



Lisa McLeod HMS III

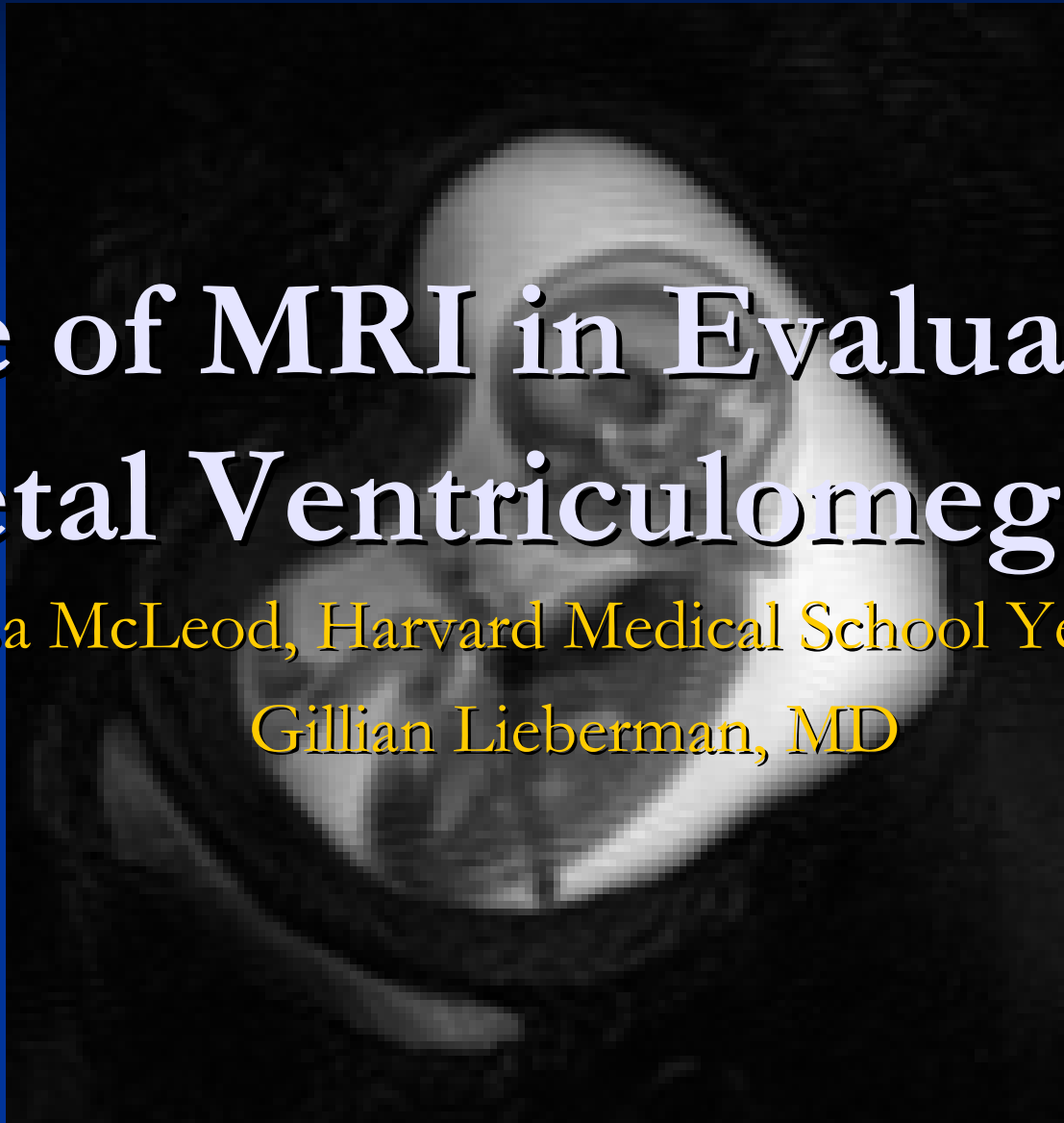
Gillian Lieberman, MD

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Use of MRI in Evaluating Fetal Ventriculomegaly

Lisa McLeod, Harvard Medical School Year III

Gillian Lieberman, MD





Objectives:

- Review *basic* fetal CNS development and neuroanatomy
- Discuss DDx of ventriculomegaly documented on fetal ultrasound
- Illustrate the use of fetal MRI in differentiating these diagnoses and its impact on management
- Identify pros and cons of Ultrasound and MRI for fetal survey
- Future directions of use of fetal MRI in diagnosis of etiology of ventriculomegaly



Landmarks of fetal brain development visible by MRI

■ Glial Cell Migration

- Visible @ 22 weeks GA
- Cells migrate from ventricular periphery toward cortical ribbon
- T2 Hypointense

