

# Platypnea-Orthodeoxia Syndrome Caused by an Intracardiac Shunt

Sanket P. Borgaonkar, MD  
Wilson W. Lam, MD  
Mehdi Razavi, MD  
Dhaval R. Parekh, MD

*Platypnea-orthodeoxia syndrome, a rare condition characterized by posture-related dyspnea, is usually caused by an intracardiac shunt, hepatopulmonary syndrome, or shunting resulting from severe pulmonary disease.*

*We report the case of a 33-year-old woman who presented with increasing dyspnea and oxygen desaturation when she sat up or arose. Our diagnosis was platypnea-orthodeoxia syndrome. A lead of a previously implanted pacemaker exacerbated a severe tricuspid regurgitant jet that was directed toward the patient's intra-atrial septum. Percutaneous closure of a small secundum atrial septal defect eliminated right-to-left shunting and substantially improved the patient's functional status. In addition to this case, we discuss this unusual condition. (Tex Heart Inst J 2020;47(4):298-301)*

**Key words:** Defibrillators, implantable/adverse effects; dyspnea/etiology; heart septal defects, atrial/ complications/diagnostic imaging/therapy; hypoxia/etiology; posture/physiology; treatment outcome; tricuspid valve insufficiency/complications

**From:** Department of Medicine (Drs. Borgaonkar, Lam, Parekh, and Razavi), Baylor College of Medicine; Department of Electrophysiology Clinical Research and Innovations (Dr. Razavi), Texas Heart Institute; and Department of Cardiology (Dr. Lam), Texas Heart Institute and Baylor–St. Luke's Medical Center, Houston, Texas 77030

**Address for reprints:** Wilson W. Lam, MD, Department of Medicine, BCM620, Baylor College of Medicine, One Baylor Plaza, Houston, TX 77030

**E-mail:** wlam@bcm.edu

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## Case Report

A 33-year-old woman was admitted for evaluation of dyspnea and chest tightness that had worsened over 3 months. Her medical history included complete congenital heart block and peripartum cardiomyopathy that had necessitated implantation of a biventricular implantable cardioverter-defibrillator (ICD) in 2010. The patient reported intermittent dyspnea that increased on minimal exertion and was not relieved by respiratory inhalation therapy. She also reported intermittent palpitations, but no chest pain, cough, sputum production, or firing of her ICD. Her medical therapy included  $\beta$ -blockers and angiotensin-converting enzyme inhibitors.

On pulse oximetry, the patient had oxygen saturation levels of 92% while supine, 83% while sitting, and lower levels during exertion. Physical examination revealed no jugular vein distention, no edema or cyanosis at rest, and a soft abdomen with no signs of hepatosplenomegaly. Auscultation revealed clear lungs, a normal  $S_1$  and  $S_2$ , and no gallops, clicks, or rubs; however, a soft grade 2/6 holosystolic murmur was heard at the left lower sternal border, and an apex beat was best palpated at the left midclavicular line. A chest radiograph showed no acute cardiopulmonary abnormalities. The patient's electrocardiogram showed biventricular pacing. Her laboratory values were within normal limits.

We suspected a cardiac or pulmonary cause of the desaturation. A computed tomogram ruled out pulmonary emboli and aortic aneurysm. Transthoracic and transesophageal echocardiograms revealed a left ventricular ejection fraction (LVEF) of 50% to 55%, severe tricuspid regurgitation (TR), and a right-to-left shunt at the atrial level that was confirmed with use of agitated saline solution. Color-flow Doppler mode showed a regurgitant jet directed toward an atrial septal defect (ASD) (Fig. 1). Coronary angiograms showed nonobstructive coronary artery disease, and the patient's right-sided pulmonary artery pressures were normal. The signs and symptoms were consistent with POS, so ASD closure was appropriate.

