A case report of platypnea–orthodeoxia syndrome: an interplay of pressure and blood flow

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Background Platypnea–orthodeoxia syndrome is a rare condition characterized by onset or worsening of dyspnoea with orthostatism (platypnea) and arterial desaturation (orthodeoxy) that is relieved by returning to a recumbent position. An acute event causing a change in the pressure relationships inside the cardiac chambers can lead to the diagnosis of a previously undiagnosed cardiac anomaly, as the following case illustrates.

Case summary A previously asymptomatic 80-year-old female patient was admitted in our hospital with a sudden onset ischaemic stroke. Initial evaluation, including 12-lead electrocardiogram and transthoracic echocardiogram, was unremarkable. During hospital stay, she develops pulmonary embolism, after which she complains of positional dyspnoea that develops upon sitting up, accompanied with refractory hypoxaemia that reverts on recumbent position. Transoesophageal echocardiogram revealed an interatrial septum with an exuberant hyperdynamic movement, and an abundant passage of contrast from the right atrium to the left, even without performing the Valsalva manoeuvre, compatible with an important patent foramen ovale. A percutaneous closure was performed, and patient has been symptom-free since then.

Discussion This is a case illustrates how an anomaly that has been present for 80 years can suddenly manifest itself with an array of different symptoms that can make the diagnosis challenging. A high degree of clinical suspicion is crucial for an accurate diagnosis and definitive treatment.

Keywords Platypnea • Orthodeoxy • Patent foramen ovale • Case report

Learning points
• Platypnea–orthodeoxia is a rare presentation of several pathologies, for which a high degree of clinical suspicion is required.
• Even though the underlying pathology can be present since birth, symptoms may appear only later in life due to a superimposed acute illness.
• Mechanism is due to a right-to-left shunt that is exacerbated when there is a shift in the anatomical relations due to changes in posture.

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Introduction

The platypnea-orthodeoxia syndrome (POS) is a rare condition which is becoming increasingly recognized, characterized by onset or worsening of dyspnoea with orthostatism (platypnea) and arterial desaturation (orthodeoxia) that is relieved by returning to a recumbent position. The pathophysiologic mechanism responsible for this syndrome is not entirely understood, but probably derives from one of the following mechanisms: intra-cardiac shunt, pulmonary vascular shunt, ventilation/perfusion mismatch, or a combination of the three.1

The most common structural anomaly associated with this syndrome is the patent foramen ovale (PFO), which usually remains asymptomatic for decades. In rare occasions, a shift in the right/left intra-cardiac pressure relationship [such as can happen in pulmonary embolism (PE)], or a change in the anatomy of adjoining structures causing a change in the direction of the venous blood inflow into the PFO (as can happen in Thoracic Kyphosis) can lead to POS.2

This following case shows illustrates how an anomaly that has been present for 80 years can suddenly manifest itself with an array of different symptoms that can make a diagnosis challenging.

Timeline

| 80 years | Female patient, previously asymptomatic |
| 19th day of hospitalization | Patient started complaining of severe positional dyspnoea with orthostatism, with associated hypoxaemia that was refractory to O2 and reverted on recumbency |
| 20th day | Ventilation/perfusion scan of the lungs showed several small peripheral emboli |
| 21st day | Transoesophageal echocardiography (TOE) showed an interatrial septum with an exuberant hyperdynamic movement, with abundant passage of contrast from the right atrium to the left |
| 25th day | Patient undergoes percutaneous closure of the foramen ovale with relief of symptoms, which allows her hospital discharge the next day |
| 6-month follow-up | Patient remains symptom-free and with normal peripheral saturation |

We present the case of an 80-year-old woman, previously healthy, that presented in our emergency department with sudden onset dysarthria and right-side hemiparesis that had over 24 h of evolution. She was admitted with the diagnosis of ischaemic stroke in the territory of the left posterior cerebral artery, as confirmed by head computed tomography (CT) scan. Her 12-lead electrocardiogram on admission was in sinus rhythm, and the transthoracic echocardiography (TTE) showed no significant structural changes besides septal hypertrophy of 17 mm. On the 19th day of hospital stay, she started complaining of positional dyspnoea that appeared whenever she sat up and reverted when on recumbent position, associated with arterial oxygen desaturation refractory to oxygen supplementation (Table 1).

