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Platypnea-Orthodeoxia Syndrome in an Elderly Patient Treated with Percutaneous Patent Foramen Ovale Closure: Report of a Case and Review of the Literature

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Abstract

Introduction: Platypnea-Orthodeoxia syndrome is characterized by dyspnea and hypoxia in the upright position, usually improving in the supine position. Two components are required: an interatrial or intrapulmonary shunt, and a functional component. Diagnosis is made by contrast ultrasonography. We report a case of Platypnea-Orthodeoxia syndrome revealed by positional dyspnea in an 87-year-old patient. The aim of this study is to describe the clinical, therapeutic and evolutionary profile of this syndrome. **Case Presentation:** This is an 87-year-old patient with a history of pulmonary embolism (PE) and stroke. He was seen for dyspnea and desaturation in orthostatism, revealing a patent foramen ovale (PFO). Progression was favorable after closure of the PFO. **Conclusion:** Platypnea-Orthodeoxia syndrome may be presented as simple exertional dyspnea. The clinician should check for improvement in symptoms and/or oxygenation during decubitus. Definitive treatment consists of percutaneous closure of the shunt.

Keywords

Syndrome, Platypnea-Orthodeoxia, Patent Foramen Ovale

1. Introduction

Platypnea-Orthodeoxia is an uncommon syndrome characterised by dyspnoea

relieved by decubitus (hence the term platypnoea) and deoxygenation following the transition from a supine to an upright position (hence the term orthodeoxia) [1].

Right-to-left shunting via an intracardiac communication such as a PFO or atrial septal defect (ASD) is the most common cause. Although previous reports have hypothesised why right-to-left shunting is exacerbated in the upright position, the underlying mechanisms are not fully understood [2].

However, this diagnosis can easily be missed. The fact that this syndrome is often accompanied by multiple other medical conditions makes it difficult to identify. Traditionally, the medical conditions associated with this syndrome are lung, liver and heart disease [3] [4] [5]. The prevalence of PFO is common in the general population, with an estimated incidence of 25% - 30%. Platypnea-Orthodeoxia syndrome is, however, likely to be under-diagnosed [6].

Transoesophageal echocardiography is the preferred diagnostic modality, offering good visualization of any defects or aneurysms that may be present in the atrial septum. Definitive treatment of SOP secondary to intracardiac shunting involves closure of the interatrial defect [7].

Progress is favorable after shunt closure, as shown in the study by Ashish H. Shah *et al.*, which demonstrated improvements in post-procedure oxygen saturation to 95.1% ± 0.5%, compared with 81% ± 8% pre-procedure [8].

In this article, we report a Platypnea-Orthodeoxia case in a patient with a history of stroke and PE.

2. Observation

This was an 87-year-old man with type 2 diabetes, hypertension, dyslipidemia, a history of diabetic neuropathy and recent hospitalization for ischemic stroke concomitant with segmental pulmonary embolism. This patient was admitted for desaturation and positional dyspnoea. On admission, he was haemodynamically stable with a heart rate of 96 bpm, blood pressure of 115/74 mmHg, saturation of 95% on 4 liter of oxygen in the sitting position and 100% on room air in the dorsal position, and temperature of 36˚. There were no signs of right or left heart failure and the heart sounds were regular with no murmur.

His ECG showed a regular sinus rhythm at 92 bpm with left anterior hemi block and right bundle branch block (**Figure 1**).

Biologically, haemoglobin was 10.7 g/dl, creatinine 80 µmol/l, BNP 851 and CRP 10. Transthoracic echocardiography (TTE) showed a non-dilated, non-hypertrophied left ventricle with septal dyskinesia. The left ventricular ejection fraction (LVEF) was preserved, and the initial aorta was moderately dilated, measured at 40 mm at the sinus, 38 mm at the sino-tubular junction and 45 mm at the tubular aorta. There was moderate aortic insufficiency, no mitral valve disease, no inter-atrial septal aneurysm, the right ventricle was undilated and normokinetic, there was no pulmonary hypertension, the inferior vena cava was undilated and compliant, and the pericardium was dry. The contrast test is posi-

tive in the basal state and in the Valsalva maneuver (Figure 2).

