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A DELICATE BALANCE – NAVIGATING NEURAXIAL ANAESTHESIA FOR CAESAREAN DELIVERY IN A PATIENT WITH MARFAN SYNDROME – CASE REPORT

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ABSTRACT Pregnancy in women with Marfan syndrome presents substantial anaesthetic challenges due to the risk of aortic dilatation, dissection, and haemodynamic instability. Anaesthetic management must prioritise meticulous control of heart rate and blood pressure to minimise stress on the aorta. We report the perioperative anaesthetic management of a 28-year-old primigravida at 34+1 weeks' gestation with Marfan syndrome and a history of valve-sparing aortic root replacement (David procedure), scheduled for an elective lower-segment caesarean section. Following comprehensive pre-anaesthetic evaluation and multidisciplinary planning, spinal anaesthesia was administered using hyperbaric ropivacaine with fentanyl. Stable intraoperative haemodynamics were maintained without episodes of hypotension or hypertension. Both maternal and neonatal outcomes were favourable. This case highlights the importance of individualised anaesthetic planning and vigilant haemodynamic control in parturient with complex cardiovascular pathology.

KEYWORDS :

INTRODUCTION

Marfan syndrome (MFS) is an autosomal dominant connective tissue disorder caused by mutations in the *FBN1* gene encoding fibrillin-1.[1] Cardiovascular involvement, particularly progressive aortic root dilatation and dissection, represents the leading cause of morbidity and mortality. Pregnancy further exacerbates cardiovascular risk due to increased blood volume, cardiac output, and hormonal effects on connective tissue integrity. Current cardiology and obstetric guidelines therefore emphasise pre-pregnancy counselling, serial echocardiographic surveillance of the aortic root, and strict blood pressure control throughout gestation. [2,3]

From an anaesthetic perspective, caesarean delivery in parturient with MFS requires strategies that avoid abrupt changes in preload, afterload, and heart rate. Both general and regional anaesthesia have been described; however, regional techniques may offer advantages by attenuating the stress response, provided hypotension is avoided. Caesarean delivery may be selected for MFS patients, particularly if the aortic root diameter enlarges beyond 40 mm. [4] We describe the anaesthetic management of an elective caesarean section in a patient with Marfan syndrome who had previously undergone valve-sparing aortic root replacement.

CASE REPORT

A 28-year-old primigravida at 34+1 weeks' gestation was scheduled for an elective lower-segment caesarean section. She had a known diagnosis of Marfan syndrome and a significant cardiac history. In 2022, she presented with acute chest pain, dyspnoea, and syncope and was found to have aortic sinus dilatation measuring 51 mm. She subsequently underwent valve-sparing aortic root replacement (David procedure) with an uneventful postoperative recovery.

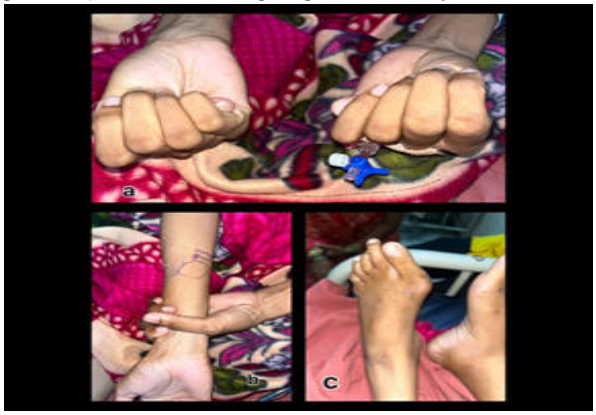

Figure 1 (a) Thumb Sign (b) Hand and wrist sign(c) valgus of foot

The patient had a strong family history of Marfan syndrome with adverse cardiovascular outcomes. During the current pregnancy, she remained hemodynamically stable. She was diagnosed with gestational diabetes mellitus at 22 weeks, managed with metformin and dietary modification. She was receiving aspirin (150 mg twice daily), which was discontinued 48 hours prior to surgery.

On examination, the patient was tall and thin (height 168 cm, weight 48 kg) with classical marfanoid features including arachnodactyly, pectus carinatum, thumb sign (figure 1a) , hand and wrist sign (figure 1b) , increased arm-span-to-height ratio, hindfoot valgus (figure 1c) , scoliosis (figure 2a) and malar hypoplasia (figure 2b). Airway examination was unremarkable.

