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CASE REPORT

Dyspnea in a nonagenarian: The usual suspects, an unexpected culprit

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Abstract Platypnea-orthodeoxia syndrome (POS) is an uncommon syndrome characterized by dyspnea and hypoxemia triggered by orthostatism and relieved by recumbency. It is often associated with an interatrial shunt through a patent foramen ovale (PFO). We report the case of a 92-year-old woman initially admitted in the setting of a traumatic femoral neck fracture (successfully treated with hip replacement surgery) in whom a reversible decline in transcutaneous oxygen saturation from 98% (in the supine position) to 84% (in the upright position) was noted early post-operatively. Thoracic multislice computed tomography excluded pulmonary embolism and severe parenchymal lung disease. The diagnosis of POS was confirmed by tilt-table contrast transesophageal echocardiography, which demonstrated a dynamic and position-dependent right-to-left shunt (torrential when semi-upright and minimal in the supine position) through a PFO. The patient underwent percutaneous closure of the PFO with an Amplatzer device, which led to prompt symptom relief and full functional recovery.

PALAVRAS-CHAVE
Síndrome platipneia-ortodeoxia;
Inclinação dinâmica;

Dispneia em doente nonagenária. Os suspeitos do costume, um culpado inesperado

Resumo A síndrome platipneia-ortodeoxia (SPO) é uma entidade rara caracterizada por dispneia e hipoxemia desencadeadas pelo ortostatismo e aliviadas pelo decúbito. Está frequentemente associada à presença de um *shunt* inter-auricular através de um *foramen* ovale patente (FOP). Relata-se o caso de uma mulher de 92 anos, internada inicialmente por fratura traumática

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do colo do fémur. Foi submetida a artroplastia da anca sem complicações. No período pós operatório inicial observou-se um declínio reversível da saturação de oxigênio de 98% em decúbito dorsal para 84% na posição ortostática. A angio-tomografia computorizada do tórax excluiu trombo-embolia pulmonar e doença grave do parênquima pulmonar. O diagnóstico de SPO foi confirmado por ecocardiografia transesofágica contrastada (soro agitado) com inclinação na mesa de tilt, que demonstrou um *shunt* direito-esquerdo dinâmico e posicional (torrencial a 45° e mínimo a 0°) através de um FOP. A doente foi submetida a encerramento percutâneo do FOP com dispositivo Amplatzer, que proporcionou alívio sintomático imediato e permitiu uma recuperação funcional total.

Introduction

Platypnea-orthodeoxia syndrome (POS) is characterized by dyspnea and hypoxemia induced by orthostatism and relieved by the supine position.[1] It is a rare but probably underestimated cause of dyspnea that results from the postural accentuation of an intracardiac or pulmonary right-to-left shunt, leading to arterial oxygen desaturation. The most common etiologic association is an interatrial right-to-left shunt through a patent foramen ovale (PFO) or an atrial septal defect.[3] Although a right-to-left pressure gradient usually drives the shunt, it can occur in the absence of an elevated right atrial pressure.[2,4] In the latter cases, embryonic remnants (such as a prominent persistent Eustachian valve or a Chiari network)[5,6] or acquired anatomical features (like pulmonary resection, aortic aneurysm, aortic elongation, pericardial effusion, constrictive pericarditis and kyphoscoliosis[3,7]) can both direct the flow from the inferior vena cava through the fossa ovalis and distort the normal atrial and septal arrangement. Individually or in association, these can create a specific anatomical and functional condition leading to a right-to-left shunt boosted by orthostatism. Although the ultimate underlying mechanisms are unknown, one possibility is that the upright position might stretch the interatrial defect, allowing streaming of systemic venous blood into the left atrium. The treatment of choice is percutaneous closure of the interatrial communication.[8]

Case report

A 92-year-old woman was admitted to hospital with a left femoral neck fracture caused by a non-syncopal fall at home.

Relevant medical history included hypertension, dyslipidemia, two previous strokes (at the ages of 75 and 82 years) without significant residual motor or cognitive deficits, osteoporosis and severe kyphoscoliosis. She was on treatment with aspirin, simvastatin and losartan. Within the year prior to the admission the patient's family had noticed a progressive functional decline, with undue fatigue and dyspnea for progressively smaller efforts, which were partially relieved by stooping and squatting.

Following admission, hip replacement surgery was undertaken on day 3 of hospital stay, with no relevant complications. During routine early post-surgical observation in the intensive care unit, she was found to be hypoxic when sitting for the first time after the intervention. Physical examination was unremarkable, but interestingly the observed breathlessness and desaturation improved after lying flat and were reproducible on subsequent mobilizations. Blood chemistry, coagulation panel and blood cell count were normal. NT-proBNP level was within the age-adjusted normal range (326 pg/dl). Blood gas analysis, taken in the upright position, showed normal pH and $PaCO_2$ (7.41 and 42 mmHg, respectively) and reduced PaO_2 (42 mmHg). The chest X-ray revealed a tortuous proximal aorta with clear lung fields. Given the clinical setting, a contrast-enhanced chest computed tomography scan was performed, which excluded both pulmonary embolism and parenchymal lung disease as potential causes. However, severe kyphosis, aortic elongation and a grossly distorted relationship of the aortic root and proximal ascending aorta with the right atrium were noticed (Figure 1). A transthoracic echocardiogram showed normal left and right ventricular systolic function and chamber dimensions, mild left ventricular diastolic dysfunction, normal sized atria, no significant valve disease and no signs of pulmonary hypertension (pulmonary artery systolic pressure of 32 mmHg). A bubble contrast study revealed a mild right-to-left atrial shunt in the supine position without Valsalva maneuver, which increased significantly when sitting (Figure 2). In order to further assess the relationship between body position and the intensity of the shunt, a tilt-table assisted transesophageal echocardiogram was subsequently performed. While the patient was lying flat, the interatrial septum was redundant and tended to bow towards the left atrium; a small separation of the septum primum and septum secundum was seen, increasing significantly to 4 mm at 45°, unmasking a large shunt by color Doppler flow (Figure 3). There was no evidence of embryonic remnants, including a prominent persistent Eustachian valve or a Chiari network. The contrast study confirmed a minimal right-to-left shunt through the PFO while lying flat, which became significantly larger in the semi-upright position (Figure 3). During the tilt-table assisted echocardiographic imaging, right and left atrial pressures were studied. Central venous pressure was measured directly in the right