■ Sulcation/Ventricles

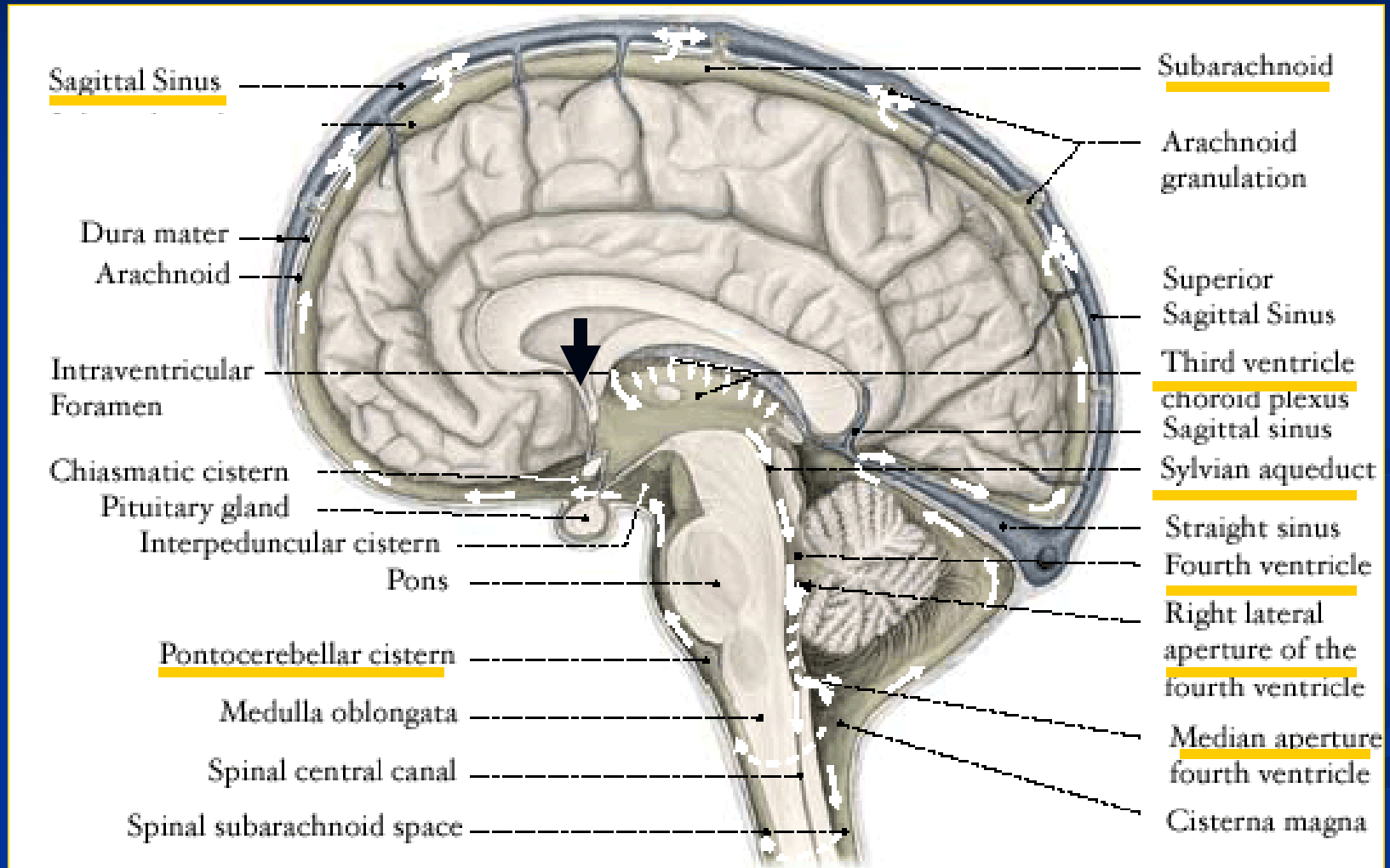
- Agyric (exc. Sylvian) until 24 weeks
- Physio Hydrocephalus resolves from 14 weeks
- Both T2 Hyperintense

■ Axonal Maturation/Myelination

- Caudal-cephalic/Dorsal-ventral
- T2 Hypointense



Ventricular CSF Circulation





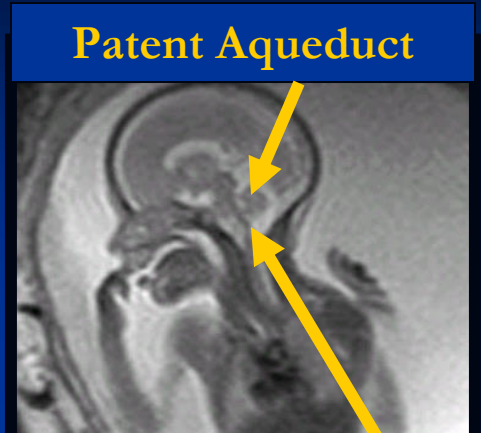
17 weeks to 23 weeks GA

- ❖ Increase sulcation (calcarine, parieto-occipital)
- ❖ Cell migration creates Intermediate layer between germinal matrix and cortical ribbon
- ❖ Reduced Ventricle size
 - ❖ Megendi & Lushka form allowing CSF flow to subarachnoid
 - ❖ Midline structures further reduce ventricle size (i.e. Corpus Call, Sept. Pallucidum)



