

Pseudo-pulmonary Embolism – A Case of Hypoxemia Associated with Right-to-left Shunt

Mariana Faustino¹, Ana Oliveira Soares¹, Fernando Rodrigues², Rui Anjos³, António Freitas¹, Victor M. Gil¹

Serviço de Cardiologia, Hospital Professor Fernando Fonseca¹, Amadora; Serviço de Pneumologia, Hospital Professor Fernando Fonseca², Amadora; Serviço de Cardiologia Pediátrica, Hospital de Santa Cruz, Centro Hospitalar de Lisboa Ocidental³, Lisboa – Portugal

Introduction

Platypnea orthodeoxia is a rare syndrome characterized by dyspnea and decreased arterial oxygen saturation when passing from the dorsal decubitus position to the sitting or upright position¹. It usually results from an intracardiac right-to-left shunt; however, the pathophysiological mechanism responsible for the positional nature of the shunt has not been clarified^{1,2}. We report a case of dyspnea and orthostatic hypoxemia, which was attributed to a right-to-left shunt due to atrial septal defect (ASD).

Case Report

The patient is a 75-year-old male with no relevant personal antecedents. He underwent elective right hip arthroplasty, and, on the fourth postoperative day, after beginning to walk, dyspnea, polypnea and low arterial oxygen saturation appeared.

His physical examination findings were as follows: arterial blood pressure 120/70 mm Hg; heart rate 68 bpm; respiratory rate 30 bpm; peripheral saturation 80%. His cardiac and pulmonary auscultations were uneventful, and neither jugular venous distention nor peripheral edema was observed.

Regarding laboratory findings, his blood gas analysis (under supplementary oxygen at 5 L/minute) stood out, with an arterial oxygen partial pressure (PaO₂) of 47 mm Hg and respiratory alkalosis. Relevant changes were observed in neither hematological nor biochemical parameters. The electrocardiogram and chest X-ray showed no significant changes.

Chest computed tomography angiography (CT-angio) revealed small and hypodense filling defects in the middle and right inferior lobar arteries, in addition to slight dilation of the ascending aorta (45 mm).

The transthoracic echocardiogram findings were as follows: cardiac chambers of normal dimensions; good left and right ventricular function; minimum tricuspid regurgitation; no pulmonary hypertension; exuberant Eustachian valve; slightly dilation of the aortic root and ascending aorta (45 mm). No ASD was identified.

Keywords

Dyspnea; Echocardiography; Transesophageal; Heart Septal Defects; Atrial.

Mailing Address: Mariana Faustino •

Avenida Marquês de Tomar, 89 6ªA. Postal Code: 1050-154, Lisboa – Portugal
E-mail: marianafaustino85@gmail.com
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Pulmonary embolism was diagnosed and enoxaparin therapy initiated. Periods of hypoxemia related to the sitting position persisted, and total normalization of peripheral saturation was obtained with the dorsal decubitus position.

Considering the hypothesis of intracardiac right-to-left shunt, transesophageal echocardiogram (TEE) with agitated saline contrast was performed, revealing exuberant atrial septal (AS) aneurysm of the *ostium secundum* type, allowing the contrast medium to pass to the left chambers, even with no Valsalva maneuver (Figure 1A).

In an attempt to clarify the positional variation of arterial oxygen saturation, TEE was also performed on a tilt table, under serial blood gas monitoring. At 60°, the prolapse of the AS aneurysm to the left atrium was maintained, and the contrast flow to the left chambers was immediate and greater than that observed in the decubitus position (Figure 1B). Concomitantly, PaO₂ decreased from an initial value of 67 mm Hg, in the decubitus position, to 43 mm Hg (under normal conditions).

After one week, the patient underwent a new high-resolution CT-angio, which showed no filling defects in the pulmonary circulation, and the review of the previous exam raised the possibility of a false-positive exam. Pulmonary ventilation/perfusion scan showed neither perfusion nor diffusion defects, and the functional respiratory tests showed no obstructive, restrictive or diffusion changes.

