defect with color Doppler (Figure 8) echocardiography is possible. Cleft in the mitral valve may be demonstrated in precordial short axis views and mitral insufficiency jet may be shown in four chamber views (Figure 8).
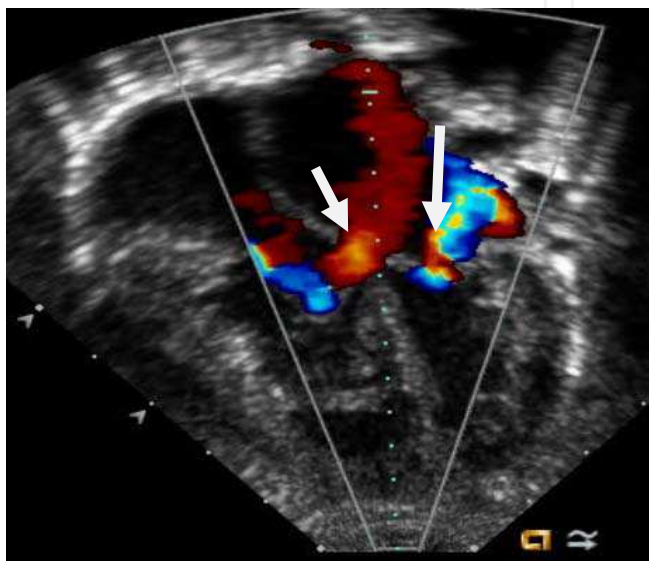

Fig. 8. Four chambered view of the heart demonstrates left-to-right shunt (red flow) across the ostium primum atrial septal defect (short arrow). Also note mitral insufficiency (long arrow).

3.2.4 Other imaging studies

Other imaging studies such as three-dimensional echo, MRI and CT may also demonstrate the defects, but are not necessary for routine cases.

3.3 Catheterization and angiography

Clinical and echocardiographic features are characteristic for the defect and cardiac catheterization is not necessary for the diagnosis. If pulmonary hypertension is suspected or if there are issues that can't be resolved by echocardiography, catheterization may be undertaken.

If catheterization is performed, step-up in oxygen saturation at the right atrial level is seen. The left heart saturations are within normal range. Because the defects are usually large, the mean pressures in both atria are equal. The right ventricular and pulmonary arterial pressures are usually normal during childhood. The left heart pressures are also normal unless there is left ventricular outflow tract obstruction. Calculated pulmonary-to-systemic

flow ratio (Qp:Qs) is used to quantify the degree of shunting and the Qp:Qs is usually in excess of 2:1. Pulmonary vascular resistance is usually normal.

Selective left ventricular cineangiography reveals a long and narrow left ventricular outflow tract resulting in what is described as goose-neck deformity, characteristic of endocardial cushion defects.

3.4 Management

The management of ostium primum ASD patients is largely dependent of the age at presentation and presence of symptoms, particularly those of congestive heart failure.

3.4.1 Medical management

Congestive heart failure is rare with ostium primum ASDs, although failure symptoms may be present in the presence of significant mitral insufficiency. In these patients anti-congestive measures (diuretics and digoxin) should be instituted. If they do not improve, surgical closure should be considered.

Transcatheter occlusion, now a standard treatment for ostium secundum ASDs, is not feasible in patients with ostium primum ASDs because there are no inferior septal rims, but more importantly because the need for addressing mitral valve cleft and the accompanying mitral insufficiency.

SBE prophylaxis is recommended and normal activity is permitted in the absence of severe mitral insufficiency.

3.4.2 Indications for closure

Although surgical correction can be performed at any age, surgery in asymptomatic patients is usually recommended at the age of 3 to 4 years. In the presence of symptoms or if there is associated severe mitral insufficiency, surgical repair may be performed at presentation, after medically controlling the heart failure.

3.4.3 Surgical management

The conventional treatment of choice of ostium primum ASDs is surgical correction. Under general anesthesia, a median sternotomy incision is made, the aorta and vena cavae are cannulated and the patient placed on cardiopulmonary bypass. Right atriotomy is made and the defect and mitral valve are exposed. Closure of the mitral valve cleft with interrupted suture material and additional reparative procedures to address observed mitral valve abnormalities (for example, annuloplasty) should be undertaken. Then the atrial defect is closed using an autologous pericardial patch and rarely other prosthetic material (Dacron or Gore-Tex). Associated ostium secundum ASD or a patent foramen ovale should also be surgically closed at the same sitting.

