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# Platypnea orthodeoxia syndrome after recent stroke: a case report of a sandwiched right atrium

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## Background

Platypnea orthodeoxia syndrome (POS) is a condition characterized by onset or worsening of dyspnoea and desaturation in upright position that is relieved by returning to a supine position. This case report illustrates a sudden onset of severe platypnea caused by compression of the right atrium (RA) due to aortic dilatation and unilateral diaphragmatic paralysis after a recent stroke.

## Case summary

A 71-year-old male patient with a medical history of recent stroke of the left hemisphere was referred to emergency department with acute dyspnoea. During observation in the emergency department, desaturation was noted in upright position. A contrast computed tomography excluded pulmonary embolism but revealed a dilated aortic root and an elevated right hemidiaphragm. The RA was compressed between these two structures (sandwiched). Given the clinical suspicion of a POS, a transoesophageal echocardiography was performed which confirmed the presence of a persistent foramen ovale (PFO) in supine position. In upright position, there was a torrential increase in right-to-left shunting. The PFO was closed using an Occlutech™ device. Directly after the procedure, the patient was symptom free.

## Discussion

A rise in RA pressure or difference in flow pattern in the RA can make a PFO become symptomatic. Elevated RA pressure was ruled out. Most anatomical pathologies influencing the flow pattern develop slowly over time. This case shows a presentation of POS after a recent stroke possible due to change in anatomy because of right hemidiaphragm paralysis in combination with the aortic dilatation.

## Keywords

Case report • Platypnea orthodeoxia syndrome • Persistent foramen ovale • Computed tomography • Transoesophageal echocardiography

## ESC curriculum

2.1 Imaging modalities • 2.4 Cardiac computed tomography • 2.2 Echocardiography • 9.7 Adult congenital heart disease

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## Learning points

- Platypnea orthodeoxia syndrome is caused by a combination of a substrate for right-to-left shunting and anatomical or haemodynamic change to the right atrium.
- Platypnea orthodeoxia syndrome can have an acute presentation due to sudden changes in anatomy (e.g. right diaphragm paralysis) or haemodynamics (e.g. pulmonary embolism, right ventricle infarction).
- Transoesophageal echocardiography in supine and upright position can confirm diagnosis of platypnea orthodeoxia syndrome.

## Introduction

Platypnea orthodeoxia syndrome (POS) is a rare condition characterized by onset or worsening of dyspnoea and desaturation in upright position that is relieved by returning to a supine position.<sup>1</sup> The most frequent intracardiac cause is a persistent foramen ovale (PFO), present in up to 25% of adult individuals.<sup>2</sup> Other shunts leading to POS has been reported from atrial septal defect, partial anomalous pulmonary venous connection, unroofed coronary sinus, and cases of complex congenital heart disease.<sup>1</sup> This case report illustrates a sudden onset of severe platypnea caused by compression of the right atrium (RA) due to aortic dilatation and unilateral diaphragmatic paralysis after a recent stroke.

## Timeline

| Time      | Event   |
|-----------|---|
| Day 0     | A 71-year-old man was admitted to the emergency department with acute dyspnoea. He had a history of hypertension and a stroke of the left hemisphere 8 days before presentation. Pulmonary embolism was ruled out. Clinical suspicion of POS. |
| Day 1     | Transoesophageal echocardiography with a bubble test showed a PFO with increased shunting in upright position.  |
| Days 2–10 | Workup with coronary angiography, right-heart catheterization, spirometry.  |
| Day 21    | Percutaneous closure of the PFO with occluder device.   |
| Day 26    | Follow-up transthoracic echocardiography with a bubble test showed no signs of cardiac shunt, symptoms fully resolved.  |

