

Management of a Parturient with Vein of Galen Malformation

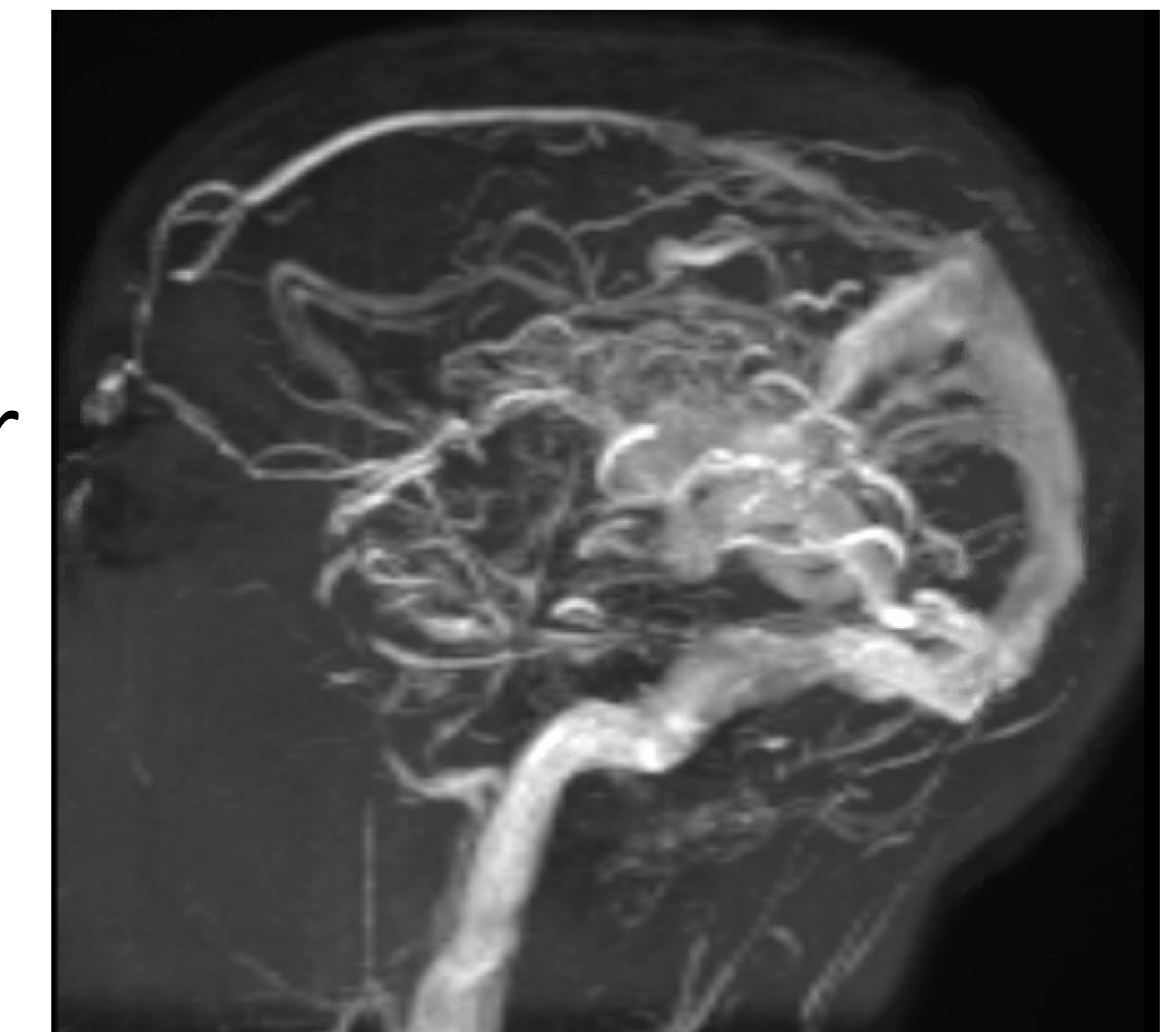
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Vein of Galen Malformations (VGAM)

- VGAM lesions are rare congenital arteriovenous malformations
 - Account for 30% of pediatric congenital vascular malformations
- Develop between 6- and 11-weeks gestation
 - About 30% diagnosed prenatally via ultrasound
- Associated with:
 - Cardiovascular Complications
 - High output heart failure (HOHF), pulmonary hypertension, coarctation of the aorta, tricuspid valve insufficiency, and Patent Foramen Ovale
 - Neurodevelopmental Complications
 - Seizure, developmental delay, macrocephaly, hydrocephalus and subarachnoid/parenchymal hemorrhage
- Prognosis: Dependent on severity and timing of symptoms
 - Most warrant endovascular intervention in childhood
- Pregnancy: At risk for cardiac chamber dilation, pulmonary hypertension, arrhythmia, heart failure, and intracranial hemorrhage
- Case: A 24 y.o. G2P0 at 14 weeks gestation presented with symptoms concerning for HOHF
 - PMH: obesity, depression, and a large, unrepaired VGAM (diagnosed in infancy)
 - Childhood: mild, stable LV dilation with preserved ejection fraction without cardiac or neurologic symptoms
 - Pregnancy: Shortness of breath and palpitations
 - Holter Monitor: ventricular ectopy (5.4%)
 - ECHO: Pertinent for LV dilation



Sagittal view of Brain MR Venogram which notes large VGAM and associated feeding vessels

Surgical and Anesthetic Management

- Multidisciplinary Collaboration
 - Neurosurgery
 - Sequential embolization in the second trimester was considered, but was not pursued due to high risk of serial embolization in pregnancy
 - Cardiology
 - Monitoring of cardiac symptoms
 - Serial ECHO and B-type natriuretic peptide
 - Metoprolol 25mg daily in the setting of ventricular ectopy
 - Maternal-Fetal Medicine
 - Initially counselled for termination of pregnancy given cardiac and neurologic risk
 - IOL with assisted-second stage vaginal delivery
 - Obstetric Anesthesiology
 - Labor: Early neuraxial placement
 - Minimize stress and pain response to labor
 - Risks of unintended dural puncture and intracranial hypotension were considered
- Cesarean section: Conversion of labor epidural to surgical anesthesia in the setting of failed IOL
 - Minimize hemodynamic fluctuations associated with induction, laryngoscopy, surgical stimulation, and emergence.
 - Epidural anesthesia allows for continuous neurologic assessment and incremental dosing
 - Optimization of multimodal analgesia for post-cesarean pain control and antiemetic prophylaxis
- Safe delivery of a liveborn neonate with weight of 4196g and Apgar score of 9 and 9 at 1 and 5 minutes, respectively.
 - Postpartum: Uncomplicated course
 - ECHO revealed stable LV size and LVEF of 55%
 - Delivery at a Level IV Maternal Care Center with access to immediate neurosurgical intervention is critical