Imaging of Craniosynostosis from Diagnosis through Reconstruction

Hesham Attaya, MD, Joel Thomas, MD, and Anthony Alleman, MD, MPH

ABSTRACT
BACKGROUND AND PURPOSE: Imaging plays a critical role in the diagnosis and management of craniosynostosis. The purpose of this article is to present a concise pictorial reference of the imaging features of the major primary craniosynostoses, emphasizing diagnostic criteria, operative repair, and postoperative follow-up imaging.

MATERIALS AND METHODS: The imaging system of a referral center with an active craniofacial team was queried to find images that best represent the classic imaging findings in metopic, sagittal, coronal (unilateral and bilateral), and lambdoid craniosynostosis. 2D and 3D-CT images of preoperative, postoperative, and delayed follow-up were selected. A review of the craniofacial imaging literature was performed to extract normative data for sutural closure and imaging features relevant to the craniofacial team.

RESULTS: This article concisely presents the imaging features relevant to the craniofacial team caring for the patient with craniosynostosis. Imaging protocols are provided. Criteria for diagnosis, including normative data for sutural closure, are summarized, and systematic evaluation of craniosynostosis imaging in the pre- and postoperative period is emphasized.

CONCLUSIONS: Primary craniosynostosis imaging is critical to the diagnosis and exclusion of craniosynostosis in the patient with abnormal calvarial shape. In the perioperative and long-term follow-up of these patients, imaging is used to detect complications and guide further interventions.

INTRODUCTION
Craniosynostosis is a disorder of skull development involving premature fusion of ≥1 of the cranial sutures and occurs in approximately 1 of 1800 live births. Normal closure times for the cranial sutures and fontanels are variable. The metopic suture starts closing as early as 3 months and is usually closed by 9 months, but it may persist into adulthood. Complete sutural closure may continue into the fourth decade of life and beyond. As a general rule, the sagittal, coronal, and lambdoid sutures should not be closed within the first year of life.

The original classification scheme devised by Virchow in 1851 proposed that an abnormality of a suture results in fusion and reduction in growth perpendicular to the plane of the suture. In general, the craniosynostoses are currently classified as either primary or secondary. The primary craniosynostoses are further classified as simple (involving a single suture) or compound (involving ≥2 sutures). The primary major suture synostoses include sagittal, metopic, coronal, and lambdoid synostoses, all of which are nonsyndromic (Fig 1). The compound synostoses include bicoronal synostosis, a nonsyndromic entity, and the syndromic synostoses, of which Crouzon and Apert syndromes are by far the most common. Fifteen percent of individuals with craniosynostosis present with syn
dromic conditions and are more severely affected than those with single-suture synostosis.

The most common genetic mutations identified in syndromic craniosynostosis involve the **FGFR** family, which influences growth of the brain and connective tissue. A mutation in 1 of these genes causes the sutures to fuse in most forms of craniosynostosis. Other genes such as transcription factors **TWIST**, **MSX2**, and **GLI3** have been associated with syndromic craniosynostosis. Secondary causes of craniosynostosis are multiple and include metabolic derangements, malformations in brain development, fetal drug exposure, and shunted hydrocephalus, among others.

In the infant with a misshapen skull, craniosynostosis must be differentiated from benign deformational plagiocephaly, also called “positional plagiocephaly,” “posterior plagiocephaly,” or “plagiocephaly without synostosis.” Benign deformational plagiocephaly is most often treated with multiple positioning techniques, including alterations to the position in which an infant is placed for sleeping, or sometimes a skull-molding helmet is prescribed for mechanical correction of the deformity. By contrast, the various craniosynostoses require surgical intervention for correction. While clinical diagnosis of the various types of cranial vault deformity can be made by physical examination and observation, imaging plays a vital role in the definitive diagnosis and in surgical and reconstruction planning.

Symptoms resulting from craniosynostosis are either caused directly by pathologic growth pattern or indirectly as a result of increased intracranial pressure. Because craniosynostosis has been associated with abnormalities in brain growth and development, early recognition not only allows correction of skull shape abnormalities but also may prevent neurologic sequelae. Abnormal head shape is, therefore, a common reason for referral to a pediatric neurosurgeon. Table 1 illustrates the characteristics and the physical appearance seen with the most common craniosynostosis types.

### Diagnosis and Imaging

The diagnosis of craniosynostosis is based on clinical features such as reduction in skull size parallel to the fused suture, skull lengthening perpendicular to the fused suture, and “ridging” of the skull along the fused suture. Growth restriction at a stenosed suture leads to compensatory exaggeration of growth at the open sutures.

