

Transoesophageal echocardiography during interventional cardiac catheterisation in congenital heart disease

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The non-surgical treatment of congenital heart malformations or iatrogenic lesions following cardiac surgery by interventional cardiac catheterisation is so well established that up to 40% of cases can be treated in this way. The most frequent procedures undertaken are balloon valvotomy or angioplasty, stenting of arterial or venous stenoses, closure of intracardiac or extracardiac communications, and balloon atrial septostomy or blade atrial septectomy. While most of these are carried out with the assistance of radiographic screening, in some circumstances transoesophageal echocardiography greatly improves the success and safety. Transoesophageal ultrasonography is an important part of imaging for closure of oval fossa atrial septal defects, occlusion of baffle fenestrations following total caval pulmonary connection, closure of congenital (or ischaemic) muscular ventricular septal defects, blade atrial septectomy, balloon mitral valvotomy, and non-surgical reduction of the ventricular septum in hypertrophic cardiomyopathy. Intracardiac ultrasound, however, may become an alternative imaging technique during device closure of atrial septal defects in older patients, avoiding the need for general anaesthesia.

Atrial septal defects

The initial selection of patients for transcatheter closure of an atrial septal defect is based on precordial echocardiography, which allows the various types to be distinguished. The final arbiter of suitability is transoesophageal echocardiography, usually performed immediately before the procedure, except in adolescents and adults with a poor echo window in whom

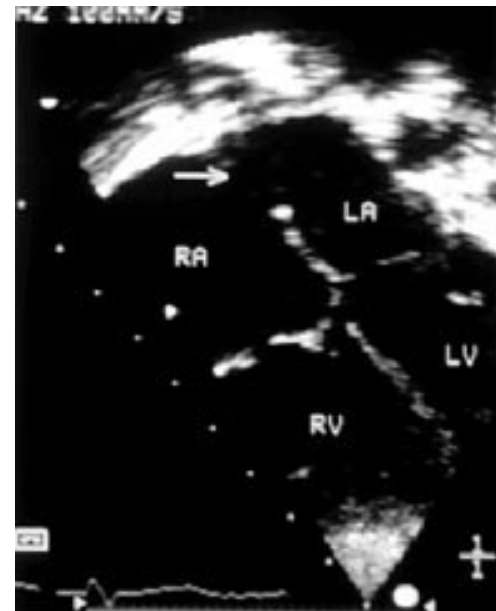


Figure 2 Four chamber section from an oval fossa atrial septal defect, in which there is a deficiency of infolding with an incomplete postero-superior rim. The arrow identifies the defect. LA, left atrium; RA, right atrium; LV, left ventricle; RV, right ventricle.

confirmation of the diagnosis will be based on prior transoesophageal echocardiography.

The true interatrial septum is the oval fossa, the majority of the remaining of tissue separating the atrial chambers being composed of an infolding of the atrial wall.¹ Defects within the oval fossa are "secundum" defects (fig 1), which may extend outside the true limits of the oval fossa when there is a deficiency of the infolding (fig 2). These defects may extend therefore posteriorly to the mouth of the coronary sinus, inferiorly to the mouth of the inferior caval vein, or superiorly into the mouth of the superior caval vein. With posterosuperior extension, the defect may extend to the mouths of the pulmonary veins, particularly the right upper pulmonary vein, while anterior encroachment is to the aorta. In contrast, a superior sinus venosus interatrial communication is found within the mouth of the superior caval vein, which has a biatrial connection, overriding the rim of the oval fossa so as to produce an "extracardiac" interatrial communication (fig 3). A coronary sinus atrial septal defect is extremely rare. It is a deficiency of the party wall between the coronary sinus and the left atrium and produces an interatrial communication through the mouth of the coronary sinus. A primum defect is part of an

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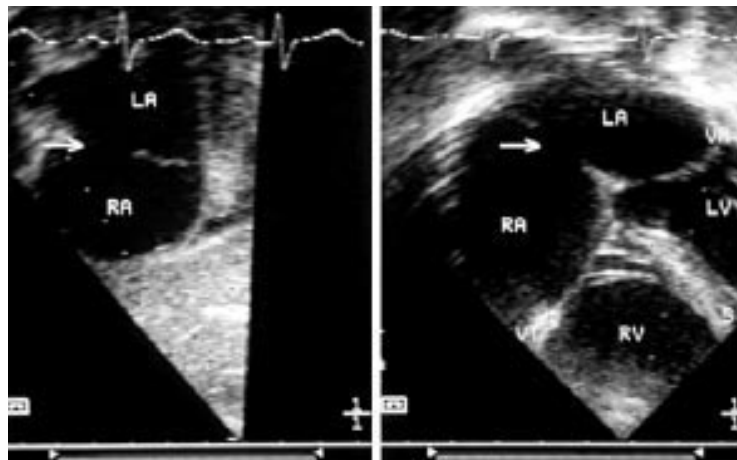


