

PLAGIOCEPHALY AND CRANIOSYNOSTOSIS TREATMENT

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INSTRUCTIONS FOR USE

This Medical Policy provides assistance in interpreting UnitedHealthcare benefit plans. When deciding coverage, the enrollee specific document must be referenced. The terms of an enrollee's document (e.g., Certificate of Coverage (COC) or Summary Plan Description (SPD)) may differ greatly. In the event of a conflict, the enrollee's specific benefit document supersedes this Medical Policy. All reviewers must first identify enrollee eligibility, any federal or state regulatory requirements and the plan benefit coverage prior to use of this Medical Policy. Other Policies and Coverage Determination Guidelines may apply. UnitedHealthcare reserves the right, in its sole discretion, to modify its Policies and Guidelines as necessary. This Medical Policy is provided for informational purposes. It does not constitute medical advice.

UnitedHealthcare may also use tools developed by third parties, such as the MCG™ Care Guidelines, to assist us in administering health benefits. The MCG™ Care Guidelines are intended to be used in connection with the independent professional medical judgment of a qualified health care provider and do not constitute the practice of medicine or medical advice.

BENEFIT CONSIDERATIONS

Essential Health Benefits for Individual and Small Group:

For plan years beginning on or after January 1, 2014, the Affordable Care Act of 2010 (ACA) requires fully insured non-grandfathered individual and small group plans (inside and outside of Exchanges) to provide coverage for ten categories of Essential Health Benefits ("EHBs"). Large group plans (both self-funded and fully insured), and small group ASO plans, are not subject to the requirement to offer coverage for EHBs. However, if such plans choose to provide coverage for benefits which are deemed EHBs (such as maternity benefits), the ACA requires all dollar limits on those benefits to be removed on all Grandfathered and Non-Grandfathered plans. The determination of which benefits constitute EHBs is made on a state by state basis. As such, when using this guideline, it is important to refer to the enrollee's specific plan document to determine benefit coverage.

When deciding coverage for this service, the enrollee-specific benefit document language must be referenced. Most Certificates of Coverage (COCs) and some Summary Plan Descriptions

(SPDs) contain an explicit exclusion from coverage for orthotic devices. The 2007 COC excludes coverage for orthotic appliances that straighten or re-shape a body part including cranial banding. Some states require benefit coverage for cranial orthotic devices and/or for treatment of congenital conditions that might otherwise be considered cosmetic. State mandates for coverage and the enrollee-specific benefit document must be used to decide coverage. For the purpose of administration of the orthotic exclusion described above, a cranial helmet is considered an orthotic when used for the non-surgical treatment of plagiocephaly. Cranial helmets used to facilitate a successful post surgical outcome are a covered health service.

Less invasive procedures including endoscopic strip craniectomy and spring-mediated cranioplasty are proven as one form of surgical treatment to repair craniosynostosis. If traditional cranial vault remodeling is available within the network, in network benefits for out of network services for less invasive procedures are not available. If out of network benefits are available, less invasive procedures including endoscopic strip craniectomy and spring-mediated cranioplasty are covered at the out of network level of benefits.

COVERAGE RATIONALE

Cranial orthotic devices are reconstructive and medically necessary for:

- a) **craniosynostosis following surgical correction**
- b) **treatment of craniofacial asymmetry in infants 3-18 months of age with severe nonsynostotic positional plagiocephaly** when all the following criteria are present (1, 2 and 3):
 1. Infant is 18 months of age or younger
 2. Severe asymmetry is present with or without torticollis
 3. There is lack of substantial improvement following conservative therapy of at least 2 months duration with cranial repositioning and stretching therapy.

Severe plagiocephaly is defined as an asymmetry of 10 mm or more in one of the following anthropometric measures: cranial vault, skull base, or orbitotragial depth; OR a cephalic index at least two standard deviations above or below the mean for the appropriate gender/age. Clinical evidence demonstrates improved surgical outcomes with the post-operative use of the orthotic.

Note: Please see [Description of Services](#) section for additional information regarding Anthropometric measurements and Cephalic Index graph.

Cranial orthotic devices are cosmetic and not medically necessary in infants with mild to moderate plagiocephaly.

There are no definitive data demonstrating that there are adverse health effects associated with a mild to moderate degree of cranial asymmetry, and, therefore, it is unclear whether treatment of these individuals provides a future health benefit, or merely a cosmetic effect. In general, severe plagiocephaly occurs in utero and is present at birth. Limited clinical evidence suggests that it may be associated with future ocular and/or oral abnormalities. Acquired plagiocephaly occurs following the placement of the infant in a supine sleeping position to prevent sudden infant death syndrome, and is ordinarily mild to moderate. Positional plagiocephaly has not been linked to future comorbidities.

Surgical treatment to repair craniosynostosis is reconstructive and medically necessary irrespective of the approach used.

Less invasive procedures including endoscopic strip craniectomy and spring-mediated cranioplasty are proven non-preferentially and medically necessary as a form of surgical treatment to repair craniosynostosis.

APPLICABLE CODES

The Current Procedural Terminology (CPT[®]) codes and Healthcare Common Procedure Coding System (HCPCS) codes listed in this policy are for reference purposes only. Listing of a service code in this policy does not imply that the service described by this code is a covered or non-covered health service. Coverage is determined by the enrollee specific benefit document and applicable laws that may require coverage for a specific service. The inclusion of a code does not imply any right to reimbursement or guarantee claims payment. Other policies and coverage determination guidelines may apply. This list of codes may not be all inclusive.

CPT [®] Code	Description
61550	Craniectomy for craniosynostosis; single cranial suture
61552	Craniectomy for craniosynostosis; multiple cranial sutures
61556	Craniotomy for craniosynostosis; frontal or parietal bone flap
61557	Craniotomy for craniosynostosis; bifrontal bone flap
61558	Extensive craniectomy for multiple cranial suture craniosynostosis (e.g., cloverleaf skull); not requiring bone grafts
61559	Extensive craniectomy for multiple cranial suture craniosynostosis (e.g., cloverleaf skull); recontouring with multiple osteotomies and bone autografts (e.g., barrel-stave procedure) (includes obtaining grafts)
21175	Reconstruction, bifrontal, superior-lateral orbital rims and lower forehead, advancement or alteration (eg, plagiocephaly, trigonocephaly, brachycephaly), with or without grafts (includes obtaining autografts)

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HCPCS Code	Description
A8000	Helmet, protective, soft, prefabricated, includes all components and accessories
A8001	Helmet, protective, hard, prefabricated, includes all components and accessories
A8002	Helmet, protective, soft, custom fabricated, includes all components and accessories
A8003	Helmet, protective, hard, custom fabricated, includes all components and accessories
L0112	Cranial cervical orthosis, congenital torticollis type, with or without soft interface material, adjustable range of motion joint, custom fabricated
L0113	Cranial cervical orthotic, torticollis type, with or without joint, with or without soft interface material, prefabricated, includes fitting and adjustment
S1040	Cranial remolding orthosis, pediatric, rigid, with soft interface material, custom fabricated, includes fitting and adjustment(s)
D5924	Cranial prosthesis

DESCRIPTION OF SERVICES

Plagiocephaly (an asymmetrical head shape) is most often the result of an infant spending extended periods of time on their back, typically during sleep. Plagiocephaly can also occur as a feature of other disorders (e.g., craniofacial disorders, torticollis, cervical anomalies) and is categorized as either positional or nonpositional (premature union of cranial sutures).

