Not so innocuous after all: a case report of platypnoea-orthodeoxia syndrome

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Dear Sir,

We report a case of platypnoea-orthodeoxia syndrome (POS), a condition characterised by a sensation of breathlessness (platypnoea) and low oxygen saturation in the upright position that improves on lying down (orthodeoxia), which was diagnosed and demonstrated elegantly on transoesophageal echocardiography. First described by Burchell et al,\(^{(1)}\) this condition is associated with patent foramen ovale (PFO), or combined with an atrial septal aneurysm (ASA).\(^{(2)}\) Up till the 20th century, only 40 cases have been reported,\(^{(3)}\) but a review of the current literature reveals at least 150 case reports to date. What was previously deemed uncommon is now increasingly recognised as an important and reversible cause of hypoxaemia.

An 86-year-old man first presented with un witnessed fall and head injury to the neurosurgical team. He had transient episodes of dyspnoea and desaturation during rehabilitation. Cardiorespiratory examinations, haematological and biochemical analysis, and resting electrocardiogram were normal. Chest radiograph revealed clear lung fields. An arterial blood gas test done in room air showed significant hypoxaemia (PaO\(_2\) 48 mmHg) and an elevated alveolar-arterial gradient of 68 mmHg (expected 21 mmHg). Computed tomography angiography of pulmonary arteries showed no evidence of pulmonary embolism or intrapulmonary vascular malformation.

Transthoracic echocardiography revealed normal left ventricular ejection function and no pulmonary hypertension. Bubble study with Valsalva manoeuvre showed a right-to-left shunt at the level of the atria. The patient was further evaluated with transoesophageal echocardiography (TEE) in the absence of sedation, which confirmed the presence of an ASA and PFO. In the supine position, the saturation was 97% on 2 litres of intranasal oxygen, and there was no significant right-to-left shunt with colour and agitated saline contrast (Fig. 1). The bed was adjusted to chair mode, and while sitting, his saturation dropped to 94% with a significant right-to-left shunt on colour Doppler and agitated saline contrast. The foramen ovale
had a significant gap and an excursion of 1 cm, and remained open for at least 60% of the cardiac cycle (Fig. 2). A diagnosis of POS was made. On the basis of reducing heart rate and the amount of right-to-left shunting, we started the patient on a trial of beta blockers, with a view to performing a transcatheter closure of the PFO if there was no improvement.

Fig. 1 Echocardiographic images (in supine position). (a) Before the bubble study, no separation of the intra-arterial septum (IAS) was observed. (b) During the bubble study, no bubbles were seen going across the IAS. RA: right atrium

Fig. 2 Echocardiographic images (in sitting position). (a) Before the bubble study, separation of the intra-arterial septum (IAS) was seen. (b) During the bubble study, significant right-to-left shunting was demonstrated, with bubbles going across the IAS. LA: left atrium; PFO: patent foramen ovale; RA: right atrium

POS occurs as a result of right-to-left shunting in the absence of pulmonary hypertension and persistent right-to-left pressure gradient. To cause this phenomenon, two components are required: an anatomical component in the form of an inter-atrial communication and a functional component that promotes abnormal shunting of blood with
positional change. In our patient, POS was likely precipitated by dehydration, reducing left atrial pressure more than the right. The upright position stretches the inter-atrial communication, allowing increased streaming of venous blood from the inferior vena cava through the defect, thus causing desaturation and dyspnoea whenever he sat up. Performing a TEE with dynamic postural manoeuvres allows for a ‘live’ demonstration of this phenomena and elegantly confirms the diagnosis of POS. Definitive treatment involves closure of the intracardiac communication\(^4\) via open surgery or transcatheter occlusion.\(^5\)

Therefore, POS should be considered in the workup of dyspnoea. TEE with postural manoeuvres readily establishes the diagnosis when POS is suspected.

Yours sincerely,

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REFERENCES