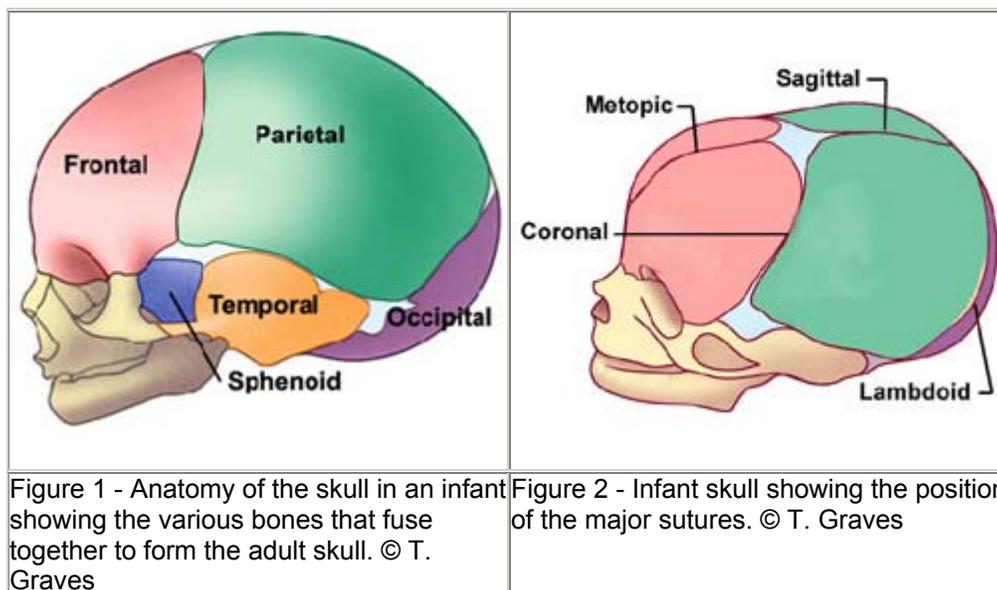


Craniosynostosis (sometimes called craniostenosis) is a disorder in which there is early fusion of the sutures of the skull in childhood. It produces an abnormally shaped head and, at times, appearance of the face. The deformity varies significantly depending on the suture or sutures involved. Surgical correction may be necessary to improve appearance and provide space for the growing brain.

Anatomy

- The bones of the skull are the frontal, parietal, temporal, occipital, and sphenoid bones (Figure 1)
- The place where two bones come together is called a suture
- At birth the adjacent bones override each other to allow the infant to pass through the birth canal. hinges called sutures, which allow the head to pass through the birth canal As the child grows the sutures allow for skull expansion to accommodate the growing brain.
- The brain doubles in size by age 6 months and again at age 2 years. These sutures normally begin to fuse around 2 years of age along with the closure of the fontanel (soft spot)
- The seam where two skull bones fuse together is called a suture. The major sutures are called sagittal, coronal, metopic and lambdoid (Figure 2)
- The sutures are completely fused by 6-8 yrs of age
- Within the skull lie:
 1. Brain
 2. Meninges
 - The dura is a membrane that lines the inside of the skull
 - The pia-arachnoid is a filmy membrane that covers the brain
 3. Blood vessels of the brain
 4. Cerebrospinal fluid (CSF), a watery fluid that is produced by and bathes the brain



Pathology

- Craniosynostosis occurs when one or more of the major sutures close early
- The cause is unknown

- It is called 'primary' when not associated with any other problem. When there is an underlying disorder it is considered 'secondary'
- It may be "simple" with only one suture closed or "compound" when multiple sutures are involved
- The result of craniosynostosis is to change the shape of the head and sometimes face
- The appearance depends on which suture or sutures close early. When a suture closes early the skull growth in that area becomes restricted. Since the growing brain needs more space there is a 'compensatory' change in the skull as the skull is pushed out in areas where the sutures are still open. The types of single suture closure are:
 1. Sagittal synostosis (Figure 3) is caused by early closure of the sagittal suture synostosis. It is the most common type of synostosis and it creates an elongated narrow skull called 'scaphocephaly' or 'dolicocephaly'. The compensatory change causes the front (frontal) and back (occipital) areas of the skull to be pushed outward causing bossing (bulging of the skull)



