Cardiology in the Young

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Sinus venosus atrial septal defect presenting with platypnea-orthodeoxia syndrome in the setting of COVID-19 infection

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Brief Report

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Abstract

Platypnea-orthodeoxia syndrome is characterised by hypoxemia and dyspnoea while upright that resolves when supine, typically associated with an interatrial communication. We present a case of platypnea-orthodeoxia syndrome associated with a sinus venosus atrial septal defect in a patient with multiple possible aetiologies of hypoxemia, including COVID-19 infection. Cardiac catheterisation with provocative maneuvers confirmed the diagnosis and symptoms resolved following defect closure. We suggest that in patients with platypnea-orthodeoxia syndrome, it is useful to obtain haemodynamic data while supine and upright

Platypnea-orthodeoxia syndrome is characterised by hypoxaemia and dyspnoea while upright that resolves when supine, typically associated with an interatrial communication. We present a case of platypnea-orthodeoxia syndrome associated with a sinus venosus atrial septal defect. Cardiac catheterisation with provocative manoeuvres confirmed the diagnosis and symptoms resolved following defect closure.

Case presentation

A 60-year-old man was hospitalised for hypoxaemia in the setting of COVID-19 infection and presumed COVID-19 pneumonia. He was treated with remdesivir and steroids and discharged on supplemental oxygen but continued to experience significant dyspnoea and hypoxaemia with two readmissions within a month. These were initially attributed to post-COVID pulmonary disease. During his third admission, a transthoracic echocardiogram with bubble study demonstrated a right-to-left intracardiac shunt. He was then transferred to our institution for consultation with adult congenital cardiology. On arrival, he had oxygen saturations of 90–97% while supine and desaturation to 80–85% when upright. There was no improvement in his hypoxaemia with supplemental oxygen. Prior to his admission for COVID-19, he was on no medications and had no major medical issues.

The differential diagnosis included CHD: atrial septal defect, patent foramen ovale, or ventricular septal defect with Eisenmenger physiology. Additionally: pulmonary hypertension, persistent effects of COVID-19 infection, and pulmonary arteriovenous malformations.

Initial laboratory results demonstrated a haemoglobin of 17.5g/dL suggestive of chronic hypoxaemia. Other laboratory results, including a brain natriuretic peptide, electrolytes, and a hepatic panel, were within normal range. Chest radiography demonstrated mild bilateral upper lung opacities representing atelectasis and/or scarring and normal cardiomediastinal silhouette. Chest CT showed partial anomalous pulmonary veins with a large sinus venosus atrial septal defect. The right upper pulmonary vein drained anomalously to the superior vena cava, and the right middle and right lower pulmonary veins drained at the level of the defect. Also, on his CT, he was noted to have a bicuspid aortic valve with a severely dilated ascending aorta (measuring 50 mm at the mid ascending level) (Supplemental Figure 1).

Given the acute onset of hypoxaemia in the setting of pulmonary disease with recent COVID-19 infection, there was uncertainty if closing the atrial septal defect would result in symptom resolution. Therefore, a cardiac catheterisation without sedation was done in the paediatric catheterisation lab with the plan to perform provocative manoeuvres.

Vascular access was obtained in the internal jugular vein for right heart haemodynamics and the femoral vein for simultaneous left heart haemodynamics across the atrial septal defect. Right and left heart catheterisations were performed with the patient lying supine. Baseline haemodynamics are shown in Figure 1. Qp:Qs was 0.8:1 with a right-to-left shunt of 0.6 L/min. His indexed pulmonary vascular resistance was 1 WU x m2.

After baseline haemodynamics showed no evidence of elevated pulmonary pressures, we performed agitated saline contrast studies in both branch pulmonary arteries, which showed no evidence of pulmonary arteriovenous malformations. We then placed the bed in Reverse Trendelenburg and the patient was sat upright (about 45 degrees from supine). Within one

