Atrial Septal Defect

Dimitrios V. Moysidis, Eleftherios Gemousakakis, Alexandros Liatsos, Christos Tsagkaris, and Andreas S. Papazoglou

Abstract

"Atrial septal defects (ASDs) are a frequent type of congenital heart disease characterized by an abnormal communication between the two atria of the heart, which leads to a left-to-right shunt." They are formed during embryological development due to disorders in atrial septation. Most ASDs are of small size and therefore are asymptomatic during infancy and childhood. However, most of them, except for some patients who have mild or nonspecific symptoms, become obvious by the age of 40 years, when they cause clinical manifestations, such as fatigue, exercise intolerance, and dyspnea. In childhood, the vast majority of isolated ASDs close spontaneously. Large and persistently symptomatic moderate-sized defects should be repaired. Intervention is also required in adulthood in symptomatic patients, right ventricular overload, left-to-right shunt, or paradoxical embolism. Transcatheter device closure is always preferred against surgical repair.

Keywords

Congenital heart disease · Atrial septal defects · Atrial septation · Ostium secundum · Crochetage sign
Left-to-right shunt · Eisenmenger syndrome
Spontaneous closure · Paradoxical embolism
Transcatheter device closure

Introduction

"Atrial septal defects (ASDs) constitute a frequent type of congenital heart disease characterized by an abnormal communication between the two atria of the heart through an insufficient interatrial septal tissue" [1] (Fig. 1). The prevalence of ASD is estimated to be approximately 1.6 per 1000 live births and 0.88 per 1000 adults while ASDs account for 7–10% of all congenital heart defects (CHDs). During the last two decades, an increase in the incidence of new ASD cases has been observed, which however is probably attributed to the widespread use of echocardiography. The prevalence of ASD is also higher in females [2]. In a large database of European patients with CHDs, conducted in the Netherlands, 67% of patients suffering from ASD were females [3].

Geographic distribution seems to be unequal too, with the highest ASD birth prevalence found in Asia, North America, and Europe [4]. Familial history of congenital heart anomalies increases the risk of a secundum ASD, which is further increased when at least one sibling has an ASD [5]. ASDs, in most cases, present as isolated defects, but they might also complicate other diseases or be a part of a syndrome, such as Down syndrome, Hurler syndrome, Noonan syndrome, chondroectodermal dysplasia, congenital rubella, and others [6].

Isolated ASDs are most commonly diagnosed in asymptomatic patients during their first two decades of life. Therefore, many patients get diagnosed incidentally during an echocardiographic investigation of a murmur. The age in which symptoms may occur depends on the size of the defect. Patients with smaller ASDs (less than 5 mm) might never present with clinically significant symptomatology. These defects are most likely to close spontaneously. Bigger defects (5–10 mm) usually lead to symptoms during the fourth or fifth decade, while defects with a diameter of more than 10 mm can become clinically obvious during the third decade of life [6].

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G. Tagarakis et al. (eds.), *Clinical and Surgical Aspects of Congenital Heart Diseases*,
https://doi.org/10.1007/978-3-031-23062-2_5

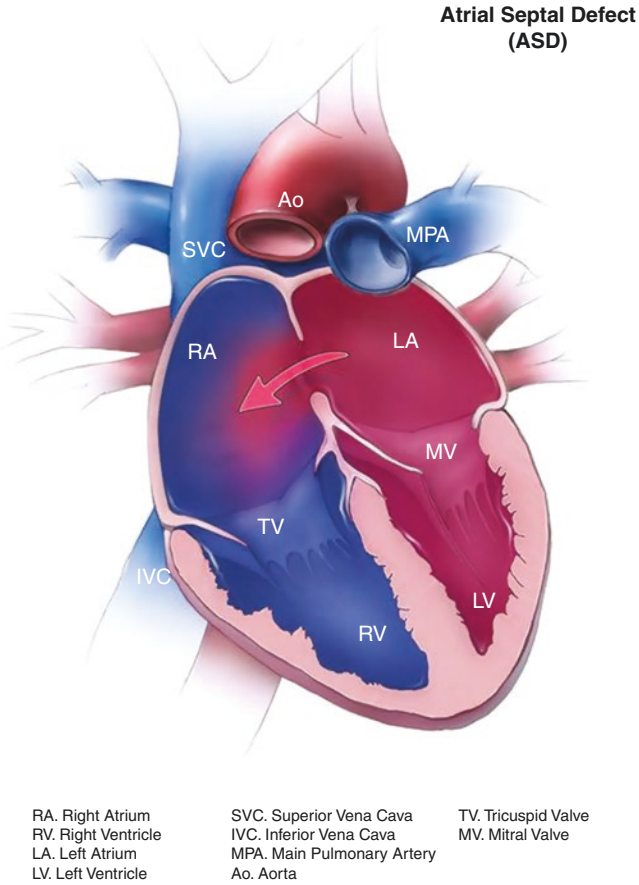

Fig. 1 Atrial septal defect illustration (*Adapted from Wikipedia commons (free copyright license)*)

Patients often present with fatigue, shortness of breath, palpitations, exercise intolerance, and syncope. If left undiagnosed and untreated, ASDs could eventually cause right-sided heart failure, manifestations of thromboembolism, and cyanosis associated with Eisenmenger syndrome. Infants with significant defects might rarely present with tachypnea, recurrent respiratory infections, and failure to thrive [7]. The most common cause of death in patients with unrepaired defects is heart failure [8]. Untreated patients and even patients, who do not receive early diagnosis and appropriate management, may experience higher rates of mortality and morbidity. On the contrary, an early and proper management approach increases event-free survival rates. However, the risk of atrial arrhythmias and thromboembolism seems to remain higher in comparison to the general population [9].

Pathophysiology

Lack of sufficient tissue to completely septate the atria is the causing factor of ASDs. Foramen ovale is vital for fetal circulation, as flow through this lesion is essential for systemic circulation perfusion. Atrial septal embryonic development is depicted in Fig. 2. Defects in any stage of this process can lead to the occurrence of ASDs. The predisposing risk factors and associated conditions (i.e., maternal exposure to chemicals, alcohol consumption during the first trimester, and gestational diabetes) have been long studied; however, their relationship has not yet been established [10, 11].

