UnitedHealthcare® Oxford

PLAGIOCEPHALY AND CRANIOSYNOSTOSIS TREATMENT

Policy Number: SURGERY 067.11 T2   **Effective Date**: October 1, 2017

Related Policies
• Cosmetic and Reconstructive Procedures
• Durable Medical Equipment, Orthotics, Ostomy Supplies, Medical Supplies, and Repairs/ Replacement
• Prosthetic Devices, Wigs, Specialized, Microprocessor or Myoelectric Limbs

INSTRUCTIONS FOR USE

This Clinical Policy provides assistance in interpreting Oxford benefit plans. Unless otherwise stated, Oxford policies do not apply to Medicare Advantage members. Oxford reserves the right, in its sole discretion, to modify its policies as necessary. This Clinical Policy is provided for informational purposes. It does not constitute medical advice. The term Oxford includes Oxford Health Plans, LLC and all of its subsidiaries as appropriate for these policies.

When deciding coverage, the member specific benefit plan document must be referenced. The terms of the member specific benefit plan document [e.g., Certificate of Coverage (COC), Schedule of Benefits (SOB), and/or Summary Plan Description (SPD)] may differ greatly from the standard benefit plan upon which this Clinical Policy is based. In the event of a conflict, the member specific benefit plan document supersedes this Clinical Policy. All reviewers must first identify member eligibility, any federal or state regulatory requirements, and the member specific benefit plan coverage prior to use of this Clinical Policy. Other Policies may apply.

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.

CONDITIONS OF COVERAGE

Applicable Lines of Business/ Products	This policy applies to Oxford Commercial plan membership.
Benefit Type	General benefits package Durable medical equipment (DME)[1]
Referral Required (Does not apply to non-gatekeeper products)	No
Authorization Required (Precertification always required for inpatient admission)	Yes[2]
Precertification with Medical Director Review Required	Yes[2]
Applicable Site(s) of Service (If site of service is not listed, Medical Director review is required)	Inpatient - Surgical treatment Home - Cranial orthotic devices
Special Considerations	[1]Cranial orthotic devices are covered only for Members with a durable medical equipment benefit or benefit rider. [2]Review by a Medical Director or their designee is required.

Before using this policy, please check the member specific benefit plan document and any federal or state mandates, if applicable.

When deciding coverage for this service, the member specific benefit plan 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. 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 member specific benefit plan 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 considered medically necessary 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.

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, 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 policy, it is important to refer to the member specific benefit plan document to determine benefit coverage.

Cranial orthotic devices are reconstructive and medically necessary for:
- Craniosynostosis (i.e., synostotic plagiocephaly) following surgical correction.
- 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 documentation of a trial of conservative therapy of at least 2 months duration with cranial repositioning, with or without 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 device.

Note:
- Please see the Description of Services for additional information pertaining to Anthropometric measurements and the Cephalic Index graph.
- Please see the list of Related Policies for information pertaining to the repair and replacement of cranial orthotic devices.

Cranial orthotic devices are cosmetic and not medically necessary for treating infants with mild to moderate plagiocephaly.
There are no definitive data demonstrating 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 and medically necessary as a form of surgical treatment to repair craniosynostosis.

APPLICABLE CODES

The following list(s) of procedure and/or diagnosis codes is provided for reference purposes only and may not be all inclusive. Listing of a code in this policy does not imply that the service described by the code is a covered or non-covered health service. Benefit coverage for health services is determined by the member specific benefit plan 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 claim payment. Other Policies may apply.

CPT Code	Description
21175	Reconstruction, bifrontal, superior-lateral orbital rims and lower forehead, advancement or alteration (e.g., plagiocephaly, trigonocephaly, brachycephaly), with or without grafts (includes obtaining autografts)
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)

CPT® is a registered trademark of the American Medical Association

CDT Code	Description
D5924	Cranial prosthesis

CDT® is a registered trademark of the American Dental Association

HCPCS Code	Description
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)

DESCRIPTION OF SERVICES

Plagiocephaly: (An asymmetrical head shape) is most often the result of 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 or positional cranial deformity (PCD), 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). Positional skull deformities are generally benign, reversible head-shape anomalies that do not require surgical intervention.

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. This can result in neurologic damage and progressive craniofacial distortion.

