Abnormal Head Shape and Crainosynostosis: State-of-the-art Management

8:45 – 9:30 a.m.

John Jensen, MD
Sean Lew, MD
I have no relevant financial relationships to disclose.
Objectives

• To review the initial evaluation of a patient with an abnormal head shape

• To understand current management techniques for positional plagiocephaly

• To obtain a basic understanding of the current surgical techniques utilized for craniosynostosis
Initial assessment

- Visual assessment from anterior, posterior, and vertex positions
- Note any preference for rotation of the neck, torticollis
- Feel for any ridging of suture lines
Abnormal head shape

Infant Skull Deformity

- Sagittal synostosis
- Metopic synostosis
- Normocephaly
- Bilateral coronal synostosis
- Unilateral coronal synostosis
- Lambdoid synostosis

Mild, nonsurgical problem of cosmetic concern only

Deformational posterior plagiocephaly
# Deformational plagiocephaly

<table>
<thead>
<tr>
<th></th>
<th>Mild</th>
<th>Moderate</th>
<th>Severe</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lateral</td>
<td>Flattening on back of skull</td>
<td>Ipsilateral ear anteriorly displaced,</td>
<td>Ipsilateral temporal skull growth</td>
</tr>
<tr>
<td>deformational</td>
<td>only</td>
<td>ipsilateral frontal bossing</td>
<td></td>
</tr>
<tr>
<td>plagiocephaly</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><em>Key findings:</em></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

| Posterior        | Central posterior deformity   | Widening of posterior skull               | Temporal bossing                          |
| deformational    |                               |                                           |                                           |
| plagiocephaly    |                               |                                           |                                           |
| (Brachycephaly)  |                               |                                           |                                           |
| *Key findings:*  |                               |                                           |                                           |

Deformational Plagiocephaly

- AKA “positional plagiocephaly”
- Multiple risk factors
  - Major
    - Torticollis
    - Supine sleep position
      - Higher association with other congenital anomalies requiring early prolonged supine positioning (e.g. surgical GI/urologic abnormalities)
      - Prolonged NICU stays
  - Minor
    - Preterm delivery, low birth weight
    - Intrauterine constraint
    - Developmental delays
      - Increased possibility of developmental delay in infants with DP, not clear that there is a causal relationship
    - Bottle-feeding (always positioning same way)
Deformational Plagiocephaly

• Prevention
    • Randomized control trial, 111 healthy newborns randomized at birth
    • Parents received detailed instructions on prevention in treatment group
    • At 3 months prevalence of DP lower in treatment group (blinded, 3D scanner): 15 vs 33% (p=0.05)
Deformational Plagiocephaly

  - Parental Instructions
    - Sleeping: Alternate head of bed, stimulation
    - Feeding: Alternate sides when feeding
    - Supervised tummy time: begin during first week of life, work up to 15-30 minutes
    - Carriers: minimize time in bouncers and car seats, alternate head position regularly
    - Play: room to move freely, avoid hanging toys (promote passivity), spread objects on floor, frequently reposition
    - Counterpositioning, stretching if head rotation preference develops
Deformational Plagiocephaly

• Treatment
  – The earlier the intervention, the better the results
  – Treat torticollis
    • Neck exercises with every diaper change
      – Rotate head side to side, hold for 10 sec
      – Tilt head side to side (ear to shoulder), hold for 10 sec
      – Rotating chair or stool technique (> 3 mos)
    • Physical therapy referral
  – Repositioning therapy
  – Counterpositioning
Deformational Plagiocephaly

• Treatment
  – Helmet (orthotic) therapy
    • Reserve for more severe or persistent cases
    • Rarely used prior to 6 months or after 12 months
    • Cost is significant, not always covered by insurance
    • High-quality evidence for efficacy is lacking
      – Most series supporting helmet therapy are Level IV evidence or worse
      – To date only one published randomized, controlled trial
Helmet therapy in infants with positional skull deformation: randomised controlled trial