Based on the location of the defect and, by extension, of the liable embryogenetic abnormality, ASDs are classified into four types (Fig. 3):

1. Ostium secundum, which is located in fossa ovalis, represents the majority of ASDs, accounting for about 70% of all cases. It is the result of either poor growth of the secundum septum or its excessive absorption [12]. It is usually an isolated congenital heart disorder, but it might also be associated with other congenital cardiac and extra-cardiac abnormalities. Holt–Oram syndrome, which is also known as the heart-hand syndrome, is the most well-described disorder frequently accompanied by a secundum ASD [13].
2. Ostium primum type, which accounts for 15–20% of ASDs, is formed when primum septum fails to fuse with the endocardial cushions at the base of the interatrial septum [12]. It can be either isolated or, most commonly, associated with atrioventricular (AV) valve anomalies, particularly a cleft in the anterior mitral valve leaflet [13, 14]. This comorbidity can also be accompanied by a contiguous defect in the inlet ventricular septum. This combination of primum ASD, cleft mitral valve, and an inlet ventricular septal defect, is known as a partial AV septal defect (AVSD), whose most severe form is the complete AV septal (or canal) defect [15].
3. Superior and inferior sinus venosus defects, which account for approximately 5–10% of all ASDs, are located in the venoatrial part of the atrial septum [13]. They represent an abnormality in the insertion of the superior or inferior vena cava, which overrides the interatrial septum. These defects are technically not considered to be ASDs since the defect is within the sinus venosus septum.
4. Coronary sinus defects, which is the most uncommon type of ASD, account for less than 1% of ASD patients.

ASDs in adults can give birth to a left-to-right shunt which generates volume overload of the right heart chambers. The severity of this shunt, as well as the direction and magnitude of blood, is determined by the size of the defect and the relative atrial pressures, which are linked to the compliances of the ventricles. These parameters also determine the natural course of isolated ASDs, which varies from spontaneous closure to asymptomatic right ventricular (RV) enlargement leading to the gradual development of symptoms with age [16].

