CLINICAL GUIDE
PAEDIATRIC HEAD DEFORMITY

Brachycephaly
Asymmetrical Brachycephaly
Plagiocephaly
Scaphocephaly
The push to put babies to sleep on their back to reduce the risk of sudden infant death syndrome (SIDS) has been associated with a decrease in the incidence of SIDS but has led to an increase in the number of babies suffering from head shape abnormalities. However, positional skull deformities are generally benign, reversible head-shape anomalies that do not require surgical intervention, as opposed to craniosynostosis, which can result in neurological damage and progressive craniofacial distortion. Although associated with some risk of positional skull deformity, healthy young infants should be placed down for sleep on their backs.

Parents are naturally concerned if they observe asymmetry or unusual flat spots on their baby’s head and abnormalities in the face, and often seek advice from their paediatrician. These concerns are valid and assessments need to include not only the back, but forehead flattening, ear shift, and orbital or facial involvement.

This guide is designed to provide information about the causes, signs and treatment strategies for managing head shape deformities in infants. This includes educating parents on methods of proactively decreasing the likelihood of the development of occipital flattening, initiate appropriate management and make referrals when necessary.

Treatment interventions include repositioning, a developmental home programme, paediatric physiotherapy for patients with torticollis and the use of a cranial remolding orthoses, such as the STARband®, to improve symmetry and normal proportion.

Why are the skulls of infants subject to deformation?

- The plasticity of the newborn’s skull makes it susceptible to external pressures in the womb, during the birth process and after birth
- The immobility of newborns and any positional neck preference can predispose infants to extrinsic skull deformities
- Intrinsic abnormalities can be caused by craniosynostosis or through genetic transmission
- Deformational forces most frequently affect the occiput, although the frontal bones and the face may be affected in severe cases
- About 24%* of babies have some type of noticeable skull deformity at birth, reducing to about 20%† by four months of age
- The abnormal shape may persist or occur if the baby spends most of the day on the back against the hard surface of infant carriers and holding devices

†Hutchison BL, Hutchison AD. Plagiocephaly and Brachycephaly in the First Two Years of Life: A Prospective Cohort Study. Pediatrics; 114;970, 2004
What are contributing risk factors for head deformation?

- Prolonged exposure to the supine position
- Lack of time on the tummy when the baby is awake
- Congenital muscular torticollis, neck weakness or restricted neck range of motion
- Males more frequently develop deformational plagiocephaly at a rate of 2:1
- Slower motor development particularly in gross motor skills
- Breech or transverse presentation
- Multiple birth infants
- Visual field deficits
- Bony abnormality in the cervical spine

What are the types of deformational head shapes?

DEFORMATIONAL PLAGIOCEPHALY

- The most common type of skull deformity in infants
- Normally noticed by caregivers at about six to 10 weeks of age
- Characterised by an asymmetrical skull shape
- Unilateral occipital flattening
- Ear is positioned more anterior on the side of the occipital flattening
- Forehead may be asymmetrical and is positioned more anterior on the side of the occipital flattening
- Facial asymmetry may be present
- May be accompanied by torticollis, limited neck range of motion, weakness and preferential head positioning
DEFORMATIONAL BRACHYCEPHALY
• Central occipital flattening
• High and sloped skull. The head is excessively wide for its length
• May be accompanied by a prominent, bossed forehead

DEFORMATIONAL SCAPHOCEPHALY
• Very elongated head shape that is excessively long for its width
• Deformational scaphocephaly caused by extrinsic forces is uncommon although it is sometimes seen in premature infants who are often positioned side lying, such as NICU infants
• Scaphocephaly caused by extrinsic positioning may be confused with sagittal synostosis, a more serious deformity that usually requires surgery to correct

