Atrial septal defects – a differential diagnosis for breathlessness in adults and the elderly

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Abstract

Atrial septal defects are a common form of congenital heart disease that can present at any age, even in the elderly. As symptoms may be non-specific (breathlessness, palpitations), a high index of suspicion should be maintained. The ECG may be normal in the absence of significant pulmonary hypertension although a chest radiograph should be helpful. The diagnosis is usually confirmed by transthoracic echocardiography, although some types of atrial septal defects may be missed in adults who are poor echo subjects. Transoesophageal echo provides definitive diagnostic information and should be undertaken in any patient with right heart dilatation of unknown cause. Whilst closure of atrial septal defects may not prevent atrial arrhythmia, it can reduce the haemodynamic consequences if episodes occur. Many atrial septal defects can now be closed with percutaneous devices, avoiding the need for sternotomy.

Key words: breathlessness, atrial arrhythmia, atrial septal defect, congenital heart disease.

Introduction

Congenital heart disease may present at any age, including in the elderly. Symptoms may be similar to those that occur in conditions more commonly associated with the older age group, such as left ventricular failure or chronic lung disease. Closure of atrial septal defects (ASDs), either with a percutaneous device or surgically, can provide dramatic improvement in symptoms. ASDs should be considered in the differential diagnosis of all patients who present with breathlessness or atrial arrhythmia.

Patient 1

A 63 year old woman was referred with a diagnosis of heart failure. She reported increasing breathlessness, such that she avoided walking and had given up playing golf. She had suffered one episode of atrial fibrillation that had made her fall to her knees. Physical examination revealed a widely split second heart sound

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Figure 1. Chest X-ray of patient 1, showing an enlarged heart with plethoric lung fields prior to treatment

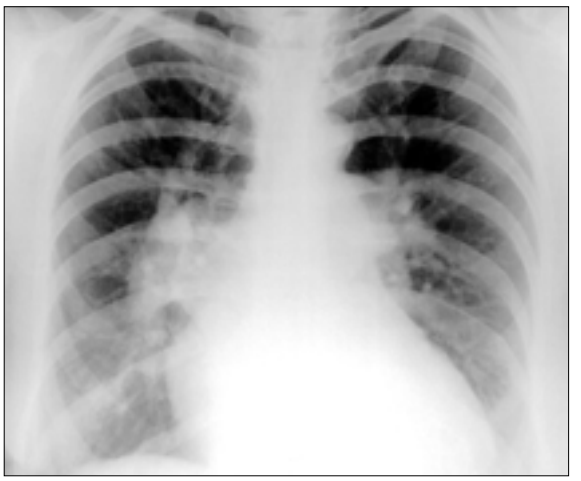


Figure 2. The post-operative chest X-ray shows the dramatic reduction in heart size and lung plethora

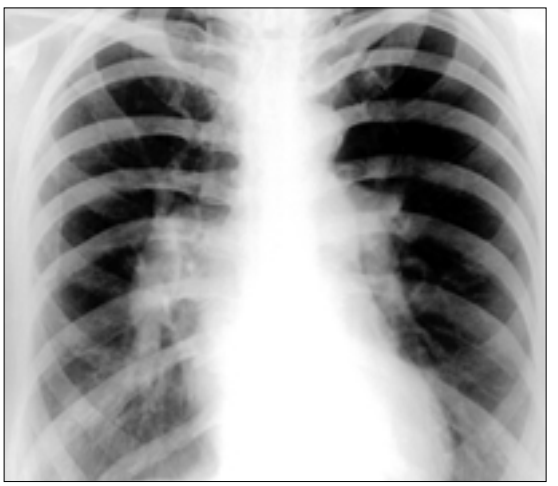


Figure 3. TOE image showing the Angel Wings™ device in the atrial septum. The left atrium is at the top of the screen and the right atrium (RA) at the bottom