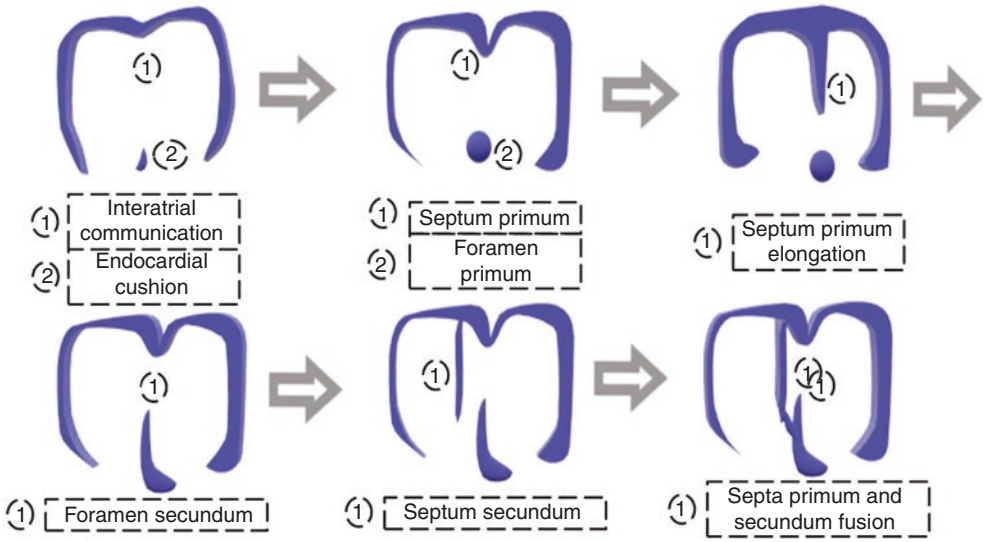

Fig. 2 Formation of atrial septum

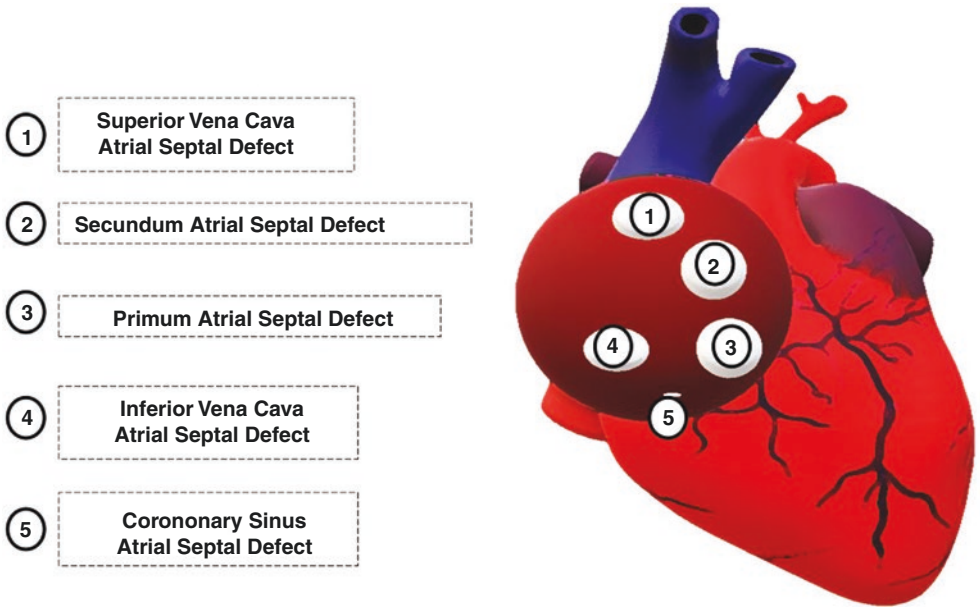

Fig. 3 Types of ASD

Spontaneous closure of ASDs has been described in approximately 40% of secundum ASDs. These small intra-atrial defects (usually less than 8 mm in diameter) limit the size of the shunt by their own resistance (pressure-separating effect) and do not cause major hemodynamic disturbances [17]. On the other hand, secundum ASDs of diameter greater than 8 mm, primum ASDs, sinus venosus defects, and coronary sinus defects do not seem to spontaneously close. On the contrary, they create size-dependent volume overload of the right heart and pulmonary arteries, which can lead later to the progressive development of pulmonary vascular obstructive disease and eventually pulmonary hypertension. This mainly occurs when the degree of shunting is significant. After years of pulmonary vascular injury, an irreversibly high pulmonary vascular resistance can gradually occur, creating a right-heart hypertrophy and consequently establishing a paradoxical right-to-left shunt (Eisenmenger syndrome).

The pressure difference between the left and right atrium can be significantly altered under several conditions. The presence of additional mitral stenosis entails a major increase in the pressure difference between the left and right atrium (Lutembacher's syndrome), which increases the left-to-right shunt with the same defect size. Furthermore, in adult patients, especially in those older than 50 years, age-induced increases in left-to-right shunting (due to relative increases in the left versus right ventricular end-diastolic pressures), systematic hypertension, and other acquired heart disease might lead to increased left ventricular (LV) end-diastolic and/or left atrial pressure, particularly in patients with mod-

erate and large ASDs [18]. These conditions might lead to ventricular diastolic dysfunction and left atrial enlargement, thereby providing fertile ground for mitral regurgitation and atrial fibrillation. These alterations in the heart's physiology increase the likelihood of developing symptoms and complications as described in the next section [17].

Diagnosis

Clinical Manifestations and Physical Findings

Most ASDs are of small size and therefore are asymptomatic during infancy and childhood. A murmur detected on physical examination or an incidental finding on an echocardiogram obtained for other reasons can lead to their evaluation and diagnosis. On the other hand, children with large ASDs occasionally suffer from heart failure (relative signs: tachypnea, rales, failure to thrive, hepatomegaly), recurrent respiratory infections, or failure to thrive. Symptoms and signs of pulmonary hypertension are uncommon in pediatric patients with ASDs but can occasionally occur.

In adult patients, ASD diagnosis should be established as soon as possible to increase the possibility of a timely repair. Even though most of these young patients have no or few symptoms on physical examination (right-sided volume overload is usually well tolerated for a long time), there is a number of findings that may give rise to clinical suspicion of an underlying ASD. Most patients with a significant shunt flow (i.e., pulmonary to systemic blood flow ratio [Qp:Qs] $\geq$1.5:1) will become symptomatic by the age of 40 years, except for some patients who have mild or nonspecific symptoms, and others who adapt to their limitations. Fatigue, exercise intolerance, and dyspnea are common clinical manifestations of ASDs in adults, while some patients develop heart failure. ASD repair reduces the severity of these symptoms. The classic physical findings of an isolated ASD in adults are typically similar to those observed in children [19–21].

Complications in patients with ASDs involve atrial arrhythmias, pulmonary hypertension (including Eisenmenger syndrome), cyanosis, and paradoxical embolism. Atrial arrhythmias (particularly atrial fibrillation, but also atrial flutter and other supraventricular tachycardias) are common in patients with ASDs after the third decade. The risk increases with age and pulmonary hypertension. Patients usually suffer from palpitations, dyspnea, and an increased risk of cardio-embolic events. The development of pulmonary vascular injury is related to the degree and duration of right heart volume overload. Pulmonary vascular damage acts as the pathogenetic substrate of pulmonary hypertension. As the disease progresses, right heart failure can be developed, and its features (elevated jugular venous pressure, hepatic congestion, tricuspid regurgitation, and pedal edema) might become obvious [16–19].

Furthermore, right heart failure might lead to elevated right heart pressures, and thus inversed (right-to-left) shunt, which is called Eisenmenger syndrome. Eisenmenger syndrome is accompanied by cyanosis and clubbing in addition to clinical signs of pulmonary hypertension. Last but not least, patients with an ASD are at risk for stroke, transient ischemic attack, or peripheral emboli due to paradoxical embolization (embolus carried from the venous side of circulation to the arterial side) [22, 23].

Electrocardiogram

An electrocardiogram (ECG) is a crucial part of the primary diagnostic evaluation of suspected heart conditions. Nevertheless, it is neither necessary nor sufficient to diagnose an ASD. Patients with small shunt or uncomplicated ASD can have a normal ECG. Sinus rhythm is observed in the majority of the patients, but the presence of atrial arrhythmias can complicate the clinical course of some patients and lead to relevant ECG evaluation and findings [13].

Right axis deviation and a "crochetage" sign (R wave notching in II, III, and aVF leads), which is correlated with the left-to-right shunt severity, can be found in secundum ASDs (Fig. 4). "Crochetage" sign involves at least one ECG lead in 73%, and all inferior leads in 27% of the ASD patients. Presence in all three inferior leads or co-occurrence with the right bundle branch block (RBBB) increases both the sensitivity and specificity of the sign [24]. Interestingly, "crochetage" sign disappears in approximately 50% of the patients within 2 weeks after ASD closure [14, 25].

Right ventricular hypertrophy and RBBB can also be observed. Left axis deviation and incomplete RBBB typically suggest the presence of a primum ASD.

Chest Radiograph

Chest radiograph findings depend on the degree of shunting. Normal chest radiograph is frequent—if not typical—in patients with small defects. Larger ASDs leading to significant left-to-right shunt often appear with right heart enlargement and prominent pulmonary vascularity (eventual signs of pulmonary arterial hypertension [PAH]). Dilation of the left atrium can also be marked, especially due to mitral regurgitation. The heart shadow often obtains a characteristic triangular appearance as the enlarged pulmonary arteries prevent the normal aortic arch from forming the heart border (Fig. 5) [19, 25, 27].

