Neurodevelopmental Implications of head positioning in term and preterm infants

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Learning Objectives:

By completion of this session participants will be able to:

» 1.) Describe neurodevelopmental implications of poor head positioning in preterm and term infants

» 2.) Demonstrate neuroprotective head positioning techniques for premature infants under 32 weeks gestation

» 3.) Identify characteristics of deformational plagiocephaly, brachycephaly, and dolichocephaly

» 4.) Demonstrate neuroprotective strategies for preventing and treating various cranial deformations
Head Positioning and IVH
Positioning and handling to prevent IVH in preterm infants

» 2003 Vermont Oxford Network focus group developed an evidence based protocol for potentially best practices to prevent IVH in VLBW infants which includes...

» Midline head (supine or sidelying) first 72 hours of life

» Head of bed elevated to 30 degrees

» No lifting of legs

» 2017 Systematic review and meta-analysis concluded there is insufficient evidence regarding the effect of head positioning and tilting on the incidence of GMH-IVH and cerebral hemodynamics and oxygenation in preterm infants. Neither recommend nor refute the use of a neutral head position and/or head tilting in order to prevent GMH-IVH.
CRANIAL DEFORMATIONS
Brain Growth and Development

➢ The rapid growth in neuronal cell number during the 10th through 18th weeks of gestation achieves near adult cell numbers.

➢ This is followed by dramatic increases in dendritic growth and arborization then myelinization.

➢ At 15 months of age the brain is roughly 65% adult size while the cerebellum has achieved adult proportion.

➢ The majority of myelinization is complete by 2 years of age.

Bronfin, 2001
Brain Growth

» A newborn's head circumference is larger than the chest circumference at birth.

» Average occipital-frontal circumference (OFC) is 35 cm in the term newborn, 45 cm at 1 year, and 55 cm in an adult.

» The OFC increases by 2 cm per month for the first 3 months of life, 1 cm per month for the second 3 months of life, and 0.5 cm per month from 6–12 months.

» The volume of the cranial vault is 65% of adult size at birth and 95% of the adult size at age 10 years. In contrast, facial size is 40% of adult size at birth and 65% at 10 years.

Bronfin, 2001
Anatomy of the Skull
Sutures

» “syndesmotic,” joints
» permit growth
» Sagittal
» Coronal
» Lambdoidal
» Metopic

Bronfin, 2001
Malformation vs. Deformation

- **Malformation**: an intrinsically altered developmental process that interferes with cell migration and differentiation through genetically programmed biochemical processes or through extrinsic chemical interference (teratogens). In essence, this process represents an error in the normal development of a part.

- **Deformation**: an alteration of a body part that is developing normally until a mechanical force is applied.

Bronfin, 2001
Cranial Malformations (Craniosynostosis)

» Craniosynostosis: the premature fusion of one or more cranial sutures.
Cranial Deformations

» Deformational Brachycephaly

» Deformational Plagiocephaly

» Deformational Dolichocephaly

» True Scaphocephaly *(Not deformational)*
Differentiation between synostotic and non-synostotic head shapes

**Deformational Brachycephaly**
- Occipital flattening
- Increased CI (wide head)
- Often have some asymmetry/plagiocephaly
- May have turricephaly (increased cranial height, limited to posterior cranium)

**Synostotic Brachycephaly**
- Bilateral coronal sutures fused
- Very rare
- Congenital not developed
- Severe forehead retrusion
- Anterior turricephaly
Differentiation between synostotic and non-synostotic head shapes

Deformational (Scapho)Dolichocephaly
• Long, slender head “toaster”
• Usually result of extreme head rotation to one side
• Premature infants positioned side-to-side in NICU
• Normal increase in width from anterior to posterior

Synostotic Scaphocephaly (Sagittal Synostosis)
• “Boat shaped” head
• Frontal bossing
• Bilateral occipital/parietal narrowing posterior to anterior fontanelle
• Decreased vertical height posterior cranium
Differentiation between synostotic and non-synostotic head shapes

