



Introduction

Ebstein's anomaly is a congenital malformation resulting in apical displacement of the septal leading to tricuspid regurgitation and atrialization of the right ventricle. Despite early treatment options, there is a subset of the adult population living with Ebstein's anomaly. Associated malformations seen in this subset include atrial septal defects (ASD), and atrial arrhythmias. Female patients with Ebstein's Anomaly can have successful pregnancies in the presence of adequate right heart function. Right heart function can be difficult to characterize in these patients, given the atrialization of the right ventricle [1].

Patient Presentation

- A 40 year old G5P4004 female 39 weeks for elective primary cesarean section and bilateral tubal ligation
- PMH: Ebstein's anomaly (unrepaired), secundum ASD (unrepaired), Wolf Parkinson White syndrome s/p ablation, and gestational diabetes
- Patient previously delivered 4 children via SVD
- Patient reports having invasive lines in previous births

Case Description

- Preoperatively:
 - Trans-thoracic echocardiogram (TTE) noted an ejection fraction of 61%, PA pressure of 29 mm Hg and severe tricuspid regurgitation.
 - Planning included timing of anticoagulant therapy, type and screen, availability of both invasive monitors and cardiac anesthesia
 - Placement of epidural with constant monitoring of non-invasive blood pressure (NIBP), oxygenation, ECG, and fetal heart tones
 - The patient remained hemodynamically stable and was transported to the OR.

Case Description (cont.)

- Intraoperatively
 - Carefully titrated epidural anesthesia was administered preoperatively with 2% Lidocaine 1:200.000 epinephrine to a total of 10ml over 30 minutes.
 - On arrival, NIBP remained close to baseline at 135/83 mm Hg.
 - Sensory level was accessed at T6 after an additional 5ml epidural aliquot.
 - 15 minutes later the patient's blood pressure fell to 101/57 mm Hg, with a decrease in SpO₂ to 82%.
 - Phenylephrine infusion and supplemental O₂ was started with immediate recovery.
 - Uterine incision to delivery of baby was within 1 minute. Oxytocin was slowly administered and excessive IVF was avoided.
 - Phenylephrine infusion was weaned off successfully.
 - Following surgical closure, 2mg of Morphine was given through the epidural prior to removal.
- Postoperatively
 - Mother was monitored in PACU with no significant changes in hemodynamics.
 - Point of Care TTE displayed findings similar to preoperative exam.