Figure 3 - Child with sagittal synostosis. Note that the head is elongated from front to back. Courtesy S. Schneider, MD

Figure 3a - Three dimensional CT of the skull of an infant with sagittal synostosis. Note that the sagittal suture is elongated between the coronal and lambdoid sutures. Courtesy S. Schneider, MD

2. Coronal synostosis (Figure 4) is caused by early closure of the coronal suture. It creates a flattening of forehead on that side called 'anterior plagiocephaly' (front skull flattening). The eye socket or orbit on that side may be misaligned and brought backward. The opposite side of the forehead may be pushed outward causing a compensatory bossing. Both coronal sutures may be closed causing a flattening of both sides of the forehead called anterior 'brachycephaly' and is often syndromic



Figure 4 - Infant with left-sided coronal synostosis. Note that the left forehead is flattened and set back and the left eyebrow is raised. Courtesy S. Schneider, MD

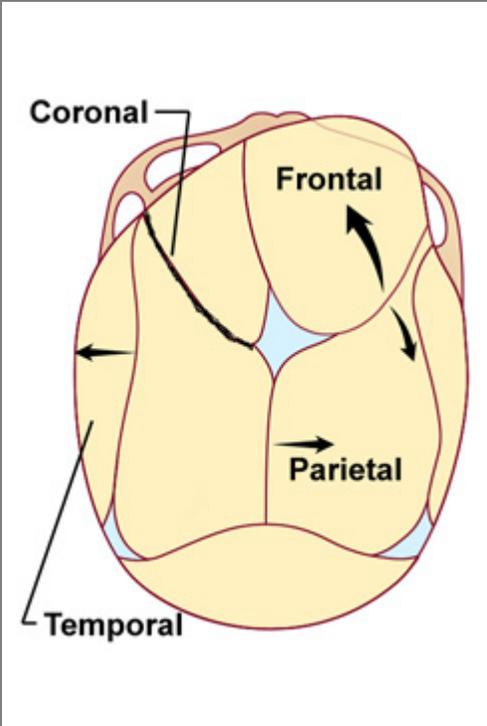
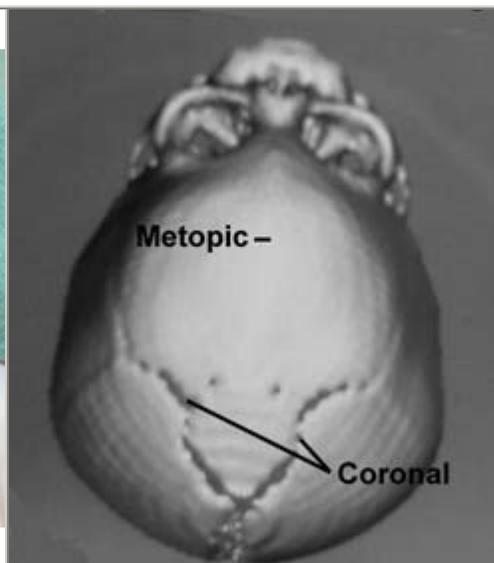


