ULTRASOUND EVALUATION OF THE FETAL CENTRAL NERVOUS SYSTEM

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OVERVIEW

- Guidelines (AIUM, ISUOG)
- Normal anatomy
- Common abnormal findings
- Specific anomalies

AIUM OBSTETRIC ULTRASOUND PRACTICE PARAMETER (2013)

• Gestational (menstrual) age assessment
  – biparietal diameter (BPD)
    • level of thalami and cavum septi pellucidi or columns of the fornix
    • cerebellar hemispheres should not be visible
    • outer edge proximal to inner edge distal skull
    • comment: head shape may be flattened (dolichocephaly) or rounded (brachycephaly) as a normal variant. Under these circumstances… measurement of head circumference (may be) more reliable than BPD for estimating gest age

AIUM OBSTETRIC ULTRASOUND PRACTICE PARAMETER (2013)

• Gestational (menstrual) age assessment
  – Head circumference
    • same level as biparietal diameter
    • around outer perimeter of calvarium
    • not affected by head shape

AIUM OBSTETRIC ULTRASOUND PRACTICE PARAMETER (2013)

• Fetal anatomic survey
  – Head, face, and neck
    • lateral cerebral ventricles, choroid plexus, midline falx, cavum septi pellucidi, cerebellum, cisterna magna, upper lip
    • comment: a measurement of the nuchal fold may be helpful during a specific age interval to assess the risk of aneuploidy
  – Spine
    • cervical, thoracic, lumbar and sacral spine
ISUOG FETAL CNS GUIDELINES

• Basic examination
  – optimized approach to fetal head and spine evaluation in fetal anatomic survey

• Fetal neurosonogram
  – Detailed evaluation for patients with increased risk for fetal CNS anomalies
  – Requires specific expertise
  – Transvaginal and/or 3-d may be useful

Ultrasound Obstet Gynecol 2007;29:109
Gynecology and Obstetrics

ISUOG FETAL CNS GUIDELINES

Basic examination

(a) transventricular
(b) transthalamic (BPD plane)
(c) transcerebellar

Ultrasound Obstet Gynecol 2007;29:109
Gynecology and Obstetrics

ISUOG FETAL CNS GUIDELINES

Basic examination

• Structures evaluated
  – head shape
  – lateral ventricles
  – cavum septi pellucidi
  – thalami
  – cerebellum
  – cisterna magna
  – spine

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ISUOG FETAL CNS GUIDELINES

Basic examination

• Lateral ventricles
  – size
  – shape
  – borders
  – content

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ISUOG FETAL CNS GUIDELINES

Basic examination

• Cavum septi pellucidi
  – fluid filled cavity between 2 membranes
  – becomes visible at approximately 16 weeks
  – obliterates near term
  – should always be visualized between 18 – 37 weeks (BPD 44-88 mm)
  – altered in several anomalies (holoprosencephaly, agenesis of the corpus callosum, severe hydrocephalus, septo-optic dysplasia)

Courtesy Alfred Abuhamad, M.D.
Gynecology and Obstetrics
ISUOG FETAL CNS GUIDELINES

Basic examination

• Cisterna magna (cisterna cerebello-medularis)
  – fluid filled space posterior to the cerebellum
  – Normally contains thin septations
  – Depth is stable 2nd half of gestation (2 – 10 mm)
  – Prior to 20 weeks cerebellar vermis has not completely covered the 4th ventricle; this may give the false impression of a vermis defect

Gynecology and Obstetrics

Ultrasound Obstet Gynecol 2007;29:109

• Measurements
  – biparietal diameter (BPD)
  – head circumference
  – internal diameter of the atrium
  – ± transverse cerebellar diameter
  – ± cisterna magna depth

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Ultrasound Obstet Gynecol 2007;29:109

• Measurement of the atrium
  – transverse plane
  – atrium distal to the transducer
  – caliper placement
    • level of glomus of choroid plexus
    • perpendicular to ventricular cavity
    • inside echoes generated by lateral walls
  – measurement stable 2nd & early 3rd trimesters (6 – 8 mm, normal < 10 mm)

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Ultrasound Obstet Gynecol 2007;29:109

• Biparietal diameter (BPD)
  – BPD and head circumference may be obtained at transventricular or transthalamic level
  – BPD caliper placement outer-outer or outer-inner depending on normogram used
  – BPD/OFD ratio: 75 – 85%
    • > 85% - brachycephaly
    • < 75% - dolichocephaly

