Atrial septal defect: a coincidental finding on a screening medical

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Abstract

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An otherwise fit, healthy medical practitioner who was a recreational diver underwent a medical assessment for a remote posting as an Antarctic Medical Practitioner at which a coincidental finding of an atrial septal defect (ASD) was made. ASDs can have health implications in extreme environments such as high altitude and is contraindicated in scuba diving. ASDs are common, being present in 1:1,500 live births and comprise 10% of all cardiac abnormalities. In this case, a percutaneous occlusive device was inserted under general anaesthetic with subsequent improvements in the practitioner's exercise capacity, return to diving and full employment, including Antarctic deployment, and right-sided heart remodelling 18 months post closure.

Key words
Right-to-left shunt; cardiovascular; radiological imaging; treatment; occupational health; case reports

Introduction

Atrial septal defect (ASD) is generally regarded as a contraindication for scuba diving because of the associated potential increased risk of decompression illness (DCI). It may also impact on occupational activities in other extreme environments such as deployment to altitude. Here, a medical practitioner reports her experience with the discovery at a routine pre-deployment medical of an ASD and its impact on her recreational and professional life.

Case report

An otherwise fit, healthy 33-year-old female medical practitioner who was a recreational diver underwent a medical assessment for a remote posting as an Antarctic Medical Practitioner (AMP). The screening included an exercise stress test (EST) because the deployment included high-altitude exposure. This EST resulted in a cascade of events that lead to a diagnosis of an ASD.

The diver recalled infrequent, brief attacks of palpitations lasting a few seconds that had begun the previous year, blamed on stress and caffeine. While she had always enjoyed an active lifestyle with regular aerobics, middle distance running and competitive Irish dancing to National levels, she expressed difficulty maintaining cadence with running owing to fatigue and shortness of breath on exertion. Past history was significant for pertussis in early infancy and mumps as a child despite immunisation, plus childhood asthma with several infective exacerbations annually. An Open Water diving certification was obtained in 2011 (with a negative bronchial provocation test in an earlier Antarctic medical). However, she had undertaken only a limited number of dives without event. Numerous extended overseas trips and several altitude exposures to 4,700 m in South America and 2,500 m in Greenland had ended otherwise uneventfully from a health viewpoint.

An ECG in 2008 for an expedition to Greenland showed P-wave inversion but no further assessment was recommended at that time, as she was otherwise asymptomatic, fit and well. The 2013 EST showed that, with exercise provocation, the P-waves reverted and the EST was completed routinely. No clinical signs were elicited on examination, and chest X-ray did not reveal cardiomegaly or prominent pulmonary vasculature. Transthoracic echocardiography (TTE) showed a secundum ASD measuring 12 mm with a shunt calculated at 1.8 (pulmonary flow/systemic flow ratio). Also of note was a mildly dilated right atrium (RA) and ventricle with an enlarged inferior vena cava to the upper limit of normal and mild pulmonary artery (PA) dilatation. Considering the size of the shunt, closure of the ASD was recommended. This diagnosis precluded medical fitness for her proposed Australian Antarctic service.

The diver travelled interstate to have an Occluotech Figulla® device inserted percutaneously under general anaesthesia with transoesophageal echocardiogram (TOE) and fluoroscopic guidance. Intraoperatively a balloon catheter was used to measure an appropriately sized device, and revealed a much larger defect, requiring a 33 mm occlusive device. There were no post-operative complications and aspirin and clopidogrel were prescribed for three months, with a subsequent three months at a twice weekly dose, then cessation. A secondary recommendation was to encourage screening for the extended family, with an older sibling also having a small ASD discovered. Coincidentally, a nephew has a congenital ventricular septal defect, which had been detected at birth.

Follow-up TTE at six weeks, three months, and a bubble contrast TTE at six months indicated no residual inter-atrial defect, which leant a green light to undertaking a dive medical and an uneventful resumption of scuba diving in mid-2014. From diagnosis until the six-week check-up, risk

Table 1
Current management recommendations for atrial septal defects

Medical Therapy
- Small ASD < 5mm[4]
- Normal RV size[4]
- No pulmonary arterial hypertension[4]

Interventional (occlusion device) and surgical closure
- Significant shunt Qp/Qs ratio > 1.5 + presence of:
  - ° Recurrent respiratory tract infections (paediatrics)[2]
  - ° Failure to thrive (paediatrics)[2]
  - ° Exercise intolerance[5,6,8]
  - ° Dyspnoea[5,6,8]
  - ° Fatigue[5,6]
  - ° Palpitations[6]
  - ° Heart failure[5,6,8]
  - ° Atrial arrhythmias[5,6,8]
- Right-sided heart enlargement ± symptoms[4]
- Paradoxical embolism[4]
- Orthodeoxia-platypnoea[4]
- Pulmonary arterial hypertension (not severe)[2]

Device closure
- < 40 mm[2,9]
- adequate rims of tissue (> 5 mm) from the defect to surrounding structures[2]

Table 2
Post-operative complications of percutaneous atrial septal defect closure; SVT – supraventricular tachycardia

Minor

Early
- Arrhythmia (atrial fibrillation/flutter; SVT)[5,11,13]
- Heart block[5]
- Femoral access site complications[5]
- Blood transfusion (rare)[13]

Late
- Nickel allergy[11]