Positional plagiocephaly, also called deformational plagiocephaly, results from external pressure (molding) that causes the skull to become misshapen. It is most often associated with infants sleeping or lying on their backs. Supine positioning is recommended as a strategy to

reduce the likelihood of sudden infant death syndrome (SIDS), and has contributed to the increased incidence of post-natal plagiocephaly. Plagiocephaly can also occur as a feature of other disorders (e.g., craniofacial disorders, torticollis, and cervical anomalies)

Craniosynostosis is a non-positional cause of abnormal head shape in infants and occurs when one or more of the sutures in the infant's skull fuse prematurely. The premature fusion of one or more sutures puts pressure on the brain, potentially restricting brain growth and exerting pressure on the other skull bones to expand out of proportion, leading to abnormal skull shape.

Non-Synostotic Positional Plagiocephaly

Positional plagiocephaly is treated conservatively and many cases does not require any specific treatment as the condition may resolve spontaneously when the infant begins to roll over and, later, to sit up. When the deformity is moderate or severe and a trial of repositioning the infant has failed, a specialist in craniofacial deformities may prescribe a cranial orthotic device to remodel the misshapen head.

Treatment for Positional Plagiocephaly

According to a systematic review (Xia, et al., 2008) repositioning therapy is preferred over molding therapy in patients who are age four months or less and in whom the severity of asymmetry is considered moderate or less. In patients who are age six months or older, or for whom the asymmetry is more than moderate (regardless of age) molding therapy is preferred. This initial treatment of positional plagiocephaly involves repositioning the infant so that there is no pressure on the flattened region of the skull. This is started as soon as the condition is diagnosed, and is most successful in babies less than 6 months of age. If the infant has torticollis, neck muscle massage and stretching is also performed to reduce the tendency of the infant to place the head in one particular position. If the asymmetry is severe, a cranial orthotic device may be used to apply pressure to prominent regions of the infant's skull. Types of orthotic devices used for this condition include a custom-made molding helmet that allows normal growth of the brain while preventing further distortion of prominent areas of the cranium, and a headband, which is based on a similar concept as the helmet but is open on the top and provides pressure to redirect skull growth.

While no accurate estimates of the incidence of positional plagiocephaly are currently available, the supine sleeping position, currently recommended by the American Academy of Pediatrics (AAP) to reduce the risk of sudden infant death syndrome (SIDS), has been associated with an increased frequency of positional plagiocephaly due to pressure of the back of the head against a firm mattress (Persing et al., 2003).

Plagiocephaly with Synostosis

Craniosynostosis is characterized by the premature closure of one or more of the fibrous joints between the bones of the skull (called the cranial sutures) before brain growth is complete. Closure of a single suture is most common. In contrast to normal skull growth, in which the skull expands uniformly to accommodate the growth of the brain, premature closure of a single suture restricts the growth in that part of the skull and promotes growth in other parts of the skull where sutures remain open. This results in an abnormal shape of the skull, but does not prevent the brain from expanding to a normal volume. However, when more than one suture closes prematurely, the skull cannot expand to accommodate the growing brain, which leads to increased pressure within the skull and impaired development of the brain (NINDS 2008).

Craniosynostosis requires surgical treatment to open the prematurely closed suture(s) in order to allow for normal brain growth., (NINDS, 2007; Sheth, (Updated 2012), Clayman et al., 2007). The major complications associated with uncorrected craniosynostosis include increased intracranial pressure and abnormal brain development.

The most common form of craniosynostosis-causes (scaphocephaly), a condition in which the head is abnormally long and narrow; it is often associated with an absent or small anterior

fontanel. Calvarial vault reconstruction (i.e., cranial vault remodeling) and fronto-orbital advancement are the mainstays of surgical treatment for craniosynostosis. Additionally, recent advancements in surgical technique to treat craniosynostosis include endoscopic-assisted surgery (e.g., strip craniectomy, strip synostectomy).

Surgical treatment of craniosynostosis has evolved from simple excision of the stenosed suture to complex procedures such as cranial vault remodeling and fronto-orbital advancement. The latter is currently considered to be the gold standard (Kane, 2004; Podda et al., 2008). Cranial vault remodeling involves removal of the fused suture, multiple osteotomies and remodeling of the skull using plates and screws as necessary. (Kane, 2004; Wolfe, et al., 2008; Clayman et al., 2007) These operations are usually recommended for infants aged 6 to 8 months, are lengthy (4-8 hours), and associated with significant blood loss (300-1500mL), need for blood transfusions and can require hospitalization for 4 to 7 days (Jimenez et al., 2002; Clayman et al, 2007). Spring-mediated cranioplasty is a minimally invasive alternative to the standard surgical procedure for craniosynostosis. Two dynamic springs made of steel are fashioned in the operating room by the surgeon and placed in the gap left by the removal of the fused suture. Three to four months after the surgery, the child has a second, smaller operation to remove the 2 springs. (Lauritzen et al., 1998; David et al., 2004)

Endoscopic strip craniectomy is another recently developed, less invasive surgery to treat craniosynostosis. The principal goal of this procedure is to remove stenosed sutures, and to allow the skull to expand into a normal shape as the brain grows. After surgery, the infant wears a customized helmet for 11 to 12 months to guide and constrain this expansion and correction process. The procedure is best performed when the infant is < 6 months old (Jimenez et al., 1998; Jimenez et al., 2002; Jimenez et al., 2004, and Jimenez and Barone, 2012).

Cranial orthotic devices have also been used after traditional surgery for craniosynostosis. The devices are used to protect the remodeled skull, prevent recurrence of the deformity, and promote corrective reshaping. In this case, they are used to maintain the remodeling accomplished by surgery, rather than to reshape the skull.

A standard method of measurement for plagiocephaly and craniosynostosis has not been adopted. Methods include the use of clinical observation and precision calipers. Other methods include the use of elastic and low temperature thermoplastic bands wrapped circumferentially around the widest point of the head which may then be digitized by photography, photocopied, or scanned to allow measurement and head shape to be analyzed. (McGarry et al., 2008)

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.