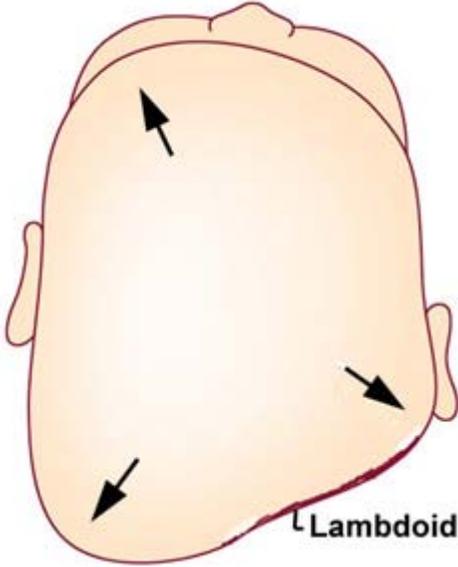
Figure 4a - Left coronal synostosis as seen from above. Note the flattening of the left forehead. The arrows indicate the direction of growth of the skull to compensate for the synostosis. © T. Graves

3. Metopic synostosis (Figure 5) is caused by early closure of the metopic suture. It creates a triangular, keel shaped and pointed forehead called 'trigonocephaly'. The eyes may appear closer together (hypoteleorism) and the sides of the head may bulge outward in a compensatory bossing of both parietal bones



<p>Figure 5 - Infant with metopic synostosis as seen from above. Note that the forehead has a keel shape. Courtesy S. Schneider, MD</p>	<p>Figure 5a - 3-D CT of an infant with metopic synostosis. Note the keel shape of the forehead and the elongated closed metopic suture. Courtesy S. Schneider, MD</p>
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4. Lambdoid synostosis (Figure 6) is caused by early closure of the lambdoid suture. It creates a flattening of the back of the head called 'posterior plagiocephaly' (back skull flattening). The ear and forehead on that side may be displaced backwards. Bilateral (both) sutures may be closed causing a flattening of entire back of the head called 'posterior brachycephaly'. Lambdoid synostosis may be confused with positional molding

	
<p>Figure 6 - Infant with lambdoid synostosis seen from behind. Note the flattened area behind the right ear. Courtesy S. Schneider, MD.</p>	<p>Figure 6a - Lambdoid synostosis as seen from above. Note that the forehead and ear are pushed back on the same side as the synostosis. The arrows indicate the direction of the skull change. © T. Graves</p>

- Positional molding (Figure7) is caused by the infant constantly lying with one side of the back of the head on a flat surface. It is also called 'deformational plagiocephaly'. It looks similar to lambdoid synostosis with the ear on the affected side usually pushed forward although the suture remains open. Unlike lambdoid synostosis, the compensatory frontal bulging of the skull is on the same side as the flattening. Both sides can be involved as in lambdoid synostosis. It may be associated with tilting of the head or torticollis

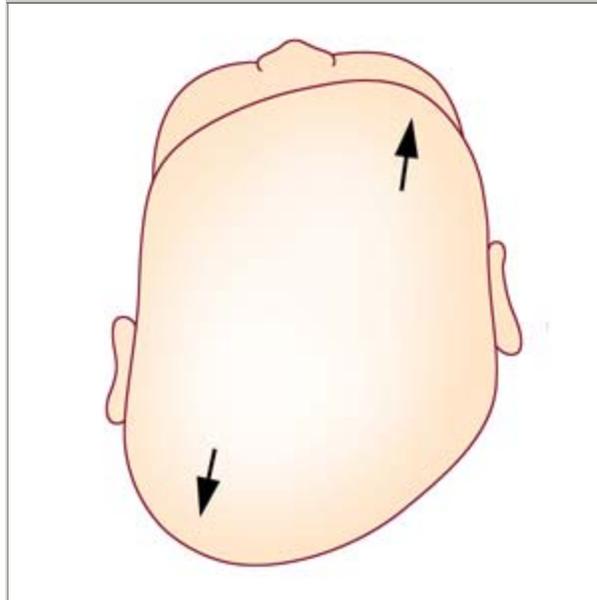


Figure 7 - Positional deformation of the right lambdoid area. The forehead and ear are displaced forward on the same side. The arrows indicate the direction of the skull change. © T. Graves

- When only one suture is involved the brain usually functions normally. More severe forms of craniosynostosis may be associated with closure of multiple sutures and/or abnormal brain formation and are called 'syndromic'
- These disorders tend to run in families. This occurs rarely when only one suture is involved, but more commonly when multiple sutures are affected. There may be associated malformations of the face, limbs and other organs