She then performed an Angio-CT scan of her lungs that showed no signs of PE and excluded lung disease. The ventilation/perfusion scan showed several small peripheral emboli in the pulmonary vascular bed. In another TTE, no significant changes were identified. According to the previous findings, suspecting POS, a transoesophageal echocardiography (TOE) was performed, which showed an interatrial septum with an exuberant hyperdynamic movement, with abundant passage of contrast from the right atrium to the left, even without performing the Valsalva manoeuvre, compatible with an important PFO (Figures 1 and 2). When Valsalva manoeuvre was performed, the patient became breathless and peripheral oxygen saturation dipped to 85%, which recovered after termination of the manoeuvre.

The patient was then referred to a tertiary centre for percutaneous closure of the PFO, in order to both treat the right-left shunt and improve symptoms and also to minimize the risk of paradoxical embolism, which was assumed to be the cause of her stroke. The device used was an Amplatzer™ PFO Occluder, size 18 mm. The day after the procedure, patient was able to stand-up with no symptoms and with 95% peripheral oxygen saturation. Upon discharge, the patient was medicated with apixaban 5 mg, atorvastatin 40 mg, and ramipril 5 mg. At 1-month follow-up after the procedure, patient remained symptom-free, with a peripheral saturation of 94% while sitting and with no oxygen supplementation. She performed a follow-up TOE, confirming the presence of the closing device in the right position, without any passage of contrast from the right to the left atrium (Figure 3). Patient still had some neurologic deficits (right side hemiparesis) but was autonomous and was still in physical rehabilitation.

### Table 1: Arterial blood gas in different postural positions

| Position   | pH  | pCO₂ (mmHg) | pO₂ (mmHg) | sO₂ (%) |
|------------|-----|-------------|------------|---------|
| Supine     | 7.44| 30.8        | 76.7       | 96.9    |
| Head at 45°| 7.41| 32.8        | 40.2       | 77      |
| Sitting    | 7.43| 32.9        | 52.5       | 88.5    |
Discussion

This case is an example of the diversity of the clinical manifestations of PFO. Platypnea–orthodeoxia syndrome is a rare syndrome caused by a limited number of clinical conditions that produce pulmonary arteriovenous shunt (arteriovenous malformation and hepatopulmonary syndrome) or right-to-left intra-cardiac shunt (PFO and/or related atrial septal defects).1

A high degree of suspicion is required to identify this condition. However, simple tests, such as arterial blood gas analysis collected in different postural positions can confirm the diagnosis of POS.3

The most frequent cause is a right-left intra-cardiac shunt, most frequently due to PFO, which affects up to 30% of the general population. PFO is usually asymptomatic because the left atrial pressure is higher than in the right atrium, and tends to keep it closed, through a valve mechanism of the septum secundum. In situations of increased pressure in the right chambers, such as in acute PE, or a directed venous flow toward the PFO, a right to left shunt may occur.2

In POS, a positional shift in the intra-cardiac anatomic relations leads to increased septal stretching, favouring the direction of flow from the inferior vena cava directly through the septal defect into the left atrium, increasing blood flow through the shunt, causing the development of dyspnoea and hypoxaemia when the patient sits up.4,5 Diagnosis of PFO is usually confirmed by an echocardiogram, preferably transoesophageal, complemented by the intravenous injection of agitated saline solution.1

In our case, the patient first presented with an acute ischaemic stroke. There is a higher prevalence of PFO in patients with cryptogenic strokes than in the general population.6 If the cause of the stroke was paroxysmal embolization, we must consider that there was previously a degree of right-left shunt that allowed for the embolus to cross to the main circulation. However, pulmonary emboli occurring during hospital stay that led to increased right ventricular and atrial pressure was probably responsible for a more significant increase in flow through the shunt, causing the patients’ symptoms.

Percutaneous closure of the PFO has been shown to be a safe and effective procedure, with immediate relief of symptoms, and low complication rates.7,8 There is lack of long-term data in clinical trials, but relieving POS symptoms in our patient was crucial for her to resume physical rehabilitation and make a full recovery from the ischaemic stroke that motivated her admission.

Lead author biography

João de Sousa Bispo received Masters’ Degree in Medicine in 2014 from the Faculty of Medicine of the University of Lisbon. Currently, he is a Cardiology Resident in Hospital of Faro, Portugal.
Supplementary material

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

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Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author’s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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