

# Platypnea and orthodeoxia syndrome as an uncommon clinical indication for a challenging percutaneous patent foramen ovale closure: a case report

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## Background

Platypnea and Orthodeoxia Syndrome (POS) is a rare clinical condition characterized by positional dyspnoea and arterial desaturation. Various mechanisms are related to this syndrome. The simultaneous presence of abnormal anatomical findings [aortic root dilatation, atrial septal aneurysm (ASA), Lipomatous septum, and patent foramen ovale (PFO)] and an occurring ventilation/perfusion mismatch can modify intracardiac haemodynamics leading to POS in elderly patients.

## Case summary

A 70-year-old man was admitted to our emergency department suffering from neurological symptoms. A brain computed tomography scan showed a subdural haematoma and the patient underwent surgical evacuation. Some days later, he experienced an acute pulmonary insufficiency (SpO<sub>2</sub> 63%) due to parenchymal basal pneumonia treated with endotracheal intubation. Two weeks later, despite pneumonia resolution, the patient's dyspnoea became worse, experiencing deep hypoxia as soon as the patient sat up with a partial resolution on recumbent position. A transoesophageal echocardiogram with bubble-test was performed showing aortic root dilatation and a lipomatous interatrial septum characterized by the presence of tunnel-like PFO with large ASA resulting in a big right to left shunt at rest with no signs of pulmonary hypertension. The patient underwent PFO percutaneous closure intervention and a few days later O<sub>2</sub> therapy was reduced and the patient decannulated.

## Discussion

This case illustrates how the presence of both intracardiac and extracardiac factors may facilitate the onset of POS in aged patients. Platypnea and Orthodeoxia Syndrome should be considered in patients with unexplained dyspnoea and arterial desaturation related to orthostatism. It has a good prognosis with an improvement of quality of life if the causal factor can be treated.

## Keywords

Patent foramen ovale • Right-to-left shunt • Platypnea-Orthodeoxia • Case report • Echocardiography

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## Learning points

- The simultaneous presence of both intracardiac anatomical findings and acute extra cardiac factor may facilitate the Platypnea and Orthodeoxia Syndrome (POS) onset in aged patients.
- Percutaneous closure of patent foramen ovale (PFO) is the treatment of choice of POS related to an interatrial right-to-left shunt.
- Atrial septal aneurysm, lipomatous interatrial septum, long tunnel-PFO represent challenging anatomical features for percutaneous closure of PFO.

## Introduction

Platypnea and Orthodeoxia Syndrome (POS) is a rare clinical syndrome characterized by positional dyspnoea (platypnea) and arterial desaturation (Orthodeoxia) related to orthostatism.<sup>1</sup> Diagnostic criteria are fulfilled if during posture change a significant drop in oxygen saturation ( $\text{SaO}_2 > 5\%$ ) and a decrease in arterial oxygen partial pressure ( $\text{PaO}_2 > 4 \text{ mmHg}$ ) is evident.<sup>2</sup> The mechanism underlying POS is related to a right-to-left shunt.<sup>2</sup> Conditions commonly associated with this syndrome are intracardiac shunt [patent foramen ovale (PFO), atrial septal defect, atrial septal aneurysm (ASA), and congenital cardiomyopathies], extracardiac shunt (pulmonary arterio-venous malformation, hepato-pulmonary syndrome, and acute respiratory distress syndrome), and ventilation-perfusion mismatch (pneumectomy, chronic obstructive pulmonary disease, interstitial lung disease, and cryptogenic organizing fibrosis).<sup>2</sup> Patent foramen ovale is the most common structural anomaly associated with POS, which usually remains asymptomatic for decades. Moreover, an acute ventilation/perfusion mismatch may facilitate POS onset.<sup>3</sup>

In addition, intrinsic cardiac anatomical findings (ASA, elongation/tortuosity of ascending aorta, lipomatous inter-atrial septum, cardiac mass, tricuspid regurgitation/stenosis, prominent Eustachian valve) may lead to POS without overlapping pulmonary hypertension.<sup>2,4-6</sup> Lastly, high pulmonary artery pressure related to different extracardiac conditions (chronic obstructive pulmonary disease, pulmonary hypertension, pulmonary embolism, constrictive pericarditis, pericardial effusion, and pneumectomy) can lead to a large right-to-left shunt through a PFO due to the significant increase of right atrium pressure.<sup>2,4-7</sup>

