Fetal obstructive hydrocephalus

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Diagnosis, treatment, and long-term outcomes of fetal hydrocephalus

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### Table 2
Classification of fetal hydrocephalus and occurrence rate.

<table>
<thead>
<tr>
<th>Classification of hydrocephalus</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary hydrocephalus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Isolated ventriculomegaly (IVM)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>True IVM</td>
<td>39</td>
<td>25</td>
</tr>
<tr>
<td>Syndromic hydrocephalus</td>
<td>18</td>
<td>12</td>
</tr>
<tr>
<td>Hydrocephalus associated with myelomeningocele</td>
<td>36</td>
<td>23</td>
</tr>
<tr>
<td>Dandy–Walker syndrome and Jobert syndrome</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>Holoprosencephaly</td>
<td>6</td>
<td>4</td>
</tr>
<tr>
<td>Cranial bifida (encephalocele)</td>
<td>9</td>
<td>6</td>
</tr>
<tr>
<td>Hydrocephalus associated with arachnoid cyst</td>
<td>12</td>
<td>8</td>
</tr>
<tr>
<td>Hydrocephalus associated with atresia of Monro</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>Corpus callosum agenesis</td>
<td>8</td>
<td>5</td>
</tr>
<tr>
<td>Fetal secondary hydrocephalus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Post-intracranial hemorrhage</td>
<td>9</td>
<td></td>
</tr>
<tr>
<td>Hydrocephalus associated with brain tumor</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>Post-infectious hydrocephalus</td>
<td>4</td>
<td></td>
</tr>
</tbody>
</table>

Extensive definition
No mention of aqueductal stenosis
Purpose

- Working definition of hydrocephalus vs ventriculomegalies
- Features of fetal obstructive hydrocephalus
- Causal, and causally associated lesions
- Brain plasticity in fetal hydrocephalus
Working definition: hydrocephalus vs ventriculomegaly

1. Disproportionate ventriculomegaly
2. Increased cephalic measurements
3. An identified obstruction

Therefore, **obstructive hydrocephalus** only
Fetal obstructive hydrocephalus: ventricular size

- Measured at the atrium, on largest side
  - normal 5-8mm throughout gestation
- Degree
  - severe >20mm: 40 cases
  - moderate 15-20mm: 11 cases
  - mild 10-15mm: 5 cases
    - 2 DW, 1 familial, 2 F/U confirmed
- Larger when late gestation, always larger on F/U studies
- Often asymmetric
Fetal obstructive hydrocephalus: cranial size

- Hydrocephalus
  - BPD ≥ 2 weeks-above-average 48/56
  - borderline in 8/56 but
    - 1 DW, 2 familial, 4 hemorrhages, 1 F/U
- Head circumference less increased than BPD
  - $HC = \frac{1}{2} (BPD + FOD) \times 3.14$
  - head becomes rounded, “turricephalic”
Fetal obstructive hydrocephalus: case series

- Fetal MR diagnosis: 56 cases
  - 44 “early” cases <32w (19.4w-29.5w)
  - 12 “late” cases >32w (32.4w-38.4w)
- Special context (early gestation only)
  - 1 family (2 siblings)
  - 9 twin gestations (20%) (general population 3.2%)
- Initial diagnosis confirmed by
  - fetopathology: 10 cases
  - pre- or post-natal MR F/Us: 10 cases
Early fetal obstructive hydrocephalus: subtypes

• Mild subtype
  – mantle thin but regular
  – pericerebral spaces maintained
  – septum pellucidum: rupture common

• Severe subtype
  – mantle thin and focally dehiscent
  – pericerebral spaces effaced
  – septum pellucidum: rupture common
Early fetal obstructive hydrocephalus: cerebral mantle

- **Mild subtype**
  - mantle thin but regular
  - pericerebral spaces maintained
  - septum pellucidum: rupture common

- **Severe subtype**
  - mantle thin and focally dehiscent
  - pericerebral spaces effaced
  - septum pellucidum: rupture common
Late fetal obstructive hydrocephalus

