Case Report

BIOMEDICAL Journal of Scientific & Technical Research

ISSN: 2574 -1241

DOI: 10.26717/BJSTR.2023.53.008412

A Hole with a Heart Around: Platypnea-Orthodeoxia Syndrome

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ARTICLE INFO

Received: October 16, 2023

Published: October 27, 2023

Citation: Martinelli E, Di Stefano A, Sapone P, Padulo F, Mondino S, Carlucci R and Cotroneo AM. A Hole with a Heart Around: Platypnea-Orthodeoxia Syndrome. Biomed J Sci & Tech Res 53(3)-2023. BJSTR. MS.ID.008412.

ABSTRACT

Platypnea-Orthodeoxia Syndrome [POS] is dyspnea and hypoxemia in the upright position that improves in the supine position. Cardiac POS is predominantly caused by congenital interatrial communications [CIC] paired with changes in the thoracic anatomy, allowing orthostatic right to left cardiac shunting. High suspicion, especially with hypoxemia without significant pulmonary disease, that does not easily correct with supplemental oxygen, should lead the clinician to obtain echocardiographic imaging, documenting right to left shunting, typically through a patent foramen ovale [PFO]. Transcatheter closure of the CIC is highly successful in relieving symptoms of dyspnea and resolving hypoxemia in the majority of patients.

Keywords: Platypnea-Orthodeoxia Syndrome; Patent Foramen Ovale; Hypoxemia; Intracardiac Shunts

Abbreviations: POS: Platypnea-Orthodeoxia Syndrome; ASD: Atrial Septal Defect; TTE: Transthoracic Echo-Cardiogram; CIC: Congenital Interatrial Communications; PFO: Patent Foramen Ovale

Introduction

Platypnea-orthodeoxia syndrome [POS] is a rare condition, characterized by positional dyspnea and hypoxemia, described for the first time in 1949 by Burchell [1]. It is uncommon, and occurs predominantly in the elderly, being more prevalent in women than men [2]. It is characterized by the appearance of dyspnea and desaturation in upright position, relieved by recumbency. It is considered to be caused by a temporary increase in intracardiac right-to-left shunt [2]. Four main causes of POS can be recognised: intracardiac shunt, pulmonary shunt, ventilation-perfusion mismatch or a combination of these [3,4]. POS occurs due to the presence of a right-to-left shunt through a PFO, less commonly through an atrial septal defect [ASD] [2,5]. More rarely, POS can be caused by ventilation/perfusion imbalances of the lung parenchyma, observed in chronic obstructive pulmonary disease, interstitial lung disease, hepatopulmonary syndrome, and pulmonary arteriovenous malformations. POS of cardiac origin can manifest insidiously as worsening dyspnea on exertion over the course of months or years, or with an episode of apparently unexplained acute dyspnea that puts the patient's life at risk [6].

Case Reports

Mrs ZD, an 85-year-old woman with a history of hyperthyroidism, arterial hypertension, vascular neurocognitive disorder, previous cerebral ischemic stroke, anxious-depressive syndrome, obesity, presented to our hospital for repeated falls and episodes of disorientation, hallucinations and cyanosis associated with new onset of hypoxemia and dyspnea. She had no history of lung disease or tobacco smoking. Upon arrival in the emergency room, the patient was alert, uncooperative. Her physical examination findings on admission were as follows: diffusely reduced vesicular murmur, rhythmic, paraphonic heart tones and severe hypoxia and cyanosis. Arterial blood gas values were: PH 7.48, pCO2 31, pO2 41, HCO3- 23, Lac 1.42. She required 10L/min of oxygen via face mask to maintain oxygen saturation in the low 90s. On chest x-ray, elevation of the right hemidiaphragm. Contrast-enhanced CT excluded the diagnosis of cerebral infarction, pulmonary embolism, pneumonia, and lung parenchymal abnormalities.