The patient underwent percutaneous ASD closure by placement of a cribriform device under fluoroscopic and intracardiac echography control.

The TEE with agitated saline was repeated and showed a well-positioned device adjacent to the AS aneurysm, with persistence of the right-to-left contrast medium flow at a lower degree than in the previous exam, and almost exclusively associated with the Valsalva maneuver. That flow did not increase at the 60° position on the tilt table; however, a PaO₂ reduction from 102 mm Hg to 75 mm Hg was observed (Figure 2).

Discussion

This is the report of a case of platypnea orthodeoxia caused by an ASD with right-to-left shunt and no evidence of pulmonary hypertension in a patient previously diagnosed with pulmonary embolism.

Platypnea can result from several cardiopulmonary processes, but its most frequent cause is a right-to-left interatrial shunt through a patent *foramen ovale* or, more rarely, a true ASD in the absence of pulmonary hypertension. Although frequent (persistence of patent *foramen ovale* in 27% of the population),

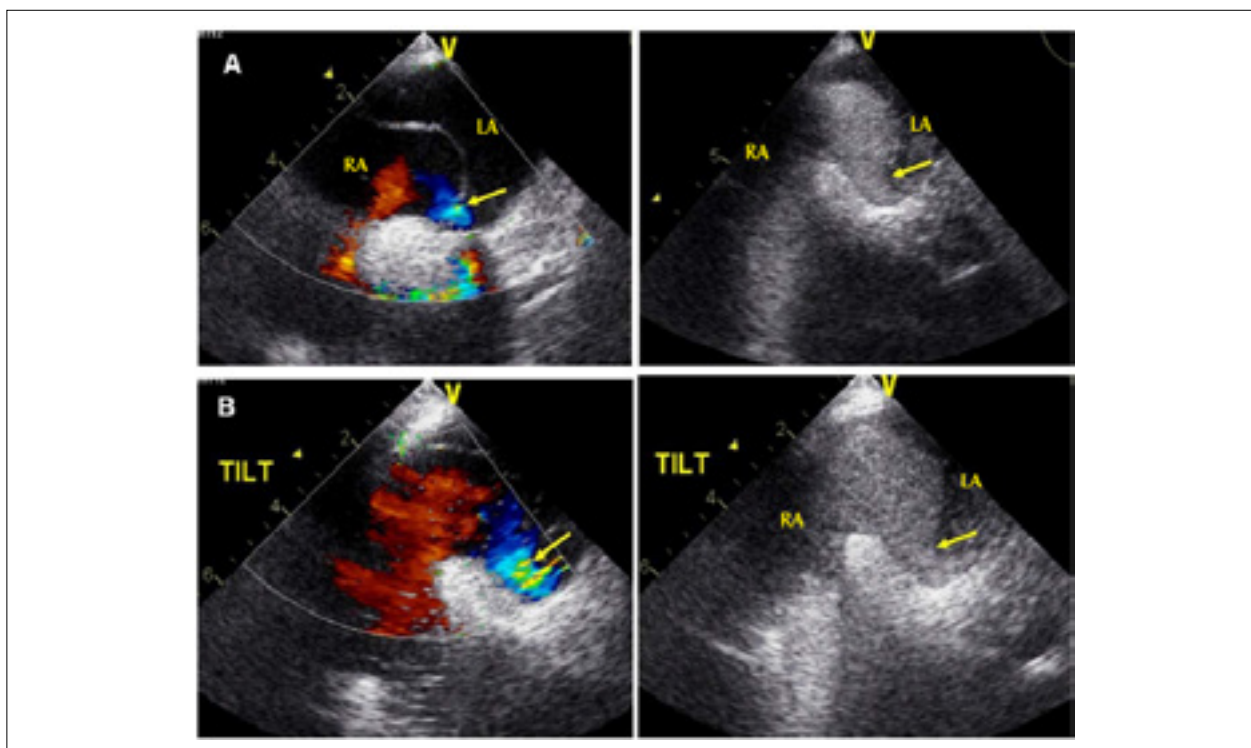


Figure 1 – (A) Transesophageal echocardiogram in the dorsal decubitus position (0°) showing an exuberant atrial septal aneurysm with right-to-left shunt (arrow) evidenced on color Doppler and contrast medium flow; (B) Transesophageal echocardiogram on the tilt table (60°) with increased right-to-left flow evidenced on color Doppler and contrast medium flow.

