

Platypnea-orthodeoxia due to osteoporosis and severe kyphosis: a rare cause for dyspnea and hypoxemia

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Abstract

Platypnea orthodeoxia is a rare disorder characterized by dyspnea and arterial desaturation, exacerbated by the upright position and relieved when the subject is recumbent. We report the case of a 79-year old woman admitted to hospital with dyspnea who was thought to have restrictive ventilatory impairment due to osteoporosis and severe kyphosis. Interestingly, the dyspnea was aggravated in the upright position, whereas the symptoms improved in the supine position. Arterial blood gas analysis confirmed orthodeoxia. The lung function test showed only a mild obstructive and restrictive ventilation disorder. Echocardiography revealed a patent foramen ovale and an aneurysm of the atrial septum protruding into the left atrium, despite normal right atrial pressure. Transesophageal echocardiography showed a prominent

Eustachian valve guiding a blood flow from the inferior vena cava directly onto the atrial septum, thereby pushing open the patent foramen ovale. Contrast-enhanced echocardiography confirmed a spontaneous right-to-left shunt through the patent foramen ovale. It was assumed that the platypnea-orthodeoxia was caused by a prominent Eustachian valve redirected to the patent foramen ovale as a result of severe osteoporosis with subsequent thoracic kyphosis and a change in the position of the entire heart. The patient underwent permanent transcatheter closure of the patent foramen ovale after hemodynamic assessment had confirmed a significant right-to-left shunt through it. After the procedure the arterial oxygen pressure increased significantly in the upright position and dyspnea improved.

(FEV1) 68%, FEV1 / VC 70%, total lung capacity 3.85 L (76%), residual volume 1.98 L (87%). Arterial blood gas testing performed in a supine position showed a pO₂ of 60 mm Hg while breathing 8 L of oxygen per minute. The pO₂ decreased to 53 mm Hg after moving into an upright position. Transthoracic echocardiography showed mildly reduced left ventricular function and an aneurysm of the atrial septum which unusually was protruding into the left atrium. Transesophageal echocardiography showed a prominent valve (Eustachian valve) of the inferior vena cava (IVC) at its junction with the right atrium. After intravenous injection of an ultrasound contrast medium (Echovist®, BayerVital), numerous contrast microbubbles were detected in the left atrium even without performing a Valsalva-maneuver (Figure 1). The appearance of contrast microbubbles in the left atrium indicates the presence of a spontaneous right-left shunt at the atrial level, as the contrast agent cannot pass through the lung capillaries.

Case Report

A 79-year old woman was admitted to hospital having experienced increasing shortness of breath over the previous months. Interestingly, the dyspnea improved with rest and lying in a supine position while the patient became severely cyanotic when upright and during minimal exercise. Her past medical history included arterial hypertension and repeated strokes with minor disabilities. The patient also suffered from severe osteoporosis of the spine with a history of several vertebral fractures. Coronary artery disease had previously been excluded by coronary angiography.

On hospital admission, the patient was almost unable to sit up. She was tachypnoeic and showed a cyanosis of the lips. The auscultation of the lungs was unremarkable. There was a grade 1/6 systolic murmur at the cardiac apex. A pronounced hyperkyphosis of the thoracic spine was noted. Lung X-ray was normal. Pulmonary embolism was excluded by computed tomography. Spirometry exhibited a mild obstructive and restrictive ventilation disorder: vital capacity (VC) 1.87 L, forced expiratory volume in one second

Color Doppler imaging showed a jet-like blood flow, arising from the IVC. The jet was redirected by the prominent Eustachian valve straight to the atrial septum, thereby pushing open the valve-closed patent foramen ovale (PFO). Interestingly, right heart catheterization with hemodynamic assessment revealed normal pressure: right atrium 8 mm Hg (mean), right ventricle 19/4 mm Hg, pulmonary artery 19/5 mm Hg. A significant right-to-left shunt was calculated by oxymetry: 20% of the systemic circulation in an upright position, and 10% while supine, respectively. To further assess the relevance of the PFO for the patient's dyspnea, the PFO was probed with a deflated Swan-Ganz catheter (7 F) from the right atrium. Subsequently, the balloon at the tip of the catheter was inflated in the left atrium and the PFO was temporarily occluded by pulling back the balloon against the atrial septum. This maneuver resulted in an immediate increase in arterial oxygen saturation from 90% to 98%. The PFO was then permanently closed with a 25 mm Amplatzer PFO occluder

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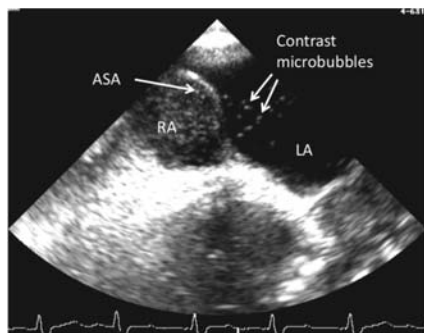


Figure 1. Transesophageal contrast echocardiography shows the right atrium and the protruding atrial septal aneurysm, both filled with intravenously injected ultrasound contrast microbubbles. Contrast microbubbles (arrows) in the left atrium indicate the presence of a right-left shunt through a patent foramen ovale.

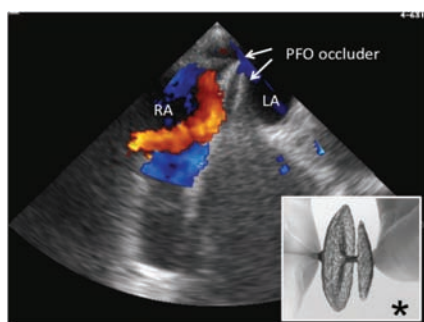


Figure 2. Transesophageal echocardiography shows the PFO occluder in the correct position sealing the PFO in the atrial septum. Colour Doppler imaging shows a venous blood stream (red) arising from the IVC and rebounding at the right-sided part of the occluder device. *Image shows a model of the Amplatzer PFO occluder used. LA, left atrium; RA, right atrium.

device (Figure 2). After the procedure the patient became less symptomatic and the arterial oxygen pressure increased significantly to 68 mm Hg in an upright position while breathing normal room air.

Discussion

A patent foramen ovale (PFO) is a remnant of normal fetal circulation. In approximately 25-30% of adults, the foramen ovale does not completely seal after birth. The vast majority of people with a PFO are asymptomatic. However, in some patients, a PFO can serve as a bypass for phenomena such as thrombi, metabolites, etc. Clinically, a PFO is linked to a variety of disease entities, including stroke, migraine headache, decompression sickness, high-altitude pulmonary edema, and platypnea-orthodeoxia syndrome. Platypnea-orthodeoxia is a rare syndrome characterized by the development of hypoxia and breathlessness in the upright position and improvement when recumbent. The most common presentation is respiratory insufficiency related to a PFO and right-to-left shunting.

Right-to-left shunting through a PFO despite normal intracardiac and pulmonary artery pressures can be explained by the flow phenomenon.¹ With aging, a prominent Eustachian valve becomes redirected to the PFO. This deformation results in a blood flow arising from the inferior vena cava towards the atrial septum and directly into the left atrium. Hyperkyphosis and spinal shortening, as found in our patient, are assumed to alter intrathoracic relationships and thereby facilitate shunting.² The upright position can

make these intrathoracic changes more pronounced and lead to a stretching of the PFO resulting in more shunting of venous blood into the left atrium. Enlargement and changes in the compliance of the heart chambers, as well as a change in heart position due to aortic root enlargement, right pneumonectomy, pericardial effusion or obesity, are other mechanisms that are thought to contribute to right-to-left shunting.³⁻⁵

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