

Anesthetic Management of a Parturient with Co-arctation of Aorta and Noonan Syndrome Undergoing Cesarean Section: A Case Report

Ashka Surve MD, Ibrahim Mohamed MD. PhD. MBA. FASA., Ryan Griffin MD., Taha Mohamad BSA.
University of Texas Medical Branch, Galveston, TX.

BACKGROUND



Heart Disease

Cardiovascular diseases are now a leading cause of maternal mortality in developed world [1].

Coarctation of the Aorta (CoA)

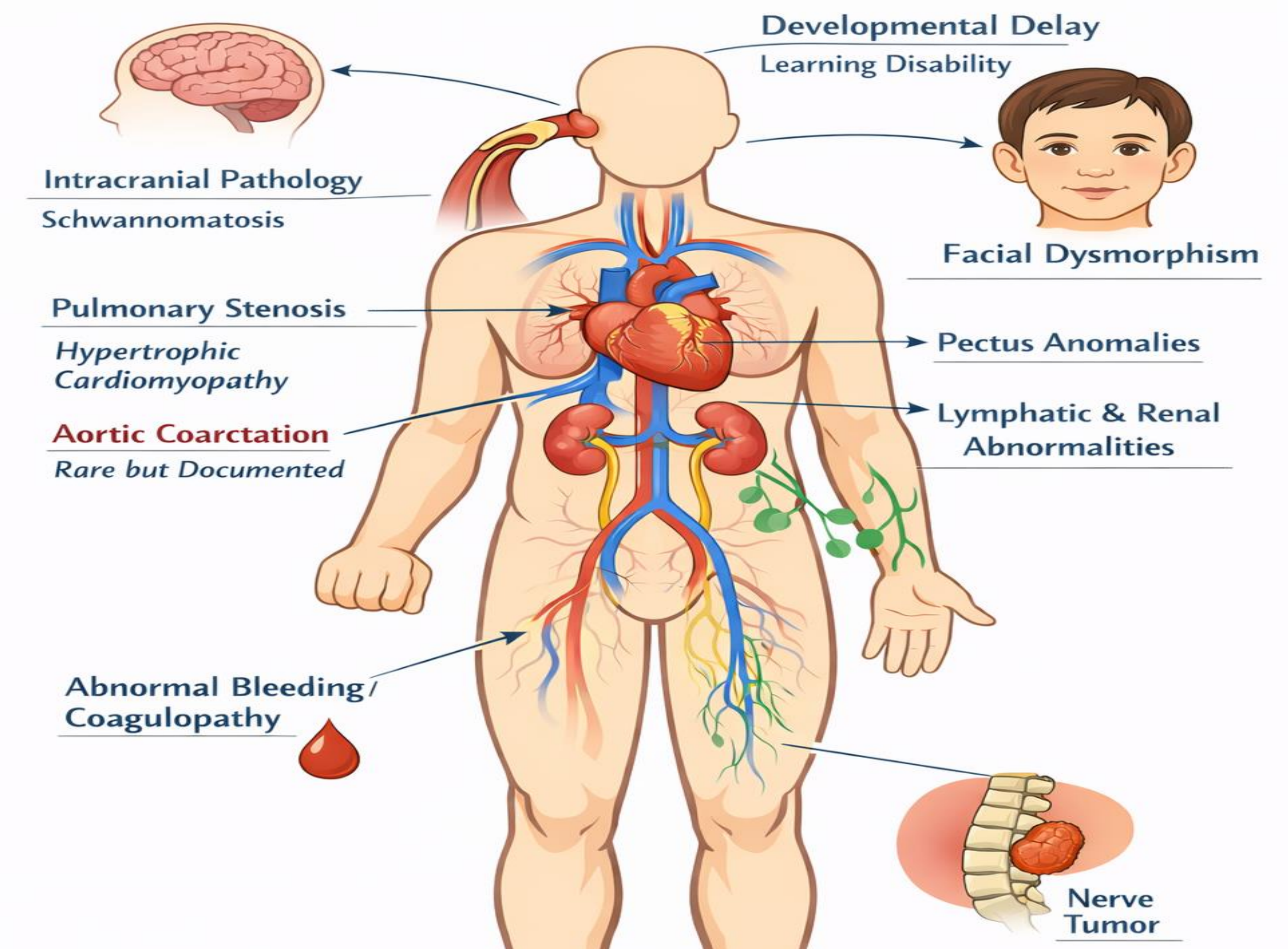
- Incidence ~1 in 10,000 live births [2,3]
- If Unrecognized in infancy → progressive intimal thickening[4]
- Hypertension, LVH, HF
- Upper > lower extremity BP gradient.
- Peak-to-peak pressure gradient between LV and aorta >20mmHg
- Renal/Hepatic complications [5]
- Screen for associated defects [5,6,7]

