

Atrial Septal Defect as Unexpected Cause of Pulmonary Artery Hypertension

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Methamphetamine abuse is an increasingly prevalent cause of pulmonary artery hypertension in the United States. Conversely, an atrial septal defect rarely presents late as pulmonary artery hypertension.

We present the case of a 44-year-old methamphetamine abuser who had a 3-month history of worsening fatigue and near-syncope. She had elevated cardiac enzyme levels and right-sided heart strain. Angiographic findings suggested methamphetamine-induced pulmonary artery hypertension; however, we later heard S₂ irregularities that raised suspicion of an atrial septal defect. Ultimately, the diagnosis was pulmonary artery hypertension and a large secundum atrial septal defect with left-to-right flow. One year after defect closure, the patient was asymptomatic.

In addition to discussing this unexpected case of a secundum atrial septal defect masquerading as methamphetamine-induced pulmonary artery hypertension, we briefly review the natural history of atrial septal defects and emphasize the importance of thorough examination in avoiding diagnostic anchoring bias. (Tex Heart Inst J 2018;45(1):42-4)

Pulmonary artery hypertension (PAH) is characterized by a progressive rise in pulmonary pressures that can lead to right-sided heart failure and death. Its estimated prevalence is 1 in 70,000 people. Recognized causes include congenital heart disease, connective-tissue disease, human immunodeficiency virus, schistosomiasis, and exposure to certain drugs and toxins.¹ Increased methamphetamine abuse during the past 25 years has led to more cases of methamphetamine-induced PAH in the United States.^{2,3} In contrast, cases of atrial septal defect (ASD) presenting late as PAH have become rare because of earlier ASD detection and repair.^{4,5} We describe our clinical decisions when treating an adult whose secundum ASD initially masqueraded as methamphetamine-induced PAH.

Case Report

In May 2015, a 44-year-old woman from northern California, a daily methamphetamine user for 23 years, presented at the emergency department with a 3-month history of progressive exercise intolerance and near-syncope. Neither the initially reported physical examination nor her vital signs revealed anything notable. Her N-terminal pro-brain-type natriuretic peptide (NT-proBNP) level was 1,641 pg/mL, her troponin I level was 0.6 ng/mL, and her urine toxicology results were positive for amphetamines. An electrocardiogram showed right-axis deviation, right ventricular (RV) hypertrophy, and incomplete right bundle branch block. A computed tomographic angiogram showed no pulmonary embolus but revealed central dilation and peripheral pruning of the pulmonary artery tree. The presumed diagnosis was methamphetamine-induced PAH. The patient was admitted for evaluation and probable pulmonary vasodilator initiation.

During bedside rounds, we palpated an RV heave in our patient and heard a wide, fixed, split S₂ with a prominent P₂ component. The unusual character of S₂ raised suspicion of an ASD as the primary reason for her symptoms. In this circumstance, sildenafil and other advanced therapies for PAH are contraindicated, because they increase blood flow to the left side of the heart and augment left-to-right shunting.^{6,7} We

therefore performed right-sided heart catheterization with an oximetry run. The results definitively established the diagnosis of PAH (pulmonary artery pressures, 79/25/43 mmHg; pulmonary capillary wedge pressure, 5 mmHg; and pulmonary vascular resistance, 4.1 Wood units) and confirmed a mid-to-high right atrial oxygen saturation step-up (superior vena cava, 64%; high right atrium, 85%; mid right atrium, 81%; low right atrium, 67%; inferior vena cava, 65%; RV, 82%; pulmonary artery, 82%; and aorta, 100%) with a pulmonary-to-systemic blood flow ratio (Qp:Qs) of 2:1. Transesophageal echocardiograms verified a 3.4-cm secundum ASD with left-to-right flow and showed severe RV enlargement with marked systolic dysfunction (Fig. 1). The patient's symptoms, the RV compromise, and the hemodynamic results (PAH with pulmonary vascular resistance <5 Wood units and Qp:Qs \geq 1.5) supported our decision to close the late-presenting ASD.⁶ The defect was too large for a percutaneous approach, so we performed minimally invasive bovine pericardial patch repair (Fig. 2).

