Cardiology in the Young (2014), **24**, 403–416
doi:10.1017/S1047951113002321

Review Article

Atrial shunts: presentation, investigation, and management, including recent advances in magnetic resonance imaging

Cheryl G. Zvaigzne,[1,2] Andrew G. Howarth,[1] David J. Patton[1,2]

[1]*Stephenson Cardiovascular Magnetic Resonance Imaging Centre, Department of Cardiology, Libin Cardiovascular Institute of Alberta, University of Calgary, Calgary, AB, Canada;* [2]*Department of Paediatric Cardiology, Alberta Children's Hospital, Calgary, AB, Canada*

Abstract Atrial shunts are a common finding in both paediatric and adult populations. Recent developments in advanced imaging have widened the options for diagnosis and evaluation of such shunts. This paper reviews the various types of interatrial communications, discusses the features of clinical presentation in adults and children, and provides an overview of the clinical assessment including physical examination, electrocardiography, echocardiography, cardiac catheterisation, computed tomography, and magnetic resonance imaging. Focus will be placed on recent developments in magnetic resonance imaging that may improve the non-invasive evaluation of atrial shunts.

Keywords: Atrial shunt; review; diagnostic imaging; magnetic resonance imaging

Received: 11 February 2013; Accepted: 24 November 2013; First published online: 7 January 2014

An atrial shunt, or interatrial communication, is any opening in the atrial septum that allows blood to flow between the right and left atria.[1] Without early detection and appropriate treatment, a patient with an atrial shunt is at risk of developing pulmonary hypertension, heart rhythm abnormalities, right-sided heart failure, and paradoxical stroke.[2–4] Management may include monitoring, percutaneous closure, or surgical closure, and depends on the size and location of the defect, as well as the presence of associated defects. Therefore, diagnostic imaging is required to evaluate the shunt and to rule out associated defects such as partial anomalous pulmonary venous anatomy.[2] Currently, most patients clinically suspected of having an atrial shunt are referred for evaluation by trans-oesophageal echocardiography.[4] Recently, advances in cardiovascular magnetic resonance imaging have produced several different techniques that can be applied in the detection and quantification of atrial shunts, offering an alternative, non-invasive imaging modality.[5]

This paper reviews the anatomy and presentation of atrial shunts in adults and children, and provides an overview of recommended clinical assessment including echocardiography, computed tomography, magnetic resonance imaging, and cardiac catheterisation. Focus will be placed on recent developments in magnetic resonance imaging that may improve the evaluation of atrial shunts and increase patient comfort.

Atrial septation and types of atrial shunts

In the foetus, elevated right atrial pressure and interatrial communication are essential to allow a bypass of the pulmonary circulation.[6] Atrial septation begins in the 6th week of development with infolding of the atrial roof to form a primary septum, led by a cap of mesenchymal cells, which migrate towards the endocardial cushions. The space between the primary septum and the endocardial cushions is the primary foramen.[7] The spina vestibula, the ridge to the right of the entrance of a single, primitive

Correspondence to: D. J. Patton, Department of Paediatric Cardiology, Alberta Children's Hospital, 2888 Shaganappi Trail NW, Calgary, Canada AB T3B 6A8. Tel: +1 (403) 955 7858; Fax: +1 (403) 955 7621; E-mail: David.Patton@albertahealthservices.ca

pulmonary vein, also grows upward to connect the primary septum with the endocardial cushions. Before complete fusion with the endocardial cushions, the septum primum undergoes apoptosis superiorly to form the foramen ovale. At the 12th week of development, a second infolding from the atrial roof, septum secundum, occurs to the right of the primary septum, and descends just past the upper edge of the primary septum.[7] This creates a one-way valve, allowing maintenance of the right-to-left shunt.

With postnatal respiration, a fall in pulmonary vascular resistance allows for increased pulmonary venous return, increasing the pressure in the left atrium relative to the right atrium so that the flap valve (upper edge of septum primum) closes against the septum secundum.[7] With time, fibrous tissue permanently seals the septal structures together.[1] A patent foramen ovale results if the two septal membranes overlap but are not adherent, allowing a transient right-to-left shunt when right atrial pressure exceeds that of the left atrium.[1,2] According to an autopsy series by Hagen et al,[8] a patent foramen ovale occurs in 25–30% of the population.

Distinct openings or holes allowing communication between the two atria are commonly known as atrial septal defects; however, it can be argued that not all are located within the true atrial septum. Recognising these distinctions as discussed below, we will use the terms atrial shunts, interatrial communication, and atrial septal defect throughout this review.[2] Atrial septal defects are recognised as the second most common congenital heart defect, with a reported incidence of 941 per million livebirths.[9] The true incidence is likely much higher, however, because many are diagnosed later in life.[2] Four types of atrial shunts other than a patent foramen ovale have been described based on their location relative to other cardiac structures.[1,10] These are illustrated in Figure 1.

The secundum atrial septal defect is the most common, accounting for 70% of all atrial shunts.[11] This defect occurs in the region of the oval fossa and results from the septum primum – the valve of the oval fossa – being absent, perforated, or too small to overlap the superior rim of the oval fossa, leaving the ostium secundum unguarded.[1] Roughly one quarter of patients with a secundum atrial septum defect also have a myxomatous abnormality of the mitral valve, which often results in mitral regurgitation.[12]

Ostium primum defects comprise 20% of atrial septal defects, and are located anterior to the fossa ovalis in the inferior part of the atrial septum, adjacent to the mitral and tricuspid valves.[13] These defects are often significant in size and result from persistence of the ostium primum. They are associated with abnormalities of the atrioventricular junction and valve, and hence are classified as

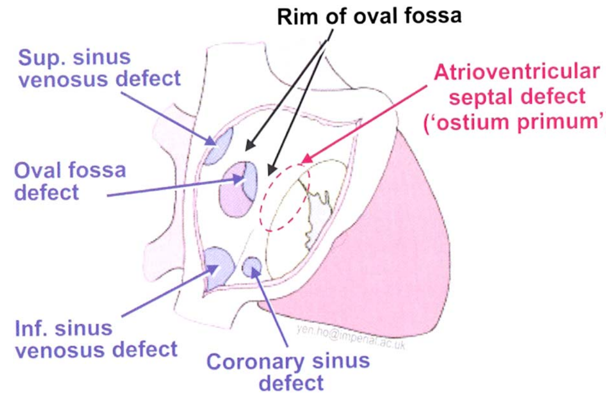

Figure 1.
Illustration of the various types of interatrial communications. The oval fossa defect referred to here is commonly known as a secundum atrial septal defect. (Reproduced with permission of Imperial College Press, publisher of Echocardiography in Congenital Heart Disease Made Simple).[10]

atrioventricular septal defects.[4,13] Evaluation and treatment of ostium primum defects will not be further discussed in this review.

The less common sinus venosus defect – 2–3% of all atrial shunts – embryologically arises from a defect between the left atrium and the superior, or rarely the inferior, caval vein.[3] Up to 85% of this type of defect are associated with partial anomalous pulmonary venous connections, where one or more of the pulmonary veins drain into the right atrium or the superior caval vein, rather than into the left atrium.[4] It is important to differentiate these defects from other atrial shunts as the majority of sinus venosus defects are not amenable to device closure and require surgical repair.

The most rare type of atrial shunt, a coronary sinus defect, occurs when an unroofed coronary sinus forms a shunt from the left atrium through the coronary sinus to the right atrium.[4] These are often associated with other defects such as left atrial connection of a persistent left superior caval vein, or, less commonly, pulmonary atresia.[10]

Pathophysiology and natural history of atrial shunts

Although interatrial communication is essential to normal foetal circulation, it does alter the path of blood flow required for postnatal physiology by allowing shunting between the normally separated pulmonary and systemic circulations. In both the normal foetal and the pathologic postnatal circumstance, the direction and magnitude of flow through the atrial septal defect is dependent on the pressure gradient between the right and left atria, and the area of the defect.[14] In turn, the pressure gradient between the atria is determined primarily by the compliance of each atrium and its associated ventricle.[14] Beyond the first few weeks of life, normal mean right atrial pressure ranges from 0 to 6 mmHg,[15] whereas left atrial pressure ranges from 4 to 12 mm of mercury.[15] Therefore, in any isolated atrial septal defect, a primary left-to-right shunt will develop. This leads to a portion of the pulmonary venous return being redirected back through the lungs, and thus an increase in flow in the pulmonary circulation with a correlated decrease in systemic flow.[2] The ratio of pulmonary to systemic flow is a reflection of the magnitude and direction of blood flow through the defect; values of 1.5:1 or greater have been used to define a haemodynamically significant atrial shunt.[16] Although shunts identified in adults are likely to remain patent, in young children it is common for an atrial septal defect to close spontaneously.[3]

Secondary to the left-to-right shunt, diastolic stress from increased right ventricular volume stimulates the ventricle to synthesise new myocardial fibres in series, which increases the end-diastolic volume and impairs function.[17] Further, the increased chamber size contributes to systolic stress, which stimulates the heart to produce new fibres in parallel, thus contributing to increased wall thickness and excessive right ventricular trabeculations.[17] Over time, dilation of the right atrium, right ventricle, the pulmonary artery, and its branches may result.[3] The tricuspid and pulmonary annuli may also become dilated and thickened, impairing their function.[1] Furthermore, ventricular interactions may occur, such that patients with right ventricular volume overload may also suffer from further decreased left ventricular output.[17] This phenomenon may be observed with cardiac imaging as diastolic septal flattening. Colan et al[17] have described immediate normalisation of left ventricular function following transcatheter closure of an atrial septal defect.