Figure 1 (Left) Short axis section of the atria, showing an oval fossa defect suitable for transcatheter closure. (Right) Four chamber section from the same patient, revealing an adequate rim of septum between the defect and the atrioventricular valves. A, left atrium; RA, right atrium; arrow, atrial septal defect; LV, left ventricle; RV, right ventricle; S, septum.

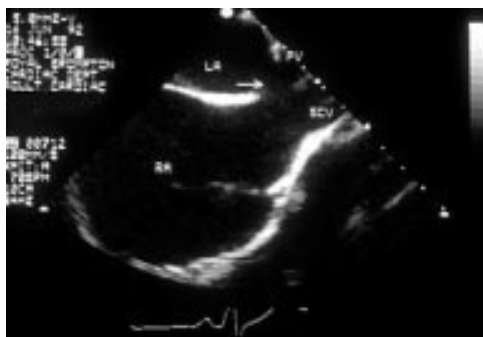


Figure 3 Transoesophageal echocardiogram in the vertical plane from a patient with a sinus venosus atrial septal defect, in which the superior caval vein (SCV) and right upper pulmonary vein (PV) override the atrial septum which divides the left atrium (LA) from right atrium (RA).

atrioventricular septal defect and is roofed superiorly by the inferior border of the oval fossa and inferiorly by the superior and inferior bridging leaflets of the common atrioventricular valve. It is important to be aware that only secundum defects are suitable for transcatheter device closure and then only when there is a 4–5 mm rim between the defect and the atrioventricular valves, the superior and inferior caval veins, and the entry of the pulmonary veins into the left atrium. Three dimensional echocardiography will demonstrate the shape and borders of an oval fossa defect.^{2–4} Depending on the location, any attempt to close one of the other types of interatrial communications is likely to result in obstruction to the systemic or pulmonary venous pathways or coronary sinus, or interference with atrioventricular valve function. Anomalous pulmonary venous drainage is also a contraindication unless more than 70% of pulmonary venous return is normal.

The most reliable imaging modalities for delivery of an atrial septal defect device into the atrial septum are fluoroscopy and transoesophageal echocardiography.² To optimise patient comfort and minimise patient movement during device placement, intubation and general anaesthesia are preferable. Biplane or omniplane echocardiography allows the exact morphology and diameter of the defect to be determined as well as the dimensions of the rim and diameter of the septum itself (fig 4). It is important to exclude a sinus venosus, coronary sinus or primum defect which will be unsuitable for device closure. In some cases, a dominant oval fossa atrial septal defect will be associated with additional fenestrations within the oval fossa, or alternatively multiple fenestrations within the oval fossa may be present, often associated with an aneurysm.²

The femoral vein is entered percutaneously and the atrial septal defect is crossed with an end hole angiographic catheter, through which an exchange guidewire can be introduced into the left lower pulmonary vein. It is important to confirm that the catheter or guidewire passes through the main and largest atrial septal defect (fig 5), or in the case of multiple fenestrations that it passes through one of the more central defects. A sizing balloon is then passed over the guidewire into the left atrium, where it is inflated to a diameter of about



Figure 4 Transoesophageal echocardiogram in which an oval fossa atrial septal defect is indicated by an arrow. LA, left atrium, RA, right atrium.