Corpus callosum

BIDMC



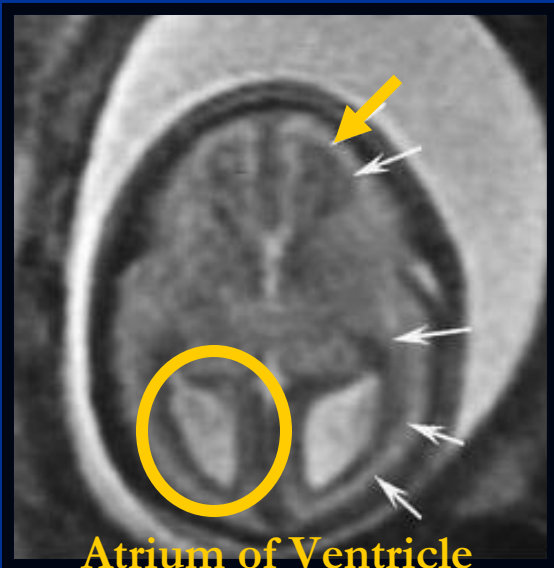
Patent Aqueduct

Brainstem Myelination

BIDMC

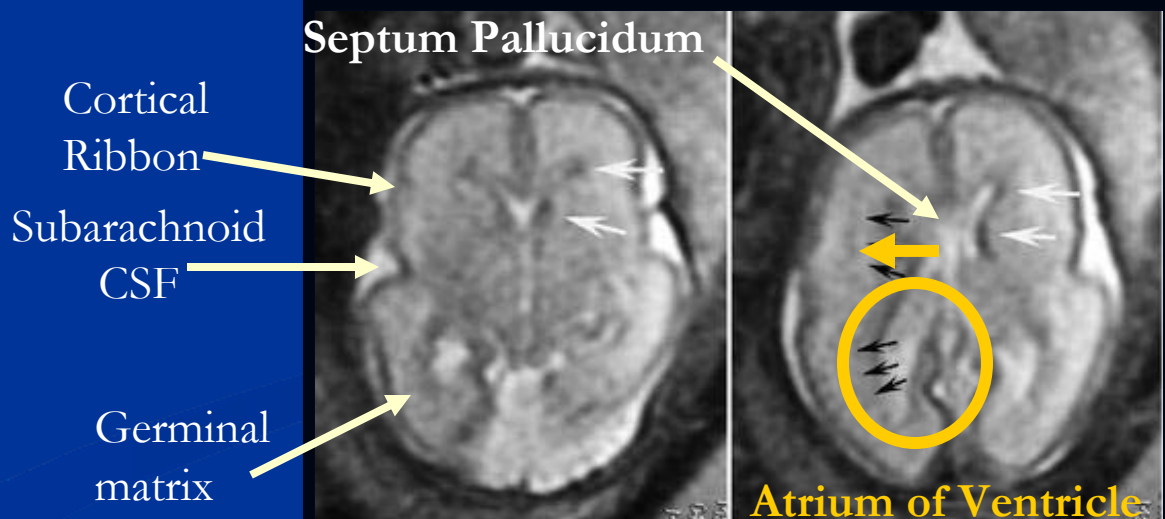
- ❖ Lower Brainstem Myelination

NL 17 Wk Fetus



Atrium of Ventricle

NL 23 Wk Fetus



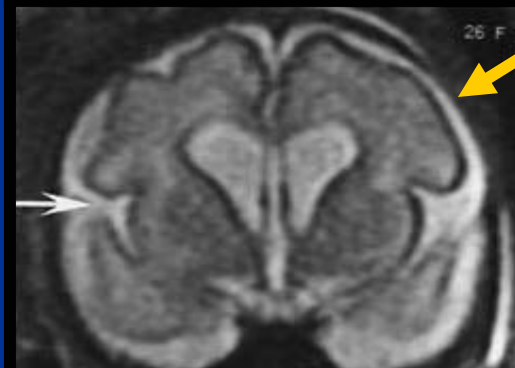
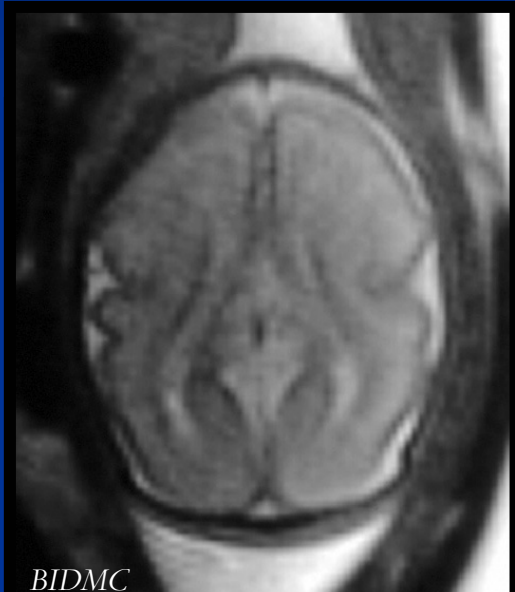
Septum Pallucidum

Atrium of Ventricle



28 Weeks to 33 Weeks GA

NL 28Wk Fetus



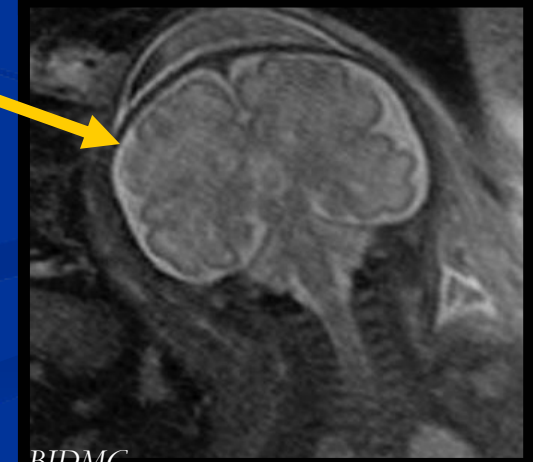
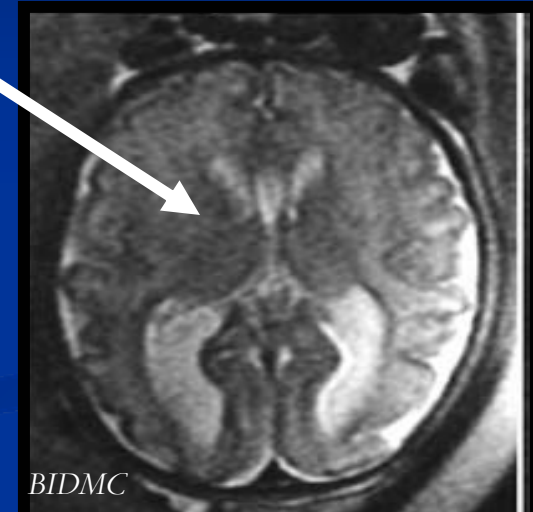
❖ Increased Axonal Myelination of Basal Ganglia

❖ Increased Sulcation (precentral gyrus, postcentral gyrus, Temporal Sulci)

❖ Maturation of Arachnoid Granulations (less subarachnoid fluid)

❖ Increased Contrast between white and grey matter

NL 33Wk Fetus





Patient K.A.:

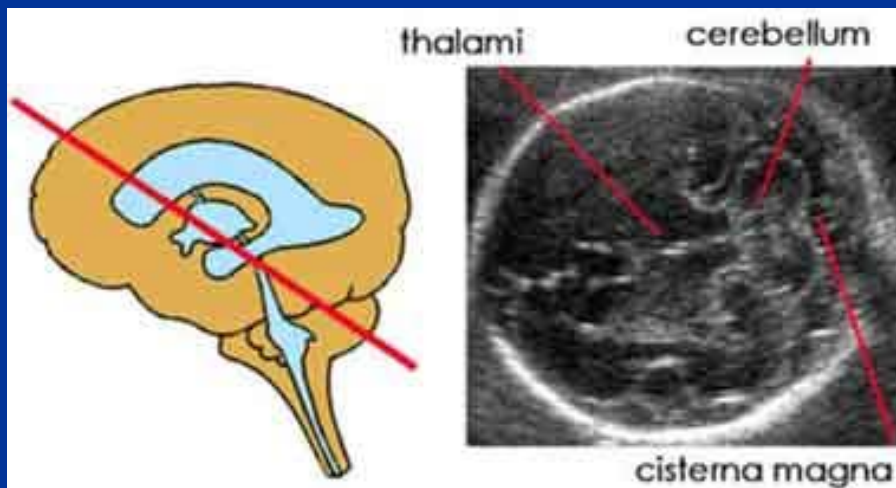
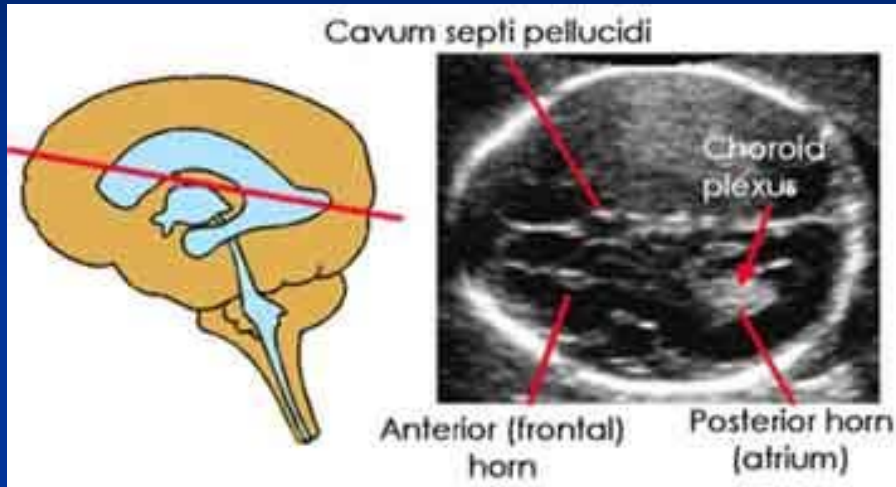
33yo F at 18 weeks GA presents for high risk ultrasound of fetus with h/o choroid plexus cysts at first trimester exam.

➤ Findings this exam:

- Persistence of abnormal choroid plexus
- Mild Borderline Ventriculomegaly (9mm prominent lateral ventricles)
- 7mm Cyst in the Posterior Fossa
- Ventricular Septal Defect

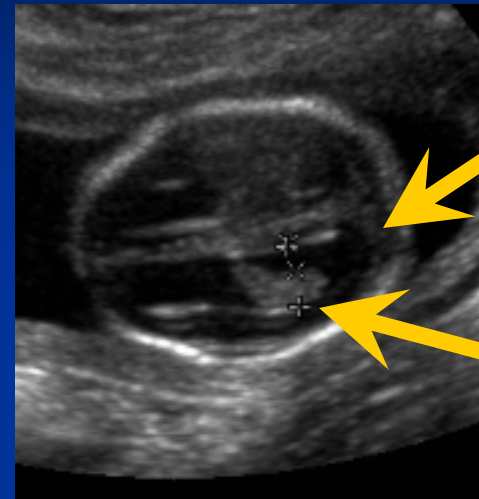


NL Patient 18 weeks



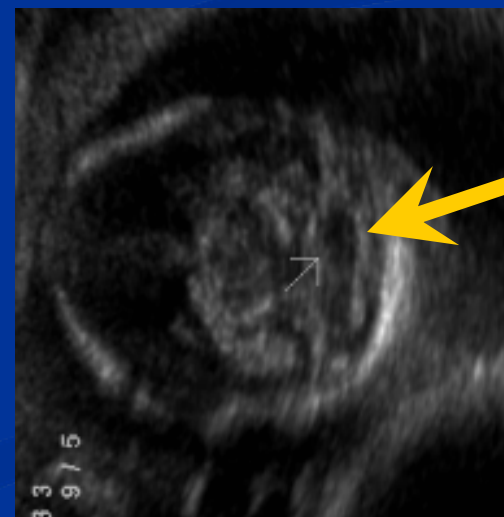
Above from <http://www.centrus.com.br>

Patient K.A. 18 weeks



Prominent ventricular atrium (cursor on medial reflection)

Dangling choroid plexus (>3mm from medial reflection)



Cyst in posterior fossa

Images from BIDMC



■ Ventriculomegaly:

- Defined as enlargement of the ventricles to greater than 10mm without an associated macrocephaly
- Frequency 0.5-2/1000 live births
- Natural History Reversible (29%), Stable (57%), or lead to Hydrocephalus (14%)*
- Prognosis – Highly dependant on etiology
 - Good when no associated malformations present. BUT Ultrasound has a 20-60% false negative rate in diagnosis of associated abnl's.
 - Bad if associated malformations, male gender, severe enlargement (>15mm), extension to 3rd/4th ventricles, or appears early in gestation.

* Values difficult to interpret given number of terminations for this finding.



Etiologies of Ventriculomegaly

- Primary causes:
 - 20% Aqueductal stenosis (*isolated ~18%*)*
 - Myelomeningocele with Chiari malformation
 - Agenesis of the Corpus Callosum (10%)
 - Dandy-Walker malformation (*prognosis variant dep.*) *
 - Holoprosencephaly*
 - Hydranencephaly
 - Lissencephaly
- Secondary causes:
 - Intraventricular hemorrhage
 - Cerebral ischemia
 - Infections (CMV, HSV, Toxo, Varicella)
 - Tumors

