Pediatric Neurosurgery: Missshapen Heads and Booties

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PEDIATRIC NEUROSURGERY:
MISSHAPEN HEADS AND BOOTIES

OVERVIEW

Spinal Dysraphism/Tethered Spinal Cord
Abnormal Head Shape

OVERVIEW

Spinal Dysraphism
Tethered Spinal Cord Syndrome
Review of different types of dysraphism
Surgical treatment
Abnormal Head Shape

DYSRAPHISM/TETHERED SPINAL CORD

Dysraphism
Failure of appropriate embryonic neural tube closure/development

Tethered Spinal Cord
Condition resulting from a dysraphic state
Anatomic and physiologic anchoring of neural elements to non-neural tissues
Clinical syndrome resulting from physiologic anchoring
Vascular phenomenon

TETHERED SPINAL CORD

Conceptual understanding
Clinical characteristics
Diagnosis
Imaging
TETHERED SPINAL CORD

Concept

Tethered Cord Syndrome

Clinical Characteristics

• Lower Extremity
  • Gait Deterioration
  • Foot/Leg Length Deformities
  • Leg Weakness/Atrophy

• Urological
  • Urinary retention

• Pain
  • Back Pain
  • Leg Numbness/Pain

• Scoliosis

When do I image?

What imaging do I order?

When do I refer to a specialist?

TETHERED SPINAL CORD

Sometimes it's pretty obvious

Thoracic Myelomeningocele

Myelomeningocele

CUTANEOUS STIGMATA

• Capillary Malformation
  • Benign (flat and small) versus Hemangioem

• Dimple/Dermal Sinus
  • Position relative to gluteal fold
  • Visualize the base

• Hypertrichosis (Faun's Tail)
  • Strongly suggestive of split cord malformation

• Subcutaneous Mass
  • Atretic Meningocele

• Aplasia Cutis (cigarette burn)

• Caudal Appendage
TETHERED SPINAL CORD

Imaging

- Ultrasound
  - Non-invasive
  - Inexpensive
  - Insensitive
  - for subtle pathology
  - Age limited (4m)

- X-ray
  - Non-invasive
  - Inexpensive
  - Little new information

- CT
  - Non-invasive
  - Expensive
  - Limited Role (Bone)

- MRI
  - Non-invasive
  - Sensitive
  - Specific
  - Expensive
  - +/- Sedation

- Feed & Sleep Technique
  - 9% require reschedule with anesthesia
  - 3.3m average age at success

OVERVIEW

- Spinal Dysraphism
  - Tethered Spinal Cord Syndrome
  - Review of different types of dysraphism
  - Surgical treatment

  - Abnormal Head Shape

SPINAL DYSRAPHISM “SPINA BIFIDA”

- Spina Bifida Aperta
  - Myelomeningocele

- Spina Bifida Occulta
  - Low Conus Medullaris
  - Dermal Sinus Track
  - Split Cord Malformation
  - Lipomyelomeningocele
  - Myelocystocele
  - Meningocele
  - Meningocele Manqué
  - Fatty Filum Terminale
  - Controversial

MYELOMENINGOCELE

- Open neural tube defect
- Almost always associated with chiari malformation, type II
- Folate supplementation
- Prenatal surgery or within 72 hours of birth
- 80% shunt dependent hydrocephalus
- Evolving Practice Patterns
- Tolerance of ventriculomegaly
- Lower with Fetal Closure

Failure of Bony Fusion is NOT associated with Tethering
- 20% of the population

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A randomized Trial of Prenatal versus Postnatal Repair of Myelomeningoceles

Shunt: 40% vs 80%

Improved:
- Hindbrain herniation
- Ambulation by 36 months

Worsened:
- Retether
- Preterm Delivery
- Uterine Dehiscence

A QUICK ASIDE ABOUT CHIARI MALFORMATIONS

Type I
- Common
- Probably largely asymptomatic
- Cerebellar Tonsils
- Usually isolated

Type II
- Exclusively in MMC
- Most do not require decompression
- Cerebellar Vermis
- Other intracranial anomalies

LOW CONUS MEDULLARIS
- Filum anchors conus caudally
- Prophylactic surgery
- 3-6 months of age (variable)

DERMAL SINUS TRACT
- Epithelialized tract from skin surface
- Incomplete disjunction of neurectoderm from surface ectoderm
- Midline from lumbosacral to tip of nose
- DDx: sacral dimple (4% of infants)
- Above the top of gluteal fold
- If you have to split the buttocks, not likely DST
- Association w/ Dermoid Tumors or Teratomas
- Meningitis risk mandates early surgery

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**SPLIT CORD MALFORMATION**
- Persistent neurenteric canal
- Type 1 – 2 dural sheaths w bony septum
- Type 2 – 1 dural sheath w fibrous septum
  - Septum acts as tethering agent
- Often associated w other lesions (MMC)
- Must also cut filum

**LIPOMYELOMENINGOCELE (SPINAL LIPOMA)**
- 3 Types (generally)
  - Dorsal
    - Mushroom off dorsal aspect of SC
    - Must also cut filum
  - Terminal
    - Below bottom of conus medullaris
  - Transitional
    - "Slurpee Spoon"
    - Most complex
    - Chaotic
    - Lipomatous mass ventral to nerve roots

**TERMINAL LIPOMA**

**TRANSITIONAL LIPOMA**

**DORSAL LIPOMA**
OVERVIEW

- Spinal Dysraphism
  - Tethered Spinal Cord Syndrome
  - Bladder, Legs/Feet, Back Pain, Scolii
- Low Threshold to Refer
  - Often saves imaging
- Correct answer for some findings isn’t known
  - Bifid Gluteal Fold, Some Dimples
- Abnormal Head Shape

ABNORMAL HEAD SHAPE

Why worry?