**Deformational Plagiocephaly**
- Frontal bossing ipsilateral
- Often associated with head preference or torticollis
- Anterior shift of ipsilateral forehead, ear, and cheek
- Eye may appear more open where forehead shows increased bossing

**Unilateral Coronal Synostosis**
- Flattening of forehead ipsilateral
- Nasal root and midfacial angulation
- Anterior displacement of ipsilateral ear
- Eye appears more open where forehead is flattened, contralateral eyelid ptosis
Differentiation between synostotic and non-synostotic head shapes

**Deformational Plagiocephaly**
*(Positional Plagiocephaly)*
- Unilateral occipital flattening
- Ipsilateral frontal bossing
- Contralateral occipital bossing
- Parallelogram (sort of)
- Ipsilateral ear shift anterior

**Lambdoidal Synostosis**
*(Synostotic Posterior Plagiocephaly)*
- Unilateral occipital flattening
- Contralateral parietal bossing
- Minimal contralateral frontal bossing
- Trapezoid, windswept
- Ipsilateral ear shift posterior

![Diagram of DP vs. LS](image)
Assessment

» No standards
» Visual Assessment
» Anthropometric Caliper measurement
» Flexicurve
» Plagiocephalometry
» 3D Photography
» Radiological imaging
» In the near future... app under development in Germany
<table>
<thead>
<tr>
<th>Measurements</th>
<th>From</th>
<th>To</th>
</tr>
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<tbody>
<tr>
<td>Length</td>
<td>Glabella</td>
<td>Opisthocranium</td>
</tr>
<tr>
<td>Width</td>
<td>Eurion (1 cm above the otobasiun superious)</td>
<td>Eurion (1 cm above the otobasiun superious)</td>
</tr>
<tr>
<td>Oblique</td>
<td>Frontotemporal point (lateral point of the ipsilateral eyebrow)</td>
<td>Lamboidal point</td>
</tr>
<tr>
<td>Circumference</td>
<td>Include Glabella and Opisthocranium (lower edge of measurement tape directly above eyebrow)</td>
<td>Include Glabella and Opisthocranium (lower edge of measurement tape directly above eyebrow)</td>
</tr>
</tbody>
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Measurements

Cranial Index (Cephalic Index) CI

- Represents the ratio of maximum cranial width to maximum cranial length
- Commonly used for isolated sagittal synostosis (ISS), but also used for any scaphocephaly/dolichocephaly or brachycephaly head shape.

Scale for Cephalic Index (CI)

- Normal: 75 – 90 mm
- Mild: 91 – 93 mm
- Moderate: 94 – 97 mm
- Severe: >97 mm
- Normocephaly or plagiocephaly = CI >76%–<90%
- Brachycephaly = CI >90%
- Dolichocephaly = CI <76%

 Ruiz-Correa 2006, Likus 2014
Deformational Dolichocephaly
Deformational Brachycephaly
Measurements

Cranial Vault Asymmetry Index (CVAI)
(used to measure plagiocephaly)

CVAI = \frac{|A-B| \times 100}{A \text{ or } B} \\
\text{(whichever is greater)}

<table>
<thead>
<tr>
<th>Level of Deformity</th>
<th>CVAI</th>
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<tbody>
<tr>
<td>1 Normal</td>
<td>&lt;3.5%</td>
</tr>
<tr>
<td>2 Mild</td>
<td>3.5%-6.25%</td>
</tr>
<tr>
<td>3 Moderate</td>
<td>6.25%-8.75%</td>
</tr>
<tr>
<td>4 Severe</td>
<td>8.75%-11.0%</td>
</tr>
<tr>
<td>5 Very Severe</td>
<td>&gt;11.0%</td>
</tr>
</tbody>
</table>
Deformational Plagiocephaly
Neurodevelopmental Implications of Synostotic Cranial Malformations

- 50% of children with nonsyndromic sagittal suture craniosynostosis had reading and/or spelling learning disability.