Intracardiac echocardiograms (ICE), obtained by using an ICE catheter and balloon sizing, showed a 12-mm ASD and right-to-left shunting (Fig. 2). Through a trans-

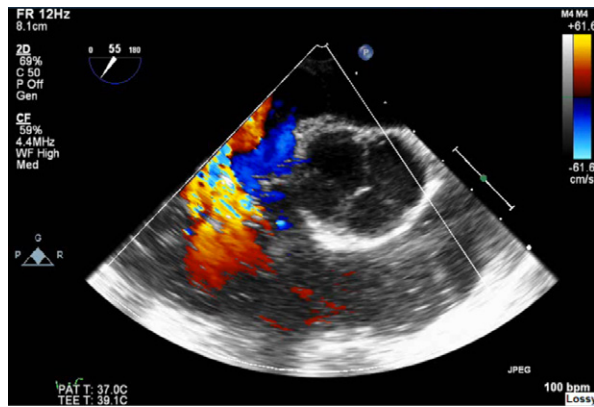
femoral venous approach, we used a 25-mm Gore Helex Septal Occluder to close the ASD (Fig. 3) and confirmed its ideal positioning by using ICE and angiography. A transthoracic echocardiogram then showed a sustained LVEF of 55% to 60% and severe TR, but no right-to-left shunting (Fig. 4). The ICD leads appeared to be in proper position. Because the shunting was stopped and

the TR jet was confined in the patient's right atrium, no further treatment was considered necessary.

The patient's functional status improved substantially. One month later, she had no signs or symptoms of POS.

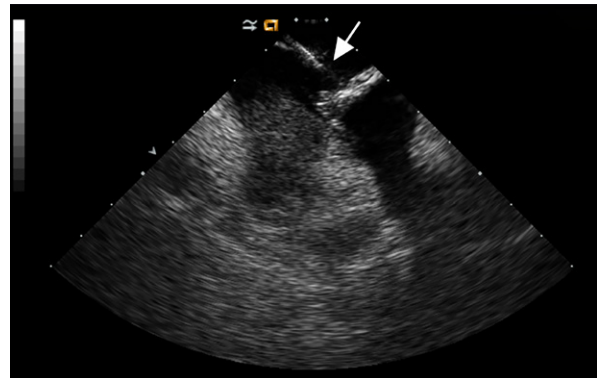
## Discussion

Although cases of POS have been reported, the condition is probably underdiagnosed. The mechanisms underlying hypoxemia in most POS cases involve both

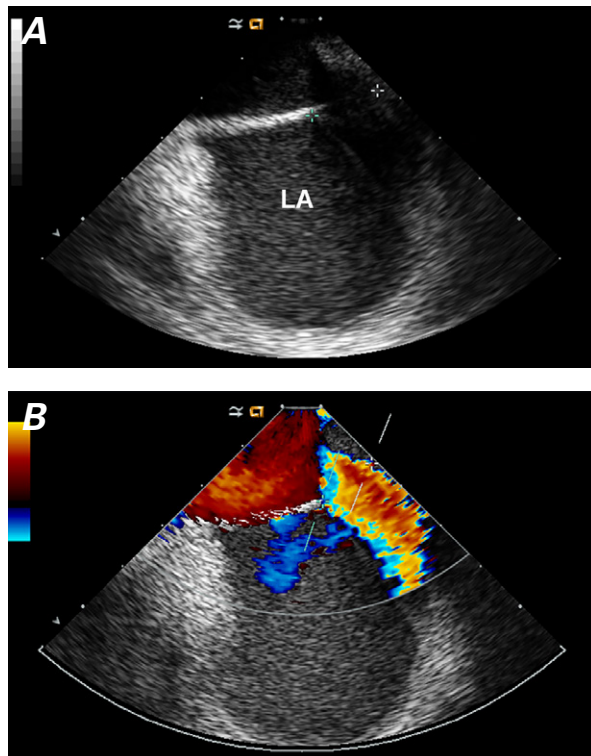


**Fig. 1** Transesophageal echocardiogram in color-flow Doppler mode (short-axis view of aortic valve at 55°) shows tricuspid regurgitation across an atrial septal defect.

Supplemental motion image is available for Figure 1.