Atrial stretch is hypothesised to play a role in the development of arrhythmias. Dilation of the right atrium has been shown to predispose patients to atrial fibrillation and atrial flutter,[4] which often produce initial symptoms such as palpitations, leading to cardiac investigation.[18] Up to 45% of adults with unrepaired atrial septal defects have also been reported to have paroxysmal supraventricular tachycardia.[19]

Chronic volume overload of the pulmonary circulation can, over time, lead to pulmonary hypertension, defined as a mean pulmonary arterial pressure >25 mmHg at rest and 30 mmHg with exercise.[20] This may progress to an irreversible increase in pulmonary vascular resistance, characterised by intimal proliferation and fibrosis, leading to right ventricular failure, exercise intolerance, reduced quality of life, and death.[20] A 1968 study of the natural history of 128 patients with a secundum atrial defect reported development of pulmonary vascular disease to be the most serious risk.[21] A more recent study[11] reported that at the time of presentation with a secundum atrial septal defect, ~9% of patients have already developed pulmonary arterial hypertension. Cherian et al[22] reported pulmonary hypertension in 13% of patients under 10 years of age, and in 14% of patients in the age group of 11–20 years. Pulmonary arterial hypertension is three times more prevalent in patients with sinus venosus defects compared with those with secundum defects,[11] as most of these patients have additional shunting through partial anomalous pulmonary venous connections. Vogel et al[11] observed that pulmonary disease develops at a younger age in these patients, warranting additional monitoring and earlier treatment.

In the presence of pulmonary hypertension, elevated right ventricular pressure causes movement of the interventricular septum towards the left. This can be visualised as systolic septal flattening so that both ventricles appear D-shaped on a short-axis image. This process may further limit left ventricular filling.[1] Pulmonary pressures can eventually rise above systemic pressures, leading to reversal of the shunt.[3] Known as Eisenmenger syndrome, this condition is characterised by cyanosis with associated findings including clubbing, polycythaemia, and coagulopathy.[3] Cherian et al[22] identified Eisenmenger syndrome in 9% of their 709 consecutive atrial septal defect patients, with little difference between younger and older patients. Although the right-to-left shunt is pathological, it is generally agreed that these shunts should not be closed in the presence of irreversible pulmonary hypertension, as they minimise progression of pulmonary disease and may help maintain systemic cardiac output.[14]

Lastly, patients with atrial shunts may have a slightly increased risk for stroke because of the possibility of a transient right-to-left shunt allowing paradoxical embolism.[2] The need to repair the shunt or to anticoagulate these patients because of this risk remains controversial.

In summary, the course of patients with an unrepaired atrial shunt is extremely variable and is dependent on the age at presentation, the type and severity of the shunt, the ratio of pulmonary-to-systemic vascular resistance, and the compliance of the left and right ventricles.[1]

Clinical presentation and initial evaluation

Symptoms

While nearly always present from birth, atrial shunts may be diagnosed at any age as the severity of symptoms, which are often non-specific, vary greatly with the significance and progression of disease.[4] Most infants with atrial septal defects are asymptomatic, although mild transient cyanosis can occur in the newborn as a result of a right-to-left shunt.[14] Children and adults with a significant shunt might complain of fatigue, dypsnoea, or exercise intolerance.[2,4] They may also experience palpitations because of atrial fibrillation or flutter, but this typically does not occur in patients before the age of 40.[18]

Clinical examination

The heart rate, blood pressure, and respiratory rate are typically normal in a patient with an atrial shunt.[2] Jugular venous pressure may be normal;[2] however, prominence of the "v" wave in comparison with the "a" wave is sometimes apparent and is considered a valuable clinical sign of an atrial septal defect.[23,24] Older children and adults with large shunts might have a precordial bulge, a right ventricular heave, and/or a prominent systolic impulse indicating an enlarged right ventricle.[1,2]

The heart sounds of a patient with an atrial septal defect are rarely normal.[2] The first heart sound at the lower left sternal border is often accentuated. A fixed and widely split second heart sound has been described as the most valuable sign of a left-to-right interatrial shunt.[2] Closure of the pulmonary valve is delayed because of volume overload of the right heart, and the delay between aortic and pulmonary valve closure is fixed because the already volume loaded right heart is unable to further increase filling with inspiration. The murmur of an atrial septal defect is typically a soft crescendo–decrescendo systolic ejection murmur at the second left intercostal space reflecting increased volume passing through the pulmonary valve. There also might be a mid-diastolic murmur at the lower left sternal border, suggesting increased flow across the tricuspid valve.[1,2,14] Pulmonary hypertension, if it has developed, will result in a loud second pulmonary sound, elimination of the widely split second heart sound and the diastolic murmur, and a shorter systolic murmur.[1,4]

Chest radiography

Chest radiography is not indicated if an atrial shunt is suspected; however, in most patients with a significant atrial shunt, a chest radiograph will demonstrate an enlarged right heart and a dilated pulmonary trunk.[2] Increased pulmonary vascular markings suggest increased pulmonary blood flow, but decreased vascularity may be seen in the presence of pulmonary vascular disease.[25]

Electrocardiogram

Patients with an atrial shunt typically have a normal heart rate with normal sinus rhythm; however, in patients older than 40 years, atrial fibrillation or flutter is common.[2] An electrocardiogram may be useful in identifying the type of atrial shunt. For example, a leftward or left superior QRS axis suggests an ostium primum defect.[4] With most secundum defects, the axis will be normal or rightward (90–170°).[2] Inverted P waves in the inferior leads may indicate a sinus venosus defect.[4] Electrocardiography is also useful in detecting associated anomalies such as right ventricular hypertrophy, indicated by RSR′ pattern in the right precordial leads.[2,4,26] First-degree atrioventricular block is not uncommon in atrial septal defect patients, and a right bundle branch block is present in half of the patients over 60 years of age.[2]

Exercise testing

Many patients with atrial septal defects have reduced cardiopulmonary exercise capacity.[27,28] Assessment of a patient's maximal oxygen uptake (VO_2 max) and "oxygen pulse" – amount of oxygen consumed per heartbeat – compared with normal age- and sex-specific values contributes additional information for select patients in whom additional assessment of the haemodynamic significance of the shunt is required, or in following improvement of symptoms post repair.[27–29]

Advanced investigations and imaging

Before recommending treatment, diagnostic imaging is required to confirm the diagnosis, and to determine the shunt location, size, and haemodynamic significance, and to rule out associated anomalies.[2] According to European Society of Cardiology guidelines,[30] diagnostic work-up generally includes transthoracic echocardiography as initial screening, and trans-oesophogeal echocardiography to complete assessment. If this is insufficient to determine a management plan, then cardiovascular magnetic resonance imaging, computed tomography, and/or cardiac catheterisation may be required.

