

CASE REPORT

Platypnea-orthodeoxia syndrome: an intriguing perioperative hypoxemia case report



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Received 7 September 2020; accepted 22 May 2021
Available online 9 June 2021

KEYWORDS

Dyspnea;
Hypoxia;
Hypotension;
Perioperative period;
Patent Foramen
Ovale;
Case report

Abstract Platypnea-orthodeoxia syndrome (POS) is a rare condition of positional hypoxemia and dyspnea. The following is a case of hypoxemia for no obvious reason in the perioperative scenario. A 70-year-old male patient was submitted to a radical prostatectomy. On several occasions after anesthetic induction, peripheral oxygen saturation decreased without any understandable cause. In the postanesthesia care unit, severe hypoxemia occurred in the upright position, oddly correcting with recumbency. An echocardiography showed a right-to-left intracardiac shunt through a patent *foramen ovale* with no pulmonary hypertension, establishing POS. Achieving this diagnosis required a broad workup with a high degree of suspicion.

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Introduction

Hypoxemia is frequent in anesthesiology practice, and can be hypoxemic, anemic, circulatory, or histiocytic in nature. Although rare, a reversible cause like Platypnea-orthodeoxia syndrome (POS) must be thought of.¹ It presents with positional hypoxemia and dyspnea in the upright position, but may have near-normal oxygen saturation levels when lying down.²⁻⁴

POS is due to three types of causes: intracardiac shunts, arteriovenous shunts, or ventilation/perfusion mismatch in the lungs.^{3,4} Cardiac conditions are among the most described causes, in particular related to patent *foramen ovale* (PFO).⁴ The mechanism of position-dependent changes in shunting remains elusive, but requires a functional and anatomical defect, and the common pathway seems to be a position-dependent opening of the PFO with right-to-left shunting.²

There are no case reports in the anesthesiology field describing the diagnosis of POS in the perioperative scenario. Our goal is to describe a case of hypoxemia without a more obvious reason in this setting. Informed consent

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from the patient and the Institutional Ethical Committee's approval were duly obtained.

Case report

A 70-year-old male patient with prostatic carcinoma was proposed for a radical retropubic prostatectomy. His background included human immunodeficiency virus infection, a one-year prior ischemic cerebellar stroke, discrete bronchiectasis, arterial hypertension, and initial signs of dementia. The patient denied dyspnea or any other cardiovascular symptoms. No previous surgical nor anesthetic interventions. The preanesthetic physical examination was innocent. His laboratory results and electrocardiogram revealed no significant changes.

The patient was submitted to the procedure under general balanced anesthesia and standard monitoring of American Society of Anesthesiologists. Preoperative peripheral oxygen saturation (SpO₂) was 95%. On several occasions after anesthetic induction, SpO₂ decreased from 100% to 86–90% without any feasible causes apart from the fall of mean arterial pressure (MAP) to 60 mmHg. The desaturation did not improve with increased fraction of inspired oxygen nor with recruitment maneuvers, only recovering after phenylephrine 0.1 mg boluses and MAP elevation to 90 mmHg. The operation lasted 60 minutes, the estimated blood loss was 700 ml, and a total of 0,5 mg of phenylephrine was administered. Anesthesia recovery and transfer to the postanesthesia care unit (PACU) was uneventful. In the PACU, even though the patient had almost no complaints of dyspnea, severe hypoxemia occurred (paO₂ 5.48 kPa, SpO₂ 73.4%), despite oxygen therapy with Venturi mask at 15 L·min⁻¹ oxygen flow. The patient empirically received intravenous hydrocortisone, intravenous aminophylline, and nebulization of ipratropium/salbutamol in standard doses. The clinical benefit was minimal.

Analytical control was carried out, showing no significant alterations. A computed tomographic angiography showed peripheral subsegmental pulmonary embolism (PE) that did not fully explain the clinical picture. Oddly, we have noticed significant hypoxemia correction with recumbency (SpO₂ raised to 94–96%) and aggravation with lifting the headboard and sitting position. A transthoracic echocardiography at the bedside was requested, and it revealed findings suggestive of a right-to-left intracardiac shunt through a PFO with no pulmonary hypertension. All intracardiac cavities showed no signs of dysfunction nor dilation, with good global systolic function. No valvular or pericardial structural changes were identified. A bubble test was performed, allowing to visualize microbubbles in the left heart cavities, clearly establishing a right-to-left shunt, and assuming the diagnosis of POS.

After the diagnosis, he maintained high flow oxygen therapy, associated with avoidance of upright position. The POS manifestations remained in the postoperative period, with a progressive reduction in oxygen requirements (SpO₂ 95% lying in bed, and SpO₂ 91–92% in the sitting position, without significant dyspnea). He was ultimately discharged to a long-term care unit, with long-term nocturnal oxygen therapy, and awaiting a cardiology consultation to guide the therapeutic approach to the POS.

