

# Large Atrial Septal Aneurysm Associated with Secundum Atrial Septal Defect



Nino Isakadze, MD, MHS, Jana Lovell, MD, Edward P. Shapiro, MD, Chun W. Choi, MD, Marlene S. Williams, MD, and Monica Mukherjee, MD, MPH, *Baltimore, Maryland*

## INTRODUCTION

Atrial septal aneurysm (ASA) is a congenital or acquired outpouching of the atrial septum. It is an incidental finding in 1% to 2.5% of the general population.<sup>1-3</sup> Although the clinical significance of isolated ASA is not certain,<sup>4</sup> it is frequently associated with the presence of other septal abnormalities, including patent foramen ovale (PFO) and atrial septal defect (ASD).<sup>4-6</sup> When associated with atrial septal abnormalities, ASA is associated with an increased risk for arterial embolism<sup>4</sup> and recurrent stroke.<sup>7</sup> Although very large ASAs are exceedingly rare, further workup and management pathways are not well defined.<sup>8</sup> Here, we describe an unusual case of a large symptomatic ASA associated with secundum ASD.

## CASE PRESENTATION

A 25-year-old woman with antineutrophil cytoplasmic antibody vasculitis complicated by pulmonary hemorrhage and polyangiitis, gastritis, and essential hypertension presented with intermittent pleurisy and postural palpitations, worsened when lying on her left side. She noted that symptoms were reduced after biannual rituximab treatments but progressively worsened a few months afterward, and subjectively, her symptoms correlated with increasing inflammatory markers. Her physical examination was unremarkable. Initial evaluation performed at an outside hospital included transthoracic echocardiogram (TTE) demonstrating a large ASA (images not available) and cardiac computed tomography (CCT), which did not show vascular abnormalities, vasculitis, or obstructive coronary disease. Demonstration on CCT of the large ASA is shown in [Figure 1](#), without evidence of interatrial shunting.

The patient also underwent cardiovascular magnetic resonance imaging (CMR), which showed a mildly reduced left ventricular ejection fraction at 51% (reference range, 57%-77%).<sup>8</sup> Additionally, there was no evidence of acute or chronic

myocarditis or pericarditis. Native T1 mapping values were mildly higher than the normal range, but the calculated extracellular volume using the modified Look-Locker inversion recovery technique was within the normal limits, suggesting no evidence of diffuse myocardial fibrosis. Normal T2 mapping values indicated no evidence of regional or global myocardial edema. On delayed enhancement imaging, there was no evidence of prior myocardial infarction or focal fibrosis. However, CMR also demonstrated a large ASA that was highly mobile and measured 2.6 cm at the orifice and 2.8 cm in length. No large interatrial shunt was observed. Given her episodic palpitations, she also underwent rhythm monitoring for 12 days, which did not show tachy- or bradyarrhythmias.

Given these findings, the patient then presented for a second opinion for further evaluation, and given the history of large ASA, TTE was performed with agitated saline to further evaluate for the presence of PFO and/or ASD. The patient had normal right atrial size, normal right ventricular size and function, and an echocardiography-derived shunt fraction of 0.7, suggestive of right-to-left shunting. Further details of right heart measurements are shown in [Table 1](#).

TTE demonstrated large mobile ASA with minimal respirophasic variation, protruding into the right atrium and measuring 2.9 cm in length and 2.0 cm in diameter. The ASA remained fixed in a rightward orientation, suggesting that a right-to-left shunt would be less likely to be demonstrated. As such, agitated saline study was indeterminate, and although no interatrial shunting was visualized with injection of agitated saline, maneuvers such as Valsalva and similar physiologic maneuvers (e.g., abdominal thrust, cough, sniff) to alter interatrial gradients that may have improved the sensitivity of right-to-left shunt were not performed ([Figure 2](#), [Videos 1 and 2](#)).

