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CASE REPORT

Successful percutaneous closure of spiral atrial septal defect

Mashail Alobaidan, A Saleem, H Abdo and J Simpson[1]

Department of Pediatric Cardiology, Armed Forces Hospital, Abubaker Alrazi Street, Sulaimania District, Riyadh 11199, Saudi Arabia
[1]Department of Congenital Heart Disease, Evelina London Children's Hospital, Westminster Bridge Road, London, UK

Correspondence should be addressed to J Simpson
Email
John.simpson@gstt.nhs.uk

Summary

The case report of a 15-year-old patient with an unusual form of atrial septal defect is described. Echocardiography showed separation of the secundum and primum atrial septums due to abnormal posterior and leftward attachment of the primum septum into the roof of the left atrium. The morphology has been variably described as a 'double' atrial septum or 'spiral' atrial septal defect. Despite the technical challenge of this form of atrial septal defect, it was effectively closed by ensuring that all relevant septal structures were incorporated between the discs of the occlusion device. This was associated with a stable position and good medium-term outcome. This contrasts with the experience of others where device embolisation or technical failure has been described.

Learning points:

- The spiral atrial septal defect is characterised by an apparently 'double' atrial septum.
- Such atrial septal defects (ASDs) have been associated with a high rate of technical failure of transcatheter closure.
- 3D echocardiography assists in understanding the anatomy of the defect.
- Following deployment of the ASD occlusion device transoesophageal echocardiography is essential to ensure that both septum primum and secundum are between the occluder discs.
- Catheter closure can be successful if close attention is paid to the morphology of the defect and incorporation of margins within the discs of the septal occluder.

Background

Device occlusion of secundum septal defects has become an accepted means of closure in cases where the anatomy of the atrial septum is judged suitable. Selection criteria have included adequacy of the rims around the defect, particularly the inferior margin and the size of the defect in relation to the total septal length. An unusual morphology of atrial septal defect has been described where there is an apparently 'double atrial septum' (1). The terminology around this lesion has been disputed with other authors attributing the appearance to a deviated or malposed primary atrial septum (septum primum) (2). More recently, the wide separation of the primary atrial septum (primum septum) from the secondary septum (septum secundum) and the 'spiral' spatial arrangement of the margins of the atrial septal defect (ASD) has led to the term spiral ASD to describe this arrangement (3). This has been associated with a high risk of device embolisation or technical failure in the placement of an occluder device (3). We report the echocardiographic findings and outcome of a patient with this form of ASD in whom percutaneous occlusion was successful.

Case presentation

A 15-year-old boy presented with a history of palpitations and shortness of breath on exertion. Physical examination was remarkable for a right ventricular heave, fixed splitting of the second heart sound and an ejection murmur in the pulmonary area. Transthoracic echocardiography confirmed a large 25 mm secundum atrial septal defect associated with right ventricular volume overload. Given these findings, it was decided to attempt percutaneous occlusion of the defect under transoesophageal echocardiographic and fluoroscopic guidance.

Investigation

Cardiac catheterisation confirmed Qp/Qs of 2.6:1 and normal pulmonary artery pressures (mean 19 mmHg). Balloon sizing of the atrial septal defect gave a dimension of 26 mm and an occlutech 27 mm device was selected. The transoesophageal echocardiographic images before initial device deployment are shown in Fig. 1a, b, c, d and e. These illustrate the clear separation of the firm margins of the atrial septal defect and the foramen ovale flap. During initial deployment of a 27 mm septal occluder, the foramen ovale flap lay between the left and right atrial discs, but the firm mitral valve rim was not, leading to an unstable device position (Fig. 2a). Subsequent deployment of a larger 30 mm device allowed a satisfactory deployment position to be achieved (Fig. 2b).

Treatment and outcome

Following successful device occlusion, the patient made an uneventful recovery with symptomatic improvement, and an uncomplicated course over 1 year of follow-up.