Renske M van Wijk PhD candidate, Leo A van Vlimmeren senior researcher in paediatric physiotherapy, Catharina G M Groothuis-Oudshoorn biostatistician, Catharina P B Van der Ploeg epidemiologist, Maarten J IJzerman professor, Magda M Boere-Boonkamp associate professor of youth health care

1Department Health Technology and Services Research, Institute of Innovation and Governance Studies, University of Twente, Drienerloaan 5, 7522 NB, Enschede, Netherlands; 2Department of Rehabilitation, Paediatric Physical Therapy, Radboud university medical center, Nijmegen, Netherlands; 3Scientific Institute for Quality of Healthcare, Radboud university medical center, Nijmegen, Netherlands; 4TNO Child Health, Leiden, Netherlands
Deformational Plagiocephaly

• Renske et al BMJ (2014)
  – 29 physiotherapy practices, 84 infants randomized between 5-6 mos of age between helmet therapy or natural course without additional treatment recommended
  – Primary outcome: change in skull shape from baseline at 24 mos
  – Secondary outcomes: ear deviation, facial asymmetry, motor development, QOL, parental satisfaction and anxiety
  – No significant difference in any outcome between groups
    • “Full recovery” in ~1/4 of patients
Deformational Plagiocephaly

• Recommendations
  – Early parental education
  – Early intervention if torticollis or plagiocephaly identified
  – Helmet therapy is an option for minority of cases (severe, or moderate without improvement with repositioning measures)
Deformational Plagiocephaly

• Wisconsin BadgerCare Plus and Medicaid Cranial Orthosis Approval Criteria
  – Between 3 and 12 months old
  – At least 2 mo failed conservative tx (PT, home exercises, adjustments, repositioning)
  – Documentaton the condition will compromise function by a CF or Neurosurgeon
  – Prescription by CF or Neurosurgeon who has completed evaluation of patient
Craniosynostosis Overview

- Premature closure of one or more sutures of the cranial vault
- Leads to abnormal head shape
- May affect brain development and growth
- 1 in 2000-2500 live births
Craniosynostosis Diagnosis

- Diagnosed usually shortly after birth by physical exam
- Milder forms may become more apparent during first year of life
- Usually distinguished from *deformational positional plagiocephaly*
- Plain radiographs not routinely required to make diagnosis
- CT scan usually obtained for surgical planning
Clinical Features

CRANIOSYNOSTOSIS
- “Trapezoid” shape
- Sutural ridging present
- Absent fontanelle +/-
- Cranial disproportion
- Brow asymmetry
- Asymmetric, mass-like bulges
- Nasal root deviated

DEF PLAGIO
- “Parallelogram” shape
- No sutural ridging
- Fontanelle usu present
- Posterior flattening
- Brows symmetric
- Mild forehead asymmetry
- Nasal root straight
“Early or delayed AF [closure] may represent normal variants in healthy children without other clinical evidence of underlying pathology.”

Craniosynostosis: Virchow

Pathologic Definition: premature fusion of cranial sutures

Synostosis + Growth = Abnormal skull form
Variable severity and morphology
Sagittal Synostosis
Metopic Synostosis
Coronal Synostosis
Lambdoid Synostosis
Synostosis secondary to arrested brain growth
Treatment

• Treatment is surgical

• Milder forms (typically sagittal or metopic) may not require treatment

• Two main options
  – Minimally invasive (endoscopic) suturectomy + postoperative helmet therapy
  – Conventional open repair
Endoscopic suturectomy
(Jimenez/Barone)
Minimally Invasive Strip Craniectomy
(Open) Cranial Vault Expansion
Hung Span Technique (McCarthy)
Open technique: Surgical Issues

- **Re-shape skull**
  - Multiple osteotomies are necessary: blood loss
- **Expand intracranial space acutely**
  - Large dead space
- **Large pieces of devitalized bone: infectious risk**
- **Dural tears are a risk**
- **Preserve structural integrity posteriorly and inferiorly**
preop CT scan (7 months old)

1 year post-op (23 months old)
Cranial Distraction Osteogenesis
Cranial Distraction Osteogenesis
The Impact of Age at Surgery on Long-Term Neuropsychological Outcomes in Sagittal Craniosynostosis

Anup Patel, M.D., M.B.A.  
Jenny F. Yang, B.S.  
Peter W. Hashim, B.A.  
Roberto Travieso, M.D.  
Jordan Terner, M.D.  
Linda C. Mayes, M.D.  
Paul Kanev, M.D.  
Charles Duncan, M.D.  
John Jane, Jr., M.D.  
John Jane, Sr., M.D., Ph.D.  
Ian Pollack, M.D.  
Joseph E. Losee, M.D.  
David J. Bridgett, Ph.D.  
John A. Persing, M.D.