At the patient's follow-up evaluation one year later, her symptoms had resolved, and auscultation revealed a physiologically split S₂ with less P₂ accentuation. Her NT-proBNP level was nearly normal (406 pg/mL), and transthoracic echocardiograms revealed no residual atrial shunt and a substantially reduced pulmonary artery systolic pressure of 30 mmHg. As of January 2017, the patient had not needed pharmacologic therapy for PAH.

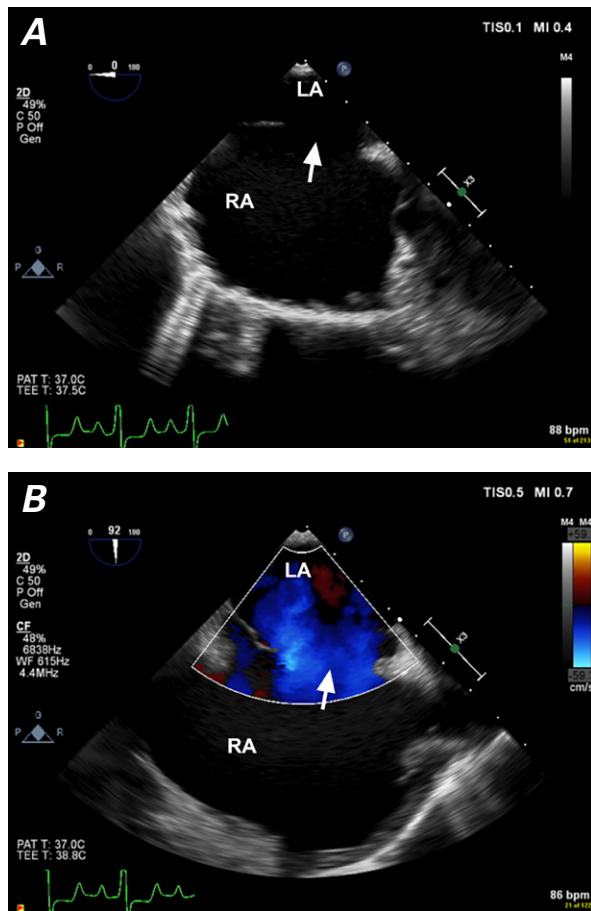


Fig. 1 Transesophageal echocardiogram shows **A**) a large secundum atrial septal defect (arrow) with **B**) a substantial left-to-right shunt (arrow) in color-flow Doppler mode.