DEFORMATIONAL BRACHYCEPHALY WITH ASYMMETRY
• Combination of brachycephalic and plagiocephalic characteristics
• The shape is disproportionately wide for its length and is also asymmetrical. May or may not include asymmetries to the forehead and facial structure
**What is craniosynostosis?**
Craniosynostosis is the premature closure of one or more cranial sutures. Craniosynostosis causes restriction of growth at the suture and abnormal growth perpendicular to the fused suture. Early referral to a specialist is always recommended. There are different types of craniosynostosis, including: sagittal, coronal, metopic and lambdoid. Infants diagnosed with craniosynostosis usually require surgery and should be referred immediately to a paediatric neurosurgeon or craniofacial specialist for further evaluation and treatment. Early diagnosis is vital. Endoscopic surgeries are often recommended before three months of age.

**Patient evaluation**
Examination of the infant skull is essential to differentiate deformational skull shape abnormalities from deformities secondary to craniosynostosis.

<table>
<thead>
<tr>
<th>Clinical indicators</th>
<th>Normal</th>
<th>Deformational skull shape</th>
<th>Craniosynostosis suspected</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fontanelles and sutures</td>
<td>Normal size for age of child, no depression or bulging, no ridging over sutures.</td>
<td>Abnormal size or other atypical appearance of fontanelle. Suture(s) may be ridged.</td>
<td></td>
</tr>
<tr>
<td>Proportion</td>
<td>Average width of the skull is 76-83% of the length of the skull.</td>
<td>Brachycephaly or brachycephaly with asymmetry: Width is &gt;91% of length.</td>
<td>Coronal synostosis: Width may be &gt;91% of length.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Scaphocephaly: Width is &lt;76% of the length.</td>
<td>Sagittal synostosis: Skull is noticeably elongated, and may be &lt;76% of length.</td>
</tr>
<tr>
<td>Occiput and parietal Bones</td>
<td>Occiput and parietal bones are symmetrical.</td>
<td>Plagiocephaly or brachycephaly with Asymmetry: Unilateral flattening that may cross midline. Occipital bossing on side opposite flattening.</td>
<td>Unilateral Lambdaid Synostosis: Impressive unilateral occipital flattening, contra-lateral parietal bossing.</td>
</tr>
<tr>
<td>Mastoid</td>
<td>Not prominent.</td>
<td>Plagiocephaly or brachycephaly with asymmetry: Mastoid is not prominent.</td>
<td>Unilateral lambdoid synostosis: Significant mastoid (not occipital) prominence on side of occipital flattening.</td>
</tr>
</tbody>
</table>

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<tbody>
<tr>
<td></td>
<td></td>
<td>Brachycephaly: Both sides of forehead abnormally bulged and prominent.</td>
<td>Bicoronal synostosis: Abnormally flat and tall forehead, turicephalic.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Metopic synostosis: Metopic suture is prominent and forehead is angulated centrally.</td>
</tr>
<tr>
<td>Eyes/Orbits</td>
<td>Eyes symmetrical.</td>
<td>Asymmetry: Eye size may be asymmetrical.</td>
<td>Flattened frontal bone is positioned higher and more open than opposite side (harlequin sign).</td>
</tr>
<tr>
<td>Ear position</td>
<td>Ears are symmetrical in height and placement.</td>
<td>Plagiocephaly or brachycephaly with asymmetry: Ear on side of occipital flattening is displaced more anterior than contralateral ear.</td>
<td>Unilambdoid synostosis: Ear on side of occipital flattening is displaced vertically and in a posterior direction toward the closed suture.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Unicoronal synostosis: Ear is displaced vertically toward the fused coronal suture.</td>
</tr>
<tr>
<td>Face</td>
<td>Forehead, eyes, cheeks, mandible and chin are symmetrical.</td>
<td>Plagiocephaly or brachycephaly with asymmetry: Forehead, eyes, cheeks, mandible and chin may be asymmetrical.</td>
<td>Unicoronal synostosis: Eyes asymmetrical, nasal root displaced toward side of fused coronal suture, chin displaced away from closed coronal suture.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Brachycephaly and scaphocephaly: Facial features are symmetrical.</td>
<td></td>
</tr>
</tbody>
</table>
Evaluating neck strength and range of motion for torticollis
• Evaluate active neck strength and range of motion to determine whether torticollis or a positional neck preference is contributing to the abnormal skull shape

