AMSER Case of the Month August 2025

2-hr-old female with hypoxemia and cardiac overload

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LOMA LINDA UNIVERSITY HEALTH SYSTEM

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Patient Presentation

- 2-hour-old ex-36-week female born via normal spontaneous vaginal delivery presents with grunting and intercostal retractions
- Vital signs positive for hypoxemia (SpO2 50-60%), which was responsive to PPV (SpO2 93%)
- Physical exam positive for a hyperdynamic precordium and continuous pulsatile bruit over the anterior fontanelle
- Echocardiogram shows dilated heart chambers and increased flow, concerning for high-output heart failure

What Imaging Should We Order?

ACR Appropriateness Criteria

Variant 3: **Child. Clinical signs or symptoms of vascular anomaly (tumor or malformation) not suggesting infantile hemangioma. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
US area of interest	Usually Appropriate	O
US duplex Doppler area of interest	Usually Appropriate	O
MRA and MRV area of interest without and with IV contrast	Usually Appropriate	O
MRI area of interest without and with IV contrast	Usually Appropriate	O
US area of interest with IV contrast	May Be Appropriate (Disagreement)	O
MRA area of interest without IV contrast	May Be Appropriate (Disagreement)	O
MRI area of interest without IV contrast	May Be Appropriate	O
Arteriography area of interest	Usually Not Appropriate	Varies
Radiography area of interest	Usually Not Appropriate	Varies
CT area of interest with IV contrast	Usually Not Appropriate	Varies
CT area of interest without and with IV contrast	Usually Not Appropriate	Varies
CT area of interest without IV contrast	Usually Not Appropriate	Varies
CTA and CTV area of interest with IV contrast	Usually Not Appropriate	Varies

