12. NEUROLOGICAL ANOMALIES

Petra Klinge, M.D.

TOPICS
- Myelodysplasia - Open vs. Closed neural tube defects
- Hydrocephalus - Congenital vs. Acquired

I. MYELOMENINGOCELE (MMC)/OPEN NEURAL TUBE DEFECT

- Single most common congenital defect of the central nervous system
- 4.5/10,000 live births
- 1500 cases/year despite dietary folate supplementation (50% reduction)
- $200,000,000 health care dollars/year

A. MMC - Embryogenesis

- Initial closure of neural tube - day 21-23
- Cranial neuropore closure - day 23-25
- Caudal neuropore closure - day 25-27
- Spinal occlusion and initial ventricular expansion - day 25-32
- Secondary Neurulation - Caudal cell mass, Cavitation/retrogressive differentiation - day 27-54

B. Unified Mechanism

D.G. McLone, M.D., Ph.D.

- Open neural tube defect and leak of CSF into amniotic fluid - AFP positive
- Loss of IV ventricular dilatation and expansion of rhombencephalon/posterior fossa
- Small posterior fossa and creation of Chiari II malformation (Arnold Chiari malformation)

C. MMC

- Open neural tube defect - exposed neural placode
- Chiari II malformation - Tectal beak, descent of IV ventricle, vermis herniation, medullary kink
- Hydrocephalus, 85% require VPS

D. MMC - Surgical Principals

- Closure of open defect/MMC - 24-72 hours, minimizing risk of meningitis
- CSF shunt/diversion for control of hydrocephalus, 80-90% cases
Spinal Dysraphism and Hydrocephalus: Neurosurgery in the Neonate

(Continuation of Surgical Principals)
• Chiari II decompression for stridor, airway obstruction, vocal cord paresis (less than 20%)

E. Closure of MMC
• Start IV antibiotics after birth
• Cover neural placode with moist telfa and plastic wrap (saran). Keep moist
• Avoid pressure to back. No peeking!
• Family discussion with true objective
• Planned, elective surgical procedure

F. Fetal MMC Repair

Fetal MMC Repair 2

Fetal MMC Repair 3: The MOMS trial and beyond

G. Encephalocele
• 1/5000 live births
• 3/4 females are occipital
• Basal, Syncipital and Convexity
• Chiari III-Cervicoocipital encephalocele with herniation of cerebellum/brainstem
• Progressive Hydrocephalus after repair
• Variable outcome

II. Closed NTD/Tethered Cord Syndrome
• Lipomyelomeningocele
• Split Cord Malformations
• Dermal Sinus Tract
• Fatty Filum
• Caudal Regression Syndrome

**Spinal Dysraphism & Hydrocephalus: Neurosurgery in the Neonate**

**A. Cutaneous Signatures**

• Hypertrichosis/hairy patch
• Dimples/Sinus tract/Benign pit
• Nevus
• Capillary hemangioma
• Lipoma/Caudal appendage/tail

**B. Lipomyelomeningocele**

• All tethered, syringomyelia common
• Chapman Classification - Dorsal, Terminal, Transitional
• Insidious neurologic deterioration- Leg weakness/atrophy, gait disturbance, sensory deficits, neurogenic bowel and bladder
• Release prior to standing (3-6 mos)

**C. Split Cord Malformation**

• Pang Classification - Type I and II
• Tethered in 2 locations
• Hypertrichosis common finding
• Hemicord may be open NTD, asymmetric lower extremity findings

**D. Dermal Sinus Tract**

• Dermal Sinus vs. Benign coccygeal pit
• Risk of meningitis/conus abscess
• Repair early/Video

**E. Fatty Filum**

**III. Hydrocephalus**

• Progressive Ventricular enlargement with increased intracranial pressure (ICP)
• Non-Communicating - Aqueductal Stenosis - Tumor - Hemorrhage - Myelodysplasia
• Communicating - Meningitis - Sinus Thrombosis

**A. Treatment Goals**

• Decrease ICP to safe, if not normal, values
• Increase the volume of brain tissue (3.5 cm of cortical mantle) maximizing the child’s potential for intellectual, emotional or motor development
• Minimize the frequency and severity of ICP elevation
• Minimize the likelihood of tx complication
• Maintain integrity of CSF pathways, if possible

*Spinal Dysraphism and Hydrocephalus: Neurosurgery in the Neonate*

**B. Surgical Procedures**

• Rickham Reservoir/Ventricular Taps
• External Ventricular Drain (EVD)
• CSF Shunt - Ventriculoperitoneal, atrial, pleural
• Third Ventriculostomy

**C. VPS**

• Minimize risk of infection
• Choose appropriate site
• Match age and weight to valve type
• Follow CSF chemistry
• Schedule electively, if possible
• Programmable vs. non-programmable valves

**D. Third Ventriculostomy**

• Non-communicating Hydrocephalus
• Late vs. early Aqueductal stenosis
• Surgical risk - Basilar Artery, Hypothalamic/Pituitary injury, Optic Nerve injury
• Post op Ventricular dilatation