Introduction

A newborn baby’s skull is made up of several bones that are not yet solidly connected to each other. This allows the infant’s skull to grow and get bigger as the baby’s brain grows. Sometimes, the baby’s skull may have become flattened or misshaped during the birthing process or for other reasons. This abnormal skull shape is called plagiocephaly. Adjustable helmets (a cranial orthotic) may be used to reshape flattened areas of a baby’s skull. However, there is no medical evidence that a child’s development is affected by a head that is not exactly the same shape on both sides. Using a helmet in this situation is cosmetic.

The skull bones may also fuse together too soon. This is dangerous, as it will not allow the brain to grow inside this solid skull. This can cause brain damage, developmental delay, and problems with thinking. Fusion of the skull bones is called synostosis. Surgery is needed to open up the space between the skull bones to allow the brain to grow normally. Helmets may be used after skull surgery to help protect the brain and reshape the bones.

This policy describes when an adjustable helmet may be medically necessary.

Note: The Introduction section is for your general knowledge and is not to be taken as policy coverage criteria. The rest of the policy uses specific words and concepts familiar to medical professionals. It is intended for providers. A provider can be a person, such as a doctor, nurse, psychologist, or dentist. A provider also can be a place where medical care is given, like a hospital, clinic, or lab. This policy informs them about when a service may be covered.
## Policy Coverage Criteria

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Medical Necessity</th>
</tr>
</thead>
</table>
| **Adjustable cranial orthosis** | Use of an adjustable cranial orthosis (cranial banding or soft shell helmet) may be considered medically necessary when the following criteria are met:  
   - Age is between 3 and 18 months of age  
   - The device is custom made and fitted for the individual  
   - Either of the following is present:  
     - The child has had surgery for crainiosynostosis, and the orthosis is needed for post-operative care  
     - The child has severe positional plagiocephaly* that has not responded to a two month trial of repositioning and/or physical therapy  

**Severe positional plagiocephaly is defined by the following (see Table 1 and 2 below):**  
   - Craniofacial asymmetry of 10 mm or more in one of the following measurements: cranial vault, skull base, or orbitotragial depth  
   - A cephalic index is at least two standard deviations above or below the mean for the appropriate gender and age.  

Note: A protective helmet (HCPCS code A800-A8004) is not a cranial orthosis/cranial remolding device. It is considered a safety device worn to prevent injury to the head. It is not addressed in this policy.
Evaluation of Plagiocephaly

The diagnosis of the type of craniosynostosis is confirmed through physical examination and imaging studies.

Anthropometric data, or the measurements used to evaluate abnormal head shape by measuring the distance in mm from one pre-designated point on the face or skull to another must document moderate to severe plagiocephaly.

The evaluation of cranial asymmetry may be based on one or more of four anthropometric measures: cranial vault, skull base, orbitotragial depth measurements or the cephalic index.

Table 1. Anthropometric Measures

<table>
<thead>
<tr>
<th>Anthropometric Measure</th>
<th>Measurement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cranial Vault</td>
<td>[left frontozygomatic point (fz) to right euryon (eu)] minus [right frontozygomatic point (fz) to left euryon (eu)]</td>
</tr>
<tr>
<td>Skull Base</td>
<td>[subnasal point (sn) to left tragus (t)] minus [subnasal point (sn) to right tragus (t)]</td>
</tr>
<tr>
<td>Orbitotragial Depth</td>
<td>[left exocanthion point (ex) to left tragus (t)] minus [right exocanthion point (ex) to right tragus (t)]</td>
</tr>
</tbody>
</table>

Evaluation of cranial asymmetry may also be based on the cephalic index, a ratio between the width and length of the head. Typically, head width is calculated by subtracting the distance from euryon (eu) on one side of the head to euryon on the other side of head and multiplying by 100. Head length is generally calculated by measuring the distance from glabella point (g) to opisthocranion point (op). The cephalic index is then calculated as:

- Head width (eu – eu) x 100
- Head length (g – op)

The cephalic index is considered abnormal if it is two standard deviations above or below the mean measurements (Farkas and Munro, 1987).
Table 2. Cephalic Index

