

A Case of Successful Elective Caesarian Birth in a 23 year old Primigravadia with vascular Ehlers Danlos

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Documented complications of vEDS in pregnancy(1):

Uterine rupture (2.6%)

Increased risk of 3rd and 4th degree vaginal and cervical tears (20%)

Arterial rupture (5.4%)

Preterm delivery (19%)

Maternal Death (6.5%)

Surgical complications (2.6%)

Background:

Vascular Ehlers-Danlos syndrome (vEDS) is an autosomal dominant condition caused by a mutation in gene COL3A1, which is responsible for encoding type III procollagen.(1)

Pregnant women with vEDS are at high risk of serious complications such as arterial dissection/rupture, uterine rupture, haemorrhage and wound dehiscence. Recent studies have found maternal mortality to be 5.3%. Severe morbidity was found in 14.5% of deliveries (including arterial dissection/rupture and uterine rupture).(1)

There are currently no guidelines for anaesthetic management for delivery in women with vEDS. However, published recommendations suggest early delivery via caesarian section with or without the use of desmopressin.(2)

Here we describe the case of a 23 year old primigravadia with vascular Ehlers Danlos who had a successful delivery via elective caesarian section at 34+1/40.

Pre-operative management

Expert opinion was sought from a specialist centre and a combined online MDT meeting was held with representatives from obstetrics, neonatology, anaesthesia, cardiac anaesthesia, vascular surgery, intensive care medicine and haematology.

Departmental awareness was of upmost importance with outcomes from the MDT being circulated throughout the department.

Perioperative Management:

Regional vs General Anaesthetic:

A combined GA and regional technique was used. An awake spinal anaesthetic was performed using 2mL hyperbaric bupivacaine and 300mcg diamorphine. General anaesthesia was maintained with sevoflurane and remifentanil to minimise blood pressure fluctuation.

Location:

The caesarian section was performed in a hybrid vascular/interventional radiology theatre with both surgical teams present in case of complications.

Monitoring:

Standard AAGBI monitoring plus arterial cannulation was chosen.

Outcome:

Estimated blood loss was 800mL.

No other significant adverse events were noted.

A live infant was delivered initially requiring CPAP in neonatal in intensive care.

The mother was initially observed in the labour ward high dependency unit but discharged to the antenatal ward the following day.

Key Learning Points:

- *This case highlights the benefits of tailoring the anaesthetic technique according to patient preference and beliefs.*

- *This case highlights the importance of multiple specialties in the care of women with rare conditions.*



References:

- 1.Murray ML, Pepin M, Peterson S, Byers PH. Pregnancy-related deaths and complications in women with vascular Ehlers-Danlos syndrome. Genet Med. 2014 Dec;16(12):874-80. doi: 10.1038/gim.2014.53. Epub 2014 Jun 12. PMID: 24922461.
- 2.Rochelson B, Caruso R, Davenport D, Kaelber A. The use of prophylactic desmopressing (DDAVP) in labor to prevent haemorrhage in a patient with Ehlers Danlos Syndrome. NY State J Med 1991;91:268-269