# CONGENITAL HEART DISEASE NEVER TOO YOUNG OR TOO OLD TO BE DIAGNOSED WITH CONGENITAL HEART DISEASE

# Limitations of Percutaneous Closure of a Complex Secundum Atrial Septal Defect



M. Scott Binder, MD, Isabel E. Binder, RDCS, and Jason R. Foerst, MD, FACC, Roanoke, Virginia

### INTRODUCTION

The first description of nonoperative closure of atrial septal defects (ASDs) occurred in 1976 from Mills and King, utilizing umbrella closure devices.<sup>1</sup> Devices, indications, and contraindications to percutaneous device closure have continued to evolve since that time. Absolute contraindications to ASD closure include pulmonary vascular resistance exceeding 8 Wood units. There are also multiple aspects of ASDs that can predict a complex closure, functioning as relative contraindications.<sup>2</sup>

Complex ASDs have multiple characteristics, but the first consideration is typically size, with cutoffs of 36-38 mm cited frequently.<sup>2,3</sup> Although one diameter is commonly reported in two-dimensional (2D) pictures, ASDs are usually not perfect circles; ovals, multiple defects, or racquet shapes have all been reported.<sup>4</sup> Additionally, guidelines have defined six rims around the ASD, and deficiencies (<5 mm length) in any of these rims can further complicate closures.<sup>5</sup> The rims are defined by their adjacent structures to include the aortic rim (superoanterior), atrioventricular valve rim (inferoanterior), superior vena cava rim (superoposterior), inferior vena cava rim (inferoposterior), posterior rim, and right upper pulmonary vein rim.<sup>5,6</sup> Rims are necessary for structural support of right and left atrial discs to ensure an adequate seal.

We present the case of a large complex ASD with absent aortic rim, resulting in necessitation of surgical closure.

### CASE PRESENTATION

A 60-year-old man presented to the pulmonary hypertension (PH) clinic at our institution with worsening exertional dyspnea over a 2-year period. He had a medical history of human immunodeficiency virus (HIV), hypertension, and latent tuberculosis (with prior 25-year incarceration) as well as risk factors for obstructive sleep apnea (OSA). He had noticed worsening dyspnea on exertion, fatigue, and weight gain of 30 pounds over the past 2 years, prompting his referral to

From the Department of Medicine (M.S.B.), Division of Echocardiography (I.E.B.), and Department of Cardiology, Structural Heart Division (J.R.F.), Virginia Tech Carilion Roanoke Memorial Hospital, Roanoke, Virginia.

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Correspondence: M. Scott Binder, MD, 1906 Belleview Avenue SE, Roanoke, VA, 24014 (E-mail: *msbinder@carilionclinic.org*).

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## **VIDEO HIGHLIGHTS**

**Video 1:** A 2D transthoracic zoomed subcostal four-chamber view of the ASD with color compare. Predominant left-to-right shunting through the defect on color flow Doppler is visualized. **Video 2:** A midesophageal 3D TEE zoomed volume-rendered image of the ASD at 135°.

**Video 3:** Color Doppler 2D TEE at 47° showing very little aortic rim tissue present, between the aortic valve and the ASD (the superoanterior aspect). The lack of aortic rim tissue may complicate percutaneous ASD closure but is not an absolute contraindication.

**Video 4:** A 2D midesophageal TEE with color Doppler visualizing residual left-to-right shunting around the 34 mm sizing balloon during maximal inflation, imaged at 65°. Lack of stopflow during maximal inflation mandates a larger device size for successful closure, which was not available at our institution.

**Video 5:** Color Doppler during the operating room 2D midesophageal TEE at 118°, showing no residual shunting across the defect following bovine pericardium patch repair of the ASD.

View the video content online at www.cvcasejournal.com.

the PH clinic. On physical examination, the patient was obese but breathing comfortably and had a 2/6 systolic murmur heard loudest at the left sternal border. No wheezing or crackles were heard on auscultation, and no lower extremity edema was present. The jugular venous pressure was not assessed during this visit.

The patient had previously completed a sleep study, which was consistent with severe OSA (apnea-hyponea index of 2.3 per hour), and had negative serologic studies. He reported good compliance with an autotitration continuous positive airway pressure machine, yet his symptoms persisted. He had previously had pulmonary function tests performed, which showed no signs of restrictive or obstructive lung disease. He had a ventilation/perfusion scan (V/Q) completed, which was a low-probability scan for pulmonary embolism or chronic thromboembolic PH. He was thought to have mixed PH due to OSA, HIV, and obesity. He had a transthoracic echocardiogram (TTE) ordered, which elucidated another potential etiology for his PH.

The TTE demonstrated a large secundum ASD with predominant left-to-right shunting. The ratio of pulmonary to systemic blood flow (Qp/Qs) was 1.9, and the right ventricular systolic pressure was 38 mm Hg. Color flow Doppler across the visualized defect illustrated an approximate size of 2 cm with predominant left-to-right shunting across the defect (Figure 1, Video 1). Given the size of the defect reported on TTE and the patient's desire to avoid surgery, plans for



Figure 1 A subcostal view of the ASD revealing an approximate size of 2.03 cm and predominant left-to-right shunting across the ASD on color flow Doppler.



