**Case Report**

**Platipnea-Orthodeoxia and PFO Syndrome: Intuition or Praise of the Practice?**

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**Abstract**

We present the case of a patient with platypnea-orthodeoxia syndrome (POS) and we discuss its clinical features and its physiopathology. The manifestations of the syndrome are dyspnea and oxygen desaturation that occur in an upright position and that improve in the supine position. In February 2018 a 26-year-old girl presented herself at our department of cardiology, reporting on palpitations and fainting. She also complained about unusual fatigue. Why not think about minor syndromes associated with PFO? After a series of examinations, the diagnostic conclusion of a patent oval foramen (PFO) associated with an interatrial aneurysm was reached.

It is believed that the syndrome remains very underdiagnosed, although cases in the literature have been described from the original description of Burtchell and Wood in 1949.

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**INTRODUCTION**

The most important clue for the diagnosis of POS is the emergence of hypoxia with postural alterations. Several previous cases have shown that the increase in shunt flow through PFO or atrial septal defect (ASD) with changes of posture from supine to straight induces hypoxia. POS is caused by the coexistence of anatomical and functional abnormalities. Anatomical abnormalities include ASD, PFO, or atrial septal aneurysm.

Because of the lower sensitivity of TTE for small shunts, the prevalence of PFO observed for TTE of contrast in the general population was lower than that observed by TEE or transcranial ultrasound; this indicates that only the TTE can fail to diagnose ASD or PFO. The POS caused by the PFO can be clearly demonstrated by the contrast technique.

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**CASE PRESENTATION**

A 26-year-old woman presented herself to our cardiology department, reporting on palpitations and fainting. His clinical history was normal - in particular, there was no history of cardiovascular disease; he also claimed to regularly engage in sports. The patient was admitted to the hospital for diagnostic tests.

His vital signs upon arrival were as follows: blood pressure, 90/60 mmHg; heart rate 59 beats / minute; respiratory rate, 18 breaths / minute; and axillary temperature, 36.9 °C. From a neurological point of view, she was attentive, time-oriented [1,2].

Blood tests revealed microcitosis (hemoglobin = 13.8 g / dL, mean corpuscular volume = 63 fl, mean corpuscular hemoglobin = 20.8 pg), a common condition. No alterations in the coagulation profile were found. No electrocardiographic changes were detected.

A transthoracic echocardiogram (TTE) revealed an interatrial aneurysm but the flow derived through the PFO on the Doppler color image was apparently not observed (Figure 1). Given the suspicion of a foramen ovale, the patient was subjected to transcranial ultrasound (Figure 2). The right middle cerebral artery is visualized, which after the injection into a peripheral vein of physiological solution and Valsalva maneuver, shows the passage of microbubbles in a significant number indicative of PFO.

The diagnosis is often delayed, so the clinical suspicion is important.

Considering these results, we concluded that the cause of hypoxia was POS with right-to-left interatrial shunting across the PFO. POS can be treated by closing the interatrial shunt...
by thoracotomy or percutaneous cardiac catheterization. The closure of the PFO has not been performed due to failure to obtain informed consent. For these reasons she was discharged after receiving a medical opinion on her posture. The patient was discharged with aspirin [3,4].

DISCUSSION

This case showed a PFO associated with an aneurysmatic interatrial septum. The PFO is a common finding in the general population and may be completely asymptomatic. Nonetheless, PFO has been associated with an increased risk of cryptogenic ischemic stroke and even more in cases of association with atrial septal aneurysm.

The mean percentage of cryptogenic strokes among ischemic strokes is 31%, of which one-third may be associated with patent foramen ovale. The foramen ovale is required for blood flow through the fetal atrial septum. In 75% of cases, its closure is complete by 2 years of age, but it may persist in 25% of patients.

There after careful clinical and instrumental evaluation a platypnea-orthodeoxia syndrome has been diagnosed.

Platypnea-orthodeoxia syndrome (POS) is an uncommon disorder characterized by dyspnea (=platypnea) and desaturation (=orthodeoxia) in upright position and improvement of symptoms and blood oxygenation in supine position. Clinical presentation is heterogeneous and often confuses the clinician.

It is characterized by a significant reduction of the arterial oxygen saturation in the transition from the supine position to the orthostatic position. It would be 'precipitated' by a reduction of the systemic arterial pressure in an upright position thus increasing the right-left shunt. In these conditions, in fact, the increase of the right-left pressure gradient resulting in an upright position leads to an increase in the deoxygenated venous blood flow between the right and left chambers and therefore the finding of a greater oximetry desaturation in orthostatism.

Once Platypnea-Orthodeoxia syndrome is diagnosed in agreement with other authors, both an anatomical and functional cause must be sought.

Additional tests will demonstrate the existence of a patent foramen ovale. This diagnosis was suitable with the patient's syndrome and was eligible for percutaneous closure procedure allowing immediate resolution of symptoms and confirming the diagnosis.

The anatomical defect consists almost exclusively in the patentcy of the oval foramen or in a misrecognized interatrial defect.
while the functional defect may be due or those that determine a preferential blood flow through interatrial communication (ex: aneurysm of the inter-atrial septum) or those causing a transient increase in pressure in the right atrium with an increase in the left right atrial gradient (ex: chronic obstructive pulmonary disease).

In the case that we described at the end we have diagnosed a misunderstood wide patency of the foramen ovale or a true interatrial aneurysm.

In fact, the aneurysm of the interatrial septum can in itself facilitate the movement of the hinge valve of the oval fossa, being able to become more horizontally in an orthostatic position, thus favoring the deoxygenated blood flow coming from the inferior vena cava.

It is recalled that the patency of the oval foramen is not a rare condition; in fact it has a prevalence of 20-35% in autopsy studies and 9.2% in patients who undergo a transesophageal echocardiography.

The correction of the patency of the oval foramen is curative for the syndrome platypnea-orthodeoxia, but there are still conflicting data [5,6].

Although the syndrome still remains undiagnosed, it will be increasingly recognized for the use of the echocardiographic method not only transthoracic but above all transesophageal and transcranial echo. Furthermore, there is an increasing awareness of the existence of the syndrome and of the healing correction if diagnosed in time.

CONCLUSIONS

Platypnea-orthodeoxia syndrome is rare and difficult to diagnose. The present case suggests that hypoxia due to postural changes should be considered a differential diagnosis of platypnea-orthodeoxia syndrome. The abnormal shunt in a vertical position can be the result of different basic conditions, which require an accurate workup and a tailor-made treatment.

Why not think of minor syndromes associated with PFO? After a series of tests, the diagnostic conclusion of a patented oval foramen (PFO) associated with an interatrial aneurysm was achieved.

Medical therapy with aspirin (and postural advice), a valid alternative not decisive in the management of young patients.

The percutaneous correction of patency of the oval foramen is curative for the platypnea-orthodeoxia syndrome.

REFERENCES