Pregnancy Concerns

- | | |
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| <ul style="list-style-type: none"> • Aortic Complications • Cardiac Decompensation | <ul style="list-style-type: none"> • Supine Hypotension • Coagulopathy • Associated Intracranial Pathology |
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Noonan Syndrome

- A disorder of RASopathy — germline Ras/MAPK pathway variants mutation[6]
- LZTR1 loss-of-function mutation. [7]
- Incidence ~1/1,000[8]



Patient Profile

22F • G1P0 • 39 weeks • ASA III

Elective primary cesarean section

Past Medical History:

- Coarctation of Aorta and Noonan Syndrome
- Aortic stenting age 4, re-dilation age 8
- Hypertension;

Cardiac Workup:

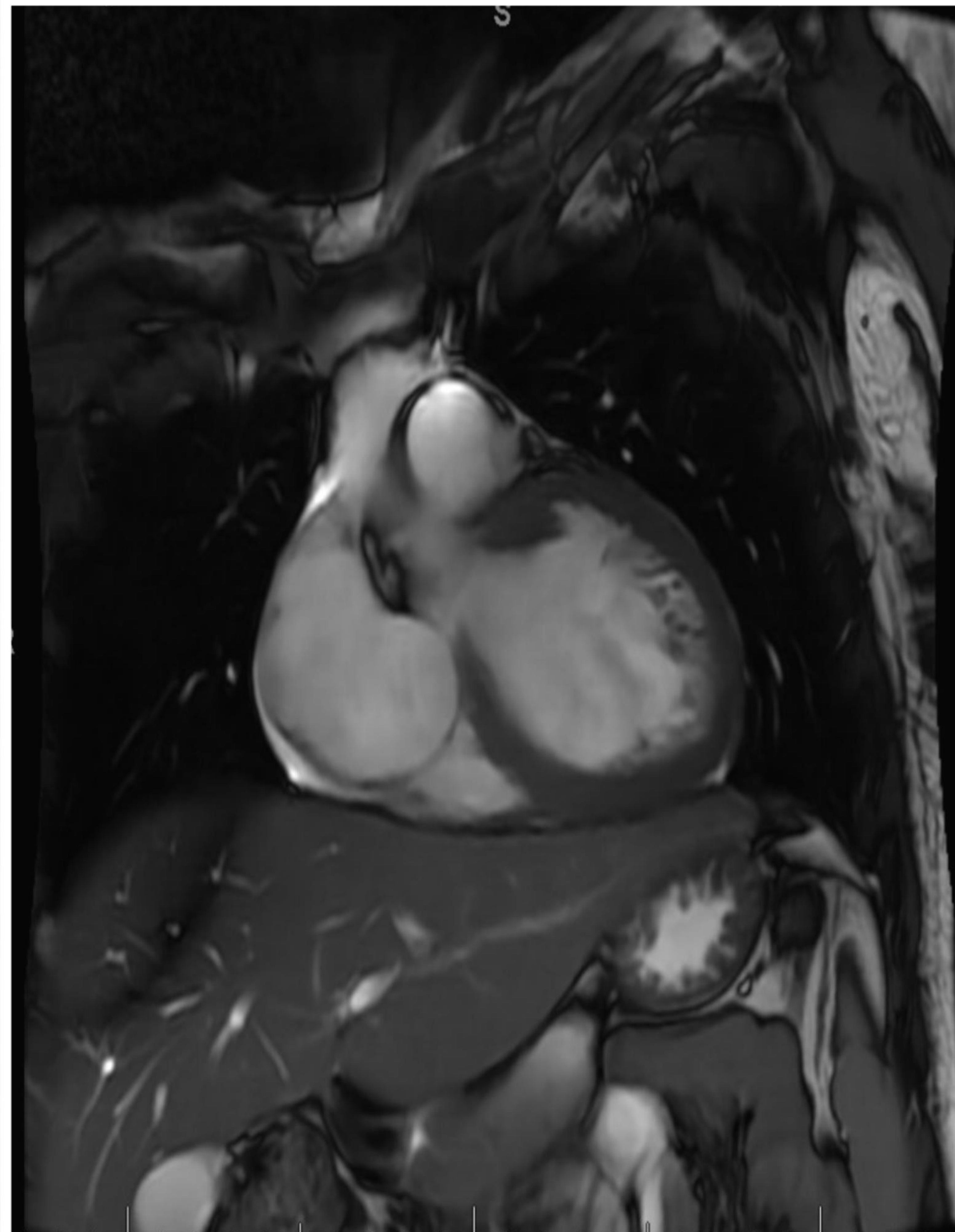
TTE (3 days pre-op):