**often associated with chromosomal abnl's*

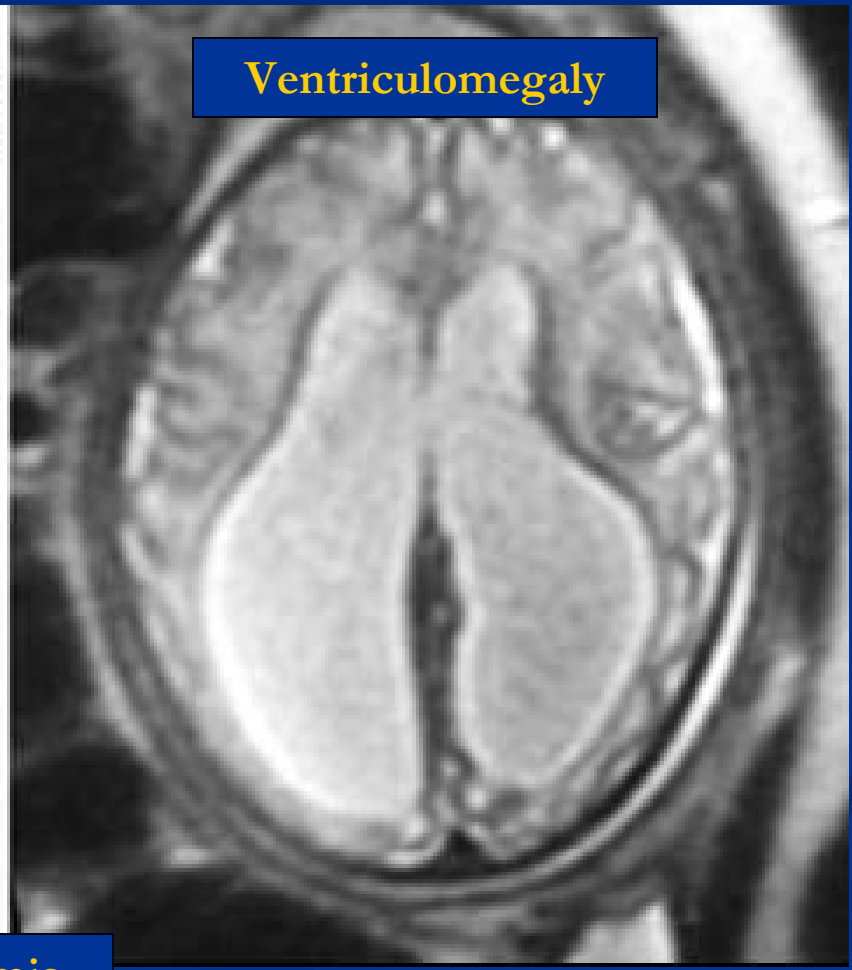
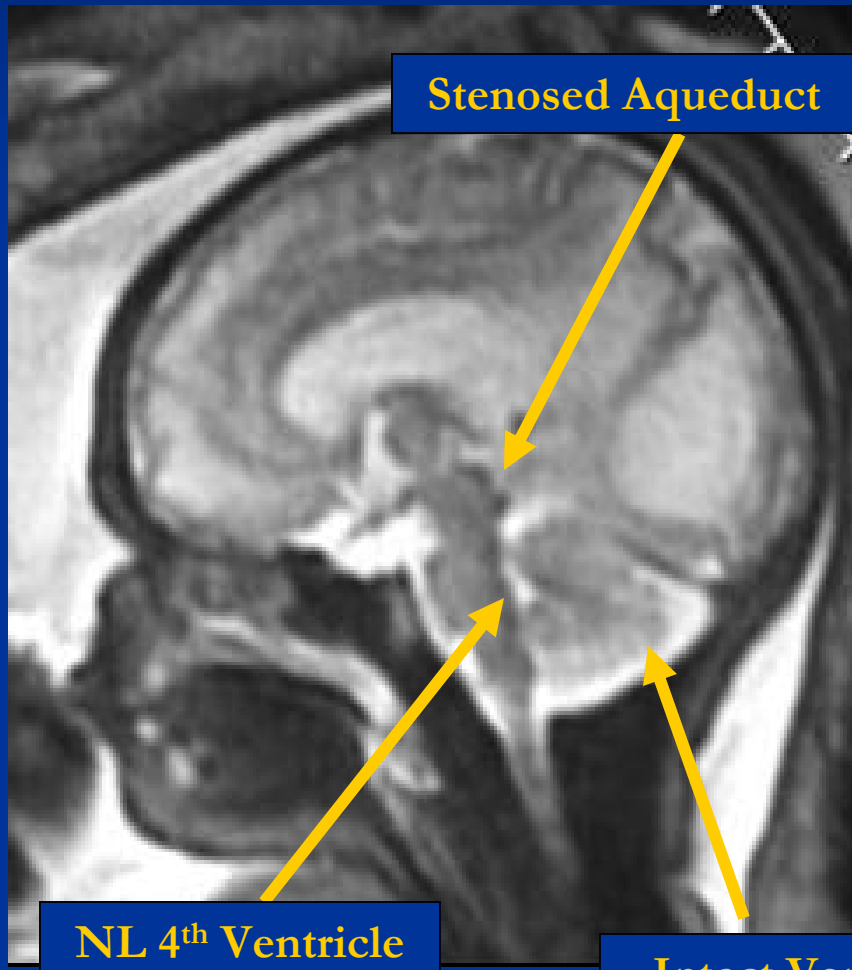


Patient work-up for Ventriculomegaly

- Maternal Blood Tests (Rubella, Parvo, HIV, Torch, anti-platelet abs)
- Karyotype of fetus
- Fetal echocardiogram
- Fetal MRI
 - CNS: Symmetry & Distribution, Cell layers, Choroid, Posterior Fossa, Aqueduct patency,
 - Extracranial: Other signs of aneuploidy