3.4.4 Results

Results are generally good with a mortality rate less than 3%. The risk factors for poor results are severe mitral insufficiency, failure to thrive and congestive heart failure.

3.5 Prognosis

The prognosis is generally good. The actuarial survival at 20- and 40-year follow-up was 87% and 76% respectively for a large group of patients that had repair of ostium primum ASDs at Mayo Clinic (El-Najdawi et al 2000). The survival was better if the mitral valve repair was performed prior to 20 years of age. Repeat surgery, mostly to address mitral valve disease was required in 11% patients. Development of sub-aortic stenosis and heart block, requiring intervention occurs in a minority of patents during long-term follow-up.

4. Sinus venosus ASDs

Sinus venosus defects constitute 5 to 10% of all ASDs and the majority of defects are located in the posterior superior portion of the inter-atrial septum, often overriding the superior vena caval orifice. These defects are frequently associated with anomalous connection of the right upper pulmonary veins to the superior vena cava or right atrium near the cavo-atrial junction. The right pulmonary veins from the entire right lung may be connected anomalously. Rarely, the defect may be located in the inferior-posterior part of the atrial septum, overriding the inferior vena caval orifice. The dilatation of right heart structures is similar to that described in ostium secundum ASDs as are the clinical features. The ECG, in addition to the findings of rsR' pattern of the QRS complex shows somewhat superiorly oriented P wave vector ($<30^0$). Echocardiogram shows right ventricular volume overloading, similar to ostium secundum ASDs, but without an obvious ASD in the secondum position. Subcostal views may show the defect. Turbulence in the right upper pulmonary veins may also help suspect this diagnosis. The indications for intervention are also similar to those discussed in the ostium secundum ASD section. However, these defects are not amenable to transcatheter closure and surgical correction is the treatment of choice. Diversion of the anomalously connected right pulmonary vein(s) into the left atrium along with the closure of the ASD should be undertaken. This may involve constructing a tunnel with an autologous pericardial patch along with enlargement of superior vena cava.

5. Coronary sinus ASDs

These are rarest types of ASDs; these are defects in the inferior and anterior portion of the atrial septum at the expected location of the orifice of the coronary sinus. These defects are often associated with a persistent left superior vena cava and unroofing of the coronary sinus, a complex described as Raghib syndrome. The defect may be seen in association with asplenia syndrome. Dilatation of right heart structures and clinical features are similar to that described in ostium secundum ASD section. Echocardiogram is useful in the evaluation and diagnosis of this anomaly. Surgical correction with patch closure of the defect, leaving the entry of coronary sinus in the left atrium is the conventional method of approach (Lee and Sade 1979). These defects are not usually amenable to transcatheter closure. However, some, particularly small, defects may be amenable to transcatheter occlusion (Di Bernardo et al 2003)

6. Patent foramen ovale

To complete the discussion of defects in the atrial septum, a brief review of PFO is in order. The foramen ovale in the fetus is kept patent because of the mechanical effect of streaming

of the inferior vena caval blood into the left atrium. At birth, a combination of increase in the left atrial pressure secondary to increased pulmonary venous return and decrease in the right atrial pressure due to eliminated placental return will result in apposition of the septum primum and septum secundum causing functional closure of the foramen ovale. Eventually anatomical closure occurs in most normal individuals.

Continued patency of the foramen ovale is critical in neonates with right heart obstructive lesions (tricuspid or pulmonary atresia) as well as left-sided obstructive lesions (hypoplastic left heart syndrome and mitral or aortic atresia) so as to allow an obligatory right-to-left or left-to-right shunt across the atrial septum, respectively. Similarly the patency of the foramen ovale is important in total anomalous pulmonary venous connection where all pulmonary and systemic venous returns come into the right atrium and consequently the systemic blood flow is entirely derived from right-to-left shunting across the PFO. In neonates with transposition of the great arteries (TGA), the circulation is parallel (instead of normal in-series circulation) and some inter-circulatory mixing is essential for survival; this is usually provided by the ASD/PFO. With any of the above scenarios the foramen ovale can become restrictive and may need enlargement either by transcatheter or surgical methodology (Rao 2007).