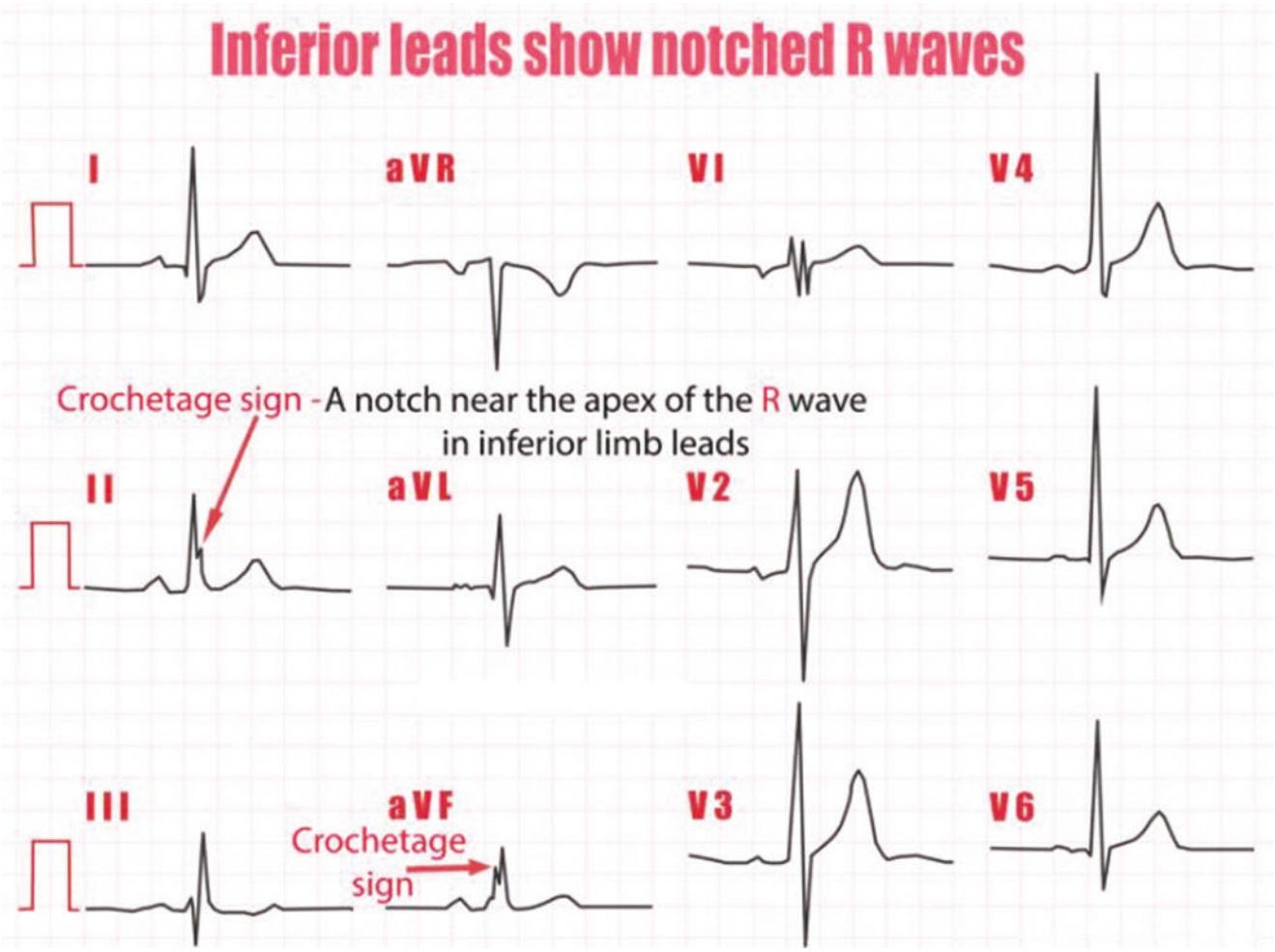

Fig. 4 Crochetage sign in ECG (*Jason Winter 2016-The ECG Educator Blog*)

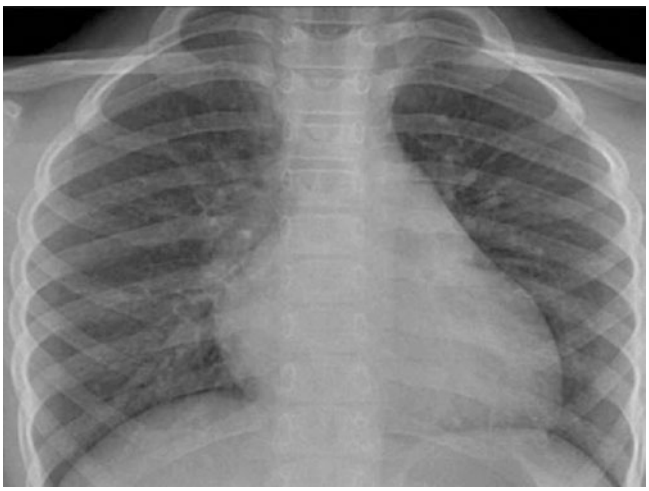

Fig. 5 Chest X-ray of a child with large ASD and hemodynamically significant left-to-right shunt (*Reprinted with permission from Arun Gopalakrishnan and Kerala Journal of Cardiology* [26])

Imaging-Based Diagnosis

Imaging is necessary to confirm the diagnosis of ASD. Echocardiography is the imaging modality of choice, utilized ante- or/and postnatally to establish the specific type, location, and size of ASD, and to identify associated abnormalities or complications.

In particular, the antenatal detection of an ASD depends on both disease type and operator. When performed by an experienced fetal echocardiographer, the third-level obstetric ultrasound can establish the diagnosis of a primum ASD at 18–22 weeks of gestation [28]. However, the most frequent ASD type, secundum ASD, cannot be reliably detected prenatally due to the sizable PFO observed in healthy fetuses. This makes the distinction between a small to moderate-size secundum ASD from a PFO with right-to-left shunt almost impossible. Yet, fetal echocardiography may still raise suspicion for a very large secundum ASD, which should be con-

firmed postnatally, as required for any other suspected prenatal ASD case. With regard to the sinus venosus and coronary sinus ASDs, although a trained fetal echocardiographer may have the ability to detect them prenatally, the sensitivity and specificity of this diagnostic method have not yet been reported for these more unusual types of ASD.

