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Platypnea-orthodeoxia syndrome: Diagnostic challenge and the importance of heightened clinical suspicion

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Introduction: Platypnea-orthodeoxia syndrome (POS) is an uncommon condition of dyspnea (platypnea) and hypoxemia (orthodeoxia) with upright position and resolving or improving with recumbency.

Case Presentation: An 83 year old woman with a history of paroxysmal atrial fibrillation, hypertension, and hyperlipidemia was admitted with several weeks of progressive positional dyspnea that worsened with standing and improved with recumbency. Initial evaluation and workup was significant for normal oxygen saturation lying flat but desaturations to the 80s sitting up or standing, require up to 15 liters-per-minute to maintain oxygen saturations above 90%. Cardiac and pulmonary physical exam were within normal limits. Resting ECG showed normal sinus rhythm and left axis deviation. Chest x-ray and pulmonary function tests were normal. The arterial blood gas demonstrated a drop in arterial oxygen tension from 93 mmHg when lying down to 54 mmHg when standing. Transthoracic echocardiogram revealed normal pulmonary pressure, normal ejection fraction, but demonstrated a bidirectional shunt at atrial level. A right heart catheterization revealed pulmonary blood flow to cardiac output ratio (Q_p/Q_s) of 0.65, signifying a hemodynamically significant right-to-left shunting in the setting of normal pulmonary pressures. A consequent transesophageal echocardiogram revealed a moderate size secundum atrial septal defect.

The patient was taken to the catheterization lab for intracardiac three dimensional imaging and defect closure, without residual shunting visualized with color-flow imaging. Post-closure arterial blood gases revealed significant improvement of standing hypoxemia, and the patient had symptomatic resolution, which has persisted at follow-up visits.

Discussion: POS is an uncommon condition associated with intracardiac shunting, pulmonary shunting, ventilation-perfusion mismatch, or a combination of these three processes. This case illustrates the classic presentation of POS in a patient with significant dyspnea sitting up/standing, which resolves with recumbency. POS is suspected when an individual has normal saturations on room air while supine with dyspnea and desaturations to less than 95% when upright. Correlation with blood-gas analysis documenting positional change in oxygen tension (PAO₂) is also helpful.

Diagnosis of POS can be delayed by the fact that the patient is hypoxemic when upright but testing is usually done when patient is supine (i.e., an echo may not show a shunt when the patient is lying down, yet a large shunt is present when the patient stands). A thorough history is of vast importance for correct diagnosis and treatment.

Patient's symptoms occurred due to gradual uncoiling of the aorta secondary to aging, selectively altering the direction of blood from the inferior vena cava to the left atrium through the atrial septal defect (ASD), creating a right-to-left shunt. When symptoms are due to shunts caused by ASD or patent foramen ovale, treatment involves intracardiac device closure.

This case has been reported to highlight the importance of taking detailed patient history and including POS on the differential for positional dyspnea. Early detection can lead to effective treatment and symptomatic resolution.

Biography

Stanislav Henkin is a 1st year Internal Medicine resident at Mayo Clinic in Rochester, MN. He graduated from Tufts University in 2008, earning Bachelor of Arts degrees in Biochemistry and Community Health, before earning his medical degree and MPH in epidemiology from Boston University School of Medicine in 2013. His research focus is long-term outcomes after percutaneous coronary intervention. His overall career goal is to be an interventional cardiologist who provides excellent high-quality and safe care; and to conduct research, especially on comparative effectiveness, with the overall goal of improving patient outcomes.

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