Swiss Cheese–Like Atrial Septal Defect

Markus Schwerzmann, MD; Stephan Windecker, MD; Bernhard Meier, MD

Atrial septal defects (ASDs), apart from the bicuspid aortic valve, are the most common congenital heart malformations, and it is not uncommon for them to go undiscovered until a patient reaches adulthood.[1] An ostium secundum ASD is the most common septal defect, comprising 75% of all these defects.[2] Elective closure should be considered in patients with signs of right heart dilatation and in those who have a presumed paradoxical event.[3] In patients with an ostium secundum ASD, percutaneous closure–first reported in 1974[4]—is now well established. Before a percutaneous closure attempt, the suitability of the defect for device implantation should be assessed with transesophageal echocardiography. Associated malformations (eg, partial anomalous pulmonary venous drainage) and the number, location(s), and the dimensions of the defect(s) should be known before the procedure. A defect in the atrial septum can exist when the flap valve derived from the septum primum does not overlap its rim (eg, because of atrial dilation) or because the flap either is too small or has a hole. The valve can also contain multiple fenestrations, creating a filigreed network across the area of the fossa ovalis.[5] All this may well be combined with patent foramen ovale (PFO).

For patients with multiple ventricular septal defects, usually in the muscular ventricular septum, the term *Swiss cheese ventricular septal defect* has been coined. The term refers to the Emmental cheese, a medium-hard cheese of Swiss origin that features characteristic holes. The holes are formed during the maturing process by fermentation of propionic acid to carbon dioxide within the body of the cheese. The cheese rind forms a barrier and prevents the gas from escaping. As a result, carbon dioxide accumulates within the body of the cheese and forms the holes.[6] In general, large holes indicate a cheese with longer maturation and more flavor. The term Swiss cheese can also be used in the setting of multiple defects in the atrial septum.[7] In our personal experience, multiple interatrial defects are present in ≈5% of all patients referred for percutaneous ASD closure.

We present 2 examples of patients referred for percutaneous ASD closure with a Swiss cheese–like ASD. In the first patient, a 44-year-old woman with previous stroke and no cardiovascular risk factors, transesophageal echocardiography showed an aneurysmatic flap valve (atrial septal aneurysm) with multiple perforations (Figure 1 and Movie I). In addition, there was a PFO. No other congenital anomaly was present. The defects within the fossa ovalis and the PFO could all be successfully addressed by percutaneous device closure, but they required the implantation of 2 different devices (Figure 2 and Movie II). One device, a 25-mm Amplatzer PFO Occluder (AGA Medical, Plymouth, Minn), was primarily used to close the PFO; the second device, a cribriform Amplatzer Occluder, was necessary to cover the multiple defects within the fossa ovalis. Transesophageal echocardiography after 6 months revealed no residual shunt. In the other patient, a 19-year-old asymptomatic man with right heart dilatation and incomplete right bundle-branch block, transesophageal echocardiography showed a 12-mm defect in the fossa ovalis extending to the anterosuperior rim (Figure 3). After careful inspection of the entire septal wall, 2 more defects were seen in a more inferoposterior view (Figure 4). There was no anomalous pulmonary venous drainage. The most centrally located defect was closed with a oversized closure device, thereby squeezing or covering the additional defects. Transthoracic echocardiography after 1 month showed a small residual interatrial shunt close to the aortic valve and regression of right ventricular chamber size.

These 2 examples of Swiss cheese–like ASDs illustrate how proper identification of multiple interatrial defects is important for successful percutaneous defect closure. Hence, it is instrumental to check for additional holes in the atrial septum once a single defect has been identified.

Disclosures

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References

1. Hoffman JI, Kaplan S. The incidence of congenital heart disease. *J Am Coll Cardiol*. 2002;39:1890–1900.
2. Brickner ME, Hillis LD, Lange RA. Congenital heart disease in adults: first of two parts. *N Engl J Med*. 2000;342:256–263.
3. Webb G, Gatzoulis MA. Atrial septal defects in the adult: recent progress and overview. *Circulation*. 2006;114:1645–1653.
4. King TD, Mills NL. Nonoperative closure of atrial septal defects. *Surgery*. 1974;75:383–388.
5. Ferreira Martins JD, Anderson RH. The anatomy of interatrial communications: what does the interventionist need to know? *Cardiol Young*. 2000;10:464–473.
6. Emmentaler Switzerland home page. Available at: http://www.emmentaler.ch. Accessed November 27, 2007.
7. Harper RW, Mottram PM, McGaw DJ. Closure of secundum atrial septal defects with the Amplatzer Septal Occluder Device: techniques and problems. *Cathet Cardiovasc Intervent*. 2002;57:508–524.