Transoesophageal echocardiography (TEE) revealed thin valves, a free left atrium and a large PFO (4 mm). On contrast, microbubbles appeared rapidly in the left atrium in the first three cardiac cycles following injection of intravenous agitated saline. The aortic valve was tricuspid and the ascending aorta moderately dilated: sinus 42 mm, junction 33 mm and tubular aorta 40 mm. Finally,

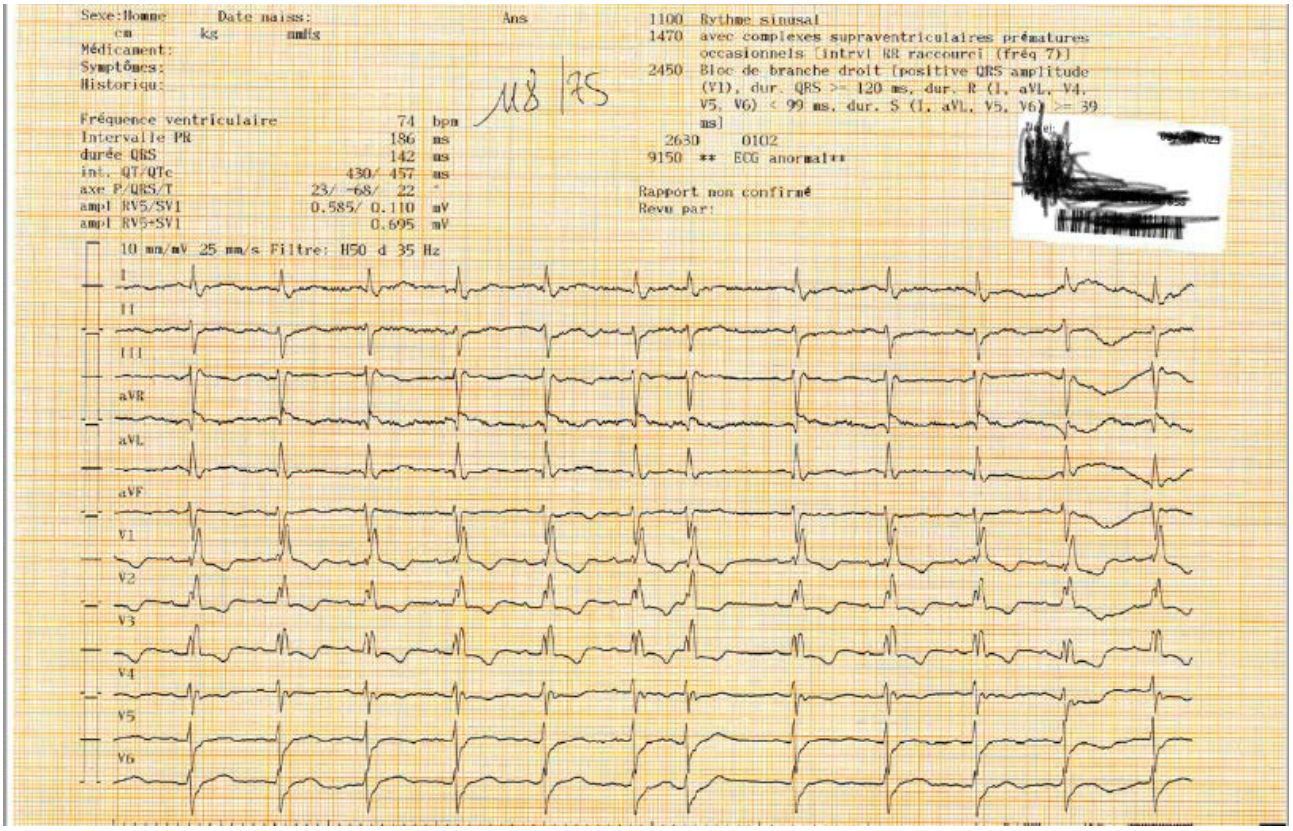


Figure 1. ECG in sinus rhythm, left anterior hemiblock, branch block droits, extrasystole atriale.

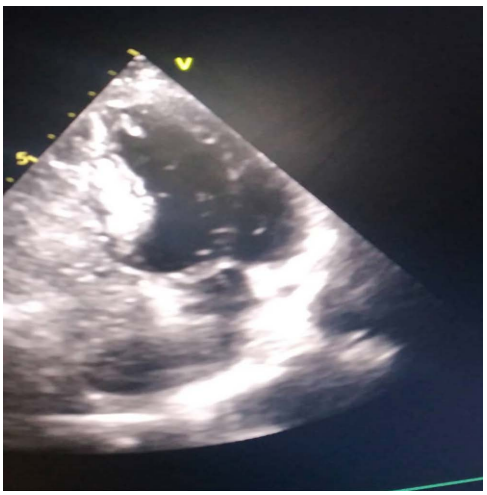


Figure 2. Four-cavity apical TTE shows opacification of the right cavities by bubbles and passage into the left atrium.

the descending thoracic aorta is moderately atheromatous around its entire circumference (Figure 3).