## Timeline

Date	Event
February 2020	Onset of neurological symptoms
29 <sup>th</sup> February 2020	Patient admitted at the emergency department: Brain computed tomography (CT) diagnosed subdural hematoma
1 <sup>st</sup> March 2020	Surgical hematoma evacuation
12 <sup>th</sup> March 2020	Onset of pulmonary insufficiency
13 <sup>th</sup> March 2020	Lung CT diagnosed pneumonia with bases consolidation CT angiography excluded pulmonary embolism
15 <sup>th</sup> March 2020	Symptoms worsening: intubation at intensive care unit
2 <sup>nd</sup> April 2020	Blood cultures negative, lung CT negative Decubitus related hypoxia continued to be observed
3 <sup>th</sup> April 2020	Transoesophageal echocardiography (Atrial septal aneurysm, dilated aortic root, tunnel like patent foramen ovale (PFO) with a large right to left shunt at rest)
6 <sup>th</sup> April 2020	Right catheterization: absence of pulmonary hypertension, Qp/Qs 0.8 Percutaneous PFO Closure (Amplatz 25 mm) with no residual shunts
10 <sup>th</sup> April 2020	Symptoms recovery and patient discharge
17 <sup>th</sup> April 2020	Follow up transthoracic echocardiogram: Device well seated, no residual shunt, New York Heart Association Class I.

## Case presentation

A 70-year-old man was admitted to the emergency department of St. Camillo Hospital in Rome, suffering from neurological symptoms including dizziness and postural instability. A brain computed tomography (CT)-scan showed a subdural haematoma and the patient underwent surgical haematoma evacuation. Some days later, he experienced acute pulmonary insufficiency associated with fever and dry cough. Physical examination revealed hypotension (80/50 mmHg), sinus tachycardia (120 b.p.m.), and oxygen arterial desaturation ( $\text{SpO}_2$  of 63%), associated with tachypnoea and intercostal recession. The cardiac auscultation was normal. There was no lower limb pain or oedema. The pulmonary physical examination revealed an increased tactile fremitus and basal coarse crackles. A subsequent CT angiography scan performed excluded pulmonary embolism. The CT thorax showed multiple parenchymal areas at basal lobes compatible with pneumonia. Two SARS Cov-2 nasal swabs were performed both with a negative result. The worsening of patient clinical status required admission to intensive care unit and intensive care management including endotracheal intubation, inotropic drugs treatment (epinephrine), antibiotics (chloramphenicol, levofloxacin, linezolid, and cotrimoxazole), and anticoagulation therapy (enoxaparin). Oxygen arterial blood saturation levels tended to worsen in the upright position while they became better in supine position (*Table 1*). Two weeks later, despite pneumonia resolution (negative blood cultures and no radiological evidence of pneumonia), the patient's dyspnoea became worse, experiencing deep hypoxia as soon as the patient sat up with a partial resolution on recumbent position. Transoesophageal echocardiography with bubble-test showed aortic root dilatation and a lipoma-

with large ASA resulting in a big right-to-left shunt at rest (Figure 1A–D) (Videos 1–3, Supplementary material online, Videos S4 and S5).

The diagnostic hypothesis was a PFO-related POS due to anatomical factors like the ASA and the aortic root dilatation and exacerbated by the ventilation/perfusion mismatch caused by previous pneumonia. The patient was scheduled for PFO percutaneous closure intervention. The right heart catheterization excluded pulmonary hypertension and confirmed the presence of an intracardiac right-to-left shunt (Qp/Qs 0.81)