The involved suture and anatomical name is listed below for the types of craniosynostosis:

- Primary craniosynostosis (PC) is a general term for the improper development and premature closure of sutures of the bones of the skull.
- Simple (or isolated) craniosynostosis classifications include:
    - Sagittal or scaphocephaly (cephal="head") – scaphocephaly (boat-shaped) – dolichocephaly (long)
    - Coronal (bilateral) – brachycephaly (short)
    - Coronal (unilateral) – plagiocephaly (diagonal)
    - Coronal (anterior plagiocephaly)
    - Metopic trigonocephaly (triangle-shaped)
    - Lambdoidal (bilateral) – posterior or occipital brachycephaly
    - Lambdoidal (unilateral) – posterior or occipital plagiocephaly
- Compound craniosynostosis

Non-Synostonic Positional Plagiocephaly

Positional plagiocephaly is treated conservatively and many cases do 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

Treatment for positional plagiocephaly is based on the age of the infant and the severity of the deformity. The optimal treatment is prevention through active counterpositioning of sleeping babies until they are able to move their heads freely during sleep, usually by six months of age.

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 SIDS, has been associated with an increased frequency of positional plagiocephaly due to pressure of the back of the head against a firm mattress. Prevention and management of positional skull deformities in infants includes anticipatory counseling for parents, mechanical adjustments, and exercises. (Laughlin, et al., 2011)

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. [National Institute of Neurological Disorders and Stroke, (NINDS) 2010]

Craniosynostosis requires surgical treatment to open the prematurely closed suture(s) in order to allow for normal brain growth. (NINDS, 2010; Clayman et al., 2007) The major complications associated with uncorrected craniosynostosis include increased intracranial pressure and abnormal brain development.

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. Cranial vault remodeling involves removal of the fused suture, multiple osteotomies and remodeling of the skull using plates and screws as necessary. (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 4-7 days. (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)

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., 2002)

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 millimeters 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)]
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:

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)

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

There are no definable standard criteria for starting helmet therapy and treatment decision is influenced more strongly by factors other than medical evidence, such as physician preference. (Kim et al., 2013)

CLINICAL EVIDENCE

Cranial Orthotic Devices

Cranial orthotic devices (COD) 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.

Lam et al. (2017) performed a retrospective chart review of their single institutional experience (2008-2014) analyzing improvements associated with various treatment modalities for positional plagiocephaly. . Univariate and multivariate analyses were used to assess the impact of these variables on the change in measured oblique diagonal difference (ODD) on head shape surface scanning pre- and posttreatment. A total of 991 infants < 1 year of age

(average age 6.2 months) were evaluated for cranial positional deformity in a dedicated clinical program. The most common deformity was occipital plagiocephaly (69.5%), followed by occipital brachycephaly (18.4%) or a combination of both (12.1%). Recommended treatment included repositioning (RP), physical therapy (PT) if indicated, or the use of a customized cranial orthosis (CO). Of the 991 eligible patients, 884 returned for at least 1 follow-up appointment and 552 patients were followed to treatment completion. Of the 991 patients, 543 (54.8%) had RP or PT as first recommended treatment. Of these 543 patients, 137 (25.2%) transitioned to helmet therapy after the condition did not improve over 4-8 weeks. In the remaining cases, RP/PT had already failed before the patients were seen in this program, and the starting treatment recommendation was CO. At the end of treatment, the measured improvements in ODD were 36.7%, 33.5%, and 15.1% for patients receiving CO, RP/PT/CO, and RP/PT, respectively. Orthotic treatment corresponded with the largest ODD change, while the RP/PT group had the least change in ODD. Earlier age at presentation corresponded with larger ODD change. The authors concluded that obtaining treatment at an earlier age as well as the type of treatment utilized impacts the degree of measured deformational head shape correction in positional plagiocephaly. This study suggests that treatment with a custom CO can result in more improvement in objective measurements of head shape.

