Atrial septal defect and concomitant significant pulmonic stenosis in an adult patient

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Abstract

An adult male had persistent hypoxemia and erythrocytosis after repair of hemodynamically significant pulmonic valve stenosis. A large ostium secundum atrial septal defect was subsequently found as the cause of these symptoms. This sequence of events warrants review of his workup at his initial visit and follow-up visit and of the diagnosis and treatment of his pulmonic stenosis and atrial septal defect. The onset of new symptoms in an adult with a history of congenital heart disease indicates that additional cardiac evaluation is needed. This particular patient had an undiagnosed right-to-left shunt through an atrial septal defect.

Keywords: ostium secundum atrial septal defect, pulmonary valve stenosis, Amplatzer septal occluder, adult

Introduction

Pulmonary valve stenosis (PS) and atrial septal defects (ASD) are common congenital heart defects. While less common in adults, ASDs make up 25 to 30% of congenital heart defects diagnosed in adulthood; 75% of these ASDs are the secundum type.[1,2] Pulmonic stenosis in adults is rarer at approximately 8%–10% of cases with valvular heart disease, and PS and a large ASD occurring simultaneously is even more uncommon.[3] When these conditions occur together, the right-to-left shunt as a result of the outflow obstruction created by the PS can protect the pulmonary vasculature from high pressures into adulthood.[2]

Case

In December 2019, a previously healthy, 39-year-old male immigrant from Honduras was referred for pulmonary balloon valvuloplasty due to significant dyspnea on exertion. Upon initial questioning, the patient disclosed that he had been told as a child in Honduras that he had a "heart problem" and a murmur and was told by those doctors that they could not treat it. His initial labs at presentation to our emergency department showed a hematocrit of 64.6%, and this value increased to >70% in the next 2 days (adjusted normal range at our hospital UCHealth Memorial Central Hospital in Colorado Springs, CO, is 39.2–53.0%). Transthoracic echocardiogram (TTE) at another facility revealed severe pulmonic stenosis, right atrial enlargement, trace tricuspid valve regurgitation, and right ventricular hypertrophy. Cardiac catheterization confirmed these findings and measured a peak gradient across the pulmonic valve of 89 mmHg. The stenosis was successfully treated with a balloon valvuloplasty (Figure 1). The post-procedure pulmonic valve pressure peak gradient was 14 mmHg with no change in tricuspid valve function; a bedside TTE showed trace tricuspid regurgitation (Figure 2).

In August 2021, after 20 months of monthly therapeutic phlebotomy with hematocrit levels ranging 49–60%, the patient was sent to our facility from an infusion center with hypoxemia. Upon arrival to our facility, he had an SpO_2 of 80% on room air and a hematocrit of 51.6%. He had no cardiac complaints

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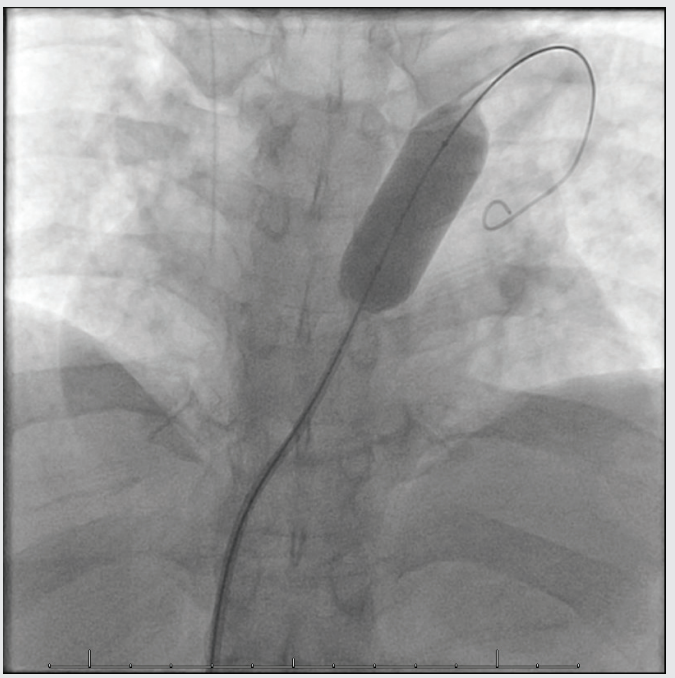

Figure 1. Fluoroscopy image taken during balloon valvuloplasty of pulmonic valve with a 25 × 4 mm balloon fully inflated and resolution of pulmonary valve indentation.

and stated that he had been feeling 'fine.' Up to this point, the patient had had infrequent cardiac or primary care follow up.

