

Dynamic echocardiography in evaluation of platypnoea-orthodeoxia

Abstract

Platypnoea-orthodeoxia is an infrequent syndrome that is usually associated with positional intra-cardiac right-to-left shunting. The authors report the case of a patient who presented with deteriorating severe dyspnoea and deoxygenation in the upright position that was partially relieved by recumbent repositioning. Dynamic echocardiography provided insight into the underlying anatomical and pathophysiological mechanisms as well as guidance for definitive therapy.

Keywords: echocardiography, orthodeoxia, platypnoea.

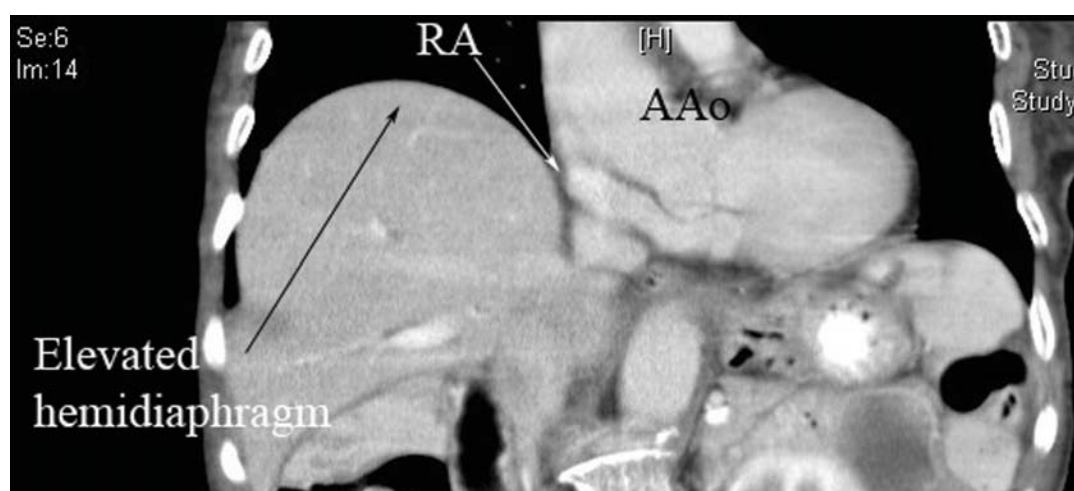


Figure 1: Chest Computed Tomography Pulmonary Angiography. Dilatation of the aortic root and ascending aorta, an eventrated anterior dome of the right hemidiaphragm containing liver and a compressed right atrium are evident.

Case report

An 83-year-old female was admitted to the high dependency unit with severe dyspnoea. She had a history of progressive dyspnoea over the last month with no other associated symptoms. Examination of the cardiovascular and respiratory system was unremarkable apart from tachypnoea. Laboratory investigations were also unremarkable with the exception of severe hypoxaemia detected on arterial blood gas analysis: PaO₂ = 30.4 mmHg, SaO₂ 60.5% and PaCO₂ = 25 mmHg. Chest x-ray revealed eventration of the right hemi-diaphragm and no visible lung pathology. Computed Tomography Pulmonary Angiography excluded significant pulmonary arterial embolism however demonstrated a large thyroid mass with retrosternal extension and without compression of great thoracic vessels, dilatation of the aortic root and ascending aorta, an eventrated anterior dome of the right hemidiaphragm containing liver and a compressed right atrium was noted (Figure 1).

It was observed that the patient repeatedly

deteriorated in the upright position and partially improved when positioned supine (PaO₂ 44 mmHg and 81 mmHg respectively). Platypnoea-orthodeoxia was further investigated with transthoracic echocardiography. It demonstrated normal size ventricular chambers and function with normal cardiac valves, dilated aortic root, small compressed right atrium and a moderate right to left shunt at the atrial level with Color Doppler (Figure 2).

A dynamic transoesophageal echocardiogram was performed in the supine and semi-erect position to further assess atrial septum anatomy and function. Spontaneous respiration was maintained during intravenous sedation administered by intermittent boluses of propofol. Patient's repositioning from supine in to semi-erect posture was performed by simply elevating the head of the patient's bed. Color Doppler, 2D imaging and agitated saline contrast studies confirmed the presence of a patent foramen ovale (Figure 3). There was a marked difference

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Figure 2: Transthoracic echocardiography, subcostal window. 2D and colour Doppler modalities. Right to left interatrial shunt is evident.