Echocardiography

Echocardiography has become the primary imaging technique for the evaluation of atrial septal defects. Right ventricular size and function can be assessed, and pulmonary arterial pressure and shunt fraction can also be estimated with transthoracic echocardiography using Doppler imaging. In children, the subcostal four-chamber view has been reported as the most diagnostic of atrial shunts (Fig 2a and b).[10] Unfortunately, in adults this view is typically too far from the heart and is not useful. Overall, the limited acoustic windows of transthoracic echocardiography do not allow complete visualisation of the atrial septum or other posterior cardiac structures, and further assessment is typically required in many older children and most adults before recommending treatment.[31,32]

Trans-oesophageal echocardiography is currently the modality of choice to evaluate an atrial shunt, as it provides an excellent view of the interatrial septum (Fig 2c) and often demonstrates pulmonary venous anatomy.[31,32] It is considered important for characterisation of an atrial shunt and for determining the best course of treatment, as well as to guide percutaneous closure of secundum defects.[33] In addition to assessment of the diameter of the defect, multi-planar trans-oesophageal echocardiography is used to assess the rims of septum surrounding the defect. The superior and inferior vena caval rims are imaged in the bicaval view, the atrioventricular valve rim in a four-chamber view, and retro-aortic rim in a short-axis plane.[33] A deficient (<5 mm) rim to the

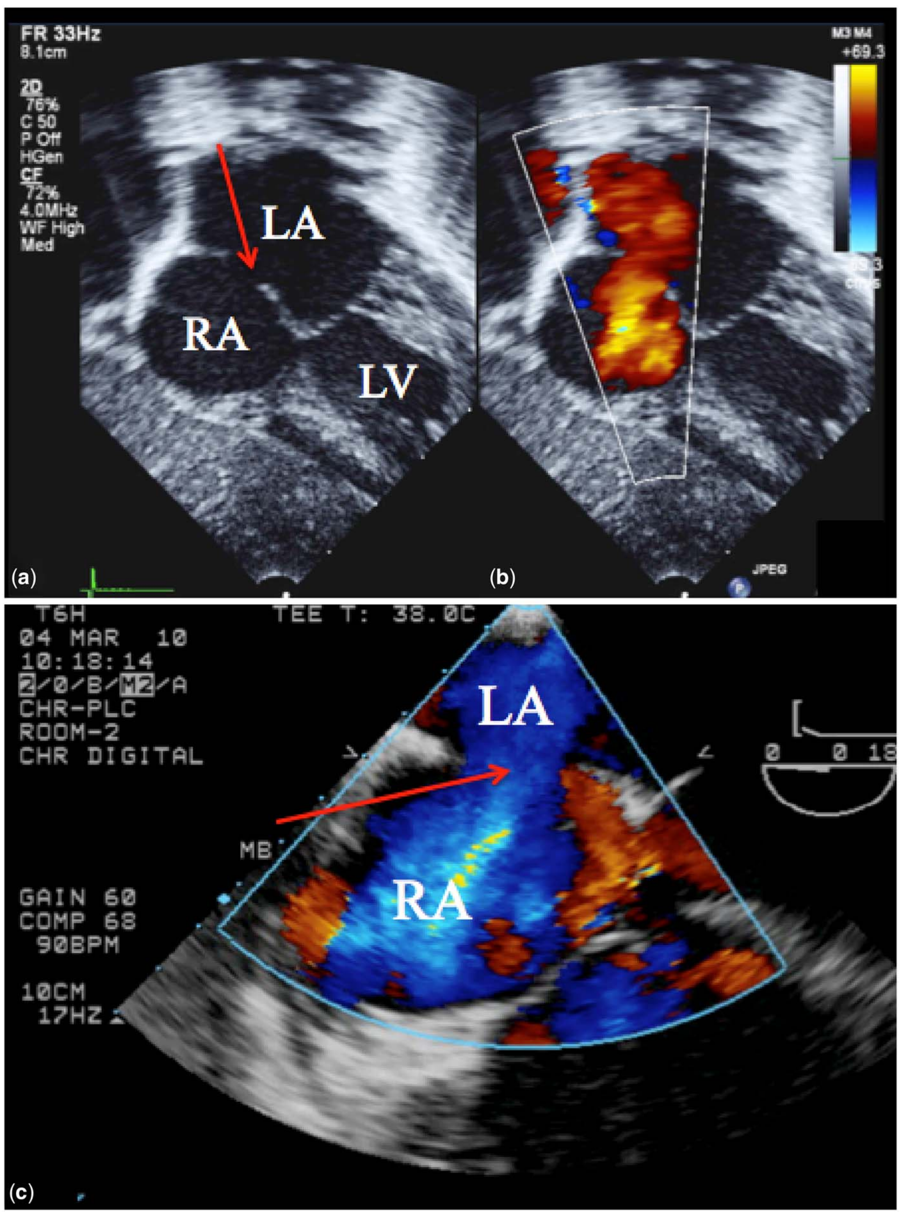

Figure 2.

Echocardiographic images of atrial shunts. (a, b) Transthoracic subcostal view of a 9-day-old infant with a large secundum atrial septal defect (arrow) with left-to-right shunt. (c) Trans-oesophageal echocardiography demonstrating a large atrial shunt (arrow) in a 28-year-old female. LA = left atrium; LV = left ventricle; RA = right atrium.

https://doi.org/10.1017/S1047951113002321 Published online by Cambridge University Press

atrioventricular valve, superior or inferior vena cava, or right upper pulmonary vein typically precludes device closure, whereas a deficient retro-aortic rim in multiple sequential views requires careful consideration of the choice of septal occlusion device versus surgical repair.[33] Visualisation of all pulmonary veins and any sinus venosus defects remains especially challenging.[13] Furthermore, the procedure can be uncomfortable for the patient, is associated with a small risk of oesophageal trauma, and requires sedation and a topical anesthetic, or general anesthesia in children.[5]

Saric et al[34] have demonstrated that three-dimensional trans-oesophageal echocardiography acquires accurate views of the atrial septum, with an additional advantage that it can provide an "en face" (in plane) view of the atrial septum. Signal dropout within the atrial septum often occurs in normal patients with three-dimensional trans-oesophageal imaging, decreasing the specificity of this technique. The inferior rim of the IVC is also not easily visualised in many patients.[34] Despite these limitations, three-dimensional trans-oesophageal echocardiography has also been shown to successfully guide percutaneous closure of atrial shunts.[35]

Intracardiac echocardiography has also been utilised in some centres for imaging atrial shunts.[36] It is not yet recognised as a routine diagnostic tool, but has proven useful in guiding transcatheter closure,[36,37] and was reported to be more comfortable for the patient than trans-oesophageal echocardiography.[38] In the axial plane of the aortic valve, the atrial septum is well seen and, according to Onorato et al,[39] the dimensions and rims of an atrial septal defect may be measured accurately. A view in the axial plane of the junction between the superior caval vein and the right atrium provides easy identification of any anomalous pulmonary veins.[39]

Catheterisation and angiography

Cardiac catheterisation has historically been recognised as among the most accurate methods of quantifying shunts and measuring pressure.[2] However, because of its invasive nature, this technique is now reserved for immediately before percutaneous closure.[40] If the pulmonary arterial pressure as estimated by echocardiography is >50% of the systemic pressure, catheterisation is indicated to assess for pulmonary hypertension.[41]

While mean right atrial pressure is typically normal in the presence of an atrial septal defect, the "v" and "a" waves are often equal.[2,24] Right ventricular and pulmonary artery pressures are also usually normal unless the defect is very large. Pulmonary arterial pressures have been observed to increase with age and shunt severity,[2] and the gradient across the pulmonary valve is increased in most atrial septal defect patients.[14] The pressure gradient between the right and left atrium is usually small, particularly in large shunts.[14,41]

Invasive oximetry measures blood oxygen saturation in various cardiac chambers. An atrial shunt is considered present when the oxygen saturation in the right atrium is at least 5% higher than that in the superior caval vein in the absence of other anomalies.[2] Applying the Fick principle allows for estimation of the shunt ratio (pulmonary flow: aortic flow). Pulmonary flow can be calculated as the oxygen consumption (ml/min) divided by the arteriovenous oxygen content difference across the lungs (ml/L), whereas aortic flow is equal to oxygen consumption (ml/min) divided by the arteriovenous oxygen content difference across the body (ml/L).[42]

Being among the oldest techniques applied in cardiac shunt detection, invasive oximetry is often used as a standard to validate newer shunt quantification techniques. However, Boehrer et al[42] argue that oximetry is unable to detect small inter-atrial shunts and provides only an estimate of shunt fraction.

Although rarely used now, the indicator dilution technique is of historical interest. It involves intravenous injection of a bolus of indicator dye, most often indocyanine green, and withdrawal of blood from the arterial circulation in order to measure the indicator concentration. Concentration of the indicator is plotted against time, and demonstrates a large primary peak followed by a normal recirculation peak in patients without a left-to-right shunt. If a left-to-right shunt is present, the primary peak will be of lesser amplitude, and a prominent early recirculation peak will be present because of blood with indicator bypassing the pulmonary circulation to return directly to the systemic circulation.[25,40,42] This technique is useful for calculating a pulmonary-to-systemic flow ratio, and has been shown to correlate well with values obtained from oximetry,[43] although oximetry provides slightly higher shunt ratio values than does the indicator dilution technique.[25] Its sensitivity in detecting small shunts is slightly better than oximetry, detecting a shunt ratio as small as 1.35; however, it fails to provide any information on shunt location.[42]

According to Boehrer et al,[42] contrast angiography is useful for detecting cardiac shunts such as ventricular septal defects, but is often insensitive to detection and quantification of atrial shunts. Nevertheless, it can be useful for depiction of pulmonary artery and pulmonary vein anatomy, as imaged from the antero-posterior or lateral views.[2]

X-Ray computed tomography

In general, cardiac computed tomography is known to provide information complementary to echocardiography or magnetic resonance imaging. Its excellent spatial resolution makes it particularly useful in detecting associated anomalies and assessing changes in the pulmonary vasculature.[13] Williamson et al[44] recently applied an electrocardiography-gated cardiac computed tomography angiography technique with a saline chaser to 20 patients, and found that computed tomography has high sensitivity in detecting a patent foramen ovale based on the presence of an atrial septal flap, a continuous contrast column between the atria, and a jet of contrast into the right atrium.