**Table 1 Blood gas analysis (BGA) parameters**

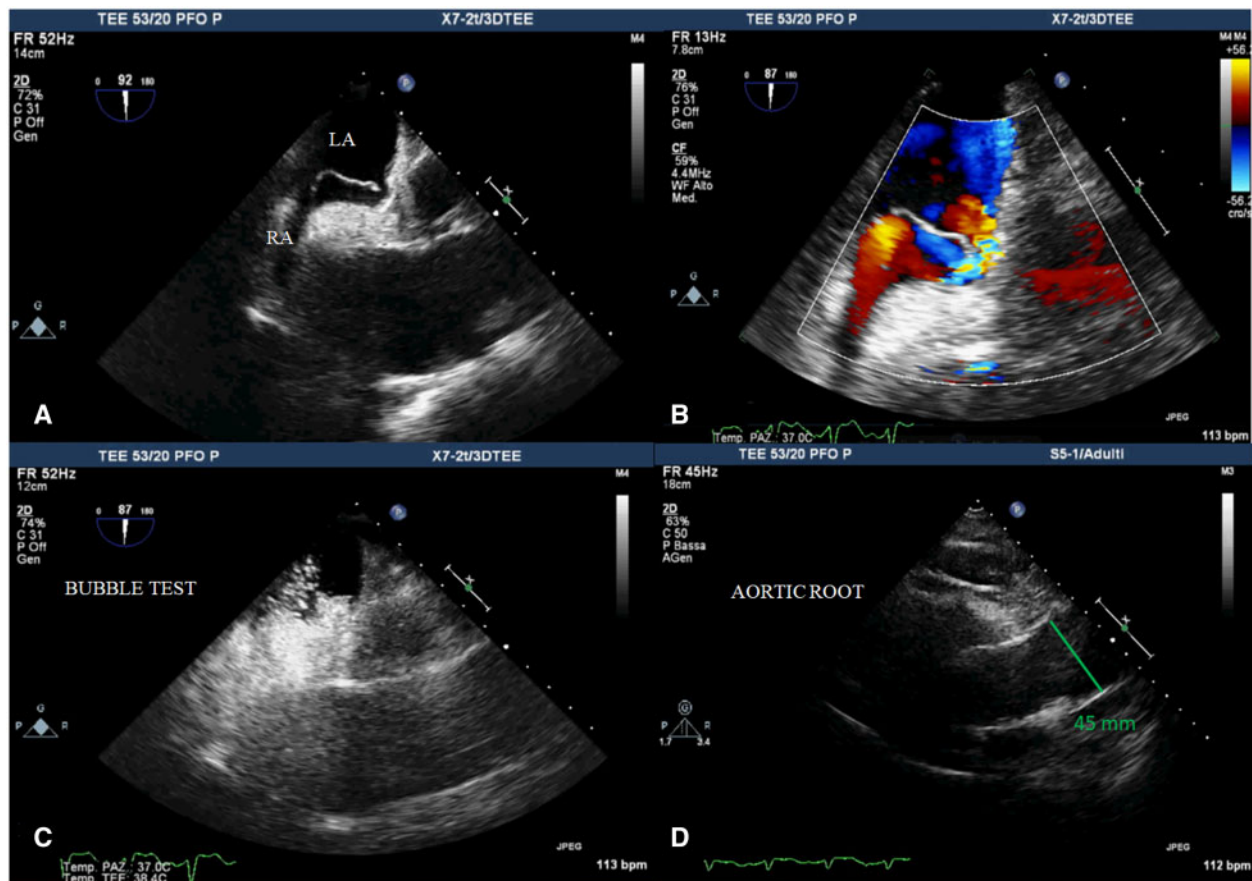
BGA parameters	Ortopnoic decubitus	Clinostatic decubitus
SpO <sub>2</sub> (%)	63	85
pO <sub>2</sub> (mmHg)	34.5	55
pCO <sub>2</sub> (mmHg)	20	20
Lac (mEq/L)	4.4	4.4

Blood gas analysis at the beginning of the acute pulmonary insufficiency. The BGA shows how the oxygen saturation levels and Oxygen blood partial pressure are influenced by the orthostatic position.