Specifications for Taking Anthropometric Measurements	
Anthropometric Measure	Measurement
Cranial Vault	[left frontozygomatic point (fz) to right euryon (eu)] minus [right frontozygomatic point (fz) to left euryon (eu)]

Specifications for Taking Anthropometric Measurements	
Anthropometric Measure	Measurement
Skull Base	[subnasal point (sn) to left tragus (t)] minus [subnasal point (sn) to right tragus (t)]
Orbitotragial Depth	[left exocanthion point (ex) to left tragus (t)] minus [right exocanthion point (ex) to right tragus (t)]

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:

$$\frac{\text{Head width (eu - eu)} \times 100}{\text{Head length (g - op)}}$$

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

Cephalic Index:

Cephalic Index (AAOP, 2004)

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

Patient Selection Criteria: No definitive selection criteria for the use of cranial orthotic devices in patients with positional plagiocephaly have been established. (Hayes, 2010)

Cranial orthotic devices are contraindicated in infants with hydrocephalus and in infants with plagiocephaly due to synostosis, although these devices may sometimes be used following surgical correction of craniosynostosis to aid in further reshaping the head. (Hayes, 2010)

CLINICAL EVIDENCE

Cranial Orthotic Devices

Cranial orthotic devices are used in infants for the treatment of positional plagiocephaly, deformation of the head that results from external pressure applied to the soft infant skull.

Evidence evaluating cranial orthotic devices for positional plagiocephaly consists of case series and systematic reviews.

A study by Hutchinson et al. (2004) showed that the prevalence of nonsynostotic plagiocephaly in healthy normal children, without helmet therapy, increases to 19.7% at 4 months but decreases spontaneously as infants grow older to 3.3% at 24 months. Hutchinson et al. (2010) later conducted a randomized controlled trial of positioning treatments by of 126 infants with

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deformational plagiocephaly and brachycephaly. No difference was found in head shape outcomes between the use of positional treatment or with the use of a Safe T Sleep positioning wrap, which is a body wrap to keep the infant on their back. By one year of age, 42% of infants had head shapes within normal range without the use of cranial orthotic devices.

Recently, a new three-dimensional (3D) digital photographic system has been used to quantify the efficacy of orthotic helmets. Lipira et al. (2010) used whole head 3D surface scans to compare outcomes of orthotic helmets and active repositioning in 70 infants with deformational plagiocephaly (DP). Helmeted (n=35) and nonhelmeted/actively repositioned (n=35) infants were matched for severity of initial deformity. Change in mean and maximum asymmetry with treatment was the basis for group comparison. The helmeted group had a larger reduction than the repositioned group in both maximum (4.0% vs 2.5%) and mean asymmetry (0.9% vs 0.5%). The greatest difference was localized to the occipital region. The authors concluded that additional studies are needed to establish the clinical significance of these quantitative differences in outcome, define what constitutes pathologic head asymmetry, and determine whether superiority of orthotic treatment lasts as the child matures.

Several studies indicate that repositioning and physical therapy can be an effective treatment for reducing cranial asymmetry in infants with positional plagiocephaly, particularly if the asymmetry is relatively mild and therapy is initiated before 6 months of age. There is also evidence that cranial orthotic devices are effective in correcting plagiocephaly, especially when therapy is instituted before the patient is 12 to 18 months of age, and that correction may occur more rapidly than with repositioning therapy, and may occur in patients who fail to respond to initial therapy with repositioning. (Mulliken et al., 1999; Pattisapu et al., 1989; Argenta et al., 1996; Pollack et al., 1997)

A systematic review by McGarry et al. (2008) suggests that the use of cranial orthoses in the management of deformational plagiocephaly is contraindicated prior to 6 months of age, since the rapid brain growth can produce dramatic spontaneous reduction in plagiocephaly. The authors further indicate that cranial orthoses are not recommended after 18 months since there is little remaining brain growth to affect head shape. (McGarry et al., 2008)

Kluba et al. (2011) enrolled 62 infants with severe positional plagiocephaly in a prospective longitudinal study. Twenty-four started helmet therapy before 6 months of age (group 1) and 38 were older than 6 months (group 2). Duration of therapy was significantly shorter in group 1 (14 weeks) compared with group 2 (18 weeks) with significantly better outcomes. The Cranial Vault Asymmetry Index in group 1 was reduced to a normal mean value while infants in group 2 did not achieve normal values. The relative improvement in asymmetry was significantly better in group 1 (75.3 percent) compared with group 2 (60.6 percent). After 4 to 11 weeks of treatment, group 1 already showed a better absolute reduction and a better relative reduction. The authors concluded that optimal starting age for helmet therapy in infants with severe positional plagiocephaly is months 5 to 6 of life. They also conclude that delaying the onset of treatment significantly deteriorates the outcome.

Two prospective controlled studies addressed cranial orthotic therapy for positional plagiocephaly. Loveday and de Chalain (2001) compared active counterpositioning (n=45) with cranial orthosis (n=29) therapy in a random sample of 74 infants referred for management of positional plagiocephaly and found no significant differences in treatment outcomes (cranial vault asymmetry index) between the groups. However, the time taken to treat positional plagiocephaly was one third shorter in the helmet group (22 weeks) than in the repositioning group (64 weeks). For both treatment groups, the posttreatment cranial vault asymmetry index was lower for nonbrachycephalic patients than for brachycephalic patients. Mulliken et al. (1999) also reported on a controlled study comparing helmet therapy with active repositioning for treatment of positional plagiocephaly. Patients (n=114) were between 2 and 9 months of age when treatment was initiated. After a mean of 4.6 months, both treatment groups had satisfactory improvement in cranial symmetry, as determined by complete anthropometric measurements, although the group

that received helmet therapy had significantly better posterior cranial symmetry than did the group treated with active repositioning.

An uncontrolled study completed by Bruner et al. (2004) confirmed a higher degree of correction with use of a cranial orthotic device.

One study evaluated treatment outcomes in groups of children with brachycephaly and positional plagiocephaly, and concluded that use of a cranial orthotic device was effective for both groups, but that more children in the plagiocephaly group were normalized after treatment (Teichgraber et al., 2004).

A study completed by St. John et al. (2002) suggested a worsening of the auricular and temporomandibular asymmetry following helmet therapy, despite improvements in overall cranial symmetry.

Graham et al. (2005) compared positioning (n=176) with orthotic therapy (n=159) in 298 consecutive infants referred for correction of head asymmetry. Thirty-seven patients were treated with initial repositioning followed by helmet therapy when treatment failed. The authors used reductions in diagonal difference (RDD) as outcome measure. They found that infants treated with orthotics were older (6.6 vs 4.8 months) and required a longer length of treatment (4.2 vs 3.5 months). Infants treated with orthosis had a mean final diagonal difference (DD) closer to the DD in unaffected infants. Orthotic therapy was more effective than repositioning (61% decrease versus 52% decrease in DD), and early orthosis was significantly more effective than later orthosis (65% decrease versus 51% decrease in DD).