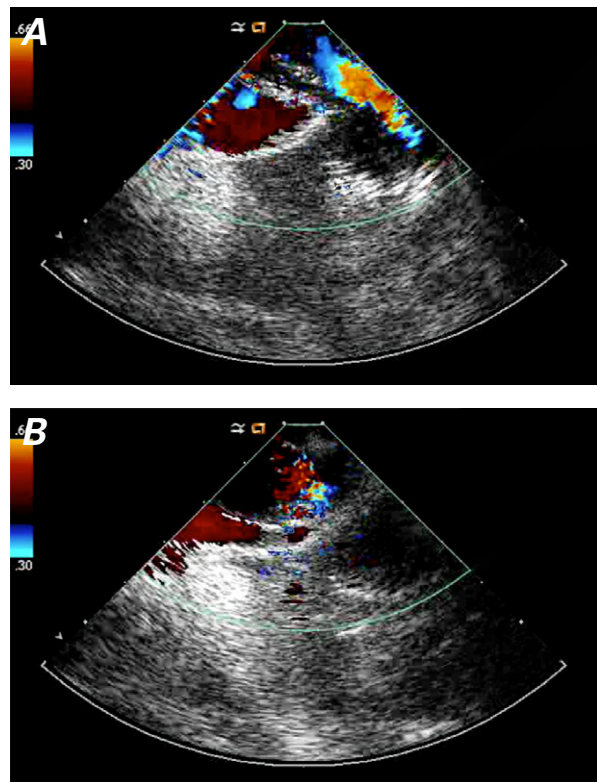


**Fig. 3** Intracardiac echocardiogram shows the septal occluder (arrow) deployed across the atrial septal defect.



**Fig. 2** Intracardiac echocardiograms show **A**) the atrial septal defect (2-dimensional image) and **B**) turbulent right-to-left flow through it (color-flow Doppler mode).

LA = left atrium



**Fig. 4** Intracardiac echocardiograms (color-flow Doppler mode) show **A**) right-to-left flow through the atrial septal defect before percutaneous closure and **B**) none after closure.

anatomic and functional defects. In cardiac POS, the anatomic defects that typically enable communication between the right- and left-sided circulation are patent foramen ovale and ASD. When POS is caused by an ASD, upright posture stretches the intra-atrial septum, and deoxygenated blood flows through the defect. The blood is preferentially shunted because of this anatomic defect and a functional defect, which our patient also had.

The absence of elevated pulmonary artery pressures differentiates cardiac POS from other causes of shunting. However, an anatomic defect by itself is typically not enough to cause desaturation. Something more, such as a functional defect, is necessary to move deoxygenated blood into the systemic circulation. A functional defect can either preferentially direct blood flow through an interatrial defect, or elevate right atrial pressure and promote blood flow across a pressure gradient. The former can be caused by pathologic conditions that induce atrial septal repositioning, such as aortic dilation, aortic aneurysm, or a history of cardiac or thoracic surgery, in particular right-sided pneumonectomy. Because the atrial septum conforms to these adverse conditions, blood flow from the venae cavae is directed toward the septal defect, causing symptoms.<sup>1</sup> In other cases, prominent eustachian valves (notable in fetal circulation) can assist right-to-left cardiac blood flow.<sup>2</sup> Increased interatrial pressures can be caused by increased pulmonary vascular resistance (such as that from pulmonary embolism, chronic obstructive pulmonary disease, or pulmonary hypertension) or by conditions that physiologically demand an elevated right-sided filling pressure to maintain cardiac output (for example, pericardial effusions or constrictive pericarditis).<sup>1</sup> Noncardiac causes of POS, most notably hepatopulmonary syndrome, are not our primary focus of discussion.

Our patient's functional defect was a TR jet aimed at the ASD. Rising from a recumbent position probably stretched her atrial septum and enabled the TR jet to shunt more strongly across the ASD. Only a few authors have reported this physiologic change as the sole cause of POS.<sup>3-5</sup> Another unusual aspect of our case was the unexpected strength of the TR jet that was caused by the right ventricular lead of our patient's ICD. The addition of the shunting to her TR caused her symptoms. When the shunting was stopped, the TR jet stayed in the right atrium and warranted no further treatment, because many patients tolerate that degree of regurgitation.

Treating POS involves closing interatrial defects. A percutaneous approach is preferable to surgery. The patient's ability to tolerate an invasive procedure, severity of symptoms, and comorbidities must be considered before intervention. A Class IIA level of evidence supports ASD closure in patients with documented POS.<sup>6</sup>

Because our patient had both an ICD and an ASD, her risk of stroke was 1% per year, so closing her ASD

was necessary to decrease the risk.<sup>7</sup> Of note, implanting a device with endocardial leads in patients who have an intracardiac shunt can increase the risk of thromboembolic events 2-fold.<sup>8</sup> Therefore, shunts should be closed before pacemaker or ICD leads are inserted, or these devices should not be inserted.<sup>9</sup> Although few investigators have examined long-term prognosis after interatrial defect closure in patients with POS, rates of shunt recurrence are low and symptomatic improvement is sustained.<sup>10,11</sup>

In summary, we think that patients who experience dyspnea and desaturation while upright should be evaluated for intracardiac shunting, and that any associated ASD should be closed.

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