IMJ
Official Journal of the Irish Medical Organisation

Ir Med J; April 2025; Vol 118; No. 4; P62
April 23rd 2025

Ehlers-Danlos Syndrome and Anaesthesia: Overcoming Challenges in Caesarean Section with Spinal Anaesthesia

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Abstract

Presentation

We present a case of a 26 year old primigravida with hypermobility type Ehler Danlos Syndrome at 37 weeks presenting for a Caesarean section.

Diagnosis

Diagnosed case of Ehler Danlos Syndrome Hypermobility type by the rheumatologist. All blood tests were within normal limits except for a raised bleeding time.

Treatment

Spinal Anaesthesia was given for Caesarean section and a healthy infant boy was delivered successfully without complications.

Discussion

Spinal anaesthesia can be a safe and effective option for patients with the hypermobility subtype of EDS giving effective pain relief while minimizing the risks of bleeing, trauma and complications related to fragilie connective tissues

Background

Ehlers-Danlos Syndrome (EDS) is a heritable connective tissue disorder characterized by genetic mutations affecting collagen synthesis. Beighton et al. described six major types of EDS, with the hypermobility type (EDS-HT) being the most common. Patients with EDS-HT typically experience joint hypermobility, frequent joint dislocations, and chronic pain and may have a family history of the disorder.[1] EDS appears to affect connective tissues in the trachea, larynx, and skin, complicating anaesthetic management. [2]

Airway management can be complicated in patients with EDS due to the collapse of fibro-elastic tissues, and they may also exhibit resistance to local anaesthetics ; mechanisms unknown.[3, 4] Despite the prevalence of EDS (approximately 1 in 5000 people), there are no standard guidelines for anaesthetic management [5]. While some studies caution against neuraxial anaesthesia due to resistance concerns, successful cases have been reported. [6, 7, 8]

Case Report

A 26-year-old woman (G1P0) with a history of EDS and gestational diabetes mellitus presented for an elective Caesarean section at 37 weeks of gestation. Her medical history included childhood asthma, treated migraines, and prior surgeries, including appendectomy and tonsillectomy. Her hypermobility syndrome had been evaluated, and she reported multiple joint dislocations, which she was able to reduce herself. She had no family history of EDS.

Her blood tests revealed a normal coagulation profile with only an increased bleeding time. Following a detailed discussion of the risks associated with neuraxial versus general anaesthesia, the patient consented to spinal anaesthesia. The risks of resistance to local anaesthetics and the potential for conversion to general anaesthesia were explained. Preparations for difficult airway management, including cervical spine hypermobility, were made.

The patient was classified as ASA II, with a Mallampati score of II and a normal neck range of motion. After standard monitoring and preparation, a 25-gauge Quincke spinal needle was inserted at the L3/L4 interspace. The patient received 10 mg of 0.5% heavy bupivacaine, along with 100 µg of morphine and 20 µg of fentanyl. A bilateral sensory block to T4 was confirmed, and the patient was positioned supine with a left lateral tilt. Intraoperatively, phenylephrine infusion was initiated to manage transient hypotension.

A healthy male infant was delivered without complications. Due to the risk of postpartum haemorrhage, tranexamic acid (1 g) was administered after the baby's delivery. Intraoperative blood loss was estimated at 600 mL. Diclofenac 100 mg and paracetamol 1 g administered rectally. The patient had uneventful recovery was discharged on postoperative day three.

Discussion

Anaesthetic management of patients with Ehlers-Danlos Syndrome hypermobility subtype (EDS-HT), presents unique challenges due to tissue fragility, airway management difficulties, and potential resistance to local anaesthetics. General anaesthesia poses risks such as tracheal collapse and cervical instability, while spinal anaesthesia can provide a safer option in obstetric cases which are a potential difficult airway [7].

Despite concerns about local anaesthetic resistance in EDS patients (mechanism unknown), successful spinal anaesthesia has been documented in this report.[9] It is essential to thoroughly evaluate each patient's history and clinical presentation to balance the risks and benefits of neuraxial anaesthesia. Literature reports that patients with EDS Vascular Subtype , epidural or spinal anesthesia can potentially cause hematoma formation or bleeding by needle

insertion and catheterization [2]. In this case, spinal anaesthesia was successfully administered without complications, highlighting its feasibility in patients with EDS-HT.

The management of patients with EDS-HT also requires preparedness for complications, including postpartum haemorrhage. In this case, proactive administration of tranexamic acid and oxytocin helped manage the risk of excessive bleeding. The patient's smooth recovery supports the viability of spinal anaesthesia in carefully selected EDS patients undergoing Caesarean section.

This case report demonstrates that spinal anaesthesia can be a safe and effective option for patients with the hypermobility subtype of EDS giving effective pain relief while minimizing the risks of bleeding, trauma and complications related to fragile connective tissues. Given the lack of standardized guidelines, further research and case reports are crucial to our understanding of the best practices for anaesthetic management in EDS patients.

Declarations of Conflicts of Interest:
None declared.

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