Ultimately, antenatal ASD imaging findings need to be confirmed postnatally through one of the following imaging-based diagnostic approaches:

Transthoracic Echocardiography

In general, the transthoracic echocardiography (TTE) with Doppler is the initial imaging test of choice for the diagnosis and evaluation of primum and secundum ASDs [29], with the diagnoses of the latter being usually definitive (Fig. 6) [30]. A comprehensive TTE includes 2D imaging as well as color-flow, pulsed-wave, and continuous-wave Doppler with multiple echocardiographic views obtained to assess the precise location and size of the ASD, as well as the hemodynamic alterations associated with the shunt [29].

A primum or secundum ASD is suspected in the presence of abrupt discontinuity, hypermobility, or drop out of the interatrial septum. On M-mode, RV dilatation and abnormal septal motion serve as signs of ASD in cases with significant shunting [31]. The subcostal four-chamber view improves the visualization of the atrial septum because the ultrasound beam is generally nearly perpendicular to the atrial septum [32]. Unfortunately, this view is suboptimal in some patients, especially in overweight and obese individuals. Parasternal short-axis and off-axis apical view, as well as other nonstandard views are commonly required to visualize the whole interatrial septum. A recent study aiming to assess the diagnostic sensitivity of the 2D echocardiographic subcostal imaging on patients with a history of ASD (mean age 31 years) demonstrated that the defect could be successfully visualized in 93 out of 105 secundum ASDs, all 32 primum ASDs, and only 7 out of 16 sinus venosus ASDs.

Moreover, it is reported that the operator can usually demonstrate through the pulsewave Doppler a classic triphasic left-to-right shunt at the level of the defect, with characteristic flow waveforms in late systole and diastole and a presystolic accentuation. An experienced echocardiographer is also able to distinguish a physiological venous flow pattern from an ASD flow pattern. In addition, color flow is used to visualize the actual shunt, with a right-to-left or bidirectional flow being developed in patients with RV diastolic dysfunction or significant tricuspid regurgitation. According to Pollick et al., the color jet width may be linked with the left-to-right shunt size [33]. It has been demonstrated that a color flow width of 1.5 cm may suggest

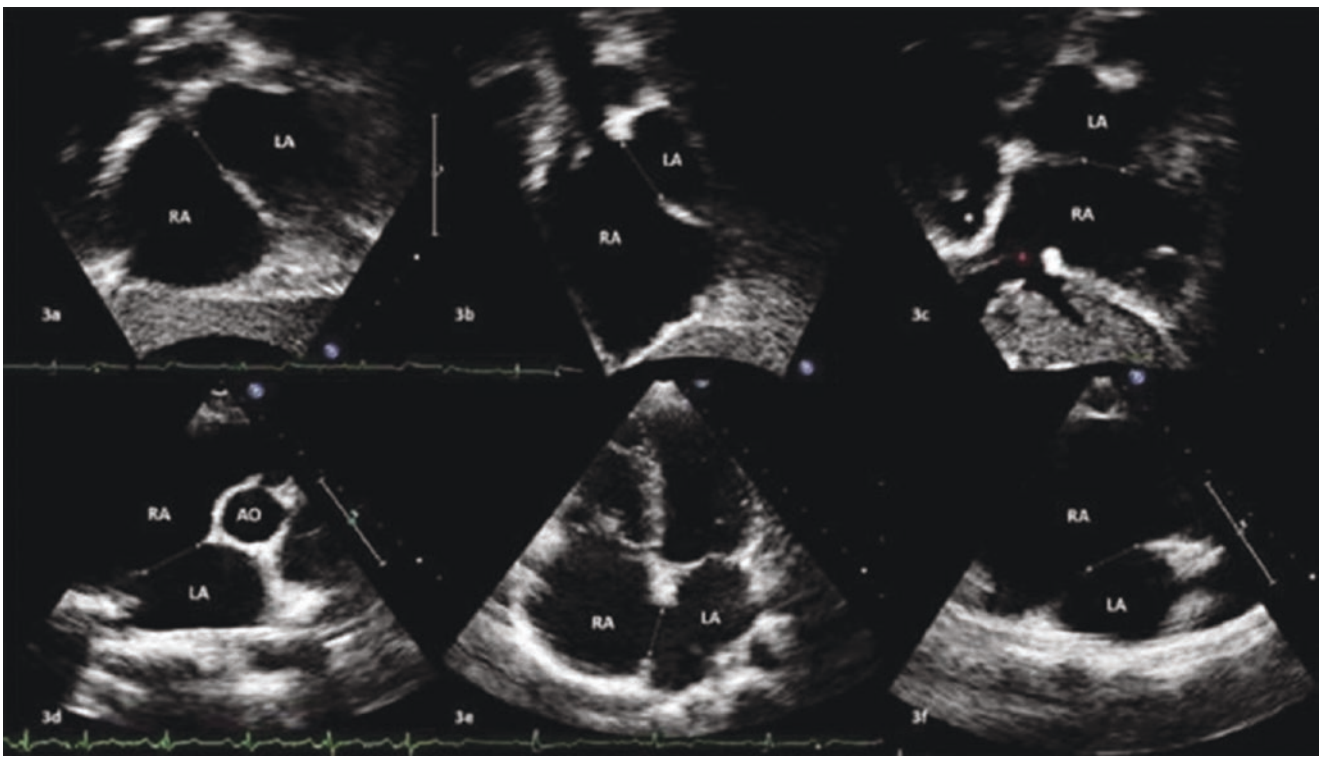

Fig. 6 Transthoracic echocardiographic images of ASD. Panels **3a**–**3c** are 2D transthoracic echocardiographic images from the subxiphoid window. Panel **3d** is the parasternal short-axis projection showing the aortic rim of the ASD. The mitral rim of the ASD is notable in the apical four-chamber view in panel **3e**. Panel **3f** is the right parasternal window showing the ostium secundum ASD (*Reprinted with permission from Arun Gopalakrishnan and Kerala Journal of Cardiology* [26])

a Qp:Qs ratio greater than 2:1, which has a major prognostic potential as discussed below [33].