On further evaluation, she was found to have worsening oxygen needs when changing from a lying to seated position and standing

up from a recumbent position. The clinical picture was consistent with platypnea-orthodeoxia. A transthoracic echo-cardiogram [TTE] revealed a normal ejection fraction, mid-apical septal hypo-akinesia; grade I diastolic dysfunction, ventricular filling pressures within limits; normal atria and interatrial septum with aneurysm of the oval fossa in the absence of visible shunts; so a TTE with intra-venously administered agitated saline contrast solution was performed and it showed a right-to-left shunt, probably through the patent foramen ovale, in the absence of signs of pulmonary hypertension and excessive dilatation of the right sections of the myocardium (Figures 1 & 2; Video 1).

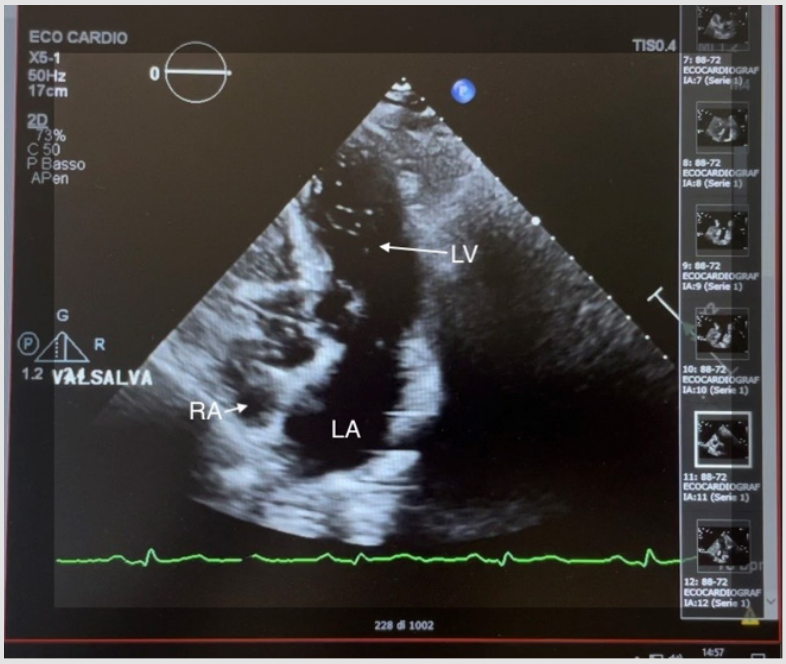


Figure 1: Transthoracic echo-cardiogram with bubble contrast showed the presence of right-to-left shunt through patent foramen ovale. LA left atrium, RA right atrium, LV Left ventricle.