In patients with large patent ductus arteriosus (PDA) or ventricular septal defect (VSD), the pulmonary blood flow is markedly increased with consequent increase in the left atrial size; this left atrial enlargement may cause stretching of the patent foramen ovale resulting in an additional left-to-right atrial shunting. However, in clinical practice, the clinician needs to address the primary cardiac problem (PDA or VSD) and the PFO will either spontaneously resolve or become small so that it may not remain clinically significant.

The prevalence of PFO, based on autopsy studies is 27%; this incidence was 34% in the first thirty years of life which decreased to 25% in 30 to 80 year-olds which further decreased to 20% in 80 to 100-year-olds (Hagen 1984). Similar prevalence was observed by TEE examinations. Consequently the PFO should be considered a normal variant. However, some these PFOs are considered to be the seat of right to left shunt causing paradoxical embolism and cerebrovascular accidents (Lechat et al 1988, Webster et al 1988, Ende et al 1996, Windecker and Meier 2003) and hypoxemia as seen in platypnea-orthodeoxia syndrome (Waight et al 2000, Rao et al 2001). Right to left shunt thru' PFO can also occur in patients who were previously treated for complex congenital cardiac anomalies (Rao et al 1997), including Fontan fenestrations as well as in patients who had right ventricular infarction (Bassi et al 2005). Decompression (Caisson's) illness (Wilmshurst et al 1996, Walsh et al 1999, Wilmshurst et al 2000) and migrane (Wilmshurst et al 2000) have also been attributed to right to left shunt across PFO. There is varying degrees of evidence regarding the benefits of transcatheter occlusion of PFOs in above described conditions; some of these issues are addressed in later chapters of this book.

7. Summary and conclusions

In this review, the clinical features and management of ASDs are discussed. Four types of defects namely, ostium secundum, ostium primum, sinus venosus and coronar sinus ASDs are included. Patients with small defects, especially in childhood, are usually asymptomatic while moderate to large defects in infancy, though rarely, may present with symptoms.

Physical findings include hyperdynamic precordium, widely split and fixed second heart sound, ejection systolic murmur at the left upper sternal border and a mid-diastolic flow rumble at the left lower sternal border. Clinical diagnosis is not usually difficult and the diagnosis can be confirmed and quantified by non-invasive echocardiographic studies. Whereas surgical intervention was used in the past, transcatheter methods are currently used for closure of ostium secondum ASDs. Surgical correction is usually necessary for the other three types of defects.

PFO is present in nearly one third of normal population and is likely to be a normal variant. In the presence of some structural abnormalities of the heart, their presence may facilitate intra-cardiac shunt to allow appropriate egress and/or mixing of blood flow. PFOs, presumed to be the seat of paradoxical embolism resulting in stroke/transient ischemic attacks deserve special consideration. Hypoxemia in post-surgical residual defects including Fontan fenestrations and right ventricular infarction may be secondary to right to left shunt across PFO. Other problems such as migraine, Caisson's disease and platypnea-orthodexia syndrome are also attributed to shunts across PFO. Evidence for benefit of transcatheter occlusion of these PFOs is variable.

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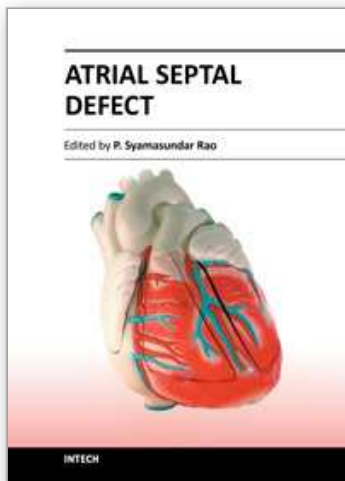
Atrial Septal Defect

Edited by Dr. P. Syamasundar Rao

Atrial Septal Defects (ASDs) are relatively common both in children and adults. Recent reports of increase in the prevalence of ASD may be related use of color Doppler echocardiography. The etiology of the ASD is largely unknown. While the majority of the book addresses closure of ASDs, one chapter in particular focuses on creating atrial defects in the fetus with hypoplastic left heart syndrome. This book, I hope, will give the needed knowledge to the physician caring for infants, children, adults and elderly with ASD which may help them provide best possible care for their patients.

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