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The online-only Data Supplement, which contains Movies I and II, is available with this article at http://circ.ahajournals.org/cgi/content/full/117/24/e490/DC1.

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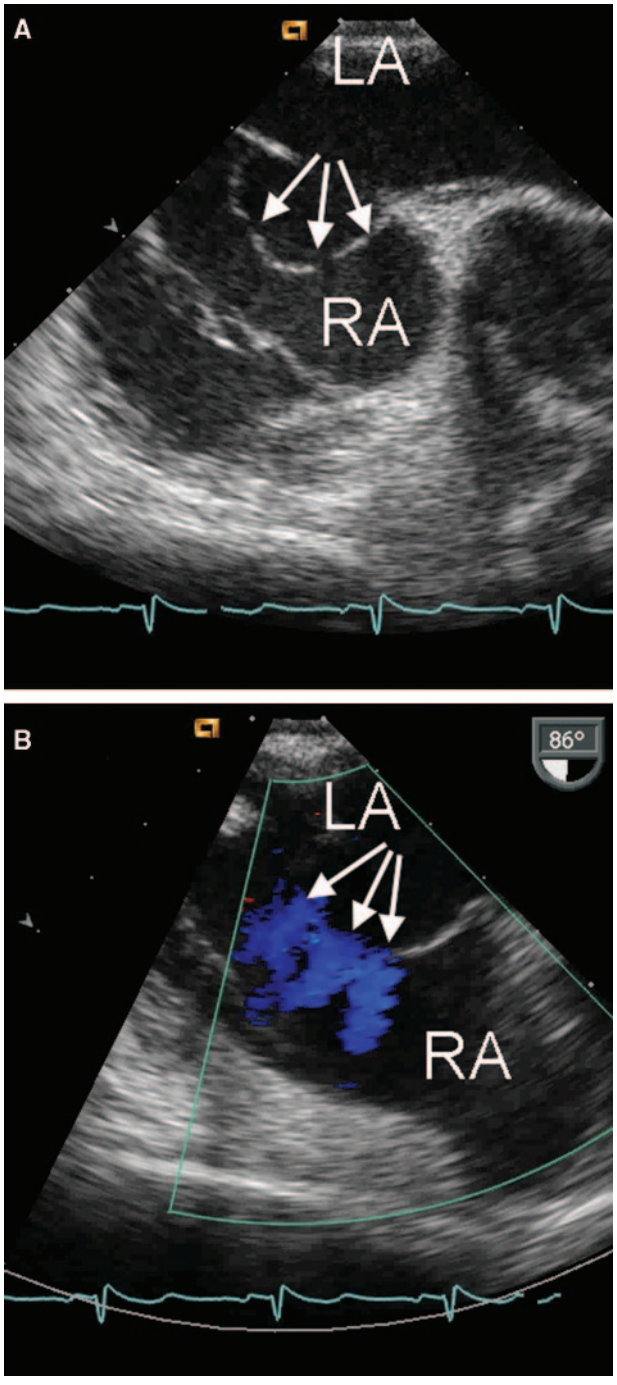

Figure 1. Multiplane transesophageal echocardiography in the 90° view showing an atrial septal aneurysm with fenestrations already identified in the B-mode (A, arrows) and clearly seen with color Doppler (B, arrows). The PFO is not depicted. LA indicates left atrium; RA, right atrium.

