Anaesthetic management of postural orthostatic tachycardia syndrome presenting during pregnancy

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Introduction

Postural orthostatic tachycardia syndrome (POTS) represents a heterogeneous group of autonomic disorders with similar clinical characteristics.[1] Postural orthostatic tachycardia syndrome is a subset of orthostatic intolerance and is associated with excessive tachycardia but a maintained blood pressure, on standing. The hallmark of the disorder was originally defined by Schondorf and Low as an increase in heart rate on standing of greater than 30 beats/min or an increase to a heart rate exceeding 120 beats/min in the absence of orthostatic hypotension.[2] It is distinguished from other sinus tachycardia conditions by both the absence of hypotension and its postural element.[3] It can be difficult to distinguish between POTS and inappropriate sinus tachycardia syndrome, however patients with POTS display more pronounced postural change in heart rate.[4]

The prevalence of POTS is approximately 0.2%.[3] The syndrome predominantly occurs in women of childbearing age, with a female:male ratio of 5:1,[5] and it can be classified as primary or secondary.[6] The primary form is often idiopathic and categorised as neuropathic or hyperadrenergic, whereas the secondary forms occur in association with a known disease. Neuropathic POTS is the most common primary form and is usually precipitated by a febrile illness, sepsis, pregnancy, surgery, or trauma.[7] Ganglionic acetylcholine receptor antibodies are present in a substantial percentage of patients with neuropathic POTS, indicating a possible autoimmune aetiology.[5]

This report describes the anaesthetic management of a parturient with post-viral POTS, associated with second-degree heart block, that had been diagnosed in pregnancy.

Case report

A 37-year-old primigravid woman with no relevant past medical history and an uncomplicated pregnancy presented to hospital at 29 weeks' gestation with a history of exertional shortness of breath, light-headedness and palpitations. A chest X-ray revealed left basal atelectasis and consolidation, and a full respiratory viral screen confirmed metapneumovirus. An electrocardiogram (ECG) revealed sinus tachycardia which was thought to be related to the viral pneumonia. She was treated conservatively but remained tachycardic with intermittent palpitations. A 24-h ECG recording showed that her heart rate varied between 65–168 beats/min, with episodes of gradual onset tachycardia or of 2:1 conduction with dropped beats, some of which was type-1 second-degree atrioventricular (AV) block. A cardiology diagnosis of post-viral tachycardia syndrome was made. In the absence of concerning symptoms, this permitted discharge home.

The woman represented at 34 weeks' gestation with worsening exertional dyspnoea and palpitations with pre-syncopal symptoms. These improved with rest and lying in the left lateral position. Limitation of her exercise capacity rendered her bed-bound. An echocardiogram and ventilation-perfusion scan were unremarkable, however lying and standing heart rate and blood pressure measurements revealed a change in heart rate from 90 beats/min to 170 beats/min, settling at 140 beats/min without a change in blood pressure and indicating a diagnosis of POTS.[3] Urinary metanephrines were within normal limits, thus excluding a phaeochromocytoma.

Medical therapy to attenuate the tachycardic response was considered inappropriate. During multidisciplinary discussions about mode of delivery, the severity of symptoms and the unusual finding of intermittent AV block caused concern that catecholamine release during labour could trigger an exaggerated and unpredictable tachycardic event, increasing the risk of emergency operative delivery in a haemodynamically unstable patient.

Elective caesarean delivery was performed at 38 +6 weeks. Direct arterial blood pressure monitoring was instituted prior to a combined spinal-epidural (CSE) technique after a 1 L crystalloid preload. The CSE was inserted with the woman in the left lateral position and a low-dose spinal component of 1.5 mL 0.5% hyperbaric bupivacaine and 0.3 mg diamorphine was used. At injection an intravenous phenylephrine

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infusion was commenced at 1600 μg/h, after which the patient was positioned supine with 15 degrees left lateral tilt. An abnormal cardiovascular response, with hypotension and sinus tachycardia, was seen rapidly and was treated by increasing the phenylephrine infusion to 2000 μg/h and with metaraminol boluses, as required. The blood pressure stabilised after two minutes and the heart rate remained at approximately 80 beats/min for the duration of the case. Cardiovascular stability was maintained as a sensory block to T4 and complete motor block of the legs was achieved using epidural 0.5% levobupivacaine 5 mL. After delivery of a live infant, intravenous oxytocin 5 U was administered over 20 min, followed by 10 U/h for four hours. Blood loss was 1400 mL in total due to uterine atony, but further uterotonics were not required.