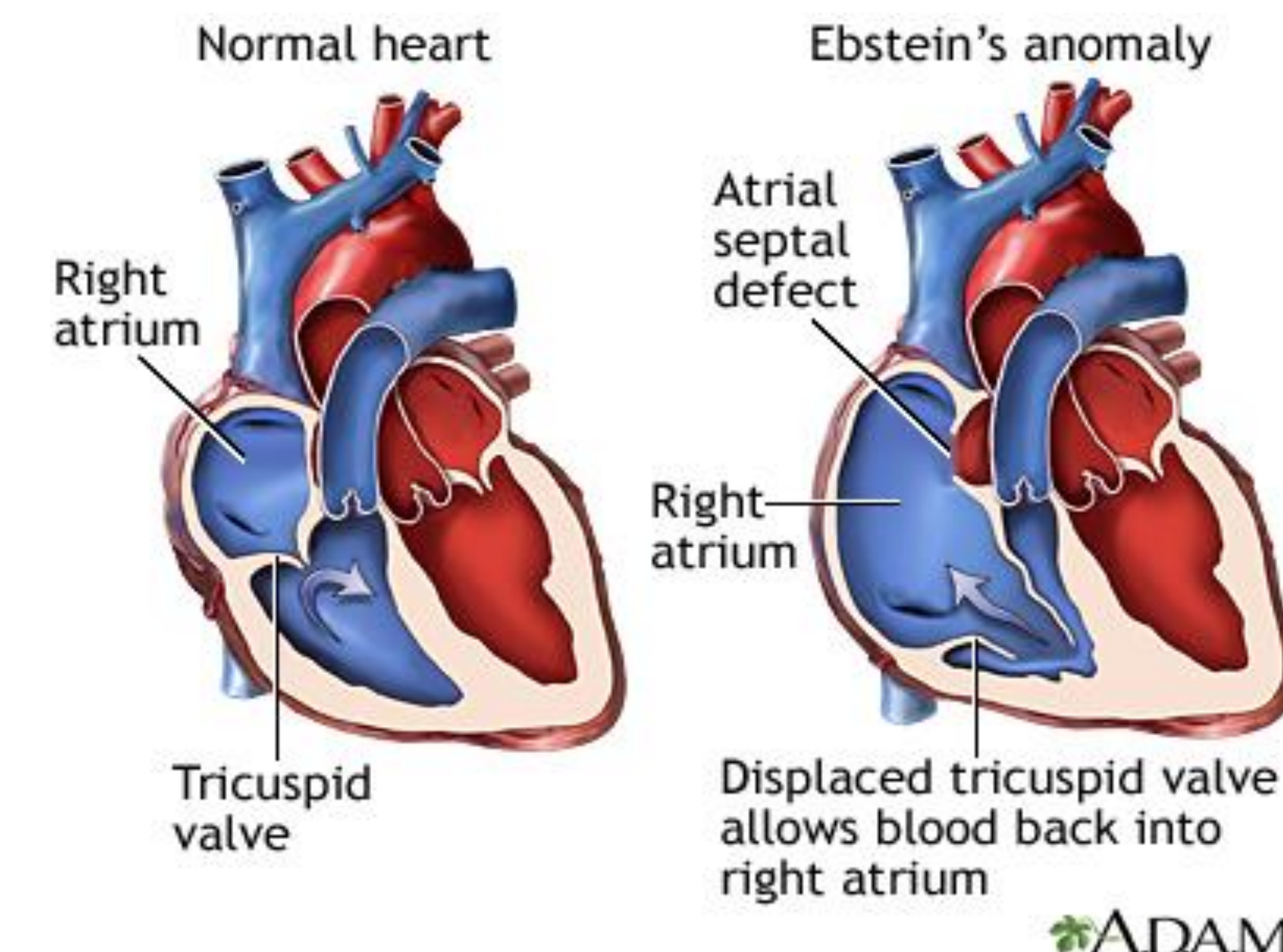


Figure 1. Depiction of Ebstein's Anomaly vs Normal Heart Anatomy (2)

Discussion

- Ebstein's anomaly, unrepaired, in a 41 year old female presenting at full term for elective cesarean section is rare.
- Unique challenges encountered during this case included placement of neuraxial anesthesia in a parturient sensitive to changes in afterload, providing adequate surgical anesthesia, attention to preload postpartum to ensure normal right heart function, and having appropriate post-operative follow-up.
- Despite careful titration of neuraxial anesthesia the patient experienced a reduction in systemic vascular resistance and afterload that led to a transient reversal of a left to right shunt and subsequent hypoxia.
- Decision made to not place invasive monitoring was made as surgical team, with all necessary equipment to place invasive lines in the OR. Future cases should weigh risks vs. benefits closely, and ultimately have necessary resources on standby

Citation

1. Kanoh, M., Inai, K., Shinohara, T., Shimada, E., Shimizu, M., Tomimatsu, H., Ogawa, M., & Nakanishi, T. (2018). Influence of pregnancy on cardiac function and hemodynamics in women with Ebstein's anomaly. *Acta Obstetrica Et Gynecologica Scandinavica*, 97(8), 1025–1031. <https://doi.org/10.1111/aogs.13373>
2. U.S. National Library of Medicine. (n.d.). *Ebstein anomaly: Medlineplus medical encyclopedia*. MedlinePlus. Retrieved April 26, 2022, from <https://medlineplus.gov/ency/article/007321.html>