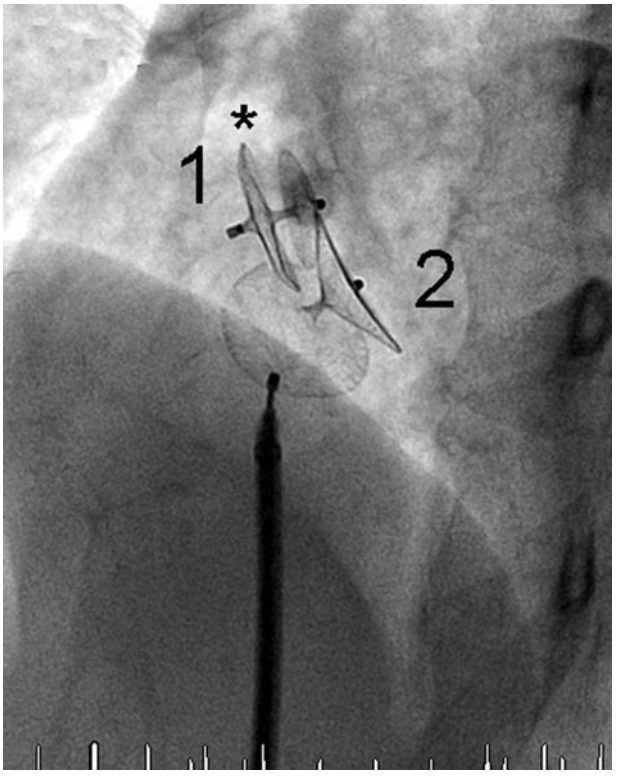

Figure 2. Percutaneous closure of the multiple defects of patient 1. Device 1 is an Amplatzer PFO Occluder in correct position, straddling the thick wedge (asterisk) of the septum secundum. Device 2 is an cribriform Amplatzer Occluder, specially designed to cover multiple defects.

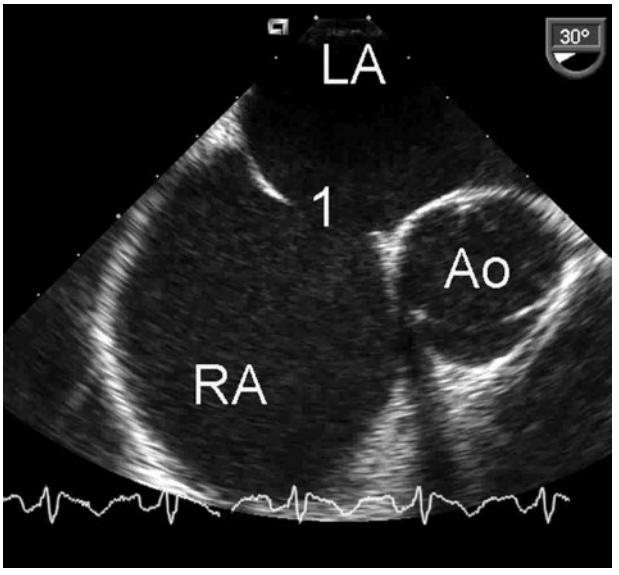

Figure 3. Multiplane transesophageal echocardiography showing a moderate-sized ostium secundum ASD (1) in a 30° view. Ao indicates ascending aorta; LA, left atrium; and RA, right atrium.

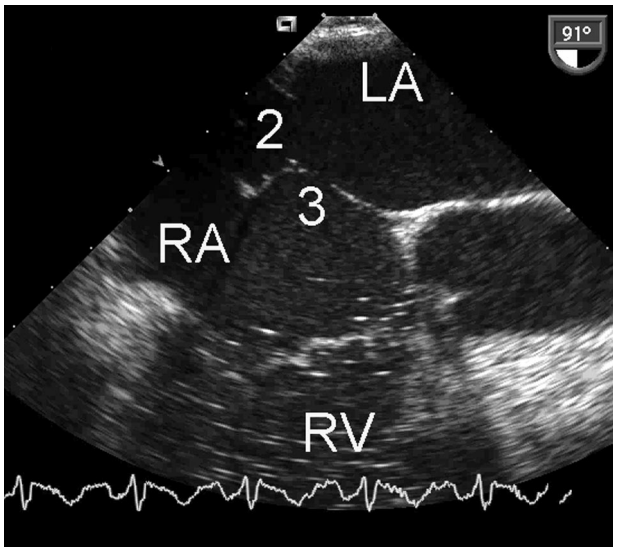

Figure 4. After pushing the echo probe into a more inferior position and rotating it posteriorly, 2 additional septal defects (2 and 3) are seen within the fossa ovalis. Ao indicates ascending aorta; LA, left atrium; and RA, right atrium.

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