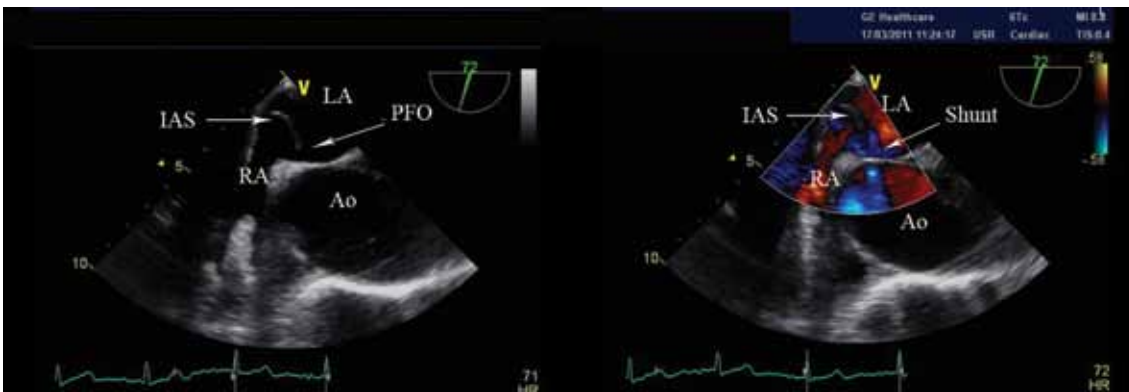


Figure 3: Transoesophageal echocardiography, 72° plane. 2D and colour Doppler modalities. PFO, dilated aortic root and right to left interatrial shunt are demonstrated.

