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Atrial septal defect

Chapter: Atrial septal defect

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Summary

Secundum atrial septal defects are usually well tolerated in childhood but may cause significant symptoms in adults. Early closure is therefore recommended and can be achieved by catheter in the

majority. Symptomatic benefit is noted at any age, but long-term follow-up is required as closure in adulthood does not prevent atrial arrhythmias.

Introduction

Atrial septal defect (ASD) is a common congenital heart defect with a birth prevalence of 1.43:1000 live births and an expected survival into adulthood of 97%.[1,2] The ostium secundum ASD is the most common (70%), followed by the ostium primum (10% and considered under atrioventricular septal defects) and the sinus venosus ASD (5–10%)[3] (Figure **17.18.1**). Patients with ostium secundum type ASDs are predominantly female (65–75%), while the gender distribution is evenly distributed in patients with ostium primum or sinus venosus ASDs.[3,4] Associated lesions include anomalous pulmonary venous connection, persistent left superior vena cava, pulmonary valve stenosis, and mitral valve prolapse. Associations with syndromes (e.g. Holt–Oram syndrome) and upper limb deformities have been described.

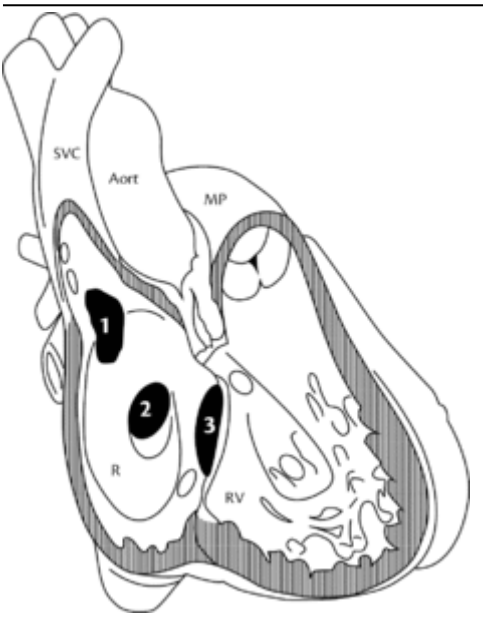

Figure 17.18.1
Types and typical locations of atrial septal defects (ASDs). In this view, the free walls of the right atrium and right ventricle have been removed, looking leftward towards the septal surface. 1, sinus venosus ASD at the junction of the superior vena cava (SVC) and right atrium; 2, ostium secundum ASD; and 3, ostium primum/atrioventricular septal defect type ASD. Aort, aorta; MP, main pulmonary artery; RV, right ventricle.

Reproduced with permission from Sommer RJ, Hijazi ZM, Rhodes JF. Pathophysiology of congenital heart disease in the adult. Part I: shunt lesions. Circ 2008;117:1090–9.

Morphology

The true atrial septum (the tissues that directly separate the atrial cavities) is confined to the floor of the oval fossa and the surrounding inferoanterior rims.[1] Defects of the true atrial septum are referred to as secundum defects (Figure **17.18.2**, Figure **17.18.3**, and Video 17.18.1(online)). Sinus venosus defects (which are often associated with abnormal pulmonary venous connections), coronary sinus defects, and ostium primum defects are communications between the atrial chambers but outside the true atrial septum.

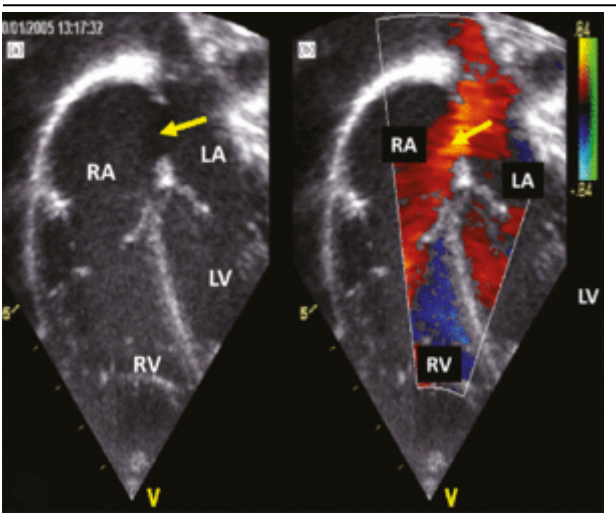

Figure 17.18.2
A large secundum atrial septal defect is demonstrated on two-dimensional and colour echocardiography. There is left-to-right flow through the defect (demonstrated by the yellow arrow). LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

