

## Complex atrial septal defect closure in children

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### Abstract

Atrial septal defect (ASD) is one of the most common congenital heart diseases (CHDs) diagnosed in children. Symptoms in ASD may be absent, but palpitations and dyspnea in children sometimes reveal a complex structural and CHD. Diagnosis is established usually by transthoracic echocardiography, but in more complex cases such as coronary sinus defect, enhanced computed tomography or cardiac magnetic resonance may be used. Indication for closure depends on the dimension and on the hemodynamical impact. There are two methods for closure: surgical and interventional. Surgery may be applied to all types of ASD, while transcatheter closure may be used only for *ostium secundum* ASD. The most important issue in the interventional closure is the anatomical aspect related to the position of the defect. Both methods may have complications. Complex cases in children usually are not recommended for interventional closure, surgery being the recommended method. Oversizing of the device to be implanted according to the sizing balloon and not to the initial defect diameter may give a solution for large ASDs. Interventional closure of large defects in small children with an aneurysmal, multi-fenestrated, malaligned septum, or with other CHD associated may be challenging. Complications are present for both types of closure, but they are relatively different.

**Keywords:** atrial septal defect, multi-fenestrated, child, interventional closure, sizing balloon.

### Introduction

Atrial septal defects (ASDs) are among the most frequent congenital heart diseases (CHDs) in children, representing 10–15% of all CHDs and in a higher percentage (20–40%) in adults. Females have a higher incidence of ASD (2:1) compared with males. There are four types of ASD (*ostium primum*, *ostium secundum*, venous sinus, coronary sinus). Any of them can be considered a complex ASD by its position, size, number of fenestrations, associated hemodynamic consequences and complications, associated lesions, technical difficulties regarding the treatment and the patient's age. The outcome is related to the complexity of the cases. The direct consequences of a large ASD are the increased pulmonary flow Qp to systemic flow Qs ratio of more than 1.5:1, right heart enlargement, atrial arrhythmias, pulmonary hypertension [1–4].

Presence of a genetic disease also increases the complexity of the case, which appears in a complex context with multisystemic manifestations. There may be present in the form of autosomal dominant (AD) or autosomal recessive (AR) diseases. ASD may be associated with Holt–Oram syndrome (AD transmission, associated with deformations of the radius – absent or hypoplastic), with Ellis–van Creveld syndrome (AR transmission, associated with polydactyly, deformations of the skeleton – short limbs and short ribs) but also in other more frequent genetic syndromes, such as Down syndrome [5]. Some of the mutations of the genes responsible for manifestation in these syndromes were identified: T-box 5 (*TBX5*),

NK2 homeobox 5 (*NKX2.5*), GATA binding protein 4 (*GATA4*) [6–10].

*Ostium secundum* ASD is the most frequent type, representing 75% of all. It is the single type which has an accepted indication for interventional closure if produces a significant hemodynamic effect. 85–90% of these defects may be closed interventional. Complexity for this type is mainly related with the anatomical position, shape, dimension of the defect, dimension of the rims, consistency of the rims, an aneurysmal aspect of the atrial septum, or multiple fenestrations.

*Ostium primum*, which represents 10–15% of all ASDs is identified as a part of the partial atrioventricular septal defect. Usually is associated with a cleft of the mitral valve, which makes the closure intervention a difficult one compared with simple surgical *ostium secundum* ASD closure.

*Sinus venosus* ASD may be superior or inferior, at the connection of the superior vena cava (SVC) and inferior vena cava (IVC) with the right atrium (RA) and represents 5–10% of all ASDs. Interventional closure in superior *sinus venosus* ASD was recently described [11]. Partial anomalous pulmonary venous drainage towards the RA, SVC or IVC is frequently associated with *sinus venosus* ASD.

Coronary sinus ASD is a very rare type of ASD. It is not a typical ASD. It consists of the unroofed coronary sinus, partially or totally, with or without persistent left superior vena cava draining into the left atrium. The innominate vein may be present (normal or hypoplastic) or absent.

The existing guidelines are not specific to different types of ASD [12]. It is known that only the secondary types of ASD can be closed interventionaly [13, 14], but also new and difficult procedures are described for closure of superior venous sinus defect or coronary sinus defect [11, 15]. Surgery was the single method to close all types of ASD since 1953, when it was performed by Gibbon up to 1976. Then, the first interventional closure was performed by King & Miller in a 17-year-old girl [16].