Nevertheless, when a defect is not apparent, contrast echocardiography with the administration of agitated saline contrast through a peripheral vein is the primary test used to determine the presence of an ASD, especially for patients with restricted acoustic windows [34]. In the case of an ASD, it shows the extravasation of saline contrast microbubbles from the RA to the LA at the level of the interatrial septum [35]. Furthermore, sinus venosus and coronary sinus ASDs can be diagnosed by the appearance of agitated saline contrast in the left atrium before it appears in the right atrium soon after the injection of contrast saline through the left arm.

Transesophageal Echocardiography

Although 2D TTE (either with Doppler or with peripheral vein injection of agitated saline) is usually adequate to identify the presence of an ASD and assess the cardiac chamber size, one should not discount that the diagnostic sensitivity of the TTE may be subject to the view obtained and operator/patient factors (i.e., optimal acoustic windows in young patients, but restricted acoustic windows in other patients due to large body habitus, and prior thoracic surgery). Hence, negative suboptimal or noncomprehensive TTEs do not exclude an ASD diagnosis. At the same time, transesophageal echocardiography (TEE) offers an excellent alternative approach in patients with poor imaging quality and is suggested if TTE fails to show an ASD in a patient with suspected ASD (Fig. 7).

TEE is highly accurate for the diagnosis of all types of ASDs in the hands of a trained echocardiographer. TEE further aids in the sizing of secundum ASDs and in detecting the most common forms of anomalous venous connections (PAPVC) [36].

When performed with color flow Doppler or with agitated saline contrast injection, TEE can detect right-to-left and left-to-right shunting with higher sensitivity than via TTE [37]. Furthermore, TEE can also detect flow through multiple ASDs. Finally, TEE imaging prior to planned surgical or percutaneous closure can determine the suitability for transcatheter device closure and prevent unnecessary therapeutic

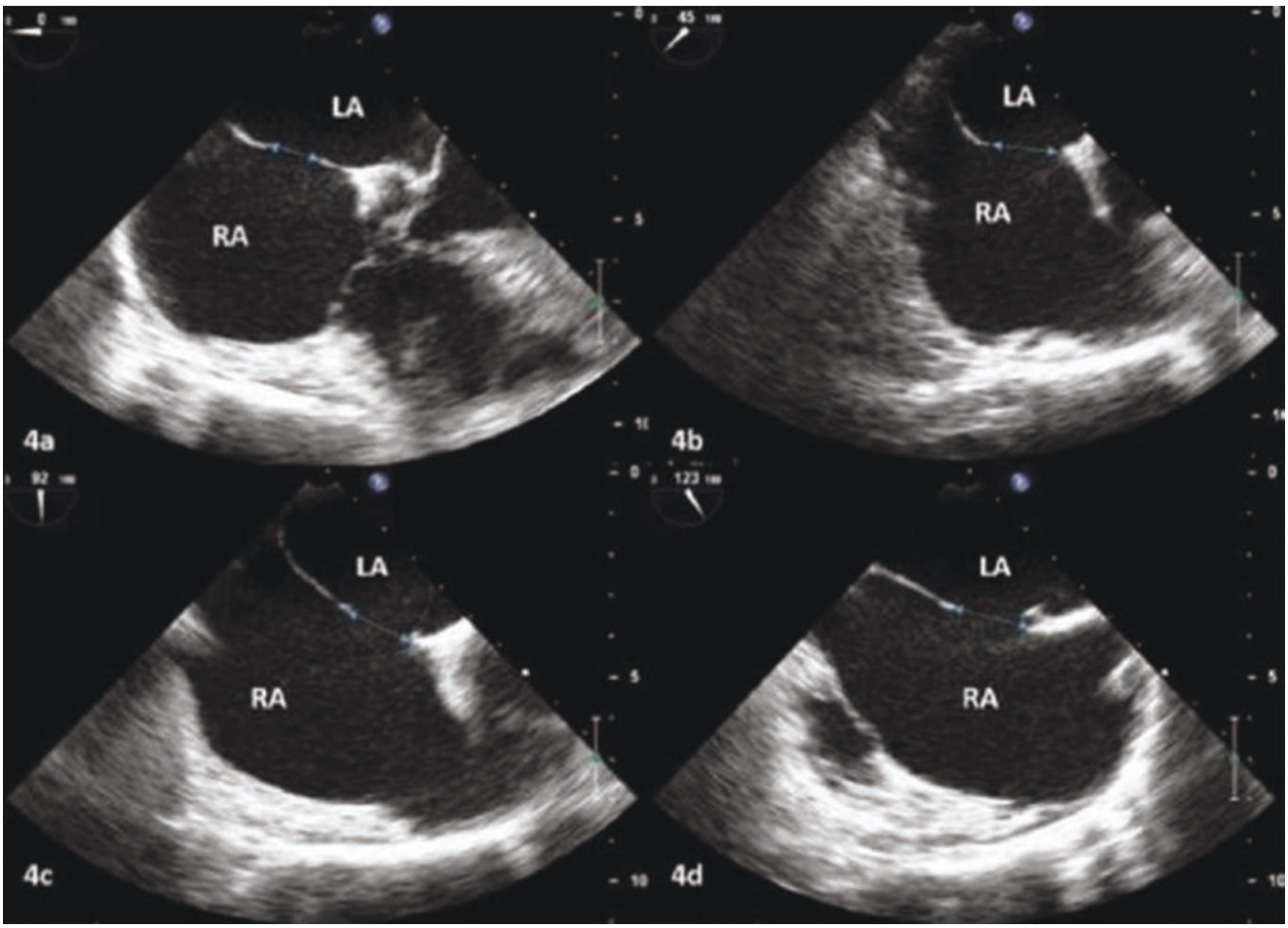

Fig. 7 Transesophageal echocardiographic images of ASD (*Reprinted with permission from Arun Gopalakrishnan and Kerala Journal of Cardiology* [26])

procedures. 3D TEE provides even better sizing and visualization of ASDs and provides important evidence with regard to their shape and their relationship to atrial superior and inferior limbic band tissue, the aortic root, and the AV valves [38–40].