This imaging modality was ordered initially

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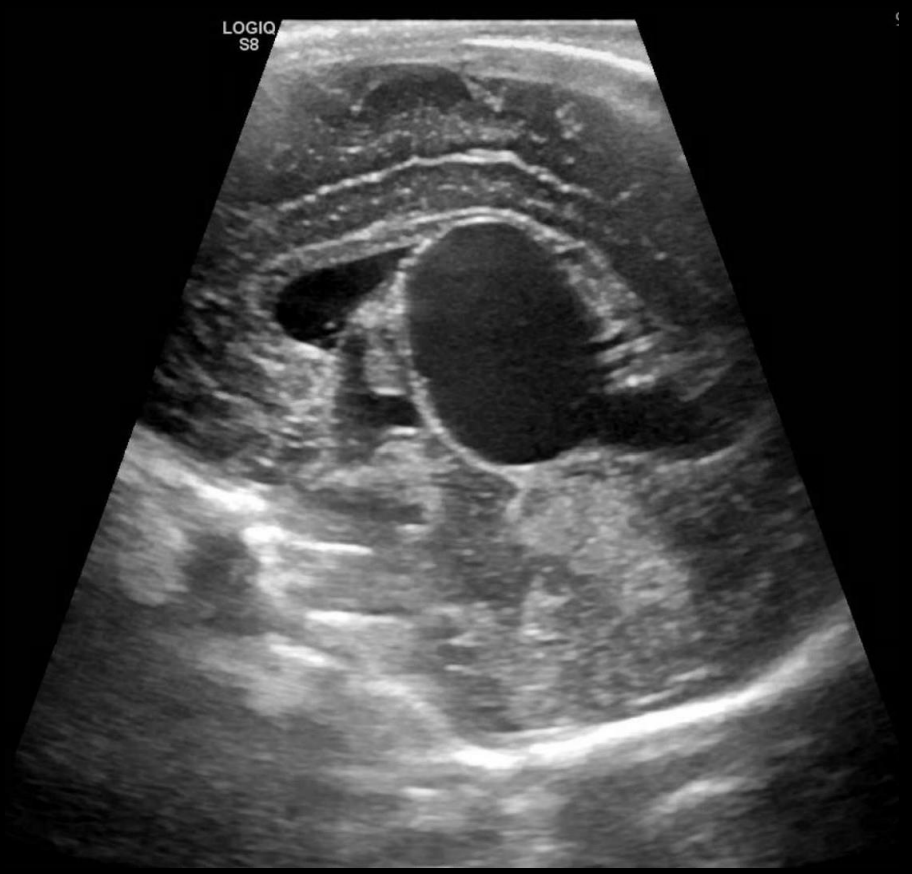
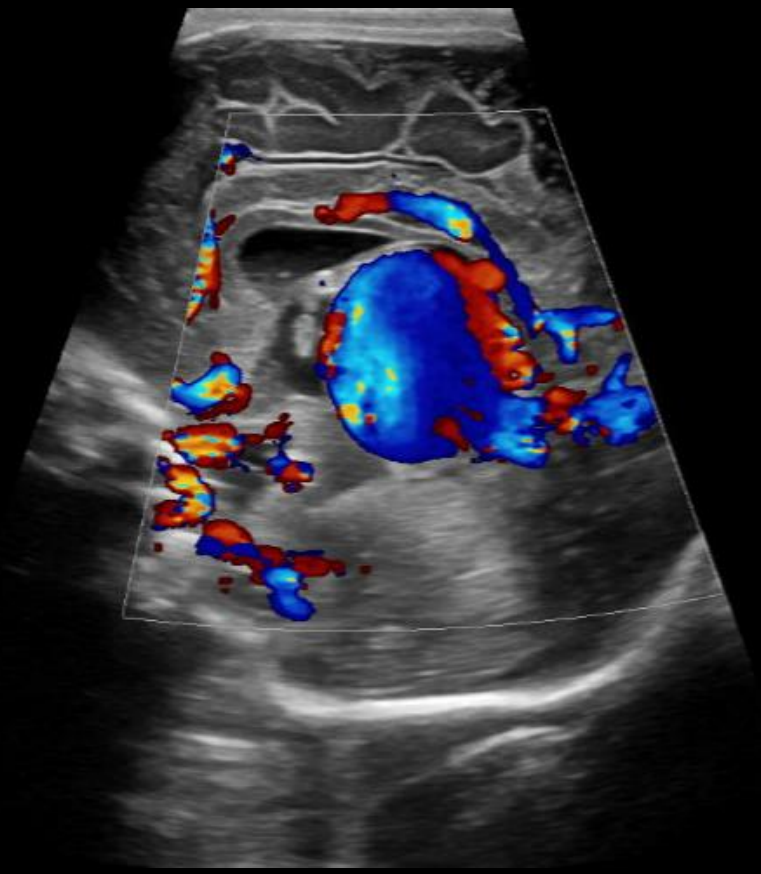
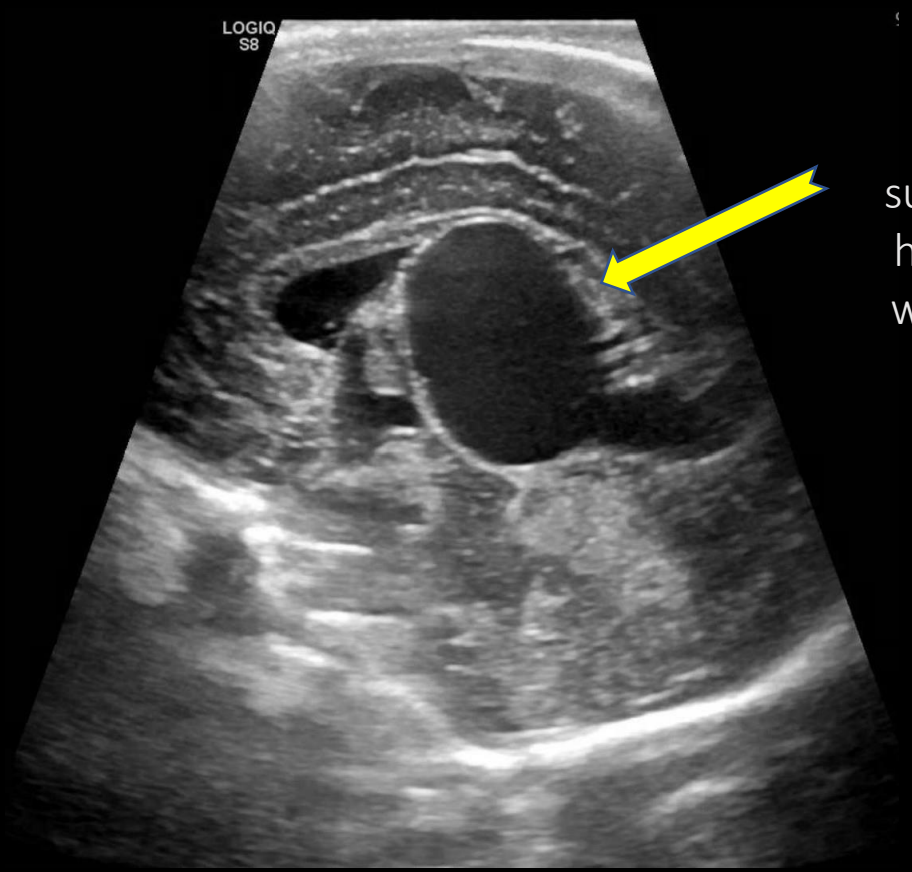
US Head with Doppler