Further workup included a TTE that showed no acute abnormality and a stable pulmonic valve without any signs of insufficiency. But shunting was suspected, and a bubble study showed a secundum ASD with large right to left shunt (Figure 3). A transesophageal echocardiogram (TEE) showed only trace tricuspid regurgitation and a large fenestrated ostium secundum ASD with bidirectional shunt (Figure 4).

After discussion of treatment options, the patient underwent percutaneous ASD closure with a 20 mm Amplatzer ASD closure device (Abbott Cardiovascular) (Figure 5). Repeat TEE 12 hours after the procedure showed the device seated securely with no residual shunting and mild tricuspid regurgitation. The patient was discharged the following morning with normal right and left filling pressures and prescribed 81 mg aspirin daily. At a 7-week follow up, he was feeling well. Repeat TEE showed a stable ASD closure device without shunting and mild tricuspid regurgitation (Figure 6). His oxygen saturation was 90% on room air. We recommended discontinuing therapeutic phlebotomy and regular follow up with his primary

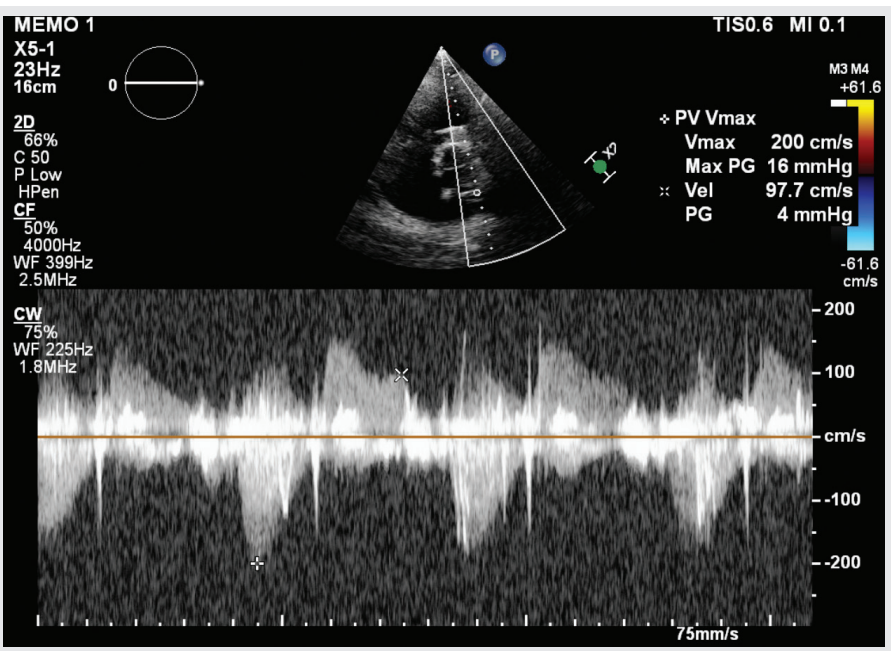

Figure 2. Post-procedure transthoracic echocardiogram showing drastically decreased gradient across pulmonic valve (max PG 16 mmHg).

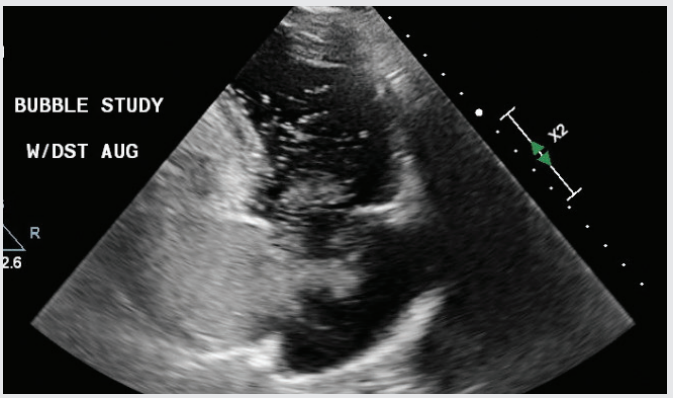

Figure 3. Bubble study with a four-chamber view showing significant right to left shunt across atrial septum.

care physician. A repeat hematocrit was not obtained post-operatively due to the patient's lack of symptoms and continued improvement in oxygen saturation.