Cardiovascular Magnetic Resonance, Computed Tomography and Cardiac Catheterization

In the presence of nondefinitive TTE and TEE findings with regard to the diagnosis of sinus venosus or unroofed coronary sinus ASDs, the assessment of PAPVC, or the presence of RV volume overload, cardiovascular magnetic resonance (CMR) or computed tomography (CT) imaging can be considered. The CT and CMR procedures require special imaging protocols and should be performed by an experienced cardiac imaging specialist, able to identify, characterize ASDs and abnormal pulmonary venous return, and also estimate shunt flow [41–43].

Additionally, recent advances in CMR and high-resolution contrast CT techniques facilitate the anatomical delineation of ASDs and the quantitative assessment of their hemodynamic complications (ventricular volumes and volumes of pulmonary and systemic flow) [44–46]. The quantification of left-to-right shunt fraction (Qp:Qs ratio) should be performed to determine whether an ASD closure is indicated or not (threshold of 1.5). This is nowadays plausible not only by means of invasive right heart catheterization (invasive oximetry), but also by means of CMR. CMR-based shunt flow calculations do not differ significantly from those of the cardiac catheterization, despite the small overestimation of the Qp:Qs ratio by CMR. Therefore, given that ASDs can be diagnosed and treated with noninvasive imaging assessment (i.e., CMR, CT, TEE, TTE+ Doppler/+saline contrast, 3D echo), cardiac catheterization in the course of the ASD clinical workup is rarely performed nowadays.

Management

Age of the patient, location and size of the defect, presence of pulmonary hypertension, and hemodynamic impact of the shunt are the main factors determining ASD management strategy. In childhood, the vast majority of small isolated secundum ASDs can spontaneously close within the first 2 years of life and in some cases as late as by 5 years of age. Hence, early closure is not indicated for these defects unless there are significant symptoms. Furthermore, there is also no high-quality evidence supporting the closure of a persistent ASD in asymptomatic children older than 2 years old with persistent small defects [47].

Moderate and large-sized secundum ASDs, and types of ASDs other than secundum, are unlikely to close spontaneously. Nevertheless, given the—small—likelihood of spontaneous closure during the first 2 years of life, intervention to close these defects tends to be postponed until after the age of 2 years in asymptomatic children. For patients with a persistent moderate or large defect, left-to-right shunting usually deteriorates with age if the defect is not repaired. Hence, the principal indication for ASD closure in children is the presence of a sizable left-to-right shunt leading to a clinically significant right heart overload. The risk of paradoxical embolism through a small ASD is yet to be quantified and the benefit of ASD closure in reducing the cardio-embolic risk has to be compared with the short- and long-term procedural risks. The preferable type of intervention (transcatheter procedure or open surgical repair) depends on the ASD type and the extent of subsequent cardiac dysfunction. Percutaneous closure is generally indicated for secundum ASDs that are not excessively large. In symptomatic infants with primum or sinus venosus or coronary sinus ASDs, surgical repair remains the gold standard [19–21, 30, 47, 48].

In terms of recommendations for intervention in adults, ASD closure in patients with RV overload and no PAH or LV disease is the most well-documented indication and is strongly recommended regardless of symptoms. In patients with PAH, closure should only be considered in the presence of significant left-to-right shunt (Qp:Qs >1.5) and pulmonary vascular resistance (PVR) ≥5 WU (persisting after targeted PAH treatment). ASD closure should not be considered as a therapeutic solution in patients with Eisenmenger physiology, and patients with PAH and concomitant PVR ≥5 WU, despite targeted PAH treatment, or desaturation on exercise. In patients with (systolic or/and diastolic) LV dysfunction, ASD closure appears to exert a deteriorating effect on heart failure and thus should be carefully evaluated. Balloon pre-interventional testing might be required to carefully weigh the benefit of eliminating left-to-right shunt against the potential negative impact of ASD closure on filling pressures of the LV [49]. In addition, patients with suspected paradoxical embolism should be referred to ASD closure, provided that the presence of PAH or LV dysfunction have been dully excluded [25, 50–53]. Finally, ASD closure is also recommended for patients with documented platypnea-orthodeoxia [48].

ASD closure can be performed either by catheter intervention or by open heart surgery. Percutaneous procedures have reported similar success and mortality rates with lower morbidity (decreased exercise capacity, shortness of breath, right heart failure) and, therefore, transcatheter device closure has become the first choice for secundum defects with a diameter of less than 40 mm, which is the case in 80% of the patients [52, 54–58]. Serious complications have been observed in ≤1% of patients [59].

Post-intervention atrial tachyarrhythmias appear to be mostly transient. Other complications include erosion of the atrial wall, anterior mitral leaflet, or the aorta, as well as thromboembolic events, but these are rarely encountered in

clinical practice [15, 27]. When this minimally-invasive procedure is not feasible or not suitable for ASD patients, particularly elderly ones, and those with primum, sinus venosus, or coronary sinus ASDs, open heart surgery is required. Nonetheless, mortality rates of surgical repair in isolated ASDs have been reported to be low (<1% in patients without significant concomitant diseases) in experienced centers, with favorable long-term outcomes when performed early (i.e., in childhood, adolescence) and in the absence of PAH [54, 55]. In general, outcomes of an ASD closure seem to be better when the repair is performed until the middle of the third decade of life [54, 55].

