

Platypnea-orthodeoxia induced by fenestrated atrial septal aneurysm

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Platypnea-orthodeoxia is a peculiar syndrome characterized by a right-to-left shunt, which occurs in the upright position. The diagnosis is made by contrast transesophageal echocardiography, paying attention to include contrast visualization in the orthostatic decubitus. The association of this syndrome with a fenestrated atrial septal aneurysm is rare and probably underlies a peculiar and also rare mechanism of shunting in presence of normal pulmonary pressure. We report of a case of a 58-year-old man with a fenestrated atrial septal aneurysm and platypnea-orthodeoxia syndrome treated by surgical closure of the atrial defect.

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Introduction

Platypnea-orthodeoxia is a syndrome described for the first time in 1949 by Burchell et al.¹, characterized by dyspnea occurring in (or exacerbated by) the upright position (i.e., platypnea), because of arterial oxygen desaturation similarly induced or accentuated by the upright position (i.e., orthodeoxia). Interatrial right-to-left shunt through an atrial septal defect (ASD) or a patent foramen ovale (PFO) is the most common cause of this syndrome^{2,3}. Here we report a case of platypnea-orthodeoxia syndrome associated with fenestrated atrial septal aneurysm.

Case report

A 58-year-old man was referred to our Institution for further investigation of an interatrial septal aneurysm associated with cyanosis. In 2003, because of the occasional finding of unexplained polycythemia (hematocrit 60%, hemoglobin 21.2 g/dl, and red blood cell count 6 330 000/mm³) he underwent a bone marrow biopsy, which was normal. In the same year he suffered a transient ischemic attack involving the left cerebral hemisphere, with no sign of ischemic scar or hemorrhage at computed tomography scan; no sources of thromboembolism were immediately evident. In the subsequent work-up, transesophageal echo-

cardiography examination revealed an atrial septal aneurysm, with right-to-left shunt documented after intravenous injection of saline. The patient was thus referred to our Institution for cardiac catheterization. On admission, the patient complained of no symptoms other than moderate postural dyspnea in the upright position. Clinical examination revealed cyanosis and digital clubbing. Auscultation of the heart revealed a paradoxical splitting of the second tone. The ECG showed left bundle branch block and a first-degree atrioventricular block. Lung fields and heart size appeared normal at chest X-ray. He was assuming only prophylactic warfarin after the transient ischemic attack. He underwent right and left heart catheterization, and coronary angiography study. Anomalous systemic venous return and pulmonary artery fistula were excluded. Coronary arteries were normal. Right heart pressures were normal: right atrium (mean 9 mmHg), right ventricle 28/4 mmHg, pulmonary artery 28/13 mmHg (mean 28 mmHg), wedge pressure 11 mmHg. Catheterization showed a right-to-left shunt (Qp/Qs 0.89), blood oxygen saturation being 87.6% in aorta, 97% in pulmonary veins, and 72.2% in pulmonary artery. Arterial oxygen saturation was 67% in orthostatic position and 80% in recumbent position. Transesophageal echocardiography was then performed with the patient still in the cath lab. Because of his symptoms of postural dyspnea, saline was inject-

ed to visualize the shunt first through the right femoral vein, and subsequently through the basilica vein of the right arm. The right-to-left shunt was more evident after right femoral than basilica vein injection (Fig. 1). An intracardiac echocardiography was then performed with a 9F-9 MHz, 110 cm long transducer (Ultra ICE, EP Technologies, Boston Scientific Corporation, San Jose, CA, USA). A very large aneurysm of the interatrial septum was observed, with a wide septal excursion.

The patient was referred to the surgeon for closure of the defect because of the very large dimension of the aneurysm (> 4 cm) and the absence of an adequate rim for percutaneous closure⁴. Surgical exploration revealed a redundant atrial septum and a cribriform aneurysm, with a main hole facing the inferior vena cava orifice; additional holes were observed in the cranial portion of the fossa ovalis. After cardioplegic cardiac arrest in extracorporeal circulation the redundant portion of the interatrial septum was removed (Fig. 2) and the hole closed with a dacron patch. The subsequent hospital course was uneventful and the patient was discharged after 6 days with normal oxygen arterial saturation and no dyspnea.

Discussion

Platypnea-orthodeoxia is a peculiar clinical entity characterized by postural hypoxemia and dyspnea induced by the upright position. Although the pathophys-



Figure 2. Interatrial septal aneurysm after surgical removal.

iological mechanism leading to orthostatic desaturation has not been fully elucidated, a prerequisite is the presence of a right-to-left shunt. Accordingly, the etiologic classification of the platypnea-orthodeoxia is based on the site of right-to-left shunt: 1) intracardiac (PFO, ASD, perforated aneurysm); 2) pulmonary vascular (pulmonary artery-pulmonary vein communications); 3) pulmonary parenchymal shunts (areas of low ventilation-perfusion ratio). Several clinical conditions have been associated with platypnea-orthodeoxia syndrome, including chronic arterio-venous shunt⁵, chronic obstructive pulmonary disease with position-dependent ventilation-perfusion mismatch, pharmacological tox-

