

Platypnea-orthodeoxia in pregnancy

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Introduction

Breathlessness in pregnancy, especially in the third trimester, is invariably common affecting up to 75% of women [1]. Ventilatory adaptations in pregnancy include increased tidal volume and subsequent minute ventilation resulting from an increase in oxygen consumption, metabolic rate, blood volume and level of progesterone. These changes, in combination with an expanding gravid uterus, lead to the common complaint of dyspnoea.

Case report

Presentation

A 32-year-old primigravida woman at 29 weeks gestation presented with breathlessness. She described worsening dyspnoea and reduced exercise tolerance over the past four weeks on a background of chronic breathlessness for 2-3 years. Usual daily activities of washing and dressing along with climbing stairs would exacerbate symptoms. She reported frequently experiencing dizziness but an absence of palpitations, syncope, cough or wheeze. Of note, she was an ex-prem baby and had a recent diagnosis of asthma and had good compliance with medication.

Examination

Chest auscultation revealed quiet breath sounds with equal air entry bilaterally, no added wheeze or crepitations. SpO₂ 90% on room air which increased to 94% on 4L/minute oxygen via nasal cannula. HR 100 bpm, BP 108/71mmHg. An ejection systolic murmur was present on cardiac auscultation. No clinical evidence of deep vein thrombosis. She had a preference for being supine as symptoms eased in this position.

Investigations

Bloods – Hb 119g/L, WCC 12x10⁹/L, CRP 5mg/L. Renal and liver function tests along with Troponin I and BNP were within normal range.
ECG - sinus rhythm, 97bpm.
Chest x-ray - congested appearance of lungs but nil else.
CTPA – nil PE or evidence of infection.
PFTs - FEV₁ and FVC of 100% and 96% respectively, FEV₁/FVC ratio 0.89.
TTE - normal biventricular size and systolic function, no diastolic dysfunction and LVEF 65%. No significant valvular abnormality or obvious colour flow across the interatrial septum visualised. No evidence of pulmonary hypertension.

Management

Initial medical management was based on the respiratory ward with regular obstetric reviews. A reduction in fetal growth velocity and an increase in oxygen requirements prompted further MDT discussions. Input from obstetric, anaesthetic, cardiology, respiratory, intensive care and neonatal teams was sought with a plan for elective caesarean section at 32 weeks.

A CSE technique with invasive BP monitoring was used for delivery along with the use of nasal high flow oxygen. Low dose oxytocin infusion and metaraminol infusion was used and she remained haemodynamically stable throughout

At seven days post-partum the patient remained significantly symptomatic with positional arterial oxygen desaturation.

A bubble contrast ECHO confirmed passage of microbubbles from the right to left atrium, which increased in volume on Valsalva manoeuvre. A diagnosis of platypnea-orthodeoxia was made.

Discussion

Dyspnoea in pregnancy is a common complaint and although it often can be attributed to physiological adaptation, the differential diagnoses to be considered are extensive. These include asthma, anaemia, pulmonary embolism, pneumonia, pneumothorax and peripartum cardiomyopathy. Of note, cardiac and thromboembolic disease continue to be leading causes of maternal mortality, highlighting the need for rapid diagnosis and treatment.

Red flags in a pregnant patient presenting with breathlessness:

🚩 Sudden-onset breathlessness	🚩 Respiratory rate > 20 breaths per minute	🚩 Orthopnoea
🚩 Breathlessness with chest pain or syncope	🚩 Oxygen saturations <94% or falls to <94% on exertion	🚩 Breathlessness with associated tachycardia

Table 1: Acute care toolkit 15 – Managing acute medical problems in pregnancy[1].

Platypnea-orthodeoxia is a rare clinical condition. It is characterised by the presence of positional dyspnoea and arterial hypoxaemia that is accentuated by being upright and relieved in the supine position.

As an underdiagnosed condition its prevalence in the general population is largely unknown. A literature review in 2017 identified 239 cases of POS in the general population [2], however reports of occurrence within the pregnant population are limited.

Its underlying pathophysiology is complex but is believed to result from right-to-left shunting of deoxygenated venous blood to arterial blood due to an abnormal anatomical and functional defect. Associated conditions are often categorised into intra-cardiac, such as, patent foramen ovale, atrial septal defect or atrial septal aneurysm, or extra-cardiac mechanisms, such as, pulmonary arteriovenous malformations and hepatopulmonary syndrome.