Discussion

The echocardiographic appearances we describe are unusual, characterised by a wide separation between the primum and secundum septal structures (1, 2). Abnormal

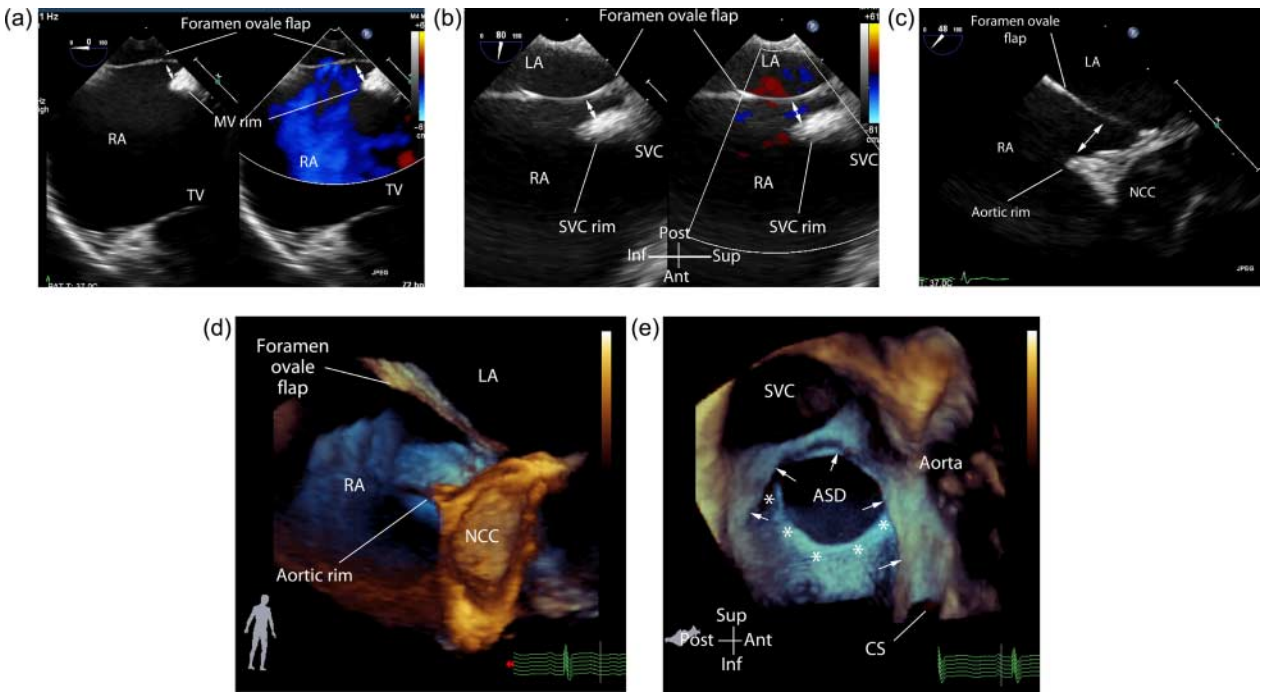

Figure 1

(a) TOE image at 0° showing separation between the foramen ovale flap (primum septum) and the mitral valve rim. The gap is shown with the double-headed arrows. (b) TOE image at 90° confirming separation of the foramen ovale flap and the superior vena cava rim of the atrial septal defect. The primum septum has attachment into the roof of the left atrium. The separation between the primum septal tissue and the firm SVC rim is indicated by the double-headed arrow. (c) TOE view at 48° showing the separation (double-headed arrow) between the aortic rim of the atrial septal defect and the foramen ovale flap. (d) Three-dimensional TOE image of the aortic rim of the atrial septal defect taken at 45° which illustrates both the plane of the firm rims of the defect starting with the aortic rim and the depth of the foramen ovale flap. The wide separation of the primum septum and the aortic rim of the defect are well seen. (e) Three-dimensional TOE image of the atrial septal defect and margins as projected from the right atrium. The firm margins of the defect including the aortic rim and superior vena cava rim are shown by the arrows and the edge of the foramen ovale flap vale by the asterisks. CS, coronary sinus; LA, left atrium; MV, mitral valve; NCC, non-coronary cusp of the aortic valve; RA, right atrium; SVC, superior vena cava; TV, tricuspid valve.

