Positional head deformity

Background

- **Positional head deformity** is a molding of the skull due to an infant preferentially lying on one side of their head. It usually presents as plagiocephaly (asymmetrical occipital flattening), but brachycephaly (symmetrical occipital flattening) or scaphocephaly (biparietal flattening) can also be seen.
- The current **prevalence** has been estimated at 13%-28%, typically peaking at age 4 months. The rate has increased since 1992, when the Academy of Pediatrics initiated the "Back to Sleep Campaign".
- **Likely risk factors** include congenital muscular torticollis, limited head mobility early in life (particularly in premature infants), prolonged supine positioning, and positional head preference.
- Plagiocephaly may be associated with **developmental delays** in infants.

Evaluation

- Infants **typically present around age 6-8 weeks** with flattening of the skull, usually of the occipital region.
- Frontal and parietal bossing, cheekbone prominence, asymmetrical eye opening, displacement of ear, forehead, and cheek, parietal widening, or expansion of anterior and posterior skull may be seen on **physical exam**.
- The **diagnosis** of a positional head deformity is made primarily based on a typical history and physical exam, and evidence of resolution over time with positional intervention. **Imaging studies** are rarely necessary and should only be ordered by specialists for infants with suspected craniosynostosis.
- Many methods are available to **quantify the head deformity** including cephalic index, oblique cranial length ratio, cranial vault asymmetry, and transcranial diagonal difference.
- The most important other condition to consider is **craniosynostosis**, which should be considered in infants with trapezoidal or other head shape not associated with positional plagiocephaly.

Management

- **Repositioning therapy** is recommended as first-line treatment for mild-to-moderate deformity.
- **Skull-molding helmets or cranial bands** may be used for at least 2-3 months for infants aged 4-12 months with moderate-to-severe deformity who are refractory to positional therapy.
  - Helmets may hasten improvement in plagiocephaly, but may not change long-term outcomes.
  - They appear most effective in infants with a transcranial difference of ≥ 10 mm.
- **Refer infants** to a pediatric neurosurgeon with craniofacial malformation expertise, a craniofacial surgeon, or a craniofacial team if the head deformity does not improve by 4-6 months of age with mechanical adjustments or if craniosynostosis is suspected.

Related Summaries
Craniosynostosis
Congenital muscular torticollis

General Information

Description

- skull molding with flattening of head due to infant preferentially lying on 1 side of head\(^1\)\(^2\)\(^3\)

Also called

- positional plagiocephaly
- positional skull deformity
- benign positional molding
- occipital plagiocephaly
- plagiocephaly without synostosis

Types

- positional head deformity characterized based on affected area of skull\(^2\)
  - plagiocephaly - asymmetrical occipital flattening
  - brachycephaly - symmetrical occipital flattening
  - scaphocephaly - parietal flattening on sides of head (more common in premature infants)
- combination of brachycephaly and some plagiocephaly is most common\(^3\)

Epidemiology

Who is most affected

- infants with preference for 1 head position when lying down (often due to congenital muscular torticollis or prematurity)\(^2\)\(^3\)

Incidence/Prevalence

- prevalence has increased since 1992, when the Academy of Pediatrics initiated the "Back to Sleep Campaign" to prevent sudden infant death syndrome\(^1\)\(^3\)
- 13%-28% reported prevalence among infants\(^1\)\(^3\)
- high range in reported prevalence due to variation in diagnostic criteria and definition of deformation\(^3\)
- prevalence typically peaks at age 4 months\(^1\)\(^3\)
- 13% prevalence of cranial flattening at birth in cohort of 183 single-born infants (Pediatrics 2002 Dec;110(6):e72)
- 22.1% prevalence of deformational plagiocephaly at age 7 weeks
  - based on prospective cohort study
  - 380 infants were evaluated at birth and at age 7 weeks for deformational plagiocephaly
  - plagiocephaly defined as oblique diameter difference index ≥ 104%
• deformational plagiocephaly observed in 6.1% of infants at birth and in 22.1% of infants at age 7 weeks
  - Reference - *Pediatrics* 2007 Feb;119(2):e408

• prevalence of positional head deforming decreases after age 4 months
  - based on prospective cohort study
  - 200 infants were evaluated for head shape from ages 6 weeks to 2 years
    - plagiocephaly was defined as oblique cranial length ratio ≥ 106%
    - brachycephaly was defined as cephalic index ≥ 93%
  - plagiocephaly or brachycephaly in
    - 16% of infants at 6 weeks
    - 19.7% of infants at 4 months
    - 9.2% of infants at 8 months
    - 6.8% of infants at 1 year
    - 3.3% of infants at 2 years
  - Reference - *Pediatrics* 2004 Oct;114(4):970

• 1%-2% prevalence of cranial abnormality in children aged 12-17 years
  - based on cross-sectional study
  - 1,045 children and adolescents aged 12-17 years were evaluated for skull deformities and abnormal facial characteristics
    - plagiocephaly was defined as cranial vault asymmetry > 1 cm
    - brachycephaly was defined as cephalic index > 0.9
  - 2% of all patients had cranial abnormality
    - plagiocephaly in 1.1% of all patients
    - brachycephaly in 1% of all patients
  - Reference - *Otolaryngol Head Neck Surg* 2012 May;146(5):823

Likely risk factors

• limited head mobility in infant early in life, particularly in premature infants (1,2,3)
• prolonged supine positioning (1)
• **congenital muscular torticollis** found in 70%-95% of infants with deformational plagiocephaly (1)
• positional head preference (1,2)