Current applications of magnetic resonance imaging in atrial shunt assessment

Cardiovascular magnetic resonance imaging can provide a valuable assessment of cardiac anatomy, function, blood flow, and tissue characteristics.[45] Furthermore, it is non-invasive and applies no ionising radiation.[4] Several cardiac magnetic resonance imaging techniques have been shown to be valuable in assessing atrial shunts.[46,47]

Steady-state free precession cine imaging

Steady-state free precession is a fast gradient echo pulse sequence used to acquire cine images during a single breath hold.[48] This sequence is excellent for determining diastolic and systolic ventricular volumes as shown in Figure 3, and for calculating ejection fraction; therefore, it is a fundamental sequence used in nearly every cardiac magnetic resonance imaging protocol.[49] If an intracardiac shunt is present, it can be quantified by calculating the biventricular stroke volume ratio, which should be equal to the ratio of pulmonary arterial flow to aortic flow (Qp/Qs) in a patient with no other cardiac shunt. The net shunt flow may be calculated as the difference between the left and right ventricular stroke volumes.[50]

In some cases, an interrupted septum may be seen in the four-chamber view and short-axis atrial stack cines (Fig 4), and flow may be visible across the defect.[47,51] A recently published study demonstrated cardiac magnetic resonance imaging to have excellent assessment of size, location, and septal rims compared with trans-oesophageal echocardiography in 20 patients using solely cardiac-gated steady-state free precession sequences.[47] The maximum and minimum size of the defect measured by steady-state free precession correlated well with trans-oesopheal echocardiography ($r = 0.87$ and 0.92), but had a weaker correlation with size of the device used in percutaneous closure ($r = 0.53$ and 0.57 for maximum and minimum size, respectively).[47] Despite these results, experts maintain that steady-state free precession images have low specificity in atrial shunt detection, and do not provide accurate defect size.[52]

Gradient-recalled echo cines with saturation band

In this technique, a pre-saturation radiofrequency pulse is applied before each image acquisition, which produces saturation of spins – preventing any signal – in a specific area or band.[52] For assessment of an atrial shunt, the saturation band is placed on either the left or the right atrium, parallel with the atrial septum. Flow through a shunt is clearly visualised by the movement of dark or bright blood into the opposing atrium, as seen in Figure 5.[52] The dimensions of the defect can be measured in both the four-chamber and

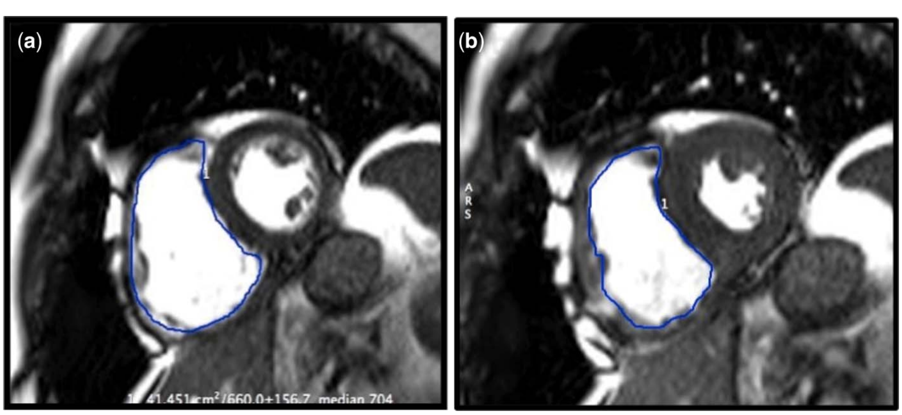

Figure 3.
One slice of a stack of ventricular short-axis steady-state free precession images in diastole (a) and systole (b) demonstrating contouring of the right ventricle in assessment of stroke volume.

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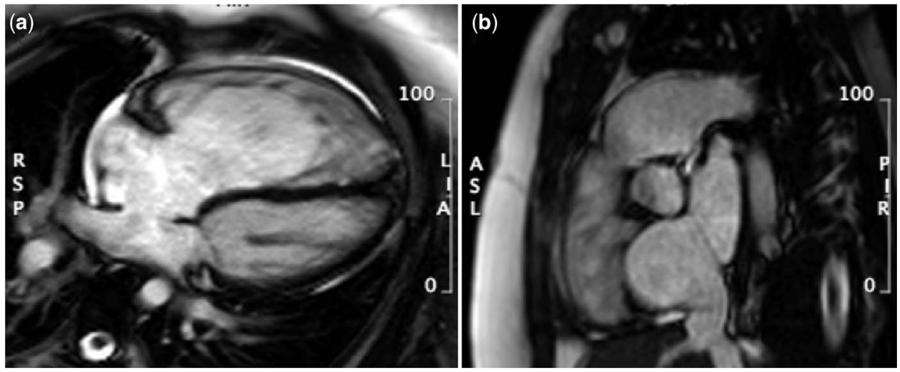

Figure 4.
Still frames of steady-state free precession MRI cines at 1.5 Tesla depicting an atrial septal defect in (a) a four-chamber view and (b) atrial short-axis view. An interrupted atrial septum and dephasing of blood is obvious in the four-chamber view, but not in the short-axis view. MRI = magnetic resonance imaging.

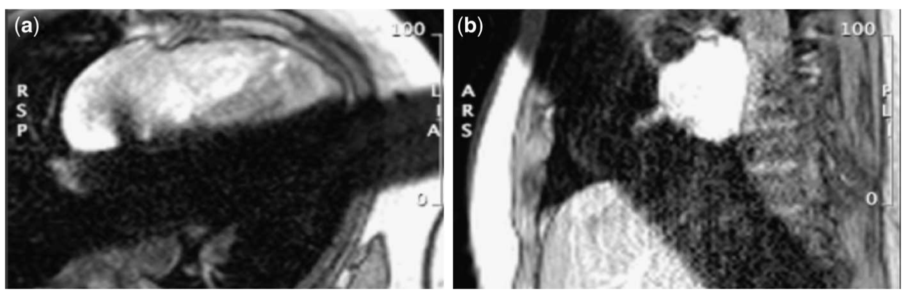

Figure 5.
Gradient-recalled echo with saturation band method shows a small shunt in four-chamber (a) and atrial short-axis (b) views.

atrial short-axis views to provide an estimate of area. Saturation bands could potentially be more sensitive than trans-oesophageal echocardiography in detecting unusually located atrial shunts.

First-pass gadolinium perfusion

Originally used for detection of coronary artery disease, first-pass gadolinium perfusion has been applied to dynamic imaging of atrial shunts.[51] A bolus of gadolinium contrast is injected intravenously and the atria and pulmonary veins are imaged rapidly using a fast gradient echo sequence. In the presence of an atrial septal defect with left-to-right shunting, dark blood can be seen entering the right atrium when it is filled with contrast, and bright blood is seen re-circulating into the right atrium from the contrast-filled left atrium (Fig 6). If contrast appears in the left atrium after the right atrium, but before the pulmonary veins, then a right-to-left shunt is present.[51] Compared with trans-oesophageal echocardiography in Hamilton-Craig's study, first-pass perfusion is less sensitive in the detection of a patent foramen ovale and tends to underestimate the size of the shunt.[51]

Phase contrast velocity

This sequence makes use of motion-induced phase shift to quantify velocity and flow either through-plane or in-plane.[53] In atrial shunts, velocity and flow are routinely quantified through the proximal ascending aorta and the main pulmonary artery in order to calculate the shunt ratio (Qp/Qs).[13,40,54] Accuracy is highly dependent on technical expertise: phase contrast velocity requires careful planning to position the imaging plane, select appropriate spatial and temporal resolution, and post-processing also requires attention to detail. Turbulence, partial volume effects, and phase-offset or background errors can contribute to inaccuracies,[55] some of which can be corrected with the use of a phantom, or by measuring the phase-offset error in a region of no flow near the area of interest, typically the chest wall.[56] Aortic and main pulmonary artery flow assessment may have additional errors because of through-plane motion of the annulus, impact of vessel compliance, and complex flow at the coronary ostia.[55] Internal validation is possible by verifying that flow through the main pulmonary artery is equal to the either the sum of the flow through the left and right pulmonary