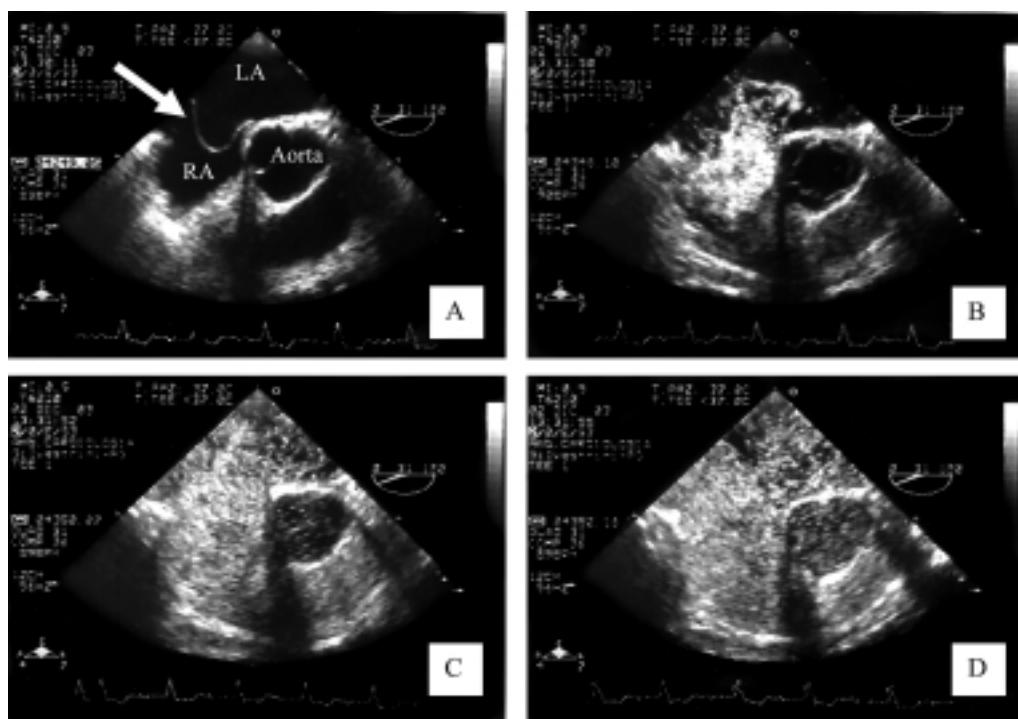


Figure 1. Transesophageal echocardiography view showing the redundant atrial septal aneurysm (arrow) (A). The sequence in B, C and D demonstrates the right-to-left shunt after injection of saline through the right femoral vein. LA = left atrium; RA = right atrium.

city, such as amiodarone and propafenone⁶⁻⁸, interatrial shunt in conjunction with pneumectomy⁹, pulmonary embolism, loculated pericardial effusion¹⁰, constrictive pericarditis, ascending aortic enlargement^{11,12}, and diabetic autonomic neuropathy¹³.

Development of platypnea-orthodeoxia in the presence of a right-to-left interatrial shunt and normal pulmonary pressures is thought to result from a combination of hemodynamic and flow phenomena. In patients with ASD or PFO and normal pulmonary pressure, a right-to-left shunt across the interatrial defect may occur during early diastole^{14,15}, because blood flowing from the inferior vena cava might cross the interatrial septal defect by kinetic energy, in the absence of a pressure gradient between the right and left atrium. Decreased right ventricular compliance might enhance the phenomenon. It is also observed in right ventricular infarction and mechanical ventilation, particularly with increased pulmonary end-expiratory pressure and it could be demonstrated by Valsalva maneuver. Another mechanism is anatomical, secondary to a prominent Eustachian valve that becomes redirected toward the foramen ovale with aging¹⁶. In this case, the blood flowing from the inferior vena cava can be directed to the left atrium by way of an over-developed Eustachian valve or a high ASD close to the superior vena cava. The last mechanism is also anatomical, but is much more rare, because it is associated (as the case we report here) with an atrial fenestrated septal aneurysm. It was described for the first time by Thomas et al.¹⁷ in 1987. The shunting occurs almost exclusively from the inferior vena cava (which is the reason why the echocontrast was assessed after injection both via the basilica vein and femoral vein) because a flap of septum secundum, adjacent to the inferior vena cava orifice, intercepts blood flow shunting into the left atrium like a "spinnaker sail", whether or not a persistent Eustachian valve coexists. This condition is exacerbated by upright position that could stretch the interatrial communication.

In conclusion, platypnea-orthodeoxia is a relatively rare syndrome generally observed in elderly subjects. A major problem with this disease is its clinical recognition, which requires a high level of suspicion. A potential hazard related to this condition is the possibility of paradoxical systemic embolism, as in the case we describe. Echocontrast transesophageal echocardiography by saline injection both in an antecubital and in a femoral vein is particularly helpful. Transcranial Doppler might represent a less invasive approach to detect atrial shunt through ASD or PFO. Transcranial Doppler has been used for PFO diagnosis by detection of air microemboli in middle cerebral arteries after peripheral injection of agitated saline. Some studies suggest that transcranial Doppler may detect clinically significant shunts^{18,19}. Treatment of platypnea-orthodeoxia is via percutaneous^{20,21} or surgical atrial septal closure, and it allows prompt correction of the hypoxemia and breathlessness.

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