Furthermore, patients with either unrepaired or repaired ASDs require further general evaluation and management. First of all, ASDs are a major predisposing factor for atrial arrhythmias, which should be managed according to standard recommendations. Cryo- or radiofrequency ablation (modified maze procedure) can be considered at the time of intervention, as device closure might later restrict electrophysiologists from accessing the left atrium. Additionally, ASDs are frequently associated with pulmonary hypertension, associated with a higher risk of complications (heart failure, cyanosis, pulmonary artery thrombosis). Advanced therapy for PAH hypertension is suitable for patients with pulmonary hypertension secondary to ASD and might improve symptoms or even render the defect operable. Moreover, patients with an ASD are at a substantial risk of infective endocarditis. Thus, appropriate guideline-driven antibiotic prophylaxis should be given when patients undergo procedures with a risk of bacteremia during the first 6 months after ASD closure. On the contrary, no treatment is warranted in isolated ASDs with no residual shunt because they have been correlated with negligible risk of infective endocarditis. All patients and their caregivers should be educated regarding the need to carefully observe oral hygiene and adhere to their medication. Finally, in terms of sports and physical activity, doctors need to provide patients with personalized recommendations based on the defect's size and concomitant symptoms.

Conclusion

ASDs constitute a characteristic type of CHD that remains "silent" and becomes obvious mostly in adulthood as most of them are small in size and therefore asymptomatic during infancy and childhood. Their clinical symptomatology, as well as diagnostic approach, remains the same in children and adults. The age of the patient, the location and the size of the defect, and the presence of pulmonary hypertension along with the hemodynamic impact of created shunts constitute the most significant parts of ASDs' clinical evaluation. Finally, their management depends on the consideration of the aforementioned parameters and demands a multilevel clinical approach by pediatric cardiologists, adult CHD cardiologists, interventional cardiologists, and cardiac surgeons.

Multiple Choice Questions

1. Which of the following is correct:
   A. The prevalence of atrial septal defects (ASDs) is estimated to be approximately 6 per 1000 live births and 0.88 per 1000 adults while ASDs account for 7–10% of all congenital heart defects.
   B. The prevalence of atrial septal defects (ASDs) is estimated to be approximately 1.6 per 1000 live births and 0.88 per 1000 adults while ASDs account for 27% of all congenital heart defects.
   C. The prevalence of atrial septal defects (ASDs) is estimated to be approximately 1.6 per 1000 live births and 0.88 per 1000 adults while ASDs account for 7–10% of all congenital heart defects.
   D. The prevalence of atrial septal defects is estimated to be approximately 0.16 per 1000 live births and 0.88 per 1000 adults while ASDs account for 27% of all congenital heart defects.
   E. The prevalence of atrial septal defects is estimated to be approximately 1.6 per 1000 live births and 0.88 per 1000 adults while ASDs account for 0.7% of all congenital heart defects.
2. Atrial septal defects are associated with:
   A. Down syndrome
   B. Noonan syndrome
   C. Chondroectodermal dysplasia
   D. Congenital rubella
   E. All the above
3. The most frequent type of atrial septal defect is:
   A. Ostium primum
   B. Ostium secundum
   C. Superior sinus venosus
   D. Inferior sinus venosus
   E. Coronary sinus defect
4. Which of the following is correct:
   A. Atrial septal defects can give birth to a left-to-right shunt, which generates volume overload of the right heart chambers.
   B. Atrial septal defects can give birth to a left-to-right shunt, which generates volume overload of the left heart chambers.
   C. Atrial septal defects can give birth to a left-to-right shunt, which generates pressure overload of the right heart chambers.
   D. Atrial septal defects can give birth to a right-to-left shunt, which generates volume overload of the right heart chambers.

E. Atrial septal defects can give birth to a right-to-left shunt, which generates volume overload of the left heart chambers.

5. Spontaneous closure of ASDs has been described in approximately:
   A. 10% of secundum ASDs
   B. 25% of secundum ASDs
   C. 40% of secundum ASDs
   D. 70% of secundum ASDs
   E. 90% of secundum ASDs
6. Atrial septal defects are not directly associated with:
   A. Arrhythmias
   B. Stroke
   C. Pulmonary hypertension
   D. Mitral stenosis
   E. Dyspnea
7. The "crochetage" sign can be found in patients with atrial septal defect and is characterized by:
   A. R wave notching in V1–V6 leads
   B. R wave notching in II, III, and aVF leads
   C. R wave notching in I, II, and III leads
   D. Q wave notching in V1–V6 leads
   E. Q wave notching in II, III, and aVF leads
8. The gold standard imaging test for the diagnosis of atrial septal defect is:
   A. Cardiovascular magnetic resonance
   B. Transesophageal echocardiograph
   C. Computed tomography
   D. Transthoracic echocardiograph
   E. Cardiac catheterization
9. Which is not an indication for performing an atrial septal defect (ASD) repair:
   A. Large asymptomatic ASDs
   B. Moderate-sized symptomatic ASDs
   C. Pulmonary hypertension with a right-to-left shunt
   D. Pulmonary hypertension with a left-to-right shunt
   E. Paradoxical embolism
10. Which of the following is correct:
   A. Percutaneous procedures have reported higher success rates than surgical repair, with lower morbidity. Serious complications in transcatheter interventions have been observed in ≤1% of patients.
   B. Percutaneous procedures have reported lower success rates than surgical repair, with higher morbidity. Serious complications in transcatheter interventions have been observed in approximately 5% of patients.
   C. Percutaneous procedures have reported similar success and mortality rates compared to surgical repair, with lower morbidity. Serious complications in transcatheter interventions have been observed in ≤0.1% of patients.
   D. Percutaneous procedures have reported lower success rates than surgical repair, with higher morbidity. Serious complications in transcatheter interventions have been observed in ≤1% of patients.
   E. Percutaneous procedures have reported similar success and mortality rates compared to surgical repair, with lower morbidity. Serious complications in transcatheter interventions have been observed in ≤1% of patients.

Answers

1. C
2. E
3. B
4. A
5. C
6. D
7. B
8. D
9. E
10. E

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