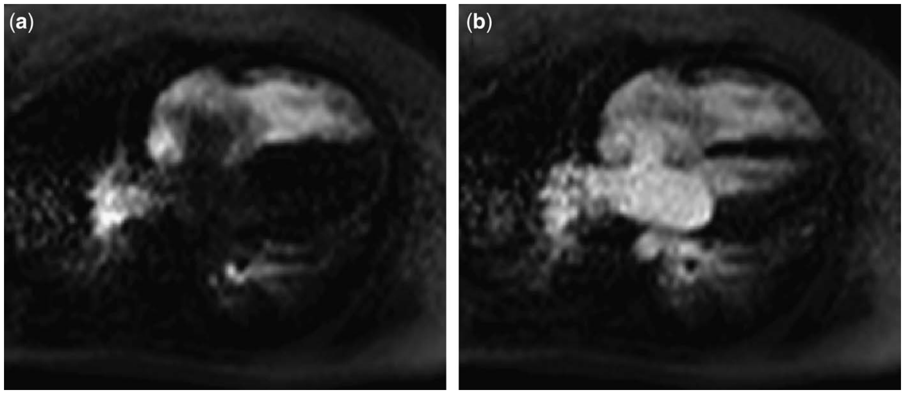

Figure 6.
Still frames of perfusion imaging clearly demonstrate a large left-to-right shunt. (a) Blood without contrast from the left atrium is seen entering the right atrium, which is filled with contrast. (b) When the contrast has entered the left atrium, it is seen recirculating into the right atrium.

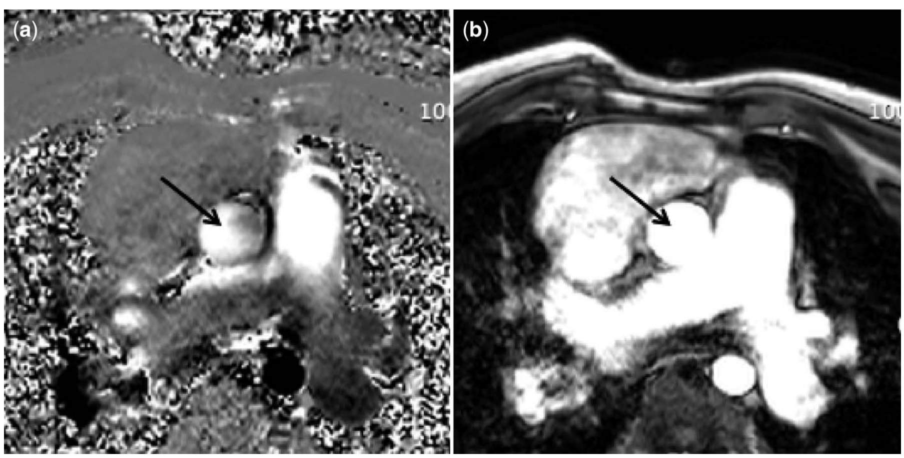

Figure 7.
Phase contrast velocity (a) and magnitude image (b) of the aorta in short-axis or through-plane view (arrow).

arteries or that pulmonary arterial flow is equal to pulmonary venous return.[40,50,55] Beerbaum et al[54,57] demonstrated that shunt ratio determination by phase contrast velocity in children is as reliable as invasive oximetry. Figure 7 demonstrates a phase contrast and a magnitude image of the aorta in through-plane view.

Phase contrast velocity can also be applied to measure flow directly through an atrial septal defect,[58] as shown in Figure 8. Thomson et al[53] recently investigated the accuracy of this method in flow quantification of secundum defects. The atrial shunt ratio was calculated in three ways: the sum of systemic and shunt flow divided by systemic flow; pulmonary flow divided by the sum of pulmonary and shunt flow; and the pulmonary-to-systemic flow ratio. They found the first shunt ratio to correlate best with oximetry results ($r = 0.89$), compared with the latter calculations ($r = 0.77$ and 0.74, respectively), suggesting that direct en face imaging of atrial shunts may provide a more accurate hemodynamic assessment.[53]

En face flow also demonstrates the shape and size of an atrial septal defect. Phase contrast cine images have been shown to be more accurate in evaluating the size and shape of atrial septal defects than both trans-oesophageal and intracardiac echocardiography.[5,53,59] In a study of 30 patients with atrial sepal defects, Holmvang et al[59] observed that phase-contrast cine images were adequate to define the defect's shape and size. The maximum diameter by phase contrast magnetic resonance imaging correlated closely with balloon sizing measurements ($r = 0.75$) and with templates cut during surgery ($r = 0.93$).[59] In 32 patients who underwent Amplatzer device closure, Thomson et al[53] found that the area of the atrial shunt measured by phase contrast velocity correlated with the size of the deployed Amplatzer device better than intracardiac echocardiography, especially in patients who received small- to medium-sized devices – area <3 cm^2. According to Colletti et al[50] this is the best method to determine the size and directionality of intracardiac shunts. A study of 54 children who underwent catheter closure with the Amplatzer septal occluder demonstrated that cardiac magnetic resonance imaging was more accurate at calculating atrial septal defect diameter relative to balloon sizing in comparison to trans-oesophageal echocardiography.[36] Finally, rim measurements can be measured with this technique, which is essential in determining whether percutaneous closure is possible.[52,58] Rim assessment using this technique is limited outside of major centres; therefore, trans-oesophageal echocardiography remains the gold standard for rim assessment.

Four-dimensional flow is a three-dimensional phase contrast velocity sequence that acquires the blood flow patterns in the entire thoracic cavity in a single acquisition, and offers particle tracing. This allows for quantification of flow through the aorta, pulmonary artery, and atrial septal defect with simplified planning, reducing overall time required and potentially increasing accuracy.[60] Valverde et al[60] highlighted these advantages in an 8 year old with a secundum atrial septal defect and anomalous pulmonary veins, which were not seen on transthoracic echocardiography. At this time, four-dimensional flow is a promising sequence, but is time-consuming and not used in most protocols for assessment of atrial shunts.

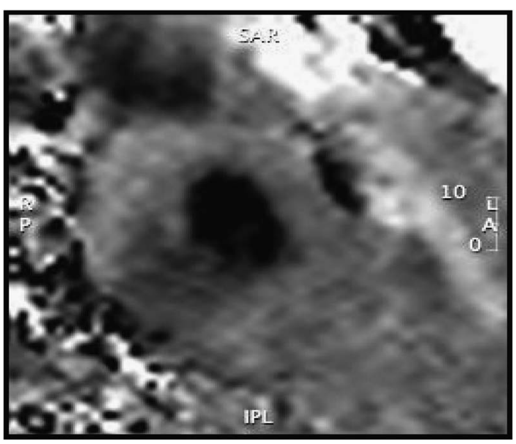

Figure 8.
Phase contrast velocity image of an atrial septal defect en face. The area was 179 mm^2.

Magnetic resonance angiography

One of the main advantages of cardiac magnetic resonance imaging over other imaging modalities is its excellent delineation of anatomy of the great arteries and pulmonary veins,[61] as seen in Figure 9, using gadolinium contrast angiography, without exposing the patient to ionising radiation.[52] This is important for detection of anomalous pulmonary venous connections, which, if present, may preclude device closure.[62]

Current time, cost, and limitations

Having multiple sequences that can acquire the same information is a major strength of cardiac magnetic resonance imaging, providing internal validation, unlike other imaging modalities.[63] The time required to complete all of the above sequences applied in atrial shunt evaluation exceeds 1 hour, although imaging time may be reduced with judicious selection of sequences. Of note, gadolinium contrast, required for first-pass perfusion and magnetic resonance imaging angiography, is contraindicated in patients with renal

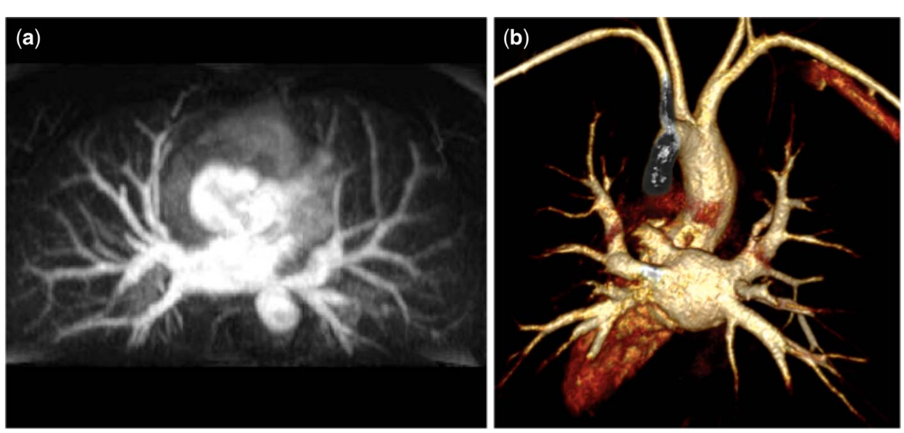

Figure 9.
Thick maximum intensity projection of pulmonary vein angiogram, transverse orientation (a). Volume-rendered display of pulmonary vein angiogram, posterior orientation (b).

