Platypnea-Orthodeoxia Syndrome:

To Shunt or Not to Shunt, That is the Question

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Platypnea-orthodeoxia syndrome is a rare disease defined by dyspnea and deoxygenation, induced by an upright position, and relieved by recumbency. Causes include shunting through a patent foramen ovale and pulmonary arteriovenous malformations.

A 79-year-old woman experienced 2 syncopal episodes at rest and presented at another hospital. In the emergency department, she was hypoxic, needing 6 L/min of oxygen. Her chest radiograph showed nothing unusual. Transthoracic echocardiograms with saline microcavitation evaluation were mildly positive early after agitated-saline administration, suggesting intracardiac shunting. She was then transferred to our center.

Right-sided heart catheterization revealed no oximetric evidence of intracardiac shunting while the patient was supine and had a low right atrial pressure. However, her oxygen saturation dropped to 78% when she sat up. Repeat transthoracic echocardiography while sitting revealed a dramatically positive early saline microcavitation-uptake into the left side of the heart. Transesophageal echocardiograms showed a patent foramen ovale, with right-to-left shunting highly dependent upon body position. The patient underwent successful percutaneous patent foramen ovale closure, and her oxygen supplementation was suspended.

In patients with unexplained or transient hypoxemia in which a cardiac cause is suspected, it is important to evaluate shunting in both the recumbent and upright positions. In this syndrome, elevated right atrial pressure is not necessary for significant right-to-left shunting. Percutaneous closure, if feasible, is first-line therapy in these patients. (*Tex Heart Inst J* 2016;43(3):264-6)

Platypnea-orthodeoxia syndrome (POS) is a rare disease defined by dyspnea and deoxygenation of arterial blood, induced by an upright position, and relieved by recumbency.¹ Frequently reported underlying causes include intracardiac shunting through some form of defect in the interatrial septum, pulmonary arteriovenous malformations, and pulmonary intraparenchymal shunts. Typically, a cause of elevated right atrial (RA) pressure—such as pulmonary embolism, pulmonary hypertension, right hydrothorax, chronic obstructive pulmonary disease, or pneumonectomy—is invoked when shunting is evident through an interatrial communication. We describe our treatment of a patient who had POS, and we review the relevant medical literature.

Case Report

In July 2014, a 79-year-old woman experienced 2 syncopal episodes while at rest. Upon evaluation in an emergency department, she was hypoxic (consuming 6L of oxygen per minute) and exhibited left hemiplegia for about one hour, before its resolution. Her chest radiograph showed nothing notable. A computed tomographic (CT) scan of her head showed no evidence of infarct, but subsequent magnetic resonance imaging (MRI) of the brain revealed an acute right thalamic infarction. A transthoracic echocardiogram (TTE) with saline microcavitation was very mildly positive and suggested intracardiac shunting (Fig. 1A).

Upon her transfer to our center for further evaluation, a pulmonary-embolism-protocol CT revealed an enlarged aorta (maximal dimension, 4.3 cm) and an acute pulmonary embolus in a subsegmental right-lower-lobe artery. Right-sided heart catheterization revealed no oximetric evidence of intracardiac shunting while the patient was supine. However, her normal oxygen saturation when lying supine immediately decreased to 78% when she sat up.

Given the discordance between the referring hospital's TTE and our cardiac catheterization findings, we performed a repeat TTE with a bubble study. When the patient was supine, there was minimal shunting upon administration of agitated saline. When she was seated, there was a dramatically positive early saline-microcavitation uptake to the left side of the heart (Fig. 1B). Positional hypoxemia occurred while the patient was upright and resolved while she was recumbent. Transesophageal echocardiograms showed an interatrial septal aneurysm and a patent foramen ovale (PFO). Right-to-left shunting through the PFO was highly dependent upon position. Shunting was minimal as the patient lay supine (Fig. 1C) but substantial when she sat upright (Fig. 1D). Cardiac MRI revealed a moderately dilated aortic root, and the heart in a horizontal position as a consequence (Fig. 2A); a prominent eustachian valve directed blood toward the interatrial septum, at the level of the PFO (Fig. 2B).

The patient underwent successful percutaneous closure of the PFO for POS with use of a 30-mm GORE® HELEX® septal occluder (W.L. Gore & Associates, Inc.; Flagstaff, Ariz). The patient tolerated the procedure well, and her oxygen supplementation was completely suspended before she left the hospital. She continued to do well during several months of follow-up after PFO closure. Her oxygen saturation on room air remained normal while supine, seated, standing, and ambulating.

Discussion

Typically, the cause of dyspnea and deoxygenation of arterial blood is discovered when the shunting of blood through a PFO is confirmed. However, in patients without elevated RA pressure, the reported underlying mechanism of dyspnea and deoxygenation of arterial blood is either 1) an enlarged aorta with a consequent shift (to a more horizontal position) of the septum pri-

Fig. 1 Transthoracic echocardiograms to evaluate shunting show **A**) minimal saline microcavitations in the LV during peak opacification of the RV (upon initial evaluation of the supine patient at another hospital) and **B**) marked saline microcavitation opacification of the LV with nearly equivalent opacification of the RV and LV with the patient seated upright (upon evaluation at our hospital). Transesophageal echocardiograms show the substantial positional dependence of intracardiac shunting. **C**) No significant shunting is seen through a patent foramen ovale (arrow) when the patient is supine; however, **D**) a marked change in septal geometry and right-to-left shunting (arrow) is seen moments later, when the patient sits at 90° (arrow).

LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle

Supplemental motion images are available for Figures 1A, 1B, 1C, and 1D.

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Fig. 2 Cardiac magnetic resonance images reveal **A)** an enlarged ascending thoracic aorta with a horizontally oriented left ventricle, and **B)** a prominent eustachian valve (*) directing the returning inferior vena cava flow toward the fossa ovalis (arrow).

num and septum secundum components of the interatrial septum, or 2) a prominent eustachian valve that directs venous blood flow toward the PFO.^{2,3}

In the case presented here, cardiac MRI revealed both of these features as the likely cause of our patient's POS. Although she did have a subsegmental pulmonary embolus, her cardiac catheterization results revealed a pulmonary capillary wedge pressure higher than the RA pressure, which renders acute elevation of RA pressure an unlikely cause.

A review of the available medical literature suggests that POS with normal RA pressure has been reported in approximately 10% of all cases of POS.¹ Some authors have labeled this normal RA pressure variant of POS "platypnea-orthodeoxia disease" (POD).¹ Review of these cases of POD has shown that onset is usually later in life (with a median in the 7th decade) and that there is no significant difference in prevalence between men and women. Furthermore, aortic abnormalities leading to a distortion in the interatrial septal anatomy are present in nearly one quarter of patients with POD. A common factor in cases of POD is the presence of a distortion in the atrial anatomy from an extrinsic cause, which can include, for example, aortic enlargement, pneumonectomy, and diaphragmatic paralysis.

Because the degree of shunting in this disease is highly dependent upon the position of the patient, it is crucial to evaluate shunting both in the recumbent and upright (sitting or standing) positions. The high degree of variability in the bubble-study findings and the absence

of shunting in a supine patient as detected by cardiac catheterization were probably caused by inconsistent application of this simple practice. Percutaneous closure of the PFO remains first-line therapy because of its safety and efficacy.⁴⁻⁶

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