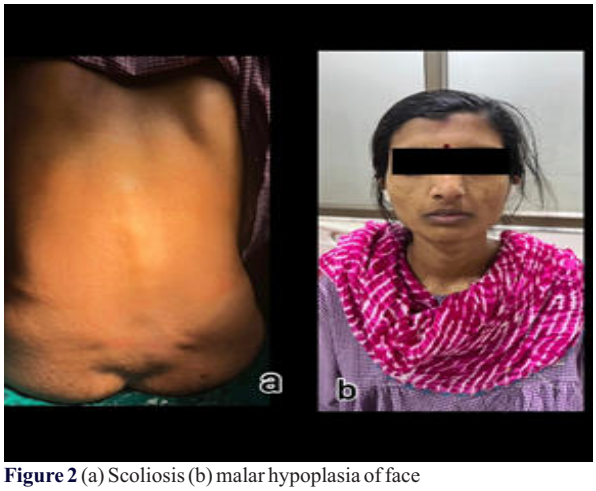

Figure 2 (a) Scoliosis (b) malar hypoplasia of face

Cardiovascular evaluation revealed normal heart sounds and sinus rhythm. ECG showed T-wave inversions in leads V2–V6. Transthoracic echocardiography demonstrated preserved left ventricular function (ejection fraction 55–60%), mild aortic regurgitation, and normal biventricular function. There were no clinical signs of heart failure. Foetal echocardiography was normal.

Cardiology consultation emphasised strict avoidance of perioperative hypertension and tachycardia to prevent stress on the aortic graft. Laboratory investigations were within normal limits. After multidisciplinary discussion, regional anaesthesia was preferred with readiness for immediate conversion to general anaesthesia if required. The primary anaesthetic goals were: Maintenance of stable heart rate and blood pressure, avoidance of sudden increases in afterload, prevention of hypotension following neuraxial block and adequate

analgesia to minimise sympathetic stimulation.

A fully equipped difficult airway trolley, emergency drugs, and crash trolley were kept ready. After confirming fasting status and obtaining informed consent, the patient was transferred to the operating theatre. Standard ASA monitors were applied, and baseline vital signs were recorded.

Spinal anaesthesia was administered in the sitting position using 2 mL of 0.75% hyperbaric ropivacaine with fentanyl 10 µg. Ropivacaine was chosen due to its favourable cardiovascular profile and reduced propensity for hypotension. The patient was positioned with a slight Trendelenburg tilt, achieving a sensory block up to T5.

A controlled co-loading strategy using Ringer's lactate was employed to maintain preload while avoiding fluid overload. Haemodynamics remained stable throughout the procedure, with an average blood pressure of 110/70 mmHg and a heart rate of approximately 78 beats/min. No vasopressors were required.

Oxytocin was administered as a slow intravenous infusion to minimise tachycardia and hypotension. Blood loss was approximately 550 mL, and urine output was adequate.

The neonate was delivered uneventfully and transferred to the Neonatal ICU (NICU) for observation on continuous positive airway pressure (CPAP).

Postoperative analgesia was supplemented with a bilateral transversus abdominis plane (TAP) block using ropivacaine to reduce opioid requirements and prevent sympathetic surges due to pain. The patient was monitored in a high-dependency intensive care unit for 24 hours with continuous haemodynamic surveillance before transfer to the ward.

DISCUSSION

Marfan syndrome affects connective tissues, causing ocular and bone defects, but nearly all adults develop cardiovascular abnormalities, which cause most early deaths.[5] Anaesthetic management of parturients with Marfan syndrome is centred on preventing aortic complications by minimising haemodynamic fluctuations. Aortic dissection is the main cause of death in patients with Marfan syndrome, approximately at 46%.[6] Dilatation of the aorta is found in approximately 60% to 80% of adult patients.[7] Even after aortic root repair, patients remain susceptible to further aortic pathology, particularly in the peripartum period. Early management of fatal changes like prevention of aortic dissection and progression of aorta dilatation can markedly increase life expectancy and quality. Even after surgical repair of the aorta, individuals with Marfan syndrome remain vulnerable to further aortic problems, such as new aneurysms, dissections, or extension of existing disease.[8] These cardiac complications are significantly increased during pregnancy due to increased circulatory volume and cardiac output. While the patient had undergone prior aortic root replacement, the cardiology team emphasized that avoiding post-delivery hypertension was critical to prevent excessive stress on the graft. Co-loading the patient with Ringer Lactate at 80ml/hr at time of induction followed by during the course of the surgery helped to maintain adequate circulating volume
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Regional anaesthesia offers several advantages, including attenuation of the stress response and avoidance of laryngoscopy-induced hypertension associated with general anaesthesia. However, neuraxial techniques carry the risk of sympathetic blockade–induced hypotension, which can compromise uteroplacental perfusion and provoke reflex tachycardia. In this case, careful drug selection, gradual block establishment, controlled fluid administration, and vigilant monitoring allowed safe conduct of spinal anaesthesia.

The use of ropivacaine, combined with fentanyl, provided adequate anaesthesia with minimal cardiovascular depression. Postoperative regional analgesia further contributed to haemodynamic stability.

CONCLUSION

This case demonstrates that elective caesarean section under spinal anaesthesia can be safely performed in parturients with Marfan syndrome and prior aortic root surgery when meticulous anaesthetic planning and vigilant haemodynamic control are employed. A multidisciplinary approach, judicious choice of anaesthetic technique and drugs, and close perioperative monitoring are essential to ensure optimal maternal and foetal outcomes.

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