In patients that experience hemodynamic consequences, such as diastolic dysfunction of the right or left ventricle or pulmonary hypertension that may decrease the outcome, partial solutions are imagined using a fenestrated device for the ASD closure [17].

The most frequent ASD associated pathologies are the patent *ductus arteriosus* (26.1%), pulmonary hypertension (3.8%) and supraventricular tachycardia (2.4%) [2]. In the same time, *ostium secundum* ASD is one of the most prevalent isolated CHD, together with a ventricular septal defect, patent *ductus arteriosus*, and pulmonary stenosis [18]. In other associated lesions, the presence of an ASD may save the patient's life as in hypoplastic left heart syndrome, D-transposition of great arteries, tricuspid atresia and total anomalous pulmonary venous return [19].

### ☞ Specific aspects in the diagnosis

In most cases, the ASDs are asymptomatic and are accidentally discovered. Sometimes, in moderate or severe forms, they may manifest as delay in somatic growth, recurrent respiratory infections, fatigability, and dyspnea. Rarely, heart failure may be present in relation to a large ASD. When the atrium dilation is important, atrial arrhythmias – atrial extrasystoles, atrial tachycardia, atrial flutter, atrial fibrillation – may appear, in up to 10% of the patients older than 40 years but also in children [19, 20].

At clinical evaluation, a fixed split first sound and a heart murmur may be heard, but usually is related to hyper-flow relative pulmonary valve stenosis.

*Ostium primum*, which is associated with a cleft of the mitral valve, may present, at auscultation, an apical systolic murmur. ASD venous sinus does not have a particular aspect, but coronary sinus defect may be associated with systemic desaturation and risk for cerebral embolization. This risk is present also in patent *foramen ovale* or *ostium secundum* ASD.

Usually, the chest X-rays show hypervascularity of the lung fields, cardiomegaly, and increased projection of the pulmonary artery.

Electrocardiographically, a right ax deviation, and minor right bundle branch block are present. The rhythm may be sinus rhythm or may show different arrhythmias. Atrial tachyarrhythmias and conduction disorders may appear. The incidence of atrial arrhythmia decreases after ASD closure. Conduction disorders are very rare, usually associated with a large shunt, but also after repair of the *ostium primum* ASD or in familial types with genetic mutations. Also, venous sinus defect may generate sinus node dysfunction [21].

Echocardiographically, by two-dimensional evaluation, it is mandatory to evaluate the type of the defect, the dimension, the hemodynamic consequences, such as right heart enlargement (right atrium, right ventricle,

pulmonary artery), associated lesions and the anatomical description of the rims and vicinity structures. In difficult cases, such as coronary sinus defect, if the images are not diagnostic according to the right heart enlargement, complementary evaluations may be necessary. Contrast echocardiography administered by the left arm, three-dimensional (3D) echocardiography, and transesophageal echocardiography (TEE) are the following choices. The defect should be evaluated from multiple views, but the subcostal is the most important.

In complex cases, some measurements and calculations may have predictive importance. For instance, a ratio of the defect/total septum  $\leq 0.35$ , or a ratio of the supero-anterior rim/defect size  $> 0.75$ , or a ratio infero-posterior rim/defect size  $> 1$  may have good predictive value [22].

Also, indications or contraindications are initially evaluated by transthoracic echocardiography [23]. After establishing the type of ASD, the surgical, interventional, or no intervention strategy is established. Indications for transcatheter closure in *ostium secundum* ASD are related with a significant hemodynamically ASD, manifested as dilation of the right ventricle with left-to-right shunt and absent/mild pulmonary hypertension with suitable anatomical characteristics (rims  $> 5$  mm, a good total length of the atrial septum). Also, the presence of paradoxical embolization is a clear indication for interventional closure. Interventional closure is totally contraindicated in the presence of sepsis, nickel allergy (except the Cocoon devices) [24], Eisenmenger syndrome, or contraindication for Aspirin or Clopidogrel. There is a special group of patients in which interventional closure can be considered. The decision of closure can be taken after a test of occlusion of the defect, depending on the pressures and systemic saturation. Those are patients with vasoactive pulmonary hypertension or left ventricle diastolic dysfunction. For these patients, fenestrated ASD devices can be implanted [23].