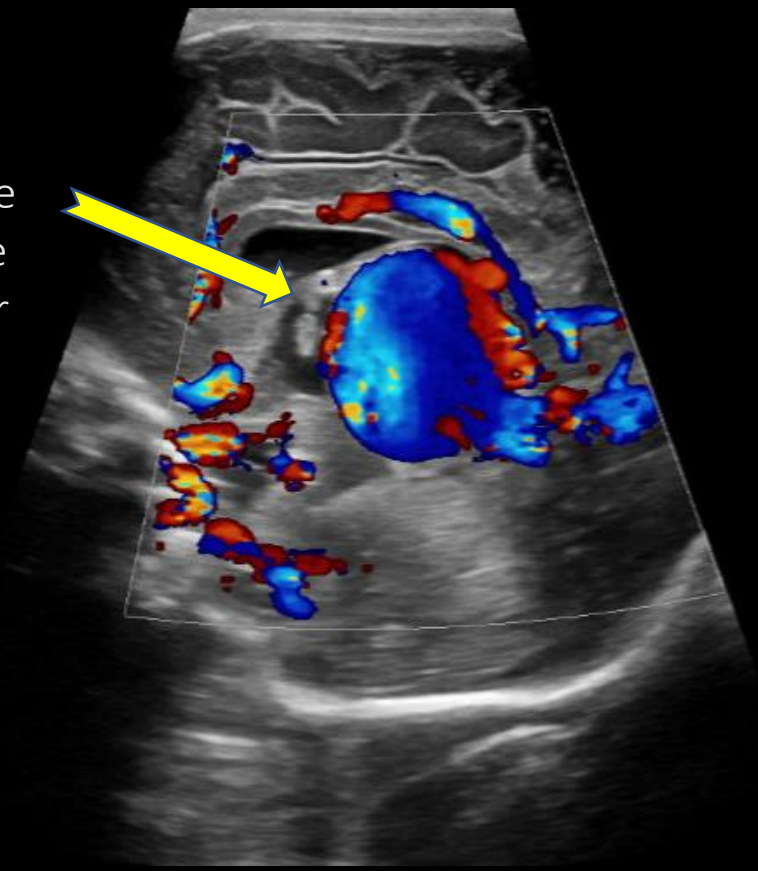




Findings



A 2.6 cm supratentorial midline hypoechoic structure with internal vascular flow



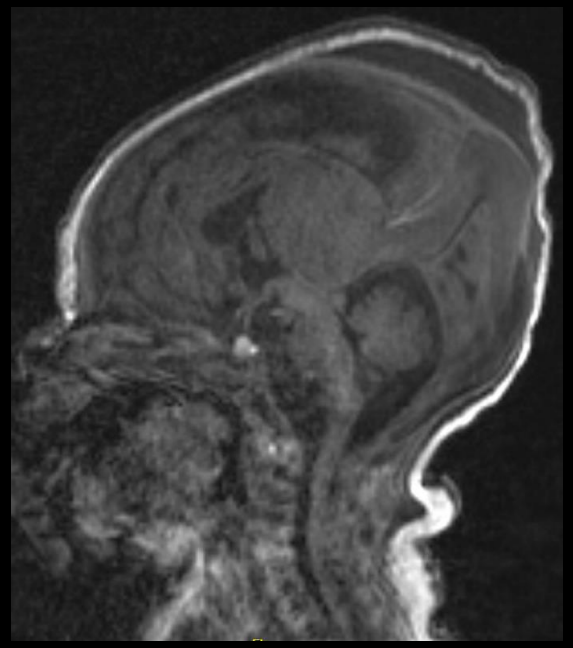
ACR Appropriateness Criteria

Variant 4: **Child. Ultrasound features raise suspicion for vascular malformation. Next imaging study.**

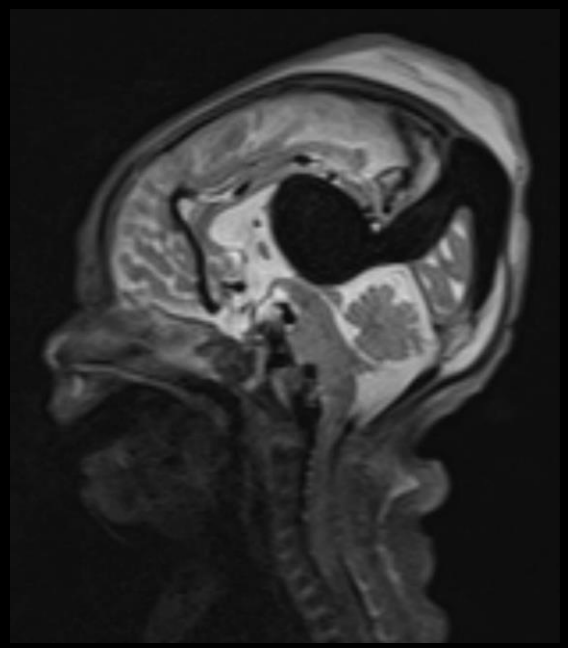
Procedure	Appropriateness Category	Relative Radiation Level
MRA and MRV area of interest without and with IV contrast	Usually Appropriate	O
MRI area of interest without and with IV contrast	Usually Appropriate	O
CT area of interest with IV contrast	May Be Appropriate	Varies
CTA and CTV area of interest with IV contrast	May Be Appropriate	Varies
US area of interest with IV contrast	Usually Not Appropriate	O
Arteriography area of interest	Usually Not Appropriate	Varies
Radiography area of interest	Usually Not Appropriate	Varies
MRI area of interest without IV contrast	Usually Not Appropriate	O
CT area of interest without and with IV contrast	Usually Not Appropriate	Varies
CT area of interest without IV contrast	Usually Not Appropriate	Varies