On the therapeutic front, the patient underwent percutaneous transcatheter closure of the PFO using a Figulla Flex Occlutech 30 closure device (Occlutech N˚ 2220193017_30).

After the procedure, we noted good positioning of the prosthesis, persistence of a moderate residual shunt and dry pericardium.

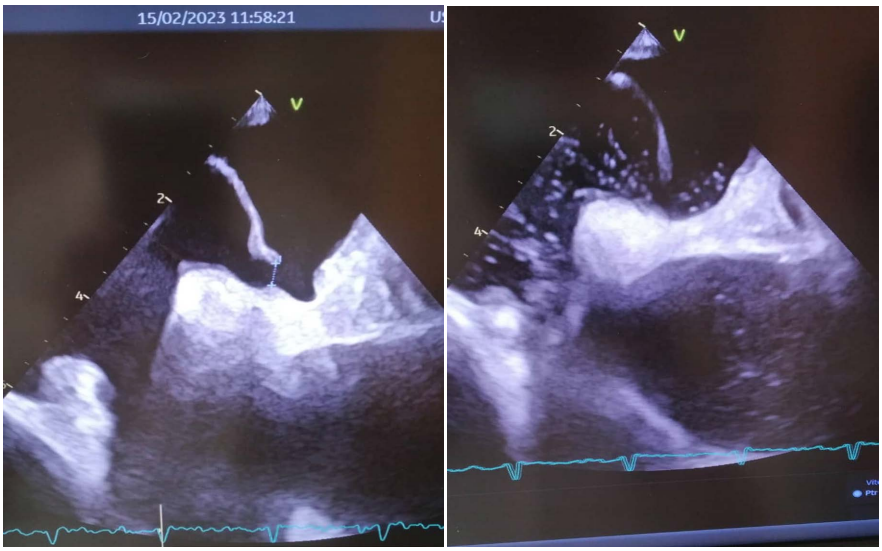
Dyspnea resolved, and oxygen saturation improved to over 94% on room air. The patient was therefore weaned off oxygen.

3. Discussion

Platypnea-Orthodeoxia syndrome is characterized by profound hypoxemia in the standing position, absent in the supine position. It is due to a right-to-left blood shunt across a PFO, often promoted by changes in thoracic anatomy and an aneurysm of the interatrial septum [9].

The causes of Platypnea-Orthodeoxia syndrome fall into three categories: intracardiac shunts, pulmonary arteriovenous shunts, and ventilation-perfusion mismatch. The most frequent cause is interatrial shunting via an AIC or PFO. When right atrial pressure is higher than left atrial pressure under conditions such as pericardial disease, PE or right heart infarction, a right-to-left shunt across the defect occurs [10] [11]. However, when there are anatomical and functional components that direct venous blood flow to the AIC or PFO, as in patients with a prominent Eustachian valve or aortic elongation or dilatation, a right-to-left shunt can occur even in the absence of elevated right atrial pressure [12].

In the study by Hayek A *et al.* [12], 16.7% of patients with Platypnea-Orthodeoxia syndrome had right heart disease (PE or tricuspid regurgitation). The same was



(a) (b)

Figure 3. ETO. (a) Visualisation of the patent foramen ovale with a diameter of 4mm; (b) Right-left bubble passage on contrast test.

true of our patient with PE.

Cases of iatrogenic Platypnea-Orthodeoxia syndrome have been reported in the literature, notably post ablation of atrial fibrillation [13].

Clinically, dyspnea and desaturation are the most common symptoms. Assessment consists of measuring oxygen saturation levels in the supine and sitting positions [14].

In our case, a patient with a potentially asymptomatic PFO suddenly presented with dyspnea. A case of positional cyanosis revealing Platypnea-Orthodeoxia syndrome has been reported by Kenny *et al.* [15].

Diagnosis requires contrast-enhanced TTE with the patient supine and upright [16]. In some cases, TTE can also be used to identify factors favouring the opening of the PFO. In the study by Singh *et al.* [5], primary pulmonary hypertension was the factor favouring PFO opening. In our patient, no right-heart abnormalities were found.

TEE is more sensitive and more likely to detect an atrial septal defect or patent foramen ovale [1]. Both TTE and TEE were performed in our patient, enabling us to measure the diameter of the PFO, which was 4 mm. Diameters of up to 6.3 mm or 15 mm have been reported in the literature [17] [18].