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failure because of an association with nephrogenic systemic fibrosis, a rare but devastating and often fatal disease.[64] Contrast is also contraindicated in patients who are pregnant or breastfeeding. Magnetic resonance imaging should not be performed in pregnant women,[65] patients with intracardiac electronic devices,[66] or extreme claustrophobia unless the benefits are deemed to outweigh the risks.

Treatment

Progression of symptoms and adverse pathophysiology can be prevented with closure of the interatrial communication, either by open-heart surgery or percutaneously with the use of a device.[67] Before recommending a treatment, a cardiologist will need to consider the patient's age and symptoms, the shunt ratio – pulmonary artery to aortic flow – the size, shape, and location of the defect, the adequacy of the septal rims, the proximity of the defect to other cardiac structures, the pulmonary vascular resistance, and associated defects.[3]

Repair of an atrial shunt is indicated if the shunt ratio is >1.5:1, or if the defect is at least 10 mm in diameter in adults, regardless of the symptoms of the patient.[4,67,68] One exception to this rule is if the patient is <2 years of age, as there is a high probability of spontaneous closure.[2] As previously indicated, these values have been recognised as causative of right heart dilation, increased pulmonary pressures, and other adverse outcomes of a chronic left-to-right shunt.[67] Repair is generally not recommended in patients with small atrial shunts; however, these patients should be monitored for development of right heart dilation or symptoms, and then closure should be considered.[2,4]

Repair of an atrial shunt is contraindicated if pulmonary hypertension has already developed (greater than eight Woods units), or if right ventricular function is severely compromised.[69] In these cases, closing the shunt is likely to worsen the patient's clinical outcome. Closure in pregnant women is typically delayed until 6 months following delivery.[4]

Anticipated benefits from atrial septal defect closure include improved exercise capacity and quality of life, prevention of pulmonary hypertension and right heart failure, and reduced risk of stroke and arrhythmias.[67] Remodelling of the right ventricle and right atrium to normal size has been demonstrated in treated patients. However, remodelling is often incomplete in older patients, with 26–29% of patients having persisting right ventricular dilation 1 year after atrial septal defect repair.[19] The prevalence of atrial fibrillation is reduced after surgical repair, especially in younger patients. Of 192 patients who had surgical repair of atrial shunt, the age of those with persistent atrial fibrillation was 47.7 ± 19 years compared with 22.9 years for resolved atrial fibrillation.[70]

Percutaneous closure

Transcatheter closure of an atrial septal defect was first performed in 1976.[71] The initial devices were not easy to place accurately within the defect and some had a high fracture rate.[2] Since the development of new occluders such as the Amplatzer device (Fig 10), percutaneous closure of atrial septal defects has significantly increased. Device closure is now as effective as surgery and the primary method of choice. Benefits include no thoracotomy, a shorter stay in hospital, less pain and discomfort to the patient, and lower cost.[2]

The primary contraindications to percutaenous closure of the atrial shunt include: too large a defect – diameter >36 mm, or if the device required is too large for the patient – inadequate septal tissue,[33] or if the defect is too close to other cardiac structures, such as the atrioventricular valves, upper pulmonary veins, or the coronary sinus. In addition, percutaneous

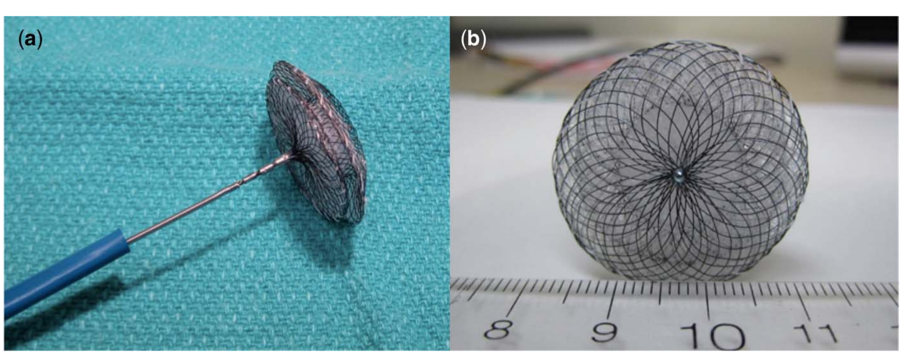

Figure 10.
Photographs of an Amplatzer Atrial Septal Occluder device. (a) View from the side demonstrating two discs connected by a thinner waist. (b) View of the device from the front demonstrating its diameter.

https://doi.org/10.1017/S1047951113002321 Published online by Cambridge University Press

closure is not recommended if the femoral veins are too small to allow access, or if the patient has intracardiac thrombi, other cardiac defects, bleeding disorders, any infection, or a contraindication to aspirin or other anticoagulants.[4]

The procedure is performed with the patient under general anaesthesia or local anaesthesia with sedation.[2] A catheter is guided into the right heart[69] where pulmonary artery pressure and pulmonary vascular resistance are determined. A sizing balloon is used to acquire the "balloon size", defined as the diameter of the balloon as measured by echocardiography when complete obstruction of Doppler flow is achieved.[33,69] Next, a delivery sheath for the device is advanced into the left atrium and the occluding device is passed through the sheath.[72] The Amplatzer septal occluder, which consists of two attached discs made of a polyester fabric encased by a woven wire mesh of nickel-titanium, is then extruded from the catheter, so that a disc resides on either side of the septum.[2,69,72,73] Alternatively, a GORE HELEX septal occluder (W.L. Gore and Associates, Flagstaff, Arizona, USA) may be used, which is a nickel-titanium wire covered with Gore-Tex.[72,73] Appropriate positioning of the device can be verified using fluoroscopy, trans-oesophageal or intracardiac echocardiography, and angiography before deployment.[69]

A known complication of percutaneous atrial shunt repair is erosion of the device through adjacent cardiac structures. To avoid this complication, it is important to assess the retro-aortic rim of the defect carefully, and to ensure the device is not significantly larger than the defect.[2] Other possible complications include embolisation of the device, tamponade from cardiac trauma, thrombosis, and arrhythmias.[2]

Surgical closure

If transcatheter atrial shunt closure is contraindicated, or if the patient prefers surgical repair, it may be necessary to perform open-heart surgery.[2] Atrial septal defects are often closed directly with sutures, or with a patch of either pericardium or synthetic material. Associated defects, if present, are usually repaired at the same time.[4] This surgery has been performed since the 1950s and now has an excellent success rate and near-zero mortality.[2,74]

Post-operative complications are rare, but can include transient or chronic arrhythmias – sinus node dysfunction, supraventricular tachycardia – or post-pericardiotomy syndrome – an inflammatory reaction.[2] Obstruction of the superior vena cava or a right-sided anomalous vein is possible following repair of a sinus venosus defect.[2]

Post-closure complications

After either method of atrial septal defect repair, ongoing concerns include mitral or tricuspid regurgitation, persistent right ventricular and atrial dilation – more commonly in older patients or those with increased pulmonary vascular resistance – and complications such as atrial fibrillation, thrombus, and sinus and/or atrioventricular node dysfunction.[3,18,75] Rarely, Budd–Chiari syndrome – occlusion of the hepatic veins – may develop.[3] Bacterial endocarditis is very rare, but cases have been reported following device closure – one at 8 weeks after the procedure and the other at 10–14 weeks after repair. Antibiotic prophylaxis is recommended for at least 6 months following repair, particularly in patients who underwent percutaneous repair.[3]

Summary and recommendations

Atrial shunts are a relatively common congenital heart defect that may lead to adverse effects such as right heart dilation, atrial arrhythmias, and pulmonary arterial hypertension. Symptoms including shortness of breath, reduced exercise tolerance, and palpitations may occur. Haemodynamically significant shunts can be repaired by percutaneous closure with an occluder, or by open-heart surgery, depending on the characteristics of the patient and of the atrial septal defect. Before a decision on treatment, a shunt must first be evaluated with advanced diagnostic imaging, and closure should be performed only after weighing the potential risks and benefits to the individual.

Transthoracic echocardiography is used as a first-line test, particularly in children, to diagnose an atrial shunt, and trans-oesophageal echocardiography is currently the primary imaging modality in its confirmation and assessment. Cardiac magnetic resonance imaging is currently used to provide additional information in unclear situations; however, evidence suggests that it approaches the sensitivity, specificity, and sizing accuracy of trans-oesophageal echocardiography with the added advantage of being non-invasive. Further research may refine the cardiac magnetic resonance imaging protocol to optimise evaluation of atrial shunts, applying only the most accurate and fastest sequences, so that it may replace trans-oesophageal echocardiography in the future.