After four hours the patient was asymptomatic and moving from the lying to sitting position elicited no tachycardic response. She remained in the obstetric high dependency unit for 24 hours, having mobilised by early evening. She remained symptom-free six months later.

Discussion

We believe this is the first case report describing post-viral POTS in which severe symptoms and concurrent second degree heart block developed during pregnancy. This condition affects predominantly women of childbearing age and is particularly relevant to the obstetric anaesthetist. Although there are case reports of POTS during pregnancy, the majority describe the course of pregnancy in women with POTS that existed prior to conception.[8–10]

The diagnosis of POTS is based on a complete history, examination, orthostatic vital signs, a 12-lead and a 24-h ECG, which together are sufficient to establish a diagnosis and initiate treatment in most cases.[3] A more extensive evaluation may include tilt-table testing and quantitative sudomotor autonomic reflex testing (QSART), however it is not recommended that these tests be performed routinely because there is no evidence that they improve the care of most patients.[3] Additionally, there is a lack of standardised data on clinical autonomic testing during pregnancy, making interpretation of the results difficult.[11]

The course of POTS during pregnancy is variable. The tachycardic response is accompanied by symptoms relating to cerebral hypoperfusion, autonomic over-reactivity, dysautonomia and sudomotor symptoms, and fainting or near-fainting in 60% of patients.[5] Symptoms may deteriorate during pregnancy due to the physiological changes that make autonomic nervous system responses to stimuli less effective.[9,10] In our patient there was a progressive worsening of symptoms. Glatter et al. described progressive worsening of symptoms beyond six months' gestation in two patients with severe POTS. This was attributed to a physiological peak increase in heart rate,[12] while other reports have described an improvement in symptoms in the later part of pregnancy because of fluid retention.[9,13]

Treatment of POTS includes non-pharmacological therapies such as compression stockings to augment peripheral vascular relaxation and adequate hydration to optimise cardiac preload. Pharmacological, therapies include the use of beta-blockers and alpha-$_1$ agonists such as midodrine, to increase vascular tone and suppress tachycardia.[8] The use of fludrocortisone, to increase blood volume by increasing sodium concentration, may be considered,[14] as can administration of cholinesterase inhibitors such as pyridostigmine.[15] Beta-blockers, the most commonly used drugs, were contraindicated in our patient due to the episodes of second-degree heart block. The cardiological opinion was that there was little evidence for the use of other drugs and that their benefit did not outweigh the risk in this patient. A small number of case reports have suggested consideration of immunoglobulin therapy but the evidence is limited.[16]

The mode of delivery for women with POTS is contentious and delivery mode depends on the symptomatology. Glatter et al.[12] reported two cases of severe POTS and recommended caesarean delivery to remove the cardiovascular stress of labour.[6] However, vaginal delivery has been found to be safe, in the absence of obstetric complications, with adequate epidural analgesia,[8–10,15,17] although these reports are confined to women with mild or well-controlled symptoms. In our patient elective caesarean delivery was performed based on concern that the severity of symptoms would render epidural analgesia insufficient to mitigate the tachycardic response during vaginal delivery. Phenylephrine is well established in treating the haemodynamic changes induced by regional anaesthesia in the obstetric patient[18] and is appropriate in POTS as it increases blood pressure by vasoconstriction rather than by increasing the heart rate.

This case describes the presentation and management of severe POTS presenting in pregnancy and the resolution of symptoms immediately following delivery. Limited evidence exists for treatment of patients developing POTS during pregnancy, particularly of those with severe symptomatology. Resolution of symptoms following delivery has not previously been described, although examples exist in which patient symptoms improved postnatally.[11,12,19] High-quality research is required to improve our knowledge of this condition and to form consensus guidance about its management during pregnancy.

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Kyphomelic dysplasia, Pierre Robin Sequence and pregnant

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ABSTRACT
We present the anaesthetic management of a parturient with kyphomelic dysplasia and Pierre Robin Sequence who underwent elective caesarean delivery. Potential anaesthetic issues and management strategies are discussed.

Keywords: Kyphomelic dysplasia; Dwarfism; Pierre Robin Sequence; Pregnancy

Introduction

Restricted growth, also referred to as dwarfism, is defined as an adult with a height of less than 148 cm.[1] It may occur in more than 400 medical conditions,[2] all of which can be broadly classified using the trunk-to-limb ratio into two main types: proportionate short stature or disproportionate short stature.[3]

Kyphomelic dysplasia (KD) is a rare prenatal skeletal disease that causes disproportionate short stature. It is characterised by short bowed long bones, irregular

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