## Case presentation

A 71-year-old male patient was referred to emergency department with acute dyspnoea. His medical history included hypertension and a recent stroke (left hemisphere) treated with intravenous thrombolysis. His symptoms started acutely the same day while bending over to tie his shoelaces. Upon presentation, he was haemodynamically stable with a blood pressure of 130/80 mmHg and a peripheral oxygenation of 95% in supine position using. Physical examination revealed no signs of heart failure. The electrocardiogram showed sinus rhythm with no signs of ischaemia. Laboratory studies showed no

abnormalities, including low NT-pro-brain natriuretic peptide and Hs troponin T levels. During observation in the emergency department, a clear difference in peripheral oxygenation in upright and supine positions was noted—dropping from 95 to 88% when sitting upright. With arterial blood gas analysis, a decrease in PO<sub>2</sub> from 8.9 kPa in supine position to 6.5 kPa in upright position was measured. A contrast computed tomography (CT) excluded pulmonary embolism and pulmonary arteriovenous shunts, but revealed a dilated aortic root (50 mm, *Figure 1A*) and an elevated right hemidiaphragm (*Figure 1B*). The RA was compressed between these two structures. In addition, contrast could be seen crossing over the atrial septum from right to left, suggestive for a PFO. Given the clinical suspicion of a POS, a transoesophageal echocardiography (TOE) was performed which confirmed the presence of a PFO in supine position (see [Supplementary material online, Video S1, Figure 1C](#)). In upright position, there was a torrential increase in right-to-left shunting (see [Supplementary material online, Video S2, Figure 1D](#)). Right-heart catheterization revealed normal right atrial pressure and no pulmonary hypertension. Using estimated Fick method, a shunt fraction of 21% was measured even in supine position. Persistent foramen ovale closure could be considered in cases of severe hypoxia after pulmonary diseases has been excluded.<sup>3</sup> The case was referred to our heart team in which the patient was accepted for percutaneous closure of the PFO. The following week, the PFO was closed using an Occlutech™ PFO Occluder 23–25 mm (see [Supplementary material online, Videos S3 and S4](#)). Directly after the procedure, the patient had a peripheral oxygenation of 95% in upright position and was symptom free. During follow-up 4 weeks later, echocardiography with bubble test revealed no signs of residual shunting and patient was free of symptoms.

## Discussion

This case report describes an elderly patient presenting with acute dyspnoea caused by POS resulting from a PFO. Under normal conditions, there is a functional closure of the PFO because left atrial pressure is higher than right atrial pressure. A rise in RA pressure or difference in flow pattern in the RA can make a PFO become symptomatic. Elevated RA pressure can be caused by pulmonary embolism, right ventricular myocardial infarction, pleural or pericardial effusion, and pulmonary hypertension from any cause.<sup>4</sup> Flow patterns may be influenced by a number of intrinsic cardiac anatomical pathologies; lipomatous hypertrophy of the inter-atrial septum, a prominent eustachian valve, tricuspid regurgitation, tricuspid stenosis, cardiac mass, persistent left superior vena cava, and aortic dilatation.<sup>1,5</sup> Changing posture from supine to upright position increases the shunt (*Figure 1C and D, Supplementary material online, Videos S1*



**Figure 1** Computed tomography aorta (A, B) with contrast showing compression of the right atrium due to aortic dilatation and an elevated right hemidiaphragm. Transoesophageal echocardiography with bubble study in supine (C) and upright position (D) with torrential increase in right-to-left shunting. AO, aortic.

and S2) probably by influencing flow patterns and inter-atrial communication. In our patient, prominent aortic dilatation was an anatomical abnormality influencing the flow pattern. Aortic dilatation develops slowly over time<sup>6</sup> and gradually development of symptoms is expected. In this case, presentation was atypical as the onset of complaints was more acute. An elevated RA pressure was ruled out by invasive measurement. Therefore, a difference in flow patterns in the RA could be the only cause. A possible explanation in this case is that right hemidiaphragm paralysis was a consequence of the stroke of the left hemisphere, which has been described in literature.<sup>7</sup>

As seen in *Figure 1A and B*, the RA is compressed (sandwiched) by the dilated aorta and the right hemidiaphragm. These two anatomical pathologies cause differences in flow patterns and explain the acute presentation days after the stroke. This case shows that POS can cause acute dyspnoea due to an anatomical change. When there is clinical suspicion of POS early imaging with CT thorax and TOE in supine and upright position should be considered.

## Lead author biography



Marijn Wiertsema was born in Groningen in 1989. After his medical training at the University of Groningen, he is currently following his residency in cardiology at the University Medical Center Groningen.

## Supplementary material

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

**Slide sets:** A fully edited slide set detailing this case 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 in line with COPE guidance.

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