Common Craniosynostosis Syndromes			
Name	Sutures involved	Common Associated manifestations	Genetics
Apert	Coronal, Sagittal, Lambdoid	Midface deficiency, Hyperteleorism, Malformations of the hands and feet	Autosomal dominant
Crouzan	Coronal, Sagittal	Midface deficiency, Hyperteleorism	Autosomal Dominant
Carpenter	All	Flat nasal bridge, mental retardation, polysyndactyly	Autosomal Recessive
Pfeiffer	Multiple	Hyperteleorism, Midface deficiency, Limb, heart and	Autosomal Dominant

		bowel malformations	
Seathre-Chotzen	Coronal	Malformations of fingers & hands	Autosomal Dominant

- Associated Problems - Craniosynostosis can be associated with other neurologic problems including hydrocephalus, Chiari I malformation and increased intracranial pressure. These are more often associated with multiple suture involvement and syndromic cases
 1. Hydrocephalus is a build up of CSF that may require the insertion of a shunt, which diverts the fluid to the abdomen where it is absorbed in the blood ([See Shunt for Hydrocephalus](#))
 2. Chiari I malformation is the displacement the lower part of the brain (cerebellar tonsils) into the upper cervical (neck) spinal canal causing pressure. Removal of bone (craniectomy - [See Craniotomy](#)) may be necessary to relieve the pressure
 3. Increased intracranial pressure is an abnormal build up of brain pressure that may occur in some cases of craniosynostosis. This pressure can be transmitted to the eyes and lead to loss of vision unless treated. Emergent surgery may be required

History and Exam

- The prenatal (prior to birth), delivery and postnatal (after birth) history are reviewed especially with regards to exposure to smoking and certain medications such as thyroid medication or methotrexate
- Time of onset of the deformity and whether or not it is worsening or improving is evaluated. Most synostoses are seen at birth or shortly thereafter and progress with time
- Deformities that are not apparent at birth and progress together with a positional preference suggest the possibility of positional molding. Sleep position preference and its relation to the deformity may be important
- The child's developmental history and attainment of milestones may be reviewed to assess possible delays
- Family history of a neurologic problems as well as abnormal head shapes suggests a possible familial syndrome
- The examination usually reveals the deformity associated with the suture or sutures closed (see above). Frequently ridges may be felt over a closed suture and at times the soft spot may also close early
- The head circumference is measured and charted in relation to the child's age
- The eyes are examined for signs of increased pressure in the head and the neurologic examination may document other associated problems
- The remainder of the physical examination is performed with special attention to the hands and feet as well as arms and legs looking for deformities
- Consultations may be obtained from specialists in genetics, ophthalmology and neurology

Tests

- Skull x-rays show which sutures are closed
- Three dimensional computerized axial tomography (CAT scan) is helpful in determining which sutures are involved and to what degree (Figure 8). The brain may also be evaluated and any brain abnormality determined
- Magnetic resonance imaging (MRI) is best for evaluating the brain and is useful for brain malformations including the Chiari type

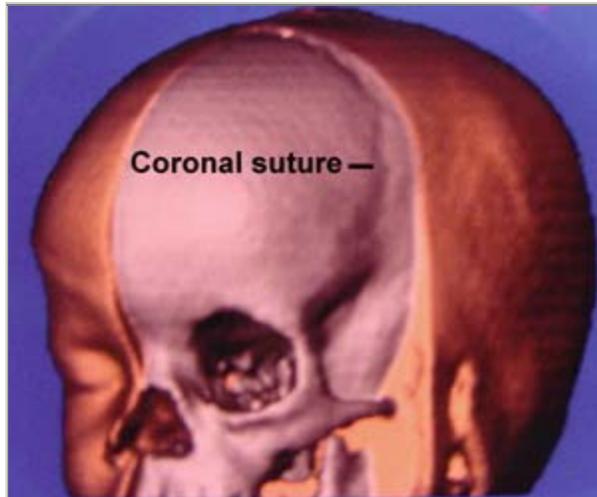


Figure 8 - 3-D CT scan showing the coronal suture and overlying skin. Courtesy S. Schneider, MD