Next step in diagnosis

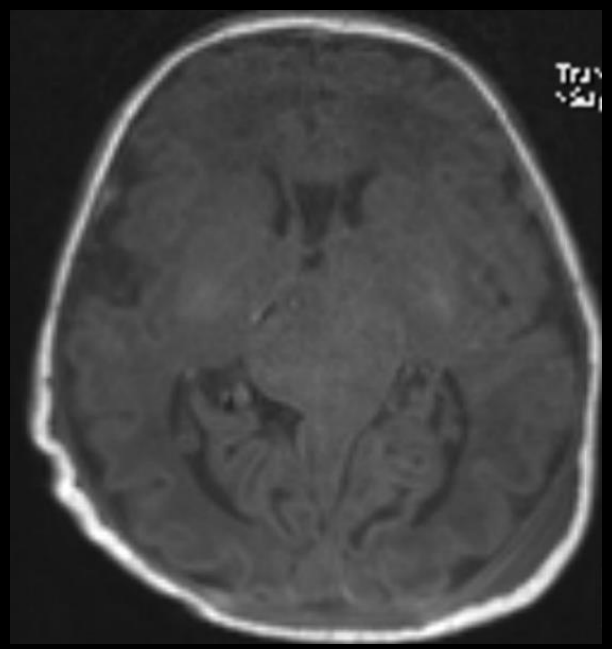
MRI Brain without Contrast



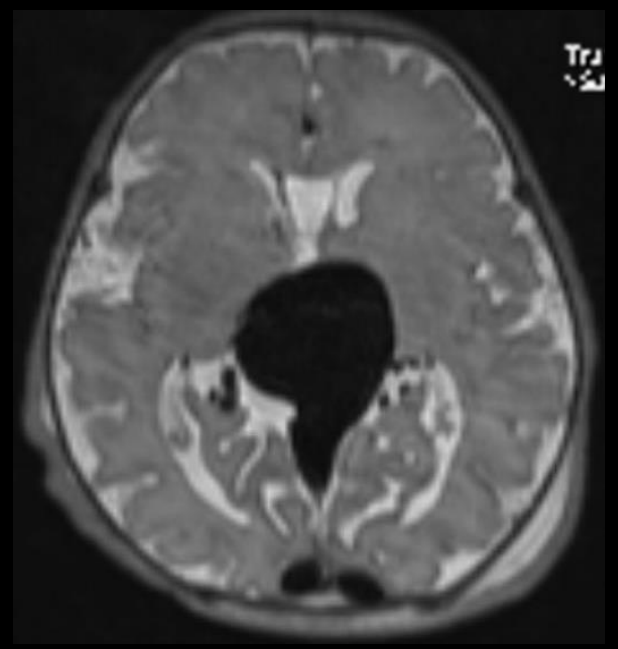
Sagittal T1



Sagittal T2

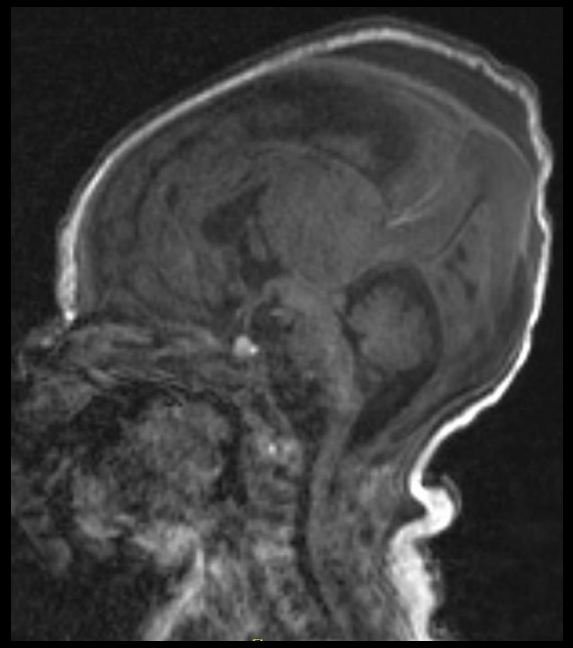


Axial T1

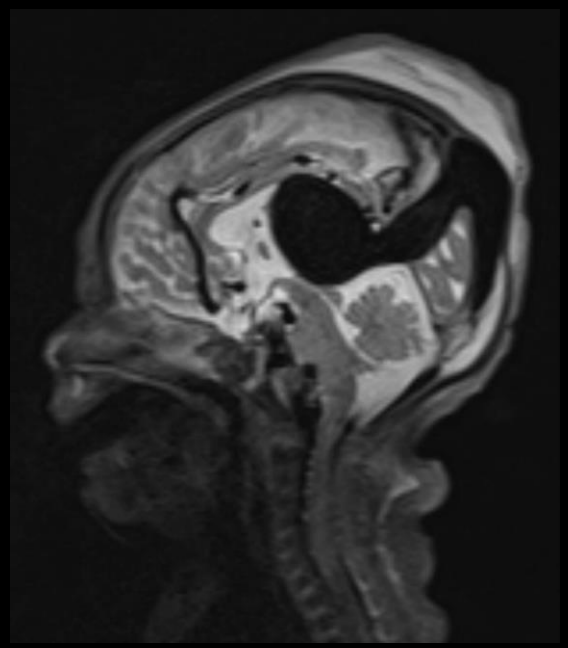


Axial T2

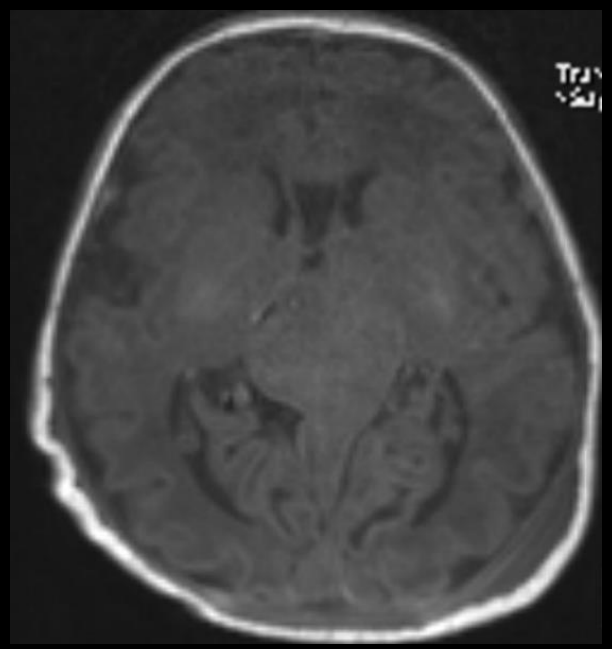
Findings



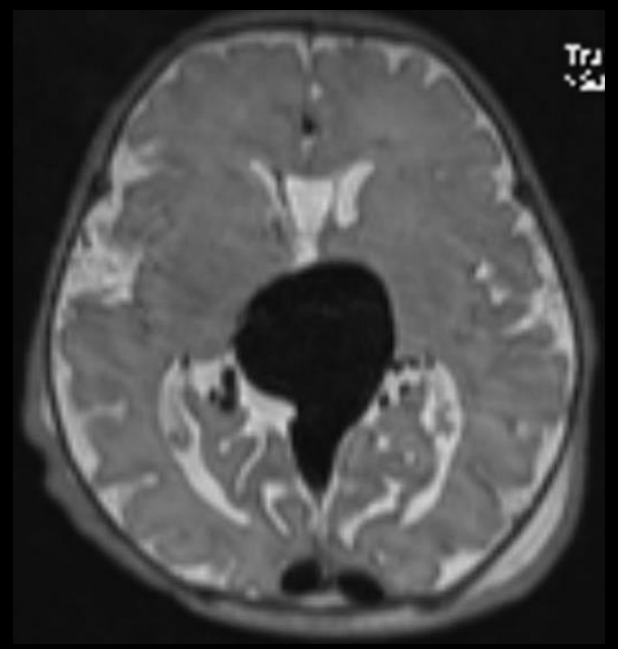
Sagittal T1



Sagittal T2



Axial T1



Axial T2

- Aneurysmal vascular structure in the pineal region draining into the superior sagittal sinus through a dilated persistent falcine sinus
- Enlargement of the venous confluence, with adjacent severe calvarial thinning vs. defect
- Hypoplastic straight sinus with prominence of the remaining dural venous sinuses

