CASE REPORT

# Postural orthostatic tachycardia syndrome complicating pregnancy: a case report with review of literature

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**Summary:** Postural orthostatic tachycardia syndrome (POTS) affects women of child-bearing age. There are little reported data on the outcomes of pregnancy in women with POTS. The most common mode of delivery reported in the literature is the caesarean section. Here we describe a woman with POTS who delivered vaginally without any complications and present a comprehensive review of the literature on pregnancy in POTS.

**Keywords:** cardiac, cardiovascular, high-risk pregnancy, maternal mortality

## INTRODUCTION

Postural orthostatic tachycardia syndrome (POTS) is an uncommon condition characterized by the development of orthostatic symptoms due to inability of the autonomic nervous system to efficiently handle the changes in blood volume and pressure when a person stands erect.[1] The cause is unknown and the symptoms include lightheadedness, weakness, visual changes, fatigue, palpitations, shortness of breath, syncope and rarely gastrointestinal disturbances that are aggravated by heat or exercise. The effect of POTS in pregnancy or vice versa is poorly understood. The course of POTS in pregnancy is variable with 80% of patients showing improvement and 60% becoming functionally normal.[2]

## CASE PRESENTATION

A 20-year-old primigravida attended our antenatal clinic in the first trimester with occasional episodes of palpitations and extreme fatigue. She was a known case of POTS, which was diagnosed one year prior when she had complaints of repeated syncopal attacks and palpitations. The diagnosis of POTS was made by the cardiologist based on her symptoms and by finding an exaggerated heart response up to 30 beats per minute without orthostatic hypotension on tilt table testing. Since then, she had been taking metoprolol 10 mg thrice daily. She had regular antenatal follow-up and remained asymptomatic throughout the pregnancy.

She was admitted at 39 weeks of gestation in early labour. She was asymptomatic, maintained a normal pulse rate throughout labour with adequate pain relief by epidural analgesia and delivered vaginally a male baby weighing 3.35 kg with a normal Apgar score. The postnatal period was uneventful and she was discharged on the third postnatal day.

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## DISCUSSION

The prevalence of POTS is unknown and is not possible to make an accurate diagnosis of POTS during pregnancy, since some of the symptoms of pregnancy may mimic POTS. All the published reports including our patient had a diagnosis of POTS prior to pregnancy. Criteria for diagnosis include an increase in heart rate $\geq$30 beats from supine to standing position, worsening of symptoms on standing and better in recumbent position, six months' duration of symptoms and a standing norepinephrine level of $\geq$600 pg/mL.[1] This tachycardic response is accompanied by symptoms related to cerebral hypoperfusion, autonomic overactivity, dysautonomia and sudomotor symptoms and fainting or near fainting has been reported in 60.5% of patients.[3] The various forms of POTS include neuropathic, hyperadrenergic, POTS with deconditioning and others.[4] Formal laboratory testing to exclude other causes of autonomic dysfunction which evaluates sudomotor, cardiovagal and adrenergic functions can be carried out. The severity of the syndrome is graded from I to IV based on symptoms, standing time and effects on activities of daily living.[4]

Our patient had grade I orthostatic intolerance. The differential diagnosis include phaeochromocytoma, hypovolaemia, inappropriate sinus tachycardia syndrome, autonomic neuropathies, medications and effects of prolonged bed rest.[1,5] A literature review of pregnancy in POTS is summarized in Table 1.