Possible risk factors

• risk factors for positional plagiocephaly at birth include (1,2)
  - multiple gestation pregnancy
  - large for gestational age
  - oligohydramnios
  - breech or transverse position
  - male sex
  - first born
risk factors for development of head deformity after birth include (1; 3)
- prematurity with associated neuromuscular immaturity
- developmental delay such as infants with hypotonia or hypertonia
- brachycephaly at birth
- early motor skill delay in upper body strength and rolling over from being placed in supine position to sleep, which resolves over time

risk factors for development of positional plagiocephaly include limited newborn passive head rotation and supine only sleeping
- based on prospective cohort study
- 200 infants recruited at birth were evaluated for head shape from ages 6 weeks to 2 years
- increased risk of positional plagiocephaly associated with
  - limited passive head rotation as newborn
    - odds ratio [OR] 6.17 for being diagnosed at age 6 weeks (95% CI 2.03-18.76)
    - OR 7.78 for being presence of plagiocephaly at age 4 months (95% CI 2.56-23.7)
  - sleeping only in supine position
    - OR 11.53 if done as newborn (95% CI 2.67-49.81)
    - OR 4.65 if done at 6 weeks (95% CI 1.82-11.89)
  - unsuccessful attempt at variation in head position when sleeping (OR 5.15, 95% CI 1.9-13.98)
  - lack of variation in head position when sleeping at 6 weeks (OR 2.75, 95% CI 1.11-6.82)
  - ≥ 21 hours of supine position per day
    - OR 3.2 if done at 6 weeks (95% CI 1.47-6.97)
    - OR 2.43 if done at 4 months (95% CI 1.47-5.2)
  - limited active head rotation at 4 months (OR 5.15, 95% CI 1.9-13.98)
- Reference - Pediatrics 2004 Oct;114(4):970

risk factors for development of deformational plagiocephaly at 7 weeks include same side head positioning and less daily tummy time
- based on prospective cohort study
- 380 infants were evaluated at birth and at age 7 weeks for deformational plagiocephaly
- risk factors for development of deformational plagiocephaly at age 7 weeks include
  - head positioned to same side (head preference) when
    - awake (odds ratio [OR] 9.5, 95% CI 5.3-17.01)
    - sleeping (OR 7.1, 95% CI 3.9-12.78)
    - bottle feeding (OR 1.9, 95% CI 1.15-3.14)
  - tummy time < 3 times per day (OR 2.7, 95% CI 1.12-6.55)
  - male sex (OR 1.8, 95% CI 1.11-2.96)
  - first born (OR 1.8, 95% CI 1.1-2.94)
- Reference - Pediatrics 2007 Feb;119(2):e408 full-text

deformational plagiocephaly appears to correlate with sleep position, with lateralization corresponding to side of head slept on
Based on retrospective cohort study

- 20,691 children treated for deformational plagiocephaly with orthotic cranial bands were assessed for risk factors and compared with expected frequency of factors in general population (based on published statistics)
- Deformational plagiocephaly significantly associated with
  - primiparity
  - nonvertex intrauterine presentation
  - twinning
  - right-sided lateralization
- Right vs. left lateralization associated with sleep position on ipsilateral side or supine position with head turned toward ipsilateral side (p < 0.0001)

Reference - Pediatrics 2009 Dec;124(6):e1126

Risk factors for development of plagiocephaly include limited change in head position when sleeping

- Based on case-control study
- 100 infants diagnosed with nonsynostotic plagiocephaly and 200 controls were evaluated at 6 weeks and mean age 25-28 weeks by surveying mothers via questionnaires
- Risk factors for plagiocephaly include
  - Infant head placement at 6 weeks
    - Head preference (adjusted odds ratio [OR] 37.46, 95% CI 8.44-66.32)
    - Limited head rotation (adjusted OR 20.4, 95% CI 5.83-71.35)
    - No variation in head position when sleeping (adjusted OR 7.11, 95% CI 2.75-18.37)
    - Unsuccessful attempt at variation in head position when sleeping (adjusted OR 7.38, 95% CI 2.43-22.42)
  - Sleeping supine at 6 weeks (adjusted OR 7.02, 95% CI 2.98-16.53)
  - < 37 weeks gestation (adjusted OR 3.26, 95% CI 1.02-10.47)
  - Reduced activity level (adjusted OR 3.32, 95% CI 1.38-7.56)
  - Developmental delay (adjusted OR 3.32, 95% CI 1.01-10.85)
  - First born (adjusted OR 2.94, 95% CI 1.23-5.16)
  - Male sex (adjusted OR 2.51, 95% CI 1.23-5.16)

Reference - Pediatrics 2003 Oct;112(4):e316

Associated conditions

- Developmental delay may be associated with plagiocephaly
  - Infants with deformational plagiocephaly may have higher rates of neurodevelopmental delay in first 3 years of life
    - Based on case-control study
    - 233 infants born ≥ 35 weeks gestation and aged 4-12 months with plagiocephaly and 167 controls were assessed by Bayley Scales of Infant Development III (BSID) at mean age 7 months
    - Comparing infants with plagiocephaly vs. infants without plagiocephaly
- plagiocephaly associated with increased rate of neurodevelopmental delay (score < 85)
  - motor delay in 19.7% vs. 9% (odds ratio 2.34, 95% CI 1.25-4.39)
  - language delay in 18.5% vs. 10.2% (odds ratio 1.84, 95% CI 1-3.39)
  - cognitive delay in 3.9% vs. 0%
- plagiocephaly associated with significantly lower mean BSID scores
  - cognitive score 102.1 vs. 107.2 (p < 0.001)
  - language composite score 92.3 vs. 97.7 (p < 0.001)
  - motor composite score 93.4 vs. 103.8 (p < 0.001)