TEE is extremely important for guiding the transcatheter device implantation in *ostium secundum* ASD, especially in small children or in complex cases with simultaneously multiple devices implantation [25]. Similar importance with TEE or higher, it has the real-time 3D Doppler TEE, which may allow visualization of the anatomical shape of the defect (a round, racquet-shape, oval, or a star) that can be important in device selection [26–28] or the spatial relationship between multiple defects followed by multiple implantations [25].

Intracardiac echocardiography may be a very good choice, during the transcatheter device implantation procedure, with a 100% rate of success, and a 2% rate of major complications. Unfortunately, it is an expensive procedure, which necessitates second vascular access of at least 8F [29, 30].

Cardiac catheterization should be performed in inconclusive case, in which pulmonary hypertension is suspected or diastolic dysfunction of the right or left ventricle.

### ☞ Interventional and surgical treatment in complex ASD

Small ASDs (less than 8 mm) have a higher rate of spontaneous closure in the first years of life [31–34],

and very rare in adolescence [35] for the *ostium secundum* type. For this reason, the indication for closure comes after the age of 3–4 years. Large ASD associated with signs of heart failure may need pharmacological treatment with diuretics and surgery even in infancy [20].

Treatment in ASD is different in relation to the four anatomical types described previously. *Ostium secundum* ASD and rare cases of ASD may be attempted interventionally, while *ostium primum* ASD and the most part of the venous sinus and coronary sinus are surgical.

Although the gold standard for the closure of the defect remains the surgery, interventional closure is the first method to attempt, because the most part of the ASD, 85–90%, are resolved without surgery [19].

There are more than 40 years since ASD interventional closure was performed by King & Miller for the first time in humans in 1976 [16]. ASD interventional closure is possible both in children and in adults with *ostium secundum* type of ASD, and it is related to the anatomical aspect of the defect [12, 36–38]. In the guidelines for pediatric interventional closure of the ASD [12] of the *American Heart Association*, interventional closure of the ASD is recommended to be performed in children which are weighing >15 kg, although the procedure can be performed even in children <2 years old

with a higher risk of major complications [3, 37]. The risks for major complications in children less than 15 kg increase almost five times (5.5% incidence of major complications) [3].

The anatomical characteristics of the septum, which make the procedures complex, refer to the rims (more than one deficient rim), to the total atrial septum length (small), aneurysmal septal tissue, multiple fenestrations, large Eustachian valve. Of a great importance, besides the anatomical characteristics, are the small age and weight of the child, the presence of significant comorbidities, pulmonary hypertension, diastolic dysfunction of the left ventricle (usually in elderly) or right ventricle (usually in patients with pulmonary atresia with intact ventricular septum at several years distance) [13, 19, 30]. Complex cases in children usually are not recommended for interventional closure being associated with a higher risk for complications [37]. Large, multiple, multi-fenestrated, aneurysmal, malaligned atrial septum, ASD with deficiency/floppy posterior or inferior rims with or without the complete absence of aortic rim, and associated lesions, such as valvular stenosis, coronary anomalies, ventricular septal defects, partial anomalous venous return, are among the most complex ASD closure procedure (Table 1) [39–43].

**Table 1 – Conditions related to transcatheter atrial septal defect (ASD) closure**

Ideal ASD	Complex ASD	Excluded ASD
• Centered	• Rim deficiency (aortic, posterior)	• Rim deficiency (inferior vena cava)
• Normal anatomy	• Malaligned septum • Multifenestrated • Aneurysmal septum • Prominent Eustachio valve	
• Small-moderate defect/big child	• Big defect (>20 mm) in a small child (<15 kg)	• Too large (>35 mm)
• <i>Ostium secundum</i>		• <i>Ostium primum</i> , venous sinus, coronary sinus
• No other congenital heart disease associated	• Severe pulmonary stenosis	• Associated anomalies: partial anomalous pulmonary venous return, divided left or right atria
• Absent pulmonary hypertension	• Reversible pulmonary hypertension	• Pulmonary hypertension with high pulmonary vascular resistance

If the interatrial shunt is bidirectional with systemic desaturation at rest >85%, a more complex closure procedure has to be performed. Initially, heart catheterization followed by balloon occlusion test. If the right atrial pressure is not increasing >20%, left atrial pressure is not decreasing >20% and the saturation increases to >94%, the patient has an indication for closure [19].