New Haven and Hartford, Conn.; Charlottesville, Va.; Pittsburgh, Pa.; and DeKalb, Ill.

Background: The relationship between surgical age and long-term neuropsychological outcomes in sagittal-suture craniosynostosis remains equivocal. Whole-vault cranioplasty and strip craniectomy are performed at various times in individuals with sagittal-suture craniosynostosis. This study used comprehensive neurological testing to examine the relationship between age at time of surgery and long-term neuropsychological function.

Methods: Seventy sagittal-suture craniosynostosis patients who had previously undergone either whole-vault cranioplasty or strip craniectomy were examined divided into three groups: treatment before 6 months (n = 41), between 6 and 12 months (n = 21), and after 12 months (n = 8). To examine long-term cognitive functioning, participants between the ages of 5 and 25 years underwent neurodevelopmental tests to evaluate intelligence, achievement, and learning disabilities.

Results: Compared with those treated between 6 and 12 months and after 12 months, patients who underwent surgery before 6 months demonstrated higher full-scale IQ (p < 0.01) and verbal IQ (p < 0.01). Patients who received surgery before 6 months also demonstrated superior abilities in word reading (p < 0.01), reading comprehension (p < 0.01), spelling (p < 0.01), and numerical operations (p < 0.05) relative to those who had surgery between 6 and 12 months old. A statistically significant higher percentage of patients treated after 6 months had one or more reading-related learning disabilities as compared with those undergoing earlier surgery.

Conclusions: This study suggests that surgery before 6 months old results in improved long-term neurological outcomes. Future studies should examine how the technique of surgery impacts these neuropsychological measures. (Plast. Reconstr. Surg. 134: 608e, 2014.)

CLINICAL QUESTION/LEVEL OF EVIDENCE: Therapeutic, III.

“This study suggests that surgery before 6 months old results in improved long-term neurological outcomes.”

The Effects of Whole-Vault Cranioplasty versus Strip Craniectomy on Long-Term Neuropsychological Outcomes in Sagittal Craniosynostosis

Peter W. Hashim, B.A.
Anup Patel, M.D., M.B.A.
Jenny F. Yang, B.S.
Roberto Travieso, M.D.
Jordan Terner, M.D.
Joseph E. Losee, M.D.
Ian Pollack, M.D.
John Jane, Sr., M.D., Ph.D.
John Jane, Jr., M.D.
Paul Kanev, M.D.
Linda Mayes, M.D.
Charles Duncan, M.D.
David J. Bridgett, Ph.D.
John A. Persing, M.D.

New Haven and Hartford, Conn.; Pittsburgh, Pa.; Charlottesville, Va.; and DeKalb, Ill.

Background: The optimal type of surgical management for isolated sagittal synostosis remains a source of significant debate. There is a paucity of data regarding possible differences in long-term neuropsychological outcomes following treatment with whole-vault cranioplasty or endoscopic strip craniectomy. This study provides the first comparative analysis examining the effects of the two techniques related to long-term intellectual functioning.

Methods: A total of 70 patients were enrolled in this multicenter study, 29 of whom had previously undergone endoscopic strip craniectomy and 41 of whom had previously undergone whole-vault cranioplasty. All patients completed a battery of neurodevelopmental tests (Beery-Buktenica Developmental Test of Visual-Motor Integration, Wechsler Abbreviated Scale of Intelligence, and Wechsler Fundamentals) to evaluate various domains of neuropsychological function.