- Mild mitral stenosis
- ↑ LV mass, mild eccentric hypertrophy
- EF 60–65%
- 25 mmHg gradient across stent
- **Cardiac MRI (2nd trimester):**
- **proximal descending aorta** stent,
- peak velocity 238 cm/s
- mean gradient 22.7 mmHg → re-coarctation

MR Angiogram/Venogram head: no abnormal findings

Labs & Pre-op: Hb 10.0 g/dl, MPG II

Medications: Aspirin 81 mg + Metoprolol XL 12.5 mg



Cardiac MRI showing LVH

Anesthesia Considerations

GA risks:

- intubation-Hemodynamic stress,
- aspiration,
- myocardial depression,
- ↑ systemic opioids use

CSE: low intrathecal dose



gradual sympathectomy



hemodynamic stability

Left uterine displacement

Monitoring: Standard ASA and **Invasive BP**

- Furosemide 20 mg IV after placenta delivery — prevent fluid overload from auto transfusion.
- Uneventful post-operative recovery.
- **Post partum** CT angiogram chest and abdomen- No changes in Aorta.

CoA in Pregnancy

- ↑ Blood volume & CO → risk of rupture, dissection, hypertensive crisis, distal hypoperfusion [1,8]
- AHA recommends monthly echo monitoring for ascending aorta changes [3]

Noonan Syndrome

- Patients with Noonan Syndrome can have phenotypic variability: no characteristic facial features in our patient – Whole Exon Sequencing was essential for diagnosis
- Multidisciplinary planning (Cardiology, MFM, Genetics, Anesthesia) is essential.
- Invasive and NIBP systolic readings may significantly diverge in CoA —mean arterial pressure is the most reliable common parameter; dual-extremity monitoring is ideal [1]

Anesthetic Strategy Comparison

- Neuraxial anesthesia with gradual sympathetic block.
- GA if necessary: Etomidate + Lidocaine to blunt laryngoscopy response, hemodynamic stability

Modified WHO cardiovascular risk stratification in pregnancy		
Class	Common Conditions	Pregnancy Risk
I	Uncomplicated small or mild pulmonary stenosis, PDA, mitral valve prolapse, successfully repaired simple lesions (e.g. ASD, VSD).	Risk no higher than general population
II	Unoperated ASD or VSD, repaired Tetralogy of Fallot, most arrhythmias.	Small increased risk of maternal mortality and morbidity
II-III	mild left ventricular impairment, hypertrophic cardiomyopathy, native or tissue valvular heart disease not considered WHO I or IV, Marfan syndrome without aortic dilatation, aorta <45 mm in aortic disease with bicuspid aortic valve, repaired coarctation	Intermediate increased risk of maternal Mortality or Moderate to Severe risk of morbidity
III	Mechanical heart valves, systemic right ventricle, fontan circulation, cyanotic heart disease	Significant increased risk of maternal mortality and morbidity
IV	Pulmonary arterial hypertension, severe systemic LV dysfunction (NYHA III-IV or EF < 30%), Marfan syndrome (aorta > 40mm)	Pregnancy contraindicated

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