Reference - *Pediatrics 2010 Mar;125(3):e537 full-text*

- similar results for rates of neurodevelopmental delay (score < 85) in follow-up for 216 case infants and 162 controls at mean age 18.5 months
  - cognitive delay in 8.3% vs. 0.6% (adjusted risk ratio [RR] 13.8, 95% CI 1.8-105.5)
  - language delay in 18.5% vs. 6.1% (adjusted RR 3.9, 95% CI 1.6-6.2)
  - motor delay in 6% vs. 1.8% (adjusted RR 3.2, 95% CI 1.1-13.1)

- similar results for rates of neurodevelopmental delay (score < 85) in long-term follow-up of 215 case infants and 163 controls at mean age 3 years
  - cognitive delay in 3.1% vs. 0%
  - language delay in 5.3% vs. 1.2% (adjusted risk ratio [RR] 7.9, 95% CI 1.8-35.1)
  - motor delay in 5.8% vs. 1.8% (adjusted RR 4.3, 95% CI 1-17.9)
- Reference - *Pediatrics 2013 Jan;131(1):e109 full-text*

- developmental delay reported in 30% of infants presenting with plagiocephaly
  - based on case series
  - 126 infants < 1 year old (mean age 5.2 months at initial assessment) with plagiocephaly had developmental assessments at 3, 6, and 12 months after enrollment
  - evaluation for communication, gross motor, fine motor, problem solving, and personal/social development using Ages and Stages Questionnaire, second edition (ASQ-2)

<table>
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<tr>
<th>Time of Assessment</th>
<th>Communication</th>
<th>Gross Motor</th>
<th>Fine Motor</th>
<th>Problem Solving</th>
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<td>12.5%</td>
<td>13.%</td>
<td>6.7%</td>
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<tr>
<td>3-month assessment</td>
<td>4%</td>
<td>28%</td>
<td>12.8%</td>
<td>10.4%</td>
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<tr>
<td>6-month assessment</td>
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<td>22.9%</td>
<td>4.1%</td>
<td>7.4%</td>
</tr>
<tr>
<td>12-month assessment</td>
<td>4.9%</td>
<td>12.2%</td>
<td>4.1%</td>
<td>8.9%</td>
</tr>
</tbody>
</table>


- left-sided plagiocephaly associated with more severe developmental delay in older children compared with right-sided plagiocephaly
based on retrospective cohort study

80 parents of patients with plagiocephaly responded to survey about cosmetic presentation and performance of patients at median age 9 years

reported developmental delay outcomes include
- language difficulty in 21%
- motor difficulty in 28%
- special education requirements in 15%

reported cognitive outcomes in left-sided vs. right-sided plagiocephaly include
- special education requirements in 27% vs. 10% (p < 0.05)
- fine motor delay in 41% vs. 22% (p < 0.05)
- speech delay in 36% vs. 16% (p < 0.05)


- no strong evidence for association with scoliosis or hip dislocation

Etiology and Pathogenesis

Causes
- consistent contact on 1 side of head due to torticollis or infant preference for 1 direction over another during time spent in supine position (such as sleeping, laying on back, and sitting in car seat)

Pathogenesis
- characteristics of normal development of infant skull that make it susceptible to positional flattening
  - cranial growth is most rapid in early infancy and slows dramatically after age 1 year
  - growth is more pronounced in occipital/parietal regions than in frontal area
  - prior to about 4 months, infants have poor muscle tone that prevents them from holding head upright unsupported and from rolling over
- flattening occurs when cranial expansion and growth are consistently resisted in specific area by external force (such as a flat surface when infant lying supine)
- other asymmetry that can result from asymmetric occipital flattening includes
  - frontal bossing on same side of flattened occiput
  - anterior shift of ipsilateral forehead, ear, and cheek
  - more open appearance of eye on side of flattening due to sagittal displacement of ipsilateral zygoma

History and Physical

History

Chief concern (CC)
- flattening of infant head usually presenting around age 6-8 weeks

History of present illness (HPI)
• ask parents about\(^1,2\)
  o skull shape at and a few days after birth
  o infant's preference for head position when lying down or sleeping, in car seat, and when feeding
    ▪ identifies infants most at risk for developing positional flattening of head
    ▪ significant preference in head rotation to 1 side is indicative of cervical imbalance or \texttt{torticollis}
  o amount of time infant spends in a supervised prone position (tummy time) each day