• Stimulate the infant to visually track an object to each side. Babies with torticollis may compensate for limited range by turning the shoulders at the endpoint of their range or tipping their head back while simultaneously jutting their chin forward

• Assess head tilt, skin fold symmetry and occipital hair growth patterns, which may indicate prolonged head/neck positioning

• Hold the infant at arm’s length and gently tip to each side. If the baby shows asymmetric head righting ability, neck weakness or limitation may be present

• Refer the infant to a paediatric physical therapist if torticollis or neck asymmetry is present, particularly if a home developmental programme fails to resolve the problem after three months of age

Can deformational plagiocephaly be prevented?
While not all deformational plagiocephaly cases will correct on their own, there are some efforts that can help prevent or reduce its effects:

• Timely diagnosis and protocol referenced treatment including acquiring accurate anthropometric data
• Comprehensive and timely parent education specially in high risk cases
• Prescribe paediatric physiotherapy or occupational therapy at three months or earlier if there is:
  • Strong positional head/neck preference
  • Torticollis
  • Developmental delay
  • Cranial deformity

• Frequent changes in body and head positions (repositioning strategies) incorporated into a tummy time home programme
• Prone positioning when the infant is awake and supervised
• Limit time in carriers and positioning devices
• Use of STAR cranial remolding orthosis when indicated
### Assessment of severity and suggested action

<table>
<thead>
<tr>
<th>Severity</th>
<th>Deformational Plagiocephaly</th>
<th>Brachycephaly</th>
<th>Brachycephaly with Asymmetry</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal–mild</td>
<td>Skull is symmetrical and well proportioned. Some mild asymmetry may be present but it is within normal limits (WNL)</td>
<td>Skull is well proportioned. It may be wider than normal but is WNL.</td>
<td>Skull may be slightly asymmetrical and wide but is WNL.</td>
</tr>
<tr>
<td>Action:</td>
<td>Document and monitor and consider physiotherapy if deformity present</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moderate</td>
<td>The skull has unilateral occipital flattening. The ear on the ipsilateral side may be displaced anteriorly.</td>
<td>The skull has central occipital flattening with compensatory growth laterally.</td>
<td>Skull is abnormally wide. Occipital asymmetry extends past midline. Ipsilateral ear may be displaced forward.</td>
</tr>
<tr>
<td>Action:</td>
<td>Refer for paediatric physiotherapy or occupational therapy and consider STARband if baby is &gt;3 months and &lt;18 months.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moderate–severe</td>
<td>Significant occipital flattening, ear on the ipsilateral side displaced anteriorly, ipsilateral frontal bone is bossed, contralateral frontal bone may appear to be more flat.</td>
<td>Central occipital flattening with compensatory growth laterally creating a very wide head with bilateral frontal bossing.</td>
<td>Occipital flattening extends beyond midline with contralateral occipital bossing. Skull is very wide for its length with significant ear shift, ipsilateral frontal bossing and contralateral frontal flattening.</td>
</tr>
<tr>
<td>Action:</td>
<td>Refer for STARband if baby is &gt;3 months and &lt;18 months.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td>Skull shape is very abnormal with unilateral occipital flattening and contralateral occipital bossing. The ipsilateral ear is displaced forward, with significant frontal asymmetry including forehead, orbits, cheeks and mandible.</td>
<td>Occiput is extremely flat, parietal bones are displaced laterally creating a very wide head, high and sloped cranial vault, and significant frontal bossing.</td>
<td>Skull is both asymmetrical and disproportionate with significant frontal changes including ipsilateral frontal bossing and asymmetry of the orbits, cheeks and mandible.</td>
</tr>
<tr>
<td>Action:</td>
<td>Refer for STARband if baby is &gt;3 months and &lt;18 months.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Treatment of plagioccephaly with a STARband® cranial remolding orthosis