Freudlsperger et al. (2016) investigated the impact of starting age and severity on the effectiveness of helmet therapy. A total of 213 pediatric patients treated for positional plagiocephaly with an asymmetry were measured according to the Cranial Vault Asymmetry Index (CVAI) using 3D-Photogrammetry. Patients were classified by age at which treatment was started: Group 1 was comprised of patients younger than 24 weeks; Group 2, those aged 24-32 weeks; Group 3, those aged >32 weeks. Groups were also categorized by severity. Mean initial CVAI was 9.8%, which reduced to 5.4%.

after helmet treatment. Group 1 (<24 weeks) showed the highest absolute and relative rate of correction. Within the groups, severity correlated positively with relative and absolute reduction of the asymmetry. A significant difference in the reduction of the CVAI depending on age was only seen in moderate and severe cases of plagiocephaly- but not in mild plagiocephaly. The authors concluded the present study confirms the effectiveness of helmet therapy for positional plagiocephaly. The use of an orthotic device is an appropriate treatment option particularly in infants with severe plagiocephaly and a start of helmet therapy before the age of 6 month is advisable.

Ho et al. (2016) conducted a retrospective review on data from a single institution (2009-2012) to investigate the effectiveness of helmet therapy compared to no helmet therapy in the treatment of positional plagiocephaly in infants under one year of age. Participants included 171 infants with positional plagiocephaly (127 males, 44 females). Eighty-four patients received a helmet while 87 were not prescribed any therapy. The decision to initiate helmet therapy was made by the clinician based on the examination, parents' wishes, and recommendation of the orthotist. Mean age at the initial consultation was 7.38 months with an average follow-up length of 5.85 months. Those with helmets had a longer mean follow-up than those without helmets (7.78 versus 3.85 months). In general, there was a reduction in overall plagiocephaly score regardless of whether or not the infant had helmet therapy. This suggests, that while the cosmetic abnormalities resulting from positional plagiocephaly improved in both groups, those with helmets may have had a greater benefit. The authors concluded that there may be a role for helmet therapy in the treatment of positional plagiocephaly, particularly in those with severe cosmetic deformity. However, further randomized controlled trials are required to produce more conclusive evidence.

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.

In a study of 1050 infants, Couture et al. (2013) reported successful use of off-the-shelf helmet therapy. Infants with an Argenta classification type I (minimal deformity) were treated with repositioning while infants with an Argenta severity rating of II - V were treated with a helmet. Correction (overall rate of 81.6%) took longer in patients with an Argenta severity of III, IV, and V compared with Argenta type II, but was not significantly affected by age.

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.

Graham et al. (2005) 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 - 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 - 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. The authors concluded that implementation of helmet treatment at a younger age resulted in more improvement in the CI.

CODs are accepted as treatment for positional cranial deformity (PCD) by major organizations. This is likely due to the large number of trials encompassing a large study population that produced consistently positive outcomes with minimal reports of complications.

Professional Societies
American Association of Neurological Surgeons (AANS)/Congress of Neurological Surgeons (CNS)
Evidence-based guidelines were developed by a multidisciplinary task force comprised of the Joint Section on Pediatric Neurosurgery of the AANS and the CNS recommend the following regarding the management of pediatric positional plagiocephaly:
- Repositioning therapy or devices may be effective as sole therapy to improve cranial asymmetry, particularly for mild-moderate deformity.
- PT is recommended and is effective over repositioning alone for reducing the prevalence of infantile positional plagiocephaly in infants 7 weeks of age.
- Helmet therapy is recommended for infants with persistent moderate-severe plagiocephaly after a course of conservative treatment (repositioning and/or physical therapy). (Flannery et al., 2016)

American Academy of Pediatrics (AAP)
The AAP endorsed the evidence-based guidelines mentioned above, developed by the multidisciplinary task force comprised of the Joint Section on Pediatric Neurosurgery of the AANS and the CNS. (Lam and Luerssen, 2016)

A clinical report published by the AAP recommends educating parents of newborns about the importance of alternating laying positions while sleeping and providing "tummy time" to prevent positional cranial deformity. If deformational plagiocephaly develops, the AAP recommends mechanical adjustments such as positioning so that the rounded side of the head is placed against the mattress or rearranging the room so that the child looks away from the flattened side of the head to see others in the room. For torticollis, neck motion exercises are recommended. If these therapies prove unsuccessful, a neurosurgical consultation is recommended to ensure a proper diagnosis and to direct subsequent management (including cranial orthotics and/or surgery). The report concludes that the best response for helmets occurs in children aged 4-12 months due to the malleability of the infant skull, and that surgery is indicated only when the deformities are severe and resistant to nonsurgical interventions. (Laughlin, et al., 2011)

Canadian Paediatric Society (CPS)
The CPS issued recommendations for the management of positional plagiocephaly. For the management of most children with mild or moderate positional plagiocephaly, the CPS recommends repositioning therapy plus PT as needed. Molding (helmet) therapy may be considered for children with severe asymmetry. The recommendations note that helmet therapy has been shown to influence the rate of improvement of asymmetry but not its final outcome. In addition, the CPS considers the evidence regarding the use of helmet therapy for the treatment of mild or moderate asymmetry insufficient. (Cummings et al., 2011. Reaffirmed 2016)

Surgical Treatment for Craniosynostosis
Surgery is recommended based on the age of the child, presence of other medical conditions, and caretaker preference.