Imaging methods such as CT or MRI can detect abnormalities such as pulmonary embolism or pulmonary arteriovenous malformation [19].

Percutaneous closure of the PFO is the most effective option, correcting the shunt and improving the New York Heart Association functional class [10] [20]. The same observation was made in our patient, whose symptoms improved after PFO closure.

The results of long-term follow-up of patients with platypnea ortthoxia syndrome published to date are limited. However, these data suggest a good prognosis, both in terms of shunt correction and symptomatic improvement [8] [16]. The series of 78 patients by Guerin *et al.* in 2005 was the largest to date. 20 at an average follow-up of 15 months, only one patient required reoperation. At 6 months, a small shunt was observed on echocardiography in only 6 patients, none of whom were symptomatic. Importantly, there were no major adverse events related to the procedure [20].

In the study by Mojadidi *et al.* [6], 64.8% of patients who received percutaneous PFO closure were classified as having improved saturation and experienced improvement or complete resolution of their dyspnoea and hypoxaemia. Patients with no change after PFO closure had predominantly a pulmonary aetiology for their hypoxia, with elevated mean pulmonary pressures measured before closure (51.4 ± 16.8 mmHg). PFO closure can resolve symptomatic postural dyspnoea and hypoxaemia and is an effective method of treating Platypnea-Orthodeoxia, but is not effective when the primary aetiology of hypoxaemia is due to a pulmonary cause [6].

4. Conclusion

Platypnea-Orthodeoxia syndrome is under-diagnosed. It is important to look for

PFO in patients with concomitant pulmonary and systemic embolisms. We believe that clinicians need to be aware of this syndrome because early diagnosis and treatment can help prevent worsening morbidity and mortality.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

[1] Gourgiotis, S., Aloizos, S., Gakis, C. and Salemis, N.S. (2011) Platypnea-Orthodeoxia Due to Fat Embolism. *International Journal of Surgery Case Reports*, **2**, 147-149. https://doi.org/10.1016/j.ijscr.2011.02.015

[2] Mima, H., Sakamoto, J., Miyake, M., Tamaki, Y., Enomoto, S., Kondo, H., *et al.* (2022) Posture-Related Change in Intracardiac Blood Flow Detected by Transesophageal Echocardiography in Platypnea-Orthodeoxia Syndrome. *CASE*, **6**, 218-222. https://doi.org/10.1016/j.case.2022.03.007

[3] Samra, G.S., Noory, A.J., Gajjar, K., Lasorda, D. and Radhakrishnan, A.U. (2022) Successful Multimodality Investigation of Refractory Platypnea Orthodeoxia Necessitating Redo Percutaneous Patent Foramen Ovale Closure. *Journal of the American College of Cardiology*, **79**, 2905. https://doi.org/10.1016/S0735-1097(22)03896-7

[4] Mizuma, K., Sugimoto, A., Mochizuki, Y., Shinke, T. and Ono, K. (2022) A Case Report of Platypnea-Orthodeoxia Syndrome: A Rare Condition Found during Diagnostic Workup of a Patient with Embolic Stroke of Undetermined Sources. *eNeurologicalSci*, **26**, Article ID: 100393. https://doi.org/10.1016/j.ensci.2022.100393

[5] Singh, V.P., Thalji, M., Singh, S., Hafeez, H., Buradkar, A., Bhat, S.P., *et al.* (2021) A Case of Platypnea-Orthodeoxia Syndrome with Primary Pulmonary Hypertension. *Journal of the American College of Cardiology*, **77**, 2790. https://doi.org/10.1016/S0735-1097(21)04145-0

[6] Othman, F., Bailey, B., Collins, N., Lau, E., Tanous, D., Rao, K., *et al.* (2022) Platypnea-Orthodeoxia Syndrome in the Setting of Patent Foramen Ovale without Pulmonary Hypertension or Major Lung Disease. *Journal of the American Heart Association*, **11**, e024609. https://doi.org/10.1161/JAHA.121.024609

[7] Knapper, J.T., Schultz, J., Das, G. and Sperling, L.S. (2014) Cardiac Platypnea-Orthodeoxia Syndrome: An Often Unrecognized Malady. *Clinical Cardiology*, **37**, 645-649. https://doi.org/10.1002/clc.22301