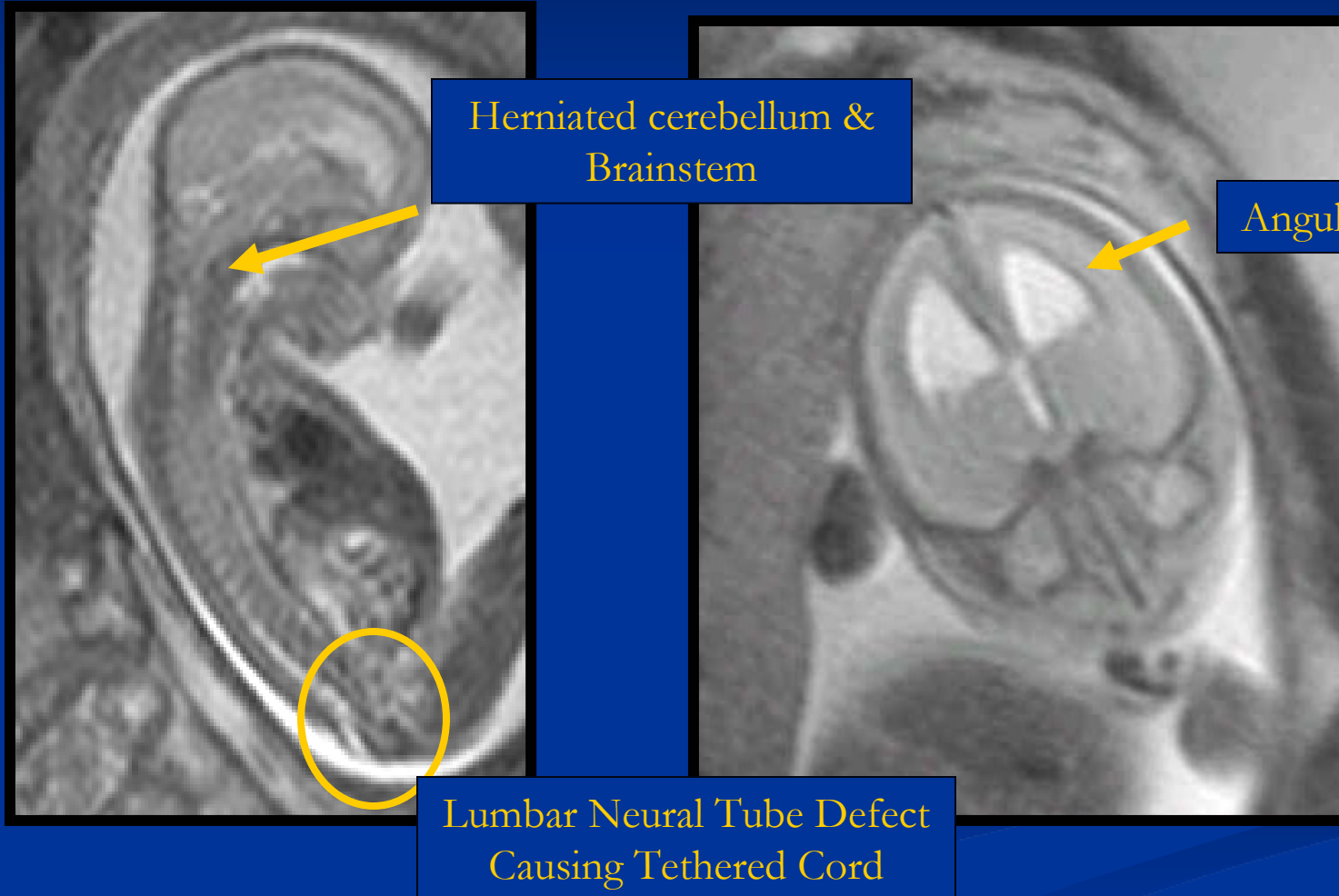


Isolated Aqueductal Stenosis in 32 Week Fetus





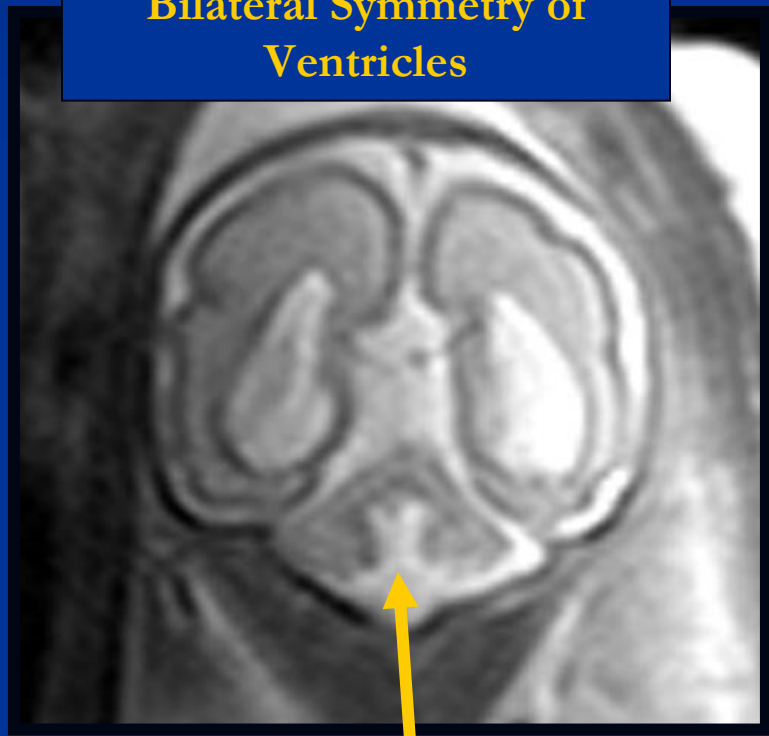
Myelomeningocele with Chiari Malformation in 23 week Fetus