Fetal US and MR imaging documentation of intrauterine craniosynostosis is possible during and after the second trimester. While conventional radiology may demonstrate sutural fusion, radiographic findings are often unreliable in the first 3 months of life. On CT, synostosis of each of the cranial sutures demonstrates characteristic presentations. Sagittal suture craniosynostosis may present with thickened bony ridges, metopic suture craniosynostosis is likely to demonstrate focal bone thickening and erosions, and lambdoid suture craniosynostosis presents with perisutural sclerosis. While plain radiographs may allow diagnosis of the major sutural craniosynostoses, axial and 3D-CT are best used to diagnose and confirm the major and minor craniosynostoses. 3D- and helical CT have improved accuracy in diagnosis to rates of 90%–100%. CT permits simultaneous evalu-

### Table 1—Terms for Skull Deformities.

<table>
<thead>
<tr>
<th>Term</th>
<th>Definition</th>
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<tr>
<td>Scaphocephaly dolichocephaly</td>
<td>“Inverted boat” shape or long narrow head with decreased width of the skull,</td>
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<tr>
<td></td>
<td>typical of sagittal synostosis</td>
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<tr>
<td>Trigonocephaly</td>
<td>Triangular-shaped forehead seen with metopic synostosis</td>
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<tr>
<td>Brachycephaly</td>
<td>Decreased AP diameter and increased width of the skull, typical of bicoronal</td>
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<tr>
<td></td>
<td>synostosis</td>
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<tr>
<td>Plagiocephaly</td>
<td>Asymmetry or flattening of 1 side of the skull; may be seen with unilateral</td>
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<tr>
<td></td>
<td>sutural fusion as in unilateral coronal or lambdoid as well as with benign</td>
</tr>
<tr>
<td></td>
<td>positional plagiocephaly, which is not associated with sutural fusion</td>
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<tr>
<td>Harlequin orbit deformity</td>
<td>Abnormal morphology of the orbit caused by superior elevation of the lesser</td>
</tr>
<tr>
<td></td>
<td>wing of the sphenoid bone resulting in proptosis and exophthalmos; this is</td>
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<tr>
<td></td>
<td>the most consistent imaging feature of unilateral coronal synostosis</td>
</tr>
<tr>
<td>Hypotelorism</td>
<td>Abnormally decreased distance between the orbits, typical of metopic synostos</td>
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http://dx.doi.org/10.3174/ng.3110013
ation of brain growth and delineation of craniofacial deformities and ventricular volumes. Additionally, CT can exclude other causes of asymmetric cranial vault growth, such as brain hemiatrophy, and can facilitate planning of surgical correction. Frequently MR imaging is used to define patients with intracranial anatomy at risk of acquired tonsillar herniation or central nervous system anomalies.11

The protocol for patients with craniosynostosis at our institution calls for supine helical 2.5-mm images in the axial plane from the inferior mandible through the entire brain and reconstruction in the axial plane at 0.625 mm with 0% overlap in the bone algorithm. The image dataset is then used by the CT technologists to generate 3D images in both surface-rendering and maximum intensity projection as single snapshots. The study is performed with a low milliampere–second setting adjusted for patient age and size, typically from 0 to 100 mAs for the 3D image acquisition. A smoothing algorithm and brain windowing are applied by the radiologist at the workstation to screen the intracranial structures. Our protocol is within Image Gently guidelines (Fig 2).

**TYPES OF CRANIOSYNOSTOSIS**

**Sagittal Synostosis**

Sagittal synostosis is the most common form of craniosynostosis representing up to 40%–60% of all craniosynostoses with a male predominance of 4:1.12,13 Approximately 80% of sagittal synostosis is nonsyndromic, with familial cases representing 6%. Twinning, increased parity, maternal smoking, uterine malformations, and intrauterine head constraint have been suggested as risk factors.14 Growth inhibition perpendicular to the affected suture leads to scaphocephaly or dolichocephaly. Growth at the patent coronal and metopic sutures leads to anterior inclination, while growth at the lambdoid suture leads to occipital bulging. Thus, patients present with an elongated cranium, flat vertex, and increased head circumference (Fig 3).15

**Radiologic Features**

Plain radiographs demonstrate ossification of the sagittal suture and anterior fontanel. The lateral view demonstrates skull elongation in the AP direction with abnormal prominence of the occiput and frontal tuberosity.16 CT demonstrates prominence of the occipital and frontal tuberosity as well as a narrowed biparietal diameter.9