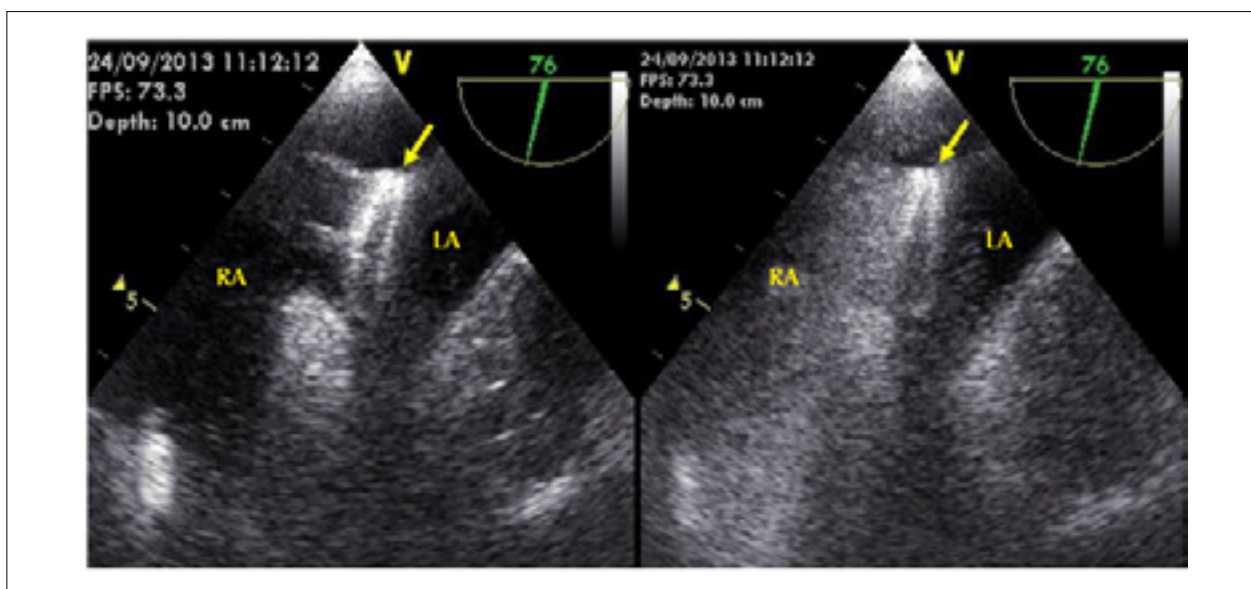


Figure 2 – Transesophageal echocardiogram after closure of the atrial septal defect, showing the occlusion device (arrow), adjacent to the atrial septal aneurysm and persistence of the contrast medium flow.

those ASDs are usually asymptomatic³. Under normal conditions, blood flows from the left side to the right side, according to the pressure gradient. Thus, a right-to-left shunt appears in the

presence of overload of the right chambers and pulmonary hypertension, and can also occur under specific circumstances, such as the Valsalva maneuver^{1,4}.

Case Report

The physiological mechanism of the positional nature of the shunt has not been fully clarified. In platypnea orthodeoxia, the pulmonary arterial pressure is typically not increased, but, in the upright position, one or more anatomical and functional factors can change the physiological flow direction^{1,5,6}.