4–5 mm greater than the measured size of the defect. The balloon is slowly withdrawn through the defect by retracting the balloon catheter, simultaneously removing fluid from the balloon during retraction if necessary (fig 6). The stretched diameter of the defect is that which indents both sides of the body of the balloon. This diameter is measured on the echocardiogram and usually exceeds the measured diameter by about 20%. Finally, the defect is completely occluded with an inflated balloon, and colour flow Doppler is used to interrogate the remainder of the atrial septum for other secondary defects. The importance of a small secondary defect is that the guide wire may inadvertently be passed through it, making correct balloon sizing impossible. Cases are occasionally encountered in which there are two large defects. It is important to balloon size both of them, because two closure devices may be required. In the presence of multiple fenestrations, however, sizing is not required and

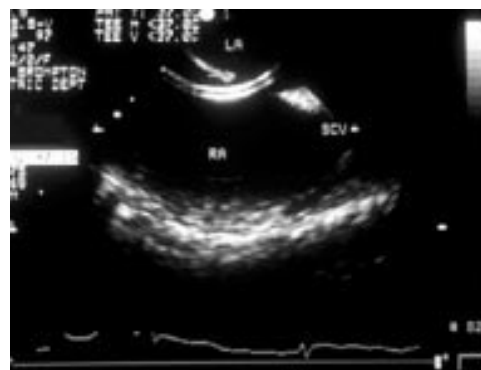


Figure 5 Transoesophageal echocardiogram identifying a catheter passing through a moderately large oval fossa atrial septal defect. LA, left atrium, RA, right atrium; SCV, superior caval vein.

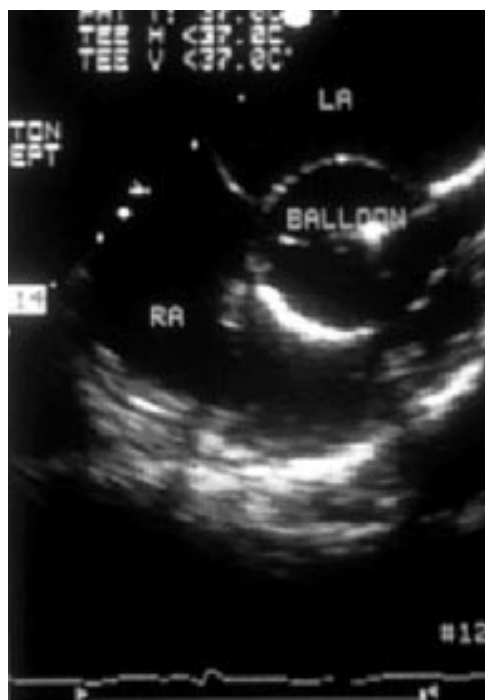


Figure 6 Transoesophageal echocardiogram recorded during balloon sizing of an oval fossa atrial septal defect. LA, left atrium, RA, right atrium.

only one device is usually needed. When there is an aneurysm of the oval fossa multiple fenestrations may occur, but closure using two devices has been described.⁵

Having determined the stretched diameter of the defect an appropriate closure device is selected and a suitable Mullins transseptal sheath is passed from the femoral vein across the defect into the mid part of the left atrium, avoiding the left atrial appendage, mitral valve or pulmonary veins. Using fluoroscopy, the left atrial disc is opened (fig 7). With cross sectional echocardiographic imaging, the left atrial disc is drawn into the defect and visualised from the four chamber horizontal section and vertical section in the plane of the superior caval vein. With the left atrial disc touching the atrial septum, and providing no part of the device is prolapsing through to the right atrium, the right atrial disc is then opened by withdrawing the Mullins sheath. Once both the discs are open, the delivery catheter or wire and the sheath are allowed to assume a neutral position with no traction. Almost always, the right and left atrial component of the device can be seen clearly, with one on either side of the atrial septum (fig 8). However, the eustachian valve appearing as a linear echo in the right atrium can be misconstrued as the atrial septum and it may be wrongly concluded that both discs have been opened in the left atrium. With the closure device in an optimal position, pulling and pushing on the delivery wire or catheter then confirms the device is secure. Finally, the device is released and colour flow Doppler is used to identify any residual defects.

Thus, transoesophageal echocardiography plays an important role in transcatheter closure of atrial septal defects. It is used before the



Figure 7 Transoesophageal echocardiogram recorded during deployment of a closure device illustrating the left atrial disc opened in the left atrium (arrow). LA, left atrium, RA, right atrium.

procedure when the precordial echocardiogram is inadequate to identify the defect and during the procedure with fluoroscopy to guide placement of the occluder. Before closure, transoesophageal echocardiography is used to determine the diameter of the defect, the length of the atrial septum from pulmonary