Major

Early
- Device malpositioning/embolisation (rare)[3,5,7,11]
- Air embolism[12]
- Device thrombus[5,7]
- Pericardial effusion with tamponade[5,7]
- Mitral valve dysfunction[3,5]
- Venous obstruction[3,5]
- Cardiac perforation (rare)[3,5]
- Stroke[7]

Late
- Device erosion[3,5,7]
- Endocarditis[7,11]
- Thromboembolism[5,11]

mitigation for working as a medical officer in a hyperbaric medicine unit included having another doctor present to undertake in-chamber duties. After the six-week check to the six-month bubble contrast echo, restricted clearance was granted to undertake quick 'dips' in the chamber to provide acute patient care. Long-term follow up plans include an annual TTE, the first of which showed resolution of RA and PA dilation, and all previous activities, including scuba diving and employment with no restrictions have been resumed, including middle distance running at 30 seconds less per kilometre with little training and she has recently returned from Antarctic deployment.

Discussion

ASDs are one of the most common congenital cardiac defects (10% of all heart abnormalities and approximately 1 in 1,500 live births, with a 2:1 female to male ratio for secundum defects).[2,3] There are four anatomical subsets of ASD: secundum (75%), primum (15%–20%), sinus venosus (5%–10%), and coronary sinus ASD (< 1%).[4] The latter three are not conducive to percutaneous device repair due to their anatomical locations.[2,4] An ASD can co-exist with a persistent patent ovale (PFO).[4] Most cases are sporadic; however, there are genetic markers for familial ASD including Holt-Oram syndrome and Ellis van Creveld syndrome (associated skeletal abnormalities), and mutations of GATA4 and NKX2.5 (associated conduction abnormalities).[3,5–7]

ASDs, regardless of size, often remain undiagnosed until later life. Generally a defect of less than < 8 mm diameter will close during infancy (approximately 4%), with spontaneous occlusion unlikely beyond early childhood.[6,8] If left untreated, defects tend to increase in size owing to left-to-right shunting across the atrial communication, causing symptoms of right heart and pulmonary vascular volume overload, which may become apparent from the third decade, with almost all lesions symptomatic by the sixth decade.[4,6,8] Symptoms include exertional dyspnoea, fatigue, syncope, and palpitations and are not correlated with shunt size.

Examination findings depend on the extent of the left-to-right shunt and its effect on the architecture of involved structures. An ASD itself does not produce an adventitious sound as there is no pressure gradient, and it is an acyanotic condition.[5,6] Only when the shunt is reversed due to increased pulmonary vascular resistance (Eisenmenger syndrome), may cyanosis and clubbing present.[6] ECG may show sinus rhythm, atrial flutter or fibrillation, with inverted P-waves indicating an absent sinus node, and potentially an incomplete right bundle branch block if right ventricular dilation is present. Chest X-ray may show cardiomegaly with prominent pulmonary vasculature, and low left heart output is evident with a small aortic notch.[3,4,6] ASDs are investigated with TTE and further detailed information achieved with TOE or contrast TTE with agitated saline.[7]

Current management recommendations for ASD are summarised in Table 1.

The first successful ASD closure was done via median thoracotomy on cardiac bypass in 1952, with the first device closure done surgically in 1974, and the first percutaneous device closure in 1991.[10,11] This has resulted in better cosmesis, cost reduction, reduced post-operative complications, and reduced hospital stay. Percutaneous closure with an endoluminal device is a safe and effective intervention for secundum ASDs < 40 mm, with low risk of intraoperative complications under expert proceduralist care (Table 2).[8]

There are currently three occluder devices on the Australian market, made of a nickel/titanium alloy (nitinol), the Amplatzer Septal Occluder (ASO), Helex Septal Occluder, and Figulla Septal Occluder (FSO).[8,9,11,12] Right-sided heart and pulmonary vasculature remodelling occurs post closure, with best overall morbidity and mortality favouring closure prior to 25 years of age.[3,6] Regardless of age at closure, patients will experience a degree of improved exercise capacity due to right ventricular remodelling, improvement in left ventricular function and decrease in pulmonary arterial pressures.[8]

Post-operative recommendations are to have a TTE at least every two years initially to ensure maintenance of device placement and evaluation of improvement in right heart size and pulmonary pressures.[5] Anticoagulation (i.e., aspirin and clopidogril) is given for several months up to a year post device closure owing to the risk of formation of thrombus until the device is epithelialized and a potential for atrial fibrillation.[3,5] There are conflicting recommendations for post-operative endocarditis antibiotic prophylaxis (risk approx. 0.8%).[4,5,7,11]

Persistence of a shunt may also occur; however, these are often small and close within 12 months post-operatively and are best assessed with a contrast TTE.[3] Recommendation for returning to normal sporting activities, including scuba diving, is within three to six months post-operatively provided that there is no evidence of right heart failure pulmonary arterial hypertension or arrhythmias.[5] Owing to a low risk profile, feasibility, and substantial symptomatic improvement due to post-operative cardiovascular remodelling, percutaneous device closure of secundum ASDs should be considered for all haemodynamically significant lesions, regardless of age.

Lessons learned by the author include a lower threshold as a doctor for discounting difficult to interpret symptoms or investigations in respect to cardiac anomalies, an appreciation for the public health system and exemplary collegiate support in times of need.

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