The patient was subsequently referred to cardiothoracic surgery for further evaluation and consideration of surgical intervention because of concern that her persistent postural palpitations were due to her large ASA. Transesophageal echocardiography (TEE) was recommended to further define anatomy and evaluate for the presence of interatrial shunt and confirmed the presence of a large ASA protruding into the right atrium, measuring up to 2.5 cm in length and 2.0 cm in diameter ([Video 3](#)). The septal tissue was redundant with multiple fenestrations ([Videos 3 and 4](#)) and evidence of right-to-left shunting with injection of agitated saline, as shown in [Video 5](#). No evidence of shunt was seen with application of color Doppler, but visualization may have been improved by lowering the Nyquist baseline velocities to  $\leq 40$  cm/sec. Three-dimensional and biplane imaging was also concerning for an ASD, as evidenced by the immobile tissue defect seen at the base of the ASA at the superior aspect of the interatrial septum ([Video 4](#)).

Using a shared decision-making approach and careful discussions among the patient, cardiology, and cardiothoracic surgery, the patient opted to proceed with surgical resection of the large ASA. Intraoperative in vivo evaluation of the interatrial septum revealed a

From the Division of Cardiology, Department of Medicine, Johns Hopkins University, Baltimore, Maryland (N.I., J.L., E.P.S., M.S.W., M.M.); and the Division of Cardiothoracic Surgery, Department of Surgery, Johns Hopkins University, Baltimore, Maryland (C.W.C.).

Keywords: Atrial septal defect, Secundum atrial septal defect, Atrial septal aneurysm, Echocardiography

Conflicts of interest: The authors report no conflicts of interest relative to this document.

Correspondence: Monica Mukherjee, MD, MPH, Division of Cardiology, Johns Hopkins University, 301 Mason Lord Drive, Suite 2400, Baltimore, MD 21224. (E-mail: [mmukher2@jh.u.edu](mailto:mmukher2@jh.u.edu)).

Copyright 2022 by the American Society of Echocardiography. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

2468-6441

<https://doi.org/10.1016/j.case.2022.02.003>

## VIDEO HIGHLIGHTS

**Video 1:** Two-dimensional transthoracic imaging of mobile ASA. Two-dimensional TTE, four-chamber apical view, showing a large (2.9 cm in length and 2.0 cm in diameter), highly mobile ASA protruding into the right atrium throughout the cardiac cycle.

**Video 2:** Two-dimensional transthoracic imaging of mobile ASA with agitated saline. TTE, four-chamber apical view, showing the agitated saline study, without evidence of interatrial shunting. However, it is important to note that further physiologic maneuvers such as Valsalva were not performed, making the findings indeterminate rather than negative.

**Video 3:** Three-dimensional transesophageal imaging of mobile ASA. Three-dimensional TEE showing a large ASA attached to the interatrial septum, which is protruding into the right atrium throughout the cardiac cycle. Multiple small fenestrations are visible throughout the redundant tissue. A tissue defect is seen at the base of the ASA, concerning for a secundum ASD. Images are acquired from the midesophageal position at 100° omniplane angle using three-dimensional multibeat acquisition.

**Video 4:** Three-dimensional transesophageal imaging of mobile ASA. Three-dimensional TEE and X-plane showing redundant and aneurysmal atrial septal tissue with multiple fenestrations. Although the ASA is large and mobile, biplane imaging demonstrates respirophasic variation and predominant positioning into the right atrium throughout the cardiac cycle. Images are acquired from the midesophageal position at 100° with biplane and simultaneous three-dimensional imaging.

**Video 5:** Two-dimensional transesophageal imaging of mobile ASA with agitated saline. Two-dimensional TEE showing agitated saline study demonstrating bubbles crossing the interatrial septum with right-to-left shunting. Bubble passage is noted at the fourth cardiac cycle after contrast appearance. Biplane imaging is obtained using the X-plane feature from the midesophageal position at 100°.

View the video content online at [www.cvcasejournal.com](http://www.cvcasejournal.com).

large ASA with multiple fenestrations throughout, as well as a large secundum ASD located at the base of the ASA in the area of the fossa ovalis. After the aneurysmal sac was resected, the large secundum ASD measured 3.5 × 2.0 cm, and a bovine pericardial patch was subsequently sewn at the site (Figures 3A and 3B).