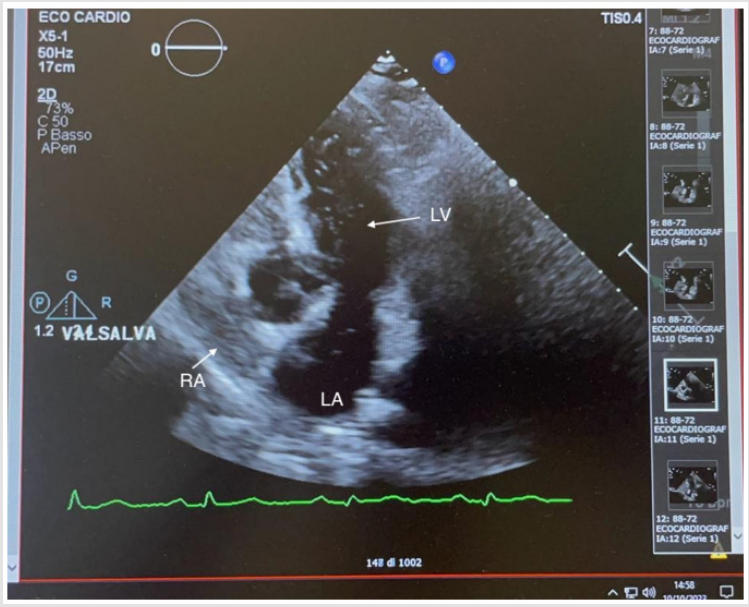
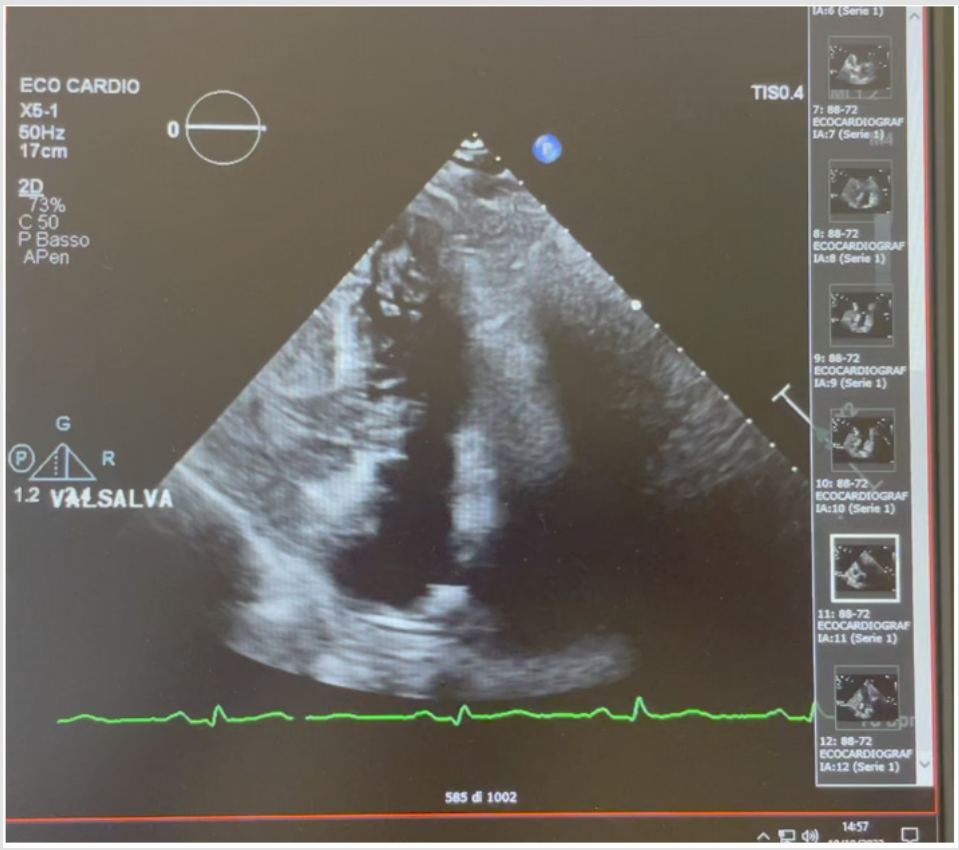


Figure 2: Transthoracic echo-cardiogram with bubble contrast showed the presence of right-to-left shunt through patent foramen ovale. LA left atrium, RA right atrium, LV Left ventricle.



Video 1: Transthoracic echo-cardiogram with bubble contrast showed the presence of right-to-left shunt through patent foramen ovale.

Considering these results, we concluded that the cause of hypoxia was POS with right-to-left interatrial shunt through PFO. POS can be treated with the closure of the interatrial shunt by thoracotomy or percutaneous cardiac catheterization [3]. Surgical closure of PFO was not performed due to the age of our patient and the clinical general condition, surgical difficulties, and failure to obtain informed consent. For these reasons she was discharged after receiving oxygen, medical therapy and advice about her posture.

