

## BACKGROUND

- Vascular Ehlers Danlos Syndrome (vEDS), aka EDS type IV
  - Caused by a mutation in *COL3A1* gene, resulting in abnormal type III collagen
  - Symptoms: acrogeria facial features, fragile blood vessels, translucent skin, and easy bruising
  - Complications: arterial dissection, colonic rupture, and uterine rupture
- Perioperative management focus on
  - Hemodynamic stability - vascular fragility
  - Positioning - joint hypermobility and skin laxity/fragility
  - Pain control – minimize opioids (risk of colonic rupture from constipation) and hemodynamic instability
- Epidural anesthesia reduces overall opioid consumption perioperatively with avoidance of bowel dysmotility/constipation
- Epidural placement in patients with vEDS is controversial due to fragility of tissue and dura
  - Limited case reports and studies regarding epidural use in patients with vEDS

## CASE

A 16-year old girl with PMH of prematurity presented with escalating abdominal pain, fever, and vomiting for three days

- Tachycardia with localized peritonitis
- CT scan: sigmoid colon abnormality

Operation: exploratory laparotomy

- Fragility of mesentery and peritoneum noted
- Extensive incision from sternum to pubis
- Colon left in discontinuity with stoma

Anesthesia: GETA, with low thoracic epidural

- Uneventful placement at T7/8 with 1 attempt



Figures 1 & 2: Translucent skin and branching blood vessels seen across patient's extremities and chest

## CASE CONTINUED

- Patient continued to recover well post-op without complications
- Genetic testing revealed *COL3A1* mutation
- Later, presented as elective procedure to the OR for bowel anastomosis and colostomy takedown
  - Surgeon requests epidural due to the great recovery from initial surgery and positive patient experience

## PROCEDURE: Thoracic Epidural

### Pros

- Reduced opioid usage and decreased risk of constipation
- Early ambulation with superior pain control
- Previous positive experience with epidural
- Easy placement of thoracic epidural in past

### Cons

- Reports of inadvertent dural puncture
- Increasing risk of bleeding due to vascular fragility
- Case reports of local anesthetic resistance and overall lack of investigation

## DISCUSSION

- Epidurals have been shown to provide effective post-operative pain control.
- The advantages include: reduced opioid consumption as well as associated side effects such as nausea, vomiting, constipation, bowel dysmotility
- vEDS is not an absolute contraindication for epidural placement
- Increased risk is due to risk of tissue perforation nerve, injury, bleeding, and inadvertent dural puncture
- There is documented resistance to local anesthetics in patients
- Each patient should be assessed with respect to clinical status, anatomy, and potential risks and benefits when devising a perioperative anesthetic plan
- Further research is needed to assess the safety and efficacy of epidural use in patients with vEDS

## REFERENCES

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