

# Filling the Holes of a Swiss Cheese Atrial Septal Defect: A Case Report and Review of the Literature

Andrew S. Kao<sup>1</sup> and Shaun Cardozo<sup>1</sup>

<sup>1</sup>Wayne State University School of Medicine

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## Abstract

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.<sup>1</sup> 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.<sup>1-2</sup> Detailed anatomy of the interatrial septum through 2D and 3D imaging serves as diagnostic and therapeutic guidance.<sup>3</sup> We describe a case of Swiss Cheese ASD complicated with an atrial septal aneurysm successfully repaired with a Cribriform Amplatzer Septal Occluder (ASO) via percutaneous transcatheter approach.

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**Authors:** Andrew S. Kao<sup>1</sup>, Shaun Cardozo MD<sup>1,2</sup>

Wayne State University School of Medicine

Detroit Medical Center, Department of Cardiology

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## Corresponding Author

Shaun Cardozo, MD

Assistant Professor of Medicine

Detroit Medical Center, Department of Cardiology

Wayne State University School of Medicine

Phone: 888-362-5000

Email: [scardozo@med.wayne.edu](mailto:scardozo@med.wayne.edu)

## Abstract:

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.<sup>1</sup> 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.<sup>1-2</sup>Detailed anatomy of the interatrial septum through 2D and 3D imaging serves as diagnostic and therapeutic guidance.<sup>3</sup> We describe a case of Swiss Cheese ASD complicated with an atrial septal aneurysm successfully repaired with a Cribriiform Amplatzer Septal Occluder (ASO) via percutaneous transcatheter approach.

## Case Presentation:

A 23-year-old male 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. 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 2D transesophageal echocardiogram (TEE) confirmed two atrial septal defects and left to right shunt (Figure 1A). 3D TEE showed an aneurysmal atrial septum and multiple defects with areas of 0.74 cm<sup>2</sup> and 0.44 cm<sup>2</sup> (Figure 1B-D). The patient subsequently underwent a successful placement of a 35-mm Cribriiform Amplatzer Septal Occluder (ASO) closure device. TTE at a 2-month follow-up revealed a well-positioned closure device without observed shunting (Figure 1E).

## Discussion:

Clear three-dimensional analyzation of these structures is crucial for proper deployment of ASO and Gore Helex Septal Occluder, which are safe and effective percutaneous transcatheter devices used in closure of fenestrated ASDs.<sup>4</sup> However, percutaneous transcatheter approach is challenging with additional structural limitations of atrial septal aneurysm, deficient ASD rims, and long inter-defect distances.<sup>1</sup> The atrial septal aneurysm may not provide sufficient rims for implantation of standardized ASD occluder and complete closure without residual shunts is difficult to achieve without the use of multiple devices.<sup>1</sup> Current literature supports that a single cribriiform device is preferred as the large discs stabilizes the aneurysmal septum, and the device itself does not rely on the septum for position stability.<sup>2,5</sup> Cribriiform 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.<sup>2</sup> Our case aims to increase clinician's awareness of Swiss Cheese ASDs complicated by additional structural defects such as aneurysmal atrial septum, the necessity of 3D imaging in analysis, and highlights the promising outcome in closure with the single Cribriiform ASO device.

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## Figure Legends:

- 1A. Two-dimensional transthoracic echocardiography with Doppler demonstrated two interatrial septal defects (white arrows) and a septal aneurysm (yellow arrow).
- 1B. Three-dimensional transesophageal echocardiography demonstrated two interatrial septal defects.
- 1C. Three-dimensional transesophageal echocardiography demonstrated one atrial septal defect measured to 0.44 cm<sup>2</sup>.

1D. Three-dimensional transesophageal echocardiography demonstrated one atrial septal defect measured to 0.74 cm<sup>2</sup>.

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

