Management of Pregnancy Termination for a Patient with Eisenmenger Syndrome and Chiari I Malformation with Associated Syringomyelia

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Background

- **Arnold Chiari Type 1** is the most prevalent Chiari Malformation, affecting **0.7%** of the population; associated with **syringomyelia** in **25% of patients** (1).
- Delivery Planning: concerns for **brainstem herniation** with unintentional dural puncture; concerns with **elevated ICP** due to Valsalva during stage 2 or with laryngoscopy.
- Theoretical concern for **worsening neurologic function** in patients with symptomatic syringomyelia after neuraxial procedures and vaginal delivery (2).

- Eisenmenger Syndrome is the end-stage result of long-standing unrepaired congenital heart disease lesions, leading to intracardiac **right-to-left shunting** due to **pulmonary hypertension**.
- Maternal mortality for women with Eisenmenger Syndrome ranges between **20-40%** (3).

- Illinois: statutory protection for **abortion as a fundamental right** (4).
- Limits on abortion after **fetal viability** are permitted (4).

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Case Timeline

Pre-op

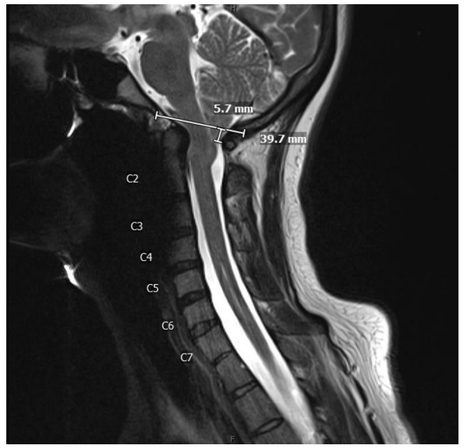
CC: 32-year-old G2P0010 female with upper extremity weakness, DOE

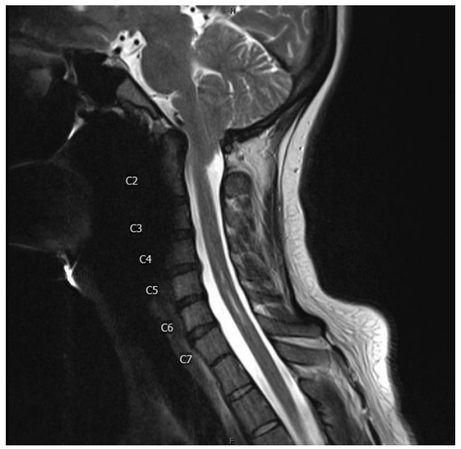
VS: SpO_2 93%

CBC: Hgb 18.5
HCG: positive (6w6d)

Stroke Code: CT brain, CT-A head/carotid with Chiari I and PDA

Admission: further workup





Chiari I (*above*) and Syrinx (*right*)

Multidisciplinary Planning

MFM
Complex Family Planning

Obstetric Anesthesiology

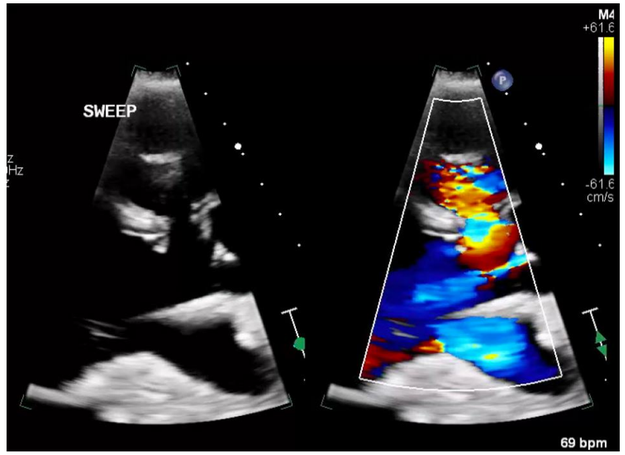
Cardiac Surgery
Cardiothoracic Anesthesiology
Perfusion

Congenital Cardiology
Pulmonary Hypertension

Operating Room Services

Dilation & Curettage

- **Anesthetic Plan:** two large-bore PIVs, minimal sedation (fentanyl, midazolam, ketorolac), paracervical block, in-person interpreter, music therapy.
- **Emergency Planning:** inhaled nitric oxide circuit, arterial line, central line, cardiac surgeon, ECMO.



Infundibular/supracristal ventricular septal defect

Post-op

- Routine PACU Care.
- Discharged **POD 1.**
- Continues to follow with congenital cardiology, pulmonology.

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Discussion and Teaching Points

- Pregnant patients with severe pHTN and Eisenmenger Syndrome face **high morbidity and mortality**.
- Pregnancy **termination should be offered**.

- **Maintain SVR**.
- **Avoid worsening PVR** (hypoxia, hypercarbia, acidosis, PEEP, hypothermia, sympathetic stimulation).
- **Neuraxial preferred** if no other contraindications.
- Continuous ECG, invasive arterial monitoring, inhaled nitric oxide or prostaglandins.

- Care coordination: **2 weeks**, more than **20 specialists** and providers.
- Ability to avoid legal concerns causing **delays in care** while respecting **patient autonomy** and **dignity**.

References

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(4) 775 Ill. COMP. STAT. 55/1-15.
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