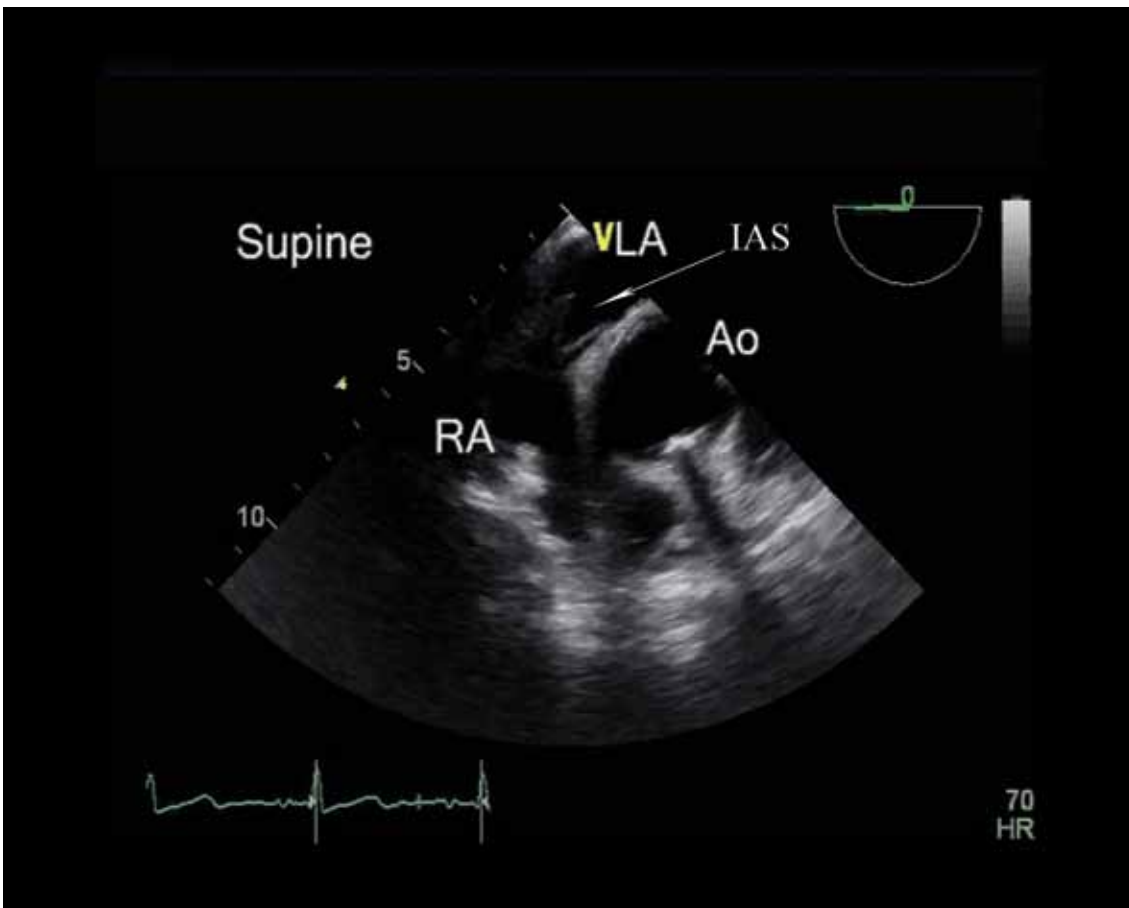


Figure 4: Transoesophageal echocardiography performed in a supine position, 0° plane. The atrial septum is intermittently buckling from the left and towards the right atrium.

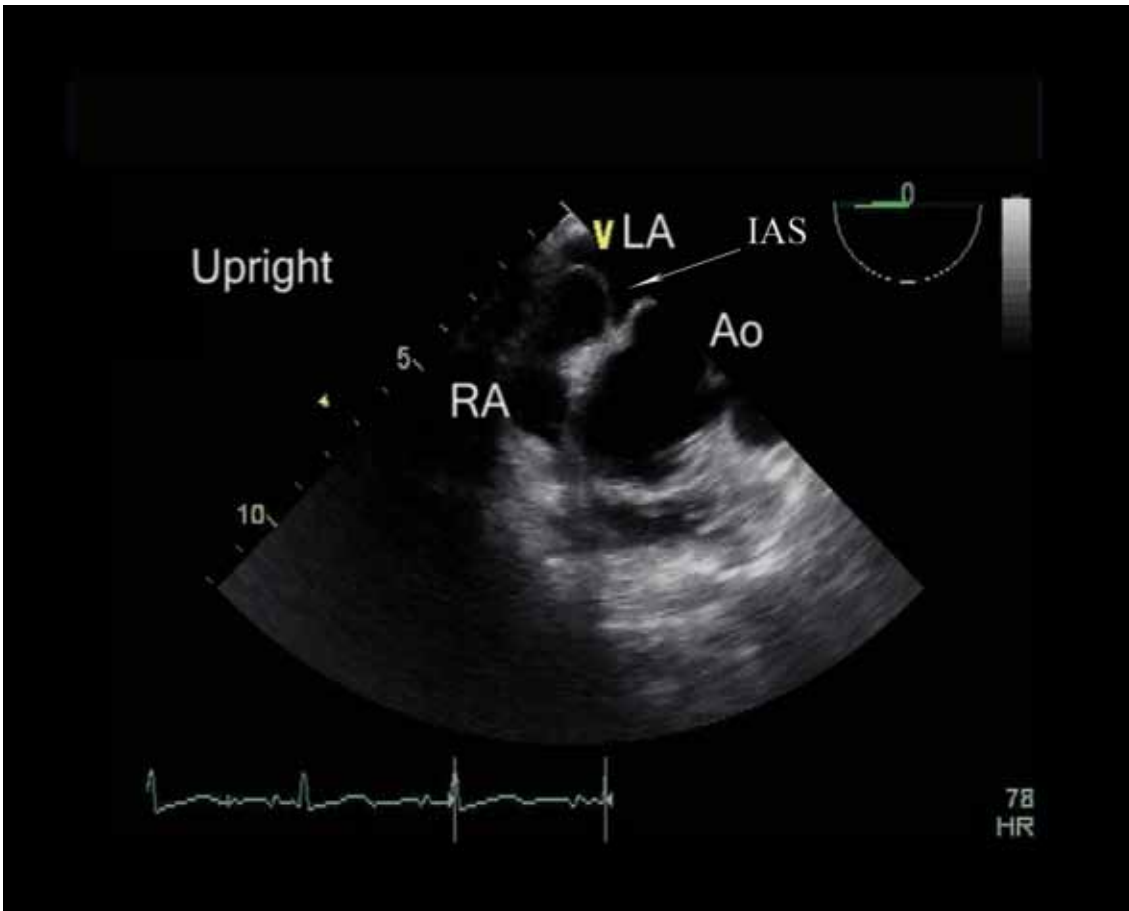


Figure 5: Transoesophageal echocardiography performed in an upright position, 0° plane. The atrial septum curvature is fixed towards the left atrium.