STAR cranial remolding orthoses derive their name from their purpose—symmetry through active remodeling
- Functions to accommodate growth, promote symmetry and improve proportion
- Refer for a STARband if the skull deformity is moderate to severe. Mild deformities should be documented and monitored
- Ideal cranial orthosis treatment results are achieved when infants begin treatment before six months of age when the skull is rapidly growing. (Please allow up to two weeks from the time of referral to the start of treatment so proper documentation can be submitted to the insurance company for coverage)
- The STARband is best prescribed during the first year of life, but can be used on infants up to 18 months of age
- Multiple studies have demonstrated that cranial remolding orthoses are more effective than repositioning in correcting skull deformities
- Many infants benefit from concurrent physical therapy and orthotic intervention, particularly if the infant has significant neck/torticollis issues
- The STARband gently molds the infant’s skull into a more symmetrical and well-proportioned shape

**Before STARband® treatment**

**STARscanner™ scan data**

**After STARband® treatment**
The STARscanner is a non-contact laser data acquisition system used by leading physicians, hospitals and treatment centres throughout the world, including Mediclinic.

- Accurate—scans to an accuracy of +/- 0.5 m
- Faster head shape acquisition
- Provides precise anthropometric data and measurements
- Infant can be scanned to follow progression of deformity prior to treatment
- Provides pre-treatment documentation for medical justification and insurance coverage
- Ongoing measurements for evaluation of progress throughout treatment
- Comparisons can be utilised to determine modifications
- Software can compare head shape changes over time
- Assessment tool to determine need for treatment

**Contraindications of cranial remolding orthoses**

Infants with head shape asymmetry are not candidates for cranial remolding orthosis treatment if:
- The infant is younger than three months
- The infant is older than 18 months
- The infant has untreated craniosynostosis
- The infant has hydrocephalus

**Key benefits:**
- Used in the leading prestigious paediatric care institutions in the world
- Scans infant’s head shape in five seconds or less
- Captures 3-D data that can be viewed in multiple planes
- Allows comparison to previous scanning results enabling outcome review
- Facilitates improved insurance and third party communication
- Facilitates improved documentation and care pathway introduction
- Detailed scan reports are filed in the patient file
Cranial deformity assessment protocols

Internationally, protocols distinguish when and what treatment is indicated, these include:

- Cephalic Index (CI)
- Children’s Health Care of Atlanta Clinical Classification Scale (CHOA)
- The Cranial Vault Asymmetry Index (CVAI)
- Cranial Vault Asymmetry (CVA)

Assessment includes all angles of the head, especially the vertex bird’s eye view. The most significant measurements used in this initial evaluation are skull base asymmetry, cranial vault asymmetry, orbitotragial depth, and cephalic index.

<table>
<thead>
<tr>
<th>Anthropometric Data</th>
<th>Measurement</th>
<th>Measures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cranial base</td>
<td>from right and left subnasal point (sn) to tragus (t)</td>
<td>measures maxillary depth or right and left morphological face height</td>
</tr>
<tr>
<td>Cranial vault</td>
<td>from frontozygomaticus point (fz) on one side of face to euryon (eu)</td>
<td>measures cranial vault asymmetry</td>
</tr>
<tr>
<td>Orbitotragial depth</td>
<td>from exocanthion point (ex) to tragus (t)</td>
<td>measures orbito-tragion depth (exocanthion)</td>
</tr>
<tr>
<td>Head width</td>
<td>from euryon (eu) on one side of head to euryon (eu) on the other side</td>
<td>measures greatest transverse diameter or maximal head width</td>
</tr>
<tr>
<td>Head length (g-op)</td>
<td>from glabella point (g) to opisthocranion (op)</td>
<td>measures maximal head depth or length</td>
</tr>
<tr>
<td>Head circumference</td>
<td>Head circumference, measured by tape: encircle the tape around the head covering glabella [g] and opisthocranion [op], do not stretch tape too tightly</td>
<td></td>
</tr>
</tbody>
</table>