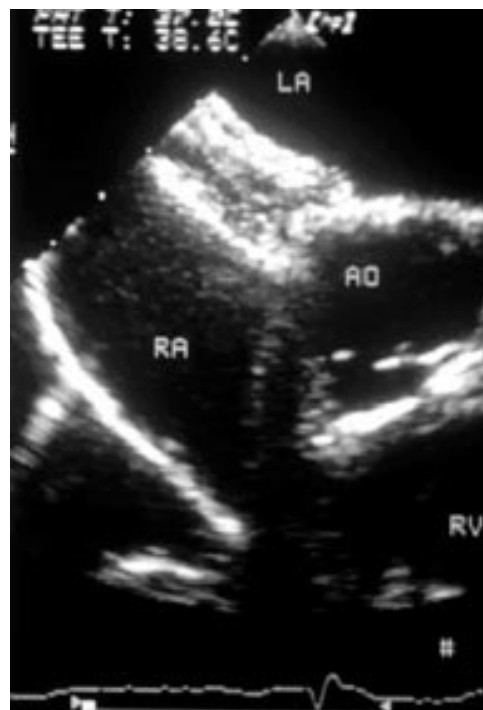


Figure 8 Transoesophageal echocardiogram recorded during device closure of an atrial septal defect with an Amplatzer occluder, showing left atrial and right atrial discs opened in their respective atria. AO, aortic root; LA, left atrium, RA, right atrium; RV, right ventricle.

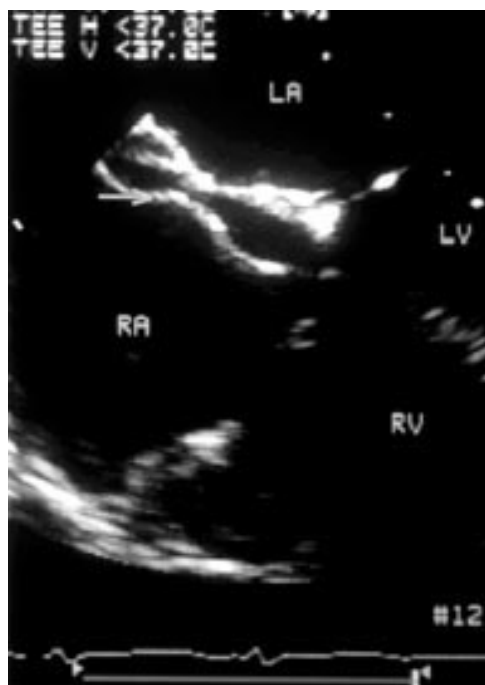


Figure 9 Transoesophageal echocardiogram recorded during closure of an oval fossa defect with a Cardioseal device (arrow). LA, left atrium; RA, right atrium; LV, left ventricle; RV, right ventricle.

vein, to the mitral hinge point, the length of rim from the edge of the defect to the atrial wall, the superior caval vein, the right upper pulmonary vein and the medial attachment of the mitral valve. Any additional defects or fenestrations should also be identified. Additional information is provided about the position of the sheath and occluder during and after deployment and the outcome of the procedure after release, particularly the presence of residual shunting, obstruction to systemic or pulmonary venous pathways, and interference with atrioventricular valve function. The four chamber long axis and short axis views are the basic echocardiographic sections used during deployment but other views may be required, depending upon the exact location of the defect. It seems likely, however, that intracardiac ultrasound could replace transoesophageal echocardiography as the optimal imaging modality.^{6 7}

The morphology of the atrial septal defect or defects is important in selecting which device to use for closure. A variety are now available⁸⁻¹⁴ and interatrial communications with a stretched diameter up to 20 mm may be closed with any of the available systems (figs 9 and 10). Larger defects, up to 40 mm, should be closed with a self-centring device, the best currently available being the Amplatzer septal occluder, although the Starflex is an adequate alternative in some cases. Multiple fenestrations in the oval fossa can be closed with a device in which the left and right atrial discs are connected by a thin central body (Cardioseal, buttoned or Helex). An aneurysm of the atrial septum need not be a contraindication to transcatheter device closure, but the left atrial disc should cover most of the atrial septum. In general terms, for all patients, the diameter of the left



Figure 10 Transoesophageal echocardiogram recorded during closure of an oval fossa atrial septal defect with an Angel Wings device. LA, left atrium; RA, right atrium.

atrial disc should not exceed the diameter of the atrial septum measured in four chamber and long axis echocardiographic sections.

