Ebstein Anomaly in the Parturient: Management Considerations

Robert Guidangen MD, Kristin Wakin MD, Joshua Abels MD
Department of Anesthesiology, University of Nebraska Medical Center

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Congenital cardiac malformation

-associations with chromosomal defects and drug exposures

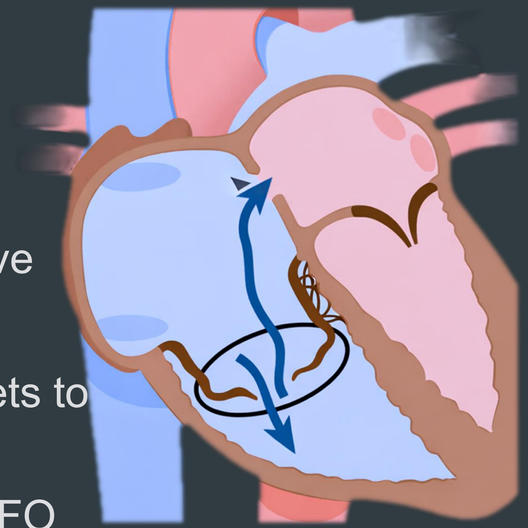
Ebstein anomaly (EA) features:

-apical displacement of the tricuspid valve

-atrialization of the right ventricle

-adherence of septal and posterior leaflets to myocardium

-often co-occurring secundum ASD or PFO



Clinical Presentation

-Signs and symptoms: tachydysrhythmias, heart failure, cyanosis, exertional dyspnea

-Prevalence of <1% of congenital heart defects and incidence of 0.2 to 0.7 per 10,000 live births[1]

-Disease severity varies from newborns who do not survive beyond infancy to asymptomatic adults diagnosed incidentally in the 6th and 7th decade of life.

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Ref.:
1. Singh DP et al (2024, Feb 28). Ebstein Anomaly and Malformation. In *StatPearls* Publishing.

Case

23yo G1P0 with intra-uterine pregnancy at 36 weeks and 6 days of gestation with EA history.

Previously repaired EA with tricuspid valve replacement and ASD closure.

IOL indicated by worsening dyspnea & lower extremity edema in the setting of worsened tricuspid stenosis and severe right ventricular dysfunction and dilation.

Anesthetic Labor Management:

- Two large bore peripheral IV catheters
- Judicious crystalloid management
- Radial arterial line placed before placement of labor epidural.
- Slow titrated continuous epidural infusion (vice PIEB) of fentanyl-bupivacaine 2mcg/mL-0.0625% started at 8mL/h. PCEA at 4mL every 10 minutes.
- Inadequate coverage can raise PCEA to 8mL

Cesarean Plan:

- 2% lidocaine-bicarbonate-epinephrine mixture hand bolused (5mL each) with close hemodynamic monitoring to activate labor epidural
- Norepinephrine as initial pressor of choice
- GA induction: etomidate as induction agent of choice and co-administered with fentanyl

Takeaway

Considerations

- Baseline cardiac function
- Co-occurring valvulopathy
- Effects of labor induction agents – *oxytocin preferred, avoidance of ergotamines*[1]
- Early anesthesiologist involvement
- Preferred mode of delivery – *vaginal delivery preferred*[1]
- Anesthetic Modality– *labor epidural early to allow for titration and adjust for hemodynamics, consider fascial plane blocks*
- Hemodynamic monitoring – *arterial line, consider CVP monitoring for C-section, telemetry at all times*
- General anesthesia – *euvolemia, normal sinus rhythm, minimize excessive tachycardia, minimize increases in PVR, consideration of ECMO candidacy*

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Ref: 1. ESC Guidelines on the management of cardiovascular diseases during pregnancy: the Task Force on the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC). Eur Heart J. 2011 Dec;32(24):3147-97