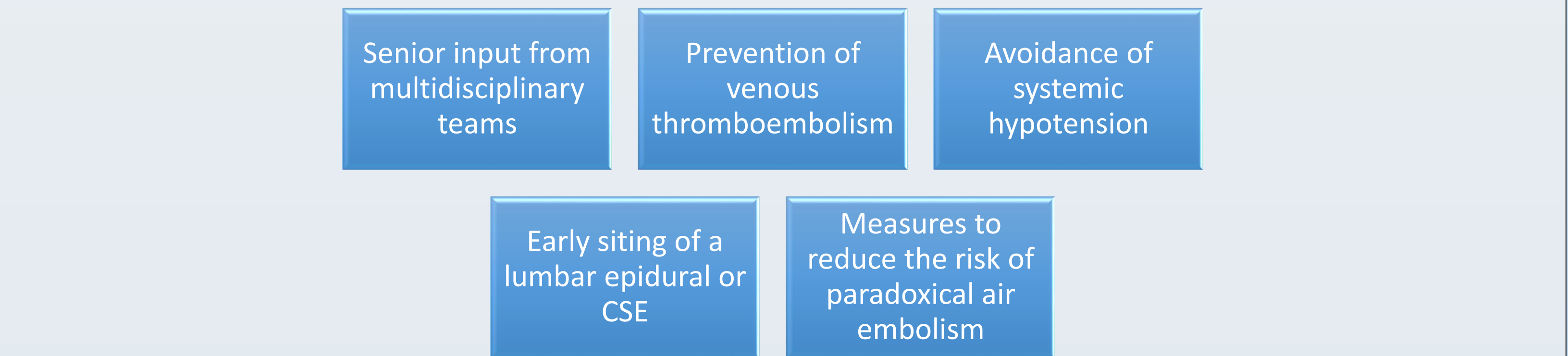
An interatrial communication, as demonstrated in this patient, usually causes a left-to-right shunt owing to the greater magnitude of pressures within the chambers on the left side of the heart and increased compliance on the right. The presence of elevated right atrial pressures or pulmonary hypertension can reverse the direction of shunt. However, crucially, right-to-left flow can still occur in the presence of normal intracardiac pressures. This is hypothesised to occur due to a functional anomaly where blood entering the right atrium via the inferior vena cava is preferentially directed across a PFO or ASD. It is often transient with a predisposition to occur in the upright position, resulting in the postural nature of symptoms.

At face value, the symptom profile of platypnea-orthodeoxia can be vague and overlaps with numerous other conditions. A comprehensive history and clinical examination along with a high degree of suspicion is the first step in making a diagnosis. Although there are no definitive diagnostic criteria, a reduction in SpO₂ >5% in the upright position in comparison to supine is suggestive of platypnea-orthodeoxia [2]. In significant right-to-left shunts, hypoxaemia may persist despite the use of escalating oxygen supplementation.

A transthoracic bubble contrast echocardiogram using agitated saline will enable differentiation between aberrant intracardiac and intrapulmonary communications. Microbubbles can be visualised directly crossing the interatrial septum or are seen in the left atrium within three cardiac cycles to confirm an intracardiac shunt. Intrapulmonary shunts will have a delay in passage of bubbles to the right chambers, usually occurring within three to six cardiac cycles.

Treatment will depend on the underlying aetiology of platypnea-orthodeoxia, including percutaneous closure of ASD or PFO; or embolisation of culprit vessels in the case of pulmonary AVMs.

In respect to management of the parturient with a new presentation of intracardiac cause of platypnea-orthodeoxia, priorities will usually align with those found within the 2018 European Society of Cardiology Guidelines for the Management of Cardiovascular Disease during Pregnancy:



Conclusion

Platypnea-orthodeoxia is a complex condition with multi-factorial aetiology. Despite its rarity, it should be considered in patients with unexplained positional dyspnoea and arterial desaturation. This will allow targeted investigation and potential curative treatment of anatomical abnormalities.

In this case the patient had likely experienced a lifelong degree of hypoxaemia which was exacerbated by the physiological demands of pregnancy.

References

- [1] Acute care toolkit 15 - Managing acute medical problems in pregnancy. Royal College of Physicians. November 2019.
- [2] Agrawal, A., Palkar, A., Talwar, A. The multiple dimensions of Platypnea-Orthodeoxia syndrome: A review. Respiratory Medicine. 129:31-38. (2017).
- [3] European Society of Cardiology Guidelines for the Management of Cardiovascular Disease during Pregnancy. (2018)