<table>
<thead>
<tr>
<th>Gender</th>
<th>Age</th>
<th>- 2 SD</th>
<th>- 1SD</th>
<th>Mean</th>
<th>+ 1SD</th>
<th>+ 2SD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>16 days – 6 months</td>
<td>63.7</td>
<td>68.7</td>
<td>73.7</td>
<td>78.7</td>
<td>83.7</td>
</tr>
<tr>
<td></td>
<td>6 – 12 Months</td>
<td>64.8</td>
<td>68.7</td>
<td>78.0</td>
<td>84.6</td>
<td>91.2</td>
</tr>
<tr>
<td></td>
<td>13 – 18 Months</td>
<td>Apply the 12 month measurements for children 13-18 months of age</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Female</td>
<td>16 days – 6 months</td>
<td>63.9</td>
<td>68.6</td>
<td>73.3</td>
<td>78.0</td>
<td>82.7</td>
</tr>
<tr>
<td></td>
<td>6 – 12 Months</td>
<td>69.5</td>
<td>74.0</td>
<td>78.5</td>
<td>83.0</td>
<td>87.5</td>
</tr>
<tr>
<td></td>
<td>13 – 18 Months</td>
<td>Apply the 12 month measurements for children 13-18 months of age</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Documentation Requirements

The patient’s medical records submitted for review should document that medical necessity criteria are met. The records should include the following:

- Child age is between 3 and 18 months old
- The requested orthosis is custom made and fitted for the child
- **AND** one of the following must be present:
  - Child had surgery for craniosynostosis (the bones in the child’s skull join together too early), and the cranial orthosis is needed for post-operative care
  - **OR**
    - The child has severe positional plagiocephaly (the child’s head is flat in the back or on one side) verified by the following measurements:
      - 10 mm or more of asymmetry in one of the following measure: cranial vault, skull base, or orbitotragal depth
      - or Cephalic index at least two standard deviations above or below the mean (for the appropriate gender and age)
Documentation Requirements

- And that the severe positional plagiocephaly has not responded to a 2 month trial of repositioning and/or physical therapy

Coding

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>S1040</td>
<td>Cranial remodeling orthosis, pediatric, rigid, with soft interface material, custom fabricated, includes fitting and adjustment(s)</td>
</tr>
</tbody>
</table>

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Related Information

Assessment of plagiocephaly in research studies may be based on anthropomorphic measures of the head, using anatomic and bony landmarks. However, there is no accepted minimum objective level of asymmetry for a plagiocephaly diagnosis. Table 3 presents normative values and the mean pretreatment asymmetries reported in large case series. These may be useful in determining if a significant variation from normal is present.

Table 3. Pretreatment Asymmetries Reported in Large Case Series

<table>
<thead>
<tr>
<th>Study</th>
<th>Cranial Base, mm</th>
<th>Cranial Vault, mm</th>
<th>Orbitotragial Distance, mm</th>
</tr>
</thead>
<tbody>
<tr>
<td>Moss (1997)¹</td>
<td>NR</td>
<td>9.2</td>
<td>7.1³</td>
</tr>
<tr>
<td>Littlefield et al. (1998)²</td>
<td>6.17</td>
<td>8.50</td>
<td>4.36</td>
</tr>
<tr>
<td>Teichgraeber et al. (2002)³</td>
<td>7.08</td>
<td>8.53</td>
<td>3.12</td>
</tr>
</tbody>
</table>
NR: not reported.
* In this report, the asymmetry was measured from the tragus to the frontozygomatic point instead of the exocanthion.

**Consideration of Age**

The ages referenced in this policy for which cranial orthoses are considered medically necessary are between 3 and 8 months. This is based on the FDA-approved age range for these helmets and the American Academy of Pediatrics (AAP) states, "The use of helmets and other related devices seems to be beneficial primarily when there has been a lack of response to mechanical adjustments and exercises, and the best response to helmets occurs in the age range of 4 to 12 months of age."