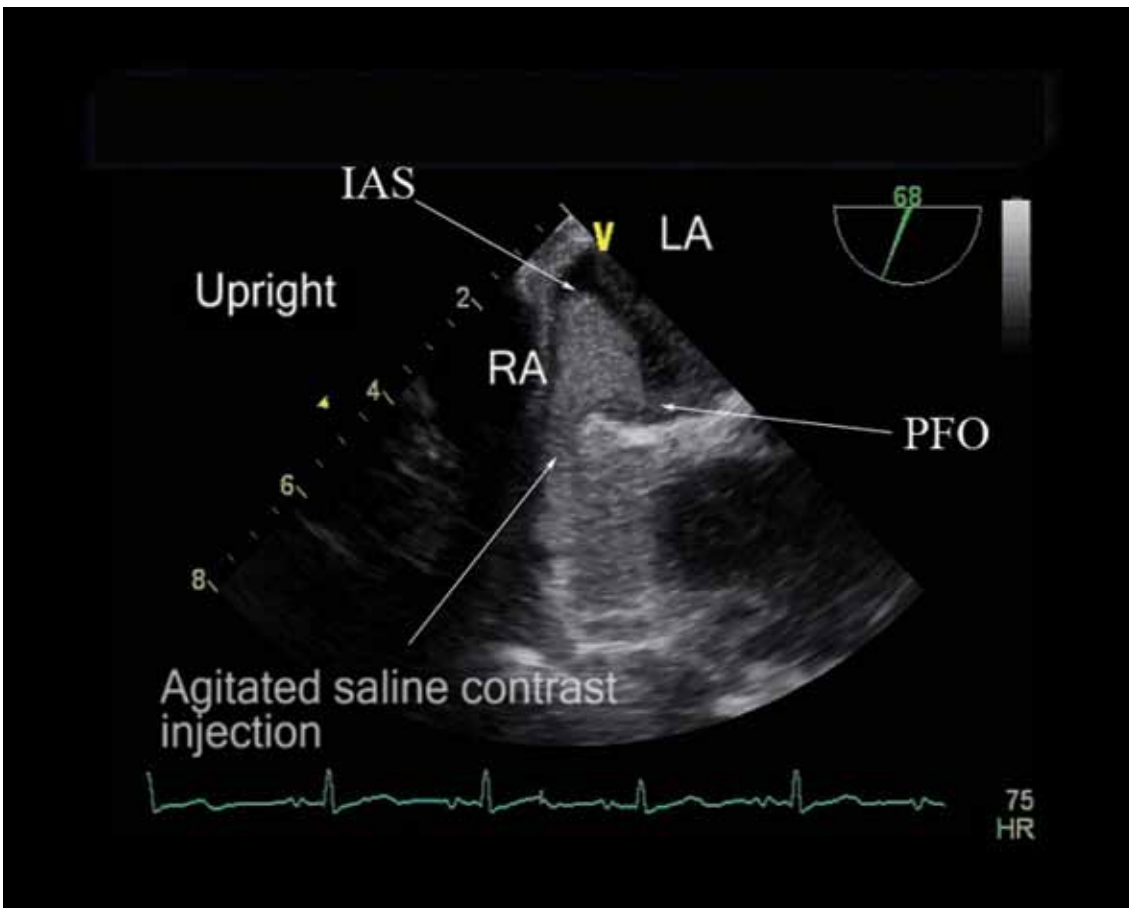


Figure 6: Transoesophageal echocardiography performed in an upright position, 68° plane. Agitated saline contrast injection confirms large right-to-left interatrial shunt.

in atrial septal curvature that was dependent on the patient's posture. The atrial septum was curved towards the left atrium with intermittent buckling to the right in the supine position (Figure 4) and persistently curved to the left when the patient was seated upright (Figure 5). Corresponding mild right-to-left shunt was evident in the supine position and a severe shunt was demonstrated semi-erect. The right atrium was small in size and appeared compressed with an ovoid shape. There was moderate lipomatous hypertrophy of the atrial septum. Agitated saline contrast study confirmed significant intra-cardiac right to left shunt at the atrial level (Figure 6).