The patient followed up with his PCP in December 2021 at an outside facility and was referred to our hematologists for re-evaluation. At his visit in the hematology clinic in January 2022, he was asymptomatic. His SpO_2 was 92% on room air, and laboratory tests showed a hematocrit of 66.2%. The hematologist

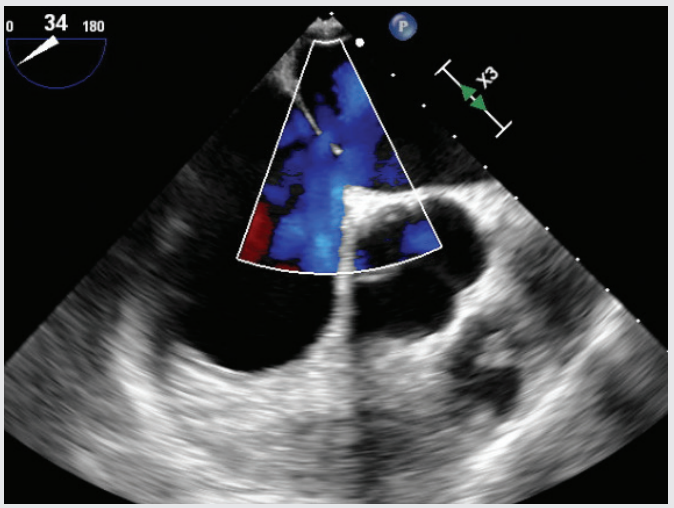

Figure 4. Transesophageal echocardiogram color Doppler displaying shunting through 1.6 cm ostium secundum atrial septal defect with superior fenestration.

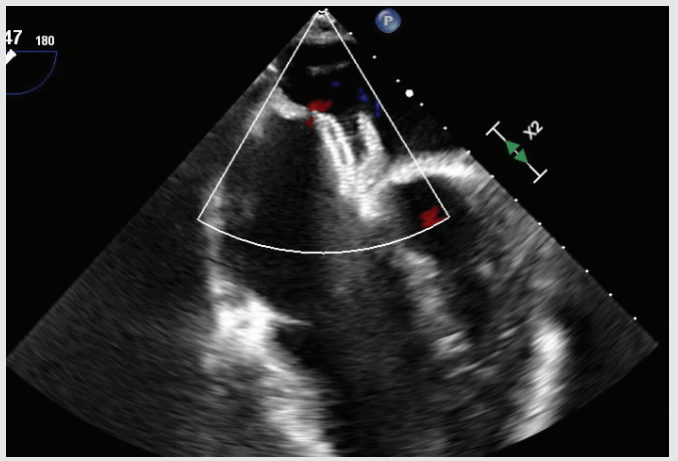

Figure 5. Peri-operative transesophageal echocardiogram showing Amplatzer Septal Occluder in place with mild residual shunt.

recommended restarting therapeutic phlebotomies with periodic blood counts.

Discussion

This patient had late onset of hypoxemia and dyspnea with a known cardiac defect, eventually

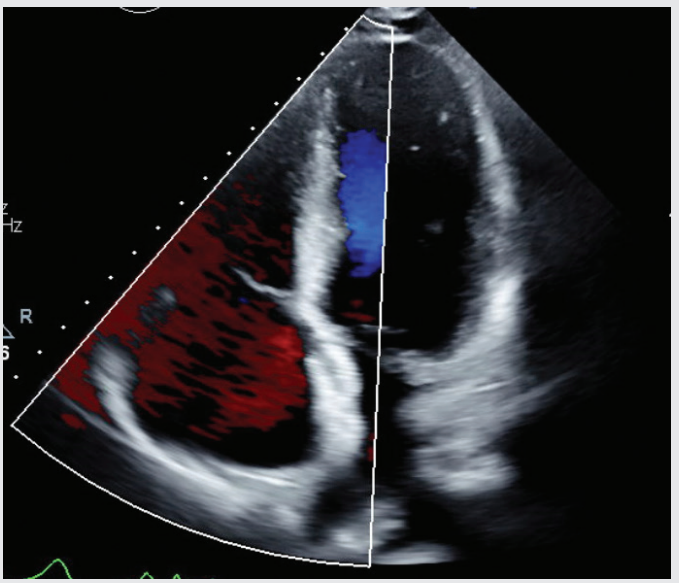

Figure 6. Transesophageal echocardiogram at an 8 week follow up. Amplatzer Septal Occluder device is seated well with no shunting across atrial septum.

