

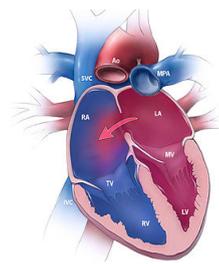
Exploring A Rare Cause of Platypnea-Orthodeoxia Syndrome

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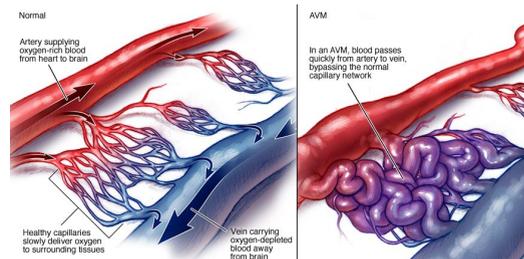
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Introduction

Platypnea-orthodeoxia syndrome (POS) is a rare, but underdiagnosed, condition. It refers to positional dyspnea and drop in PaO₂, observed with assuming an upright position and resolved with supine position. This is in contrast to the more familiar concept of orthopnea, in which a patient's dyspnea is relieved with upright positioning. POS is often due to an underlying right to left shunt, either at intracardiac or intrapulmonary level, or more rarely, a ventilation perfusion mismatch. With intracardiac shunts, there is stretching of atria with upright positioning which causes the defect – whether it be ASD or PFOs – to widen, which subsequently increases shunting of deoxygenated blood. With arteriovenous malformations (AVMs), adopting an upright position expands the lung volume and opens up more AVMs, causing increased blood flow in lung bases. This leads to more shunting of deoxygenated blood through pulmonary AVMs, resulting in worsening hypoxemia and dyspnea.^{1,3}



Atrial Septal Defect



Arteriovenous malformation

Anasarca has been studied as a rare etiology of POS; mechanism is attributed to the dynamic collapse and expansion of pulmonary vasculature with positional changes. Severe anasarca causes collapse of lower lung fields while in supine position; when upright, there is increased perfusion at lung bases due to gravity however the formerly collapsed areas remain poorly ventilated, causing increased physiological shunting of deoxygenated blood to enter the systemic circulation. Here, we present a case of severe, yet reversible, POS, occurring secondary to anasarca in the absence of any underlying intracardiac shunts or pulmonary AVMs.

Case

A 49-year-old African American male presented with progressively worsening dyspnea over four months. Patient also reported swelling, which initially was limited to his lower extremities, then later progressed to involve his whole body. Patient's weakness and dyspnea worsened to the point that he became bed bound for three weeks prior to hospitalization. Patient was a formerly obese individual, who underwent a Roux-En-Y gastric bypass surgery an year prior to presentation. On admission, patient's O₂ saturations were 95% while supine and 70% upon sitting. Patient was symptomatic with shortness of breath and palpitations upon sitting up. Admission labs revealed a low serum albumin of less than 1.5.

Case (continued..)

CT chest angiography was negative for pulmonary embolism, AVMs, and findings suggestive of pulmonary hypertension. Echocardiogram showed normal LV and RV function; bubble study did not reveal any intracardiac shunt. Patient's anasarca was attributed to severe malnutrition secondary to gastric bypass. Review of patient's imagings revealed that patient's gastro-jejunal anastomosis was performed at a more distal site than what is done routinely, which likely led to reduced absorptive surface causing patient's malnutrition. Patient was subsequently started on total parenteral nutrition, with improvement of anasarca and resolution of POS.

Patient was to be considered for gastric bypass revision, pending improvement of his functional status.

Discussion & Conclusion

This case report focused on investigating anasarca secondary to malnutrition as the etiology of a patient's POS. In patients with anasarca, there is compression of the lung tissue by the accumulated fluid. Upon assuming an upright position, there is increased perfusion due to gravity dependent blood flow in the lower lung fields. However, the alveoli in the lower lung fields remain collapsed and poorly ventilated even with upright positioning, resulting in a V/Q mismatch, as seen in pulmonary AVMs. Subsequently, deoxygenated blood enters the systemic circulation, leading to the observed phenomenon of situational hypoxia, worsened in the upright position.

POS remains an underdiagnosed and rare disease entity. Obtaining a detailed history of factors eliciting one's dyspnea and evaluating a patient's oxygen saturations with positional changes are crucial in determining the underlying etiology. Insufficient workup may lead to missing the diagnosis of POS and therefore, improper management, causing patients to suffer from persistent dyspnea. On the other hand, proper and timely diagnosis leads to appropriate management by addressing the underlying etiology of POS, leading to resolution of patient's initial dyspneic symptoms.

References

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