Postoperatively, the patient recovered from surgery well and on follow-up stated that she was no longer experiencing symptoms of palpitations and was feeling “wonderful.” Although the resolution of her symptoms suggests that her clinical presentation was likely related to the ASA and secundum ASD, we cannot entirely exclude that her symptoms were unrelated. TTE 7 weeks after surgery showed a repaired interatrial septum with a minimal residual ASD demonstrated by Doppler and an agitated saline study, which will likely resolve over time.

**Table 1** Right heart chamber size and function

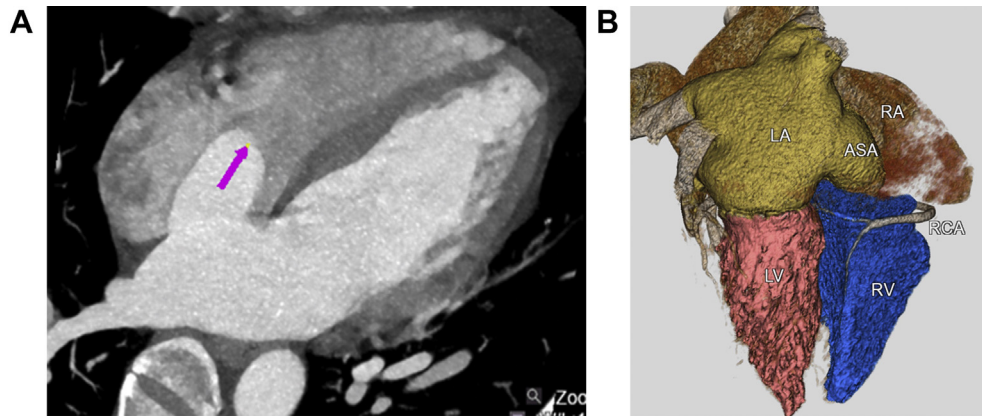
Echocardiographic parameter	Patient measurements
RVIDD, cm	2.23
RVOT, cm	1.90
Right atrial area, cm <sup>2</sup>	14.7
Right atrial volume indexed to BSA, mL/m <sup>2</sup>	19.0
Basal dimension, cm	3.70
Midventricular dimension, cm	2.20
Longitudinal dimension, cm	8.27
RVEDA, cm <sup>2</sup>	18.4
RVESA, cm <sup>2</sup>	8.44
Fractional area change, %	54.1
TAPSE, cm	2.44
Q <sub>p</sub> /Q <sub>s</sub>	0.70

BSA, Body surface area; Q<sub>p</sub>/Q<sub>s</sub>, ratio of pulmonary to systemic blood flow; RVEDA, right ventricular end-diastolic area; RVESA, right ventricular end-systolic area; RVIDD, right ventricular internal diastolic dimension; RVOT, right ventricular outflow tract; TAPSE, tricuspid annular plane systolic excursion.

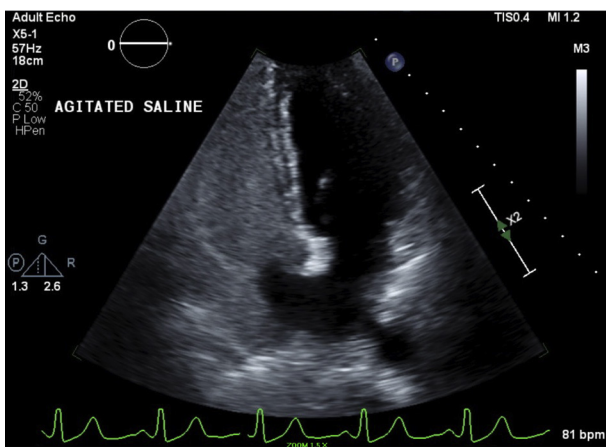
Right heart chamber size and function derived from transthoracic echocardiographic parameters are shown.