For more information on the Starscanner or orthotic management of cranial deformity, or to refer a patient, please contact Mediclinic Orthotics Services, Mediclinic Beach Road at +971 4 379 7711 or email mcme.oandp@mediclinic.ae or visit www.starbandkids.com.

A full list of references are available from the contacts above.
Cephalic Index (CI) or cephalic ratio

- Ratio that measures the proportion of the head
- Width divided by length of the head x 100
- Babies with brachycephaly and scaphocephaly should be assessed using this scale
- Babies with a cephalic index greater than two standard deviations above or below the mean should be referred for a cranial orthotic
- As the cephalic ratio approaches 100%, the head shape becomes more abnormal
- Ears may be symmetrical

\[ CI = \left( \frac{\text{Width}}{\text{Length}} \right) \times 100 \]

<table>
<thead>
<tr>
<th>Level</th>
<th>Clinical presentation</th>
<th>Recommendation</th>
<th>CVAI</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Normal</td>
<td>All symmetry within normal limits</td>
<td>No treatment</td>
</tr>
<tr>
<td>2</td>
<td>Mild</td>
<td>• Minimal asymmetry in one posterior quadrant • No secondary changes</td>
<td>Repositioning program • If 2 month of repositioning has failed, cranial remolding orthosis at parent’s request for cosmetic purposes</td>
</tr>
<tr>
<td>3</td>
<td>Moderate</td>
<td>• Two quadrant involvement • Moderate to severe posterior quadrant flattening • Minimal ear shift and/or anterior involvement</td>
<td>Repositioning program or Cranial remodeling orthosis-based on age and history</td>
</tr>
<tr>
<td>4</td>
<td>Severe</td>
<td>• Two or three quadrant involvement • Severe posterior quadrant flattening • Moderate ear shift • Anterior involvement including noticeable orbit asymmetry</td>
<td>Cranial remolding orthosis</td>
</tr>
<tr>
<td>5</td>
<td>Very severe</td>
<td>• Three or four quadrant involvement • Severe posterior quadrant flattening • Severe ear shift • Anterior involvement including orbit and cheek asymmetry</td>
<td>Cranial remolding orthosis</td>
</tr>
</tbody>
</table>

Children’s Healthcare of Atlanta (CHOA), Cranial Vault Asymmetry Index (CVAI), and Cranial Vault Asymmetry (CVA) severity scales

\[ CVAI = \frac{(A - B) \times 100}{A \text{ or } B} \]

(Whichever is greater)

\[ CVA = A - B \]

Normal ≤ 3mm
Mild/Moderate < 12mm
Moderate/Severe ≥ 12mm
Measuring the cranial vault
Take a measurement from the FZ (frontozygomatic point) on one side of the face to the EU (eurion) on the opposite side of the head. The frontozygomaticus is the most lateral point on the frontozygomaticus suture and is located at the upper and outer corner of the orbit. The eurion is the most lateral point on the head. It is located in the parietal region and is found by taking spreading calipers and finding the widest medial-lateral points on the head. The difference between these two measurements is the cranial vault asymmetry.