Results: In a group comparison of those treated before 6 months of age, whole-vault patients obtained higher scores relative to endoscopic strip craniectomy patients on visuomotor integration, full-scale intelligence quotient, verbal intelligence quotient, word reading, and reading comprehension ($p < 0.05$ for all). When compared against strip craniectomy performed before 5 months of age, the whole-vault group still showed significantly higher scores in verbal intelligence quotient, reading comprehension, and word reading ($p < 0.05$ for all).

Conclusions: The type of surgical intervention for isolated sagittal synostosis impacts long-term neuropsychological outcomes. Patients undergoing early whole-vault cranioplasty attained higher intelligence quotient and achievement scores relative to those undergoing strip craniectomy. Surgical management with whole-vault cranioplasty performed before 6 months of age provides the most favorable long-term intellectual outcomes in patients with isolated sagittal synostosis. (Plast. Reconstr. Surg. 134: 491, 2014.)

CLINICAL QUESTION/LEVEL OF EVIDENCE: Therapeutic, II.

“Patients undergoing early whole-vault cranioplasty attained higher intelligence quotient and achievement scores relative to those undergoing strip craniectomy.”

<table>
<thead>
<tr>
<th>Minimally invasive</th>
<th>Conventional open</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Optimal surgery age 2-3 months</strong></td>
<td><strong>Optimal surgery age 6-9 months</strong></td>
</tr>
<tr>
<td>Multiple small incisions</td>
<td><strong>Single large incision</strong></td>
</tr>
<tr>
<td>Overnight hospital stay</td>
<td><strong>3-4 day hospital stay</strong></td>
</tr>
<tr>
<td>Blood transfusion rare</td>
<td><strong>Blood transfusion routine</strong></td>
</tr>
<tr>
<td>Requires 6-9 mos of cranial molding helmet therapy</td>
<td><strong>No requirement for postoperative helmet use</strong></td>
</tr>
<tr>
<td>More frequent follow-up appointments</td>
<td><strong>Less frequent follow-up appointments</strong></td>
</tr>
</tbody>
</table>
Technique Comparison

• No advantage between techniques in terms of risks
• Typically ~90% favorable outcome with either technique
• Theoretical concerns/unknowns
  – Longer anesthetic with open surgery vs. anesthesia at younger age with minimally-invasive
  – Effect of helmet therapy on brain development
  – No randomized studies comparing techniques
Craniosynostosis at Children’s Hospital of Wisconsin

- Joint procedure by neurosurgery and craniofacial
- All referred patients to either specialty receive consultations from both neurosurgery and craniofacial
- Surgical procedures performed as a team
- Referral must be made early in life for patient to have option of minimally-invasive techniques. Diagnosis and such consultations will be “fast-tracked” accordingly
Craniosynostosis Team

Craniofacial

Arlen D. Denny, MD
John N. Jensen, MD

Neurosurgery

Andrew B. Foy, MD
Bruce A. Kaufman, MD
Sean M. Lew, MD
What makes us unique?

• We have an exceptionally good track record with regard to postoperative complications
  – 10 year review of 333 craniosynostosis cases at CHW
    • 9 unplanned returns to OR for minor issues (hematoma evacuation, minor infections)=2.7%
    • 0 major complications (stroke, death)

• Long term followup

• Offer all techniques (traditional procedure, minimally invasive, distraction osteogenesis)
Future Goals

• Currently enrolling patients to determine functional benefit of open procedure with regard to resting connectivity

• Plan for expansion of study to include a comparison of techniques
Locations

Referred patients can be seen at:

• Delafield

• Main Campus - Milwaukee
Appointments

• Patient families can call central scheduling to schedule or be connected to our nurse triage line

• Referring providers can call our nurse triage line direct at (414) 266-7525

• If you have an urgent appointment request, call our nurse triage line
Contact Information

John Jensen, MD
(414) 266-7525

Sean Lew, MD
(414) 266-6435

Physician Referral Consultation: (800) 266-0366
Cephalohematoma

Extent of hematoma can be clue to level of bleed

Vaccuum-assisted Delivery
Cephalohematoma