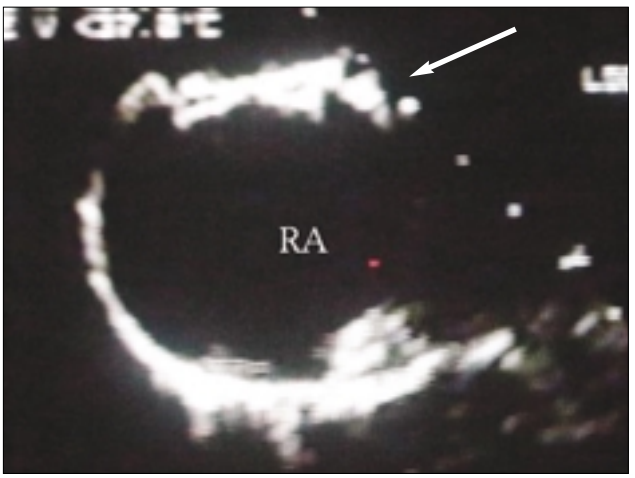
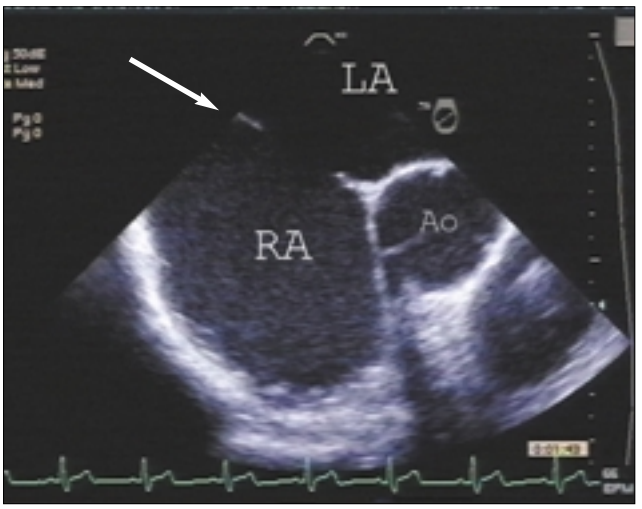


Figure 4. TOE appearance of an untreated secundum ASD (RA = right atrium, LA = left atrium)



and a pulmonary flow murmur. Chest radiography (figure 1) demonstrated an enlarged heart, large pulmonary arteries and pulmonary plethora. The electrocardiogram (ECG) showed right bundle branch block. Echocardiography was performed, which confirmed the clinical diagnosis of ASD. At catheterisation the pulmonary pressure was found to be raised at 55/15 mmHg (mean 29), in addition to a left-to-right shunt of 3.3:1 and normal coronary arteries.

Surgical repair was undertaken. This confirmed the diagnosis, revealing a large ASD extending from close to the inferior vena cava to the superior aspect of the foramen ovale. The ASD was closed with an autologous pericardial patch. Post-operatively her heart size returned to normal and the chest X-ray findings largely resolved (figure 2).

Ten years later she remains well and active. Her only symptoms relate to occasional paroxysms of atrial fibrillation that are well tolerated.

Patient 2

A 63 year old woman presented to the physicians with palpitations associated with presyncope. She had a history of recurrent palpitations that were associated with near collapse. Over the previous 10 years she had become increasingly short of breath on exertion. A 24-hour tape confirmed paroxysmal atrial flutter and chest X-ray was consistent with an atrial septal defect. The atrial septal defect was confirmed by echocardiography and moderate right ventricular volume overload was noted.

ASD closure was undertaken with the Das Angel Wings™ device (figure 3). Following the procedure the patient was less short of breath and was also much less troubled by her palpitations. Although the palpitations occurred as frequently as before, she was only vaguely aware of them and was able to go about her daily chores without interruption.We would speculate that, prior to ASD closure, the onset of atrial flutter led to a sudden increase in left atrial pressure, an increase in left-to-right shunting and a fall in systemic cardiac output. After closure of the ASD the fall in cardiac output was reduced and symptoms were minimised.

Discussion

Breathlessness is a common symptom among adults presenting to general practitioners and physicians. The differential diagnosis naturally includes chronic lung disease and left ventricular failure, but most adult physicians forget that congenital heart disease may present in middle-aged and elderly patients. As the heart ages the left ventricle can develop diastolic dysfunction (becoming 'stiffer'), leading to increased left atrial pressure. In the presence of an ASD this can lead to an increased left-to-right shunt (particularly on exercise), and can precipitate the onset of symptoms.

Atrial fibrillation (AF) and other atrial arrhythmias are a common late development in patients with an ASD. A paroxysm of AF further impairs LV filling, which may lead to a drop in systemic cardiac output, occasionally associated with syncope. In both case reports described above the patients suffered an atrial arrhythmia that caused haemodynamic compromise. In the second patient, closure of the ASD did not prevent the arrhythmia but attenuated the haemodynamic effects so that she was only minimally inconvenienced by the arrhythmia following closure.