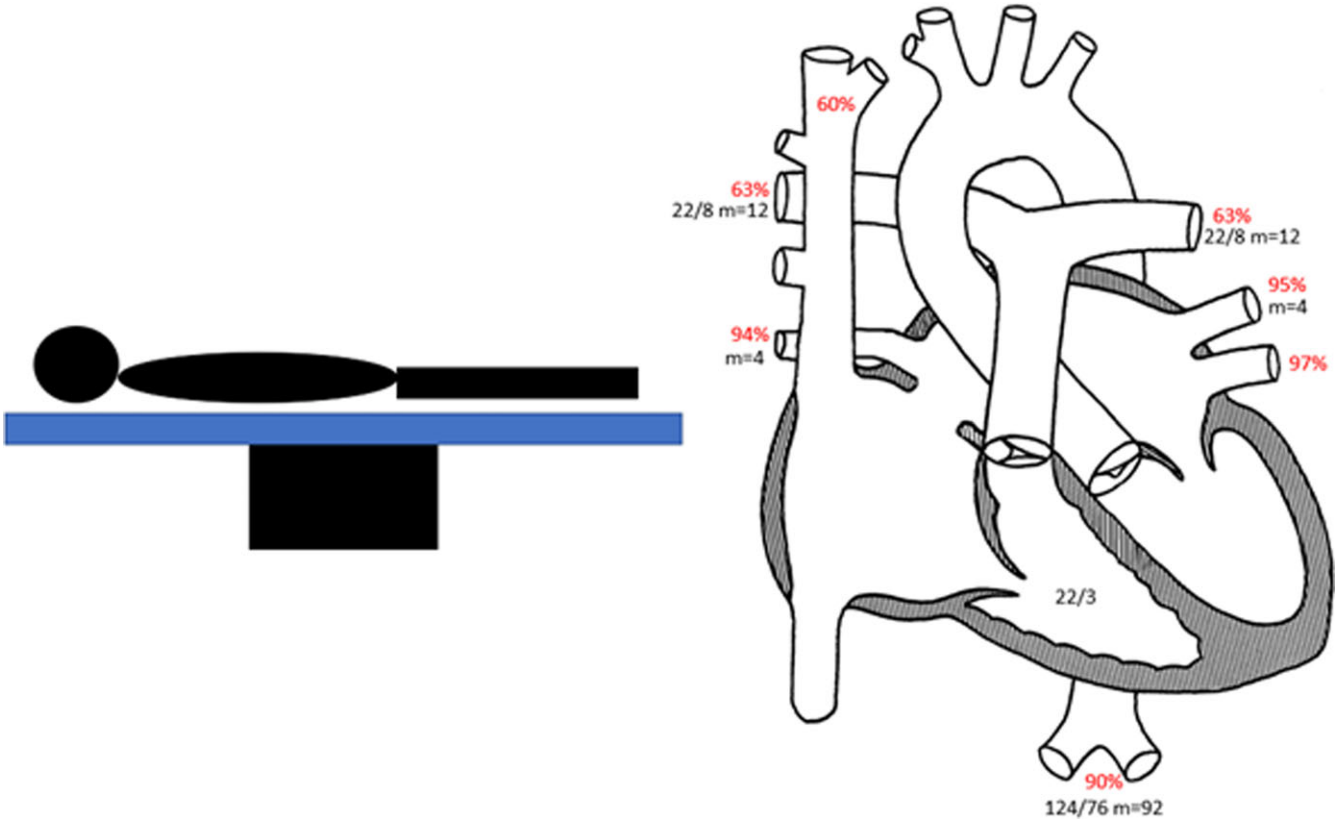

Figure 1. Patient positioning and supine haemodynamics. m = mean pressure.

minute he reported feeling dyspnoeic and his oxygen saturation decreased to 67%. Haemodynamics were repeated (Fig. 2) and demonstrated a decrease in his Qp:Qs to 0.6:1 and increase in right-to-left shunt to 1.2 L/min. Given the significant increase in his right-to-left shunt with stable pulmonary venous saturations while upright, it was clear that the interatrial shunt was the primary aetiology of hypoxaemia.

Based on these results, the decision was made to surgically close the atrial defect with baffling of the anomalous pulmonary veins via a Goretex patch and replace the aortic valve, aortic root, and ascending aorta with a 25 mm On-X valved conduit with coronary reimplantation. His postoperative course was unremarkable, he transferred out of the ICU on postoperative day 7 and discharged on postoperative day 9. The patient was symptom-free with normal oxygen saturations following surgery. He healed well from surgery and remained asymptomatic with no hypoxaemia or dyspnoea at outpatient follow-up.

Discussion

Platypnea-orthodeoxia syndrome is uncommon and can be seen rarely in cases of right-to-left interatrial shunting in patients with an atrial-level communication. However, since most patients with an interatrial shunt do not develop platypnea-orthodeoxia syndrome, it is hypothesised that it may be related to elevated right-sided-filling pressures or pulmonary hypertension.[1,2] Another theory is that there are conformation changes in the interatrial septum in the upright position that lead to increased right-to-left shunting in symptomatic patients.[3–5] Reports have described aortic root dilation or elongation in patients with platypnea-orthodeoxia syndrome and hypothesised that root enlargement changes the atrial septal conformation such that more systemic venous flow is directed across the septum.[3,4] A study from Bertaux et al. found that an increase in the diameter of the aortic root was significantly correlated with increased compression and mobility of the atrial septum as well as increased shunting at the atrial level.[5] We suspect that our patient's symptoms were most likely related to this phenomenon given his significantly dilated ascending aorta and normal right heart pressures. COVID-19 infection could have acted as a triggering event for hypoxaemia due to either right-to-left shunting from transient pulmonary hypertension or parenchymal lung injury as has been described with silent hypoxaemia syndrome following COVID infection.[6,7]