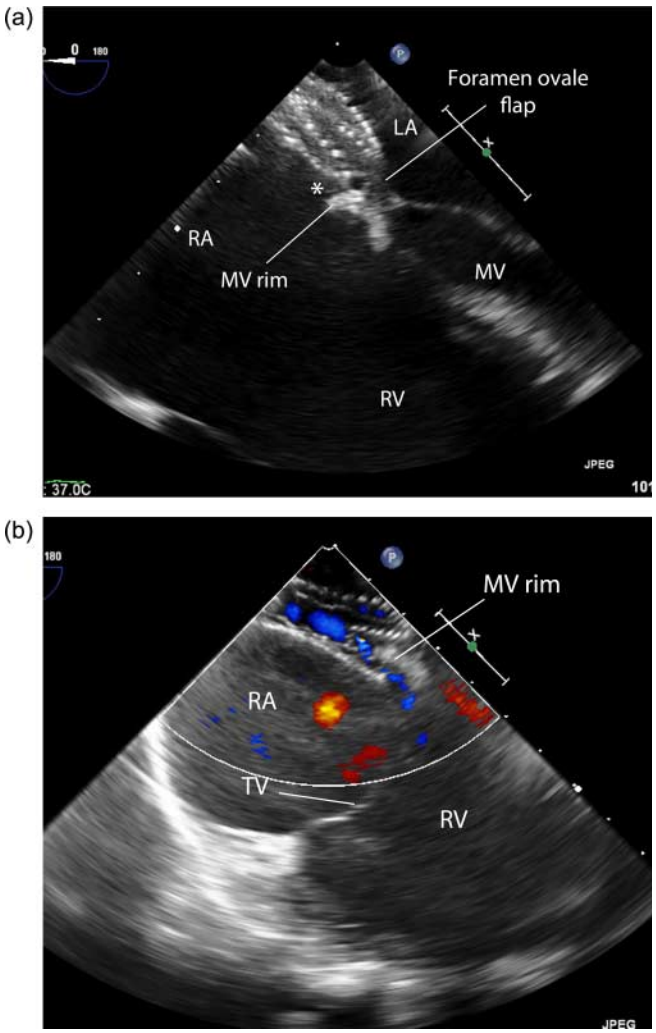

Figure 2
(a) TOE image obtained following initial deployment of 27-mm occluder device. This shows that the left atrial disc is well deployed in the right atrium but that the right disc is apposed to the foramen ovale flap valve but not the firm mitral valve rim. The asterisk indicates where the firm mitral valve margin is not gripped between the left and right atrial discs. (b) TOE image following deployment of a larger 30-mm occlusion device. This shows that the mitral valve rim is now gripped effectively between the left and right atrial discs. LA, left atrium; MV, mitral valve; RA, right atrium; RV, right ventricle; TV, tricuspid valve.

posterior and leftward attachment of the primum septum in the roof of the left atrium was evident in our case, consistent with the description of others (3). This poses a significant challenge for device closure, which depends on placement of left and right atrial discs on either side of the atrial septum. In the case we describe, the device had to appose the primum and secundum septal structures, to achieve a stable device position. Initial device deployment was unsatisfactory because the right atrial disc gripped only the primum septum leading to rocking of the device which was deemed unsatisfactory. Oversizing of the device permitted the right atrial disc to be deployed against the firm margins of the septum secundum and obliteration of the space between the primum and secundum septums. This was associated with a stable device position which has been maintained. Thus, although some authors have preferred a surgical approach and cautioned against device closure (3), effective device closure is feasible in selected cases with transoesophageal guidance and an understanding of this unusual morphology.

Declaration of interest
The authors declare that there is no conflict of interest that could be perceived as prejudicing the impartiality of the research reported.

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Patient consent
Written consent has been obtained for publication.

Author contribution statement
Dr M Alobaidan, Dr A Saleem, Dr H Abdo and Prof. J Simpson all contributed to the clinical care of the patient and the ASD occlusion procedure. Prof. J Simpson was responsible for the 3D echocardiographic assessment. All authors contributed the content of the manuscript.

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