[8] Shah, A.H., Osten, M., Leventhal, A., Bach, Y., Yoo, D., Mansour, D., *et al.* (2016) Percutaneous Intervention to Treat Platypnea-Orthodeoxia Syndrome: The Toronto Experience. *JACC*: *Cardiovascular Interventions*, **9**, 1928-1938. https://doi.org/10.1016/j.jcin.2016.07.003

[9] Mojadidi, M.K., Gevorgyan, R., Noureddin, N. and Tobis, J.M. (2015) The Effect of Patent Foramen Ovale Closure in Patients with Platypnea-Orthodeoxia Syndrome. *Catheterization and Cardiovascular Interventions*, **86**, 701-707. https://onlinelibrary.wiley.com/doi/full/10.1002/ccd.25953 https://doi.org/10.1002/ccd.25953

[10] Fenster, B., Nguyen, B. and Buckner, J. (2013) Effectiveness of Percutaneous Closure of Patent Foramen Ovale for Hypoxemia. *The American Journal of Cardiology*, **112**, 1258-1262. https://doi.org/10.1016/j.amjcard.2013.06.022

[11] Takiguchi, H., Niimi, K., Aoki, T., Ogiya, R., Ohno, Y., Nakazawa, G., *et al.* (2013) Platypnea-Orthodeoxia Syndrome Caused by a Latent Atrial Septal Defect. *Internal Medicine*, **52**, 1809-1811. https://doi.org/10.2169/internalmedicine.52.0578

[12] Hayek, A., Rioufol, G., Bochaton, T., Rossi, R., Mewton, N., Paccalet, A., *et al.* (2021) Prognosis after Percutaneous Foramen Ovale Closure among Patients with Platypnea-Orthodeoxia Syndrome. *Journal of the American College of Cardiology*, **78**, 1844-1846. https://doi.org/10.1016/j.jacc.2021.08.050

[13] Torres, R., Wiener, P.C. and Salacup, G. (2020) Platypnea-Orthodeoxia Syndrome Post Atrial Fibrillation Ablation. *Journal of the American College of Cardiology*, **75**, 3316. https://doi.org/10.1016/S0735-1097(20)33943-7

[14] Agrawal, A., Palkar, A. and Talwar, A. (2017) The Multiple Dimensions of Platypnea-Orthodeoxia Syndrome: A Review. *Respiratory Medicine*, **129**, 31-38. https://doi.org/10.1016/j.rmed.2017.05.016

[15] Kenny, C.M., Murphy, C.E., Clough, L. and Ashley, D. (2019) Positional Cyanosis Reveals Platypnea-Orthodeoxia-Syndrome. *Respiratory Medicine Case Reports*, **28**, Article ID: 100941. https://doi.org/10.1016/j.rmcr.2019.100941

[16] De Vecchis, R., Baldi, C. and Ariano, C. (2016) Platypnea-Orthodeoxia Syndrome: Multiple Pathophysiological Interpretations of a Clinical Picture Primarily Consisting of Orthostatic Dyspnea. *Journal of Clinical Medicine*, **5**, Article 85. https://www.mdpi.com/2077-0383/5/10/85/htm https://doi.org/10.3390/jcm5100085

[17] Uchihashi, M., Makino, M., Kaimoto, S., Imai, Y., Hadase, M., Kurata, H., *et al.* (2020) Platypnea-Orthodeoxia Syndrome Associated with Spontaneously Ruptured Chordae Tendineae of Tricuspid Valve. *CASE*, **4**, 90-92. https://doi.org/10.1016/j.case.2019.10.006

[18] Kuzma, M., Bowers, J. and Farbaniec, M. (2020) Platypnea-Orthodeoxia Syndrome with Normal Right Heart Pressures in an Octogenarian. *JACC*: *Case Reports*, **2**, 324-325. https://doi.org/10.1016/j.jaccas.2019.12.020

[19] Hagen, P.T., Scholz, D.G. and Edwards, W.D. (1984) Incidence and Size of Patent Foramen Ovale during the First 10 Decades of Life: An Autopsy Study of 965 Normal Hearts. *Mayo Clinic Proceedings*, **59**, 17-20. https://doi.org/10.1016/S0025-6196(12)60336-X

[20] Guérin, P., Lambert, V., Godart, F., Legendre, A., Petit, J., Bourlon, F., *et al.* (2005) Transcatheter Closure of Patent Foramen Ovale in Patients with Platypnea-Orthodeoxia: Results of a Multicentric French Registry. *CardioVascular and Interventional Radiology*, **28**, 164-168. https://doi.org/10.1007/s00270-004-0035-3