Ventricular septal defects

The first description of transcatheter closure of a ventricular septal defect was that of Lock and colleagues in 1988.¹⁵ There have been a small number of subsequent publications¹⁶⁻¹⁹ and device closure of a ventricular septal defect remains the subject of research protocols. The paucity of information available may be a reflection of the technical difficulties which can be encountered. Suitable candidates are those with a muscular ventricular septal defect, a residual defect at the patch margins following cardiac surgery, and ischaemic defects following myocardial infarction.^{20 21} Because of their close proximity to the aortic and tricuspid valves, perimembranous ventricular septal defects are generally unsuitable for transcatheter device closure unless there is a contraindication to open heart surgery. Even then, only perimembranous defects opening to the inlet of the right ventricle might be suitable.¹⁶ Other defects unsuitable for device closure include those which are doubly committed or associated with aortic valve prolapse.

The majority of ventricular septal defects are closed with a device delivered through a long transeptal sheath introduced through the right internal jugular vein. The exception is some muscular inlet or perimembranous defects in which the sheath is inserted into a femoral vein. It is often safer to have an arteriovenous guidewire passing from the femoral artery to the internal jugular or femoral vein for introduction of the sheath. An alternative approach is to use a femoral venous entry site and puncture of the oval fossa so that an end

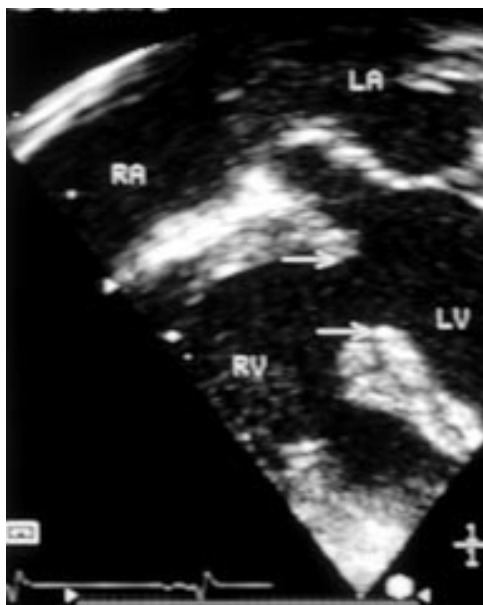


Figure 11 Transoesophageal horizontal section of a large muscular ventricular septal defect, whose borders are identified by the arrows. LA, left atrium; RA, right atrium; LV, left ventricle; RV, right ventricle.

hole catheter can be passed first to the left atrium and then to the left ventricle, across the ventricular septal defect into the right ventricle. Thus, a guide wire and long sheath can then be introduced from the femoral vein, through the left side of the heart and into the right ventricle. Whichever route is used, the guidewire must be identified traversing the main defect and in the case of multiple defects crossing one that is situated centrally in the ventricular septum. In similar fashion to atrial septal defects, a combination of fluoroscopy and transoesophageal echocardiography is required.

Balloon sizing of the defect is essential, but the type of closure device selected will depend on many factors, including the preference and experience of the operator, the thickness of the

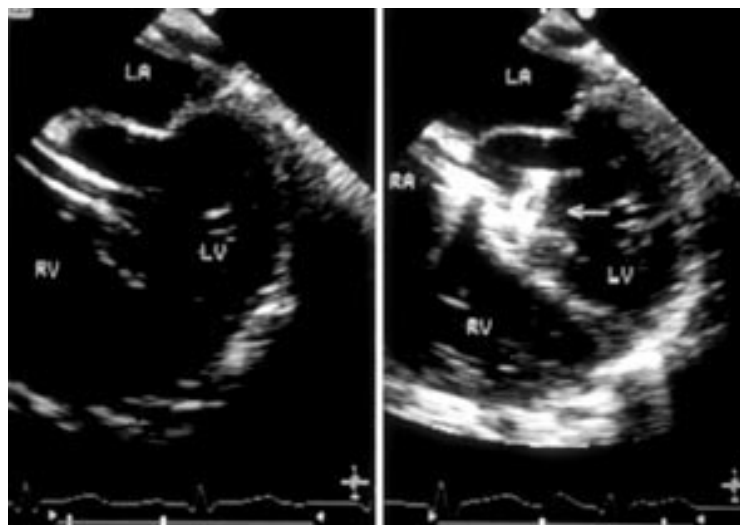


Figure 12 (Left) Transoesophageal echocardiogram recorded during device closure of a perimembranous ventricular septal defect, in which the sheath can be seen crossing the defect from right ventricle (RV) to left ventricle (LV). (Right) Horizontal plane transoesophageal echocardiogram recorded during device closure of a perimembranous ventricular septal defect, in which the left ventricular disc (arrow) can be seen to open within the left ventricle. LA, left atrium; RA, right atrium.