The same authors also compared the effect of repositioning versus helmet therapy on cephalic index (CI) in 193 infants referred for brachycephaly. Among the subgroup of 96 infants treated by repositioning from an average age of 4.6 months to 7.7 months, the mean initial CI was 86.3% and the mean final CI was 85.7%. The change in CI was not significant. Among the subgroup of 97 infants treated with helmets from an average age of 6.0 months to 10.3 months, the mean initial CI was 91.5% and the mean final CI was 88.4%. The change in CI for this group was significant. In the helmet group treatment at a younger age resulted in more improvement in the CI. (Graham et al., 2005b)

Bialocerkowski et al (2006) conducted a systematic review of the literature published from 1983 to 2003 to determine the effectiveness of conservative interventions for infants with positional plagiocephaly. The authors selected 16 papers including 12 case series and four comparative studies for detailed review. They found that the methodological quality of the studies was moderate to poor, thus their results should be interpreted with caution. A consistent finding was that counterpositioning with or without physiotherapy or helmet therapy may reduce skull deformity; however, it was not possible to draw conclusions regarding the relative effectiveness of these interventions.

Katz et al. (2010) asked parents of 61 patients with plagiocephaly to rate their children's head shape and ear position before and after helmet therapy (a score of 1 being abnormal and 10 being normal). Topographic laser head scans for an aged-matched cohort of 91 patients with deformational plagiocephaly were acquired. Parent ratings before and after molding, respectively, were head shape 2.99 ± 1.50 versus 7.88 ± 1.64 and ear position 3.75 ± 2.5 versus 7.73 ± 2.34 . Measurements before and after molding were cephalic ratio 0.89 ± 0.07 versus 0.87 ± 0.08 , overall symmetry index 0.87 ± 0.05 versus 0.90 ± 0.04 , radial symmetry index 59.9 ± 26.9 mm versus 46.3 ± 25.1 mm, cranial vault asymmetry index 7.2 ± 3.75 versus 4.8 ± 2.8 , and ear offset 5.7 versus 5.5 mm. The authors concluded that helmet molding produces reproducible changes in head shape. Despite relatively small actual changes on topographic laser imaging, parents perceive a large correction in head shape and ear position following helmet molding.

Professional Societies

American Academy of Pediatrics (AAP)

According to a recently updated clinical report published by the American Academy of Pediatrics (Laughlin, et al., 2011; Persing, et al. 2003) parents are recommended to have preventive counseling during the newborn period, including instruction on alternating the newborn's laying position while sleeping and giving the newborn some "tummy time" while awake. If deformational plagiocephaly is diagnosed, the AAP statement recommends mechanical adjustments, including positioning so that the rounded side of the head is placed against the mattress and a change in the layout of the room to cause a child to look away from the flattened side of the head to see parents or others in the room. For torticollis, neck motion exercises are recommended. If these therapies prove unsuccessful, the AAP statement recommends a neurosurgical consultation to ensure a proper diagnosis and to direct subsequent management, including consideration of molding helmets and surgery. The AAP statement concludes that the best response for helmets occurs in children aged 4 to 12 months due to the malleability of the infant skull. Surgery is only indicated when the deformities are severe and resistant to nonsurgical interventions.

In 2005 the AAP published a policy statement: The changing concept of Sudden Infant Death Syndrome that addresses positional plagiocephaly. The AAP statement reports a recent case-control study that has shown that many cases of plagiocephaly are associated with supine sleeping position. Children with developmental delay and/or neurologic injury have increased rates of plagiocephaly, although a causal relationship has not been demonstrated. One study showed that the incidence of plagiocephaly in healthy normal children decreases spontaneously from 20% at 8 months to 3% at 24 months of age.

In 2009, the AAP reviewed data on > 20,000 children (13,158 male and 6527 female patients) with deformational plagiocephaly (DP). The reviewers found that several factors were associated with DP such as intrauterine presentation, sleep position, and lateralization with sleep position being the single greatest predictor of lateralization. (Joganic, 2009)

Surgical Treatment for Craniosynostosis

The published evidence on surgical treatment for craniosynostosis consists of single-center prospective and retrospective case series reporting outcomes of a variety of procedures. Agraval et al. (2006) and Fearon et al. (2006) followed a very small number of patients treated for single sagittal synostosis for more than 10 years. They found that surgery leads to a significant improvement in the cephalic index, which is most marked in the early postoperative period. There is currently insufficient evidence that craniosynostosis surgery prevents or reduces risk of neurobehavioral impairment (Spelz et al., 2004) There are no randomized controlled studies that compare safety and efficacy of different treatment modalities for craniosynostosis. A few studies compared two different surgical procedures based on retrospective chart reviews. (Boop et al., 1996) There is some evidence that better correction of morphology occurs after cranial vault remodeling for sagittal craniosynostosis than with strip craniectomy. Panchal et al. (1999a) found that subtotal calvarectomy achieved normal cephalic index in the majority of the children, at least when performed within the first 13 months of life, extended strip craniectomy did not, even when performed before 4 months of age. However, photographic assessment by lay and professional observers has shown no advantage of one technique over the other. (Panchal et al., 1999b)

Spring-mediated Cranioplasty

David et al. (2010) conducted a study to compare the outcomes of the first 75 cases of spring-assisted surgery (SAS) for the treatment of sagittal with a prospectively collected group of patients treated with cranial expansion (cranial vault remodeling [CVR]). All patients successfully underwent SAS without significant complications with a mean follow-up of 46 months. Perioperative variables including odds ratio, time, blood loss, transfusion requirements, intensive care unit and hospital stay lengths, and hospital costs differed significantly in favor of SAS. The mean cephalic index improved from 69 preoperatively to 75.4 after SAS, comparable with the change from 66 to 72.5 for CVR. This correction was maintained at 3- and 5-year follow-ups. Anterior frontal bossing was corrected on three-dimensional scan volume measurements.

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Taylor et al. (2011) retrospectively compared the safety and efficacy of spring-mediated cranioplasty (SMC) and minimally invasive strip craniectomy with parietal barrel staving (SCPB) analyzing the hospital records of the first 7 SMCs and the last 7 SCPBs. All 14 patients successfully underwent cranial vault remodeling with significant improvement in cephalic index. Demographics, length of stay in the intensive care unit, preoperative cephalic index, and postoperative cephalic index were similar between SMC and SCPB. Spring-mediated cranioplasty had statistically significantly shorter operative time, less estimated blood loss and shorter length of hospital stay as compared with SCPB. Complications included 1 spring dislodgment in an SMC that did not require additional management and 1 undercorrection in the SCPB group. The authors stated that spring-mediated cranioplasty has become the predominant means of treatment of scaphocephaly in patients younger than 9 months because of its improved morbidity profile.

