Fetal Ultrasound Buffet: Bananas and Lemons of Arnold-Chiari Malformation

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Our Patient: Clinical Presentation

- 25 yo female
- Irregular cycle since August
- Suspected pregnancy, G1P0?
Diagnostic Imaging: Fetal US

Fetal Ultrasound

• Sound waves
• Transducer emits and receives sound waves
• 1st Trimester: transabdominal and transvaginal
• 2nd and 3rd Trimesters: transabdominal

Patient Preparation:

• 1st trimester: full urinary bladder to displace gas
• 2nd and 3rd trimesters: no prep necessary

Patient Comfort Level:

• Generally painless; minor discomfort with transvaginal US

Contraindications:

• No contraindications to obstetrical US

The Wishard: http://www.wishard.edu/health-library/Content?contentTypeID=92&contentId=P09031
Diagnostic Imaging: Fetal US

Fetal US examination needs to be standardized based on guideline created and revised by:

- American Institute of Ultrasound in Medicine (AIUM)
- American College of Radiology (ACR)
- American College of Obstetrics and Gynecology (ACOG)
- Society of Radiologists in Ultrasound (SRU)
Our Patient: Indications for US during 1st trimester

1. Confirmation of the presence of an intrauterine pregnancy
2. Evaluation of a suspected ectopic pregnancy
3. Defining the cause of vaginal bleeding
4. Evaluation of pelvic pain
5. Estimation of gestational (menstrual) age
6. Diagnosis or evaluation of multiple gestations
7. Confirmation of cardiac activity
8. Imaging as an adjunct to chorionic villus sampling, embryo transfer, and localization and removal of an IUD
9. Assessing for certain fetal anomalies in high risk pts
10. Evaluation of maternal pelvic masses and/or uterine abnormalities
11. Measuring the nuchal translucency (NT) when part of a screening program for fetal aneuploidy
12. Evaluation of a suspected hydatidiform mole
Our Patient: Findings on fetal US during 1st trimester

- Intrauterine gestational sac
- Single living embryo
- Gestational sac (4wk), yolk sac (5wk), heart beat (>6wk)
- Crown-rump length (CRL) = 12mm = gestational age 7w4d

*Normal: 5mm to 12mm at 7wk
Diagnostic Imaging: Fetal US during 2\textsuperscript{nd} & 3\textsuperscript{rd}

- Fetal anatomy can be adequately assessed > 18wks
  - Limiting factors: fetal size, position, movement, abdominal scars, maternal abdominal thickness

Standard Examination according to the OB US Guidelines (2013):

<table>
<thead>
<tr>
<th>Head, face, and neck</th>
<th>Chest</th>
<th>Others</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lateral cerebral ventricles</td>
<td>Heart</td>
<td>Spine</td>
</tr>
<tr>
<td>Choroid plexus</td>
<td>Four-chamber view</td>
<td>Extremities</td>
</tr>
<tr>
<td>Midline falx</td>
<td>Left ventricular outflow tract</td>
<td>Sex</td>
</tr>
<tr>
<td>Cavum septi pellucidi</td>
<td>Right ventricular outflow tract</td>
<td></td>
</tr>
<tr>
<td>Cerebellum</td>
<td>Abdomen</td>
<td></td>
</tr>
<tr>
<td>Cistern magna</td>
<td>Stomach (presence, size, situs)</td>
<td></td>
</tr>
<tr>
<td>Upper lip</td>
<td>Kidneys</td>
<td></td>
</tr>
<tr>
<td>Nuchal fold* if indicated</td>
<td>Urinary bladder</td>
<td></td>
</tr>
</tbody>
</table>

Abdomen
- Umbilical cord insertion site
- Umbilical cord vessel number
Fetal US: Head examination during 2\textsuperscript{nd} & 3\textsuperscript{rd} trimester

Three planes of view of the head:
- Transventricular
- Transthalamic
- Transcerebellar

Anatomical structures to examine:

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International Society of Ultrasound in Obstetrics and Gynecology Education Committee, 2007
Our Patient: Transventricular plane at 17wk

Pause to view the image and continue to see the findings.
Our Patient: Transventricular view findings

- Lateral ventricle width: 11.4mm
- Dangling choroid plexuses

Cardoza et al., 1988
Dangling Choroid Plexuses: Ventriculomegaly

Ventriculomegaly: lateral ventricles greater than 11mm wide

Our Patient

Cardoza et al., 1988
Our Patient: Transventricular view findings

Lemon Sign: Frontal bones lose their normal convex contour and appear flattened or inwardly scalloped

ScienceKids: http://www.sciencekids.co.nz/experiments/invisibleink.html
Our Patient: Transcerebellar plane at 17 wk

Pause to view the image and continue to see the findings.
Our Patient: Transcerebellar plane findings

Banana Sign: Anterior curving of the cerebellum due to small posterior fossa
Our Patient: Obliterated cisterna magna

Banana Sign = Small Posterior Fossa

Spina Bifida? (Lemon sign)