**Definition of Terms**

**Anthropomorphic Assessment of Plagiocephaly**

**Brachiocephaly:** Shortened front to back dimension of the skull that results from premature fusion of the coronal suture

**Coronal suture:** Skull joint that goes across the top of the skull and separates the front and back halves of the skull

**Cranial base:** Asymmetry of the cranial base is measured from the subnasal point (midline under the nose) to the tragus (the cartilaginous projection in front of the external auditory canal

**Cranial index:** The cranial index, which describes a ratio of the maximum width to the head length expressed as a percentage, is used to assess abnormal head shapes without asymmetry. The maximum width is measured between the most lateral points of the head located in the parietal region (i.e., euryon). The head length is measured from the most prominent point in the median sagittal plane between the supraorbital ridges (i.e., glabella) to the most prominent posterior point of the occiput (i.e., the opisthocranion), expressed as a percentage. The cranial index can then be compared to normative measures.

**Cranial vault:** Asymmetry is assessed by measuring from the frontozygomaticus point (identified by palpation of the suture line above the upper outer corner of the orbit) to the euryon, defined as the most lateral point on the head located in the parietal region.
**Craniosynostosis:** Fusion of at least two of the skull bones before the brain has fully formed.

**Metopic suture:** Skull joint that separates the left and right halves of the forehead.

**Orbitotragial depth:** Asymmetry of the orbitotragial depth is measured from the exocanthion (outer corner of the eye fissure where the eyelids meet) to the tragus (the cartilaginous projection in front of the external auditory canal)

**Plagiocephaly:** Flattening of the skull on the back or one side of the head.

**Sagittal suture:** Skull joint that separates the left and right halves of the skull.

**Synostosis:** Fusion of two bones

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### Evidence Review

#### Description

Cranial orthoses involve an adjustable helmet or band that progressively molds the shape of the infant cranium by applying corrective forces to prominences while leaving room for growth in the adjacent flattened areas. A cranial orthotic device may be used to treat postsurgical synostosis or positional plagiocephaly in pediatric patients.

#### Background

**Craniosynostoses**

An asymmetrically shaped head may be synostotic or nonsynostotic. Synostosis, defined as premature closure of the sutures of the cranium, may result in functional deficits secondary to increasing intracranial pressure in an abnormally or asymmetrically shaped cranium. The type and degree of craniofacial deformity depends on the type of synostosis. The most common is scaphocephaly, a narrowed and elongated head resulting from synostosis of the sagittal suture that separates the left and right halves of the skull. Trigonocephaly, in contrast, is premature fusion of the metopic suture that separates the left and right sides of the forehead and results in a pointed, triangular shape of the forehead. Unilateral synostosis of the coronal suture that
separates the front and back halves of the skull results in an asymmetric distortion of the forehead called plagiocephaly, and fusion of both coronal sutures results in brachycephaly. Combinations of these deformities may also occur.

**Treatment**

Synostotic deformities associated with functional deficits are addressed by surgical remodeling of the cranial vault. The remodeling (reshaping) is accomplished by opening and expanding the abnormally fused bone.

In a 2008 review of the treatment of craniosynostosis, Persing indicated that premature fusion of one or more cranial vault sutures occurs in approximately 1 in 2500 births.\(^1\) Of these craniosynostoses, asymmetric deformities involving the cranial vault and base (eg, unilateral coronal synostosis) will have a higher rate of postoperative deformity, which would require additional surgical treatment. Persing suggested that use of cranial orthoses postoperatively may serve 2 functions: (1) they protect the brain in areas of large bony defects, and (2) they may remodel the asymmetries in skull shape, particularly when the bone segments are more mobile.

**Plagiocephaly**

Plagiocephaly without synostosis, also called positional or deformational plagiocephaly, can be secondary to various environmental factors including, but not limited to, premature birth, restrictive intrauterine environment, birth trauma, torticollis, cervical anomalies, and sleeping position. Positional plagiocephaly typically consists of right or left occipital flattening with advancement of the ipsilateral ear and ipsilateral frontal bone protrusion, resulting in visible facial asymmetry. Occipital flattening may be self-perpetuating in that once it occurs, it may be increasingly difficult for the infant to turn and sleep on the other side. Bottle feeding, a low proportion of “tummy time” while awake, multiple gestations, and slow achievement of motor milestones may contribute to positional plagiocephaly. The incidence of plagiocephaly has increased rapidly in recent years; this is believed to be a result of the “Back to Sleep” campaign recommended by the American Academy of Pediatrics, in which a supine sleeping position is recommended to reduce the risk of sudden infant death syndrome. It has been suggested that increasing awareness of identified risk factors and early implementation of good practices will reduce the development of deformational plagiocephaly.
Treatment

It is estimated that about two-thirds of plagiocephaly cases may auto-correct spontaneously after regular changes in sleeping position or following physical therapy aimed at correcting neck muscle imbalance. A cranial orthotic device is usually requested after a trial of repositioning fails to correct the asymmetry, or if the child is too immobile for repositioning.