- Abnormal pressure on brain ➔ very rare
- Neurologic compromise ➔ extremely rare
- Sequelae of cosmetics ➔ OK

ABNORMAL HEAD SHAPE

- Normal Anatomy and Growth
  - Plagiocephaly versus Craniosynostosis
  - Characteristic head shapes
    - Surgical treatment

CRANIAL SUTURES

- Sagittal
- Coronal
- Metopic
- Lambdoid
- Squamosal
- Skull base

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LOOK AT THE BIG BRAIN ON BRAD
- Relative to birth size
  - 50% brain growth at 2 months
  - 100% at 6 months
  - 200% 10 months
  - 90% of adult size at 12 months
  - Virtually complete by 2 years
  - Normal sutures fuse by 6-8 years
  - Brain drives skull growth

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PLAGIOCEPHALY
- Oblique Contouring of the Skull
- Posterior (occipital) or Anterior (frontal)
- True craniosynostosis versus positional molding
  - Posterior is most commonly positional
  - Anterior is most commonly true craniosynostosis
CRANIOSYNOSTOSIS

**Premature fusion of one or more sutures**
- Incidence 0.4/1000
- Usually sporadic
- Compensatory cranial enlargement along open sutures
- Virchow’s Law
  - Perpendicular growth restraint, parallel growth enhancement
  - Predictable patterns of abnormality

CRANIOSYNOSTOSIS (cont'd)
- Multiple etiologies
  - Syndromic vs. non-syndromic
  - Metabolic disorders
  - Chromosomal abnormalities
- Radiographs – limited utility

POSITIONAL PLAGIOCEPHALY

- Most common asymmetric head shape
- Occipital flattening
- Ear positioned forward
- +/- frontal bossing
- Peri-sutural sclerosis on x-ray
  - (don’t get x-rays)
- Torticollis

POSITIONAL PLAGIOCEPHALY

- Behavioral modification - positional changes
- Questionable significance if face is symmetrical
- Virtually no late complaints from patients
  - as far as we know

POSITIONAL PLAGIOCEPHALY

- Worn for months
- Refitted
- Expensive ($150-4,000)
  - Not covered by insurance
  - Lack of outcomes data
  - Sweet fashion statement

ABNORMAL HEAD SHAPE

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CRANIOSYNOSTOSIS

- Correct congenital deformity
- Essentially cosmetic
  - Benefit to early operation in some cases
- Neurologic function mostly unaffected
  - No effect on seizures or cognitive delay
- Rarely, elevated intracranial pressure is relieved

SAGITTAL SYNOSTOSIS

- Most common pattern
  - Sagittal suture fused
- Scaphocephaly—long narrow skull
  - Sagittal ridge and bossing
- Dolichocephaly—abnormally long skull
  - Normal variant

SAGITTAL SYNOSTOSIS imaging

Don't need imaging...
  - (some debate)

SAGITTAL SYNOSTOSIS surgery

...because you treat based on appearance

CRANIOSYNOSTOSIS Surgical Options

Open
- 2-5 day hospitalization
- Pediatric anesthesia is crucial
- Complications should be few
- Periorbital swelling
- Blood transfusions
- Post-op Cranial irregularities
  - Persistent cranial defects in 5%

Endoscopic
- 1-3 day hospitalization
- Pediatric anesthesia is crucial
- Complications should be few
- +/- Periorbital swelling
- +/- Transfusion
- Post-op Helmet
  - until approx 12 months
A: Sagittal craniectomy  
B: Biparietal morcellation; panels of bone created with osteotome are left attached to the dura

**SAGITTAL SYNOSTOSIS**

- Minimally invasive surgery
- Treatment
  - Operation at 3 m or less
    - Easier, Safer, Better Outcome
  - Open versus Endoscopic – similar outcomes
    - Open
      - Vertex craniectomy vs. morcellation vs. Pi procedure vs. calvarial reconstruction
    - Endoscopic
      - Shorter procedure, Smaller Incision
      - Helmet until 12m

**ENDOSCOPIC STRIP CRANIECTOMY**
HELMETS

CORONAL CRANIOSYNOSTOSIS

• Unilateral = Anterior plagiocephaly
  • Contralateral frontal bossing
  • Flat frontal bone & Short orbit
• Bilateral is usually syndromic

CORONAL SYNOSTOSIS

Don’t need imaging...
• (some debate again)

Harlequin sign
• Looks cool
• Doesn’t make the Dx

UNICORONAL SYNOSTOSIS

• Optimal age 3-6 months
• Unilateral or bilateral craniotomy
• Unilateral or bilateral orbital rim reconstruction
• Lateral canthal advancement

Open versus Endoscopic

METOPIC CRANIOSYNOSTOSIS

• Wide spectrum of severity
• Pointed forehead with triangle shape
  • Trigonocephaly
  • Lateral orbital rims recessed
• Increased association with brain/chromosomal abnormalities
• CT or MRI is optional
• May improve cosmetically with time

METOPIC SYNOSTOSIS

Imaging
**METOPIC SYNOSTOSIS**

- Optimal at age 6-8 mos.
- Bilateral frontal craniotomy
- Orbital rim reconstruction
- Bilateral lateral canthal advancement

**POSTERIOR DEFORMITIES**

- Lambdoid synostosis is extremely rare
  - Lambdoid synostosis vs. occipital positional plagiocephaly
  - Does it matter?

**CRANIOSYNOSTOSIS**

- Diagnosed by inspection
- Surgery to correct congenital deformity
  - Primary outcomes related to cosmesis
  - Imaging usually not necessary
  - Rarely need CT or MRI
  - Earlier diagnosis provides opportunity for earlier surgery
  - Endoscopic versus open
  - Pediatric anesthesia very important

**THANK YOU**

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