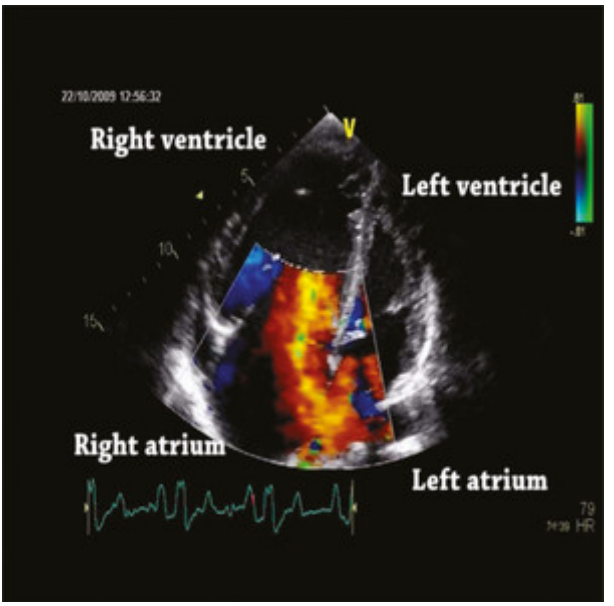

Figure 17.18.3
Secundum atrial septal defect with left-to-right shunt, dilated right atrium, and right ventricle.

[This video cannot be viewed in PDF format. To view it, please go to the original web version of this chapter.]

Video 17.18.1(online) Transthoracic two-dimensional echocardiogram showing secundum type atrial septal defect. Colour flow mapping demonstrates the left-to-right shunt across the defect.

Pathophysiology

The left-to-right shunt across the ASD results from the greater compliance of the right ventricle than the left ventricle.[7,8,9] As left ventricular compliance decreases with age, or in the presence of conditions that may raise left atrial pressure (e.g. hypertension, ischaemic heart disease, cardiomyopathy, aortic and mitral valve disease), the left-to-right shunt through the ASD may increase. Conditions that reduce right ventricular compliance (including pulmonary arterial hypertension, pulmonary stenosis, right ventricle disease, tricuspid valve disease), will reduce left-to-right shunting and may eventually reverse the shunt, causing cyanosis.

A left-to-right shunt results in right ventricular volume overload and this will cause right ventricular dilatation. It is well tolerated throughout childhood, despite a pulmonary-to-systemic flow ratio which may exceed 3:1. The pulmonary vasculature is also able to accommodate the increased blood flow at low levels of pulmonary artery pressure for many years. A persistently large left-to-right shunt will result, from late childhood, in increased right atrial and right ventricular dilatation, predisposing to arrhythmia and a progressive increase in pulmonary vascular resistance in some patients. Severe pulmonary vascular disease is rare (<5%) unless there are other associated factors.

Clinical presentation and natural history

Most patients remain well throughout childhood, although they can present with failure to thrive, breathlessness, recurrent chest infections, or rarely heart failure. By the fourth decade, most patients develop symptoms with palpitation, shortness of breath, chest infections, and occasionally right heart failure. Patients often deteriorate when atrial arrhythmias (atrial fibrillation or flutter) develop. Patients may occasionally be identified in chest clinics. Life expectancy is reduced without treatment and patients benefit from closure, even at an advanced age.

Key clinical findings are fixed splitting of the second heart sound, right ventricular heave, ejection systolic murmur, and mid-diastolic murmur. A loud second heart sound with cyanosis indicates pulmonary hypertension.

Investigations

◆ *Electrocardiogram* (*ECG)*: the ECG usually shows right axis deviation and incomplete right bundle branch block. Right ventricular hypertrophy may be present. In primum ASD, there is left axis deviation.

◆ *Chest radiography*: chest radiography shows right atrial/ventricular dilatation with increased pulmonary vascularity.

◆ *Transthoracic echocardiogram*: this is the best technique for diagnosis and quantification, which is based on right heart enlargement. Doppler assessment of tricuspid regurgitation provides an estimate of pulmonary artery pressure.

◆ *Transoesophageal echocardiogram*: two- and three-dimensional transoesophageal echocardiography will assist both in diagnosis and assessment of suitability for catheter closure. This is based on size, position, rim size, and quality. Transoesophageal echocardiography is an important part of catheter ASD closure (Figure **17.18.4**).[5]

◆ *Cardiovascular magnetic resonance imaging/cardiac computed tomography*: may be useful if echocardiography is not diagnostic and may assist in quantification of cardiac dimensions and function, as well as definition of pulmonary venous connections in sinus venosus ASD.

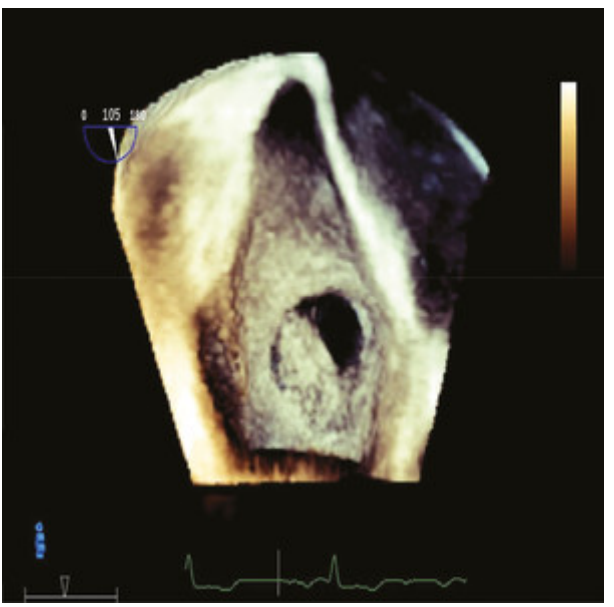

Figure 17.18.4
Three-dimensional view of the atrial septal defect from the right atrium, obtained by transoesophageal echocardiography.