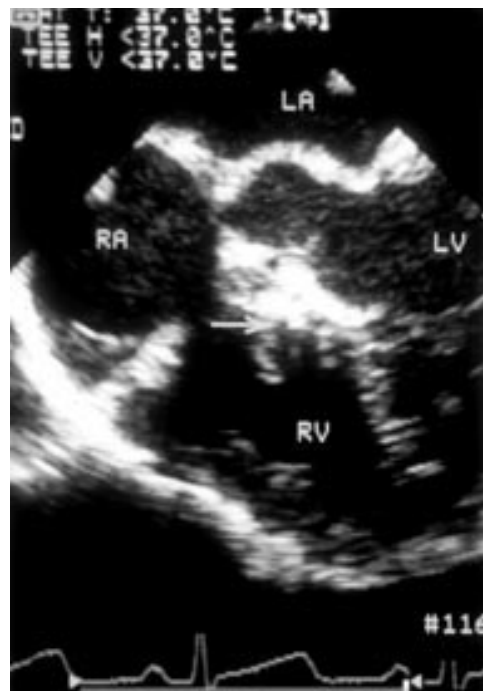


Figure 13 Horizontal transoesophageal echocardiogram following device closure of a perimembranous ventricular septal defect. An arrow identifies the position of the closure device within the ventricular septal defect.

ventricular septum, and the exact morphology of the defect. Multiple muscular defects are best closed with a Cardioseal device or others with a thin central connecting body. My own preference for solitary defects is to use one of the various Amplatzer occluders. A device specifically designed for ventricular septal defects is available,²¹ although in some cases the Amplatzer ductal occluder or atrial septal defect occluder may be preferable. The Cardioseal, however, has been used frequently to close solitary defects. An Amplatzer device, designed specifically for perimembranous defects, is currently undergoing clinical trials.

Ventricular septal defects cannot be closed safely without the use of transoesophageal echocardiography, which is used to determine the morphology and diameter of the defect (fig 11) and the relation to the semilunar and atrioventricular valves and their tensor apparatus. It is essential to obtain the echocardiographic sections which adequately display the defect. There is frequently a need for the transgastric short axis view. Important information is provided about the position of the wire, sheath, and occluder during (fig 12) and after (fig 13) deployment, particularly any interference with atrioventricular or semilunar valve function before release. With the closure device positioned in the long sheath, the distal disc is opened in the appropriate ventricle. When using the internal jugular vein as the entry site, the tip of the sheath will be in the body of the left ventricle. With echocardiographic imaging, the tip of the sheath and the opened distal disc are pulled into the ventricular septal defect. Retracting the sheath over the delivery wire or catheter then allows the remainder of the



Figure 14 Transgastric short axis echocardiographic section following transcatheter closure of a muscular ventricular septal defect, showing the closure device within the muscular defect. The right ventricular disc is well seen within the right ventricle (RV), although the left ventricular disc within the left ventricle (LV) is imaged less well.

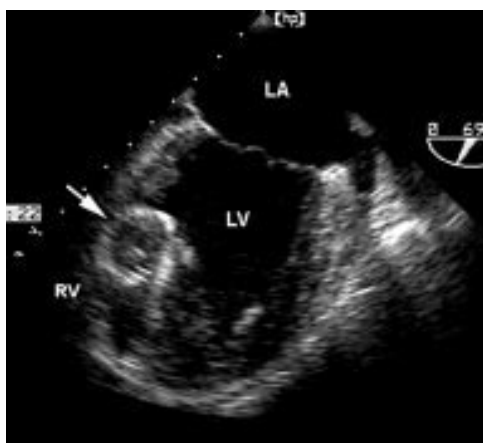


Figure 15 Transoesophageal echocardiogram recorded immediately following device closure of an ischaemic muscular ventricular septal defect in a patient presenting with cardiogenic shock. An arrow identifies the Amplatzer closure device. LA, left atrium; LV, left ventricle; RV, right ventricle.

device to open within the defect and right ventricle. Pulling and pushing gently on the delivery wire or catheter confirms the device is secure. Only when echocardiography confirms optimal device position is it released (figs 14 and 15). There is a significant risk of a periprocedural haemopericardium which can be recognised immediately.