Traditional Surgery ("Calvarial Vault Remodeling")
Thwin et al. (2015) conducted a systematic review and meta-analysis to identify and synthesize the best available evidence on the morphological, functional and neurological outcomes of craniectomy compared to cranial vault remodeling in infants on or before the mean age of 24 months. Twenty-seven studies were considered suitable for this review, and all were descriptive in nature. Authors concluded that neither procedure offers a clear long-term advantage over the other in short, medium and longer term time points relative to cephalic index and longer follow-up is required to compare outcomes further. Patients who have surgery of any type for isolated sagittal synostosis may have deficiencies in different subdomains at later school-age testing and be at risk for learning disorders, but maintain an age-appropriate global intelligence quotient (IQ) and school performance. There is no evidence suggesting that surgery of either type imparts any benefit relative to functional or neurological outcomes. Higher quality primary

research comparing the morphological and functional outcomes of craniectomy and cranial vault remodeling in primary sagittal synostosis is necessary, measuring both short and long term outcomes.

Fearon et al. (2006) followed a very small number of patients treated for single sagittal synostosis for more than 10 years. They concluded that surgery leads to a significant improvement in the cephalic index, which is most marked in the early postoperative period.

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.

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 (Minimally Invasive Surgery) with Cranial Helmet Molding

In an in-depth retrospective study, Jimenez and Barone (2013) reported their experience (1996-2012) of treating coronal synostosis with minimally invasive endoscopy-assisted suture osteotomies in young infants, followed by cranial molding using cranial orthoses. During the 16-year period, a total of 115 patients underwent successful operations without any deaths and were followed closely for an average of 7.7 years. The authors' experience conveyed that early treatment of coronal synostosis with endoscopy-assisted craniectomy and postoperative molding helmets leads to significant correction of craniofacial abnormalities, including vertical dystopia, nasal deviation, sagittal misalignment, and ipsilateral proptosis. It was their opinion that the endoscopic approach was superior to traditional calvarial remodeling techniques, citing reduced intraoperative and postoperative risks as well as optimal long term benefits. They concluded that endoscopic treatment of coronal synostosis is a safe, cost-effective, and highly efficacious method of treatment, and this option should be considered and offered to parents of children affected by the condition.

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.

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.

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.

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.

Professional Societies

American Association of Neurological Surgeons (AANS)

In a Patient Fact Sheet on craniosynostosis and craniofacial disorders, the AANS states that when it is indicated, most experts recommend surgical treatment between the ages of 3 - 8 months, depending on the case and surgical procedure. They state that early intervention is beneficial for several reasons, including prevention of further deformities: the bones are most malleable at this age, bone re-growth is quicker and more likely, and rapid brain growth benefits from skull remodeling. However, The AANS does not endorse any particular treatments or procedures specifically.

American Academy of Pediatrics (AAP)

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

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.

There are multiple open clinical trials studying craniosynostosis. For more information, go to www.clinicaltrials.gov.

U.S. FOOD AND DRUG ADMINISTRATION (FDA)

Cranial orthoses are classified by the FDA as Class II devices. This classification requires special controls, including prescription use, biocompatibility testing, and labeling (contraindications, warnings, precautions, adverse events, and instructions for physicians and parents). 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 May 22, 2017)

The foregoing Oxford policy has been adapted from an existing UnitedHealthcare national policy that was researched, developed and approved by UnitedHealthcare Medical Technology Assessment Committee. [*2017T0031O*]

American Association of Neurological Surgeons (AANS) Web site. Craniosynostosis and craniofacial disorders - (Patient Fact Sheet information).

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.

Collett BR, Gray KE, Starr JR, et al. Development at Age 36 Months in Children With Deformational Plagiocephaly. Pediatrics 2013;131;e109.