**Family history (FH)**
• ask about family history of \texttt{craniosynostosis}

**Physical**

**HEENT**
• perform examination of head to assess for\(^1,2\)
  o frontal (forehead) and parietal bossing
  o cheekbone prominence
  o displacement of ear, forehead, and cheek
  o symmetrical occipital flattening
  o parietal widening
  o flattening on side of head
  o expansion anteriorly and posteriorly
• while looking down at top of head, assess approximate shape formed by position of ears and cheekbones\(^1\)
• look for subtle facial deformities, such as\(^1,3\)
  o contralateral facial flattening
  o more open appearing eye on side of head with increased forehead bossing
• methods to quantify head deformity\(^2,3\)
  o cephalic index
    ▪ most commonly used for brachycephaly and scaphocephaly
    ▪ defined as width of the head divided by the anteroposterior length \(\times 100\)
      o cranial width/breadth - maximum horizontal biparietal diameter of head
      o cranial length - distance from glabella to opisthocranion
    ▪ Reference - \texttt{Neurosurg Focus 2013 Oct;35(4):E2}
  o oblique cranial length ratio
    ▪ may be used for plagiocephaly
    ▪ defined as ratio of longer cranial cross-diagonal to shorter cross-diagonal
    ▪ in North America, plagiocephaly typically defined as oblique cranial length ratio of \(\geq 105\%\) or \(106\%\)
- limitation of oblique cranial length ratio is that asymmetry will artificially appear to improve as head size grows
- normal values for oblique cranial length ratio and cephalic index vary among cultures, so cutoff values for defining mild, moderate, or severe deformity and when to treat also vary
  - transcranial diagonal difference
    - used to quantify plagiocephaly
    - defined as difference between longer cranial cross-diagonal and shorter cross-diagonal
    - calculated from 2 oblique head measurements obtained using anthropometric caliper
    - in absence of anthropometric caliper, degree of flattening can be measured using fingertips
      - place fingertips on opposite sides of occiput in parallel lines posterior to anterior lobe
      - estimate offset between sides of posterior cranium (typical finger width is 8-11 mm)
  - cranial vault asymmetry
    - distance between left frontozygomatic point and right euryon point (defined as point on either parietal bone marking either end of greatest transverse diameter of skull) - distance between right frontozygomatic point and left euryon point
    - cranial vault asymmetry index - cranial vault asymmetry/shorter frontozygomatic-euryon distance × 100
  - other methods used to measure plagiocephaly may include
    - 3-dimensional (3-D) digital head shape measurements acquired using noninvasive laser scanner (STARscanner) (J Craniofac Surg 2006 Nov;17(6):1084)
    - full axial projection radiographs
    - Ezeform ring mold of cranium or Heads-Up band digitally photographed and analyzed by software
    - thermoplastic material wrapped around widest part of infant's head, traced onto paper, and measured
    - elastic headband wrapped around infant's head and digitally photographed
    - Reference - Dev Med Child Neurol 2008 Aug;50(8):568

Neck
- assess for torticollis
  - look for limitation of active rotation of head away from flattened occiput
  - palpate sternocleidomastoid for nodule
  - consider performing rotating chair/stool test (examiner rotates on rotating chair or stool while holding infant facing parent, while parents attempts to keep gaze of infant)
  - see also Congenital muscular torticollis

Diagnosis
Making the diagnosis

- Diagnosis made primarily based on history, physical exam, and resolution over time with positional intervention\(^1\,^3\)
  - Plagiocephaly is diagnosed when there is ipsilateral frontal (forehead) and parietal bossing, cheekbone prominence, and displacement of ipsilateral ear, forehead, and cheek
    - Parallelogram shape due to unilateral occipital flattening with ipsilateral frontal bossing is common
    - Anterior displacement of ear may be indicative of either positional plagiocephaly or craniosynostosis
    - Posterior displacement of ear more common with craniosynostosis
  - Brachycephaly is diagnosed when there is symmetrical occipital flattening and compensatory parietal widening
  - Scaphocephaly is diagnosed when there is flattening on side of head and compensatory expansion anteriorly and posteriorly

Differential diagnosis

- Craniosynostosis\(^1\,^3\)
  - Premature fusion of 1 or more cranial vault sutures resulting in a variety of craniofacial abnormalities
  - May appear at birth, but diagnosed at mean age 4.1 months (J Craniofac Surg 2013 Jan;24(1):96)
  - May be suspected with trapezoidal shape (due to occipital and frontal flattening) looking down on top of head
  - Unilateral coronal synostosis (premature closure of 1 coronal suture) causes anterior plagiocephaly
    - Flattening of forehead and superior orbital rim with protrusion of anterior globe
    - Nasal root and midfacial angulation
    - Anterior displacement of ipsilateral ear, though may also be posterior
    - Eye that appears more open on the same side as the flattened forehead
  - Lambdoidal synostosis (more rare) may present with
    - Asymmetric cranial height with a shorter flattened side (wind-swept appearance of head)
    - Mastoid bossing on affected (flattened) side
    - Parallelogram head shape with affected ear displaced posteriorly and inferiorly (in contrast to anterior placement with deformational plagiocephaly)
  - Also consider in utero or intrapartum head molding due to\(^1\,^3\)
    - Uterine constraint (especially in multigestational pregnancy)
    - Forceps or vacuum-assisted birth
    - Breech delivery
    - Large for gestational age
    - Oligohydramnios
Testing overview

- Imaging (skull x-ray or cranial computed tomography) is unnecessary in most situations\(^1\)\(^2\)
  - Avoid imaging in infants born with normal head shape that changes progressively during first few weeks of life
  - Rarely indicated for infants with torticollis, unless progressive or associated with other clinical findings

- Reserve imaging (usually cranial computed tomography) for infants with suspected craniosynostosis, including infants\(^1\)
  - Born with deformities
  - With no improvement after several weeks with repositioning

- Imaging studies should be ordered by craniofacial surgeon\(^1\)\(^2\)

Treatment

Treatment overview

- Consider parents’ degree of concern along with physical exam when evaluating treatment options and timing\(^2\)

- Repositioning therapy is recommended as first-line treatment for mild-to-moderate deformity

- Skull-molding helmets or cranial bands may be used for infants aged 4-12 months
  - With moderate-to-severe deformity if refractory to positional therapy
  - Most effective if used for at least 2-3 months with transcranial difference ≥ 10 mm