A retrospective study of 23 metopic synostosis patients operated with spring-assisted correction conducted by Maltese et al. (2007). The authors used a spring used together with a cranioplasty for the correction of both hypotelorism and orbital shape in trigonocephaly. Preoperative mean bony interorbital distance was 10.6 mm (range, 7.7 to 13.2 mm). It increased to 15.7 mm (range, 10.4 to 22 mm) at 1.5 months postoperatively and to 16.2 mm (range, 10.9 to 24.5 mm) 5 months postoperatively. Results as judged clinically ranged from little effect to a definitive overcorrection. The fronto-orbital axis was improved in every case. Average fronto-orbital axis was -4 degrees (range, -33 to 23 degrees) preoperatively and 28 degrees (range, 11 to 46 degrees) postoperatively

Endoscopic Strip Craniectomy with Cranial Helmet Molding

There is a moderate body of published literature available reporting the single center experience with endoscopic strip craniectomy for craniosynostosis. There are no randomized controlled trials on this therapy, and most studies performed retrospective analysis of the data.

In a retrospective study, Jimenez and Barone (2012) reported their 16-year experience (1996-2012) of treating sagittal synostosis with endoscopy-assisted techniques and postoperative cranial orthotic therapy in 256 patients. A wide-vertex craniectomy with bilateral barrel stave osteotomies of the temporal and parietal bones using small scalp incisions and endoscopic viewing techniques was performed. Author observations included: 1) Correction begins within 6 weeks of surgery; 2) Helmet use for up to 12 months maintains the correction and normalized cephalic indices; 3) Cephalic shape and morphology stabilize at approximately 18 months; and 4) Their longest follow-up of 15 years corroborates these findings.

Shah et al. (2011) prospectively observed 89 children less than 12 months old who were surgically treated for a diagnosis of isolated sagittal synostosis between 2003 and 2010. The endoscopic procedure was offered starting in 2006. There were 47 endoscopically treated patients with a mean age at surgery of 3.6 months and 42 patients with open-vault reconstruction whose mean age at surgery was 6.8 months. The mean follow-up time was 13 months for endoscopic versus 25 months for open procedures. The mean operating time for the endoscopic procedure was 88 minutes, versus 179 minutes for the open surgery. The mean blood loss was 29 ml for endoscopic versus 218 ml for open procedures. Three endoscopically treated cases (6.4%) underwent transfusion, whereas all patients with open procedures underwent transfusion, with a mean of 1.6 transfusions per patient. The mean length of stay was 1.2 days for endoscopic and 3.9 days for open procedures. Of endoscopically treated patients completing helmet therapy, the mean duration for helmet therapy was 8.7 months. The mean pre- and postoperative cephalic indices for endoscopic procedures were 68% and 76% at 13 months postoperatively, versus 68% and 77% at 25 months postoperatively for open surgery.

Jimenez et al. (2007) conducted a study with a total of 100 patients who presented with synostosis of the metopic or coronal sutures were consecutively treated during a 6-year period using minimally invasive endoscopic-assisted suturectomies. After surgery, all patients were fitted

with custom-made cranial helmets for up to 12 months. The coronal group consisted of 50 patients, 26 females and 23 males with a mean age of 3.78 months. The metopic group consisted of 50 patients, 35 males and 16 females with a mean age of 4.1 months. For the entire cohort, the mean estimated blood loss was 34 cc (5-250 cc). The mean estimated percent of blood volume lost was 5.2% (1-26%). There were no intraoperative blood transfusions and five postoperative for a total transfusion rate of 6.7%. The mean surgical time was 56 min. All but one patient (99%) was discharged on the first postoperative day. Complications included two dural tears and four pseudomeningoceles. There were two cases of incomplete reossification of the craniectomy. There were no infections, mortalities, hematomas, or visual injuries. There were no complications related to helmet therapy except three superficial skin breakdowns that cleared immediately with helmet non-use for 3-4 days. Using anthropometric measurements and extensive photographic and physical assessments, excellent results were obtained in 84%, good results in 9%, and poor results in 7% of patients.

Barone and Jimenez (2004) also reported on outcomes in 72 patients with coronal synostoses, with a reported follow-up of 7 years. Among these patients initial surgical results were similar and all were discharged on the first postoperative day. There was little blood loss and there were no blood transfusions. It was noted that vertical dystopia resolved within 1 year, but that frontal plagiocephaly improved more slowly. The best results were obtained in those aged < 3 months. Continued improvement and correction was noted up to 4 years after surgery in older patients. Small cranial defects remained in 3 patients. Authors reported that failure to achieve adequate correction of the deformities was the result of parental noncompliance with helmet therapy.

The same group described the efficacy and safety of endoscopic strip craniectomy along with a postoperative molding helmet in 185 young infants with variety of stenoses (107 sagittal, 42 coronal, 37 metopic, and 7 lambdoid, for a total of 198 suture). The mean blood loss was 29.4 cc, and only two patients underwent intraoperative blood transfusion. Fourteen patients underwent postoperative blood transfusion; none was life-threatening. There were no deaths, complications, neurological injuries, or infections. All but six patients were discharged on the first postoperative day. A majority of the patients achieved or approached normocephaly. (Cartwright et al., 2003) The earlier experience of this center in 100 patients with similar results were report in 2002 (Jimenez et al., 2002) Ninety-four of the 100 (94%) patients achieved normocephaly; all but 6 scaphocephalic patients have achieved normocephaly. Four of those patients were older (> 6 months), and 2 had significant compliance problems with the use of the helmet.

Tobias et al. (2001) investigated the incidence of venous air embolism (VAE) during endoscopic strip craniectomy using Doppler monitoring in a series of 50 consecutive patients. Four of 50 (8%) had a single episode of VAE, which resolved spontaneously. The authors report that the incidence of VAE in open craniectomy procedures can be significantly greater at > 80%, but advised the use of monitoring for VAE during endoscopic procedures despite the relatively low incidence.

Murad et al. (2005) reported the results of endoscopic strip craniectomy in 19 patients with sagittal (11) metopic (5), coronal (2) or sagittal and metopic (1) synostoses. Patients were operated on using a technique similar to that described by Jimenez and colleagues. Helmets were fitted 1 to 2 weeks after surgery and worn for a minimum of 12 months. Seventeen of 19 patients (89.5%) had good cosmetic results with 1 surgery, but 2 patients required > 1 operation. Most patients were discharged on postoperative day 1. The mean operative time was 97 minutes, with an estimated blood loss of mean 39 mL. Two of 19 patients (10.5%) underwent transfusion intraoperatively and 3/19 (15.8%) postoperatively; complications were minimal.

