Neonatal Cranial Sonography
Congenital Anomalies

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GOALS
• Describe stages of brain development
• Review common congenital lesions
  – Clinical and pathologic features
  – Sonographic findings
  – CT and MR correlation

Stages of Brain Development
• CYTOGENESIS  molecules into cells
• HISTOGENESIS  cells into tissues
• ORGANOGENESIS  tissues into organs
  – Neural tube closure  3-4 weeks
  – Diverticulation   5-6 weeks
  – Neuron migration   2-6 months
  – Organization      (6 mos-postnatal)
  – Myelination       (postnatal)

Tube Closure Defects
• Anencephaly
• Encephalocele
• Chiari II malformation
• Dandy-Walker
• Callosal agenesis

Anencephaly
• Absent cranium and cerebral hemispheres
• Lethal
  – 75% still born
• Diagnosis with prenatal US

Encephalocele
• Herniated meninges or brain tissue through a calvarial defect
  – occipital 70%
  – frontothemoidal or sphenoidal (30%)
• US: CSF filled scalp mass
Occipital Encephalocele

Chiari II Malformation: Clinical
- Presents in neonates
- Clue-Meningocele—“Always” Present

Chiari II Malformation: Anatomy
- Caudally displaced cerebellum and vermis
- Compressed 4th ventricle
- Hydrocephalus
  - especially after myelomeningocele repair

Chiari Malformation

Dandy-Walker Malformation
Clinical Features in the Neonate
- Increased head size (50% birth)
- Signs of hydrocephalus
- Bulging occipital bone
- CNS anomalies
- Renal anomalies
Dandy-Walker Malformation

- **Classic**
  - Large posterior fossa
- **Variant**
  - Normal posterior fossa volume

Dandy-Walker Malformation

- Cystic dilatation 4th ventricle, hypoplastic vermis & cerebellum, large posterior fossa

Dandy-Walker Variant

- Mildly enlarged 4th ventricle, inferior vermis hypoplasia, normal posterior fossa volume

Dandy-Walker Variant

- Image via Mastoid Foramen
  - Normal inferior vermis & cerebellum

Callosal Agenesis

- Result of ischemic or infectious insult
- Absence may be complete or partial
  - posterior absence in partial agenesis
- Clinical findings
  - asymptomatic if isolated
  - seizures, other CNS anomalies

Callosal Anatomy

- Normal
- Agenesis

bundles of Probst
**Callosal Agenesis: US**

- Absent corpus callosum
- Widely separated lateral ventricles
- Colpocephaly (dilated occipital horns)
- High-riding 3rd ventricle
- “Sunburst” sulcal pattern on medial surface of hemispheres

**Congenital Brain Malformations**

- **ORGANOGENESIS** tissues into organs
  - Neural tube closure 3-4 weeks
  - Diverticulation 5-6 weeks
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  - Organization (6 mos to postnatal)
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**Holoprosencephaly**

- Failure of differentiation and cleavage of the prosencephalon (forebrain)(5th week)
- Associations: trisomy 13 (70%) and 18
- DeMyer classification:
  - alobar, semilobar, lobar

**Alobar Holoprosencephaly**

- Most severe
- Pancake-like cerebrum
- Single ventricle
- Fused thalami
- Absent third ventricle
- Normal posterior fossa
Alobar Holoprosencephaly

- “Face predicts the brain”
- Microcephaly
- Midline facial deformities
- Hypotelorism
- Cyclopia

DDX: Hydranencephaly

- Fluid-filled supratentorial sac
- Falx and fissures present
- Normal basal ganglia

Semilobar Holoprosencephaly

- Intermediate severity
- Anterior brain, frontal horns, thalami still fused
- Separate occipital horns
- Small 3rd ventricle
- Mild facial anomalies

Semilobar Holoprosencephaly

- Fused frontal horns
- Small 3rd vent
- Occipital horn

Lobar Holoprosencephaly

- Mildest form
- Division nearly complete
- Fused frontal horns
- Small frontal lobes
- Need CT/MR for diagnosis
Lobar Holoprosencephaly

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Disorders of Neuronal Migration

- Lissencephaly
- Schizencephaly
- Heterotopias
- (Polymicrogyria)
- Retardation, seizures

Lissencephaly

- "Smooth brain"
  - No or few gyri
- US/MR features
  - Smooth cortex
  - No gyri or sulci
  - Thick gray matter
  - Wide Sylvian fissures

Schizencephaly

- Clefs from lateral ventricles to cortical surface
  - gray matter lined
- In-utero porencephaly
- Two types:
  - Closed lip
  - Open lip

Neurons migrate from lateral ventricle to cortex

Unilateral Schizencephaly
- Fluid filled cleft
- Absent septum pellucidum

Gray Matter Heterotopia
- Normal gray matter not located in cortex
  - Subependymal
  - Subcortical
- Present with seizures

Summary
- Lesion type varies with stage of brain development
- US useful for diagnosis of errors in early development
- CT/MRI indicated for diagnosis of errors in tissue organization and myelination