- Refer patient to pediatric neurosurgeon with craniofacial malformation expertise, a craniofacial surgeon, or craniofacial team if head deformity does not improve by age 4-6 months with mechanical adjustments or if craniosynostosis is suspected

Activity

- Repositioning therapy recommended as first-line treatment option for mild-to-moderate deformity\(^1\)\(^2\)
  - Positional adjustments include
    - Alternating supine head position (left vs. right occiput) nightly
    - Reposition crib in room to require child to look away from flattened side of head to see people in room
    - Awake infant should spend ≥ 30-60 minutes/day in a supervised prone position (tummy time)
    - Minimize time infant spends in car seats and swings
  - No evidence comparing positional adjustments vs. no intervention for positional plagiocephaly ([Dev Med Child Neurol 2005 Aug;47(8):563 PDF](#))

- Physical therapy intervention may reduce risk for severe deformational plagiocephaly at age 6 and 12 months compared to parent education ([level 2 [mid-level] evidence](#))
  - Based on randomized trial with unclear allocation concealment and without long-term outcome comparison
65 infants aged 7 weeks with positional preference randomized to maximum 8 sessions of physical therapy by age 6 months plus written basic preventive measures vs. written basic preventive measures provided to parent
  • physical therapy sessions included
    o exercises to reduce positional preference and to stimulate motor development
    o parental counseling about counterpositioning, handling, and nursing infant
  • written basic preventive measures were provided to parents in both groups

severe plagiocephaly was defined as oblique diameter difference index (ODDI) score ≥ 104%

physical therapy intervention associated with reduced risk for severe deformational plagiocephaly
  • at 6 months (relative risk 0.54, 95% CI 0.3-0.98)
  • at 12 months (relative risk 0.43, 95% CI 0.22-0.85)


• if torticollis present
  o perform neck-stretching exercises with each diaper change
  o if no improvement after 2-3 months, refer patient to physical therapist

Surgery and procedures

• surgery not recommended for positional skull deformity, unless severe deformity persists despite mechanical adjustments and other causes are suspected (such as craniosynostosis)

Orthosis

• custom-made orthotic helmets may be used for patients with moderate-to-severe deformity
  o most effective if used for 2-3 months in patients aged 4 to 12 months with transcranial difference ≥ 10 mm
    ▪ may be used up to age 18 months
    ▪ correction may take 6-8 months
  o rarely disrupt sleep patterns
  o helmets may reduce plagiocephaly more quickly than standard repositioning therapy, but evidence for superior long-term outcomes with helmets lacking
    ▪ orthotic helmets or cranial bands may be more effective in short-term for reducing plagiocephaly than repositioning methods, but long-term efficacy unknown
      • based on systematic review of observational studies with methodologic limitations
      • systematic review of 11 observational studies (9 cohort and 2 case-control) comparing repositioning methods and orthotic helmets and cranial bands for treatment of posterior positional plagiocephaly
      • methodologic limitations included
        • high or differential loss to follow-up
• orthosis used immediately in some studies and after unsuccessful treatment by repositioning in others
• intervention duration ranged from 1.2 months to 16 months and varied between orthosis and positioning
• mean age at start of treatment ranged from 3 months to 9 months
• mean age at end of treatment ranged from 5 months to 24 months
• only 2 studies extended follow-up beyond end of the intervention
  o superior short-term efficacy for orthotic management in treating plagiocephaly reported in 8 studies with 1,032 patients
  o mean age at start of treatment ranged from 3 months to 9 months
  o mean age at end of treatment ranged from 8 months to 12 months
  o similar efficacy between repositioning and orthotic treatments reported in 3 studies
• Safe T Sleep mattress and repositioning did not improve cranial asymmetry more than repositioning alone after 12 month treatment with results limited by low adherence to mattress use in 1 study with 126 patients
• cranial bands did not improve cranial asymmetry more than repositioning alone after 4.5-month treatment with results limited by different measures of cranial asymmetry used for each group in 1 study with 112 patients
• helmet did not improve cranial asymmetry index more than repositioning with results limited by 3-fold differences in treatment duration (15 months for helmet group vs. 5.5 months for repositioning group) in 1 study with 74 patients
  o Reference - Ann Phys Rehabil Med 2013 Apr;56(3):231
• in infants aged 5-6 months helmet therapy is not more effective in improving positional skull deformation at age 2 years than no helmet ([level 1 [likely reliable] evidence])
  o based on cluster randomized trial
  o 84 term infants aged 5-6 months with moderate-to-severe positional skull deformation were randomized to helmet therapy vs. no helmet and followed to age 2 years
  • helmet was to be worn for 23 hours/day to age 1 year, or less if results were considered satisfactory
  • no infants had muscular torticollis, craniosynostosis, or dysmorphic features
  • all infants had already received physiotherapy prior to enrollment
  o additional treatment for skull deformation was discouraged in both groups
  o outcome assessors were blinded
  o 24% in helmet therapy group received treatment until age 12 months (mean age at discontinuation 10 months)
  o full recovery (defined as oblique diameter difference index < 104% and cranioproportional index < 90%) in 26% with helmet vs. 23% with no treatment (not significant)
  o no significant differences in
  • changes in plagiocephaly or brachycephaly
  • infant ear deviation, facial asymmetry, occipital lift, motor development, or quality of life
• parent satisfaction
  o parental anxiety was less in the helmet therapy group (p = 0.04)
  o side effects of helmet included skin irritation, sweating, pain, and hindered cuddling between parent and child
  o Reference - HEADS trial (BMJ 2014 May 1;348:g2741 full-text)