**Multiple or multi-fenestrated ASDs** represent 10–15% of all ASDs [44]. Often, they associate aneurysmal atrial septum, which is defined by more than 10 mm excursion. In children, age and weight are very important when it comes to the interventional closure of an ASD. In children, large ASD was defined as a ratio of the ASD diameter/body surface area (BSA) >15 mm/m<sup>2</sup> [45] or by a ratio ASD diameter/weight >1.2 [37]. Deficiencies of the anterior, posterior or superior rims are considered also risk factors for unfavorable outcome [37, 39, 45]. Aortic erosion, cardiac perforation, atrioventricular block or device-related aortic regurgitation may happen with oversizing of the device [36, 37, 39, 44, 45]. There are authors who recommend oversizing by 2 mm, for one deficient rim and by 4 mm for two deficient rims, usually

less than 20–30% of the measured diameter either by TEE or by balloon sizing [39, 46].

Multi-fenestrated ASDs are not straight-ahead cases for interventional cases because most of them associate aneurysmal septum, floppy, deficient rims, heart dilatation or multiple devices implantation may be necessary [25].

The **total length of the septum** (TLS) is very important in children with large ASD and septal aneurysm and it is preferred for evaluation instead of using balloon sizing for associated risks (hemodynamic compromise due to obstruction of the flow from the IVC and inflow of the ventricles with low cardiac output and hypotension) [36]. The TLS is measured in four-chamber view by TTE, preferably and/or before initiation of the procedure. TLS should be superior to the left atrial diameter of the device intended to be used [36].

If a **single device** can be used, or if **multiple devices** are needed, it is an extremely important and widely discussed issue in the medical literature. Each of the possibilities has advantages and disadvantages. To use a single device, it advocates a shorter procedure time with exposure to lower time X-ray radiation, lower embolization

and erosion risks, a financial advantage, and the procedure has a lower degree of technical difficulty [38]. When using multiple devices, the procedure is more technically complicated and requires more experience, it is more expensive, and the risks of embolization or erosion are greater [47]. In a study of 33 patients using more than one device, Awad *et al.* reported a 6% complication rate (one embolization case and one case of erosion), with 97% immediate success rate. A distance higher than 7 mm was considered favorable for implantation of two devices [47]. In the case of multiple device implantation, firstly the larger defect has to be closed [47], but this may be different depending on the school. In another study of 34 children with multiple ASDs, 19 (55.9%) children received single-device ASD closure, 15 children received more than one device (two devices per 14 and three devices in one patient). It is noted that the one who received three devices (31 years old) died at home 30 days after the procedure with cardiac tamponade but without signs of erosion [48]. Seventy-five percent of cardiac erosions occur within the first five days after the procedure [47].

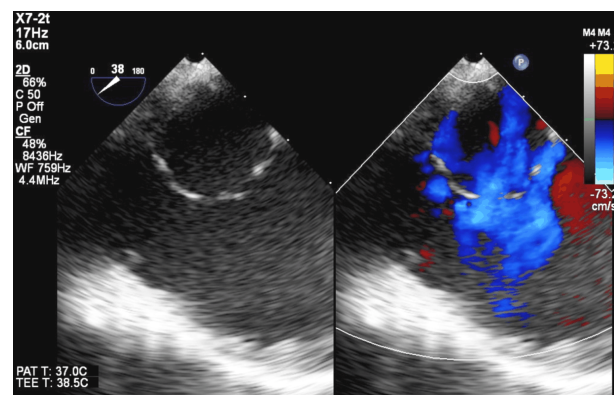
In a study published by Butera *et al.*, in case of multi-fenestrated defects associated with aneurysm (as in Figure 1) (36 patients), in 2/3 (69%) of the patients one device was used [Amplatzer Cribriform Occluder (ACO) or Amplatzer Septal Occluder (ASO)] and in 1/3 of the patients two devices were used either ACO or a combination of an ASO with an ACO. A trivial residual shunt was present in 12% of the cases [44]. In the same study, there were three patients with three openings and three devices implanted. Almost the same percentage (63.5%) for single device use was obtained in another study performed by Masseli *et al.* on 148 patients with multiple ASDs [49]. A higher percentage for single device uses for complex ASD closure was reported by Santoro *et al.* (95.8%), in a study on 83 patients with complex ASD anatomy. Initial ASD closure rate was in 59.7% of the patients, reaching 95.9% at the last follow-up [50]. There are authors who consider that ACO is not the best option for closure aneurysmal multi-fenestrated ASD due to the fact that the waist is narrowed (connecting pin) and does not offer any support for a large device in an aneurysmal tissue [51].