Summary of Evidence

For individuals who have open or endoscopic surgery for craniosynostosis who receive a postoperative cranial orthosis, the evidence includes case series. Relevant outcomes are change in disease status, morbid events, functional outcomes, quality of life, and treatment-related morbidity. Overall, the evidence on the efficacy of cranial orthoses following endoscopic-assisted or open cranial vault remodeling surgery for craniosynostosis is limited. However, functional impairments are related to craniosynostosis, and there is a risk of harm from additional surgery when severe deformity has not been corrected. Because cranial orthoses can facilitate remodeling, use of a cranial orthosis is likely to improve outcomes after cranial vault remodeling for synostosis. The evidence is sufficient to determine that the technology results in a meaningful improvement in the net health outcome.

For individuals who have positional plagiocephaly who receive a cranial orthosis, the evidence includes a comparative study and case series. Relevant outcomes are change in disease status, morbid events, functional outcomes, quality of life, and treatment-related morbidity. Overall, evidence on an association between positional plagiocephaly and health outcomes is limited. The largest controlled study found no difference in function between infants with plagiocephaly and age-matched concurrent controls. Taking into consideration the limited number of publications over the past decade and the likelihood of both study and publication bias in uncontrolled studies, the scientific literature does not support an effect of deformational plagiocephaly on functional health outcomes. The evidence is insufficient to determine the effects of the technology on health outcomes. However, when positional plagiocephaly is severe and the child has not responded to a two month trial of repositioning and/or physical therapy, a cranial orthosis may help avoid the need for surgery in some cases.
Ongoing and Unpublished Clinical Trials

A currently ongoing trial that might influence this policy is listed in Table 4.

Table 4. Summary of Key Trials

<table>
<thead>
<tr>
<th>NCT No.</th>
<th>Trial Name</th>
<th>Planned Enrollment</th>
<th>Completion Date</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ongoing</td>
<td>NCT02370901* Cranial Orthotic Device Versus Repositioning Techniques for the Management of Plagiocephaly: the CRANIO Randomized Trial</td>
<td>226</td>
<td>Nov 2020</td>
</tr>
</tbody>
</table>

* Denotes industry-sponsored or cosponsored trial
NCT: national clinical trial.

Clinical Input Received From Physician Specialty Societies and Academic Medical Centers

While the various physician specialty societies and academic medical centers may provide appropriate reviewers who collaborate with and make recommendations during this process, input received does not represent an endorsement or position statement by the physician specialty societies or academic medical centers, unless otherwise noted.

In response to requests, input was received from 3 physician specialty societies (4 reviews) and 2 academic medical centers while this policy was under review in 2008. Input was mixed about whether the use of helmets or adjustable banding for treatment of plagiocephaly or brachycephaly without synostosis should be considered medically necessary or not medically necessary. Clinical input agreed that cranial orthoses may be indicated following cranial vault surgery.
Practice Guidelines and Position Statements

**Congress of Neurological Surgeons et al.**

In 2016, the Congress of Neurological Surgeons, American Association of Neurological Surgeons, and American Academy of Pediatrics published a joint evidence-based guideline on the role of cranial molding orthosis therapy for patients with positional plagiocephaly.\(^{25,26}\) They provided level II recommendations (uncertain clinical certainty) on the use of helmet therapy “for infants with persistent moderate to severe plagiocephaly after a course of conservative treatment (repositioning and/or physical therapy)” and “for infants with moderate to severe plagiocephaly presenting at an advanced age.” The recommendations were based on a randomized controlled trial, 5 prospective comparative studies, and 9 retrospective comparative studies (all rated as class II evidence).

