Anesthetic Management of a Parturient with Ebstein Anomaly and Tethered Cord Syndrome: A Case Report

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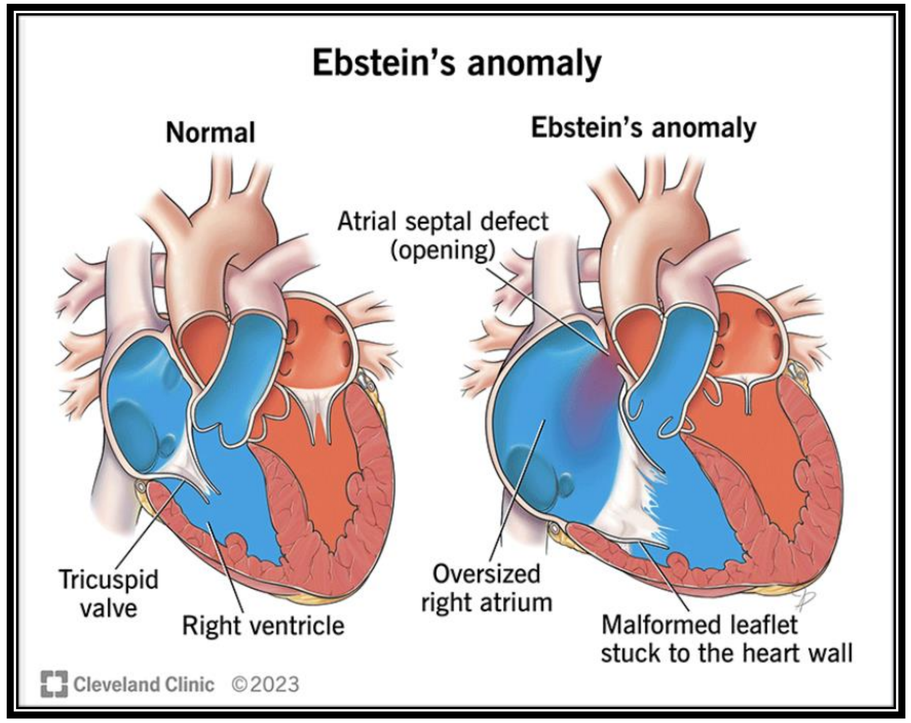
BACKGROUND

- **Ebstein anomaly** – a rare congenital malformation resulting in apical displacement of the *tricuspid valve*
  - Results in a poorly contractile right ventricle, enlarged right atrium, and tricuspid regurgitation
  - Can lead to right heart failure and cardiac arrhythmias (e.g. WPW syndrome)

- **Tethered cord syndrome** – abnormal tissue attachment to spinal cord
  - Often associated with spina bifida



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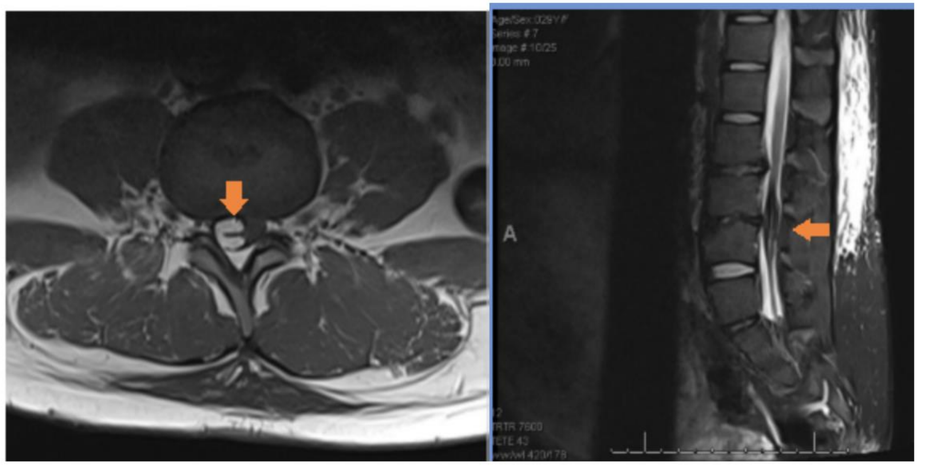
SOAP Society for Obstetric Anesthesia and Perinatology

CASE

30 YO G1P0000 @ 23w1d presented with PPROM and acute heart failure exacerbation

PMH: Ebstein anomaly, dextrocardia, WPW syndrome, spina bifida occulta with tethered cord syndrome, hepatic cirrhosis, bicornuate uterus

PSH: 3 open-heart surgeries and tricuspid valve replacement x2, dual-chamber pacemaker/ICD placement (DDI mode)



Patient's Lumbar MRI showing intradural lipoma (orange arrow) which is where the tethered cord terminates.

Admission

Treated for HF exacerbation with diuresis

Multidisciplinary meeting

Workup

Fetal ultrasound showed breech presentation

TTE revealed severely dilated RA, dilated IVC with hepatic vein flow reversal suggesting elevated right atrial pressures. Normal LV size and function. Bubble study (-)

MRI lumbar spine showed a tethered cord terminating in an intradural lipoma at L4-L5, with thickened filum extending inferiorly

Hospital Day 69 (32w6d)

C-section for concern for Triple I

Pre-induction arterial line and 2 large-bore PIVs

Sterile magnet placed over abdominal pacemaker

Arrhythmias not requiring treatment

Vasopressor with phenylephrine

Bilateral TAP blocks placed

POD1

Continued diuresis

Minimal pain requirements

POD6

Discharged home on oral diuretics

DISCUSSION

- Physiologic effects of pregnancy on cardiac function
- General vs neuraxial anesthesia in high-risk cardiac patients
- Neuraxial in parturient with spinal dysraphism
  - Imaging-guided epidural
- Importance of multidisciplinary preoperative planning
- Appropriate monitors and contingency plans
  - E.g. availability of procainamide

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