

Anaesthesia for the obstetric patient with Ehlers Danlos Syndrome.

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Background

EDS is an umbrella term encompassing thirteen subtypes of connective tissue diseases, each with varying severity and associated implications [1]. This syndrome presents many challenges to the obstetric anaesthetist. The condition has many perioperative considerations due to the wide range of physical manifestations including musculoskeletal, cardiac, vascular, haematological, joint and skin involvement. There have also been cases of local anaesthetic resistance reported in EDS. The exact cause is unclear.

Epidemiology

For many years EDS has been considered to be a rare disease affecting 1 in 5,000 individuals. A recent UK based study has seen an increased rate of diagnosis over the last 27 years, estimating the prevalence of EDS to be 1 in 500 individuals and 70% more common in women [2].

Patient Description

We present a case of a 35-year-old female (G5 P4) 30 weeks gestation who required an out of hours category 2 emergency lower segment caesarean section (LSCS). This was her second emergency LSCS who previously required conversion to general anaesthesia (GA) due to unsuccessful spinal anaesthesia. Past medical history included Ehlers Danlos Syndrome type 3 (EDS) and neurofibromatosis type 1 (NF1).

Case Presentation

This patient was admitted to Cork University Maternity Hospital High Dependency Unit with threatened pre-term labour and evolving scar tenderness from a previous LSCS. An emergency caesarean section was planned given her presentation in addition to the potential for uterine rupture in patients with EDS. During previous pregnancies there had been multiple epidurals sited with no analgesic effect and attempts at spinal anaesthesia which were unsuccessful requiring conversion to general anaesthesia (GA). Given the urgency of the case the decision was made not to attempt a combined spinal epidural (CSE) and ultrasound was not used as the spinous processes were clearly visible and easily palpable. Spinal was straightforward; first pass, clear cerebrospinal fluid (CSF), good flow and aspirating freely with barbotage performed at the beginning and end of injection. Intrathecal injection of 11.5mg of hyperbaric bupivacaine, 15mcg fentanyl and 150mcg morphine was performed.

Fifteen minutes post spinal there was no motor block (Bromage score 1) or sensory block. At this stage decision taken to convert to general anaesthetic with protective measures to prevent hyperextension of the cervical spine given the propensity of EDS patients to joint laxity and dislocations. Intubation and GA were uneventful and LSCS proceeded uneventfully with minimal blood loss and a healthy infant was born weighing 2.5kg. Post-operative pain was a significant issue for this patient which was ultimately managed with multimodal analgesia paracetamol, non- steroidal anti-inflammatory drugs, ketamine, clonidine and Morphine Patient Controlled Analgesia. Her anaesthetic record also demonstrated significant pain after previous GA LSCS following an unsuccessful neuraxial anaesthesia where the same dose of intrathecal morphine and fentanyl was injected. During a retrospective chart review of this patient it was found that an MRI was conducted in 2016, investigating potential causes of unsuccessful neuraxial anaesthesia. It reported dural ectasia with widening of the spinal canal at S1 relative to L4 and bilateral S1 perineural cysts. This report was not available on the CUMH patient information system.

Discussion

At present there are no guidelines published on anaesthetic management of the parturient with EDS [1]. Recommendations have been made to avoid neuraxial blockade in patients with vascular EDS (type 4), however for other subtypes anaesthetic management options include neuraxial anaesthesia, the decision to do so made upon the patient's disease severity and associated co morbidities [3]. One case report has suggested the use of continuous spinal anaesthesia in a patient with a history of failed single dose spinals to avoid a GA [1].

This case demonstrates the importance of anaesthetic involvement in the antenatal period in order to establish the subtype, severity of EDS, examine extent of systemic involvement, and any previous anaesthetic complications. This would also offer an opportunity to source appropriate imaging like in this case, given the central role of neuraxial anaesthesia within obstetric anaesthetic practice. EDS has a known association with dural ectasia, a dilatation of the lumbrosacral sac, which can complicate neuraxial anaesthesia [3].



This case offers another example of failed neuraxial anaesthesia in EDS. It supports the suggestion that a registry of perioperative complications for patients with EDS would undoubtedly improve patient safety in the obstetric population [1].

References

1. Wloch K, Simpson M, Gowrie-Mohan S. Local anaesthetic resistance in a patient with Ehlers-Danlos Syndrome undergoing caesarean section with continous spinal anaesthesia. Anaesthesia Reports. 2020; 8: 56-58.
2. Demmler JC, Atkinson MD, Reinhold EJ, et al. Diagnosed prevalence of Ehlers-Danlos syndrome and hypermobility spectrum disorder in Wales, UK: a national electronic cohort study and case–control comparison. BMJ Open 2019;9:e031365.
3. Wiesmann T, Castori M, Malfait F, Hinnerk W. Recommendations for anaesthesia and perioperative management with Ehlers-Danlos syndrome (s). Orphanet Journel of Rare Diseases. 2014; 9: 109.