In pregnancy, due to physiological cardiovascular changes, symptoms of POTS might deteriorate because of inefficient autonomic nervous system.[6] The course of POTS during pregnancy is variable. In our patient there was no worsening of symptoms. Similarly, the symptoms remained unchanged in 84% and 88% of parous and nulliparous patients, respectively, in a study by Kimpinski *et al*.[2] However, Glatter *et al*.[7] showed progressive worsening beyond six months gestation in two of his patients with severe POTS which was attributed to physiological peak increase in heart rate which will worsen POTS. In contrast, some studies showed improvement in the

Table 1 **Postural orthostatic tachycardia syndrome (POTS) complicating pregnancy: previous cases reported in the literature**

| Reference | Number of cases | Maternal age (years) | Parity | Severity of POTS | Medications prior to pregnancy | Medications during pregnancy | Duration of POTS (years) | Course of POTS in pregnancy | Comorbidity | Antenatal complications | Mode of delivery | Labour analgesia | Neonatal outcomes | Postnatal improvement |
|---|---|---|---|---|---|---|---|---|---|---|---|---|---|---|
| Glatter *et al.*[7] | 2 | 26 | – | Severe | Midodrine | Midodrine | 3 | Deterioration >24 weeks | – | Hyperemesis | Elective caesarean section | Epidural | 3.2 kg (both) | Symptoms improved, drug dosage↓ |
| | | 24 | – | Severe | Midodrine and $\beta$-blocker | None | 6 | | | | | | | |
| Corbett *et al.*[9] | 1 | 30 | N | Hyperadrenergic form | $\beta$-Blocker | ↑ Dose of $\beta$-blocker | 3 | Worsened | Mitral valve prolapse with regurgitation | – | Emergency caesarean section | Epidural | 3.65 kg Apgar – 1 and 5, minute – 8, 9 | Improved |
| McEvoy *et al.*[10] | 1 | 18 | P | – | Fludrocortisones and $\beta$-blocker | Unchanged | – | Unchanged | – | Pregnancy-induced hypertension | Forceps | Epidural | Apgar – 1 and 5 minute – 6, 8 | – |
| Kodakkatil and Das[6] | 1 | 21 | N | Grade III POTS | None | None | 5 | Unchanged | – | Threatened preterm labour | Vaginal | Epidural | – | – |
| Kanjwal *et al.*[8] | 22 | 30 ± 7 | – | 21 – partial dysautonomic form; 1 – hyperdrenergic form | – | $\beta$-blocker (18%), midodrine (31%), SSRI's (31%), fludrocortisone (13%), Combination (40%), none (18%) | – | Unchanged – 3 (13%), improved –12 (55%), worsened – 7(31%) | Migraine 36%, joint hypermobility syndrome 9%, factor V deficiency 9% | Hyperemesis – 1 (4.5%), complete heart block – 1 (4.5%) | Vaginal – 18 (82%), caesarean – 4 (18%) | – | 1 – atrial septal defect, 1 – ventricular septal defect, 1 – ventricular septal defect | Stable – 15 (69%), worse – 7 (13%) |
| Jones and Ng[11] | 1 | 34 | N | – | – | – | 7 | Worsened | Ehlers–Danlos syndrome type III | – | Elective caesarean section | Epidural + spinal | Apgar score – 1and 5, minute – 9, 10 | Improved |
| Powless *et al.*[5] | 7 (9 pregnancies) | – | N and P | – | 4(6) – $\beta$-blockers, 1 – pyridostigmine and Fludrocortisones, 2 – not on drugs | 1(2) – drugs unchanged, 2(3) – ↑dose of $\beta$-blocker, 1 – addition of drugs, 2 – not on drugs | – | 4(5) – exacerbation | – | 1 – oligohydramnios, 1 – chronic placental abruption, 1 – PPROM*, preterm labour, 1 – gestational hypertension | Vaginal – 7, caesarean – 2 | 5 – epidural (vaginal delivery) | 3.15 kg (mean) | – |
| Kimpinski *et al.*[2] | 51 (116 pregnancies) | P – 33.6 ± 8.4, N –29.6 ± 10.1 | N and P | – | – | – | P – 3.7 ± 2.6, N – 2.1 ± 2.2 | Static P (51) 84% N (61) 88%<br>Progressive 8% 0<br>Improved 4% 6%<br>Relapse–remitting 2% 5%<br>Unknown 2% 0 | Migraine (P – 24%, N – 26%) | Miscarriage, placenta previa, placental abruption, malpresentation, peripartum hysterectomy | – | – | 2.995 kg ± 0.834 kg, Apgar score 1 minute – 8.2 ± 0.8, 5 minutes – 9.2 ± 0.4 | Improved |
| Kimpinski *et al.*[12] | 1 | 26 | P | – | – | – | Diagnosed 3 weeks postpartum | | Peripartum cardiomyopathy | – | Elective caesarean section | – | – | – |