## DISCUSSION

ASDs represent a continuum of persistent interatrial communications and are distinct from a PFO, in which a flap is present and there is intermittent communication, typically with respirophasic variation.<sup>5</sup> The echocardiographic guideline criterion for defining ASA is >1.0-cm excursion of the septal tissue into the right or left atrium from the atrial septal plane or total right or left excursion of 1.5 cm.<sup>9</sup> Secundum ASDs represent the vast majority and are positioned in the fossa ovalis. Less common are ostium primum ASDs, located along the inferior pole of the interatrial septum, and sinus venosus defects, positioned near the caval entry. The presence of both septum primum<sup>2</sup> and septum secundum<sup>3,10</sup> ASDs has previously been reported with ASA. One study showed a higher prevalence of ASDs when ASAs involve the entire atrial septum, but a more recent, larger study showed a similar prevalence of ASDs when ASAs involve the entire septum or fossa ovalis alone.<sup>3,10</sup> Olivares-Reyes *et al.*<sup>11</sup> described five distinct subtypes of ASAs on the basis of location during cardiac cycle. Further subtypes include 2L, in which the ASA protrudes toward the left atrium throughout the cardiac cycle; 3RL, in which there is more excursion toward the right atrium than the left atrium; 4LR, in which there is more excursion over the left atrium than the right atrium; and 5, in which bidirectional movement is present, with similar distance toward right and left atria. Our patient had a type 1R ASA, as it protruded into the right atrium throughout the cardiac cycle, with minimal respirophasic variation. ASAs may be difficult to visualize, as they tend to be highly mobile and occur within the thin region of the fossa ovalis. Furthermore, without dedicated imaging to evaluate for shunting and significant technical expertise in interpretation, large and highly mobile ASAs, such as in our case, may be missed. In fact, only with TEE were we able to truly appreciate the extent of the ASA and the evidence of



**Figure 1** CCT of ASA. Cardiac computed tomographic images demonstrate redundant atrial septal tissue with a large ASA protruding into the right atrium (RA). **(A)** Multiplanar reconstruction, oblique axial orientation, apical four-chamber display of the interatrial septum. Temporal resolution of CCT results in a degree of blurring of the image at the tip of the ASA, and grossly, no interatrial shunt is visualized. **(B)** Three-dimensional volume-rendered reconstruction image of the whole heart, highlighting the large ASA. LA, left atrium; LV, left ventricle; RCA, right coronary artery; RV, right ventricle.

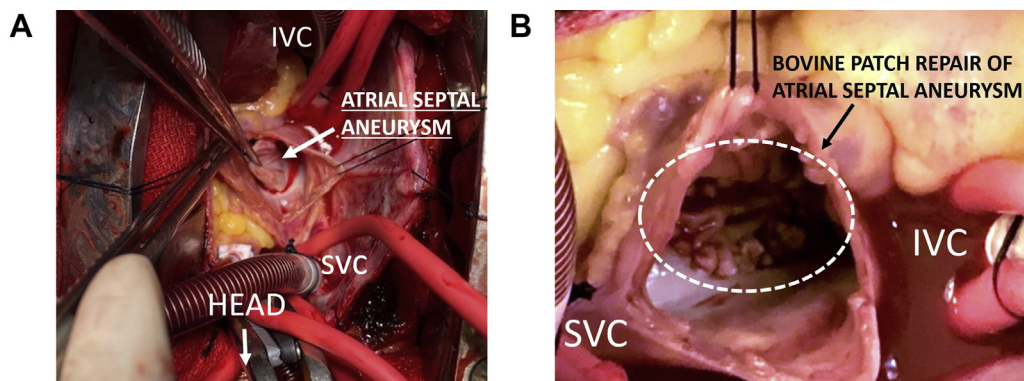


**Figure 2** Apical four-chamber view of ASA. A still frame of the apical four-chamber view on TTE is shown. Following agitated saline injection, there is evidence of a well-delineated large ASA protruding into the right atrium.