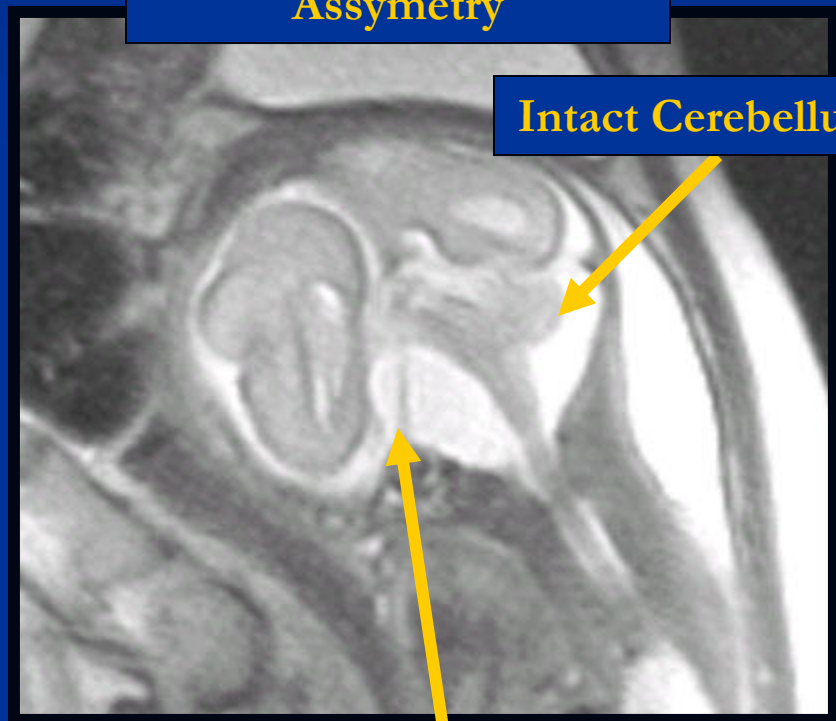
Dandy Walker Variant Vs. Arachnoid Cyst in 26 Week Fetuses

Bilateral Symmetry of Ventricles



Agenesis/Dysgenesis of Cerebellar Vermis

Assymetry

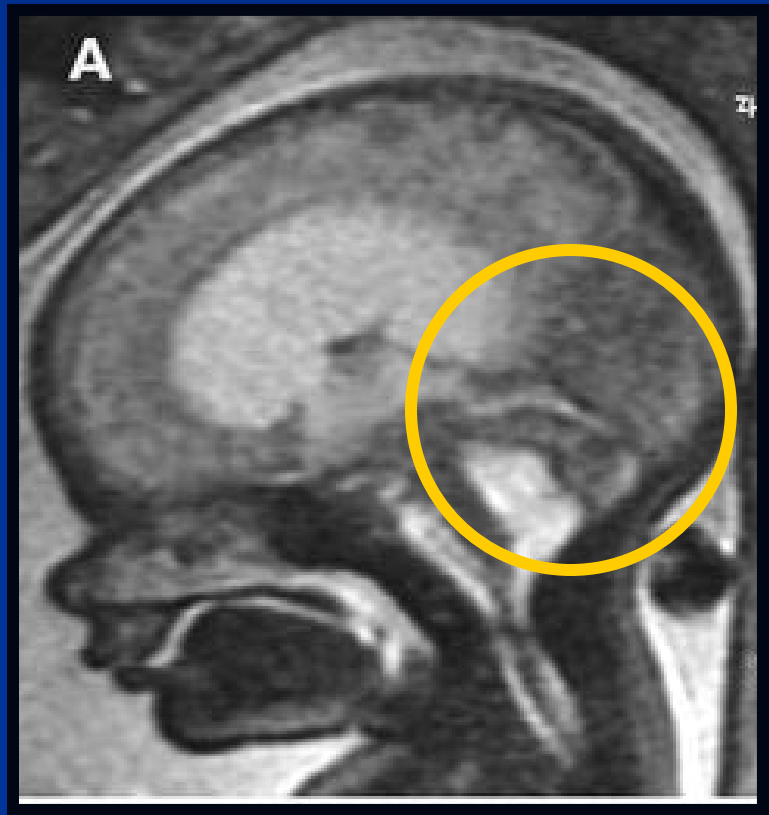


Intact Cerebellum

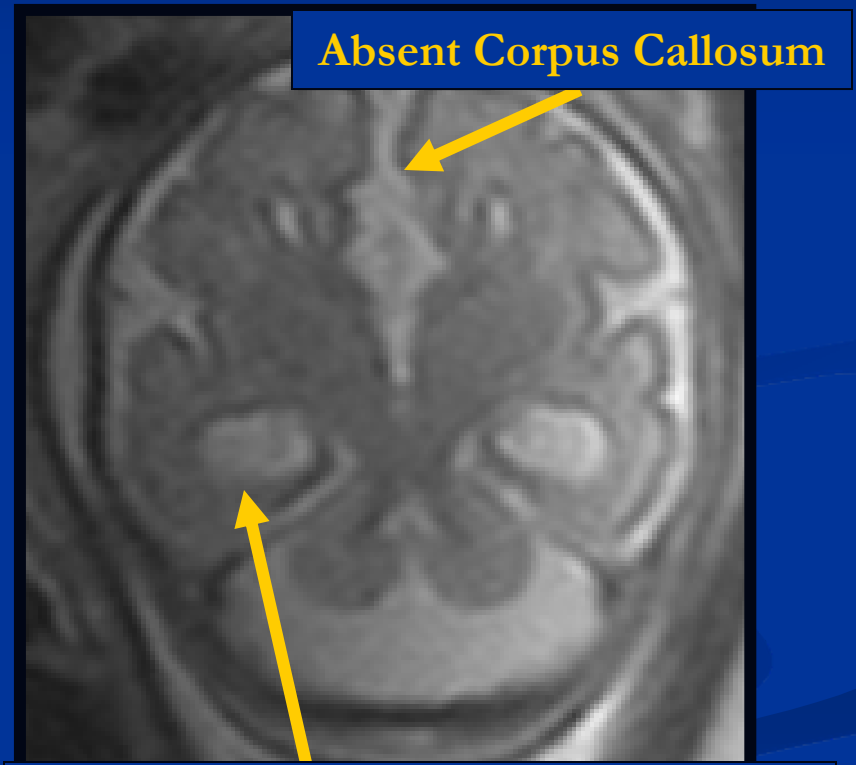
Septation and Mass effect on Adjacent tissues



