

# Walking on a tight rope: Anaesthetic Management of a Patient with Vascular Ehlers Danlos Syndrome for an Elective Caesarean Section

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Vascular Ehlers-Danlos Syndrome (EDS) is a rare autosomal dominant genetic disorder caused by structural abnormalities in type-III collagen.<sup>1</sup>  
Pregnancy is not advised due to significant maternal risks and mortality<sup>2</sup>  
These patients present unique challenges due to their heightened risk of vascular fragility, bleeding, hyperextensibility and uterine rupture.

## CASE REPORT

20 year old primiparous woman diagnosed with vEDS prenatally  
History: Excessive bruising, prolonged bleeding from minor wounds.  
Transferred Care: Referred at 19 weeks’ gestation

## ANAESTHETIC PLAN

General anaesthesia based on patient preference and multidisciplinary counselling.

## MANAGEMENT

Standard AAGBI with invasive blood pressure monitoring\*  
GA technique tailored to minimise airway trauma and haemodynamic instability.  
Blood conservation strategies: Prophylactic Tranexamic Acid and Desmopressin along with Intraoperative cell salvage.

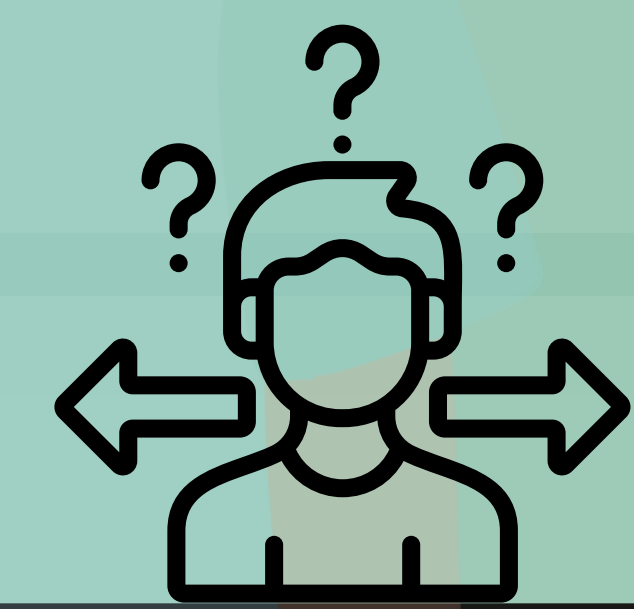
## OUTCOME



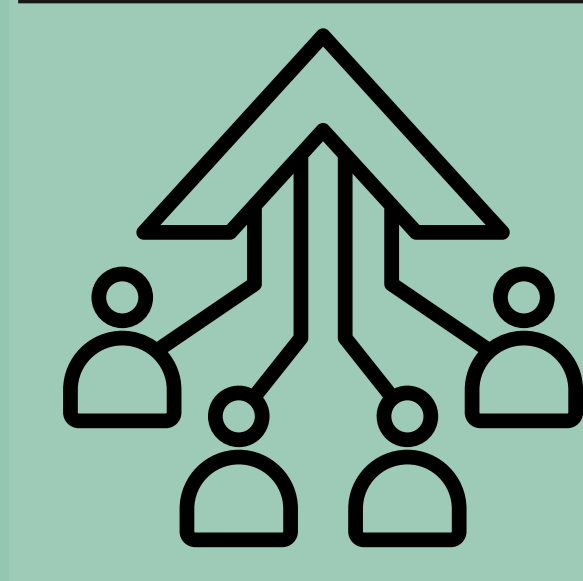
- Healthy female infant delivered at 34 weeks
- Mother transferred to maternal high dependency unit for observation

## DISCUSSION

Anaesthetic management remains controversial



| Consideration          | General Anaesthesia                       | Regional Anaesthesia                |
|------------------------|-------------------------------------------|-------------------------------------|
| Airway Management      | Trauma – due to tissue fragility          | Avoids airway instrumentation       |
| Bleeding Risk          | Increased blood loss, vascular dissection | Neuraxial Haematoma, dura fragility |
| Haemodynamic Stability | Controlled Blood pressure management      | Neuraxial hypotension               |



- A collaborative approach - careful risk benefit analysis
- Individualised Decision-making
- Multidisciplinary Team Involvement
- Meticulously planning

**References**  
1. Malfait F, Francomano C, Byers P, et al. The 2017 international classification of the Ehlers-Danlos syndromes. Am J Med Genet C Semin Med Genet. 2017.  
2. Pepin M, Schwarze U, Superti-Furga A, Byers PH. Clinical and genetic features of Ehlers-Danlos syndrome type IV. N Engl J Med. 2000.