Prior case reports on platypnea-orthodeoxia syndrome often describe cardiac catheterisation data but to our knowledge, few have performed invasive haemodynamics with patients both supine and upright. Given that our patient's initial clinical picture was compounded by the diagnosis of COVID-19, we felt that performing provocative manoeuvres during catheterisation was necessary to determine if surgical closure would address his symptoms. The haemodynamic data allowed us to determine with certainty that his hypoxaemia was originating from a position-dependent increase in his right-to-left shunt in the absence of elevated right heart pressures. Our findings suggest that in patients with platypnea-orthodeoxia syndrome, there is diagnostic utility in obtaining haemodynamic data while patients are supine and sitting upright, particularly when there is uncertainty about the underlying aetiology.

Supplementary material. The supplementary material for this article can be found at https://doi.org/10.1017/S1047951124000416.

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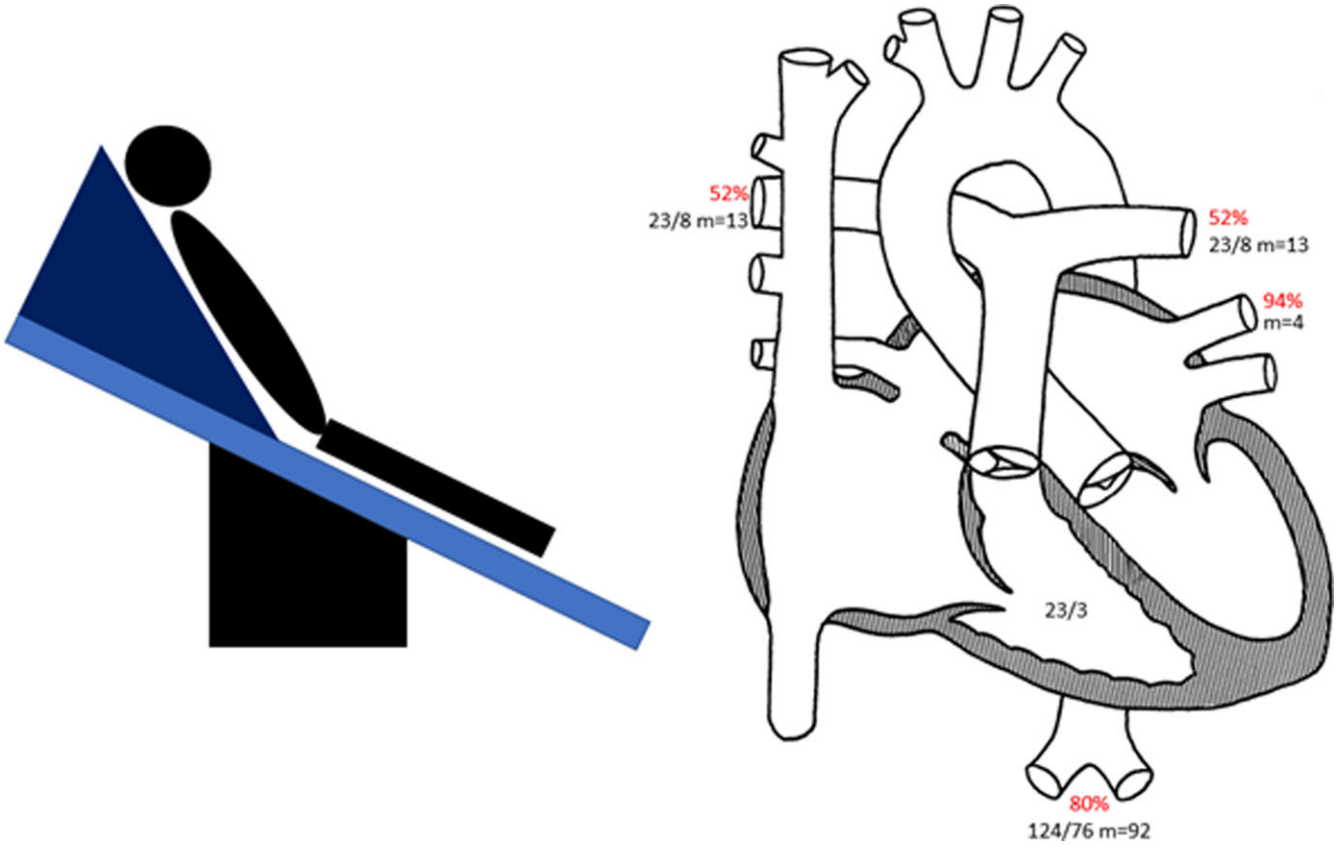

Figure 2. Patient positioning and semi-supine haemodynamics. m = mean pressure.

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Ethical standards. The patient provided consent for the publication of this report.

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