The electrocardiogram in a patient with an ASD classically demonstrates incomplete right bundle branch block but it can be normal. With the onset of pulmonary hypertension there will be evidence of right ventricular hypertrophy. Transthoracic echocardiography will usually be able to detect a secundum ASD (figure 4), by far the most common type of ASD, but may not detect the rarer sinus venosus defect. Right heart dilata-

Figure 5. An Amplatzer septal occluder *in situ,* closing the defect between left atrium (LA) and right atrium (RA)

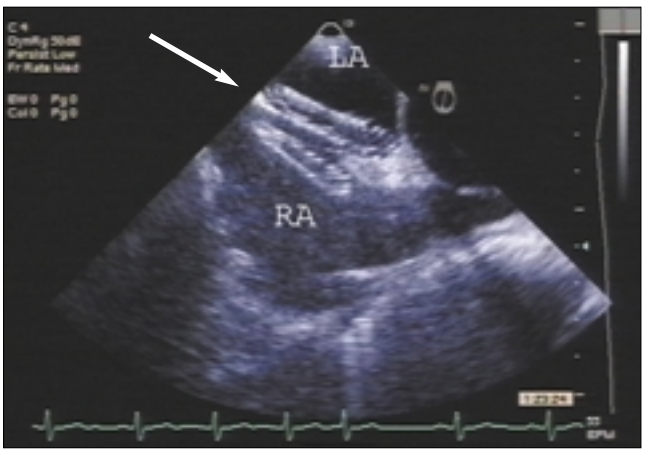
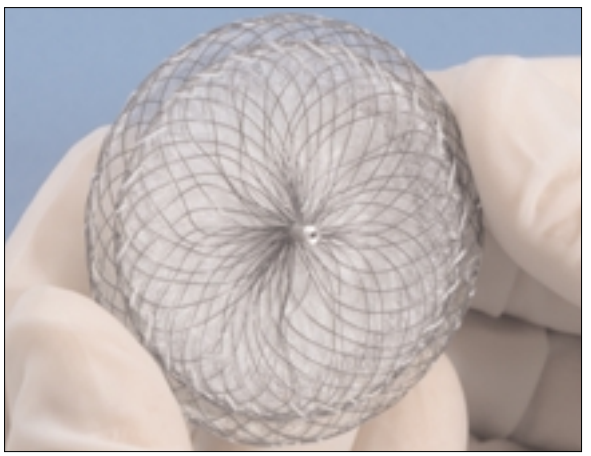


Figure 6. A 34 mm Amplatzer device, viewed from the right atrial surface, demonstrating the screw thread in the centre of the right atrial disc, that connects the device to the delivery system



tion may be apparent which, in the absence of overt lung disease, should provoke a search for alternative diagnoses. Bubble contrast echocardiography can be useful and may be used in combination with a Valsalva manouevre, when contrast may be visualised in the left heart. If, however, there is still unexplained right heart dilatation, transoesophageal echocardiography (TOE) should be considered as this has a high diagnostic yield for atrial septal defects and the pulmonary venous abnormalities that often accompany sinus venosus ASD. If TOE is not available, right heart catheterisation can be performed to rule out a left-to-right shunt and to measure the pulmonary artery pressure and vascular resistance directly.

Thermodilution cardiac output can be used to measure pulmonary blood flow when an ASD is present by placing the proximal port (through which the cold saline is injected) in the right ventricle (confirmed by pressure tracing or contrast injection). By using this method to measure pulmonary flow in combination with pulmonary artery and wedge or left atrial pressure, the pulmonary vascular resistance can be accurately calculated. Lung function tests may give a clue if there is a significant left-to-right shunt since, in the presence of normal ventilatory volumes, there may be an elevated rate of uptake of carbon monoxide (K_{CO}) due to the high lung blood flow.

Elderly patients with ASD who have symptoms and right heart dilatation should be considered for closure of the defect. Percutaneous closure using a device (figures 5 and 6) can be performed for many ASDs (even multiple ASDs) though surgical closure is currently necessary for sinus venosus defects and if anomalous pulmonary venous drainage is present. Percutaneous device closure reduces hospital stay and convalescence time and should be offered for appropriate patients.

Dramatic symptomatic improvement can occur following ASD closure at any age and the chest X-ray changes can also be dramatic (figures 1 and 2). Hence ASD is a highly treatable cause of breathlessness in all age groups. ASD closure may not reduce the incidence of atrial arrhythmia but, by reducing the haemodynamic consequences, it may lead to a dramatic reduction in patients' symptoms. Since late atrial arrhythmia following surgical closure may involve circuits around the atriotomy scar, it is possible that device closure may in future be shown to reduce this complication.

Key messages

- Patients with ASD may present at any age
- The ECG can be normal in patients with ASD
- Patients with a dilated right heart of unknown cause should have transoesophageal echocardiography
- Many ASDs can be closed without the need for open surgery
- The haemodynamic consequences of atrial arrhythmia may be reduced by ASD closure

Editors' note

This is the first in a series of articles on congenital heart disease management. The management of patent foramen ovale is considered in a future issue.