**National Institute of Neurological Disorders and Stroke**

The National Institute of Neurological Disorders and Stroke (2017) has stated that “treatment for craniosynostosis generally consists of surgery to improve the symmetry and appearance of the head and to relieve pressure on the brain and the cranial nerves [although] for some children with less severe problems, cranial molds can reshape the skull to accommodate brain growth and improve the appearance of the head.”\(^{27}\)

**National Health Service Quality Improvement**

In 2007, Scotland’s National Health Service Quality Improvement issued an evidence note on the use of cranial orthosis treatment for infant deformational plagiocephaly.\(^{28}\) No evidence-based conclusions could be reached due to the limited methodologic quality of available trials. The evidence note concluded that randomized controlled trial would be needed to determine the true effectiveness of cranial orthoses.
American Academy of Pediatrics

In 2011, the American Academy of Pediatrics (AAP) revised its 2003 policy on the prevention and management of positional skull deformities in infants. AAP indicated that, in most cases, the diagnosis and successful management of deformational plagiocephaly can be assumed by the pediatrician or primary health care clinician and that mechanical methods, if performed early in life, may prevent further skull deformity and may reverse existing deformity. In most cases, an improvement is seen over a 2- to 3-month period with repositioning and neck exercises, especially if these measures are instituted as soon as the condition is recognized. AAP indicated that use of helmets and related devices seems to be beneficial primarily when there has been a lack of response to mechanical adjustments and exercises, and the best response to helmets occurs in the age range of 4 to 12 months of age.

In a 2011 policy statement, AAP indicated that consideration should be given to early referral of infants with plagiocephaly when it is evident that conservative measures have been ineffective, because orthotic devices may help avoid the need for surgery in some cases. AAP also recommended placing infants on their backs for sleep with supervised “tummy time” for the prevention of plagiocephaly.

Medicare National Coverage

There is no national coverage determination (NCD). In the absence of an NCD, coverage decisions are left to the discretion of local Medicare carriers.

Regulatory Status

Several devices cleared for marketing by the U.S. Food and Drug Administration through the 510(k) process are intended to apply passive pressure to prominent regions of an infant’s cranium to improve cranial symmetry and/or shape in infants from 3 to 18 months of age. FDA product code: MVA.

References


### History

<table>
<thead>
<tr>
<th>Date</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>12/08/15</td>
<td>New Policy. Adopting to support medical necessary indications; excluded in contract language. Policy effective date is May 1, 2016 following provider notification.</td>
</tr>
<tr>
<td>04/20/16</td>
<td>Annual review. Policy updated with literature review. Coverage criteria expanded; assessment information moved from policy guidelines to policy section.</td>
</tr>
<tr>
<td>11/08/16</td>
<td>Minor update. Language added to the Rationale section to indicate that the applicable age range of this policy is based on FDA-approval for these helmets and is supported by the American Academy of Pediatrics (AAP).</td>
</tr>
<tr>
<td>Date</td>
<td>Comments</td>
</tr>
<tr>
<td>------------</td>
<td>-------------------------------------------------------------------------------------------------------------------------------------------</td>
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<tr>
<td>02/01/17</td>
<td>Annual Review, approved January 10, 2017. Policy updated with literature review through September 26, 2016; no references added. Policy statements unchanged.</td>
</tr>
<tr>
<td>03/24/17</td>
<td>Policy moved into new format; no change to policy statements.</td>
</tr>
<tr>
<td>05/01/18</td>
<td>Annual Review, approved April 18, 2018. Policy updated with literature review through January 2018; no references added. Minor edits for clarity. Otherwise, policy statements unchanged</td>
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<tr>
<td>09/01/18</td>
<td>Minor update. Re-added Consideration of Age information; it was inadvertently removed in a previous update.</td>
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</table>

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Email AppealsDepartmentInquiries@Premera.com

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U.S. Department of Health and Human Services
200 Independence Avenue SW, Room 509F, HHH Building
Washington, D.C. 20201, 1-800-386-1019, 800-537-7697 (TDD)

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Oromoo (Cushite):


Lakkoofsa bibiliala 800-722-1471 (TTY: 800-842-5357) ti bibiliala.

Italiano (Italian):