Gynecology and Obstetrics

Ultrasound Obstet Gynecol 2007;29:109
Fetal neurosonogram

- Additional coronal and sagittal planes

Common abnormal findings

- Lateral ventricles: abnormal size, shape, appearance
- Failure to visualize cavum septi pellucidi
- Cerebellum: abnormal size, shape, appearance
- Cisterna magna: abnormal size, shape, appearance
- Abnormal appearance of spine

Ventriculomegaly

- Enlargement of the cerebral ventricles (internal atrial diameter > 10 mm)
- Incidence 0.3 – 1.5 / 1000 livebirths
- Present in a variety of CNS anomalies
- Prognosis depends upon severity and presence of associated abnormalities

Outcomes in 176 cases

<table>
<thead>
<tr>
<th>Atrial Width (mm)</th>
<th>Alive (%)</th>
<th>Normal (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>10 – 12 mm</td>
<td>98%</td>
<td>93%</td>
</tr>
<tr>
<td>12.1 – 14.9 mm</td>
<td>80%</td>
<td>75%</td>
</tr>
<tr>
<td>&gt; 15 mm</td>
<td>33%</td>
<td>62%</td>
</tr>
</tbody>
</table>

Mild ventriculomegaly

- Atrial width 10 – 15 mm
- Good outcome if isolated
- ↑ risk of developmental delay (up to 1/3)
  - may represent early manifestation of brain damage from heterogeneous causes
- ↑ risk of abnormal outcome
  - female fetus
  - atrial width ≥ 12 mm
  - second trimester diagnosis

Overt ventriculomegaly

- Isolated
  - aquaductal stenosis
  - communicating hydrocephalus
  - 70% survival / 54% normal IQ
- Associated anomalies
  - neural tube defects
  - midline anomalies
FETAL CNS ULTRASOUND
Ventriculomegaly management
- Detailed anatomic survey
- Fetal echocardiography
- Fetal karyotype
- Fetal infection evaluation
- Consider MRI
- Serial sonograms
- Counseling

FETAL CNS ULTRASOUND
Abnormal appearance of ventricles
- Increased echogenicity, thickness, irregularity of wall
  - hemorrhage, infection, neuronal migration disorder
- Periventricular echogenicity, echogenic foci, or cysts
  - infection
- Abnormal shape
  - agenesis of corpus callosum (teardrop)

FETAL CNS ULTRASOUND
Failure to visualize cavum septi pellucidi
- Agenesis of the corpus callosum
- Holoprosencephaly
- Septo-optic dysplasia
- Absent cavum septi pellucidi
- Schizencephaly
- Secondary to disruption
  - Aqueductal stenosis, Chiari II malformation, porencephaly, hydranencephaly

FETAL CNS ULTRASOUND
Holoprosencephaly
- Complex brain abnormality caused by failure of cleavage of the prosencephalon
- Prevalence
  - 1/2500 – 1/1600 livebirths
  - 1/250 1st & 2nd trimester terminations
- Spectrum
  - Complete fusion of cerebral hemispheres within single ventricle (alobar)
  - Partial fusion of frontal lobes (lobar)

FETAL CNS ULTRASOUND
Dandy-Walker complex
- Ventriculomegaly
- Large cisterna magna
- Cerebellar vermis defect

FETAL CNS ULTRASOUND
Dandy-Walker complex
- Classic Dandy-Walker malformation
  - enlarged posterior fossa
  - complete or partial agenesis of the vermis
  - elevated tentorium
- Dandy-Walker variant
  - variable hypoplasia of the vermis
  - ± enlargement of posterior fossa
- Megacisterna magna
  - normal vermis and 4th ventricle
FETAL CNS ULTRASOUND
Classic Dandy-Walker malformation

- Mortality up to 25%
- Subnormal intelligence 40 – 70%
- High incidence of associated abnormalities
  - CNS
  - non-CNS
  - abnormal karyotype

FETAL CNS ULTRASOUND
Dandy-Walker complex

- Heterogeneous group
- Poorly correlated with postmortem pathology (~ 50%)
- False positive diagnoses
  - gestational age < 18 – 20 weeks
  - Blake’s pouch
- False negative diagnoses
  - partial vermian agenesis
  - normal cisterna magna