## **Background and Importance**



- Mortality as high as 30-50% despite increasing technology (up to 56% in 1990s)<sup>1</sup>
- Contraindication to pregnancy<sup>2</sup>
- Physiologic changes put pregnant women at risk: <sup>3</sup>
  - Increased plasma volume and CO
  - Overloaded cardiopulmonary system, reduced SVR
  - Increased risk of thrombosis
  - Reduced PA compliance + increase in RV afterload and EDV  $\rightarrow$  ultimately lead to RV dysfunction
    - 1. J Am Coll Cardiol. 1998;31(7):1650-1657
    - 2. Obstetrics & Gynecology 133(5):p e320-e356
    - 3. Integr Blood Press Control. 2022;15:33-41



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## Patient & Case

- 23yo G1P0 admitted at 27w6d in the setting of an unplanned, but desired pregnancy
- PMH: BMPR2 mutation → Type 1 pulmonary arterial HTN (functions in the antiproliferative signaling pathway)
- Worsening RV function throughout pregnancy requiring transition from oral pulmonary vasodilators to continuous IV therapy
- At 25w2d her estimated PAP on TTE was 123mmHg with flattened septum, by week of delivery it had improved to 52mmHg
- At 30w6d she required emergent cesarean for pre-term labor under GA due to inability to safety perform neuraxial (AC not held, thrombocytopenia from Treprostinil up titrations)

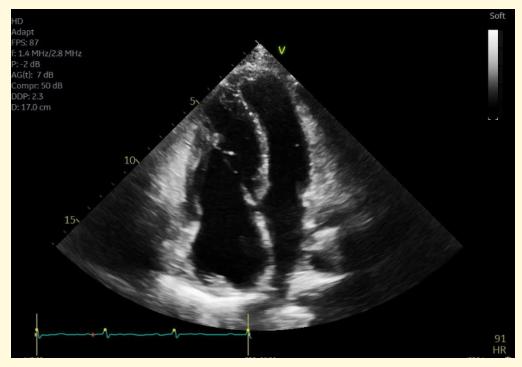


Figure 1. Apical four chamber view with severe RV dilation.

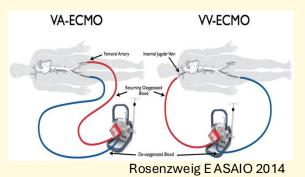
- ECMO was available with CT surgery in OR.
- Preinduction arterial line, PAC and swan were placed.
- iNO was added after intubation.
- PA pressures were suprasystemic by delivery and beyond into PP period, reaching 140-180mmHg at times. No vasopressor was required during case.
- ECMO was considered daily but ultimately never cannulated due to reassuring end organ perfusion.



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## **Discussion and Take Aways**



- Multidisciplinary care team at a center with experience in PHTN in pregnancy is paramount. (phone tree, back up plans)
- Must include expertise from MFM, NICU, PHTN, OB anesthesia, CT anesthesia, CT surgery, ICU, pharmacy, and ECMO cannulation team.
- Greatest risk is RV dysfunction and failure in the immediate postpartum period up to 72hrs after delivery and ECMO should be continually considered if eligible.
- This case contributes to the growing knowledge based due to her gestational age achieved in the setting of her disease severity and eventual morbidity despite maximal efforts by a multidisciplinary team.
- Our team has also considered ECMO earlier in the course and more preemptively in our cases following this patient. We must strive to stay vigilant during the postpartum period and work collaboratively with ICU team for best possible outcome.



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