Obstetric Management of Cardiac Disease in Alagille Syndrome

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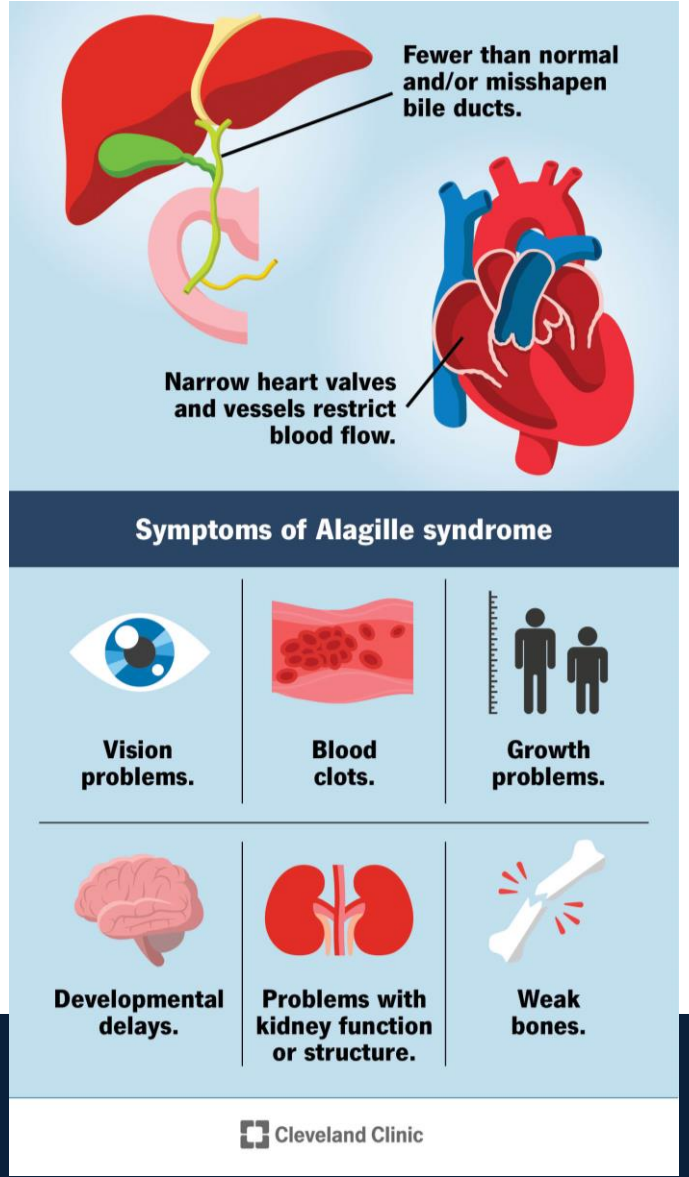
Background

- Alagille syndrome (ALGS) – autosomal dominant connective tissue disease
  - Variable involvement of liver, heart, eyes, skeleton, face, brain, kidneys, and vasculature
  - Mutation in the *JAG1* and *NOTCH2* genes
  - Incidence of 1 : 30,000
- Cardiac disease – present in 85-98% of ALGS
  - R lesions are most common, i.e. pulmonary artery stenosis (PAS)
  - Others – tetralogy of Fallot, supravalvular aortic stenosis, L lesions
  - 15% mortality in ALGS deaths
- Anesthetic considerations –
  - Airway concerns d/t facial anomalies
  - Neuraxial considerations 2/2 vertebral defects, coagulopathy
  - Cirrhosis, portal hypertension, esophageal varices (i.e. rupture with expulsive efforts)



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Case Presentation

- 24yo G1P0 with bilateral PAS, chronic hypertension, biliary diversion at age 7
  - Fam Hx: mother with ALGS-related heart disease, maternal grandmother s/p heart and liver transplant
  - Evaluation for pulmonary angioplasty underway prior to pregnancy