Discussion

The most important clue for diagnosis of POS is the emergence of hypoxia with postural changes. Several previous cases have demonstrated that the increased shunt flow across PFO or ASD with posture changes from supine to upright induce hypoxia [7]. The standing upright position can directly change the anatomical route from the vena cava to an intracardiac defect in a straight line, which increases right-to-left shunt [3]. POS is caused by the coexistence of anatomical and functional abnormalities [3]. Anatomical abnormalities include ASD, PFO, or fenestrated atrial septal aneurysm. However, the presence of only these anatomical factors cannot induce POS because they usually cause left-to-right shunting. In addition, functional factors, such as right hemidiaphragmatic paralysis, pericardial effusion, constrictive pericarditis, emphysema, arteriovenous malformation, pneumonectomy, liver cirrhosis, aortic aneurysm, or elongation changes of shunt flow can determine POS [8]. The causes can be or a hemodynamic phenomenon with an interatrial pressure gradient, or a selective blood flow from the vena cava to the left atrium passing through an intercardiac defect [9]. TTE has low sensitivity for small shunts and so the prevalence of PFO observed in the general population was reported to be lower than that observed by transoesophageal echocardiogram [TOE] [14.9% versus 24.3%] [10]; this indicates that only TTE can fail the diagnosis of ASD or PFO. POS caused by PFO can be clearly demonstrated by the technique of contrast TOE [11].

There are previous reports where surgical closure of ASD or PFO were successfully performed [12-14]; however, percutaneous transcatheter closure devices have recently come into use because of their reduced invasiveness and are easy and safe compared with open surgery [11,15].

Conclusion

The pathophysiology of POS is not yet well clarified; however it is believed that age-related mechanical and physiological factors are able to act on a pre-existing congenital interatrial communication,

determining a right-left interatrial shunt. This shunt would become more conspicuous with the assumption of the upright position due to the presence of tethering forces capable of making the communication between the two atria wider [16].

Normally in adults, the pressure in the left atrium is slightly higher than the pressure in the right atrium so, even in the presence of interatrial communications, a right-left shunt does not occur. This shunt becomes evident when the pressure in the right atrium becomes acutely [for example in the case of pulmonary embolism, ventricular acute myocardial infarction, acute pericardial effusion] or chronically [any condition of chronic pulmonary hypertension] greater than that in force in the left atrium [17]. However, in the majority of patients with POS of cardiac origin, the pressure in the right atrium is not elevated [18,19]; this eventuality should therefore not distract from the possible diagnosis of POS.

It should also be taken into account that not all people with atrial septal defect will develop POS. So, what happens in patients suffering from this syndrome? It is not always possible to identify the triggering event, however it is necessary to consider the possibility of POS for every patient with dyspnea and desaturation in orthostatism, especially when the symptoms are poorly responsive to oxygen therapy and are alleviated by assuming the supine position. In such patients, although it may seem absurd to look for a congenital pathology at an advanced age, the presence of atrial septal defects must be firmly investigated. These conditions are in fact often susceptible to treatment percutaneously [transcatheter implantation of a closure device] or surgically with resolution of the symptoms [20-22]. In patients in whom POS is due to extracardiac causes [rarer], depending on the pathogenesis, the treatment may be represented by arterial embolization of pulmonary arteriovenous malformations, surgery/lung transplant and, in cases of hepato-venous syndrome lung in liver transplantation.

Acknowledgements

The authors sincerely thank the patient, who gave consent for publication of this case report. The authors sincerely thank Doctor Davide Forno, Cardiologist of Maria Vittoria Hospital for the kind and precious collaboration.

Conflict of Interest

The authors declare that they have no competing interests.