**Coronal Synostosis**

Unilateral or bilateral fusion of the coronal suture is the second most common form of craniosynostosis and accounts for 20%–30% of all craniosynostosis cases.12 Unilateral coronal synostosis in infancy is considered sporadic and nonsyndromic (Fig 4), while bilateral coronal synostosis is commonly syndromic (Fig 5). Unilateral coronal synostosis is more commonly seen in females with the right side having a greater frequency than the left.13 Patients with unilateral coronal synostosis present with a palpable suture, ipsilateral exophthalmos, uplifting of the ipsilateral eyebrow, and flattening of the frontal bone, resulting in anterior plagiocephaly. Patients with bilateral coronal synosto-
sis have symmetric bone growth along the sagittal and metopic sutures, resulting in brachycephaly.17

Radiologic Features
Plain radiographs demonstrate absence of normal coronal sutural radiolucency on the affected side with deviation of the nasal root. The most consistent imaging feature of coronal synostosis, the harlequin deformity of the orbit, occurs because of superior elevation of the lesser wing of the sphenoid with resultant proptosis and exophthalmos, which can be severe. Anterior flattening of the ipsilateral forehead and orbit and compensatory bulging of the contralateral side is a common presentation leading to anterior plagiocephaly. Involvement of the nasomaxillary suture leads to deviation of the nasal root to the side of the stenosed suture.18

Metopic Synostosis
Premature closure of the metopic suture produces a keel-shaped triangular frontal deformity known as trigonocephaly. Metopic synostosis comprises <10% of craniosynostosis overall, with the suture normally closing around 9 months to 2 years of age.12 Although most of the cases arise spontaneously, several syndromes and associated malformations have been described. Genetic testing is warranted when an infant presents with this defect.19 Males greatly outnumber females with a ratio of slightly less than 3:1.13

Radiologic Features
The imaging hallmark of early metopic synostosis is trigonocephaly. The skull base narrows as fusion starts at the glabella. As a result, features of metopic synostosis include hypoplastic ethmoidal sinuses, hypotelorism, and anterior bowing of the coronal sutures. Evaluation of the metopic suture is possible after 9 weeks and during the second trimester on fetal US due to the expansion of the frontal bone ossification centers. Fetal US readily identifies early metopic suture closure or the presence of an additional large midline frontal bone (Fig 6).11
Lambdoid Synostosis
Lambdoid craniosynostosis may be clinically mistaken for deformational plagiocephaly. The incidence of deformational plagiocephaly is approximately 1 in 300 live births compared with lambdoid synostosis, which occurs in 3 in 100,000 live births.²⁰²¹ Plagiocephaly has been associated with intrauterine pelvic constraints, repetitive sleep positioning, and myoneural dysfunction. More common since the recommendation that infants sleep on their backs, deformational plagiocephaly is not associated with sutural fusion and infrequently requires surgical correction.²² Repositioning therapy and cranial orthoses are the 2 main nonsurgical treatment options for deformational plagiocephaly. Repositioning therapy includes positioning the child in the prone position while the child is awake and alternating sleep positions, thus limiting the time in which the patient is in the supine position.²³²⁴ An additional option is a cranial orthosis, which is a helmet used between the ages 6 and 18 months, which creates a round surface over the flattened skull. Constant pressure of the growing brain against the concave surface helps correct the flattening.²⁵²⁶

Radiologic Features
On plain films, obliteration of the lambdoid suture and compensatory contralateral parietal and frontal bossing is observed in cases of lambdoid suture craniosynostosis. Fusion of the lambdoid suture leads to ipsilateral occipital flattening and enlargement of the ipsilateral mastoid process. In lambdoid suture synostosis, the ipsilateral ear is displaced posteriorly toward the fused suture (trapezoidal shape) (Fig 7). Deformational plagiocephaly is diagnosed by ipsilateral occipital bone flattening and contralateral frontal flattening (parallelogram shape) (Fig 8). In unilateral lambdoid craniosynostosis, the angle formed between lines drawn through the midlines of the anterior and posterior fossae is >7°. Additionally, sutural fusion is demonstrated in the axial and 3D-CT images in lambdoid synostosis. This is statistically significant compared with that observed in posterior plagiocephaly resulting from postural molding, which is <7°.²⁷²⁸

Multiple Sutural Craniosynostosis
Although bilateral coronal craniosynostosis is the most common of the multiple sutural craniosynostoses, multiple major and minor sutures may fuse prematurely. Multiple suture synostosis is more frequently syndromic, with Crouzon and Apert (FGFR gene mutations) being the most common (Fig 9). Children with syndromic craniosynostoses demonstrate multiple cranial facial abnormalities. The growth progresses forward toward the forehead with the parietooccipital areas having the least growth. The orbits typically have a short
roof leading to exophthalmos and proptosis. The maxilla fails to progress downward and forward, and this type of growth can lead to airway compromise and nasopharyngeal constriction. Although Crouzon, Apert, and Pfeiffer syndromes are the most common, >180 syndromes involving craniosynostosis have been identified.29,30