Small Posterior Fossa
   (Banana sign)

Chiari II Malformation
   (Arnold-Chiari Malformation)

Ventriculomegaly
   (Dangling CP)

Definitions

- Symptomatic hindbrain herniation
  - Contents herniate through foramen magnum
  - Cerebellar compression
- Almost always associated with myelomeningocele
Disease: Pathophysiology of Chiari II

Oligo-CSF theory
1. Myelomeningocele
2. Leakage of CSF
3. Insufficient CSF volume to extend ventricular system
4. Small posterior fossa

Crowding theory
1. Restricted growth of posterior fossa
2. Squeezed through foramen magnum

Hydrodynamic pulsion theory
1. Early progressive hydrocephalus
2. Cerebellum pushed down
Disease: Chiari II Malformation

Classic findings on fetal US

- Posterior fossa compression
- Small or obliterated cisterna magna
- "Banana" sign if severe
- Frontal bone concavity ("lemon" sign)
- Mild ventriculomegaly in 50%
- Open neural tube defect (ONTD)

Image Interpretation Pearls

Don’t wait for the banana sign!

ONTD harder to find than cranial findings, Consider MR if ONTD not seen
Imaging Algorithm for Chiari II

- US finding of ventriculomegaly/hydrocephalus detected
  - US finding of myelomeningocele detected
    - CMII
  - US finding of myelomeningocele absent
    - MRI
      - Tight posterior fossa detected
        - Myelomeningocele detected
          - CMII
          - CMII (with open defect)
        - Myelomeningocele absent
          - CMII not likely
            - Other than CMII (craniosynostosis) (swollen hindbrain) (tumor)

US: ultrasonography, CMII: Chiari type II malformation

Ando et al., 2007
Imaging Algorithm for Chiari II: Our Patient

US finding of ventriculomegaly/hydrocephalus detected

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      - (craniosynostosis)
      - (swollen hindbrain)
      - (tumor)

- US finding of myelomeningocele absent
  - MRI
  - Tight posterior fossa absent
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US: Ultrasonography, CMII: Chiari type II malformation

Ando et al., 2007
Disease: DDx given the 2nd trimester US findings

**Aqueductal Stenosis**
- Obstruction of aqueduct of Sylvius
- Noncommunicating hydrocephalus
- Posterior fossa remains normal

**Craniosynostosis**
- Premature suture fusion
- Spine most often normal

**Dandy-Walker**
- Partial or complete agenesis of cerebellar vermis
- 4th ventricle communicates with cisterna magna
- Cisterna magna is enlarged
Companion Patient: Dandy-Walker Malformation

- Partial or complete agenesis of cerebellar vermis
- 4th ventricle communicates with cisterna magna
- Cisterna magna is enlarged
Chiari II: Epidemiology

Age

- Advanced maternal age at slightly higher risk
  - ≥ 35 years at time of delivery
  - Secondary to association with T18 and T13

Epidemiology (U.S. data)

- 1 in 25,000-35,000 LB
- 3% of all spontaneous abortions
- 1-2% recurrence risk
Chiari II: Natural History

High morbidity and mortality
- 35% LB die within first 5 years
- 50% with IQ < 80

Obstructive hydrocephalus

Musculoskeletal dysfunction
  25% complete lower limb dysfunction

Gastrointestinal/genitourinary dysfunction
  Only 17% with normal continence
Chiari II: Available Treatments

**Cesarean section delivery at term**
- ↓ infection rate
- ↓ meningomyelocele sac rupture rate

**Immediate postnatal ONTD surgery**
- Cover exposed spinal cord

**In utero surgery**

**Pros:**
- Chiari II can reverse
- ↓ shunt dependence

**Cons:**
- Paralysis and continence rates unchanged
- ↑ preterm delivery risk
Chiari II: Available Treatment – In utero surgery

Fig. 1. Photographs. A: Fetal MMC exposed through a hysterotomy. The placode is oriented in a vertical direction and is surrounded by a relatively large CSF-filled sac. The arachnoid over the lateral margins of the sac is extremely thin and translucent. Uterine staples can be seen at the edge of hysterotomy. B: The skin is closed primarily with a running absorbable suture. C: After delivery, the skin has healed completely. The associated skin hemangioma is visible surrounding the midline incision.

Gupta et al., 2012
Chiari I

Tonsilar herniation

Chiari III

Portion of cerebellum or brainstem herniate into high cervical encephalocele (6yo female – cerebellum, occipital lobe)

Thomas et al, 1999 (Chiari I) and Rutherford et al., 2001 (Chiari III)
Acknowledgments

- Dr. Pauline Bishop
- Dr. Gillian Lieberman
- Ms. Claire Odom
References

Images

  <http://commons.wikimedia.org/wiki/File:Cervelletto_e_cisterna_magna_ecografia_ad_ultrasuoni_Dr._Wolfgang_Moroder.jpg>.
  <http://www.wishard.edu/health-library/Content?contentTypeId=92&contentId=P09031>.