Dilation of the ascending aorta is a frequently cited pathophysiological mechanism³. In the upright position, the effect of gravity on the dilated aortic root leads to the forward and downward displacement of that vessel, resulting in elevation of the right atrial pressure and stretching of the AS, with enlargement of the ASD orifice^{2,6}. It also contributes to AS horizontalization, directing the blood flow from the inferior vena cava preferentially to the AS orifice. Other factors, such as kyphoscoliosis and pneumonectomy, contribute to AS horizontalization^{6,7}.

Persistence of the Eustachian valve also contributes to direct blood flow from the inferior vena cava to the AS orifice, as reminiscence of its intrauterine function². The AS aneurysm disturbs the laminar blood flow on the AS, making it turbulent, and directs it to the ASD orifice⁸.

In the case here reported, the ascending aorta dilation developed over time, the voluminous AS aneurysm, and the prominent Eustachian valve might have played an important role in the pathophysiology of platypnea orthodeoxia.

On the other hand, even in the absence of pulmonary hypertension, conditions causing a localized increase in right atrial pressure and/or a reduction in the compliance of right chambers, such as pulmonary embolism, constrictive pericarditis, pericardial effusion, tricuspid stenosis, right atrial myxoma, and right ventricular dysfunction, play a role in the etiology of right-to-left shunt^{5,6,9}.

Initially, the hypothesis of pulmonary embolism was considered for our patient, but that diagnosis was not confirmed

later. However, a small pulmonary embolism, totally resolved with anticoagulant therapy, could have been the factor revealing the clinical findings of platypnea orthodeoxia.

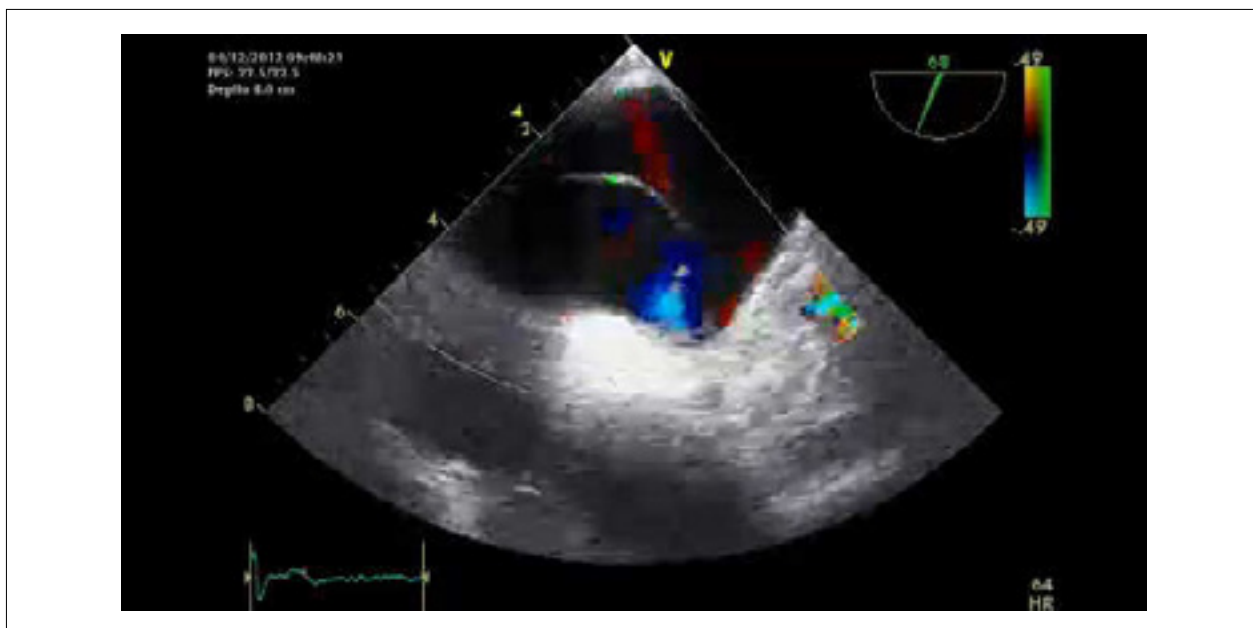
In that case, the patient's degrees of dyspnea and hypoxemia were not in accordance with the hemodynamic stability and lack of echographic signs of overload of the right chambers, and that diagnosis would be insufficient to explain the positional variations of saturation, raising the suspicion of another concomitant process. The CT-angio suggestion of peripheral pulmonary embolism of the distal pulmonary artery branches should be considered with reserve, because of the low resolution of that exam for such diagnosis, contrarily to that which occurs for major trunks.