Calculating the Cephalic Index
The cephalic index measures the overall proportion of the vault of the skull. It is the ratio of the maximum width of the skull to the maximum length. The measurement is acquired by measuring EU (eurion) to EU with calipers which provides the maximum width, measuring the G (glabella) to OP (opisthocranion) with calipers which provides the maximum length, and performing a mathematical calculation. Cephalic Index = Maximum width (eu-eu) x 100 divided by the maximum length (g-op)

Misconceptions
Paediatric head deformity will resolve itself
It is a misconception that the infant’s head “will round out on its own as the child becomes more active, begins to roll over, and learns to sit up.” This is based in part on outdated scales of motor development and a lack of understanding on the effect of supine sleep positioning. The pattern of early motor development is affected by sleep position. On average, supine sleepers attain common motor milestones later than prone sleepers. Prior to 1992, infants’ heads often corrected in the first few months of life because infants that were placed prone to sleep were generally in a variety of positions during the day, thus avoiding prolonged time in one position. Now that supine is the position of choice and there is a four to six week delay in the acquisition of head and trunk control, infants’ heads often do not “round out” as they did previously. The role of thorough parental education, repositioning, and paediatric physical therapy when ROM issues are present needs emphasis.

It is only a cosmetic issue
Classifying cranial deformity as a cosmetic issue is oversimplified and not evidence based. When left untreated, moderate to severe deformity may lead to significant cosmetic and functional-neurological and psychological consequences. For example, the pressure exerted on the intraorbital muscles and nerves, among others, can result in sensory and motor disturbances. As a consequence, infants with head deformities attempt to compensate for the head’s abnormal orientation in space which can result in ocular and vestibular impairment.

Skull deformities are well known for inducing an inferiority complex in childhood. As adults, social issues as a result of the visible deformity are compounded by difficulties in wearing glasses and auricular asymmetries, hair style problems, temporo-mandibular joint asymmetries and teeth alignment problems.

The window of opportunity and correction is up to 12 or even 16 months of age
Though correction can be achieved up to 16 months of age, it’s important to understand the skull undergoes 85% of its postnatal growth within the
first year of life. Early recognition and treatment within this small window of opportunity is paramount. Paediatric physical therapy can greatly aid motor development, and assist in reducing the effects of torticollis presentations. In the “severe” deformation group, the earlier the cranial orthotic treatment started, the higher symmetry ratio recovery obtained. Treatment is especially effective when started in four-month old infants. The “mild” deformation group showed that cranial orthotic remoulding was most effective if treatment started before six months of age.

**Cranial orthotic management is more successful in Western and European populations as the orthotics are designed for this patient geographic**

Despite the structural and physiological differences from infants of other ethnicity, cranial orthotic remoulding is effective in all infants, provided that intervention, timing and recognition conditions are met.

**The deformity is only on the back of the head, and probably will be covered with hair**

The deformation of one element leads to compensatory deformation and displacement of all other connected elements. This can include facial deformation, mandibular asymmetry, congenital and/or acquired muscular torticollis, abnormal eye placement, external ear deformity and misalignment, orbital asymmetry resulting in strabismus and other ocular problems, and epicanthal fold on the side of flatness.

In addition abnormal cranial height, abnormal cranial width/breadth, and occipital flattening with ipsilateral forehead bossing may be present. This compensation for the head’s abnormal orientation in space results in ocular and vestibular impairment and distortion of the orbits with pressure on the extraocular muscles and nerves, resulting in sensorimotor disturbances.

**Cranial remoulding orthoses are uncomfortable to the baby and might be unsafe**

The vast majority of infants have very few problems tolerating the orthotic. As can be expected, specialised care and proper fitting insures comfort and compliance. Clarren et al. and numerous other studies documented the safety and efficacy of cranial remoulding orthosis for positional plagiocephaly and other positional cranial deformities.

**Cranial orthotics management is not approved by insurance companies**

In the event protocols are adhered too, early repositioning attempted and chronological referenced anthropometric measurements indicate, most insurance companies consider cranial remodeling orthotics as medically necessary for treatment of moderate to severe positional head deformities.

For additional information or to request an appointment please contact Mediclinic Orthotic Services, call 04 435 9999 or email mcme.oandp@mediclinic.ae

Unresolved asymmetric cranial deformity.