Editorial for the article:
“Percutaneous Intervention to Treat Platypnea – orthodeoxia Syndrome: The Toronto Experience” by Ashish H Shah, Mark Osten, Andrew Leventhal, Yvonne Bach BScH, Daniel Yoo, Danny Mansour, Lee Benson, William M Wilson, and Eric Horlick

Platypnea-Orthodeoxia Syndrome: An overlooked cause of hypoxemia.

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Platypnea-orthodeoxia syndrome (POS) is one of those onomatopoeic medical terms that is difficult to say but packs a lot of information into a short phrase. It describes a condition where patients are significantly hypoxemic and short of breath in the upright position but may have near-normal oxygen saturation levels when supine. It is a rare disorder, most commonly associated with right-to-left shunting through a patent foramen ovale.[1] Since 20% of all humans have persistent patency of the foramen ovale after birth and into adulthood, there has to be something anatomically unusual about those people who develop the platypnea-orthodeoxia syndrome. This could be due to distorted anatomy, changes in right atrial hemodynamics or a prominent Chiari network and Eustachian valves which have been suggested as mechanisms for more right to left shunting and increased incidence of paradoxical emboli and POS.[2]

The accompanying article by the structural heart disease group at Toronto's Hospital for Sick Children, adds significant information to our database knowledge about this relatively rare condition. They report on 52 patients who had percutaneous closure for this syndrome over 18 years. As with many other conditions in medicine, this is an "orphan" disorder and there will never be a randomized clinical trial to prove what therapy is best. Using a combination of the Amplatzer PFO occluder and the Amplatzer ASO septal devices, this group achieved an 80% complete closure rate. Even in those patients who did not have complete closure, there was a significant improvement in the oxygen saturation (from 81% to 95%) as well as symptomatic clinical improvement. This is a gratifying result for these highly symptomatic patients, which emphasizes the benefits of percutaneous treatment of structural heart disease without the need for open heart surgery.[3]

This study was performed over 18 years and there have been significant changes in imaging and devices within that time period, such as the use of intracardiac ultrasound imaging or softer and more efficient closure devices such as the Gore Cardioform (Gore & Associates, Inc. Flagstaff AZ, USA) device. The use of intra-cardiac echocardiography (ICE) imaging as the series progressed obviates the need to have patients intubated such as during TEE guidance. With careful catheter manipulation one can ensure a reliable and safe closure with minimal sedation.[4]
The take-home message from this article is to be cognizant of platypnea-orthodeoxia as a potential explanation for hypoxemia in those patients who do not have severe pulmonary parenchymal disease. In this regard, Shah and Horlick described a clever way of distinguishing hypoxemia from pulmonary disease versus right to left shunting by measuring the oxygen saturation in individual pulmonary veins. This can be accomplished if a PFO is present by passing the catheter across the atrial septum and into the orifices of the individual pulmonary veins as they drain into the left atrium.[5,6]

The intra-atrial septal anatomy can have a significant effect on the level of shunting across the septum.[7] A useful insight provided in this paper, is the description of the possible mechanism by which platypnea-orthodeoxia syndrome can occur due to the abnormal angle of the atrial septum based on their findings from CT imaging in 14 patients. However, the abnormal angle was only present in those patients who had atrial septal aneurysms or large PFOs that required the Amplatzer ASO device. As noted in other papers, the platypnea-orthodeoxia syndrome is often associated with rotational distortion of the thoracic anatomy due to pneumonectomy, ascending aortic dilatation, or diaphragmatic paralysis.[7] Measurement of the angle in this study was done after deployment of the closure devices, which itself could have affected the inter-atrial angle measured.

Quantitation of the degree of right-to-left shunting can be performed with transcranial Doppler (TCD) studies, which is more sensitive than other imaging modalities.[8] It was noted in this paper that an aneurysmal septum was observed in 66% and was more likely to be present in patients who required a non-PFO closure device. Another option is to use balloon sizing for large PFOs, a long tunnel, or a mobile aneurysmal septum to determine the device size and avoid the need to change these expensive devices. A proposed PFO classification scheme can help guide the closure technique and stratify the difficulty of the intervention.[9]

The authors also raise a clever hypothesis that the mechanism of developing this syndrome is due to a vortex of flow created by the spatial orientation of venous return from the inferior vena cava where the blood is directed superiorly and posteriorly, and the superior vena cava where the blood is directed anteriorly and inferiorly. This circular
motion may be responsible for hemodynamically opening up the PFO flap. At this point, this concept is only hypothetical but potentially could be studied with 4 dimensional MRI in the supine versus the upright position.

In conclusion, this article is helpful as a reminder to look for a patent foramen ovale and consider the platypnea-orthodeoxia syndrome in patients with dyspnea and hypoxemia. This is pertinent when progressive dyspnea develops after surgery that may otherwise be attributed to pulmonary disease or in patients with hypoxemia that is out of proportion to their known pulmonary disease. A simple screening test such as TCD and a relatively safe percutaneous closure device can alleviate significant suffering associated with the platypnea-orthodeoxia syndrome.
References:


