Craniosynostosis Surgical Options

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APPLICATION STATEMENT

The application of the Clinical Coverage Guideline is subject to the benefit determinations set forth by the Centers for Medicare and Medicaid Services (CMS) National and Local Coverage Determinations and state-specific Medicaid mandates, if any.

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BACKGROUND

The normal skull consists of several plates of bone that are separated by sutures. The sutures (fibrous joints) are found between the bony plates in the head. As the infant grows and develops, the sutures close, forming a solid piece of bone, called the skull. Craniosynostosis is a condition in which the sutures close too early, causing problems with normal brain and skull growth. Premature closure of the sutures may also cause the pressure inside of the head to increase and the skull or facial bones to change from a normal, symmetrical appearance. The condition occurs in one out of 2,200 live births and affects males slightly more often than females. Craniosynostosis is most often sporadic (occurs by chance). In some families, craniosynostosis is inherited and is a feature of many
different genetic syndromes that have a variety of inheritance patterns and chances for reoccurrence, depending on the specific syndrome present.\textsuperscript{1,8}

Types of plagiocephaly include: \textsuperscript{2}

- **Anterior plagiocephaly** involves fusion of either the right or left side of the coronal suture that runs from ear to ear. Coronal synostosis causes the normal forehead and the brow to stop growing, producing a flattening of the forehead and the brow on the affected side - the forehead tends to be excessively prominent on the opposite side. The eye on the affected side may also have a different shape and there may be flattening of the back area (occipital). Unilateral lambdoidal suture synostosis causes posterior plagiocephaly.

A common cause of plagiocephaly is deformational (or positional) plagiocephaly refers to the asymmetrical head from repeated pressure to the same area of the head. Deformational plagiocephaly is not craniosynostosis and surgery is not required. It results when the occipital bone that is dependent (in one position) flattens due to pressure on the skull when a child is sleeping. The number of infants with deformational plagiocephaly has risen, likely due to the "Back to Sleep" campaign promoted by the American Academy of Pediatrics to help prevent sudden infant death syndrome (SIDS). Treatment options will be determined depending on severity.

- **Trigonocephaly** is a fusion of the metopic (forehead) suture which runs from the top of the head down the middle of the forehead, toward the nose. Early closure of this suture may result in a prominent ridge running down the forehead. The forehead looks pointed, like a triangle, with closely placed eyes (hypotelorism).

- **Scaphocephaly** is an early closure of fusion of the sagittal suture which runs from front to back, down the middle of the top of the head. This causes a long, narrow skull (from front to back and narrow from ear to ear).

The involved suture and anatomical name is listed below for the types of craniosynostosis:\textsuperscript{13,14,15,16,17}

1. **Primary.** Primary craniosynostosis (PC) is a general term for the improper development of the bones of the skull, which can result in an abnormal head shape in affected individuals. Severity of PC varies in patient to patient, although intelligence is usually unaffected. PC may occur as an isolated finding or as part of a syndrome. The main treatment is surgery, but not all affected children will require surgery. The exact cause of PC is unknown, although the skull abnormalities result from the abnormal hardening (ossification) of the cranial sutures. PC is distinguished from secondary craniosynostosis (SC), which occurs because of a primary failure in brain growth.\textsuperscript{17}

2. **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

3. **Compound craniosynostosis** classifications include:
   - Nonsyndromic.
     - Bicoronal
   - Syndromic. In addition to craniofacial malformations, syndromic craniosynostosis involves multiple systems (e.g., cardiac, genitourinary, musculoskeletal). Many patients have a family history of abnormal head shape. Clinical examination of infants with craniofacial malformations should include careful evaluation of the neck, spine, digits, and toes. Crouzon’s disease and Apert’s syndrome occur more frequently than the other syndromes associated with craniosynostosis.\textsuperscript{13}
     - Crouzon's disease is a birth defect characterized by abnormalities in the skull and facial bones, caused by a fusing of both sides of the coronal suture; it often causes the skull to be short in the
Front and the back. Flat cheek bones and a flat nose are also typical of this disorder. Crouzon’s disease is inherited through an autosomal-dominant pattern; nearly 60 percent of cases are new mutations, and many are associated with paternal age older than 35 years. Crouzon’s disease occurs in one of every 25,000 live births and accounts for 5 percent of cases of craniosynostosis. Nucleotide alterations causing amino-acid substitutions at the FGFR2 gene on chromosome 10 lead to the Crouzon phenotype. Clinical findings include brachycephalic craniosynostosis, significant hypertelorism, proptosis, maxillary hypoplasia, beaked nose and, possibly, cleft palate. Intracranial anomalies include hydrocephalus, Chiari 1 malformation, and hind-brain herniation (70 percent). Pathology of the ear and cervical spine is common. Infants with Crouzon’s disease do not have anomalies of the hands and feet as do infants with Apert’s syndrome.

- **Apert’s syndrome** (acrocephalosyndactyly) is a autosomal dominant disorder and craniofacial abnormality characterized by an abnormal head shape, small upper jaw, and fusion of the fingers and toes. It is caused by nucleotide alterations resulting in amino-acid substitutions, leading to a mutation in the FGFR2 gene located on chromosome 10. Craniosynostosis and symmetric syndactyly of the extremities are hallmarks of this syndrome. Clinical features include misshapen skull caused by coronal suture synostosis, wide-set eyes, mid-face hypoplasia, choanal stenosis, and shallow orbits. Intracranial anomalies include megalencephaly, hypoplastic white matter, and agenesis of the corpus callosum, leading to cognitive impairment. Cardiac anomalies, including atrial septal defect and ventricular septal defect, and renal anomalies such as hydronephrosis occur in 10 percent of these patients. It occurs in one of every 160,000 live births.

- **Pfeiffer’s syndrome** is a birth defect characterized by abnormalities of the skull, hands, and feet; it results in wide-set, bulging eyes, an underdeveloped upper jaw, and a beaked nose due to the head being unable to grow normally.

- **Saethre-Chotzen syndrome** is a birth defect characterized by abnormalities of the skull and facial bones. It results in wide-set eyes and droopy eyelids, and fingers may be abnormally short and webbed.

- **Brachycephaly** is a birth defect characterized by the disproportionate shortness of the head and is caused by a premature fusing of the coronal suture. It is commonly associated with a number of syndromes, such as Apert’s, Crouzon’s, Pfeiffer’s, Saethre-Chotzen, and Carpenter’s. Deformational brachycephaly can also occur from infant positioning during sleep.

4. Other

- **Multiple sutures**: pansynostosis (all), oxycephaly (conical), acrocephaly (pointed), turricephaly or turmschadel (tower), cloverleaf skull or kleeblattschadel
- **Encephalocele**: Characterized by a protrusion of the brain or its coverings through the skull.
- **Kleeblattschadel syndrome**: A birth defect characterized by abnormalities of the skull and facial bones. It is caused by a premature fusing of the fibrous sutures. Also called “cloverleaf skull.”
- **Oxycephaly**: A birth defect characterized by abnormalities in the skull and facial bones. This syndrome causes the top of the skull to be pointed or cone-shaped. It is caused by a premature fusing of the coronal and sagittal sutures. Also known as turricephaly or high-head syndrome.
- **Pierre Robin syndrome**: A birth defect characterized by abnormalities in the facial bones, resulting in a smaller than normal lower jaw or receding chin. The tongue often falls back in the throat causing difficulty breathing.
- **Torticollis**: Also known as wryneck, is a twisting of the neck that causes the head to rotate and tilt at an odd angle. It most often results from tightness of one of the neck muscles.

*Indications and Contraindications to Intervention*

In addition to the optimization of the growth potential of the brain in the early perinatal period, additional indications for surgical intervention in members with craniosynostosis include prevention of:
• Intracranial hypertension and its associated sequelae that occur in some with uncorrected synostosis
• Progression of the calvarial deformity
• Progression of the facial deformity

Surgery is advocated in early infancy because most brain growth occurs in the first year of life. Therefore, if the deformed sutures are not released, the deforming vectors of the continually growing brain result in progression of the deformity with increasing age. The osseous defects following surgery undergo reossification more completely if surgery is performed in those younger than 1 year compared with later. The calvarium in an infant aged 3-9 months is much more malleable, making it easier to shape and providing a better outcome.4

The only absolute contraindication to surgical intervention is the presence of microcephaly. Calvarial sutures close secondary to the lack of expansile force from the underlying brain. Surgical intervention just to release the fused sutures is associated with high rate of re-fusion and thus is not appropriate. Although surgery for craniosynostosis improves calvarial shape, it should not be considered cosmetic.4

The key to treating craniosynostosis is early detection and treatment. Some forms of craniosynostosis can affect the brain and development of a child. The degree of the problems is dependent on the severity of the craniosynostosis, the number of sutures that are fused, and the presence of brain or other organ system problems that could affect the child. Genetic counseling may be recommended by the physician to evaluate the parents of the child for any hereditary disorders that may tend to run in families. A child with craniosynostosis requires frequent medical evaluations to ensure that the skull, facial bones, and brain are developing normally. The medical team works with the child's family to provide education and guidance to improve the health and well-being of the child.3

Surgical Options

Traditional procedures involve some form of open surgery to remove the fused suture and remodel the skull, using plates and screws as necessary. These operations are usually recommended in infants 6 to 8 months of age, are lengthy (4-8 hours in duration), and can require postoperative hospitalization for 4 to 8 days. Confirmation of diagnosis and presurgical evaluation requires a multidisciplinary team that may involve geneticists, pediatric neurologists, plastic surgeons, neurosurgeons, endocrinologists, and ophthalmologists.6

Endoscopic surgery, such as endoscopy-assisted strip craniectomy or suturectomy, is less invasive than open cranial vault reconstruction and is best performed when the infant is younger than 6 months old. The surgeon uses an endoscope to remove the fused suture(s), which allows the growing brain to expand. After surgery, the infant wears a customized helmet that helps to mold the skull into a proper shape as the brain grows. A pediatric neurosurgeon or plastic surgeon makes two incisions in the scalp at each end of the fused suture, and a dissecting space is created beneath the scalp in a bloodless fashion using a rigid endoscope and an electrocautery device. The surgeon incises the skull at each end of the suture, and after separating the dura (tough outer membrane covering the brain) from the overlying skull, makes two more incisions to form a rectangular strip. This strip of bone containing the fused suture is removed through the scalp incisions in one or two pieces. The operation normally lasts between 1 and 2 hours and most patients are discharged in 1 to 2 days. Within 1 week of surgery, the infant is fitted with a customized helmet that applies pressure on abnormal bulges in the skull, but allows for expansion in the correct direction. The helmet helps to guide the expanding skull to a normal shape over 11 to 12 months. Most patients typically need one or two helmets during the treatment period. Follow-up appointments occur every 2 to 3 months.8

Children with syndromic deformities require careful follow-up into adulthood because they may develop recurrent stenosis and/or raised intracranial pressure from other causes (e.g., hydrocephalus, hindbrain hernia). Papilledema, if present initially, may herald a recurrence if seen on follow-up. Children with syndromic forms of craniosynostosis (e.g., Crouzon or Apert syndrome) can be affected by a variable degree of developmental delay. Long-term care and surveillance by an integrated neurosurgical and craniofacial multidisciplinary team is essential.15

Surgical interventions in non-syndromic craniosynostosis include:13

• Sagittal Synostosis. Involves either strip craniectomy or cranial vault remodeling with excision of the frontal, parietal, and occipital bones, which are trimmed, reshaped, and affixed with absorbable plates. Recently,
minimally invasive endoscopic strip craniectomy, which involves significantly less blood loss and a shorter hospital stay, has been successful.

- **Coronal Synostosis.** Objective is to increase anteroposterior dimensions of the calvaria.
- **Metopic Synostosis.** Objective is to increase the volume of the anterior cranial fossa.

The management of craniofacial syndromes includes correction of craniosynostosis between three and six months of age, and correction of limb defects between one and two years of age. When the patient is a young adult, surgeries to normalize appearance and correct malocclusion are done.

Potential intraoperative complications include massive blood loss and air embolism. Mortality rates are low according to recent reports. Careful follow-up of the patient is necessary after surgery to ensure that the sutures do not re-fuse. Postoperative monitoring of head circumference and checking for signs and symptoms of increased intracranial pressure are necessary.

**Professional Organizations**

The **American Academy of Orthotists and Prosthetists** draft consensus statement on "Orthotic management of deformational plagiocephaly stated that "very young infants who have not developed midline head control, rolling, or sitting, may require a second orthosis to prevent regression of the head shape". The AAOP stated that a second orthosis is rarely required but may be used in cases of increased severity, extenuating circumstance (infant with multiple health issues), or a very young infant (less than 3 months). Criteria for use of a second orthosis include ill-fitting orthosis after multiple attempts to adjust, age/severity indicators with a willingness to continue by the family, post-operative adjunct/ preventative measures. The guideline also noted that termination of the orthotic treatment program is recommended, without weaning, when head shape falls within normal limits. If unresolved torticollis exists or if sleeping patterns are poor (same side as flatness), use is continued for an additional 2 to 4 weeks. Furthermore, unshunted or uncontrolled hydrocephalus as well as craniosynostosis are contraindications for cranial remodeling orthosis.⁵

The **American Academy of Pediatrics (AAP)** advises that infants with craniosynostosis be treated by a pediatric neurosurgeon with expertise in craniofacial malformations.⁶ Additional guidelines, recommendations, or position statements were not identified on the AAP web site.

The **American Association of Neurological Surgeons (AANS)** states:⁷

Most experts recommend that babies undergo surgery between the ages of 3 to 8 months, depending on the case and surgical procedure. Early intervention is beneficial for several reasons, aside from 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.

Regarding endoscopic surgery, the AANS states the following:

A newer less invasive form of surgery utilizes endoscopy, but is only a viable option in specific cases of craniosynostosis. The preferred age for this surgery is 3 months, but the infant should be no older than 6 months, to obtain optimal results.

The **American Association for Pediatric Ophthalmology and Strabismus (AAPOS)** management guidelines provide recommendations in evaluating and treating pediatric patients:¹²

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<th>Age Category</th>
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<td>Birth – 12 months</td>
<td><strong>Isolated and syndromic craniosynostosis</strong></td>
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<td><strong>Diagnostic Work-Up:</strong> After diagnosis is made and before and after significant craniofacial surgery, a complete exam should be completed to assess for orbital/canthal dystopia, ptosis, proptosis/exorbitism, quality of lid closure, abnormal head position, forehead retrusion, visual acuity, pupil reactivity, strabismus, anterior segment/cornea, optic nerve and retina, refractive error.</td>
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<td><strong>Treatment Options:</strong> Exam schedule may be more frequent based on severity of visual and ocular abnormality.</td>
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<td><strong>Treatment Options:</strong> Early ptosis surgery for severe sight threatening ptosis, nasolacrimal system surgery, tarsorraphy if indicated, e.g. for spontaneous globe luxation or exposure keratopathy, spectacles for high or asymmetric refractive errors, ambylopia treatment, artificial tears or lube for exposure keratopathy. Consider strabismus surgery if no impending surgical orbital manipulation.</td>
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Chan and colleagues\(^9\) noted that craniosynostosis results in characteristic skull deformations. Correction of craniosynostosis has traditionally involved an open cranial vault remodeling (CVR) procedure. A technique recently developed endoscope-assisted cranietomy (EAC) repair in conjunction with a post-operative molding helmet to guide cranial growth. Few studies compared these 2 approaches to the treatment of the various forms of craniosynostosis. This study was a retrospective review of 57 patients who underwent craniosynostosis repair by either the endoscope-assisted or open techniques; and compared operating room times, blood loss, volume of transfused blood, length of hospital stay, and overall costs. The endoscopic technique was performed on younger children (4.7 months versus 10.6 months, \(p = 0.001\)), has shorter operating room times (2 hours 13 minutes versus 5 hours 42 minutes, \(p = 0.001\)), lower estimated blood loss (74.4 ml versus 280.2 ml, \(p = 0.001\)), less transfused blood (90.6 ml versus 226.9 ml), shorter hospital stays (1.2 days versus 4.9 days, \(p = 0.001\)), and decreased cost ($24,404 versus $42,744, \(p = 0.008\)) relative to the traditional open approach. The authors concluded that issues with the endoscope-assisted procedure primarily concerned the post-operative helmet regimen, specifically patient compliance (17.1 % non-compliance rate) and minor skin breakdown (5.7 %). The endoscope-assisted repair with post-operative helmet molding therapy was a cost-effective procedure with less operative risk and minimal post-operative morbidity. This was a valuable treatment option in younger patients with compliant care-givers.

Vogel and associates\(^10\) stated that the surgical management of infants with sagittal synostosis has traditionally relied on open CVR techniques; however, minimally invasive technologies, including EAC repair followed by helmet therapy (HT, EAC+HT), is increasingly used to treat various forms of craniosynostosis during the 1st year of life. These researchers determined the costs associated with EAC+HT in comparison with those for CVR. They performed a retrospective case-control analysis of 21 children who had undergone CVR and 21 who had undergone EAC+HT. Eligibility criteria included an age less than 1 year and at least 1 year of clinical follow-up data. Financial and clinical records were reviewed for data related to length of hospital stay and transfusion rates as well as costs associated with physician, hospital, and outpatient clinic visits. The average age of patients who underwent CVR was 6.8 months compared with 3.1 months for those who underwent EAC+HT. Patients who underwent EAC+HT most often required the use of 2 helmets (76.5 %), infrequently required a 3rd helmet (13.3 %), and averaged 1.8 clinic visits in the first 90 days after surgery. Endoscope-assisted cranietomy plus HT was associated with shorter hospital stays (mean of 1.10 versus 4.67 days for CVR, \(p < 0.0001\)), a decreased rate of blood transfusions (9.5 % versus 100 % for CVR, \(p < 0.0001\)), and a decreased operative time (81.1 versus 165.8 minutes for CVR, \(p < 0.0001\)). The overall cost of EAC+HT, accounting for hospital charges, professional and helmet fees, and clinic visits, was also lower than that of CVR ($37,255.99 versus $56,990.46, respectively, \(p < 0.0001\)). The authors concluded that EAC+HT were a less costly surgical option for patients than CVR. Furthermore, EAC+HT were associated with a lower utilization of peri-operative resources. Findings suggested that EAC+HT for infants with sagittal synostosis may be a cost-effective 1st-line surgical option.

Hinchcliff et al.\(^11\) stated that the current treatment of craniosynostosis is open surgical excision of the prematurely fused suture and CVR. Due to the change in skull morphology and the increase in volume, some tension on the skin flaps is noted with closure. Although complete wound breakdown is rare, it can be a devastating complication. These researchers presented their experience with the use of the SPY imaging system to visualize and record

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1 year – 9 years

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Medical Literature Review

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blood flow within the flaps of a 1-year old patient with anterior plagiocephaly. The authors concluded that intra-operative indocyanine green angiography has the potential to be a significant advantage in such cases, providing a safe and objective method to assess intra-operative scalp perfusion, allowing the surgeon to take additional measures to ameliorate any ischemic problems.

General Principles of Surgical Repair

Metopic synostosis is characterized by trigonocephaly. The forehead appears ridged, and the patient has hypotelorism and proptosis. This condition is repaired by advancing the orbital rims, which are noted to be recessed, in addition to removing the fused metopic suture. The forehead requires careful reconstruction. Some institutions perform an endoscopic strip suturectomy through a small incision and then helmet the child to reshape the head as the child grows.\textsuperscript{15}

Sagittal synostosis is characterized by a skull that is long and narrow. Correction requires reconstruction of the skull so that it is shorter and wider. One factor that must be taken into account during preoperative planning and repair is compensatory growth, which can be anterior, posterior, or both. Frontal bossing can be quite significant, particularly when compensatory growth is anterior. Surgical goals are to shorten and widen the skull. Additionally, a bifrontal craniotomy is required to correct the frontal bossing. In similar fashion, if the compensatory growth involves the occipital bone, then an occipital craniotomy is required. If compensation involves both the frontal and occipital bones, then surgery often needs to be performed in a modified prone position and should include both a bifrontal and occipital craniotomy.\textsuperscript{15}

Unilateral coronal synostosis produces a forehead that is typically bossed on one side and recessed on the other. In this case, a bifrontal craniotomy is required with reconstruction of the frontal bone. In particular, the bossed area needs to be recessed and reduced. An orbital rim advancement is also required.\textsuperscript{15}

Bilateral coronal synostosis produces a skull that is excessively tall and short. The surgery to correct this should produce a skull that is longer in the anterior-posterior dimension and shorter in the superior-inferior dimension. As with the unilateral coronal synostosis case, an orbital rim advancement is required.\textsuperscript{15}

POSITION STATEMENT

Applicable To: Medicaid

Refer to HS 009: Cranial Remodeling With Orthotic Devices for coverage information on protective, soft, or prefabricated helmets for children.

Surgery is recommended based on the age of the child, presence of other medical conditions, and caretaker preference. The following are considered medically necessary for craniosynostosis:\textsuperscript{15}

- **Traditional Surgery (“Calvarial Vault Remodeling”).** During surgery, an incision is made in the infant’s scalp. The shape of the head is corrected by moving the area that is abnormally fused or prematurely fused and then reshaping the skull so it can take more of a round contour. Surgery can last up to eight hours and requires one night in the intensive care unit (ICU) plus an additional 3-5 days in the hospital for monitoring. Surgery is ideal for babies beyond 6 months of age, because of expected blood loss. There is the possibility that a blood transfusion will be necessary during surgery. Temporary facial swelling may occur following surgery. Unlike other surgical options, there are no additional steps post-surgery unless a recurrence of craniosynostosis is found. Follow up with the surgeon within one month post-surgery to check incision site. Additional follow-up visits may occur at 6 and 12 months post-surgery.

- **Endoscopic Surgery (Minimally Invasive Surgery).** This minimally invasive surgery is performed when the infant is much younger (less than 3 months old). Using an endoscope, the surgeon can look through and see immediately inside and outside the skull through very small incisions in the scalp. The surgeon opens the suture to enable the baby’s brain to grow normally. The surgery takes approximately one hour and involves less blood loss compared to the calvarial vault remodeling leaving a small chance of requiring a blood transfusion. Infants typically require an overnight stay in the hospital prior to being released.
This type of surgery is followed by the use of a molding helmet. After the surgery is performed, a molding helmet is prescribed. The neurosurgeon should follow up with the member every 3 months for the first year post-surgery to check progress of the helmet reshaping the skull. (Additional appointments will be necessary with the helmet provider for fitting the helmet).

NOTE: Deformational plagiocephaly is not craniosynostosis and surgery is not required. It can result when the part of the skull (occipital bone) that is dependent (in one position) flattens out due to pressure, as when sleeping on that part of the skull.¹

Endoscopic surgery for primary craniosynostosis is typically performed by a pediatric neurosurgeon, pediatric plastic surgeon, or a pediatric surgeon.⁶

Endoscopic Strip Craniectomy and Spring-Mediated Cranioplasty are considered medically necessary as a form of surgical treatment to repair craniosynostosis.

Medical Necessity Criteria

Craniosynostosis is considered medically necessary based on the following:

1. **Child's age.**
   - The best time to intervene is between 3 and 9 months of age; infants with symptoms and signs of increased intracranial pressure require urgent decompression.³,¹³ Optimal time to perform surgery is before age 1 as the bones are still very soft and easy to work with. Surgery can sometimes be performed as early as 1 month of age. Early diagnosis and consultation with a specialist are important.³
   - Traditional procedures involve a form of open surgery to remove the fused suture and remodel the skull, using plates and screws as necessary; these are usually recommended in infants 6 to 8 months of age, are lengthy (4-8 hours in duration), and can require postoperative hospitalization for 4 to 8 days. Confirmation of diagnosis and pre-surgical evaluation requires a multidisciplinary team that may include geneticists, pediatric neurologists, plastic surgeons, neurosurgeons, endocrinologists, ophthalmologists.⁸

   NOTE: Surgery for craniosynostosis is considered urgent regardless of age, as long as medical necessity criteria are met.

2. **Overall health and medical history**³,¹³
   - Clinical history should include complications of pregnancy, duration of gestation, and birth weight.
   - Infant's sleeping position history (to differentiate craniosynostosis from plagiocephaly without synostosis)

3. **Radiographic evidence.** Plain radiography is the first step in the evaluation of suspected craniosynostosis and is sufficient for diagnosing single-suture craniosynostosis. Anteroposterior and lateral views of the skull are usual. Evaluate the entire length of each suture because only a small segment may be involved.⁴ The diagnostic value of the CT scan outweighs that of plain radiography because the sutures can be identified more accurately on a CT scan. CT scanning also aids in the evaluation of the brain for structural abnormalities (e.g., hydrocephalus, agenesis of the corpus callosum) and in excluding other causes of asymmetric vault growth (e.g., brain hemiatrophy, chronic subdural hematoma). Three-dimensional surface reconstruction using CT scanning can help the surgeon to accurately delineate the craniofacial deformity and plan surgical reconstruction.¹³

   Typically, each patient has early postoperative plain radiographs, a CT scan in the first few months after operation, and, in syndromic cases, MRI scans at yearly intervals to evaluate for the development of a hindbrain hernia. In patients with maxillary hypoplasia, yearly plain radiographs may be needed to help assess the progress of the deformity when the issue of possible midface advancement is considered.¹⁵

   Signs of craniosynostosis on plain radiography include bony bridging across the suture that produces beaking or heaping up of bone; sclerosis, straightening and narrowing of the suture; and loss of suture clarity.

4. **Diagnosis.** Member must have a diagnosis of **one** of the following (as described on p. 2):¹,³,¹³
   - Primary Craniosynotosis
• Secondary Craniosynostosis
• Simple Craniosynostosis
  o Sagittal or scaphocephaly
  o Coronal (bilateral)
  o Coronal (unilateral)
  o Coronal (anterior plagiocephaly)
  o Metopic (trigonocephaly)
  o Lambdoidal (bilateral, including posterior or occipital brachycephaly)
  o Lambdoidal (unilateral, including posterior or occipital plagiocephaly)
• Compound Craniosynostosis
  o Nonsyndromic (bicoronal or brachycephaly)
  o Syndromic
    ▪ Crouzon disease
    ▪ Apert syndrome
    ▪ Pfeiffer syndrome
    ▪ Saethre-Chotzen syndrome
    ▪ Brachycephaly
    ▪ Carpenter syndrome
    ▪ Cloverleaf skull
    ▪ Baller-Gerold syndrome
    ▪ Antley-Bixler syndrome

NOTE: Deformational plagiocephaly is not craniosynostosis and surgery is not required. It can result when the part of the skull (occipital bone) that is dependent (in one position) flattens out due to pressure, as when sleeping on that part of the skull.\(^3\)

5. **Extent of the craniosynostosis** is to be determined via radiographic evidence (e.g., CCT and/or x-rays).

*Ongoing Treatment and Evaluation*

Long-term care and surveillance by an integrated neurosurgical and craniofacial multidisciplinary team is essential.\(^1\) A child with craniosynostosis requires frequent medical evaluations to ensure that the skull, facial bones, and brain are developing normally. The medical team works with the child's family to provide education and guidance to improve the health and well-being of the child.\(^3\)

*Orthotic Devices*

Cranial remodeling with orthotic devices (helmets or bands) for members with moderate to severe positional (nonsynostotic) plagiocephaly is considered medically necessary when ALL of the following criteria are met:

1. Positional plagiocephaly is associated with ANY of the following:
   - Premature birth; OR,
   - Restrictive intrauterine positioning; OR,
   - Cervical abnormalities; OR,
   - Birth trauma; OR,
   - Torticollis (shortening of the sternocleidomastoid muscle); OR,
   - Sleeping positions

2. Remodeling is initiated at 4-12 months of age; AND,

3. A 2-month trial of conservative therapy (repositioning of the head such that the child lies opposite to the preferred position) has failed to improve the deformity and is judged unlikely to do so; AND,

4. There is documentation of EITHER of the following criteria*:
   - Cephalic index ± at least two standard deviations from the mean for the appropriate gender/age; OR,
• Asymmetry of 6 mm or more in ONE of the following measures:
  o Cranial vault; **OR**,
  o Skull base; **OR**,
  o Orbitotragial depth

### COVERED ICD-10-CM DIAGNOSIS CODES

- Q67.0 Congenital facial asymmetry
- Q67.1 Congenital compression facies
- Q67.2 Dolichocephaly
- Q67.3 Plagiocephaly
- Q67.4 Other congenital deformities of skull, face and jaw
- Q75.0 Craniosynostosis
- Q75.1 Craniofacial dysostosis
- Q75.2 Hypertelorism
- Q75.3 Macrocephaly
- Q75.8 Other specified congenital malformations of skull and face bones
- Q75.9 Congenital malformation of skull and face bones, unspecified

### REFERENCES


MEDICAL POLICY COMMITTEE HISTORY AND REVISIONS

<table>
<thead>
<tr>
<th>Date</th>
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<tbody>
<tr>
<td>7/6/2017, 9/27/2016</td>
<td>• Approved by MPC. No changes.</td>
</tr>
<tr>
<td>8/6/2015</td>
<td>• Approved by MPC. Clarified language re: coverage for members regardless of age when criteria met.</td>
</tr>
<tr>
<td>11/6/2014</td>
<td>• Approved by MPC. New.</td>
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