- 37% of children with isolated sagittal synostosis had speech and/or language impairment of whom 71% had moderate or severe impairments.

- Children with nonsyndromic craniosynostosis had significantly different standardized distribution of BSID-II PDI=0% accelerated, 43% normal, 48% mild delay and 9% significant delay.

- Consistently lower mean neurodevelopmental scores in children with single-suture craniosynostosis compared with controls. These results provide further support for neurodevelopmental screening in young children with single-suture craniosynostosis.

Magge, 2002; Shipster, 2003; Panchal, 2001; Starr, 2012
Neurodevelopmental Implications of Synostotic Cranial Malformations

• “Craniosynostosis is no more considered merely an aesthetic disorder, as numerous reports of neurodevelopmental outcomes are being published. Although there are many issues to be addressed and confounding variables to take into account, patients and their families are making more complex demands not only in terms of cosmetic appearance but also in terms of cognitive results. The clinician's point of view should be geared toward comprehensive management, embracing appearance as well as function.”

• “Overall, findings suggest that isolated craniosynostosis is associated with a 3-5 fold increase in risk for cognitive deficits or learning/language disabilities”

Shim, 2016; Collett, 2005
Neurodevelopmental Implications of Nonsynostotic Cranial Deformations

Incidence of Otitis Media

• “…structural differences in the anatomy of the middle ear and eustachian tube can result in an increased susceptibility to otitis media.”

• A retrospective study using a parent questionnaire comparing 1259 patients with deformational plagiocephaly to patient data from CDC. Questionnaire showed a trend directly correlating otitis media and severity level of DP. (not statistically significant)

• In addition a subset of infants with DP were administered a tympanogram to assess state of middle ear. Tympanogrammetry showed a marked percentage of infants with DP to have eustachian tube dysfunction.

• "The more severe cases (types IV-V) of plagiocephaly had a higher percentage of otitis media than the less severe cases (types I-III),"

• "The significantly high percentage of tympanogram readings that pointed to otitis media...suggests an overall malfunction of the middle ear drainage function of the eustachian tube in these children”

Purzycki, 2009
Neurodevelopmental Implications of Nonsynostotic Cranial Deformations

Auditory ERPs and Plagiocephaly

• “It is suspected that the developmental delay in school-aged children diagnosed as infants suffering from plagiocephaly is caused by the modification of the skull form.”

• This study compared auditory ERPs in infants with DP and healthy controls.

• “Differences between the patients and control subjects indicate that already at this early age the presence of the plagiocephalic skull signals compromise of brain functioning.”

• “The present data suggest that most of the plagiocephalic infants have an elevated risk of auditory processing disorders.”

• This study demonstrated, “for the first time, that the central sound processing, as reflected by ERPs, is affected in children with plagiocephaly.”

Balan et al.
Figure 1: Typical auditory ERPs of plagiocephalic (solid line) and control (dashed line) children of 6 to 16 months of age. The P150 and N250 deflections are visible. Balan et al.
Neurodevelopmental Implications of Nonsynostostotic Cranial Deformations

ERPs and Deformational Plagiocephaly

• In contrast to prior study, infants with deformational plagiocephaly did not differ from controls with respect to the maximum P150 or N450 signals.
• “This study marks the first electrophysiologic examination to demonstrate that deformational plagiocephaly is not likely associated with significant impairments in language function.”
• This study used phonemes rather than tones as the external stimulus to directly examine language processing, not just auditory processing.
• “The scope of this investigation was limited to language systems and other areas of brain functioning may be at risk in infants with deformational plagiocephaly.”
Aggregate event-related potential waveforms from plagiocephalic and control infants over the left frontal scalp illustrating the P150 and N450 components.
Neurodevelopmental Implications of Nonsynostostotic Cranial Deformations

Visual Field Defects

• two abnormal conditions were identified: hemifield asymmetries, in which the breadth of the left and right hemifields differed by more than 20 degrees; and constricted hemifields, in which a given hemifield was at least 20 degrees less than those established for normal controls of the same age.

