Craniosynostosis and Plagiocephaly

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    • Dr. Laurie Ackerman at Riley Hospital for Children

Agenda

• Craniosynostosis vs positional plagiocephaly
• Anterior fontanel closure
• Microcephaly
• Benign macrocrania
Craniosynostosis

• Premature fusion of ≥1 cranial suture(s)
• Incidence 1:2000 to 1:2500 live births
• Some syndromic/familial, most not
• Fibroblast growth factor receptor pathways
• Skull cannot grow perpendicular to the suture so it grows parallel (Virchow’s law)
• Differentiate from positional plagiocephaly

Normal Sutures

Positional Plagiocephaly

• Posterior flattening
• Anterior ear and forehead displacement on side of flattening
• Parallelogram shape
• Not craniosynostosis
Sagittal Craniosynostosis

- Long (AP)
- Narrow (lateral)
- Frontal bossing
- Occipital bossing
- Fontanel open or closed
- Scaphocephaly

Bilateral Coronal Craniosynostosis

- Short (AP)
- Wide (lateral)
- Syndromic?
- Symmetric positional flattening?
- Brachycephaly
Bilateral Coronal Craniosynostosis

Unilateral Coronal Craniosynostosis

- Forehead flattening
- Orbit drawn up
- Fontanel displaced
- Nasal root deviation

Harlequin sign
Unilateral Coronal Craniosynostosis

- Pointed forehead
- Narrow forehead
- Triangle shaped
- Hypotelorism
- Ridging alone may be followed
- Trigonocephaly

Metopic Craniosynostosis

- Pointed forehead
- Narrow forehead
- Triangle shaped
- Hypotelorism
- Ridging alone may be followed
- Trigonocephaly
Lambdoid Craniosynostosis

- Posterior flattening
- Posterior ear and forehead displacement
- Trapezoid shape
- Ear forced down
- Rare

Lambdoid Craniosynostosis

- Posterior flattening
- Posterior ear and forehead displacement
- Trapezoid shape
- Ear forced down
- Rare

Lambdoid vs Positional
Diagnostic Uncertainty

- Skull x-rays
  - Phased out in favor of...
- CT Head “limited skull very low dose”
  - Same or less radiation than 4 view skull x-rays
  - Provides 3D skull images, brain not seen
  - If shows open sutures, then no craniosynostosis
  - (Epic orderable is RADCT400)

Natural History

- Left untreated, there is a risk for:
  - Worsening head shape
  - Overall head growth restriction leading to increased intracranial pressure (ICP)
Elevated ICP

Treatment Goals

• Unlocking the bones
• Reshaping the skull
• Open vs Endoscopic vs Springs

Craniosynostosis Treatment

<table>
<thead>
<tr>
<th></th>
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<th>Endoscopic</th>
<th>Spring-Assist</th>
</tr>
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<tbody>
<tr>
<td>≥ 6 mo</td>
<td>2.5 to 3.5 mo</td>
<td>3 to 6 mo</td>
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<tr>
<td>Incision ear to ear</td>
<td>One or two 2cm incisions</td>
<td>One 5cm incision for sagittal</td>
<td></td>
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<tr>
<td>Eyes swollen shut</td>
<td>No periorbital swelling</td>
<td>Usually no periorbital swelling</td>
<td></td>
</tr>
<tr>
<td>Transfusion always</td>
<td>Transfusion rare</td>
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<td></td>
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<tr>
<td>ICU 1d, Floor 3d</td>
<td>Floor 1d</td>
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<td>No helmet</td>
<td>Helmet required</td>
<td>No helmet, but second surgery 3m</td>
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Open

Fronto-orbital Advancement  Posterior CVR

Sagittal Synostosis Repair Options

Strip Craniectomy  Pi Procedure

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Endoscopic

Endoscopic Strip Craniectomy with Helmet

Helmet

- Hard shell
- Moldable foam inside
- Gaps where skull growth desired
- 23 hours per day
- Every day
- Managed by orthotist
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Springs

- Stainless steel
- Force is variable
  - Patient age
  - Bone thickness
  - Deformity severity
- Second surgery for removal at 3m
Cranial Expansion with Normal Calvarial Shape

Distraction Osteogenesis

- Bicoronal incision, craniotomy, leave bone on
- Implant distractors, wait 4-5 days
- Distract 1 mm/day x 30 days
- Remove posts, close skin (in office)
- Wait 3 months
- Remove distractors (in OR)
Cranial Expansion with Normal Calvarial Shape

Preop: headache & papilledema
Postop: both resolved

Ilizarov and Distraction Osteogenesis: Applications in Syndromic Synostosis

Plagiocephaly Treatment

- Cosmetic issue, does not affect brain devel
- Parents concerned?
- Alternate head position for sleep, holding
- Tummy time
- PT for torticollis
- Development of head control
- Helmet
Pumpkin s/p Helmet in Circleville

Anterior fontanel closure

- Median time to closure 14 months
- Wide variation in closure times
  - 1% closed at 3 months
  - 38% closed at 1 year
  - 96% closed at 2 years

“Early” fontanel closure

- Assess for craniosynostosis
- Head growth along a curve?
- Signs of high ICP?
  - HA, N/V, lethargy, upgaze palsy
- Developing normally?
Microcephaly

• Assess for craniosynostosis
• Brain growth drives skull growth
• Neurology referral

Benign Macrocrania

• Head circumference initially crosses curves but then parallels a curve, often > 95%
• Familial - large heads run in the family

Benign Macrocrania

• Assess for craniosynostosis
• Head growth along a curve?
• Signs of high ICP?
  • HA, N/V, lethargy, upgaze palsy
  • Bulging, tense fontanel when upright
• Developing normally?
  • Can be some delay in head control due to size
Diagnostic Uncertainty

- Head US
  - No radiation
  - Will definitively assess for hydrocephalus
  - Large subarachnoid spaces are expected
- Limited MRI brain (at NCH)
  - If fontanel closed
  - No radiation and no sedation

Thank you