Discussion

Although it likely remains underdiagnosed, the recognition of POS is increasing.² In the preanesthetic evaluation, we should look for an association between breathlessness and upright position. POS mostly occurs in older people, although, in most cases, it is linked to a congenital heart defect, especially PFO.³ SpO₂ also needs to be analyzed in the lying and the upright position. A drop in the SpO₂ of > 5% in the upright position with improvement on recumbency, should elicit the diagnosis of POS.⁴

In the perioperative period, hypoxia is most likely caused by an inadequate oxygen supply; hypoventilation; alveolar ventilation/perfusion mismatch; right-to-left shunting; and reduction in oxygen carrying and using capacity (decreased cardiac output; anemia and histiocytic hypoxia).¹ The clinical presentation of POS can be subtle and difficult to attain under general anesthesia, so it must be reminded in the differential diagnosis of refractory hypoxemia, after exclusion of classic etiologies in the perioperative setting. As we verified, these patients have a minimal or no response at all to oxygen supply in the upright position,^{2,4} nor to traditional therapies for lung and heart dysfunction.⁴

PFO has a prevalence of 25% in general adult population.^{2,3} Most people with an isolated PFO are asymptomatic because they do not have right-to-left shunt, since the left atrial pressure is higher than the right atrial pressure, leading to a functional closure.^{3,4} Right-to-left atrial shunting despite normal intracardiac pressures and normal pulmonary function through a PFO has still not been completely elucidated, but requires both an anatomical and functional flaw in order to occur.^{2,4} Functional defects act through transient reversal of the left-to-right pressure gradient, because they are capable of elevating right atrial pressure to the point that the gradient reverses for certain parts of the cardiac cycle, particularly when the patient is standing.² Such phenomenon can be physiologic (posture, inspiration, cough, or Valsalva maneuver)³ or pathological, producing an increased pulmonary vascular resistance or leading to decreased right-sided compliance, frequently age-related (PE, ascending aortic dilatation, diaphragmatic paralysis, obstructive sleep apnea, chronic obstructive pulmonary disease, right ventricular infarction, constrictive pericarditis, pericardial effusion, right atrial myxoma, eosinophilic myocardial disease, etc.).² At the same time, interatrial septum and PFO may acquire a conformational transformation during adulthood that causes blood flow to preferentially route from the right atrium to the left, especially when seated upright, via repositioning of the atrial septum, putting it directly in line with the blood flow from the inferior vena cava.⁴

Our patient had never exhibited symptoms of POS until this intervention. We have seen important desaturation during hypotensive periods in the intraoperative phase. A drop in systemic vascular resistance and significant blood loss probably raised intracardiac shunt fraction, as it has been demonstrated that hypovolemia can change ventilation-perfusion matching and the right-to-left shunt seems to be more likely to develop when in the dehydrated state.³ Also, desaturation was promptly resolved after phenylephrine boluses, a potent direct-acting alpha-1 adrenergic agonist.

Intravenous α agonists were already used to establish a rapid diagnosis of shunting, upon rapid correction of hypoxemia. Also, it is not uncommon to use adrenergic agonists in cardiac surgery to reduce right-to-left shunting in children with congenital heart defects. In this case, we believe to have temporarily reversed the intracardiac right-to-left shunt by causing a sharp rise in left ventricular end-diastolic and left atrial pressures.³

In our patient's case, we confirmed the harmful influence that PFO exerts, as it can be associated with stroke, paradoxical embolism and POS.^{3,4} In the perioperative period, patients with PFO are exposed to various physiological threats, such as hemorrhage, mechanical ventilation, patient positioning, and the fall in systemic vascular resistance. This increases the vulnerability to stroke due to hemodynamic changes that favors right-to-left shunting, hypercoagulability, and formation of venous thromboemboli.⁵ We have detected a PE that could have led us to stop our investigation, assuming it as the single reason for hypoxemia. However, PE is frequently coexisting with POS.¹ The anesthesiologist must be aware that an otherwise clinically silent PFO, can manifest with POS during a PE. On the other hand, the presence of POS should compel us to seek, not only the presence of an intracardiac communication, but also a possible concomitant disorder.

Transesophageal echocardiography is the gold standard for diagnosing this syndrome. It allows the direct visualization of any cardiac defects or aneurysms that may be present in the atrial septum. However, a transthoracic echocardiography can be useful too. Associating a bubble study with intravenous agitated saline can help us to assess the presence of right-to-left shunting, suggesting an intracardiac shunt if those bubbles appear within three cardiac cycles, and should be the first test in the diagnostic algorithm. These examinations should ideally be executed with the patient in lying and upright positions. Rarely, the shunt can only be seen during a Valsalva maneuver.²

Closure of the atrial septal defect is the best treatment for POS, with a fast relief of symptoms. Still, caution is needed when deciding for definitive treatment, considering the benefit-risk profile for each patient.²⁻⁴

In conclusion, our patient had POS secondary to a PFO without elevated right atrial pressures, making it highly unusual. After excluding more common conditions, especially in elderly patients, where symptoms may otherwise be attributed to ventilation impairment, it is essential to have a high index of suspicion to consider POS, because desaturation and dyspnea are common events in our practice. An echocardiography can suggest the diagnosis and the treatment can signify a great improvement in the patient's quality of life.

Conflicts of interest

The authors declare no conflicts of interest.

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