Just as the major cranial sutures can fuse prematurely, minor sutures can similarly fuse prematurely as well. Abnormal fusion of the minor sutures may be symmetric or asymmetric. Premature fusion of the skull base sutures can lead to significant changes in the craniofacial axis of a child. Minor sutures include the squamosal sutures and the synchondroses of the mid and posterior skull base: paired sphenoccipital, anterior intraoccipital, posterior intraoccipital, petrooccipital, and occipitomastoid sutures.31 Craniosynostosis may also be secondary to metabolic abnormalities such as rickets, bone dysplasias, and effects of fetal teratogens, such as hydantoin. Secondary sutural closure may occur after ventriculoperitoneal shunt surgery, which reduces the expansile forces on the calvaria (Fig 10).

**Reconstruction**

The most worrisome complication of the craniosynostoses that may lead to abnormal neurologic development is increased ICP. The greater the number of sutures prematurely fused, the greater likelihood of increased ICP. The incidence of intracranial hypertension doubles after the first year of life.32,33 Early surgical correction of craniosynostosis is performed in the hopes of limiting or preventing neurodevelopmental sequelae of craniosynostosis and establishing a more aesthetic cranial contour.

Additionally, the bones are also more malleable in infancy; therefore, early surgical repair is preferred, typically within the first 3–12 months of life. The basic principles for surgical correction of craniosynostosis include re-establishing sutural patency, allowing growth and development, and restoring symmetry. Surgical correction, usually performed

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Fig 8. In contrast to craniosynostosis, deformational plagiocephaly in a 6-month-old boy is not associated with suture fusion. Anterior (A), superior (B), posterior (C) 3D renderings, and axial CT (D).

Fig 9. An 8-month-old girl with Crouzon syndrome (a FGFR2 mutation). Anterior (A), posterior (B), superior (C), and left lateral (D) 3D renderings demonstrate multiple suture synostosis, including metopic, inferior portion of the coronals and the squamosal. Patients with Crouzon syndrome commonly demonstrate midfacial hypoplasia, proptosis, beaked nose, and cleft palate.

Fig 10. A 5-month-old girl with a ventricular-peritoneal shunt placed at 3 days of age secondary to intracranial hydrocephalus. Anterior (A), right lateral (B), left lateral (C), and posterior (D) 3D renderings demonstrate multiple sutural craniosynostosis. The patient had sagittal craniosynostosis at birth.
as a cooperative procedure by pediatric neurosurgical and craniofacial plastic surgery teams, involves a combination of craniectomy, or removal of the abnormally fused suture, and cranial vault remodeling.

The procedures currently used to correct sagittal synostosis include strip craniectomy and cranial vault remodeling. Cranial vault remodeling involves removal of the frontal, parietal, and occipital bones; radial osteotomies; and surgical molding before replacement of the skull. The bones are fixed in place with absorbable plates and screws. Strip craniectomy involves removing just a narrow strip of bone along both sides of the sagittal suture without bone remodeling or replacement. Newer endoscopic-assisted techniques for strip craniectomy have also been developed. Distraction osteogenesis of cranial strip osteotomies may also be used. Differences in opinion exist between some neurosurgeons and plastic surgeons regarding the merits of the individual procedures.

Coronal synostosis reconstruction involves fronto-orbital advancement, with bifrontal craniotomy, removal of the fronto-orbital bars, remodeling of the orbits to obtain symmetry, replacing and advancing the fronto-orbital bars, and molding the frontal bones before reattachment. Absorbable fixation plates, bone grafts, and absorbable sutures are also used (Fig 11). Metopic synostosis is corrected in a similar fashion; however, the metopic suture is also excised (Fig 12).

Lambdoid synostosis reconstruction involves excision of the fused lambdoid suture, including the ipsilateral asterion, to the level of the sagittal junction. Newer techniques involve cranial vault remodeling with parietooccipital bone rotational flaps.

Imaging in the early postoperative period may be required to detect complications (hemorrhage, intracranial migration of hardware, and so forth.). Imaging performed weeks to months postoperatively may be used to assess cranioplasty patency or closure and calvarial shape and to rule out complications such as hardware migration or hydrocephalus.

REFERENCES