Teichgraeber et al. (2009) treated a cohort of 67 infants with nonsyndromic sagittal, unicoronal, bicoronal, and metopic craniosynostosis, either with the microscopic (n = 40) or the open (n = 27) approach. In the microscopic approach, incisions were placed over the premature suture, and using a surgical microscope, the appropriate synostectomy was performed. The open approach used a traditional coronal incision with cranial vault reconstruction. Both groups of patients had

postoperative molding helmet therapy. The median surgical times for microscopic and open approaches were 108 and 210 minutes, the volumes of blood loss were 75 and 220 mL, the durations of hospital stay were 2 and 4 days, the numbers of helmet were 2 and 1, and the durations of helmet therapy were 10.5 and 8 weeks, respectively. The analysis of variance for repeated measures showed that there was no statistically significant difference between the 2 groups in any of the craniosynostoses.

ONGOING STUDIES: No registered ongoing studies using endoscopic surgery for primary craniosynostosis were identified on the ClinicalTrials.gov online database, which is sponsored by the National Institutes of Health (ClinicalTrials.gov, 2012).

In summary, the perioperative variables and clinical outcomes reported for endoscopic strip craniotomy followed by extensive use of cranial orthotic devices demonstrate that the procedure is safe and effective. Long-term follow-up data using objective measures are still limited.

Cranial Orthotic Devices after Traditional Surgery for Craniosynostosis

Seymour-Dempsey et al. (2002) retrospectively compared the results of surgery alone (n=6) versus surgery and postoperative banding (n=15) in 21 children with sagittal synostosis. The authors used the cephalic index (CI) and the divergence of the CI from the norm (DFN) as objective outcome measures. Correction toward a normal CI was seen in the banded group throughout the course of treatment, while this trend was not present in the non-banded group.

Littlefield et al (2005) summarized the experience of Cranial Technologies, Inc (manufacturer) with the DOC band in 305 infants presented for postoperative treatment. Sixteen patients were excluded from the study due to noncompliance issues or for entering treatment more than four weeks after surgery was performed. Of the remaining 289 infants, nearly all forms of craniosynostosis were represented including 162 sagittal (56.1%), 44 metopic (15.2%), 38 unilateral coronal (13.1%), 12 bicoronal (4.2%), 15 unilateral lambdoid (5.2%), and 18 multiple suture synostosis (6.2%). A full range of surgical procedures were reported including minimally invasive endoscopic-assisted craniectomy, strip and extended strip craniectomy, and many variations of cranial vault remodeling. Mean entrance age was 6.9 months and mean treatment time was 3.3 months. Statistically significant improvement in cephalic index was documented with a mean improvement of 13.0% when considering all cases, and a 16.8% improvement when considering only sagittal synostosis.

Professional Societies

American Academy of Pediatrics (AAP)

The American Academy of Pediatrics (AAP) advises that infants with craniosynostosis be treated by a pediatric neurosurgeon with expertise in craniofacial malformations (Laughlin et al., 2011).

U.S. FOOD AND DRUG ADMINISTRATION (FDA)

Since 1998 cranial orthoses have been classified in the 510(k) database as class II (special controls) devices. They are intended for medical purposes to apply pressure to prominent regions of an infant's cranium in order to improve cranial symmetry and/or shape in infants from 3 to 18 months of age, with moderate to severe nonsynostotic positional plagiocephaly, including infants with plagiocephalic-, brachycephalic-, and scaphocephalic-shaped heads. The FDA has approved a large number of cranial orthoses. Additional information, under product code MVA, is available at: <http://www.accessdata.fda.gov/scripts/cdrh/cfdocs/cfPMN/pmn.cfm>. Accessed June 20, 2014

Additional Products

Specific devices include but are not limited to the following:

- Dynamic Orthotic Cranioplasty (DOC) Band, Cranial Technologies, Inc. (Phoenix, AZ)
- Ballert Cranial Molding Helmet, Ballert Orthopedic (Chicago, IL)

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- RHS Cranial Helmet, Restorative Health Services, Inc. (Nashville, TN)
- Cranial Shaping Helmet, Lone Star Orthotics, Inc. (Houston, TX)
- Hanger Cranial Band, Hanger Orthopedic Group, Inc. (Bethesda, MD)
- P.A.P. Orthosis (Plagiocephalic Applied Pressure Orthosis), Personal Performance Medical Corp.
- Static Cranioplasty Orthosis, Luke Stikeleather Co. (Fairfax, VA)
- & P Cranial Molding Helmet, Orthotic and Prosthetic Lab, Inc. (Evansville, IN)
- Cranial Solutions Orthosis CSO, Cranial Solutions (Pompton Lakes, NJ)
- Cranial Symmetry System, Beverly Hills Prosthetics Orthotics, Inc. (Los Angeles, CA)
- STARband[™], STARlight[™] Cranial Remolding Orthosis, and the Clarren Helmet, Orthomerica Products, Inc. (Newport Beach, CA)
- ECA orthosis, Eastern Cranial Affiliates (Arlington, VA)
- COPC Band, Center for Orthotic and Prosthetic Care of KY (Louisville, KY)
- cranial orthoses manufactured by:
 - Children's Health Care Services, Fountain Valley Prosthetics and Orthotics Inc. (Fountain Valley, CA)
 - Precision Prosthetics & Orthotics (St Louis, MO)

CENTERS FOR MEDICARE AND MEDICAID SERVICES (CMS)

Medicare does not have a National Coverage Determination (NCD) for cranial orthotic devices used in the treatment of plagiocephaly or surgically corrected craniosynostosis. Local Coverage Determinations (LCDs) do not exist at this time.

(Accessed June 25, 2014)

REFERENCES

American Academy of Orthotists and Prosthetists (AAOP). Orthotic Treatment of Deformational Plagiocephaly, Brachycephaly and Scaphocephaly. Clinical Standards of Practice (CSOP) Consensus Conference on Orthotic Management of Plagiocephaly, 2004. Available at: <http://www.oandp.org/olc/lessons/html/200508-22/display.asp?fmTermId=Deviations1>. Accessed June 20, 2014.

American Academy of Pediatrics Task Force on Sudden Infant Death Syndrome. The changing concept of sudden infant death syndrome: diagnostic coding shifts, controversies regarding the sleeping environment, and new variables to consider in reducing risk. *Pediatrics*. 2005 Nov;116(5):1245-55. Epub 2005 Oct 10.

American Academy of Pediatrics (AAP). Surgical Advisory Panel. Guidelines for referral to pediatric surgical specialists. *Pediatrics*. 2002;110(1 Pt 1):187-191. Available at <http://pediatrics.aappublications.org/content/110/1/187.full.pdf+html>. Accessed June 20, 2014.

American Academy of Pediatrics (AAP) Website. Clinical report. Prevention and management of positional skull deformities in infants. December 2011.