• Interestingly, there was no statistical correlation between the side of the visual-field defects relative to the side of the occipital flatness. However, for the majority of the 16 infants for whom these comparisons were investigated, the visual defects were ipsilateral to the flattened occiput. A correlation between the severity of the hemifield constriction and the severity of the cranial asymmetry was also observed.

• These observations do not speak to whether these defects are a product of plagiocephaly itself or the result of a more global developmental delay

Siatakowski et al
Neurodevelopmental Implications of Nonsynostotic Cranial Deformations

Visual Field Defects

- Eight of 93 patients (9%) with deformational plagiocephaly had unilateral astigmatism, and 14 (15%) had bilateral astigmatism.

- Children with deformational plagiocephaly do not have an increased prevalence of strabismus compared with the general population but do have an increased prevalence of astigmatism, whereas children with nonsyndromic craniosynostotic plagiocephaly have an increased prevalence of strabismus and astigmatism.

Gupta et al.
Neurodevelopmental Implications of Nonsynostotic Cranial Deformations

Mandibular Symmetry

• Observations made at the Washington University School of Medicine, St. Louis, Missouri in 1996. At that time CT scans were commonly obtained.
• 23 scans available to analyze the shape and volume of the mandible, which, although not directly affected by positional forces, appeared to be affected by the altered shape of the cranium.
• This analysis found that the volume of the mandible on the affected side (i.e., the side of the occipital flatness) was roughly 4 percent larger than on the contralateral side.
• Similarly, small but consistently significant differences were observed in several other defined linear measurements between the affected and non-affected sides of the mandible.
• Unfortunately, given the preliminary nature of the article, neither the possible effects of these mandibular asymmetries on oral functions nor their response to interventions such as remolding orthoses were investigated.

Kane et al
Neurodevelopmental Implications of Nonsynostostotic Cranial Deformations

Developmental Delay

• Children with DP were more likely to require special education services in school than their non-affected siblings (34.9% vs. 6.6%). Required services included speech therapy, occupational therapy, and physical therapy.
• 36% of children with deformational plagiocephaly had delays in one or more domain on the ASQ (Ages and Stages Questionnaire)
• before any intervention, infants with deformational plagiocephaly show significant delays in both mental and psychomotor development. Also of particular note is that no child with deformational plagiocephaly showed accelerated development.
• Children with DP scored lower on all scales of the BSID-III than children without DP. Differences were largest in cognition, language and adaptive behavior and smallest in motor development. These findings do not imply that DP causes developmental problems, but may nonetheless serve as a marker of developmental risk. Clinicians should screen children with DP for developmental concerns to facilitate early identification and intervention.
Neurodevelopmental Implications of Nonsynostotic Cranial Deformations

Developmental Delay

• Australian infants with DP displayed significantly weaker motor skills than normative estimates, males> females, at very least a diagnostic marker of neurodev. problems

• A positive association between plagiocephaly and developmental delay was reported in 13 of 19 studies, including 4 of 5 studies with “strong” methodological quality.

• This review suggests plagiocephaly is a marker of elevated risk of developmental delays. Motor delay was the most commonly affected domain reported in high-quality papers. Clinicians should closely monitor infants with plagiocephaly.

• These studies suggest that “infants with DP are at increased risk for developmental delays in infancy, and the level of risk is comparable to or even greater than the risk level for infants with craniosynostosis.”

• Results of these studies “very tentatively suggest that DP is associated with increased risk for developmental delay; however a causal association should not be presumed.”