For the **balloon sizing technique** of the largest defect, the disappearance of the left-to-right flow is followed with the inflation of the second balloon, and if the distance is less than 5–7 mm between the two openings, a single occluder is chosen (Figures 2–4) [44, 47, 52]. At the diameter obtained by balloon sizing at stop-flow, 2 mm should be added to avoid device erosion. This measure should be less than the total septum measured in the four-chamber view [36]. An interesting possibility was expressed by Carano *et al.*, who performed atrial balloon septostomy in cases that could not be closed either by using a large device or by two to three smaller devices [53]. However, the risk to obtain an unpredictable enlargement of the hole that can interfere subsequently with the interventional closure of the new defect. The effect of balloon stretching on the largest ASD in multi-fenestrated aneurysmal ASD was studied by Baruteau *et al.* on 161 children. The balloon-stretched diameter increased from 15 mm/m<sup>2</sup> (TEE)–17 mm/m<sup>2</sup> (TTE) to

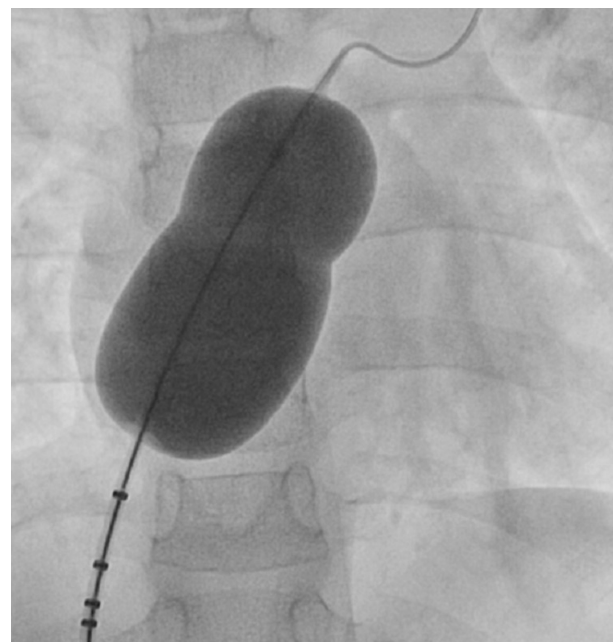
26.3±6.3 mm/m<sup>2</sup>. Extreme caution should be used when the ASO/left atrial (LA) length ratio is >1, given the risk of cardiac deformation and interference with adjacent structures. By stretching the defect to 26.3±6.3 mm/m<sup>2</sup>, Baruteau *et al.* obtained a closure rate of 92.6% [45].

After transcatheter device closure, antiplatelet treatment (single or dual) has to be followed for at least six months.

**Surgical closure** was the first approach in ASD and remain still the only solution for all types of ASD except *ostium secundum* ASD. Also, large *ostium secundum* ASDs have as single solution the surgery [1]. Surgery of the ASD is associated with reduced mortality and morbidity, but it is more invasive, having associated the risk of the cardiopulmonary bypass [54]. The procedure is taking place under general anesthesia, followed by median sternotomy, arterial and venous cannulation, cardiopulmonary bypass, and cardiac arrest. A right atrial incision is performed. Following the type and the dimension of the ASD, the techniques may vary.