Argenta LC, David LR, Wilson JA, et al. An increase in infant cranial deformity with supine sleeping position. *J Craniofac Surg*. 1996;7(1):5-11.

Barone CM, Jimenez DF. Endoscopic approach to coronal craniosynostosis. *Clin Plast Surg*. 2004;31(3):415-422.

Bialocerkowski AE, Vladusic SL, Howell SM. Conservative interventions for positional plagiocephaly: a systematic review. *Dev Med Child Neurol*. 2005 Aug;47(8):563-70. Review.

Boop FA, Shewmake K, Chadduck WM. Synostectomy versus complex cranioplasty for the Plagiocephaly and Craniosynostosis Treatment: Medical Policy (Effective 10/01/2014)

treatment of sagittal synostosis. *Childs Nerv Syst* 1996; 12: 371-5.

Brent R. Collett, Kristen E. Gray, Jacqueline R. Starr, et al. Development at Age 36 Months in Children With Deformational Plagiocephaly. *Pediatrics* 2013;131:e109

Bruner TW, David LR, Gage HD, et al. Objective outcome analysis of soft shell helmet therapy in the treatment of deformational plagiocephaly. *J Craniofac Surg.* 2004;15(4):643-650.

Cartwright CC, Jimenez DF, Barone CM, et al. Endoscopic strip craniectomy: a minimally invasive treatment for early correction of craniosynostosis. *J Neurosci Nurs.* 2003 Jun;35(3):130-8.

Clayman MA, Murad GJ, Steele MH, et al. History of craniosynostosis surgery and the evolution of minimally invasive endoscopic techniques: the University of Florida experience. *Ann Plast Surg.* 2007 Mar;58(3):285-7.

ClinicalTrials.gov [online database]. Search for Clinical Trials [craniosynostosis AND endoscopy]. 2012. Available at: <http://www.clinicaltrials.gov>. Accessed June 20, 2014.

David LR, Plikaitis CM, Couture D, et al. Outcome analysis of our first 75 spring-assisted surgeries for scaphocephaly. *J Craniofac Surg.* 2010 Jan;21(1):3-9.

Farkas LG, Munro IR, editors. *Anthropometric facial proportions in medicine.* Springfield, IL, Charles C. Thomas; 1987.

Fearon JA, McLaughlin EB, Kolar JC. Sagittal craniosynostosis: surgical outcomes and long-term growth. *Plast Reconstr Surg.* 2006 Feb;117(2):532-41.

Graham JM Jr, Gomez M, Halberg A, et al. Management of deformational plagiocephaly: repositioning versus orthotic therapy. *J Pediatr.* 2005a Feb;146(2):258-62.

Graham JM Jr, Kreutzman J, Earl D, et al. Deformational brachycephaly in supine-sleeping infants. *J Pediatr.* 2005b Feb; 146(2):253-7.

Hayes, Inc. Health Technology Brief. Endoscopic Strip Craniectomy with Cranial Helmet Molding for Primary Craniosynostosis. Lansdale, PA.: Hayes, Inc.; May 2007. Last updated May 21, 2009. Archived June 13, 2010.

Hayes, Inc. Health Technology Brief. Endoscopic Surgery for Primary Craniosynostosis. Lansdale, PA.: Hayes, Inc.; November 2012.

Hayes, Inc. Hayes Medical Technology Directory. Cranial Orthotic Devices. Lansdale, PA: Hayes, Inc.; Updated February 2013.

Hutchison BL, Hutchison LA, Thompson JM, et al. Plagiocephaly and brachycephaly in the first two years of life: a prospective cohort study. *Pediatrics.* 2004 Oct;114(4):970-80.

Hutchison BL, Stewart AW, de Chalain TB, et al. A randomized controlled trial of positioning treatments in infants with positional head shape deformities. *Acta Paediatr.* 2010 May 19.

Jimenez DF, Barone CM. Endoscopy-assisted wide-vertex craniectomy, "barrel-stave" osteotomies, and postoperative helmet molding therapy in the early management of sagittal suture craniosynostosis. *Neurosurg Focus.* 2000 Sep 15; 9(3):e2.

Jimenez DF, Barone CM. Early treatment of anterior calvarial craniosynostosis using endoscopic-assisted minimally invasive techniques. *Childs Nerv Syst.* 2007 Dec;23(12):1411-9.

Plagiocephaly and Craniosynostosis Treatment: Medical Policy (Effective 10/01/2014)

Jimenez DF, Barone CM, Cartwright CC, et al. Early management of craniosynostosis using endoscopic-assisted strip craniectomies and cranial orthotic molding therapy. *Pediatrics*. 2002 Jul;110(1 Pt 1):97-104.

Jimenez DF, Barone CM. Multiple-suture nonsyndromic craniosynostosis: early and effective management using endoscopic techniques. *J Neurosurg Pediatr*. 2010;5(3):223-231. Available at: <http://thejns.org/doi/pdf/10.3171/2009.10.PEDS09216>. Accessed June 20, 2014.

Joganic JL, Lynch JM, Littlefield TR et al. Risk Factors Associated With Deformational Plagiocephaly. *Pediatrics* 2009;124:e1126-e1133.

Kabbani H, Raghuvver TS. Craniosynostosis. *Am Fam Physician*. 2004 Jun 15;69(12):2863-70. Review.

Kane AA. An overview of craniosynostosis. *JPO*. 2004;16(4S):50-55. American Academy of Orthotists & Prosthetists (AAOP). Available at: http://www.oandp.org/jpo/library/2004_04S_050.asp. Accessed June 20, 2014.

Katzel EB, Koltz PF, Sbitany H, Giroto JA. Treatment of plagiocephaly with helmet molding therapy: do actual results mimic perception? *Cleft Palate Craniofac J*. 2011 Mar;48(2):205-9. Epub 2010 Apr 21.

Kluba S, Kraut W, Reinert S, Krimmel M. What is the optimal time to start helmet therapy in positional plagiocephaly? *Plast Reconstr Surg*. 2011 Aug;128(2):492-8.

Laughlin J, Luerksen 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.

Lauritzen C, Sugawara Y, Kocabalkan O, et al. Spring mediated dynamic craniofacial reshaping: case report. *Scand J Plast Reconstr Surg Hand Surg* 1998; 32: 331-8.

Lipira AB, Gordon S, Darvann TA, Hermann NV, Van Pelt AE, Naidoo SD, Govier D, Kane AA. Helmet versus active repositioning for plagiocephaly: a three-dimensional analysis. *Pediatrics*. 2010 Oct;126(4):e936-45. Epub 2010 Sep 13.

Littlefield TR, Hess RM, Pomatto JK. Postoperative use of the cranial remodeling orthosis. American Academy of Orthotists & Prosthetists (AAOP) 2005 Journal of Proceedings. Available at: <http://www.oandp.org/publications/jop/2005/2005-2.asp>. Accessed June 20, 2014.