Knight, Martinuk, Collett
Incidence of Deformational Plagiocephaly

- The incidence of plagiocephaly varies widely and is based on anecdotal evidence of increase in the number of referrals to specialty clinics. Five studies have produced varying results, indicating that the incidence of plagiocephaly ranges from 3.1% to 61.0%.
- This is the first study to estimate the incidence of positional plagiocephaly using 4 community-based data collection sites in infants ranging from 7 to 12 weeks of age. The estimated incidence of positional plagiocephaly was found to be 46.6%.
- Of the 440 infants assessed, 205 were observed to have some form of plagiocephaly.
- The incidence of plagiocephaly in infants at 7 to 12 weeks of age was estimated to be 46.6%.
- Of all infants with plagiocephaly, 63.2% were affected on the right side.
- Of all infants with plagiocephaly 78.3% had a mild form

Mawji et al.
Risk Factors for Deformational Plagiocephaly

- Premature
- Gender (boys)
- Birth rank (first)
- Only bottle feeding
- Same side when bottle feeding
- Infant head preference in supine sleeping
- Prone positioning on monitors (tummy time < 3 times/day when awake)

“Children with occipital VP shunts are at significant risk of developing contralateral positional plagiocephaly, particularly in the first 12 months of life.”

Mechanism of Cranial Deformation

Several Hypotheses:

1.) Infant’s head is “soft” or “malleable” (water balloon model)
   -No immediate deformation takes place (severity peaks ~ 4 months)
   -not all infants have their “soft” heads deformed

2.) Inherent problem with bone mineralization
   -no evidence to support this hypothesis either

3.) Hereditary
   -identical twins
   -human cerebrum is not naturally flat or asymmetric

Rogers, 2011
History of Cranial Deformation

» Intentional cranial deformation:
  Common in ancient cultures of Peru, N. American Chinook Indians, French aristocracy and others.
Modern Cranial Deformation

» Incidence of deformational plagiocephaly and brachycephaly has risen drastically since 1996 and the introduction of safe sleep protocol

» Container Babies

» New life saving respiratory equipment
Newton’s first law for cranial deformation

» An object at rest (infant’s head on bed), there will be an equal, but opposite force from the bed to the infant’s head.

» This counterforce will resist cranial growth in the area of contact, and consequently, volume increases will be displaced to areas where there is no resistance.

» The force applied by the head to the resting surface equals the weight of the infant’s head multiplied by the force of gravity. \( (F=mg) \)

» The process is fastest in early infancy and tapers dramatically even after the first year of life.
The pumpkin analogy.

A stationary pumpkin growing against a firm planar surface will become flat over time. The degree of deformation is proportionate to the rate of growth against a constant and fixed external force. This may explain why infants with larger average head sizes (e.g., males and larger infants) and those with rapid rates of head growth (e.g., premature infants) are more likely to develop DP and DB.

Rogers, 2011
Proposed Developmental Pathways for Mechanism of DP and Developmental Delay
Prevention and Treatment Strategies

» 1. Alternating infant’s head of bed direction
» 2. Changing crib position from parallel to perpendicular to headwall
» 3. Rotating infants’ position in bed on schedule between supine, prone, and left/right side-lying
» 4. Providing cares from both sides of bed
» 5. Providing daily prone positioning (when awake at home, on monitors at hospital)
» 6. Alternate feeding positions (use both arms)
» 7. Use supportive equipment for respiratory tubing as necessary
» 8. Assess size and fit of respiratory caps every cares (adjust as necessary)
» 9. Weekly assessment, monitoring and education by physical therapist
» 10. Helmet therapy if severe  (best at 5/6 months-12 months of age, still helps up to 18 months)

“A 4-month standardized pediatric physical therapy program to treat positional preference significantly reduced the prevalence of severe deformational plagiocephaly compared with usual care.”

Danner-Bowman 2015, Rogers, 2011, Vlimmeren 2008
Equipment and Positioning Aides

» Alternative sleeping surfaces
» Custom fabricated foam cups
» Tortle hat
» Cuddle Cup (coming soon from DandleLion)
» Specialized pillows
» Orthotic Helmet/Band
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