Couture DE, Crantford JC, Somasundaram A et al. Efficacy of passive helmet therapy for deformational plagiocephaly: report of 1050 cases. Neurosug Focus 2013; 35(4):E4.

Cummings C. Canadian Paediatric Society Community Paediatrics Committee. Positional plagiocephaly. Paediatr Child Health. 2011;16(8):493-496.

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.

Flannery AM, Tamber MS, Mazzola C, et al. Congress of Neurological Surgeons Systematic Review and Evidence-Based Guidelines for the Management of Patients With Positional Plagiocephaly: Executive Summary. Neurosurgery. 2016 Nov;79(5):623-624.

Freudlsperger C, Steinmacher S, Saure D, et al. Impact of severity and therapy onset on helmet therapy in positional plagiocephaly. J Craniomaxillofac Surg. 2016 Feb;44(2):110-5.

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. Hayes Medical Technology Directory. Cranial Orthotic Devices for the Treatment of Positional Cranial Deformity. Lansdale, PA: Hayes, Inc.; July 2014. Updated July 2016.

Ho JP, Mallitt KA, Jacobson E, et al. Use of external orthotic helmet therapy in positional plagiocephaly. J Clin Neurosci. 2016 Jul;29:46-51.

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. Early treatment of coronal synostosis with endoscopy-assisted craniectomy and postoperative cranial orthosis therapy: 16-year experience. J Neurosurg Pediatr. 2013 Sep;12(3):207-19.

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.

Kim SY, Park MS, Yang JI, et al. Comparison of helmet therapy and counter positioning for deformational plagiocephaly. Ann Rehabil Med. 2013Dec;37(6):785-95.

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

Lam S, Pan IW, Strickland BA, et al. Factors influencing outcomes of the treatment of positional plagiocephaly in infants: a 7-year experience. J Neurosurg Pediatr. 2017 Mar;19(3):273-281.

Lam SK, Luersssen TG. New guidelines review evidence on PT, helmets for positional plagiocephaly. AAP News. October 27, 2016.

Laughlin J, Luerssen TG, Dias MS. Committee on Practice and Ambulatory Medicine Section on Neurological Surgery. Prevention and management of positional skull deformities in infants. Pediatrics. 2011 Dec;128(6):1236-41.

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, et al. Helmet versus active repositioning for plagiocephaly: a three-dimensional analysis. Pediatrics. 2010 Oct;126(4):e936-45.

Littlefield TR, Hess RM, Pomatto JK. Postoperative use of the cranial remodeling orthosis. American Academy of Orthotists & Prosthetists (AAOP) 2005 Journal of Proceedings.

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.

Naidoo SD, Skolnick GB, Patel KB, et al. Long-term outcomes in treatment of deformational plagiocephaly and brachycephaly using helmet therapy and repositioning: a longitudinal cohort study. Childs Nerv Syst. 2015 Sep;31(9):1547-52.

National Institute of Neurological Disorders and Stroke (NINDS). Craniosynostosis Information Page. Last updated October 26, 2010.

Orthoses in the treatment of infants with deformational plagiocephaly. Dev Med Child Neurol. 2008 Aug;50(8):568-76.

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

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, 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.

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, et al. . Microscopic minimally invasive approach to nonsyndromic craniosynostosis. J Craniofac Surg. 2009 Sep;20(5):1492-500.

Thwin M, Schultz TJ, Anderson PJ. Morphological, functional and neurological outcomes of craniectomy versus cranial vault remodeling for isolated nonsyndromic synostosis of the sagittal suture: a systematic review. JBI Database System Rev Implement Rep. 2015 Sep;13(9):309-68.

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.

van Wijk RM, van Vlimmerean LA, Groothuis-Oudshoorn CG et. al. Helmet therapy in infants with positional skull deformation: randomised controlled trial. BMJ 2014; 348:g2741.

POLICY HISTORY/REVISION INFORMATION

Date	Action/Description
10/01/2017	• Updated coverage rationale; replaced language indicating "cranial orthotic devices are cosmetic and not medically necessary *in* infants with mild to moderate plagiocephaly" with "cranial orthotic devices are cosmetic and not medically necessary *for treating* infants with mild to moderate plagiocephaly" • Updated supporting information to reflect the most current description of services, clinical evidence, and references • Archived previous policy version SURGERY 067.10 T2