

12. NEUROLOGICAL ANOMALIES

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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

- Myelomeningocele Repair in utero: A Report of 3 Cases. Tulipan N, Bruner JP., *Pediatr Neurosurg.* 1998, 28:177
- Reduced Hindbrain Herniation after Intrauterine MMC Repair: A report of 4 Cases. Tulipan N, Hernanz-Schulman M, Bruner JP. *Pediatr Neurosurg.* 1998, 29:274

Fetal MMC Repair 2

- Improvement in Hindbrain Herniation Demonstrated by Serial Fetal MRI Following Fetal Surgery for MMC. Sutton LN, Adzick S, et al. *JAMA.* 1999, 383:1826
- Fetal Surgery for MMC and the Incidence of Shunt-Dependent Hydrocephalus. Bruner JP, Tulipan N, et al. *JAMA.* 1999, 282:1819

G. Encephalocele

- 1/5000 live births
- 3/4 females are occipital
- Basal, Syncipital and Convexity
- Chiari III-Cervicooccipital 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, syrinx 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

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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