

# Three-Dimensional Cardiac Imaging: A Necessity to the Diagnosis and Treatment of Swiss-Cheese Atrial Septal Defect

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## Abstract

Swiss-cheese atrial septal defect (ASD) is a malformation characterized by multi-fenestrated interatrial defects. Here, we describe a vignette of a 23-year-old man with Swiss-Cheese ASD characterized by two defects with areas of 0.74 cm<sup>2</sup> and 0.44 cm<sup>2</sup>, complicated with an atrial septal aneurysm successfully repaired with a cribriform amplatzer septal occluder (ASO) via the percutaneous transcatheter approach. This case emphasizes the importance of attaining a clear view of three-dimensional structures for proper device selection and deployment in repair as additional structural defects such as concomitant aneurysms impose significant challenges.

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**Categories:** Cardiology

**Keywords:** atrial septal defect (asd) diagnosis, atrial septal defect, percutaneous transcatheter repair, cardiology, three-dimensional echocardiography

## Introduction

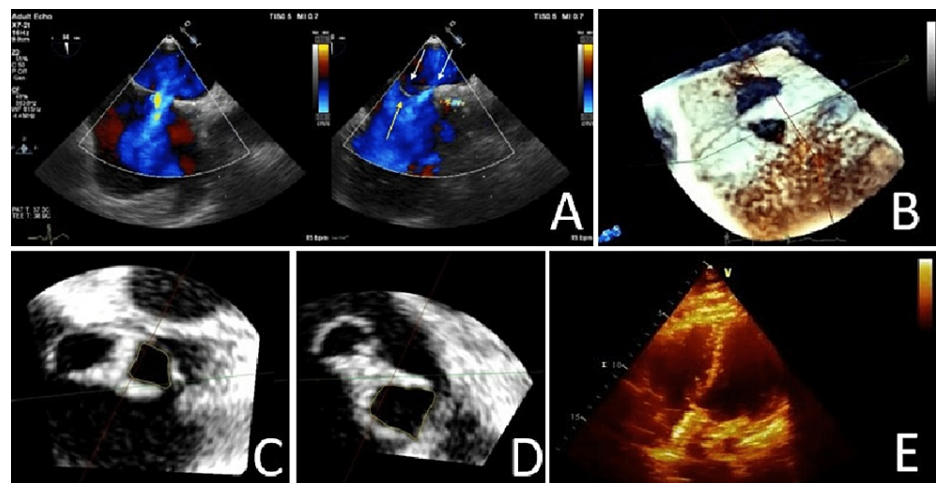
Swiss-cheese atrial septal defect (ASD) is a structural finding of the heart used to characterize multiple or fenestrated interatrial defects found in approximately 10% of patients with ASD [1]. The presence of an aneurysm in the atrial septum is an additional malformation associated with fenestrated ASD that further increases the risk of arrhythmia and thromboembolic events [1-2]. Detailed anatomy of the interatrial septum through two- and three-dimensional imaging serves as diagnostic and therapeutic guidance [3]. Current available treatment alternatives include a percutaneous transcatheter approach and open surgical repair. The open surgical repair exhibits similar efficacy but is associated with more complications and is only preferable in the setting of ostium primum, sinus venosus, and coronary sinus defects [4].

## Case Presentation

A 23-year-old man with a history of malignant migraine and two prior episodes of transient ischemic attacks presented for an elective closure of his suspected patent foramen ovale based on a prior bubble study and negative neurological workup. A prior transthoracic echocardiogram (TTE) revealed an interatrial shunt based on a positive bubble study, with right atrial enlargement, and mild tricuspid regurgitation. A subsequent two-dimensional transoesophageal echocardiogram (TEE) confirmed a left to right shunt with two ASDs and an atrial septal aneurysm (Figure 1A). A three-dimensional TEE confirmed these findings and measured the two interatrial defects with areas of 0.74 cm<sup>2</sup> and 0.44 cm<sup>2</sup> (Figure 1B-D). The patient subsequently underwent successful placement of a 35-mm cribriform amplatzer septal occluder (ASO) closure device. TTE at a two-month follow-up revealed a well-positioned closure device without observed shunting (Figure 1E).

### How to cite this article

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**FIGURE 1: Echocardiography images**

1A. Two-dimensional TTE with Doppler demonstrated two interatrial septal defects (white arrows) and a septal aneurysm (yellow arrow).

1B. Three-dimensional TEE demonstrated two interatrial septal defects.

1C. Three-dimensional TEE demonstrated one ASD measured at 0.44 cm<sup>2</sup>.

1D. Three-dimensional TEE demonstrated one ASD measured at 0.74 cm<sup>2</sup>.

1E. Post-procedural TTE demonstrated stable positioned occluder and septum.

## Discussion

A clear three-dimensional analysis of these structures is crucial for the proper deployment of ASO and Gore Helex Septal Occluder, which are safe and effective percutaneous transcatheter devices used in the closure of fenestrated ASDs [5]. ASO is a widely used device comprised of heavy metallic structures on both sides that permanently fuse with the atrial septal wall, whereas the Gore Helex Septal Occluder is softer with less metallic content that can close a defect up to 17 mm in diameter [6]. However, the percutaneous transcatheter approach with either device is challenging with additional structural limitations of an atrial septal aneurysm, deficient ASD rims, and long inter-defect distances [1]. The atrial septal aneurysm may not provide sufficient rims for implantation of standardized ASD occluder, and, thus, complete closure without residual shunts is difficult to achieve without the use of multiple devices [1]. Current literature supports that a single cribriform device is preferred as the large discs stabilize the aneurysmal septum, and the device itself does not rely on the septum for position stability [2,7]. Cribriform ASO device is available in 18, 25, or 35 mm sizes, and its use had successfully repaired 13 out of 16 cases of fenestrated ASD with aneurysmal atrial septum in one study [2]. In comparison to the surgical approach, percutaneous transcatheter repair is less invasive and requires less time for convalescence. Surgery is indicated in the setting of individual preference to avoid foreign device implantation, large ASD defect size, or unfavorable rim structural limitations for any available device [8]. The utility of three-dimensional imaging is that the limited orthogonal-planar view of two-dimensional imaging may fail to reveal additional fenestrations until residual shunts are observed in the post-deployment of a standardized occlude device [9]. Future studies to investigate the disparities of defect size between two- and three-dimensional imaging are warranted. Our case aims to increase clinician's awareness of Swiss-Cheese ASD complicated by additional structural defects such as the aneurysmal atrial septum, the necessity of a three-dimensional analysis to guide proper device selection, and highlights the promising outcome in closure with the single Cribriform ASO device.

## Conclusions

Swiss-Cheese ASD is an uncommon malformation of multi-fenestrated interatrial defects. Evaluation with three-dimensional echocardiography prior to repair is critical to assess for additional structural defects that can limit proper device deployment. Repairing with the cribriform ASO device via the percutaneous transcatheter approach is an effective method to repair Swiss-Cheese ASD complicated with an atrial septum aneurysm.

## Additional Information

## Disclosures

**Human subjects:** Consent was obtained or waived by all participants in this study. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** This article was previously posted to the medRxiv preprint server on January 25, 2023.

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