• in infants aged 6 months with plagiocephaly helmet therapy may improve cranial asymmetry by mean age 10.2 months more than supportive interventions did by mean age 18.5 months (level 2 [mid-level] evidence)
  o based on prospective cohort study with differential follow-up and without long-term outcomes
  o 128 infants with positional plagiocephaly were evaluated after treatment with or without helmet therapy
    • 62 infants started helmet therapy at mean age 6.3 months and evaluated at mean age 10.2 months
    • 66 infants without helmet started study at mean age 6.2 months and evaluated 12 months later at mean age 18.5 months
    • parents in both groups were instructed to use supportive interventions (physiotherapy, osteopathy, repositioning)
  o cranial vault asymmetry index (CVAI) was calculated from cranial diagonal measurements taken with anthropometric metal cranial caliper
    • CVAI of 0% defined perfect symmetry
    • CVAI > 3.5% defined as significant asymmetry according to Loveday and de Chalain
    • CVAI ≥ 5% defined as significant asymmetry according to Regelsberger
  o comparing infants with helmet therapy at end of treatment vs. infants without helmet therapy at end of treatment (p < 0.0001)
    • CVAI at end of treatment 4.1% vs. 6.3%
    • absolute median reduction in CVAI from baseline 9.2% vs. 2.7%
    • relative median reduction in CVAI from baseline 68.3% vs. 30.7%

• younger patients reported to achieve higher rates of correction of head deformity within 6 months using helmet therapy than older patients, but > 50% of patients initiating helmet therapy at age > 12 months reported to achieve correction of head deformity (level 3 [lacking direct] evidence)
  o based on case series
  o 1,050 children aged 19 days to 22 months who had helmet therapy for Argenta Type II-V plagiocephaly followed up for mean 6.3 months
    • Argenta levels of deformity
      o Type I defined as asymmetrical occipital flattening only
      o Type II defined as Type I plus malposition of affected ear
      o Type III defined as Type II plus forehead deformity
      o Type IV defined as Type III plus malar deformity
      o Type V defined as Type IV plus cranial decompression vertically or temporally
    • correction defined as achievement of Type I or 0 deformity
overall correction rate 81.6%
- rate of correction to Type I or 0 deformity increased with reduced age at time of therapy initiation (no p values reported)
  - 93.3% of 60 patients ≤ 3 months old
  - 82.9% of 584 patients aged 4-6 months
  - 81.5% of 330 patients aged 7-9 months
  - 65.6% of 61 patients aged 10-12 months
  - 53.3% of 15 patients > 12 months old
- mean time to correction decreases with severity
  - 3.1 months for Type II (177 patients)
  - 4.7 months for Type III (560 patients) (p < 0.0001 vs. Type II)
  - 5.4 months for Type IV (265 patients) (p < 0.0001 vs. Type II)
  - 5.6 months for Type V (48 patients) (p < 0.0001 vs. Type II)
- Reference - Neurosurg Focus 2013 Oct;35(4):E4

- infants with plagiocephaly that start helmet therapy at age 5 months or younger may have more improvement in cranial asymmetry after 7 months (level 2 [mid-level evidence])
  - based on prospective cohort study without long-term outcomes
  - 108 infants aged 3-43 months (mean 9 months) with plagiocephaly treated with helmet therapy for mean duration 6.6 months evaluated with 3-dimensional (3-D) scans and cranial measurements using caliper
  - comparing initial vs. final scores (no p values reported)
    - mean cranial vault asymmetry 16 mm vs. 4.7 mm
    - mean cranial vault asymmetry index 10.7% vs. 3%
  - cranial vault asymmetry scores and cranial vault asymmetry index scores improved most with
    - younger starting age (≤ 5 months)
    - best compliance (≥ 20 hours of helmet wearing/day)
  - infants with poor helmet compliance (< 15 hours/day) or severe deformity required longest helmet therapy duration (mean > 7 months)

- adverse effects with helmet use
  - reported short-term complications include
    - occasional malodorous perspiration
    - minor skin irritation/contact dermatitis
    - social stigma
    - cost
    - pressure spots and hair loss
    - parental dissatisfaction
  - no long-term complications have been reported
pressure sores and skin reactions may occur in children with plagiocephaly and/or brachycephaly treated with helmet orthosis
  o based on case series
  o 410 children with plagiocephaly treated with helmet orthosis evaluated for complications associated with therapy
  o 22.4% had complications including
    • pressure sores in 10%
    • skin reaction due to accumulation of cleaning fluids in 6.3%
    • deficient fitting in 5.9%
    • failure of treatment in 1.2%
    • skin infection in 0.5%
    • bacterial abscess in 0.24%
  o no significant difference in complication rate among severities of cranial deformity

- infant sleep positioners not recommended
  o FDA and Consumer Product Safety Commission (CPSC) warn consumers not to use infant sleep positioners due to possibility of suffocation (*FDA MedWatch 2010 Sep 29, FDA Press Release 2010 Sep 29*)
  o 13 infant deaths (most ≤ 3 months old) associated with use of infant sleep positioners in United States during 1997-2011 (*MMWR Morb Mortal Wkly Rep 2012 Nov 23;61:933 full-text*)
- a review of helmet therapy for treating plagiocephaly can be found in *Neurosurg Focus 2013 Oct;35(4):E2*

Consultation and referral
- if head deformity does not improve by age 4-6 months after mechanical adjustments, refer patient to pediatric neurosurgeon with craniofacial malformation expertise, a craniofacial surgeon, or craniofacial team[^1]
- if torticollis does not improve after 2-3 months of neck-stretching exercises, refer patient to physical therapist[^1]