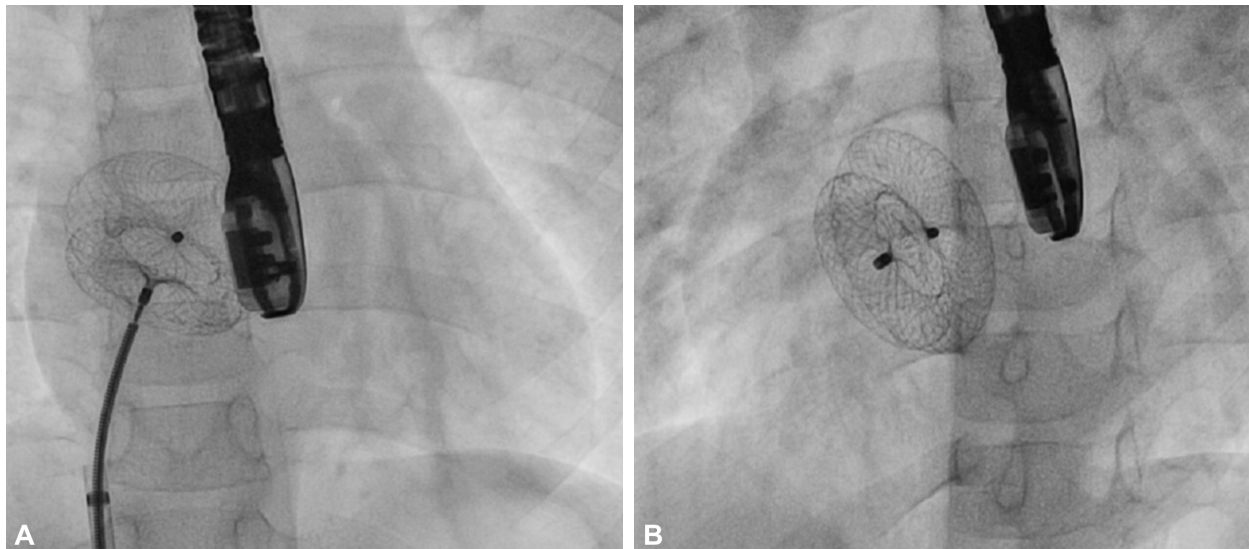


**Figure 1 – Atrial septal defect using the transesophageal echocardiography dual two-dimensional and color Doppler imaging. The images show five openings into the aneurysmal atrial septum in a 7-year-old girl who presented with palpitations in our Clinic.**

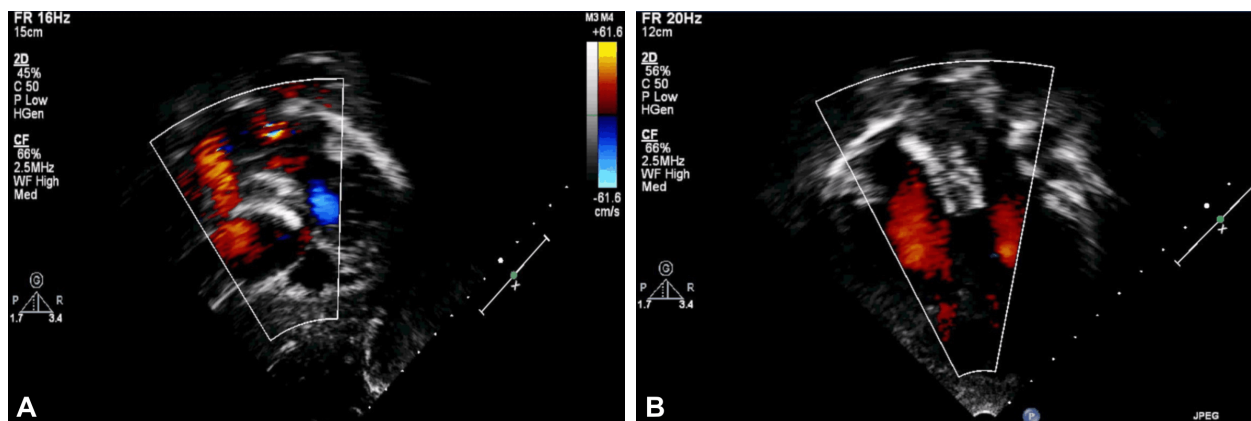


**Figure 2 – Sizing balloon on angiographic antero-posterior image showing the real measurement of the atrial septal defect (ASD), difficult to evaluate in multi-fenestrated five openings ASD of the same patient.**





