Late emergence of platypnea orthodeoxia: Chiari network and atrial septal hypertrophy demonstrated with transoesophageal echocardiography

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Platypnea orthodeoxia is a rare syndrome that describes breathlessness on standing that resolves on lying flat. We present a previously healthy patient who developed platypnea orthodeoxia in her eighth decade of life. Cardiovascular imaging demonstrated an atrial septal defect, extensive Chiari network and atrial septal hypertrophy. We propose the development of lipomatous atrial septal hypertrophy led to altered atrial compliance and a baffle to direct flow preferentially to the left heart on standing.

Case description
A 74-year-old woman was referred for echocardiography with breathlessness and saturations of 80% on room air. Respiratory investigations, including CT pulmonary angiogram, had been normal. Transthoracic echocardiography demonstrated normal left ventricular size and function with no significant valve abnormalities. Right heart was of normal size and function, with normal right ventricular systolic pressure. An atrial septal defect was identified with colour flow mapping. However, flow across the defect was from left-to-right with no evidence of a significant right-to-left shunt on agitated saline contrast injection. During the supine examination, the patient’s oxygen saturations were noted to be 97% on room air. When the patient stood up again saturations fell back to 80%. Agitated saline injection with the patient standing now resulted in striking opacification of the left ventricle consistent with a significant right-to-left shunt (Figure 1A and B, see Supplementary material online, Video 1A and B). Platypnoea orthodeoxia was diagnosed.

Transoesophageal echocardiography demonstrated a 24-mm secundum atrial septal defect. Of interest, there was an extensive mobile Chiari network (Figure 2A, see Supplementary material online, Video 2A), which moved dynamically with posture relative to a rigid, hypertrophied septum. With the patient supine the geometric arrangement of the Chiari network and septum appeared to favour preferential flow from the inferior vena cava into the right atrium. In a semi-supine position the Chiari network and inferior septum shifted towards the left atrium (Figure 2B, see Supplementary material online, Video 2B).

Platypnoea orthodeoxia describes dyspnea while ‘upright’, relieved on resumption of a supine posture.1,2 It was unclear why our patient had developed this phenomenon in the eighth decade of her life. Despite the atrial septal defect our patient had no changes in right heart pressure that would contribute to a right-to-left shunt. No previous imaging was available to know whether the size of the atrial septal defect or shunt had changed over time. However, one of the defining features of platypnea orthodeoxia is a right-to-left shunt in the presence of normal intracardiac pressures. Both an anatomical means for the right-to-left shunt—in our patient the atrial septal defect—and a functional component that alters with posture is required.3 Figure 2 demonstrates the dynamic relationship of the Chiari network with the hypertrophied septum following small changes in posture. This observation is consistent with the proposal that platypnea orthodeoxia can occur due to postural changes in atrial anatomy that preferentially direct vena caval inflow towards the left atrium.3,4 It is possible that changes in right atrial inflow also cause regional variation in right atrial pressure that contributes to the right-to-left shunt. We propose the development of atrial septal hypertrophy, due to lipomatous change, in someone with a pre-existing, extensive Chiari network accounts for the late emergence of platypnea orthodeoxia. The septal

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hypertrophy alters atrial compliance and creates a baffle for flow to be directed to the left heart.

Supplementary material

Supplementary material associated with this article can be found in the online version.

Conflict of interest: none declared.

References

Figure 2 (A) and Video 2A: Transoesophageal echocardiography with the patient lying supine demonstrates the extensive Chiari network, hypertrophied atrial septum and atrial septal defect. The Chiari network extends across the right atrium and generates laminar flow across the septum within the right atrium (arrow). (B) and Video (B) A small change in posture to the semi-supine position alters the geometry of the septum, Chiari network and septal defect with laminar flow more likely to be directed to the left atrium (arrow).