Follow-up
- monitor head shape at routine well-child visits until infant can sit up, crawl, shows no signs of torticollis, and generally spends less time in supine position[^1]
- **assess for developmental abnormalities** closely at routine well-child visits

Complications and Prognosis

Complications
- persistent, unidirectional positioning and limited neck motion may result in torticollis or *cervical dystonia*[^1]
- no strong evidence for positional skull deformity causing[^1,2]
  o otitis media
  o temporomandibular joint syndrome
- mandibular asymmetry
- abnormal vision development

**Prognosis**

- benign condition with good cosmetic prognosis (1-2)
- most deformities improve spontaneously within first few months of life if infant avoids resting head predominantly on flattened area of skull
- if deformity does not resolve spontaneously, mechanical adjustments and exercises for 2-3 months typically yield significant improvement by age 6 months
- progression of resolution
  - in infants small noticeable differences in facial projection typically disappear with growth
  - in older children asymmetry is typically less noticeable
  - some adolescents may have mild head deformity
  - most adults have slight craniofacial asymmetry
- head shape abnormalities and developmental delay associated with deformational plagiocephaly or brachycephaly in infancy usually resolve by age 4 years (level 2 [mid-level] evidence)
  - based on long-term follow-up of cohort study
  - 129 children, mean age 5.8 months, diagnosed with deformational plagiocephaly or brachycephaly were reevaluated at age 3-4 years with head shape measurements of cephalic index (CI) and oblique cranial length ratio (OCLR) used to define severity of deformity
  - all parents were counseled about repositioning strategies, and infants were referred for physiotherapy if neck muscle dysfunction present
- degrees of deformity
  - normal defined as CI < 93% and OCLR < 106%
  - mild defined as CI between 93% and 96% and OCLR ≤ 108%, or CI ≤ 93% and OCLR between 106% and 108%
  - moderate defined as CI between 96% and 99% and OCLR ≤ 110%, or CI ≤ 96% and OCLR between 108% and 110%
  - severe defined as CI > 99% or OCLR > 110%
- comparing initial vs. follow-up assessment
  - ≥ 1 area of developmental delay in 41% vs. 11% (p < 0.05)
  - head shape measurements (no p values reported)
    - normal range in 0% vs. 61%
    - mild range in 22% vs. 26%
    - moderate range in 31% vs. 9%
    - severe range in 47% vs. 4%
  - Reference - Arch Dis Child 2011 Jan;96(1):85

**Prevention and Screening**

**Prevention**
• provide counseling to parents about measures to prevent skull deformity or to prevent progression; recommendations include:
  o beginning at birth, alternate supine head position (left vs. right occiput) nightly
  o awake infant should spend ≥ 30-60 minutes/day in a supervised prone position (tummy time)
  o minimize time infant spends in car seats and swings
  o continue above measures until infant can sit up independently
• infant sleep positioners not recommended
  o FDA and Consumer Product Safety Commission (CPSC) warn consumers not to use infant sleep positioners due to possibility of suffocation (FDA MedWatch 2010 Sep 29, FDA Press Release 2010 Sep 29)
  o 13 infant deaths (most ≤ 3 months old) associated with use of infant sleep positioners in United States during 1997-2011 (MMWR Morb Mortal Wkly Rep 2012 Nov 23;61:933 full-text)

Screening
• screen for positional skull deformity at each health checkup until age 1 year

Guidelines and Resources

Guidelines

United States guidelines
• Cincinnati Children's Hospital Medical Center (CCHMC) Best evidence statements (BESt) on
  o use of care giver education to prevent positional plagiocephaly can be found at CCHMC 2012 Jun PDF or at National Guideline Clearinghouse 2012 Oct 29:37637
  o prognosis of infant development with plagiocephaly, torticollis can be found at CCHMC 2011 Jun PDF or at National Guideline Clearinghouse 2012 Jan 2:34044

Canadian guidelines
• Canadian Paediatric Society (CPS) guidelines on positional plagiocephaly can be found in Paediatr Child Health 2011 Oct;16(8):493 full-text [English, French]

European guidelines
• Swedish expert guideline on child health care nurses to prevent nonsynostotic plagiocephaly can be found in J Pediatr Nurs 2011 Aug;26(4):348

Review articles
• review can be found in Childs Nerv Syst 2011 Nov;27(11):1867
• review of positional plagiocephaly guidelines can be found in Neurosurg Focus 2013 Oct;35(4):E1
• review of assessment and diagnosis of deformational plagiocephaly can be found in J Pediatr Health Care 2012 Jul-Aug;26(4):242
• review of management of deformational plagiocephaly can be found in J Pediatr Health Care 2012 Sep-Oct;26(5):320
• review of diagnosis and management of deformational plagiocephaly can be found in J Neurosurg Pediatr 2009 Apr;3(4):284, commentary can be found in J Neurosurg Pediatr 2010 Feb;5(2):219
• review of prevalence, risk factors, and natural history of positional plagiocephaly can be found in Dev Med Child Neurol 2008 Aug;50(8):577
• review of plagiocephaly and awareness, prevention, and treatment can be found in Community Pract 2008 Apr;81(4):38

MEDLINE search
• to search MEDLINE for (Plagiocephaly) with targeted search (Clinical Queries), click therapy, diagnosis, or prognosis

Patient Information
• handout from American Association of Neurological Surgeons
• handout from Children’s Hospital & Research Center Oakland PDF
• handout from Kids Health
• handout from Cincinnati Children’s Hospital Medical Center or in Spanish