interatrial shunting and concern for a secundum ASD, as evidenced by a tissue defect at the base of the ASA on biplane imaging. Direct inspection at the time of surgical resection led to appreciation of multiple fenestrations within the ASA and a large tissue defect at the base of the ASA in the area of the fossa ovalis, consistent with secundum ASD. Visualization of secundum ASD on two-dimensional TTE has sensitivity of 80%.<sup>12</sup> Presence of right-sided cardiac overload along with a mobile atrial septum on TTE can be an initial clue for the presence of ASD and can be further evaluated using color flow Doppler, which can help visualize persistent interatrial shunting.<sup>5</sup> In contrast, PFOs are characterized by left-to-right shunting without the presence of a true tissue defect in the interatrial septum, and they are located mostly in the anterior or superior portion of the fossa ovalis. Further evaluation

and quantification of shunting can be performed using pulsed-wave<sup>13</sup> and continuous-wave Doppler imaging to obtain further measurements, including the direction, velocity, and timing of the flow.<sup>9</sup> Application of color Doppler with scale reduction of the Nyquist limit to 35 to 40 cm/sec can help in capturing low-velocity flow across small fenestrations. If a suspected ASD is not well visualized on two-dimensional imaging with color Doppler, further investigation with agitated saline along with physiologic maneuvers to increase right-to-left shunting should be considered. This technique has almost 100% sensitivity to identify ASDs<sup>14</sup> and further highlights the importance of adopting and strictly adhering to specific guideline-driven protocols for interatrial septal imaging, such as dedicated two-dimensional and three-dimensional biplane imaging, Nyquist limit adjustment, and physiologic maneuvers during agitated saline injection. In our case, although agitated saline study was performed during TTE, the size and mobility of the ASA prevented visualization of significant interatrial shunt. However, TEE has advantages and provides better definition and visualization of the interatrial septum, and it is helpful when TTE is not diagnostic. TEE further aids with potential procedural planning, as it allows comprehensive visualization of the interatrial structure and anatomy, especially if three-dimensional imaging is implemented.<sup>5</sup>

Tzimas and Papadopoulos<sup>7</sup> reported a case of a large ASA that was not visualized on TTE in a patient undergoing coronary artery bypass grafting surgery for severe left anterior descending coronary artery stenosis. After induction of anesthesia, TEE visualized the ASA with a PFO filled with thrombus, which was dislodged during the case, resulting in multiple embolic events and a fatal outcome. Kumagai *et al.*<sup>15</sup> also described a case of a large ASA seen on CCT and CMR, which was incidentally found in a patient undergoing evaluation for chest pain. Imaging demonstrated a 3.0 × 3.0 cm ASA protruding into the right atrium along with thrombus. Given the presence of thrombus, the patient underwent surgical resection of the ASA. The same team also reported a case of an ASA associated with a PFO, and in this case, an interatrial shunt was visualized on TTE. TEE also showed the ASA and PFO, trivial tricuspid regurgitation, and a gradient. Thus, CCT and CMR were performed and subsequently



**Figure 3** Surgical pathology of ASA and secundum ASD. Gross images of surgical pathology visualizing large ASA before and after surgical repair. **(A)** Direct visualization of the large ASA. **(B)** Bovine patch repair of the secundum ASD noted at the base of the ASA.

showed a large ASA with associated thrombus. This patient also ultimately underwent surgical ASA resection.

Although the data are very sparse, our case, along with a prior case report,<sup>8</sup> suggests that TTE, CCT, or CMR may fail to demonstrate the presence of interatrial shunts, especially when the size of an aneurysm is large. Furthermore, the risk for thromboembolic complications is not well established for individuals with large ASAs and interatrial shunts. Surgical or interventional closure of a fenestrated ASD may require medical optimization to avoid the development of acute left heart failure in older individuals with increased left-to-right shunting. Fortunately, and likely because of her age, our patient did not have right-sided cardiac overload and adverse remodeling of the right heart chambers.<sup>9</sup> Thus, determination for surgical management should be made on an individual basis with shared decision making with the patient and clinical team.

## CONCLUSION

We demonstrate in the present case that TEE is superior to other imaging modalities (TTE, CCT, and CMR) in detecting presence of an intra-atrial shunt, although the degree of interatrial communication may be underestimated. Large ASAs are frequently accompanied by concomitant defects of the atrial septum, and close interrogation during TEE allows anatomic delineation.