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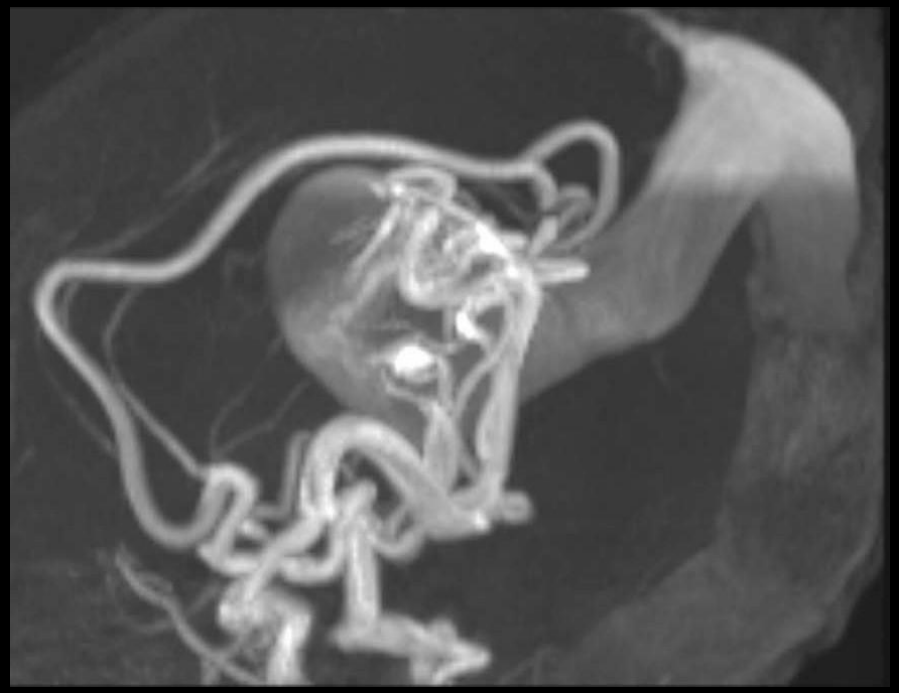
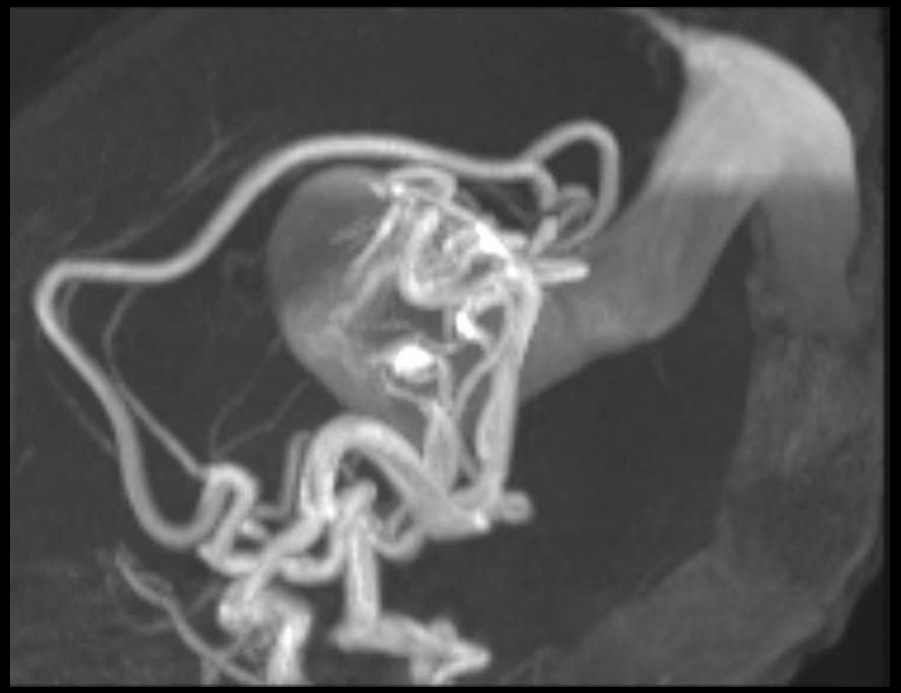
MRA Brain without Contrast



Findings



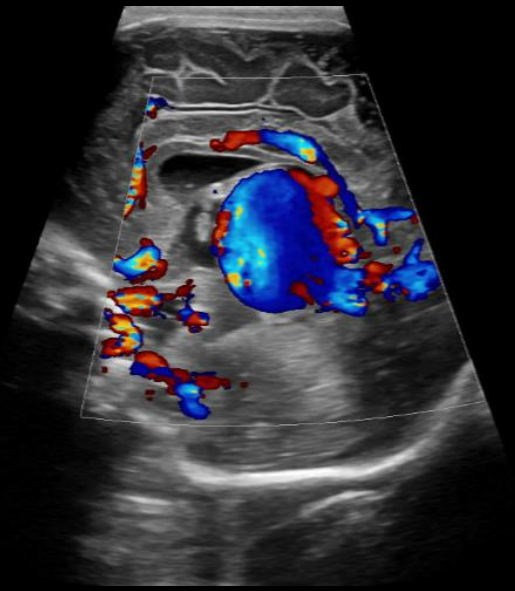
- Aneurysmal vascular structure in the pineal region with (1) enlarged feeding arteries arising from the right pericallosal artery and distal branches of the bilateral posterior cerebral arteries and (2) prominent draining veins into the superior sagittal sinus
- Prominent, tortuous bilateral posterior communicating arteries and basilar artery

Final Dx:

Vein of Galen Aneurysmal Malformation

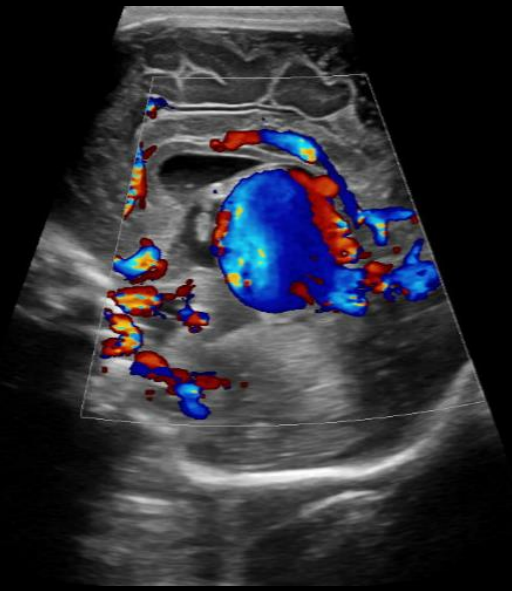
Vein of Galen Aneurysmal Malformation

- Definition:
  - High-flow arteriovenous fistula between the median prosencephalic vein and deep choroidal arteries
  - Develops between 6-11 weeks of gestation
  - Prevents the normal regression of the median prosencephalic vein
  - Misnomer: Not a true aneurysm of the vein of Galen, which fails to form due to the arteriovenous fistula



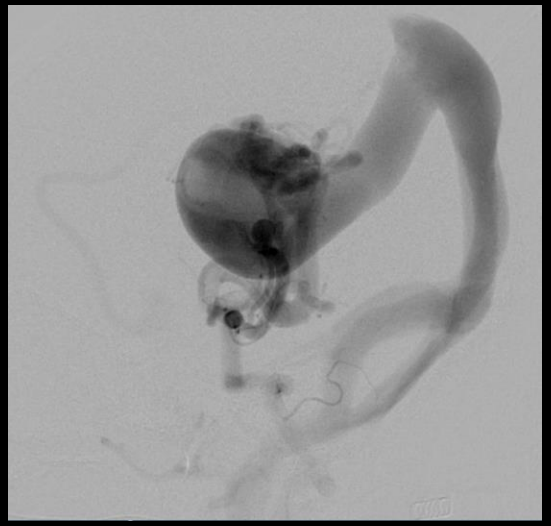
Vein of Galen Aneurysmal Malformation

- Demographics:
  - M:F = 2:1
  - 30% of pediatric cerebral vascular malformations
- Presentation:
  - High-output heart failure
  - Cranial bruit
  - Hydrops fetalis
  - Developmental delay



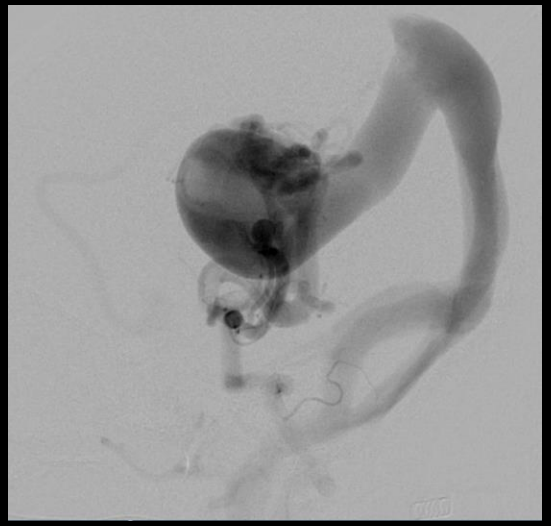
Vein of Galen Aneurysmal Malformation

- Imaging Characteristics:
  - US: Anechoic extra-axial structure at midline posteriorly with associated prominent Doppler flow
  - T1: Pulsation artifact possible from the varix; flow void or heterogeneous signal from turbulent flow
  - T2: Prominent flow voids from dilated feeding arteries and draining veins
  - MRA: Delineates arterial feeders and arterialized flow in the draining veins



Vein of Galen Aneurysmal Malformation

- Treatment:
  - Ideally, embolization is deferred until 6 months of age to allow the cavernous sinus to mature
  - If cardiac failure is refractory to medical management, embolization may be necessitated in the neonatal period
  - Arterial or venous embolization is possible
  - Coils and glue can be used in a single procedure or staged approach

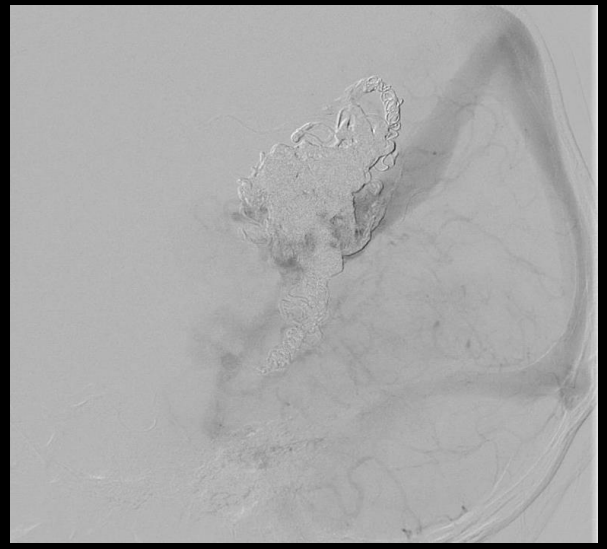


Treatment

- This patient underwent multiple endovascular embolizations via an arterial feeder approach.
- She is currently awaiting her 8th embolization.