ICD-9/ICD-10 Codes

ICD-9 codes
• 754.0 congenital musculoskeletal deformities of skull, face, and jaw
• 756.0 anomalies of skull and face bones

ICD-10 codes
• Q67 congenital musculoskeletal deformities of head, face, spine and chest
  o Q67.0 facial asymmetry
  o Q67.1 compression facies
  o Q67.2 dolichocephaly
  o Q67.3 plagiocephaly

References

General references used
• 1. Laughlin J, Luerssen TG, Dias MS, Committee on Practice and Ambulatory Medicine, Section on Neurological Surgery. Prevention and management of positional skull deformities in infants. Pediatrics. 2011 Dec;128(6):1236-41 full-text, correction can be found in Pediatrics 2012 Mar;129(3):595

Synthesized Recommendation Grading System for DynaMed Plus
- DynaMed systematically monitors clinical evidence to continuously provide a synthesis of the most valid relevant evidence to support clinical decision-making (See 7-Step Evidence-Based Methodology).

- Guideline recommendations summarized in the body of a DynaMed topic are provided with the recommendation grading system used in the original guideline(s), and allow DynaMed users to quickly see where guidelines agree and where guidelines differ from each other and from the current evidence.

- In DynaMed Plus (DMP), we synthesize the current evidence, current guidelines from leading authorities, and clinical expertise to provide recommendations to support clinical decision-making in the Overview & Recommendations section.

- We use the Grading of Recommendations Assessment, Development and Evaluation (GRADE) to classify synthesized recommendations as Strong or Weak.
  - Strong recommendations are used when, based on the available evidence, clinicians (without conflicts of interest) consistently have a high degree of confidence that the desirable consequences (health benefits, decreased costs and burdens) outweigh the undesirable consequences (harms, costs, burdens).
  - Weak recommendations are used when, based on the available evidence, clinicians believe that desirable and undesirable consequences are finely balanced, or appreciable uncertainty exists about the magnitude of expected consequences (benefits and harms). Weak recommendations are used when clinicians disagree in judgments of relative benefit and harm, or have limited confidence in their judgments. Weak recommendations are also used when the range of patient values and preferences suggests that informed patients are likely to make different choices.

- DynaMed Plus (DMP) synthesized recommendations (in the Overview & Recommendations section) are determined with a systematic methodology:
  - Recommendations are initially drafted by clinical editors (including ≥ 1 with methodological expertise and ≥ 1 with content domain expertise) aware of the best current evidence for benefits and harms, and the recommendations from guidelines.
  - Recommendations are phrased to match the strength of recommendation. Strong recommendations use "should do" phrasing, or phrasing implying an expectation to perform the recommended action for most patients. Weak recommendations use "consider" or "suggested" phrasing.
  - Recommendations are explicitly labeled as Strong recommendations or Weak recommendations when a qualified group has explicitly deliberated on making such a recommendation. Group deliberation may occur during guideline development. When group deliberation occurs through DynaMed-initiated groups:
    - Clinical questions will be formulated using the PICO (Population, Intervention, Comparison, Outcome) framework for all outcomes of interest specific to the recommendation to be developed.
    - Systematic searches will be conducted for any clinical questions where systematic searches were not already completed through DynaMed content development.
    - Evidence will be summarized for recommendation panel review including for each outcome the relative importance of the outcome, the estimated effects comparing intervention and comparison, the sample size, and overall quality rating for the body of evidence.
    - Recommendation panel members will be selected to include at least three members that together have sufficient clinical expertise for the subject(s) pertinent to the
recommendation, methodological expertise for the evidence being considered, and experience with guideline development.

- All recommendation panel members must disclose any potential conflicts of interest (professional, intellectual, and financial), and will not be included for the specific panel if a significant conflict exists for the recommendation in question.
- Panel members will make **Strong recommendations** if and only if there is consistent agreement in a high confidence in the likelihood that desirable consequences outweigh undesirable consequences across the majority of expected patient values and preferences. Panel members will make **Weak recommendations** if there is limited confidence (or inconsistent assessment or dissenting opinions) that desirable consequences outweigh undesirable consequences across the majority of expected patient values and preferences. No recommendation will be made if there is insufficient confidence to make a recommendation.
- All steps in this process (including evidence summaries which were shared with the panel, and identification of panel members) will be transparent and accessible in support of the recommendation.
  - Recommendations are verified by ≥ 1 editor with methodological expertise, not involved in recommendation drafting or development, with explicit confirmation that Strong recommendations are adequately supported.
  - Recommendations are published only after consensus is established with agreement in phrasing and strength of recommendation by all editors.
  - If consensus cannot be reached then the recommendation can be published with a notation of "dissenting commentary" and the dissenting commentary is included in the topic details.
  - If recommendations are questioned during peer review or post publication by a qualified individual, or reevaluation is warranted based on new information detected through systematic literature surveillance, the recommendation is subject to additional internal review.

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**Special acknowledgements**

- Daniel A. Ostrovsky, MD (Assistant Professor of Medicine and Pediatrics, Duke University School of Medicine; North Carolina, United States) provides peer review.
- Esther Jolanda van Zuuren, MD (Head of Allergy, Dermatology, and Venereology, Leiden University Medical Centre; Netherlands; Editor, Cochrane Skin Group) provides peer review.
- Alan Ehrlich, MD (Executive Deputy Editor; Assistant Professor of Family Medicine, University of Massachusetts Medical School; Massachusetts, United States).
How to cite

- National Library of Medicine, or "Vancouver style" (International Committee of Medical Journal Editors):