## SUPPLEMENTARY DATA

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.case.2022.02.003>.

## REFERENCES

- Di Tullio MR, Sacco RL, Sciacca RR, Jin Z, Homma S. Patent foramen ovale and the risk of ischemic stroke in a multiethnic population. *J Am Coll Cardiol* 2007;49:797-802.
- Silver MD, Dorsey JS. Aneurysms of the septum primum in adults. *Arch Pathol Lab Med* 1978;102:62-5.
- Mugge A, Daniel WG, Angermann C, Spes C, Khandheria BK, Kronzon I, et al. Atrial septal aneurysm in adult patients. A multicenter study using transthoracic and transesophageal echocardiography. *Circulation* 1995; 91:2785-92.
- Belkin RN, Waugh RA, Kisslo J. Interatrial shunting in atrial septal aneurysm. *Am J Cardiol* 1986;57:310-2.
- Martin SS, Shapiro EP, Mukherjee M. Atrial septal defects—clinical manifestations, echo assessment, and intervention. *Clin Med Insights Cardiol* 2014;8(suppl 1):93-8.
- Turc G, Lee JY, Brochet E, Kim JS, Song JK, Mas JL, et al. Atrial septal aneurysm, shunt size, and recurrent stroke risk in patients with patent foramen ovale. *J Am Coll Cardiol* 2020;75:2312-20.
- Tzimas P, Papadopoulos G. Huge interatrial septal aneurysm: a coincidental but rather fatal finding. *Case Rep Med* 2011;2011:705260.
- Petersen SE, Khanji MY, Plein S, Lancellotti P, Bucciarelli-Ducci C. European Association of Cardiovascular Imaging expert consensus paper: a comprehensive review of cardiovascular magnetic resonance normal values of cardiac chamber size and aortic root in adults and recommendations for grading severity. *Eur Heart J Cardiovasc Imaging* 2019;20:1321-31.
- Silvestry FE, Cohen MS, Armsby LB, Burkule NJ, Fleishman CE, Hijazi ZM, et al. Guidelines for the echocardiographic assessment of atrial septal defect and patent foramen ovale: from the American Society of Echocardiography and Society for Cardiac Angiography and Interventions. *J Am Soc Echocardiogr* 2015;28:910-58.
- Hanley PC, Tajik AJ, Hynes JK, Edwards WD, Reeder GS, Hagler DJ, et al. Diagnosis and classification of atrial septal aneurysm by two-dimensional echocardiography: report of 80 consecutive cases. *J Am Coll Cardiol* 1985;6:1370-82.
- Olivares-Reyes A, Chan S, Lazar EJ, Bandlamudi K, Narla V, Ong K. Atrial septal aneurysm: a new classification in two hundred five adults. *J Am Soc Echocardiogr* 1997;10:644-56.
- Shub C, Dimopoulos IN, Seward JB, Callahan JA, Tancredi RG, Schattenberg TT, et al. Sensitivity of two-dimensional echocardiography in the direct visualization of atrial septal defect utilizing the subcostal approach: experience with 154 patients. *J Am Coll Cardiol* 1983;2:127-35.
- Vick GW III, Murphy DJ Jr, Ludomirsky A, Morrow WR, Morriss MJ, Danford DA, et al. Pulmonary venous and systemic ventricular inflow obstruction in patients with congenital heart disease: detection by combined two-dimensional and Doppler echocardiography. *J Am Coll Cardiol* 1987;9:580-7.
- Stout KK, Daniels CJ, Aboulhosn JA, Bozkurt B, Broberg CS, Colman JM, et al. 2018 AHA/ACC guideline for the management of adults with congenital heart disease: executive summary: a report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. *J Am Coll Cardiol* 2019;73:1494-563.
- Kumagai M, Nishizawa J, Takehara M, Shinoda E, Minatoya K. Surgical management of a giant atrial septal aneurysm. *Clin Case Rep* 2021;9:e04125.