Management

Closure of the defect has been shown to reduce cardiac mortality[6,7] and morbidity.[6,8,9] Although there have been concerns regarding the benefit of closure in older patients,[7,10,11,12] surgical repair has been shown to be superior to optimal medical management in ASD patients older than 40 years.[9] ASD closure results in symptomatic and haemodynamic improvement even in older patients.[13] A haemodynamically significant ASD (shunt ratio >1.5 or right ventricular dilatation) will generally be closed in childhood. A small ASD, the presence of pulmonary arterial hypertension that is too advanced, or older patients with left ventricular dysfunction are reasons not to close the ASD.[3]

Surgical repair has very low risk in the absence of co-morbidities and has an excellent long-term prognosis when performed early and in the absence of pulmonary hypertension.[11,14] Transcatheter device closure has become the treatment of choice for secundum ASD when it is feasible (Figure **17.18.5**, Video 17.18.2(online), and Video 17.18.3(online)). Serious complications are very rare (<1% of patients), and comparative studies with surgery have shown lower morbidity and shorter hospital stay. A number of different devices are now available. Antiplatelet treatment to allow for device endothelialization, is required for 6 months. Patients with large defects (stretched diameter >38 mm), inadequate septal rims (<5 mm, except towards aorta), or proximity to the atrioventricular valves, coronary sinus, or vena cavae should be referred for surgery.

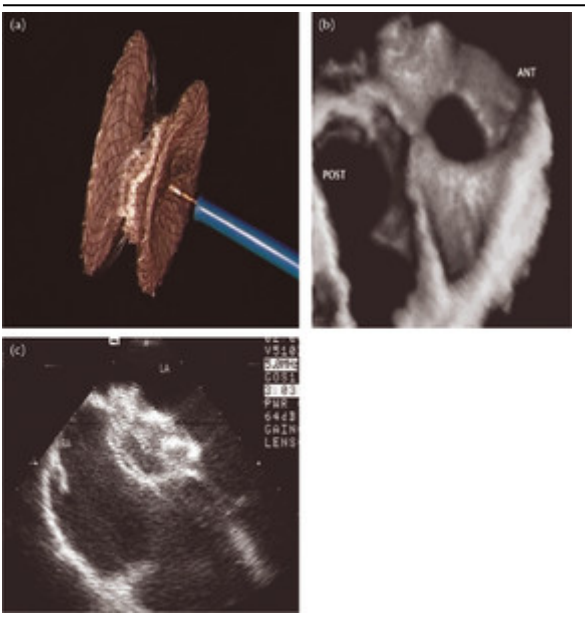

Figure 17.18.5
Transcatheter occlusion of atrial septal defect (ASD). (a) Amplatz ASD device attached to delivery wire and shown outside long delivery sheath. (b) Three-dimensional echocardiogram demonstrating ASD seen from right atrial aspect with clear superior and inferior margins. (c) Transoesophageal echocardiogram following delivery and release of Amplatz ASD device across defect. LA, left atrium; RA, right atrium.

[This video cannot be viewed in PDF format. To view it, please go to the original web version of this chapter.]

Video 17.18.2(online) Occlusion of a secundum type atrial septal defect being demonstrated in a model heart viewed from the left and right sides using an Amplatz septal occluder.

[This video cannot be viewed in PDF format. To view it, please go to the original web version of this chapter.]

Video 17.18.3(online) Transoesophageal two-dimensional echocardiogram showing Amplatz septal occluder across the atrial septum post release with colour flow mapping confirming the absence of any residual left-to-right shunt across the atrial septum.

Long-term outcome

Untreated patients have decreased life expectancy, although they may reach an advanced age without closure.[15,16,17] In contrast to unrestricted ventricular septal defect/patent ductus arteriosus, only 10–20% of all patients with ASD progress to pulmonary arterial hypertension, probably reflecting the different pulmonary vascular response to volume and pressure overload. The incidence of atrial fibrillation and of atrial flutter increase steeply with age, and are associated with increased morbidity and mortality. Closure is thus best undertaken in childhood and adolescence as atrial arrhythmias may persist or recur when ASD closure is performed in adulthood. Despite this, patients benefit symptomatically from closure of a significant ASD at any age. Nevertheless, in elderly patients with co-morbidities, who are not suitable for device closure, surgical risk must be carefully considered against potential benefits.

Additional considerations

◆ *Exercise/sport*: no restrictions required in the absence of pulmonary arterial hypertension.

◆ *Pregnancy*: risk is low in the absence of pulmonary arterial hypertension, but it is contraindicated in patients with severe pulmonary arterial hypertension or Eisenmenger syndrome.

◆ *Infective endocarditis*: does not occur in isolated ASD and prophylaxis is not required. It may be recommended for 6 months after device closure.

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