Only a small number of patients with a ventricular septal defect are suitable candidates for

device closure, which should be performed with intubation and general anaesthesia. The procedure is technically much more difficult than closure of an atrial septal defect and an experienced team of cardiologist, anaesthetists, and surgeons should be available to ensure that there is adequate patient safety. Instruction by an individual experienced in all aspects of the technique is highly desirable for those contemplating performing the procedure for the first time. Recently, we have used the techniques described above for the successful closure of ischaemic defects in patients presenting with cardiogenic shock.

Miscellaneous lesions

Transoesophageal echocardiographic imaging should be used for device closure of patent foramen ovale associated with transient cerebral ischaemic attacks²² and for occlusion of a baffle fenestration made at the time of a total caval pulmonary connection²³ (fig 16), and for the closure of a baffle leak following the Mustard or Senning operation. The techniques used are very similar to those for closure of a conventional atrial septal defect.

In the majority of cases of congenital mitral stenosis, surgical treatment provides the best outcome. When balloon mitral valvotomy is performed, however, transoesophageal echocardiography is essential in small children for safe perforation of the atrial septum and for precise positioning of the balloon dilatation catheter across the mitral valve, avoiding unnecessary damage to the tensor apparatus.²⁴ Blade atrial septectomy in infants and small children is a hazardous procedure, which is best monitored with a combination of fluoroscopy and transoesophageal echocardiography.²⁵ The procedure is usually reserved for patients with a restrictive atrial septal defect in the setting of mitral atresia, absence of the left atrioventricular connection, or severe mitral stenosis associated with a hypoplastic left ventricle. Finally, it is our policy, in children undergoing non-surgical myocardial reduction for hypertrophic obstructive cardiomyopathy using alcohol ablation,²⁶ always to monitor the procedure with transoesophageal echocardiography. It allows precise imaging of the site of injection of alcohol by first delineating the area

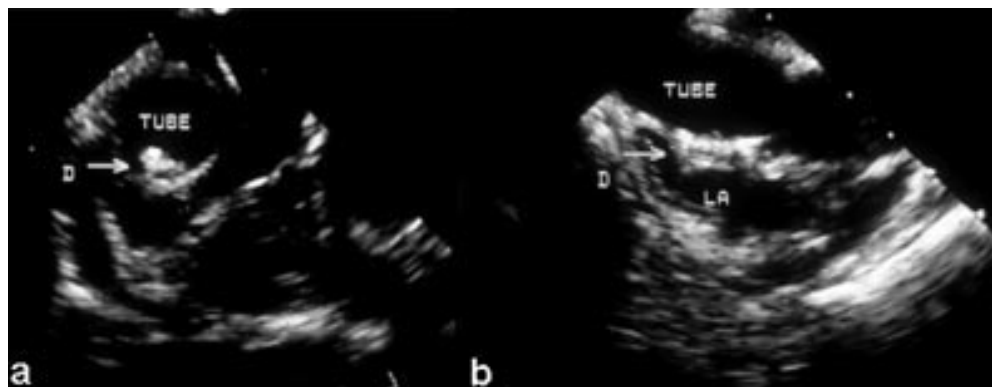


Figure 16 Transoesophageal echocardiograms immediately following transcatheter closure of a fenestrated lateral tunnel (tube) in a patient who had undergone a previous total caval pulmonary connection. The closure device (D, arrow) is seen in (a) horizontal and (b) vertical sections. LA, left atrium.

with a myocardial contrast agent, injected directly through the coronary catheter.