Acknowledgements

The authors wish to acknowledge the Alberta Children's Hospital Research Institute and the Stephenson Cardiovascular Magnetic Resonance Imaging Centre for their support of this work.

References

1. Porter CEW. Atrial septal defects. In: Allen HD, Clark EB, Gutgesell HP, Driscoll DJ (eds). Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adults, 7th edn. Lippincott Williams & Wilkins, Philadelphia, PA, 2008, pp 632–645.
2. English R, Anderson R, Ettedgui J. Interatrial communications. In: Anderson RH, Baker EJ, Penny DJ, Redington AN, Rigby ML, Wernovsky G (eds). Pediatric Cardiology, 3rd edn. Churchill Livingstone, Elsevier, Philadelphia, PA, 2010, pp 523–546.
3. Veldtman GR, Freedom RM, Benson LN. Atrial septal defect. In: Freedom R, Yoo S, Mikailian H, Williams WG (eds). The natural and modified history of congenital heart disease. Wiley-Blackwell, Elmsford, NY, 2003, pp 31–43.
4. Webb G, Gatzoulis MA. Atrial septal defects in the adult: recent progress and overview. Circulation 2006; 114: 1645–1653.
5. Piaw CS, Kiam OT, Rapaee A, et al. Use of non-invasive phase contrast magnetic resonance imaging for estimation of atrial septal defect size and morphology: a comparison with transesophageal echo. Cardiovasc Intervent Radiol 2006; 29: 230–234.
6. Whitaker KB. Embryologic development of the cardiopulmonary system. In: Whitaker KB (ed). Comprehensive Perinatal and Pediatric Respiratory Care. Delmar, Thompson Learning, Albany, NY, 2001, pp 18–20.
7. Anderson RH, Brown NA, Webb S. Development and structure of the atrial septum. Heart 2002; 88: 104–110.
8. Hagen PT, Scholz DG, Edwards WD. Incidence and size of patent foramen ovale during the first 10 decades of life: an autopsy study of 965 normal hearts. Mayo Clin Proc 1984; 59: 17–20.
9. Hoffman JI, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol 2002; 39: 1890–1900.
10. Ho SY, Rigby ML, Anderson RH. Interatrial communications. In: Ho SY, Rigby ML, Anderson RH (eds). Echocardiography in Congenital Heart Disease Made Simple, 1st edn. Imperial College Press, London, 2005, pp 67–75.
11. Vogel M, Berger F, Kramer A, Alexi-Meshkishvili V, Lange PE. Incidence of secondary pulmonary hypertension in adults with atrial septal or sinus venosus defects. Heart 1999; 82: 30–33.
12. Joy J, Kartha CC, Balakrishnan KG. Structural basis for mitral valve dysfunction associated with ostium secundum atrial septal defects. Cardiology 1993; 82: 409–414.
13. Hoey ET, Gopalan D, Ganesh V, Agrawal SK, Screaton NJ. Atrial septal defects: magnetic resonance and computed tomography appearances. J Med Imaging Radiat Oncol 2009; 53: 261–270.
14. Vick GW. Defects of the atrial septum including atrioventricular septal defects. In: Garson A, Bricker JT, Fisher DJ, Neish SR (eds). The Science and Practice of Pediatric Cardiology, 2nd edn. Williams & Wilkins, Baltimore, Maryland, 1998, pp 1142–1177.
15. Bustin D. Hemodynamic Monitoring for Critical Care. Appleton-Century-Crofts, Norwalk, Connecticut, 1986.
16. Sanders SP, Yeager S, Williams RG. Measurement of systemic and pulmonary blood flow and QP/QS ratio using Doppler and two-dimensional echocardiography. Am J Cardiol 1983; 51: 952–956.
17. Colan SD. Ventricular function in volume overload lesions. In: Fogel M (ed.). Ventricular Function and Blood Flow in Congenital Heart Disease. Blackwell, Malder, MA, 2005, pp 205–221.
18. Gatzoulis MA, Freeman MA, Siu SC, Webb GD, Harris L. Atrial arrhythmia after surgical closure of atrial septal defects in adults. N Engl J Med 1999; 340: 839–846.
19. Meijboom F, Hess J, Szatmari A, et al. Long-term follow-up (9 to 20 years) after surgical closure of atrial septal defect at a young age. Am J Cardiol 1993; 72: 1431–1434.
20. Haworth SG, Rabinovitch M. Pulmonary circulation. In: Anderson RH, Baker EJ, Penny DJ, Redington AN, Rigby ML, Wernovsky G (eds). Paediatric Cardiology, 3rd edn. Churchill Livingstone, Elsevier, Philadelphia, PA, 2010, pp 117–141.
21. Craig RJ, Selzer A. Natural history and prognosis of atrial septal defect. Circulation 1968; 37: 805–815.
22. Cherian G, Uthaman CB, Durairaj M, et al. Pulmonary hypertension in isolated secundum atrial septal defect: high frequency in young patients. Am Heart J 1983; 105: 952–957.
23. Tavel ME, Stewart J. Usefulness and limitations of precordial phonocardiography and external pulse recordings. Cardiovasc Clin 1975; 6: 41–55.
24. Parikh DN, Fisher J, Moses JW, et al. Determinants and importance of atrial pressure morphology in atrial septal defect. Br Heart J 1984; 51: 473–479.
25. Rees A, Farru O, Rodriguez R. Phonocardiographic radiological, and haemodynamic correlation in atrial septal defect. Br Heart J 1972; 34: 781–786.
26. Cohen JS, Patton DJ, Giuffre RM. The crochetage pattern in electrocardiograms of pediatric atrial septal defect patients. Can J Cardiol 2000; 16: 1241–1247.
27. Giardini A, Donti A, Formigari R, et al. Determinants of cardiopulmonary functional improvement after transcatheter atrial septal defect closure in asymptomatic adults. J Am Coll Cardiol 2004; 43: 1886–1891.
28. Suchon E, Podolec P, Tomkiewicz-Pajak L, et al. Cardiopulmonary exercise capacity in adult patients with atrial septal defect. Przegl Lek 2002; 59: 747–751.
29. Radojevic J, Rigby M. Atrial septal defect, Chapter 25. In: Gatzoulis MA, Webb GD, Daubeney PEF (eds). Diagnosis and Management of Adult Congenital Heart Disease, 2nd edn. Elsevier Saunders, Philadelphia, 2011, pp 183–184.
30. Baumgartner H, Bonhoeffer P, De Groot NM, et al. Task Force on the Management of Grown-up Congenital Heart Disease of the European Society of Cardiology (ESC); Association for European Paediatric Cardiology (AEPC); ESC Committee for Practice Guidelines (CPG). ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010; 31: 2915–2957.
31. Gnanapragasam JP, Houston AB, Northridge DB, Jamieson MP, Pollock JC. Transoesophageal echocardiographic assessment of primum, secundum and sinus venosus atrial septal defects. Int J Cardiol 1991; 31: 167–174.
32. Fischer G, Stieh J, Uebing A, Hoffmann U, Morf G, Kramer HH. Experience with transcatheter closure of secundum atrial septal defects using the Amplatzer septal occluder: a single centre study in 236 consecutive patients. Heart 2003; 89: 199–204.
33. Lerakis S, Babaliaros VC. Transesophageal echocardiography guided percutaneous closure of secundum atrial septal defects. Echocardiography 2010; 27: 724–727.
34. Saric M, Perk G, Purgess JR, Kronzon I. Imaging atrial septal defects by real-time three-dimensional transesophageal echocardiography: step-by-step approach. J Am Soc Echocardiogr 2010; 23: 1128–1135.
35. Lodato JA, Cao QL, Weinert L, et al. Feasibility of real-time three-dimensional transoesophageal echocardiography for guidance of percutaneous atrial septal defect closure. Eur J Echocardiogr 2009; 10: 543–548.
36. Durongpisitkul K, Tang NL, Soongswang J, Laohaprasitiporn D, Nanal A. Predictors of successful transcatheter closure of atrial septal defect by cardiac magnetic resonance imaging. Pediatr Cardiol 2004; 25: 124–130.
37. Zanchetta M, Rigatelli G, Pedon L, Zennaro M, Carrozza A, Onorato E. Catheter closure of perforated secundum atrial septal defect under intracardiac echocardiographic guidance using a single Amplatzer device: feasibility of a new method. J Invasive Cardiol 2005; 17: 262–265.
38. Hernandez F, Garcia-Tejada J, Velazquez M, Albarran A, Andreu J, Tascon J. Intracardiac echocardiography and percutaneous closure of atrial septal defects in adults. Rev Esp Cardiol 2008; 61: 465–470.