Hemorrhage Vs. Agenesis of Corpus Callosum in 26 Week Fetuses



Hypointense Parenchyma = Hemorrhage/clot blocking outflow tract



Colpocephaly: Prominent Occipital Horns

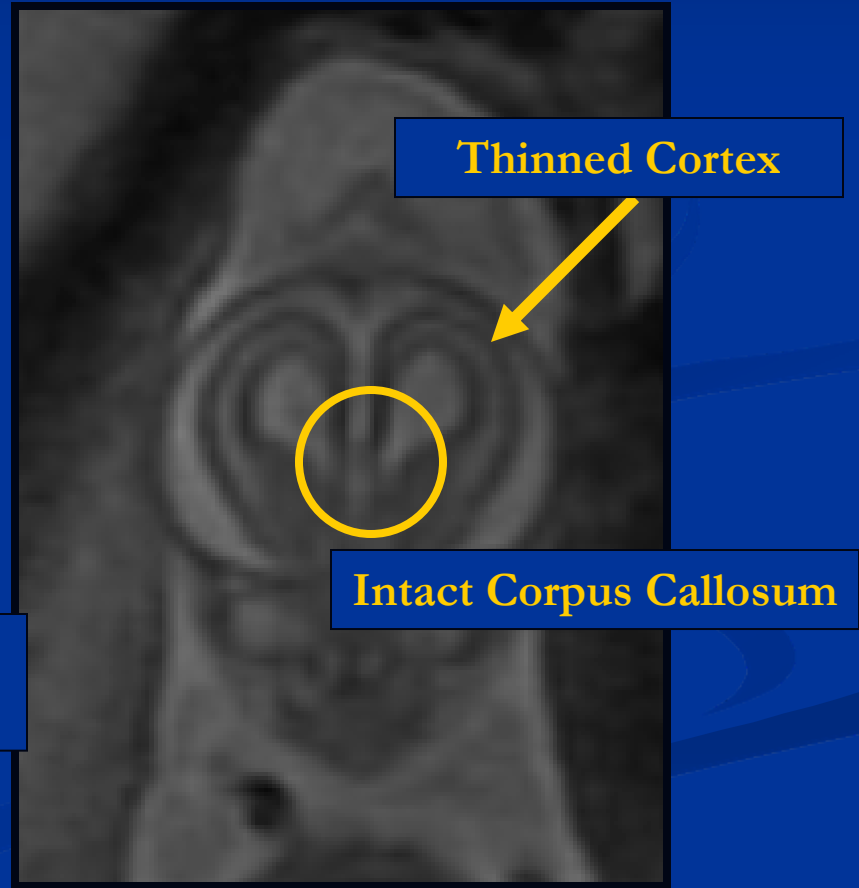
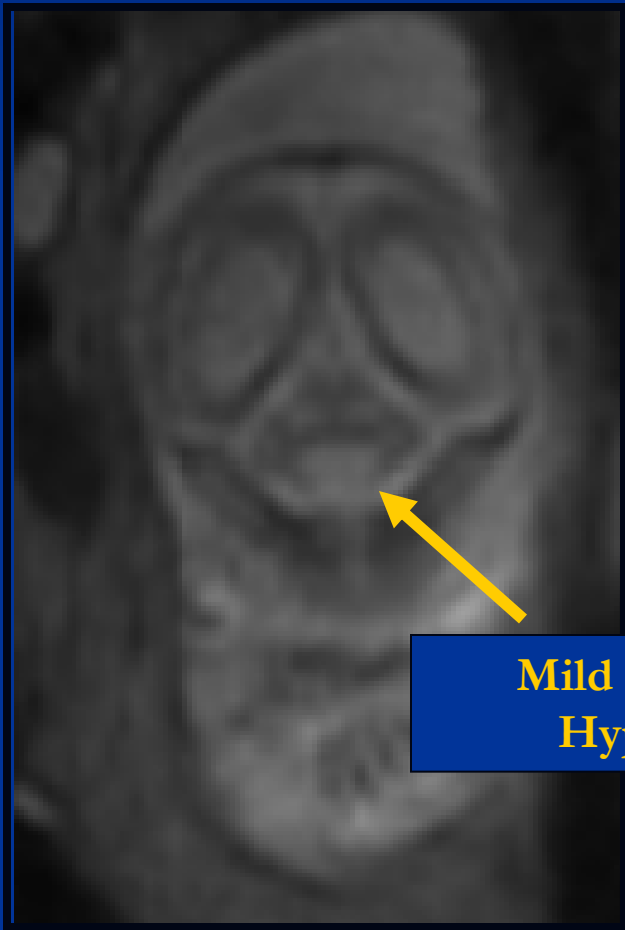


Back to Patient K.A.....

- Posterior fossa difficult to conclusively assess
- What is the origin of the posterior cyst?
- Why are the ventricles so prominent?
- What is this child's prognosis?
- Since ultrasound could not conclusively dx, same day fetal MRI ordered.



Fetal Findings Were: Dandy Walker Variant with Cortical Atrophy





How Should K.A. Be Counseled?

- Depending on mother's wishes, amniocentesis should be recommended
- Dandy Walker variant can have mild prognosis
- Cortical thinning implies perturbed brain development
- Given ventricular prominence plus associated malformations (VSD) prognosis is poor



■ When to use MRI:

- Obese mothers
- Low position of head
- Calcification of cranium
- CNS anomalies not diagnosable by US
- When HASTE ultra fast spin echo MRI available

■ When NOT to use MRI:

- Too much fetal movement
- Suspected cardiac anomalies
- Early gestational age (too many incidental findings)
- Absolute contraindications (claustrophobia, metal)



Future Uses of Fetal CNS MRI:

- Help Guide Patient Counseling When Abnormalities are Found
- New outlook into patient selection for in utero interventions:
 - High probability of good outcome for cases of isolated ventriculomegaly/hydrocephalus
- Useful correlations between Ventricle morphology and underlying soft tissue defects:
 - Colpocephalus → Agnesis of Corpus Call.
 - Angular Anterior Horns → Meningomyelocele
 - Fused Anterior Horns → Absence of Sept pallucidum



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Suggested Reading

SD Brown, Children's Hospital and Massachusetts General Hospital, Boston, MA; JA Estroff and CE Barnewalt, Children's Hospital, Boston, MA. Fetal MRI. *Applied Radiology* 2004; 33(2) 9-25.



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