This case evidences the need for a high index of suspicion to establish the diagnosis of platypnea orthodeoxia, and it should be considered in the differential diagnosis of unexplained or non-totally explained hypoxemia.

Platypnea orthodeoxia was first described in 1949, and 228 cases have been reported, of which, 215 have an intracardiac origin. However, it might still be underdiagnosed^{1,4}.

Its therapy implies either percutaneous or surgical ASD closure; the former is used in approximately 75% of the cases reported, although the latter is more effective, mainly for fenestrated ASD and complex defects⁴. However, the decision to intervene depends on the severity of symptoms and morbidities associated. In our case, the defect closure was not complete: the right-to-left shunt and of the PaO₂ reduction associated with the upright position persisted, although to a lesser extent and asymptomatic.

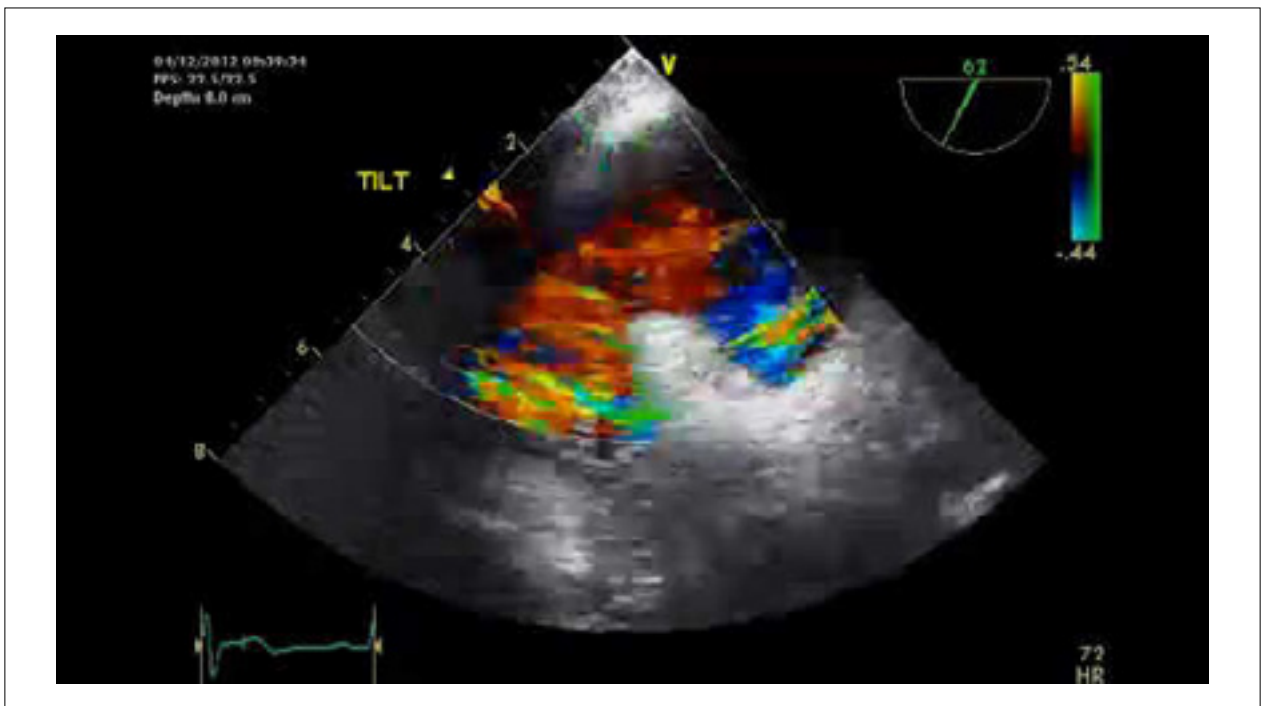
In addition, this case showed the usefulness of the TEE performed on a tilt table to establish the correlation between PaO₂ and the anatomical and functional changes induced by the upright position on the atrium.