P, parous; N, nulliparous; SSRI's, selective serotonin reuptake inhibitors; PPROM, preterm premature rupture of membranes

later part of pregnancy due to increased fluid retention which occurs during this period.[8]

In a study by Kanjwal *et al*.[8] three of 22 offspring of women with POTS had congenital abnormalities including atrial septal defect, ventricular septal defect and Down's syndrome. Other studies have shown good neonatal outcomes with no stillbirths or any congenital abnormalities.[1,5] Our patient did not have any fetal complications.

The treatment of POTS involves volume expansion with high salt and fluid intake. Additional pharmacological therapies include fludrocortisone, midrodrine or acetylcholineterase inhibitors. Beta blockers are used to treat adrenergic symptoms.[5] Glatter *et al*.[7] was the first to describe the use of midodrine, a category C drug, in pregnancy in his patients. A study by Powless *et al*. showed that the patients who did not require treatment for POTS prior to conception remained so with a less likely chance of exacerbation. Among those women who had exacerbation of their symptoms in pregnancy, a majority needed an increased dosage of pre-existing medications although very few required the addition of new drugs.[5] Our patient was on metoprolol on the same dosage prior to as well as throughout pregnancy. Exercise training and re-conditioning is emerging as a very important strategy. In a study by Glatter *et al*.[7] lifting the baby and caring for the infant forced women to develop their upper body strength that attributed to the improvement in symptoms as well as a positive psychological outlook at six months postpartum. Treatments must be carefully tested due to medication sensitivity often associated with POTS patients, and each patient will respond to different therapies in different ways. Even though the first line of treatment is volume expansion, our patient showed a good response to beta blockers, which block the effects of epinephrine and norepinephrine released by the autonomic nervous system, as compared with volume expansion.

The major controversy lies in the mode of delivery in POTS with pregnancy. Glatter *et al*.[7] was the first to report pregnancy in POTS patients and as the patients worsened after six months' pregnancy, elective caesarean section was performed to attenuate the stress of labour and avoid consequent triggering of a tachycardic response. But, subsequently published studies have shown that vaginal delivery with adequate labour analgesia is safe in these patients and caesarean section should be reserved for obstetric indications.[5,6,8] In a study by Powless *et al*.[5] two patients delivered vaginally without epidural anaesthesia without any complications. Our patient also delivered vaginally with adequate epidural analgesia without any complications.

The largest study published to date on the effect of pregnancy on POTS compared the clinical presentation, autonomic dysfunction and pregnancy outcomes in parous and nulliparous women with POTS and concluded that the long-term impact of pregnancy on POTS does not appear to be clinically important. However, there does appear to be a trend towards improvement in the short-term postpartum period. Adverse pregnancy events were similar to those seen in the general public and do not present a barrier to women with POTS who want to have children.[2]

## CONCLUSION

From the above-published data, POTS does not seem to pose an increased risk for women during pregnancy and child birth. They can safely undergo vaginal delivery with a successful obstetric and neonatal outcome, caesarean section being reserved for obstetrical indications. A multidisciplinary approach is required to avoid potential maternal or fetal injury resulting from syncope in these patients.

### DECLARATIONS

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**Contributorship:** HS had the idea for the study. Dr Shyjus Nair collected the data. HS and NP analysed the data and NP wrote the first version of the paper to which all authors contributed. All authors had full access to the data and contributed to interpretation of data and approved the final version.
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