References

1. Burchell HB (1949) Reflex orthostatic dyspnea associated with pulmonary hypotension. Am J Physiol 159: 563-564.
2. Agrawal A, Palkar A, Talwar A (2017) The multiple dimensions of Platypnea Orthodeoxia syndrome a review. Respir Med 129: 31-38.
3. Cheng TO (1999) Platypnea-orthodeoxia syndrome etiology, differential diagnosis, and management. Catheter Cardiovasc Interv 47 (1): 64-66.
4. Chen GP, Goldberg SL, Gill EA Jr (2005) Patent foramen ovale and the platypnea-orthodeoxia syndrome. Cardiol Clin 23(1): 85-89.
5. Henkin S, Negrotto S, Pollak PM, Cullen M, OCochlain F, et al. (2015) Platypnea-Orthodeoxia Syndrome: Diagnostic Challenge and the Importance of Heightened Clinical Suspicion. Tex Heart Inst J 42(5): 498-501.
6. Rudy CC, Ballard C, Broberg C, Hunter AJ (2017) Platypnea-orthodeoxia syndrome: a case of chronic paroxysmal hypoxemia. J Gen Intern Med 32(1): 127-130.
7. Bonpei T, Yoshihiro T, Hidemi H (2012) Importance of Platypnea orthodeoxia in the differential diagnosis of dyspnea. Intern Med 51: 1651-1652.
8. Cheng TO (2002) Mechanisms of platypnea-orthodeoxia: what causes water to flow uphill? Circulation 105: e47.
9. F Godart, C Rey, A Prat, A Vincentelli, A Chmaït (2000) Atrial right to left shunting causing severe hypoxaemia despite normal right sided pressures. Eur Heart J 21(6): 483-489.
10. Miriam Roxas Timonera, Cristina Larracas, Deborah Gersony, Marco Di Tullio, Andrew Keller, et al. (2001) Patent foramen ovale presenting as platypnea-orthodeoxia: Diagnosis by transesophageal echocardiography. J Am Soc Echocardioqr 4(10): 1039-1041.
11. Neill J, Lin H (2017) A review of transcatheter closure of patent foramen ovale. Methodist Debakey Cardiovasc J 13: 152-159.
12. Ryota Fukuoka, Tomohiro Kawasaki, Hisashi Koga, Kyoko Umeji, Yoshiya Orita, et al. (2014) Platypnea-orthodeoxia syndrome associated with a thoracic vertebral fracture following a car accident. Intern Med 53(1): 35-38.
13. Nobutaka Hirai, Takashi Fukunaga, Hiroaki Kawano, Osamu Honda, Tomohiro Sakamoto, et al. (2003) Platypnea-orthodeoxia syndrome with atrial septal defect. Circ J 67(2): 172-175.
14. Yasuhiro Nagayoshi, Kensuke Toyama, Hiroaki Kawano, Ikuo Misumi, Shinzo Miyamoto, et al. (2005) Platypnea-orthodeoxia syndrome combined with multiple congenital heart anomalies. Intern Med 44(5): 453-457.
15. Yoichi Takaya, Teiji Akagi, Yasufumi Kijima, Koji Nakagawa, Manabu Taniguchi, et al. (2014) Transcatheter closure of right to left atrial shunt in patients with platypnea-orthodeoxia syndrome associated with aortic elongation. Cardiovasc Interv and Ther 29(3): 221-225.
16. Layoun ME, Aboulhosn JA, Tobis JM (2017) Potential role of patent foramen ovale in exacerbating hypoxemia in chronic pulmonary disease. Tex Heart Inst J 44(3): 189-197.
17. Zanchetta M, Rigatelli G, Ho SY (2005) A mystery featuring right-to-left shunting despite normal intracardiac pressure. Chest 128: 998-1002.
18. Homma S, Messé SR, Rundek T, Sun YP, Franke J, et al. (2016) Patent foramen ovale. Nat Rev Dis Primers 2: 15086.
19. Knapper JT, Schultz J, Das G, Sperling LS (2014) Cardiac platypnea-orthodeoxia syndrome: an oftenunrecognized malady. Clin Cardiol 37: 645-649.
20. Guerin P, Lambert V, Godart F, Legendre A, Petit J, et al. (2005) Transcatheter closure of patent foramen ovale in patients with platypnea-orthodeoxia: results of a multicentric French registry. Cardiovasc Intervent Radiol 28(2): 164-168.
21. Kay JD, OLaughlin MP, Ito K, Wang A, Bashore TM, et al. (2004) Five-year clinical and echocardiographic evaluation of the Das AngelWings atrial septal occluder. Am Heart J 147: 361-368.
22. Rao PS, Palacios IF, Bach RG, Bitar SR, Sideris EB (2001) Platypnea-orthodeoxia: management by transcatheter buttoned device implantation. Catheter Cardiovasc Interv 54(1): 77-82.

ISSN: 2574-1241

DOI: 10.26717/BJSTR.2023.53.008412

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