diagnosed as PS and ASD. Up until this point, he had been compensating well without any important limitations, so why did he decompensate acutely at the age of 39? We hypothesize that this was due to a recent elevation change. He immigrated from Honduras at an elevation of approximately 2,250 feet to our location in Colorado Springs, CO, at approximately 6,000 feet of elevation, 6 months prior to his initial visit. With acclimation to high altitude, the physiologic response is decreased cardiac output, decreased work of the left ventricle, and increased work of the right ventricle.[4] The increased viscosity of the blood due to his increasing hematocrit likely increased pulmonary pressures, potentially decreasing his right ventricular outflow and increasing his right-to-left shunt.[5] Eisenmenger syndrome was also considered as a possible cause of his right-to-left shunt, but this seemed unlikely given his significant improvement after pulmonic balloon valvuloplasty.[6]

This case demonstrates the difficulty in understanding the potential causes of symptoms in an adult patient with acute on chronic hypoxemia. The patient initially presented with an obvious pulmonic stenosis by TTE. He seemed to be asymptomatic until presenting to our facility. There was little immediate suspicion for shunting given the obvious pulmonic stenosis and the lack of atrial shunting on TTE doppler. Also, with the immediate improvement in symptoms, no ASD was suspected. Following the procedure, the patient had infrequent cardiac follow up. After another acute on chronic hypoxemia episode occurred, his persistent erythrocytosis and low O_2 saturations were worked up, and this led to a diagnosis of a concurrent atrial septal defect.

The secundum ASD was not seen on TTE at the patient's initial presentation and no bubble study was performed during his first hospital stay. His PS was treated, and he improved, but it would have been beneficial for the patient to have his ASD corrected at his initial presentation. There have been several cases and studies showing success with simultaneous percutaneous corrections of secundum type ASDs and PS.[7,8] Based on the severity of the stenosis and symptom onset at 39-years-old, suspicion should be raised about the presence of a possible ASD, which may have helped keep the patient asymptomatic for so long.[2] This case suggests that a bubble study should be considered in adult patients with severe pulmonic stenosis, especially if they have erythrocytosis. This patient had persistent erythrocytosis after his cardiac procedures which should have eliminated right to left shunting. He will need long-term follow-up to determine the exact cause.

Conclusion

This case suggests that adult patients with severe pulmonic stenosis should be investigated thoroughly for the presence of an atrial communication and shunting.

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References

1. Geva T, Martins JD, Wald RM. Atrial septal defects. Lancet 2014;383(9932):1921–1932.
2. Ruckdeschel E, Kim YY. Pulmonary valve stenosis in the adult patient: pathophysiology, diagnosis and management. Heart 2019;105(5):414–422.
3. Adler DH. Atrial septal defect workup: Laboratory Studies, Imaging Studies, other tests. Medscape. https://emedicine.medscape.com/article/162914-workup#showall. Accessed 2/26/2022.
4. Bärtsch P, Gibbs JS. Effect of altitude on the heart and the lungs. Circulation 2007;116(19):2191–2202.
5. Naeije R, Dedobbeleer C. Pulmonary hypertension and the right ventricle in hypoxia. Exp Physiol 2013;98(8):1247–1256.

6. Arvanitaki A, Giannakoulas G, Baumgartner H, et al. Eisenmenger syndrome: diagnosis, prognosis and clinical management. Heart 2020;106(21):1638–1645.
7. Medina A, de Lezo JS, Delgado A, et al. Combined percutaneous atrial septal defect occlusion and pulmonary balloon valvuloplasty in adult patients. Texas Heart Inst J 2000;27(2): 216–217.
8. Xu XD, Ding XY, Liu SX, et al. Immediate- and medium-term effects of simultaneous percutaneous corrections of secundum type atrial septal defect combined with pulmonary valve stenosis in local anesthesia and without transesophageal echocardiography guidance. J Cardiol. 2015;65(1): 32–36.