Use of MRI in Evaluating Fetal Ventriculomegaly

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http://bidmc.harvard.edu/content/departments/radiology/files/fetalatlas/default.htm
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

http://carecure.rutgers.edu/spinewire/Articles/SCIischemia/Sagittal_brain1.gif
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)
- Lower Brainstem Myelination

Lower images from http://www.radnet.ucla.edu/residents/chief/residentrounds1.htm
28 Weeks to 33 Weeks GA

- 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

http://www.radnet.ucla.edu/residents/chief/residentrounds1.htm
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

Patient K.A. 18 weeks

Prominent ventricular atrium (cursor on medial reflection)

Dangling choroid plexus (>3mm from medial reflection)

Cyst in posterior fossa

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

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,
  - Exacranial: Other signs of aneuploidy
Isolated Aqueductal Stenosis in 32 Week Fetus

Images from BIDMC
Myelomeningocele with Chiari Malformation in 23 week Fetus

Herniated cerebellum & Brainstem

Angular Ventrices

Lumbar Neural Tube Defect Causing Tethered Cord

Images from BIDMC
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

Images from BIDMC
Hemorrhage Vs. Agenesis of Corpus Callosum in 26 Week Fetuses

Hypointense Parenchyma = Hemorrhage/clot blocking outflow tract

Absent Corpus Callosum

Colpocephaly: Prominent Occipital Horns

Images from BIDMC
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

- Mild Cerebellar Hypoplasia
- Thinned Cortex
- Intact Corpus Callosum

Images from BIDMC
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 contrindications (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 ➔ Agenesis of Corpus Callosum
  - Angular Anterior Horns ➔ Meningomyelocele
  - Fused Anterior Horns ➔ Absence of Septum Pellucidum
References:

- Oi S Diagnosis, Outcome, and Management of Fetal Abnormalities: Fetal Hydrocephalus Child’s Neuro 19(7-8):508-516
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.
Acknowledgements:

- Dr. Deborah Levine
- Dr. Michelle Swire
- Dr. Ilse Castro-Aragon
- Dr. Gillian Lieberman
- Pamela Lepkowski
- Webmaster Larry Barbaras