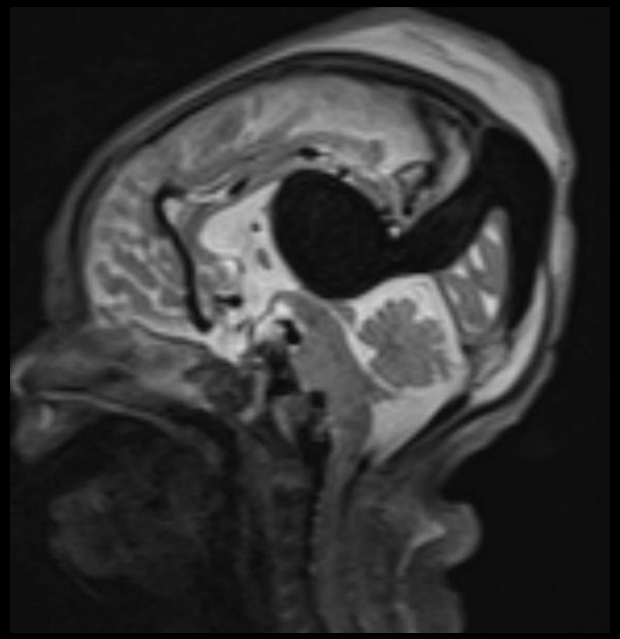
1st embolization

3rd embolization

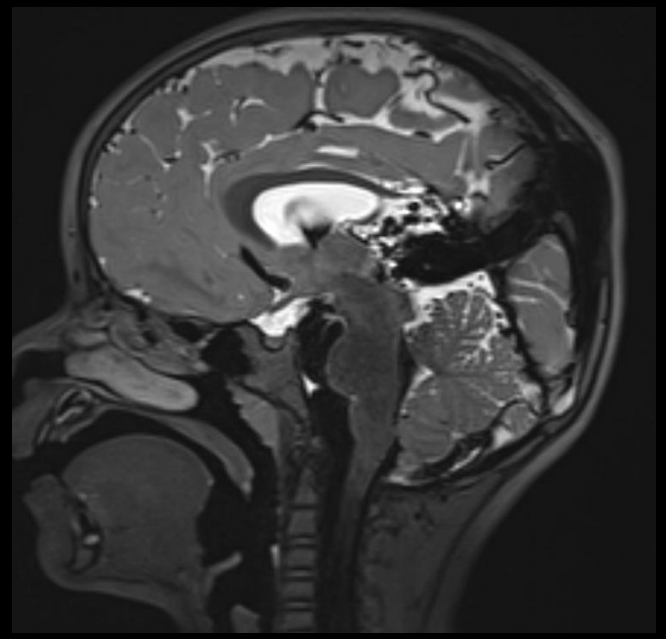


7th embolization

Pre- vs. Post-Embolization
MRI Brain without Contrast



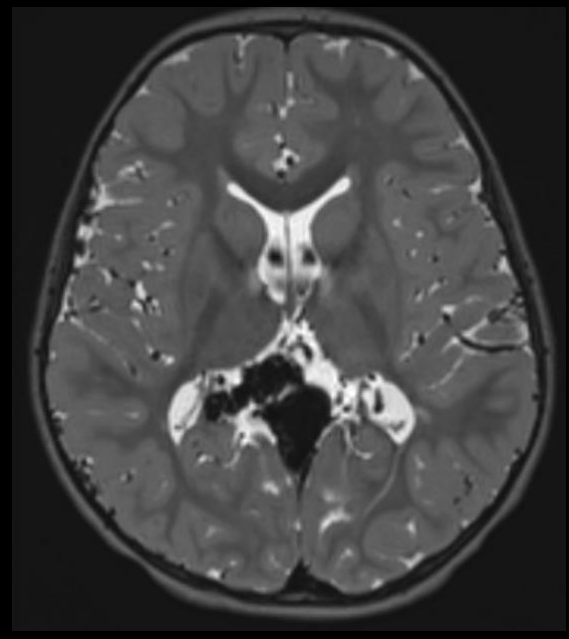
Pre



Post

Sagittal T2

Pre



Post

Axial T2

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References:

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