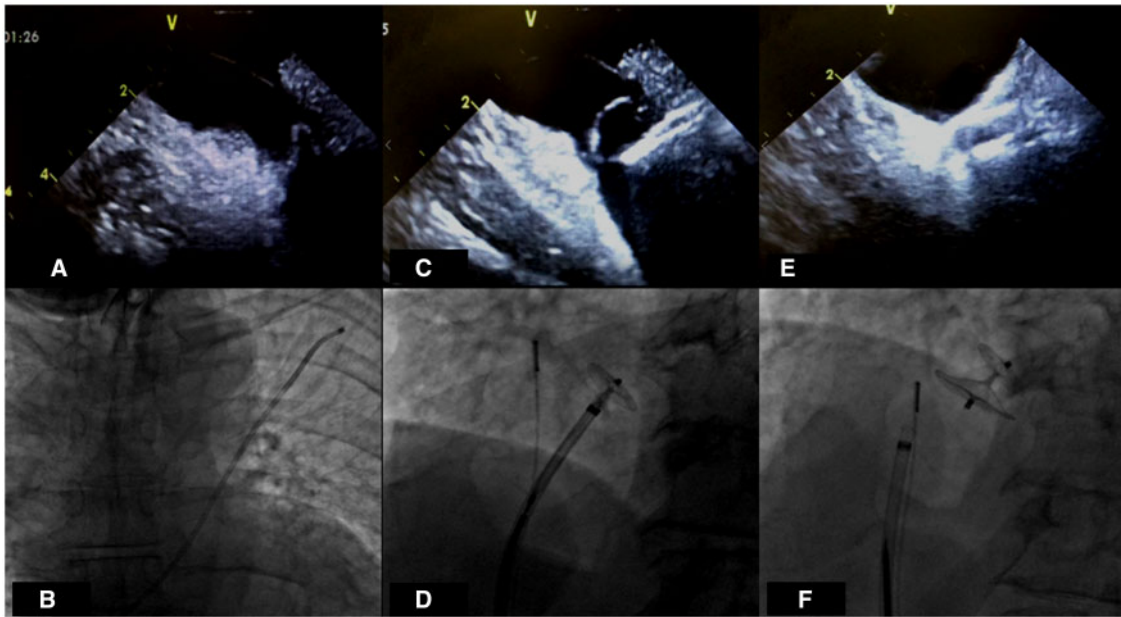
(Table 2). An Amplatzer PFO occluder device 25 mm was successfully implanted under intracardiac echocardiography (ICE) guidance without any residual shunt (Figure 2A–F) (Supplementary material online, Videos S6–S8). Device implantation was challenging due to the simultaneous presence of three complex anatomical features of the interatrial septum (ASA, long tunnel PFO, and lipomatous interatrial septum).<sup>8</sup> A few days later, oxygen therapy was reduced and the patient was discharged. The 2-week follow-up echocardiography showed a well seated PFO closure device without any residual shunt either at rest and during Valsalva manoeuvre. The patient reported a better exercise tolerance without any limitation of daily physical activity (New York Heart Association I).

## Discussion

We describe a case of a PFO-related POS without pulmonary hypertension or right heart failure that was successfully treated by catheter-based closure of the PFO. The most common POS related cause is the presence of an intracardiac shunt (PFO).<sup>2,3</sup> Although PFO can be undetectable and asymptomatic for decades, an occurrence of ventilation/perfusion mismatch may reveal POS in elderly patients.<sup>2,9</sup>



**Figure 1** Transoesophageal echocardiogram. (A) Lipomatous interatrial septum, atrial septal aneurysm, and tunnel-like patent foramen ovale. (B) Right-to-left shunt at colour Doppler. (C) Saline contrast study with bubbles in left side. (D) Aortic root dilatation (green line, 45 mm).