39. Onorato E, Casilli F, Zanchetta M. Intracardiac echocardiography by ultra ICE. In: Sievert H, Qureshi S, Wilson N, Hijazi Z (eds). Percutaneous Interventions for Congenital Heart Disease. Informa Healthcare, United Kingdom, 2007, pp 49–59.
40. Hundley WG, Li HF, Lange RA, et al. Assessment of left-to-right intracardiac shunting by velocity-encoded, phase-difference magnetic resonance imaging. A comparison with oximetric and indicator dilution techniques. Circulation 1995; 91: 2955–2960.
41. Levin AR, Spach MS, Boineau JP, Canent RV Jr, Capp MP, Jewett PH. Atrial pressure-flow dynamics in atrial septal defects (secundum type). Circulation 1968; 37: 476–488.
42. Boehrer JD, Lange RA, Willard JE, Grayburn PA, Hillis LD. Advantages and limitations of methods to detect, localize, and quantitate intracardiac left-to-right shunting. Am Heart J 1992; 124: 448–455.
43. Jarmakani JM. Cardiac catheterization. In: Moss AJ, Adams FH, Emmanouilides GC (eds). Heart disease in infants, children and adolescents, 2nd edn. Williams & Wilkins, Baltimore, 1977, pp 116–117.
44. Williamson EE, Kirsch J, Araoz PA, et al. ECG-gated cardiac CT angiography using 64-MDCT for detection of patent foramen ovale. AJR Am J Roentgenol 2008; 190: 929–933.
45. American College of Cardiology Foundation Task Force on Expert Consensus Documents, Hundley WG, Bluemke DA, et al. ACCF/ACR/AHA/NASCI/SCMR 2010 expert consensus document on cardiovascular magnetic resonance: a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents. J Am Coll Cardiol 2010; 55: 2614–2662.
46. Pennell DJ, Sechtem UP, Higgins CB, et al. Clinical indications for cardiovascular magnetic resonance (CMR): consensus panel report. J Cardiovasc Magn Reson 2004; 6: 727–765.
47. Teo KS, Disney PJ, Dundon BK, et al. Assessment of atrial septal defects in adults comparing cardiovascular magnetic resonance with transoesophageal echocardiography. J Cardiovasc Magn Reson 2010; 12: 44–49.
48. Lamb HL, Kozerke S, Doornbos J, Bax JJ, de Roos A. Clinical approach to cardiovascular magnetic resonance techniques. In: Higgins CB, de Roos A (eds). MRI and CT of the Cardiovascular System. Lippincott Williams & Wilkins, Philadelphia, PA, 2006, pp 5–8.
49. Sandner TA, Theisen D, Bauner KU, Picciolo M, Reiser MF, Wintersperger BJ. Cardiac functional analysis with MRI. Radiologe 2010; 50: 514–522.
50. Colletti PM. Evaluation of intracardiac shunts with cardiac magnetic resonance. Curr Cardiol Rep 2005; 7: 52–58.
51. Hamilton-Craig C, Sestito A, Natale L, et al. Contrast transoesophageal echocardiography remains superior to contrast-enhanced cardiac magnetic resonance imaging for the diagnosis of patent foramen ovale. Eur J Echocardiogr 2011; 12: 222–227.
52. Johnson TR, Fogel MA. Abnormalities of the atria and systemic veins. In: Fogel MA (ed.). Principles and Practice of Cardiac Magnetic Resonance in Congenital Heart Disease. Wiley-Blackwell, United Kingdom, 2010, pp 110–119.
53. Thomson LE, Crowley AL, Heitner JF, et al. Direct en face imaging of secundum atrial septal defects by velocity-encoded cardiovascular magnetic resonance in patients evaluated for possible transcatheter closure. Circ Cardiovasc Imaging 2008; 1: 31–40.
54. Beerbaum P, Korperich H, Barth P, Esdorn H, Gieseke J, Meyer H. Noninvasive quantification of left-to-right shunt in pediatric patients: phase-contrast cine magnetic resonance imaging compared with invasive oximetry. Circulation 2001; 103: 2476–2482.
55. Fratz S, Chung T, Greil G, et al. Guidelines and protocols for cardiovascular magnetic resonance in children and adults with congenital heart disease: SCMR expert consensus group on congenital heart disease. J Cardiovascu Magn Reson 2013; 15: 51–76.
56. Gatehouse PD, Rolf MP, Graves MJ, et al. Flow measurement by cardiovascular magnetic resonance: a multi-centre multi-vendor study of background phase offset errors that can compromise the accuracy of derived regurgitant or shunt flow measurements. J Cardiovasc Magn Reson 2010; 12: 5.
57. Beerbaum P, Parish V, Bell A, Gieseke J, Korperich H, Sarikouch S. Atypical atrial septal defects in children: noninvasive evaluation by cardiac MRI. Pediatr Radiol 2008; 38: 1188–1194.
58. Beerbaum P, Korperich H, Esdorn H, et al. Atrial septal defects in pediatric patients: noninvasive sizing with cardiovascular MR imaging. Radiology 2003; 228: 361–369.
59. Holmvang G, Palacios IF, Vlahakes GJ, et al. Imaging and sizing of atrial septal defects by magnetic resonance. Circulation 1995; 92: 3473–3480.
60. Valverde I, Simpson J, Schaeffter T, Beerbaum P. 4D phase-contrast flow cardiovascular magnetic resonance: comprehensive quantification and visualization of flow dynamics in atrial septal defect and partial anomalous pulmonary venous return. Pediatr Cardiol 2010; 31: 1244–1248.
61. Wang ZJ, Reddy GP, Gotway MB, Yeh BM, Higgins CB. Cardiovascular shunts: MR imaging evaluation. Radiographics 2003; 23 (Spec No): S181–S194.
62. Prasad SK, Soukias N, Hornung T, et al. Role of magnetic resonance angiography in the diagnosis of major aortopulmonary collateral arteries and partial anomalous pulmonary venous drainage. Circulation 2004; 109: 207–214.
63. Fogel MA. Assessment of ventricular function and flow. In: Fogel MA (ed.). Principles and Practice of Cardiac Magnetic Resonance in Congenital Heart Disease: Form, Function, and Flow. Wiley-Blackwell, United Kingdom, 2010, pp 63–72.
64. Juluru K, Vogel-Claussen J, Macura KJ, Kamel IR, Steever A, Bluemke DA. MR imaging in patients at risk for developing nephrogenic systemic fibrosis: protocols, practices, and imaging techniques to maximize patient safety. Radiographics 2009; 29: 9–22.
65. Ain DL, Narula J, Sengupta PP. Cardiovascular imaging and diagnostic procedures in pregnancy. Cardiol Clin 2012; 30: 331–341.
66. Marinskis G, Bongiorni MG, Dagres N, et al. Performing magnetic resonance imaging in patients with implantable pacemakers and defibrillators: results of a European Heart Rhythm Association survey. Europace 2012; 14: 1807–1809.
67. Therrien J, Warnes C, Daliento L, et al. Canadian cardiovascular society consensus conference 2001 update: recommendations for the management of adults with congenital heart disease part III. Can J Cardiol 2001; 17: 1135–1158.
68. Daniel WC, Lange RA, Willard JE, Landau C, Hillis LD. Oximetric versus indicator dilution techniques for quantitating intracardiac left-to-right shunting in adults. Am J Cardiol 1995; 75: 199–200.
69. Fu Y, Cao Q, Hijazi Z. Closure of secundum atrial septal defect using the Amplatzer septal occluder. In: Sievert H, Qureshi S, Wilson N, Hijazi Z (eds). Percutaneous Interventions for Congenital Heart Disease. Informa Healthcare, United Kingdom, 2007, pp 265–288.
70. Oliver JM, Gallego P, Gonzalez AE, et al. Surgical closure of atrial septal defect before or after the age of 25 years. comparison with the natural history of unoperated patients. Rev Esp Cardiol 2002; 55: 953–961.
71. King TD, Thompson SL, Steiner C, Mills NL. Secundum atrial septal defect. nonoperative closure during cardiac catheterization. JAMA 1976; 235: 2506–2509.
72. Gervasi L, Basu S. Atrial septal defect devices used in the cardiac catheterization laboratory. Prog Cardiovasc Nurs 2009; 24: 86–89.
73. Bjornstad PG. The role of devices in the closure of atrial septal defects in the oval fossa. Cardiol Young 1998; 8: 285–286.
74. Alexi-Meskishvili VV, Konstantinov IE. Surgery for atrial septal defect: from the first experiments to clinical practice. Ann Thorac Surg 2003; 76: 322–327.
75. Luermans JG, Post MC, Yilmaz A. Late device thrombosis after atrial septal defect closure. Eur Heart J 2010; 31: 142.