**Figure 3 – Closure of the multi-fenestrated ASD by a device with a 22 mm ASO: (A) Before releasing the device; (B) After releasing the device. ASD: Atrial septal defect; ASO: Amplatzer Septal Occluder.**



**Figure 4 – A 22 mm Amplatzer Septal Occluder was implanted, with a stable position and no residual shunt, at distance from the atrioventricular valves and the walls of the atria: (A) Subcostal view; (B) Apical four chambers view.**

In *ostium primum* ASD, the corrective patch has to be attached to the atrioventricular septum, between the tricuspid and mitral valve. Also, the mitral cleft has to be repaired or a mechanical prosthesis has to be implanted in the most complicated cases.

In superior venous sinus ASD, there is also a partial anomaly of the pulmonary venous drainage (96% of the cases), which has to be redirected towards the left atrium. The connection of the anomalous pulmonary vein is towards the right atrium, towards the connection of the right atrium with the superior vena cava, or with the superior vena cava (in most cases). Three techniques are described for this type of ASD, the single patch, the two-patch technique and the Warden repair [55, 56].

Inferior *sinus venosus* defect is a very rare type of ASD, which has a more difficult repairing technique [57]. In a study published on 45 patients with this type of defect, a worse operator outcome was present, a higher reintervention rate (9%), and prolonged total cardiopulmonary bypass time. Also, the preoperative diagnosis was correct in only 36% of the patients [58].

The coronary sinus defect is the rarest type of ASD. There are many corrective techniques, and they are chosen according to the anatomy, partially or completely unroofed coronary sinus, with or without persistent left superior

vena cava (PLSVC) into the left atrium. Mainly, consists in patching the defect, rerooting the coronary sinus and redirecting the PLSVC into the right atrium (using the right appendage), superior vena cava, or left pulmonary artery.

More and more minimally invasive surgery with a reduced dimension of the incision is used for surgical closure of the ASD for cosmetic reasons. Also, different locations of the incision, different from the medial sternotomy are described: submammary, hemisternotomy, limited lateral thoracotomy or limited sternotomy. Technically, it is more difficult to realize the closure of the ASD, but this approach is not associated with decreased mortality or morbidity. In an intervention on 37 patients, the results were excellent [59–61].

There are minimal differences in the outcome in a comparative evaluation between surgery and interventional closure. After transcatheter closure, only by echocardiography, some differences could be noticed. The left atrial volume index, the left ventricle myocardial performance index and the right ventricle performance index (RVPI) improved, while by surgery the RVPI was not influenced [62]. In another study using strain-rate imaging in similar groups, it was noticed that both atrial functions were preserved only for the interventional approach at nine

years from intervention [63]. The surgery is a cheaper solution associated with a prolonged hospitalization comparing with the transcatheter intervention, which remains a cosmetic solution, less traumatic [64, 65]. There is no difference between the two interventions (surgical and transcatheter) regarding the survival, the functional capacity, atrial arrhythmias or embolic neurological events [66].

### ❏ Specific aspects related to the complications

Natural survival rate after the age of 50 years is approaching 50%. Complications may be related to the natural history of the disease, such as heart failure, atrial arrhythmias, pulmonary hypertension, paradoxical embolism, and associated cerebral ischemia, or with the corrective procedures. Immediately after the closure procedure, there is an immediate improvement in the clinical manifestation more in patients in which the procedure was performed before 25 years old [67, 68]. Preexisting atrial arrhythmias and neurological events may remain a long-term risk after the procedure [69]. Early surgical intervention in infants is associated with the risk for late supraventricular tachycardia.

Severe preexisting pulmonary hypertension with reversed shunt may have a worse prognosis if the closure is attempted. In rare cases with reversible pulmonary hypertension or diastolic dysfunction of the right or left ventricle, fenestrated devices may be implanted after the test occlusion. However, fenestrations may close spontaneously in 50% of the cases in up to 20 months [17].

Interventional closure may be associated with a procedure-related complication, such as thrombus formation, infection, pericardial effusion, transitory ischemic attack, device embolization, cardiac perforations and erosions or new complete atrioventricular block, supraventricular tachycardia (atrioventricular nodal reentrant tachycardia) or ventricular extrasystoles [19, 70–72].