Video 1 – Transesophageal echocardiogram in the dorsal decubitus position (0°) showing an exuberant atrial septal aneurysm with right-to-left flow evidenced on color Doppler.

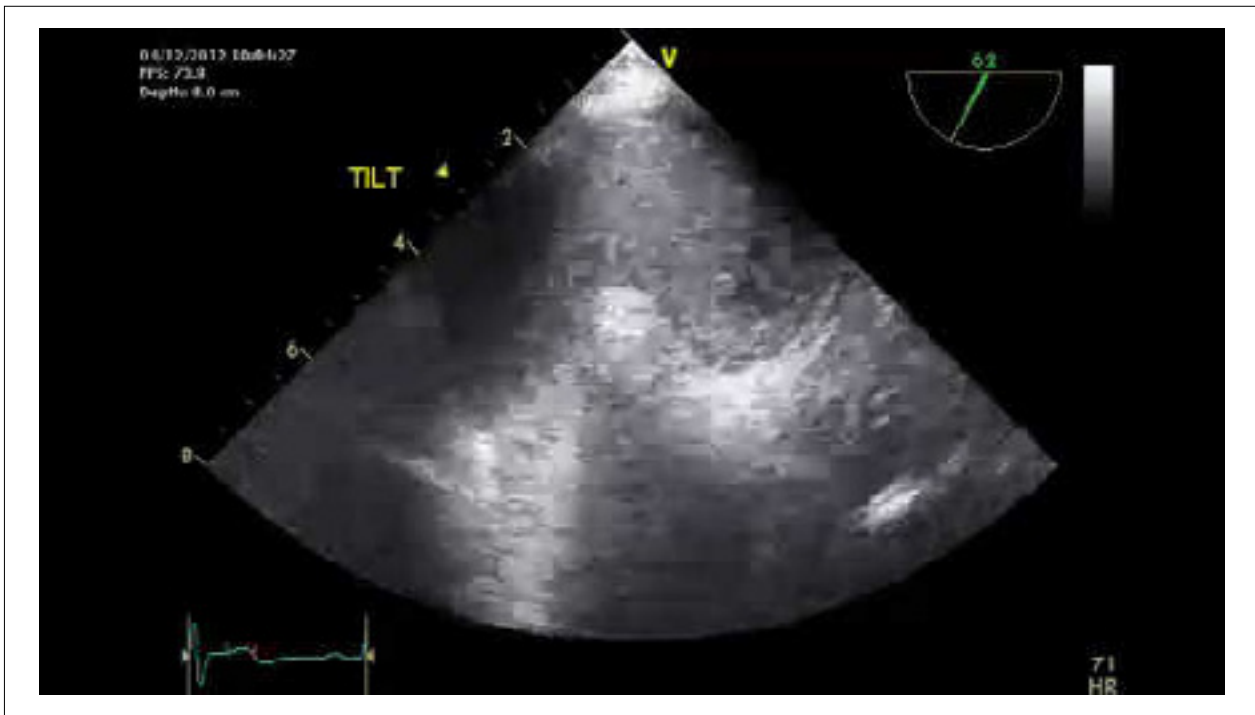


Video 2 – Transesophageal echocardiogram with agitated saline in the dorsal decubitus position (0°) showing an atrial septal aneurysm with right-to-left contrast medium flow.