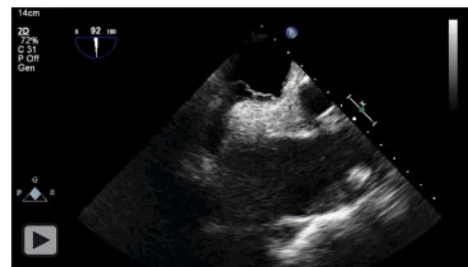


**Figure 2** Intracardiac echocardiography and fluoroscopic guidance. (A). Intracardiac echocardiography view of Amplatz Super Stiff Guidewire crossing the aneurismatic fossa ovalis. (B) Fluoroscopic view of a multipurpose in left upper pulmonary vein crossing the fossa ovalis. (C,D). Intracardiac echocardiography and fluoroscopic view of left disc positioning on the left side. (E,F) Intracardiac echocardiography and fluoroscopic view of the device (Amplatzer patent foramen ovale occluder) released.

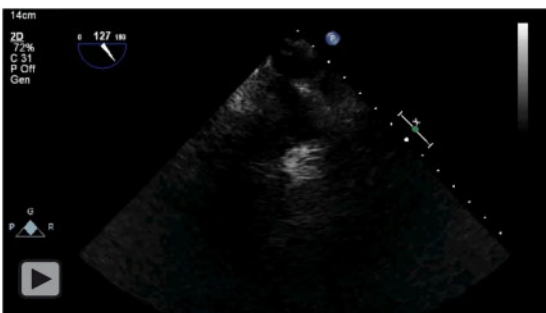
**Table 2** Right heart catheterization

Right heart catheterization (Qp/Qs 0.81)	SpO <sub>2</sub> (%)
Caval vein	50
Pulmonary artery	52.2
Pulmonary vein	98
Aorta	91

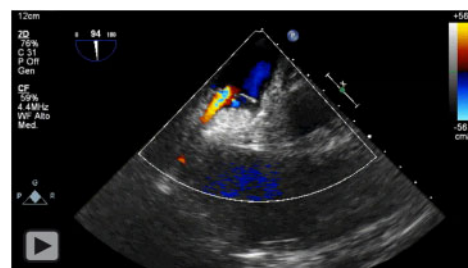
SpO<sub>2</sub> levels detected by the right heart catheterization confirm the presence of a right-to-left large intracardiac shunt.



**Video 2** TEE view of long tunnel.



**Video 1** TEE view of atrial septum aneurism.



**Video 3** Right to left shunt at colour-Doppler.

In literature, several case reports showed parenchymal lung diseases such as emphysema, interstitial lung disease, or basal lung consolidation occasionally facilitate POS onset due to severe ventilation/perfusion (V/Q) mismatch from bases to apices. When a lung base disease occurs, an upright positioning worsens the ventilation/perfusion mismatch at bases and create a physiologic shunt without gas exchange.<sup>2</sup>

In addition, it has been reported how aortic root dilatation is a degenerative and age-dependent process; this is why symptoms of POS can appear in late adult life in the presence of PFO with a huge aortic root dilatation.<sup>4,7</sup> Furthermore, a lipomatous septum can favourite the right-to-left shunt due to atrial compliance changes, especially when associated with other anatomical findings like a well-represented Chiari Network.<sup>10</sup>

## Conclusion

We describe a case of PFO-induced platypnea-orthodeoxia syndrome with a large right-to-left shunt at rest associated with the dilated aortic root, atrial septum hypertrophy, large ASA, and exacerbated by pneumonia with basal lung consolidation. No evidence of pulmonary hypertension, pulmonary embolism, heart failure, elevated filling pressures, or hepatic failure was reported. It was successfully treated with percutaneous PFO-closure under intracardiac echocardiography guidance.

Although POS is rare, we consider pivotal taking into consideration this diagnostic option in patients with unexplained dyspnoea and arterial oxygen desaturation in orthostatism. Platypnea and Orthodeoxia Syndrome has a good prognosis if PFO is the main causal factor and percutaneous closure is the treatment of choice.<sup>11</sup>

## Lead author biography



Francesco Dipasquale graduated in 2015 at the Faculty of Medicine of Catania, Italy. Recently he got the postgraduate diploma in Cardiology at the University of Catania. In the

last year, he did an Interventional Cardiology fellowship at the St. Camillo Hospital, Rome, Italy.

## Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

**Slide sets:** A fully edited slide set detailing these cases and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient.

**Conflict of interest:** None declared.

**Funding:** None declared.

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