The patient underwent percutaneous patent foramen ovale (PFO) closure with an Amplatzer device under combined fluoroscopy and transoesophageal echocardiography guidance. Right atrial and pulmonary artery pressures measured invasively prior to PFO closure were normal. PFO closure required upsizing to a 30 mm Amplatzer PFO occluder device after a 25 mm device was demonstrated to be unstable on transoesophageal echocardiography at its attachment to the lipomatous portion of the interatrial septum following the Minnesota Wiggle manoeuvre. Following PFO closure there was immediate improvement in clinical signs. The patient was able to sit upright without a significant drop in arterial oxygen saturation and without severe dyspnoea. Postural deoxygenation progressively diminished over the next few days with pulse oximetry measurements indicating oxygen saturation of 98% in the supine position and 93% in erect position on day four. Repeat echocardiography prior to discharge revealed mild residual right to left shunt in the supine position of no clinical significance.

Discussion

Platypnoea-orthodeoxia is a condition associated with symptoms of dyspnoea and deoxygenation recorded in the upright position with symptom improvement in the recumbent position. It was first described in 1949¹ with over 50 other case reports to follow.

The physiological mechanism is thought to be related to an increase in shunting of venous blood to the arterial circulation. A number of medical conditions are linked to platypnoea-orthodeoxia, including pulmonary disease (pneumonectomy, emphysema etc), liver disease (cirrhosis), large vascular malformations, and pericarditis etc.² The most frequent cause however is patent foramen ovale with shunting at the atrial level.³

It is suggested that two components must co-exist to ensure clinical symptomatology: anatomical and functional.⁴ From the anatomical point of view, an abnormal communication between right and left sides of the circulation must exist and be of a sufficient size to allow for a significant shunt. This could be a single veno-arterial communication or multiple communications in the lungs, liver or indeed inside the heart. From a functional point of view, when an atrial communication is present, an increase in right sided pressures must be sufficient to exceed left sided pressures to ensure significant right-to-left shunt and thus deoxygenation of arterial blood due to venous admix.

Many authors have attempted to explain physiological changes leading to the anatomical and functional conditions predisposing individuals to develop postural changes consistent with platypnoea-orthodeoxia.⁵ Stretch of the interatrial

communication, prominent Eustachian valve and thoracic aortic aneurysm⁶ have been linked to the anatomical changes.⁷

In our case there was a combination of several anatomical factors which attributed to the development of symptomatic platypnoea-orthodeoxia.

First, our patient had eventration of the anterior dome of right hemidiaphragm, containing liver tissue. This pathology was likely a congenital anatomical abnormality. CT chest demonstrated anatomical contact between this part of the diaphragm and the right atrium with resulting compression of the right atrium.

Second, our patient had a dilated aortic root which further contributed to the compression of the right atrium between the elevated hemi-diaphragm and aortic root, resulting in reduced right atrial volume and elevated pressure.

Third, the patient had a patent foramen ovale, which was functional in both the supine and upright positions.

Fourth, the patient had a large thyroid mass, which potentially contributed to the mediastinal mass-effect compressing the right atrium with gravity in the upright position.

The changes observed in the atrial septum curvature during the dynamic echocardiographic assessment confirmed that only transient excess of right atrial pressure over the left was present during the cardiac cycle when the patient was in a recumbent position. This allowed for only a small transient right-to-left shunt. It was markedly different when the patient was positioned upright, immediately leading to a change in the curvature of the atrial septum being fixed and bulging towards the left atrium throughout the cardiac cycle. This change supports continuous elevation of the right atrial pressure over the left when the patient was sitting or standing. Gross right-to-left shunting was also seen with agitated saline contrast studies when performed in both positions, however more marked in the semi-erect position.

Platypnoea-orthodeoxia caused by a patent foramen ovale has previously been demonstrated with contrast echocardiography performed on a tilt-table.⁸ Head of bed elevation was used in our patient to invoke postural changes during transoesophageal echocardiography.

Percutaneous PFO occlusion is the standard treatment for patients with platypnoea-orthodeoxia where PFO is shown to be the cause. Our patient underwent an uneventful procedure. Intraprocedural transoesophageal echocardiography proved useful in confirming the mechanism and need to appropriately upsize the Amplatzer PFO occluder device.

We conclude that echocardiography is an essential technique for diagnosis and treatment of patients with platypnoea-orthodeoxia. By performing dynamic echocardiographic studies in the recumbent and erect positions we were able to demonstrate not only the presence of a PFO as a cause of symptoms in our patient but also explain a combination of intricate anatomical and physiological changes leading to the symptoms. We suggest that dynamic echocardiography should be routinely employed in the investigations of patients with platypnoea-orthodeoxia.

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