HPI

- Pre-E w/ SF at 33 wks
- BMZ, Mg
- Nifedipine, labetalol for cHTN

Echo

- L PA – 91mmHg
- R PA – 43mmHG
- Normal EF
- RV hypertrophy
- Normal fetal echo

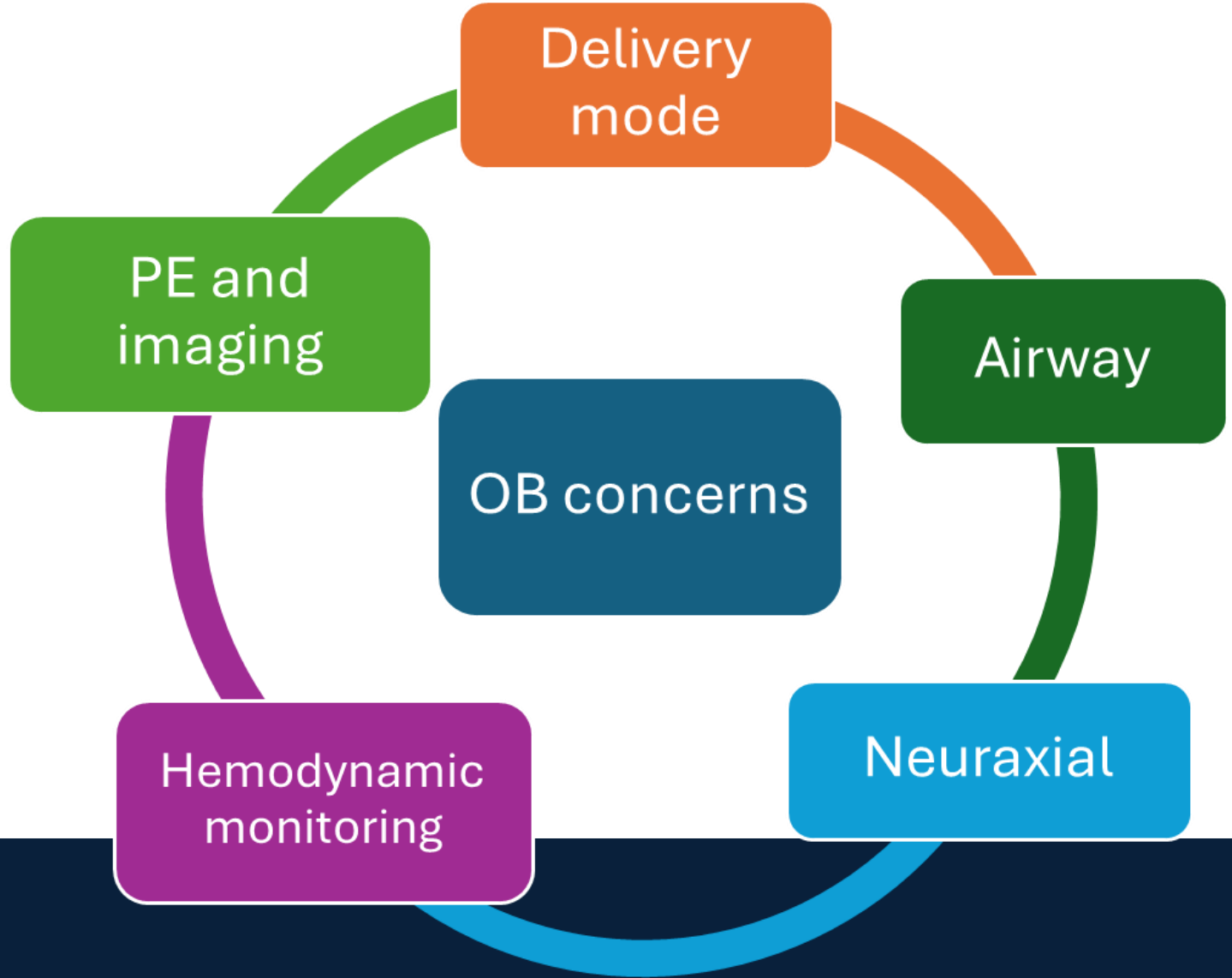
Delivery

- IOL at 34 wks
- A-line, epidural (x2)
- Sustained SRBP, prolonged decels
- Emergent CS
- APGAR 6 and 8, 1680g

Post-Partum

- Tele x 12 hrs
- 48-hr TTE – L PA 100 mmHG, R PA 48 mmHG
- Nifedipine dose increased
- Asx, discharged

Summary - Learning Points



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