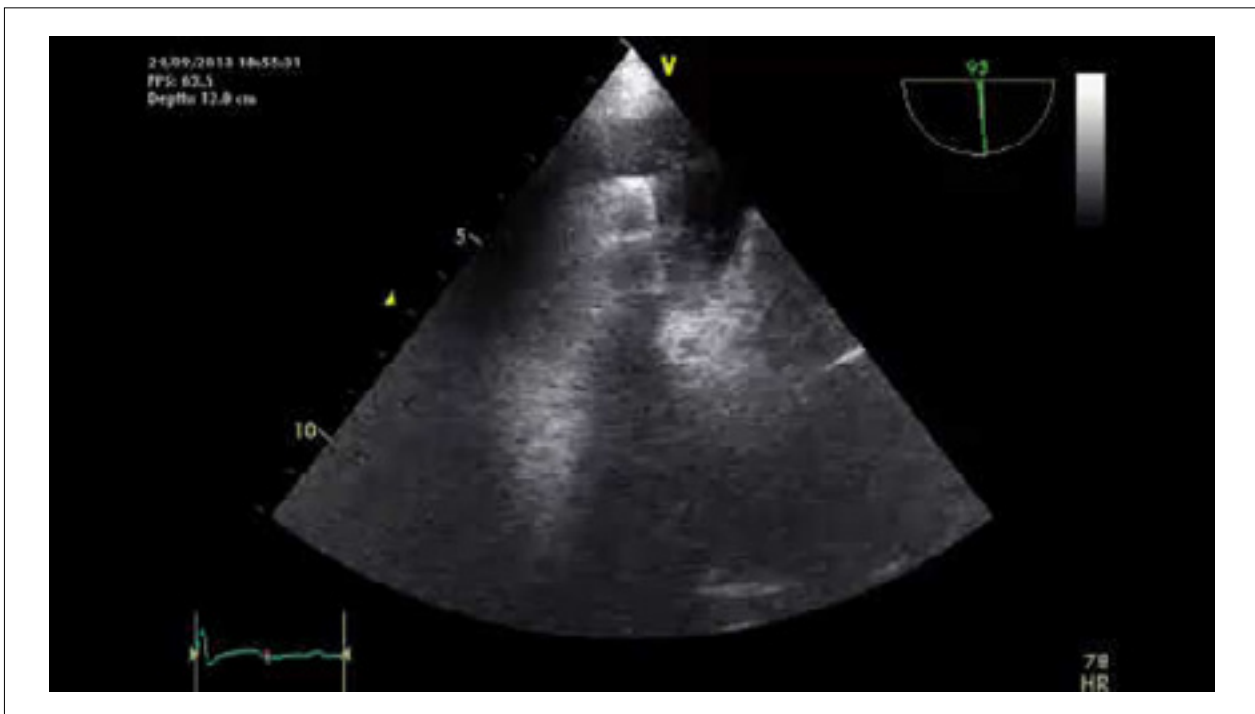


Video 3 – Transesophageal echocardiogram on the tilt table at 60°: prolapse of the atrial septal aneurysm to the left atrium was maintained, and increased right-to-left contrast medium flow as compared to that in the decubitus position.

Case Report



Video 4 – Transesophageal echocardiogram with agitated saline on the tilt table at 60°: increased right-to-left contrast medium flow as compared to that in the decubitus position.



Video 5 – Transesophageal echocardiogram with agitated saline after atrial septal defect closure: occlusion device adjacent to the atrial septal aneurysm, and persistent but reduced right-to-left contrast medium flow.

Author contributions

Conception and design of the research: Faustino M; Soares AO; Freitas A; Gil VM; Acquisition of data: Faustino M; Soares AO; Freitas A; Analysis and interpretation of the data: Faustino M; Soares AO; Rodrigues F; Anjos R; Freitas A; Gil VM; Writing of the manuscript: Faustino M; Critical revision of the manuscript for intellectual content: Faustino M; Soares AO; Freitas A; Gil VM.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any thesis or dissertation work.

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