Figure 2 Three-dimensional TEE image of the secundum ASD, demonstrating an oval shape with very large size,  $38 \times 28$ mm.

percutaneous closure with transesophageal echocardiogram (TEE) guidance were made.

The initial TEE 2D measurements suggested a large secundum defect of 40 mm ( $36 \times 40$  mm). With three-dimensional (3D) imaging, the ASD measured  $38 \times 27$  mm (Figures 2 and 3; Video 2). Although the 2D and 3D measurements correlated well in this example, it is important to understand that 3D measurements, although allowable with new ultrasound systems, should not be considered routine practice or without limitations. Paralax may cause inaccurate measurements on direct 3D volume-rendered displays.

Therefore, these should be considered estimates and are not as accurate as carefully performed 2D measurements obtained using 3D imaging–guided alignment. The TEE also showed very little aortic rim present (Figure 4, Video 3) and a predominantly oval shape. The patient was balloon sized with TEE, but color Doppler demonstrated no evidence of stop flow with maximal inflation of a 34 mm sizing balloon (Abbott, Santa Clara, CA), precluding adequate closure with the largest available ASD closure device at our institution (Figure 5, Video 4). Using the Fick principle, the Qp/Qs ratio was 4.74 on catheterization (pulmonary arterial oxygen saturation, 95.2%; mixed venous oxygen saturation, 80.9%; pulmonary venous oxygen saturation, 99.4%). His pulmonary vascular resistance was normal at 1.15 Wood units.

The patient was referred to cardiac surgery and underwent bovine patch closure of his ASD with no residual shunt present (Figure 6, Video 5) and was discharged home on room air with no complications on postop day 8. At his 1-month follow-up clinic appointment, the patient noted that his breathing and dyspnea on exertion have improved. This has allowed him to increase his exercise capacity and has substantially improved his quality of life.

#### DISCUSSION

Transthoracic echocardiography was essential in discovering one of the underlying causes of this patient's symptoms. Other etiologies certainly may have contributed (HIV, OSA, obesity), but the large size of the defect, late discovery, enlarged right heart, and shunt fraction suggest that this defect played a substantial role.

Three-dimensional imaging of the atrial septum and ASD was crucial in this case, to allow for more accurate size and shape analysis



Figure 3 Two-dimensional TEE midesophageal images of the secundum ASD, measured at 0° (*left*) and 55° (*right*). Maximal orthogonal dimensions were similar at 36 mm and 40 mm, respectively.



Figure 4 Two-dimensional TEE color Doppler imaging, demonstrating very little aortic rim tissue present.

of the ASD and to reassess following balloon inflation. Guidelines have standardized ASD and atrial septum imaging on TEE, and recent studies have shown that accuracy is much greater with 3D TEE imaging when compared with 2D TTE or TEE comparisons.<sup>7,8</sup> Older studies have shown differences in 2D TEE measurements of size compared with balloon measurement during catheterization, so thorough 3D or balloon dilated 2D imaging is essential for accuracy.<sup>9</sup> A study by Johri *et al.*<sup>10</sup> enrolled 24 patients with ASDs. They found that in patients with residual shunting after ASD closure, the size of the defect was 27% larger on 3D imaging than on 2D imaging and that the majority were complex in shape.<sup>10</sup> The same study also found that 3D measurements correlated well with 2D balloon-stretched dimensions, and some single-center studies have questioned whether balloon sizing is still necessary, but further research is needed to clarify this point.<sup>10,11</sup>

The deficient aortic rim was another factor complicating percutaneous closure in this patient, along with the very large size and



Figure 5 Percutaneous closure attempt of the ASD, with residual shunting (*white arrow*) visualized around the 34 mm sizing balloon during maximal inflation (*red arrow*). *LA*, Left atrium; *RA*, right atrium.

irregular shape. Studies have shown decreased immediate response rates with deficient rims (74% vs 92%).<sup>12</sup> The location of the defect can allow some deficiencies to be overcome, but superior and posterior rim deficiencies have been associated with increased procedural failure.<sup>12-14</sup> Mathewson *et al.*<sup>13</sup> studied 76 patients with complex ASD closures who had absent aortic or inferior vena cava rims and found that absent aortic rims could be closed with special techniques, but monitoring for disc prolapse and ensuring perpendicular orientation was obligatory before deployment. Kijima *et al.*<sup>15</sup> found that even though procedural success was lower with multiple rim deficiencies, if deployed successfully, there is no difference in cardiovascular event occurrence over a mean of 2 years of follow-up. Had a larger device size been available at our institution, closure of this complex defect, even with an absent aortic rim, may have been possible.



Figure 6 Color Doppler after bovine pericardial patch repair of the ASD, with no residual shunt visualized.

#### CONCLUSION

We presented the case of a patient with an extremely large ASD ( $38 \times 28$  mm) with associated aortic rim deficiency and irregular oval shape. Percutaneous closure under TEE guidance was attempted, but the large size precluded adequate percutaneous intervention.

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#### SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi. org/10.1016/j.case.2022.03.008.

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