LA = left atrium; RA = right atrium

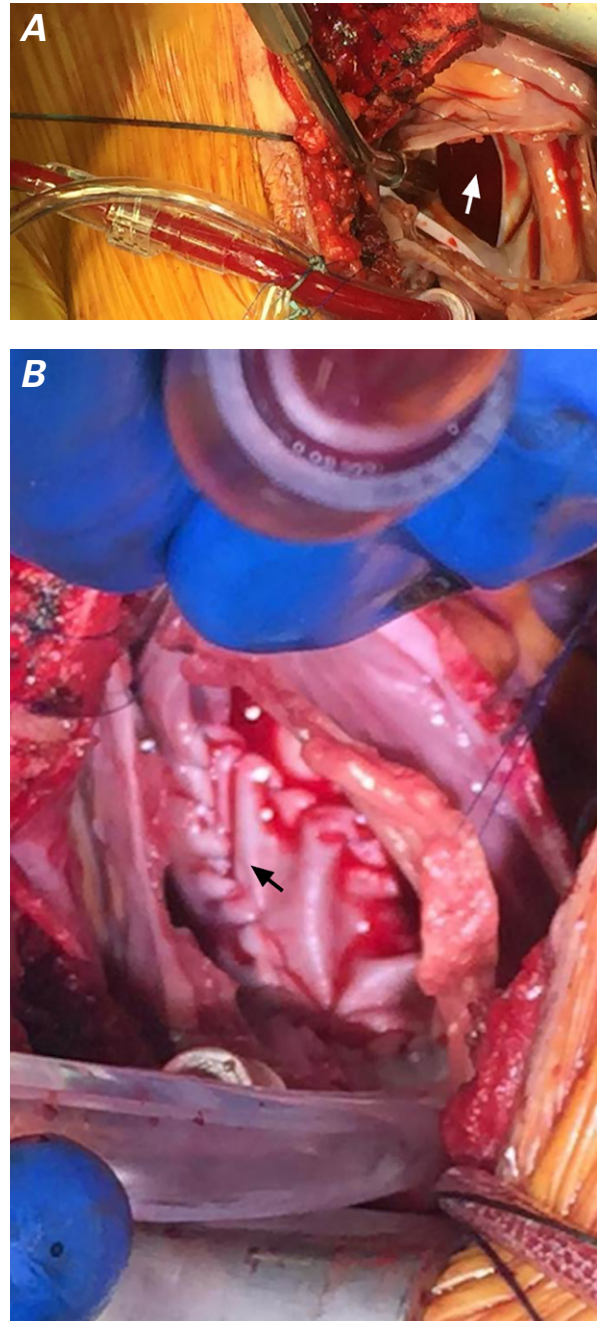


Fig. 2 Intraoperative photographs. **A**) Right atriotomy exposes a 2.5 × 4.5-cm hole in the interatrial septum (arrow) and blood in the left atrium. **B**) Bovine pericardial patch closure (arrow) restores normal left atrial geometry.

Discussion

The “duck test” is frequently applied to clinical reasoning on the wards to establish a diagnosis and to prevent over-testing and defensive medicine.* However, without comprehensive and accurate data, this principle can lead to an anchoring bias with potentially harmful consequences. Our applying the duck test to our patient’s presentation led to a singular diagnosis of methamphetamine-induced PAH—a well-established entity of increasing prevalence in the U.S. and at our clinic in California.^{2,3} However, we avoided the pitfall of anchoring by reviewing the physical examination: careful auscultation led to our discovering an ASD that had caused continuous left-to-right shunting since our patient’s birth and thus was presumably the culprit, although her methamphetamine abuse had also probably contributed to her PAH. In light of these new data, we did not prescribe empiric sildenafil therapy—which, in the presence of left-to-right shunting, would have increased pulmonary blood flow, possibly worsening her symptoms and causing arrhythmia. Instead, we expedited analysis and repair of the ASD.^{6,7}

The reported prevalence of ASDs at birth is 0.2%, and secundum defects constitute approximately 70% of these (~1 in 700 births).⁸ However, the natural history of ASDs has evolved consequent to earlier detection and repair, improvements in medical therapy and closure techniques, and repair in older patients. Before 1970, fewer than half of patients with ASDs reportedly survived beyond 40 years, and only 10% lived to 60 years; end-stage PAH and Eisenmenger syndrome were frequent causes of death.⁹ Conversely, as of 1995, the estimated 10-year survival rate was 95% in middle-aged patients (mean age, 57 yr) who underwent surgical repair.⁵ The prevalence of ASD-induced PAH was estimated to be 6% to 7%, although exact comparison between time periods is difficult because of varying diagnostic criteria for PAH.^{4,5} Secundum ASDs can be closed either surgically or percutaneously; the class I indication for repair is evidence of RV compromise with or without symptoms. Closure is contraindicated in the presence of irreversible PAH without left-to-right shunting.⁶ Our patient had a compatible hemodynamic status and met the class I indication for repair. Just one year after surgery, her estimated pulmonary pressures were nearly normal, indicating the high plasticity of the pulmonary artery tree.

This unexpected case of a secundum ASD presenting late as PAH underscores the importance of examining patients carefully, to avoid anchoring bias and to guide

appropriate clinical reasoning and diagnosis. Otherwise, the whole heart of the matter might be missed—or, as almost happened in this case, the hole itself.

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* *When I see a bird that walks like a duck and swims like a duck and quacks like a duck, I call that bird a duck.*
— widely attributed to James Whitcomb Riley