Device embolization is a complication that may occur in 1% of the cases usually related to an undersizing of the defect, in the presence of the soft, floppy rims or reduced operator experience. Usually, it happens immediately or in the next 24 hours but also late displacement was described [68, 73].

Device implantation may be associated with the risk of rhythm (1–4%) and conduction problems. If a device related complete atrio-ventricular block (AVB) appears, which persist after corticoid therapy, then the device has to be removed. Risk factors for complete AVB device related are a deficient posteroinferior rim, the use of larger devices or pre-existing conduction abnormalities.

Cardiac erosions may appear in 0.1–0.3% of the cases, usually early (within one year after device closure), but also late after the procedure [19, 74–77]. The walls that are most frequently affected are the roof of the right or left atrium, or the atrial junction with the aorta. Erosion and perforation may lead to hemopericardium, pericardial tamponade or aortic fistula. As manifestation, they present with chest pain, shortness of breath, hemodynamic collapse or sudden cardiac death. Pericardial effusion at discharged echocardiography should prolong the hospitalization for several days for echocardiographic

follow-up [78]. The mortality rate for this complication is 10% [75]. Treatment consists of emergency cardiac surgery. Risk factors for cardiac erosion are oversized device, deficient anterior and superior rims, multiple attempts to position the device, occluder type (Amplatzer, Occlutech, and Cardia), adult age, movement of the device into the heart [19]. Oversized devices on a floppy atrial septum may lead also to tissue dissection or arrhythmogenicity [38].

After device implantation, the patient needs to receive antiplatelet therapy for at least six months in order to prevent thrombus formation. During this period endothelialization appear, even in multiple device implantation with device overlapping [79]. In specific categories of patients (resistance to Aspirin, thrombophilia disorders, atrial fibrillation), thrombus may appear (1.2–70% at four weeks after implantation) [80]. Some authors recommend dual antiplatelet therapy (Aspirin and Clopidogrel) [81]. Thrombus formation is not only a transcatheter complication [82, 83]. It may appear also after surgical closure of an ASD [84].

Residual shunts may persist in 20% of the cases but in >90% they are small [85, 86].

The surgical mortality rate is around 1% in younger patients in the absence of moderate or severe pulmonary hypertension and depends also, on the severity of the associated lesions [87, 88]. The rate of surgical complications varies with the type of ASD and type of intervention. Cardiac arrhythmias may be present in up to 36% after surgical closure of ASD [72, 89].

For venous sinus defects, postoperative complications are related to the stenosis of the superior vena cava or of the pulmonary veins, with residual shunting and arrhythmias of the sinus node (low atrial or junctional rhythm) more frequently on the patients with the two-patch repair. For the patients with a partial pulmonary venous connection to the superior vena cava, the Warden procedure is the most used, and for the partially pulmonary venous connection to the RA or the RA–SVC junction, the single patch technique is the best option [55, 90, 91]. Surgery for coronary sinus defect usually has a very good outcome.

### ❏ Conclusions

Complex ASDs are not very rare in clinical practice. For non-*ostium secundum* ASDs, the only indication is surgery although for specific types of ASD (superior venous sinus and coronary sinus defects) attempts for interventional closure were done. In the *ostium secundum* type, depending on the anatomical aspect, the therapeutic algorithm starts with the evaluation of the possibility of interventional closure. Interventional closure has the advantage of a good cosmetic appearance and reduced hospitalization duration. Using a sizing balloon is mandatory for complex defects to control the real diameter. Oversizing according to the sizing balloon and not to the initial defect diameter may give a solution in such cases. Interventional closure of aneurysmal multi-fenestrated septum defect is challenging but is the ideal therapeutic method. Possible complications (cardiac erosion and perforation, device embolization, stroke) are severe but rare. For patients with a large ASD or if they have contra-

indications for the transcatheter procedure, surgery remains the only possibility. Surgery has a very low mortality rate that tends to zero, is cheaper, but it has the disadvantage of having a chest scar, even for minimally invasive interventions, and a longer hospitalization rate.

#### Conflict of interests

None declared.

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