- 1 Ho S, McCarthy K, Rigby M. Anatomy of atrial and ventricular septal defects. *J Intervent Cardiol* 2000;13:475-85.
- 2 Magni G, Hijazi Z, Pandian N, et al. Two- and three-dimensional transesophageal echocardiography in patient selection and assessment of atrial septal defect closure by the new DAS-Angel Wings device. *Circulation* 1997;96:1722-8.
- 3 Marx G, Fulton D, Pandian N, et al. Delineation of site, relative size and dynamic geometry of atrial septal defect by real-time three-dimensional echocardiography. *J Am Coll Cardiol* 1995;25:482-90.
- 4 Dall'Agata A, McGhie J, Taams M, et al. Secundum atrial septal defect is a dynamic three-dimensional entity. *Am Heart J* 1999;137:1075-81.
- 5 Ewert P, Berger F, Vogel M, et al. Morphology of perforated atrial septal aneurysm suitable for closure by transcatheter device placement. *Heart* 2000;84:327-31.
- 6 Hwang B, Lee P, Chiu P. Intracardiac ultrasound assessment of atrial septal defect: comparison with transthoracic echocardiographic, angiocardiographic and balloon-sizing measurements. *Cardiovasc Intervent Radiol* 2001;24:84.
- 7 Hijazi Z, Wang Z, Cao Q, et al. Transcatheter closure of atrial septal defects and patent foramen ovale under intracardiac echocardiographic guidance: feasibility and comparison with transesophageal echocardiography. *Catheter Cardiovasc Interv* 2001;52:194-9.
- 8 Lock J, Rome J, Davis R, et al. Transcatheter closure of atrial septal defects. *Circulation* 1989;79:1091-9.
- 9 Zamora R, Lloyd T, Beekmann R, et al. Follow-up results of the multi-institutional phase 1 FDA supervised clinical trials of transcatheter occlusion of atrial septal defects with buttoned device [abstract]. *J Am Coll Cardiol* 1997;29:143A.
- 10 Carminati M, Hausdorf G, Tynan M, et al. Initial clinical experience of transcatheter closure of secundum atrial septal defect with a septal occlusion device [abstract]. *Eur Heart J* 1997;19(suppl):136.
- 11 Das G, Shrivastava S, O'Laughlin D, et al. Intermediate term follow up of patients after percutaneous closure of atrial septal defects with the Das angel wings device [abstract]. *Circulation* 1996;94:1-56.
- 12 Rieckers C, Hamm C, Stern H, et al. Percutaneous closure of secundum atrial septal defect with a new self-centering device ("angel wings"). *Heart* 1998;80:517-21.
- 13 Rhodes J, Dobrolet N, Lane G, et al. Atrial septal defect closure with the Helix septal occluder device: the FDA phase I feasibility trial. *Cardiol Young* 2001;11(suppl):71.
- 14 Pedra C, Pihkala J, Lee K-J, et al. Transcatheter closure of atrial septal defects using the Cardio-Seal implant. *Heart* 2000;84:320-6.
- 15 Lock J, Block R, McKay R, et al. Transcatheter closure of ventricular septal defects. *Circulation* 1988;78:361-8.
- 16 Rigby M, Redington A. Primary transcatheter umbrella closure of perimembranous ventricular defect. *Br Heart J* 1995;73:368-71.
- 17 Laussen P, Hansen D, Perry S, et al. Transcatheter closure of ventricular septal defects: hemodynamic instability and anesthetic management. *Anesth Analg* 1995;80:1076-82.
- 18 Das G, Hijazi Z, O'Laughlin M, et al. Initial results of the VSD/PFO/ASD closure trial [abstract]. *J Am Coll Cardiol* 1996;27:119A.
- 19 Sideris E, Walsh K, Haddad J, et al. Occlusion of congenital ventricular septal defects by the buttoned device. *Heart* 1997;77:276-9.
- 20 Pesonen E, Thilen U, Sandstrom S, et al. Transcatheter closure of post-infarction ventricular septal defect with the Amplatzer septal occluder device. *J Scand Cardiovasc* 2000;34:446-8.
- 21 Thanopoulos B, Tsaousis G, Konstadopoulou G, et al. Transcatheter closure of muscular ventricular septal defects with the Amplatzer ventricular septal defect occluder: initial clinical applications in children. *J Am Coll Cardiol* 1999;33:1395-9.
- 22 Daniel W. Transcatheter closure of patent foramen ovale. Therapeutic overkill or elegant management for selected patients at risk. *Circulation* 1992;86:1013-15.
- 23 Redington A, Rigby M. Transcatheter closure of inter-atrial communications with a modified umbrella device. *Br Heart J* 1994;72:372-7.
- 24 Spevak P, Bass J, Ben-Sacher G, et al. Balloon angioplasty for severe congenital mitral stenosis. *Am J Cardiol* 1990;66:472-6.
- 25 Perry S, Lang P, Keane J, et al. Creation and maintenance of an adequate interatrial communication in patients with left atrioventricular valve atresia or stenosis. *Am J Cardiol* 1986;58:622-6.
- 26 Sigwart U. Alcohol ablation for mid-ventricular obstruction: straying from the path of righteousness? *Eur J Echocardiogr* 2000;1:227-8.

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