Loveday BP, de Chalain TB. Active counterpositioning or orthotic device to treat positional plagiocephaly? *J Craniofac Surg*. 2001;12(4):308-313.

Maltese G, Tarnow P, Lauritzen CG. Spring-assisted correction of hypotelorism in metopic synostosis. *Plast Reconstr Surg*. 2007 Mar;119(3):977-84.

McGarry A, Dixon M, Greig R, et al. Head shape measurement standards and cranial orthoses in the treatment of infants with deformational plagiocephaly. *Dev Med Child Neurol*. 2008 Aug;50(8):568-76.

Mulliken JB, Vander Woude DL, Hansen M, et al. Analysis of posterior plagiocephaly: deformational versus synostotic. *Plast Reconstr Surg*. 1999;103(2):371-380.

Murad GJ, Clayman M, Seagle MB, et al. Endoscopic-assisted repair of craniosynostosis. *Neurosurg Focus*. 2005 Dec 15;19(6):E6.

Plagiocephaly and Craniosynostosis Treatment: Medical Policy (Effective 10/01/2014)

National Institute of Neurological Disorders and Stroke (NINDS). Craniosynostosis Information Page. Last updated September 16, 2008. Available at: <http://www.ninds.nih.gov/disorders/craniosynostosis/craniosynostosis.htm>. Accessed June 20, 2014.

Panchal J, Marsh JL, Park TS, et al. Sagittal craniosynostosis outcome assessment for two methods and timings of intervention. *Plast Reconstr Surg*. 1999a May;103(6):1574-84.

Panchal J, Marsh JL, Park TS, et al. Photographic assessment of head shape following sagittal synostosis surgery. *Plast Reconstr Surg*. 1999b May;103(6):1585-91.

Pattisapu JV, Walker ML, Myers GG, et al. Use of helmets for positional molding. *Concepts Pediatr Neurosurg*. 1989; 9:178-184.

Persing J, James H, Swanson J, et al. Prevention and management of positional skull deformities in infants. American Academy of Pediatrics Committee on Practice and Ambulatory Medicine, Section on Plastic Surgery and Section on Neurological Surgery. *Pediatrics*. 2003;112(1 Pt 1):199-202.

Podda S, Wolfe A, Kordestani RK, et al. Craniosynostosis management. Updated September 17, 2008.

Podda S, Ciminello F, Wolfe A. Congenital, Synostoses. Updated August 18, 2010. eMedicine [website].

Pollack IF, Losken HW, Fasick P. Diagnosis and management of posterior plagiocephaly. *Pediatrics*. 1997;99(2):180-185.

Rekate HL. Occipital plagiocephaly: a critical review of the literature. *J Neurosurg*. 1998;89(1):24-30.

Seymour-Dempsey K, Baumgartner JE, Teichgraeber JF, et al. Molding helmet therapy in the management of sagittal synostosis. *J Craniofac Surg* 2002;13:631-635.

Shah MN, Kane AA, Petersen JD, Woo et. Al. . Endoscopically assisted versus open repair of sagittal craniosynostosis: the St. Louis Children's Hospital experience. *J Neurosurg Pediatr*. 2011 Aug;8(2):165-70.

Sheth RD, Iskandar BJ. Pediatric craniosynostosis. Updated. May 2012. eMedicine [website]. Available at: <http://www.emedicine.com/neuro/topic80.htm>. Accessed June 20, 2014.

St. John D, Mulliken JB, Kaban, LB, Padwa BL. Anthropometric analysis of mandibular asymmetry in infants with deformational posterior plagiocephaly. *J Oral Maxillofac Surg*. 2002;60(8):873-877.

Taylor JA, Maugans TA. Comparison of spring-mediated cranioplasty to minimally invasive strip craniectomy and barrel staving for early treatment of sagittal craniosynostosis. *J Craniofac Surg*. 2011 Jul;22(4):1225-9.

Teichgraeber JF, Baumgartner JE, Waller AL, Reis SM, Stafford MT, Hollinger LE, Gateno J, Xia JJ. Microscopic minimally invasive approach to nonsyndromic craniosynostosis. *J Craniofac Surg*. 2009 Sep;20(5):1492-500.

Teichgraeber JF, Seymour-Dempsey K, Baumgartner JE, et al. Molding helmet therapy in the treatment of brachycephaly and plagiocephaly. *J Craniofac Surg*. 2004;15(1):118-123.

Plagiocephaly and Craniosynostosis Treatment: Medical Policy (Effective 10/01/2014)

Tobias JD, Johnson JO, Jimenez DF, et al. Venous air embolism during endoscopic strip craniectomy for repair of craniosynostosis in infants. *Anesthesiology*. 2001;95(2):340-342.

Xia JJ, Kennedy KA, Teichgraeber JF, Wu KQ, Baumgartner JB, Gateno J. Nonsurgical treatment of deformational plagiocephaly: a systematic review. *Arch Pediatr Adolesc Med*. 2008 Aug;162(8):719-27.

van Wijk RM, van Vlimmerean LA, et. al. Helmet therapy in infants with positional skull deformation: r09 andomised controlled trial. *BMJ* 2014; 348:g2741

POLICY HISTORY/REVISION INFORMATION

Date	Action/Description
10/01/2014	<ul style="list-style-type: none"> • Reorganized policy content • Updated benefit considerations; added language for <i>Essential Health Benefits for Individual and Small Group</i> plans to indicate: <ul style="list-style-type: none"> ○ For plan years beginning on or after January 1, 2014, the Affordable Care Act of 2010 (ACA) requires fully insured non-grandfathered individual and small group plans (inside and outside of Exchanges) to provide coverage for ten categories of Essential Health Benefits (“EHBs”) ○ Large group plans (both self-funded and fully insured), and small group ASO plans, are not subject to the requirement to offer coverage for EHBs; however, if such plans choose to provide coverage for benefits which are deemed EHBs (such as maternity benefits), the ACA requires all dollar limits on those benefits to be removed on all Grandfathered and Non-Grandfathered plans ○ The determination of which benefits constitute EHBs is made on a state by state basis; as such, when using this guideline, it is important to refer to the enrollee’s specific plan document to determine benefit coverage • Updated coverage rationale: <ul style="list-style-type: none"> ○ Reformatted and relocated information pertaining to medical necessity review; added language to indicate if service is “medically necessary” or “not medically necessary” to applicable proven/unproven statement ○ Added reference to the <i>description of services</i> section of the policy for additional information regarding anthropometric measurements and Cephalic Index graph • Updated list of applicable HCPCS codes; added A8003 • Updated supporting information to reflect the most current description of services, clinical evidence, CMS information and references • Archived previous version 2013T0031K