Not really different from post-natal ("congenital") hydrocephalus
• 1/12 mild hydrocephalus: superior vermian mass
• 11/12 severe hydrocephalus (partial mantle loss)
<table>
<thead>
<tr>
<th>Etiology</th>
<th>Early: N (%)</th>
<th>Late: N (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isolated aqueductal stenosis</td>
<td>19 (43)</td>
<td>1</td>
</tr>
<tr>
<td>Rhombencephalosynapsis</td>
<td>11 (25)</td>
<td>82%</td>
</tr>
<tr>
<td>Ependymal nodules</td>
<td>6 (14)</td>
<td>1</td>
</tr>
<tr>
<td>Midline cysts</td>
<td>3 (7)</td>
<td>4 (33)</td>
</tr>
<tr>
<td>Hemorrhages</td>
<td>4 (9)</td>
<td>5 (42)</td>
</tr>
<tr>
<td>Others</td>
<td>1 (avf)</td>
<td>1 (mass)</td>
</tr>
<tr>
<td></td>
<td><strong>44 (100)</strong></td>
<td><strong>12 (100)</strong></td>
</tr>
</tbody>
</table>

- Etiologies similar in mild and severe subtypes
- Etiologies of late hydrocephalus same as “congenital” hydrocephalus
Early fetal obstructive hydrocephalus: etiologies

- Aqueductal stenosis in 82%
  - isolated 43%
  - associated with other abnormalities
    - rhombencephalosynapsis (& midbrain, oosterior thalamus)
    - ependymal nodules
- Midline cyst, hemorrhage much less common
• Isolated aqueductal stenosis common = 43%
  – classic explanations
    • secondary to hydrocephalus
    • undocumented TORCH, hemorrhage, dormant mass
    • maldevelopment (D. Russel, DH Padget)
  – recent literature data: animal, human fetus
    • ependymal denudation +/- SCO changes
      – ependymal nodules (6 cases) (Ferland 2011, Rodriguez 2102)
      – factor of rupture of septum pellucidum?
      – cause or effect of hydrocephalus?
Ependymal denudation, SCO and nodules

Rhombencephalosynapsis

- Rhombencephalosynapsis (associated with aqueductal stenosis)
  - fused cerebellar hemispheres & dentates, no vermis
  - fused colliculi, posterior thalami
- Present in 11/44 early fetal hydrocephalus (25%)
  - otherwise very rare: 100 cases published (Passi 2015)
Rhombencephalosynapsis

  - mesencephalo- / diencephalosynapsis, aqueductal abnormalities

  - 42 postnatal cases

  - 20 postnatal cases
Early fetal hydrocephalus: evolution

- Ventriculomegaly increases (in weeks-above-average)
- Morphotype – mild or severe – does not change with advancing gestation
- Fetal hydrocephalus does not prevent gyration to develop
  - except for the dehiscent mantle segment
- When treated early, fetal hydrocephalus seems to recover a reasonable mantle thickness
  - except for the dehiscent mantle segment
  - cellular lineages?
Early fetal aqueductal stenosis

Late fetal aqueductal stenosis
Hypomyelination

Plasticity: mantle restoration in early hydrocephalus

Neonatal (early fetal) aqueductal stenosis
Poor myelination
Early hydrocephalus and brain plasticity

- Persistent expression of signaling pathways for axonal growth and branching
- Axonal progression and branching mostly subcortical
- Lack of myelination
  - myelin: most potent inhibitor of axonal development
  - myelin-associated inhibitors limit axon development

Fetal hydrocephalus: summary

- Fetal hydrocephalus triad
  - ventriculomegaly, increased BPD, obstruction
    - ? communicating hydrocephalus
- Aqueductal stenosis, rhombencephalosynapsis & ependymal nodules surprisingly common
- Pathology: ependymal denudation common
  - cause vs effect of hydrocephalus
  - ependymal nodules & ruptured septum pellucidum
- Good morphologic recovery possible (role of lack of myelination?)