

The platypnea-orthodeoxia syndrome: an unusual case of dyspnea

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Dear Editor,

We read with great interest the brief case report by Carbonelli et al. [1] that highlights the pivotal role of a correct physical examination in the diagnosis of platypnea-orthodeoxia syndrome (P-OS) that occurred after a sublobar lung resection. The P-OS is a rare clinical manifestation characterized by dyspnea with a decrease in arterial blood oxygen saturation, which occur when the patient is sitting or standing up, and is relieved in the supine position [2]. It is frequently related to a patent foramen ovale (PFO), but it is itself not sufficient to produce clinically detectable issues (Fig. 1). Conditions leading to an increase in right heart pressures, such as pulmonary arteriovenous shunt, chronic obstructive pulmonary disease, surgical lung resection, pulmonary embolism, constrictive pericarditis, and kyphoscoliosis, can increase the right-to-left shunting, and may lead to a P-OS [3]. However, it is known that not all patients with P-OS have elevated right heart pressures.

We recently experienced a case of progressive dyspnea that worsened in the orthostatic position in an 83-year-old woman. She was affected by sarcoidosis, arterial hypertension, diabetes and chronic HBV infection. The physical examination revealed: blood pressure, 120/80 mmHg; pulse rate, 99 beats/min, and regular; bilateral medium basilar inspiratory crackles on chest auscultation; and no cardiac murmurs. Peripheral blood oxygen saturation was

99 % while supine, and 80 % in the upright position on 60 % oxygen with face mask. Electrocardiogram demonstrated a sinus rate of 99 beats/min, with a right bundle branch block pattern. Laboratory evaluation revealed no significant abnormalities. On chest X-ray study, bilateral pleural effusions and aortic elongation without cardiomegaly were found. A computed tomographic pulmonary angiogram showed a mild lung fibrosis and bilateral pleural effusion with no evidence of pulmonary embolism or arteriovenous malformation. A transthoracic echocardiogram revealed an interatrial septal aneurysm with to-and-fro flow, preserved biventricular systolic function and dilatation of the ascending aorta. The transesophageal echocardiogram with bubble test confirmed atrial septal aneurysm with PFO and right–left microbubble flow at rest and after Valsalva maneuver. Microbubble flow increased in the upright position. Right ventricular systolic pressure was estimated to be in the high end of the normal range, based on the velocity of the tricuspid regurgitant jet, with no evidence of pulmonary hypertension. A right-heart catheterization confirmed normal pulmonary pressure values and transcatheter closure of the PFO was performed. Post-procedure saturation testing revealed that resting arterial oxygen saturation had increased to 95 % in both the supine and standing position with no need for supplemental oxygen.

The physiologic mechanism responsible for right-to-left shunting induced by postural changes in patients with PFO is not fully understood. Furthermore, it is more difficult to explain right-to-left shunting when pulmonary pressures are normal. Alterations of atrial geometry, like elongation of the ascending aorta or atrial septal aneurysm, that affect the position and anatomy of the heart, may enhance the communication between the atria, potentially leading to right atrial remodeling and persistent shunting [4]. An

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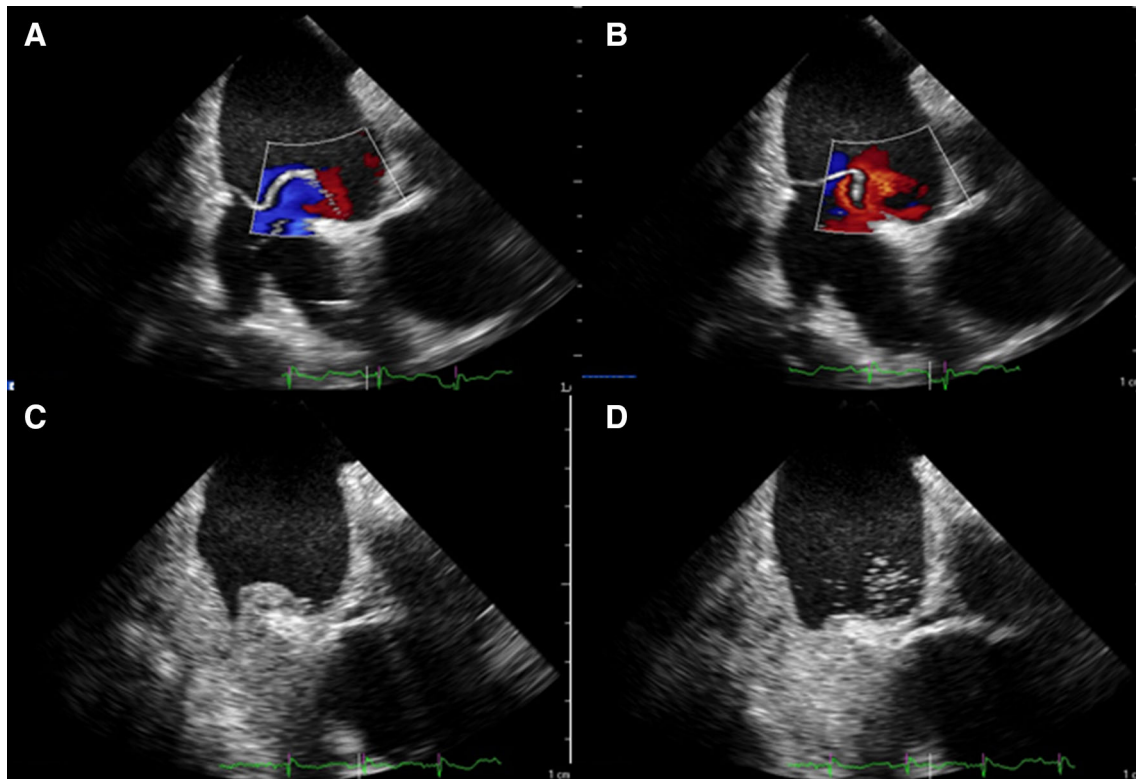


Fig. 1 a, b Atrial septal aneurysm with patent foramen ovale (PFO). c, d Micro-bubble flow from the right atrium to left atrium through the PFO

enlarged ascending aorta can give a horizontal direction to the atrial septum and a counterclockwise rotation of the heart, twisting the right atrium and favoring caval inflow against a PFO, especially in the sitting and orthostatic position [2]. In our case, the main anatomical factors that concurred to transient, right-to-left shunting induced by postural changes were the enlarged aortic root and the atrial septal aneurysm. Indeed, the aortic root enlargement distorted interatrial septum and reduced the distance between the aorta and the atrial posterior wall, favoring caval inflow. The concomitant presence of an atrial septal aneurysm facilitated the movement of the flap valve of the fossa ovalis and determined the maintenance of the foramen ovale wide open [5].

In conclusion, this is a typical case of P-OS with normal pulmonary pressure, where the concomitant presence of atrial geometry alterations and PFO leads to a transient right-to-left shunt. The clinical history of dyspnea worsening in the orthostatic position, together with the finding of arterial desaturation on accurate physical examination can raise the clinical suspicion of P-OS and guide the diagnostic and therapeutic approach.

Conflict of interest The authors declare that they have no conflict of interest.

Statement of human and animal rights All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with human and animals performed by any of the authors.

Informed consent None.

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