Indications/Contraindications

- In cases of single suture synostosis correction of the deformity and/or progressive deformity are the primary indications for surgery
- The age at time of operation varies depending on the suture involved and the degree of deformity
- Occasionally there are signs of increased intracranial pressure which requires surgery
- In cases of multiple synostosis preservation and/or restoration of neurologic function may require surgery
- Surgery for Chiari decompression or insertion of a shunt for hydrocephalus may be necessary
- Not all cases require surgery. Mild metopic synostosis and deformational plagiocephaly are not considered surgical unless of a severe nature that does not improve with time
- Certain medical conditions such as infection or other problem may require delay or even preclude surgery

Surgical Procedures

- All children are screened to ensure they are medically fit for surgery
- The procedure is a craniotomy or craniectomy for craniofacial reconstruction. The craniotomy involves the surgical removal of sections of bone (bone flap) including the involved suture, which are rearranged and replaced. If the bone is not replaced it is called a craniectomy
- Many different and varied procedures have been performed for craniosynostosis depending on the age, suture and pattern of involvement
- The child is placed under anesthesia ([See Anesthesia](#)) with the insertion of intravenous lines and catheter in the bladder
- The incision is dependent on the suture involved and the preference of the surgeon
- The scalp is lifted and the skull exposed for reconstruction. Burr holes are placed to allow a bone window to be created ([See Craniotomy](#))
- The bones may be reconstructed with sutures, wire or plates
- In the areas where bone has been removed the body restores the bone with time
- Blood transfusion is frequently necessary and blood is kept available for possible transfusion ([See Blood Transfusion](#))

- Sagittal synostosis
 1. Strip (sagittal syndectomy) or vertex craniectomy for sagittal synostosis. The fused sagittal suture is removed. There are several variations depending on the amount of bone removed. Additional cuts in the skull (osteotomies) may be carried out to allow the skull to be immediately reshaped in the older child. The bone re-grows as the skull is reshaped. (Figure 9)
 2. Pi Procedure for sagittal synostosis. A section of skull is removed shaped like the Greek letter pi (μ). There are variations of the procedure depending on the type of compensatory changes that have occurred. The procedure allows for immediate improvement (Figure 10)
 3. Endoscopic or endoscope -assisted procedure. The procedures may be combined with an endoscope (a lighted tube and lens system through which instruments may be passed) to perform cuts in the bone through smaller incisions with less blood loss. Usually the children require a remodeling helmet to be worn following the operation to allow the reshaping process to continue (Figure 11)

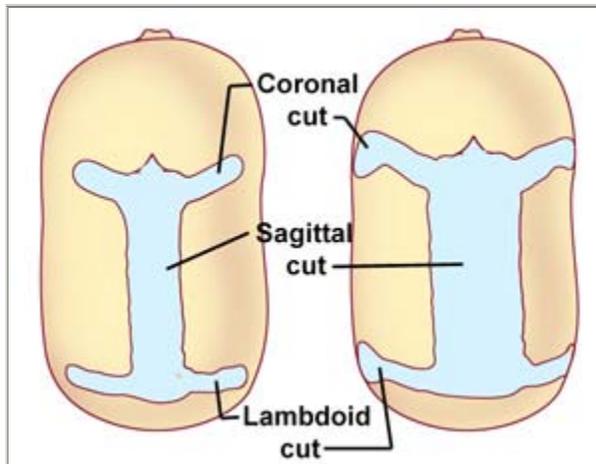


Figure 9 - Two variations of strip craniectomies for sagittal synostosis. Part of the coronal and lambdoid sutures may also be removed. © T. Graves

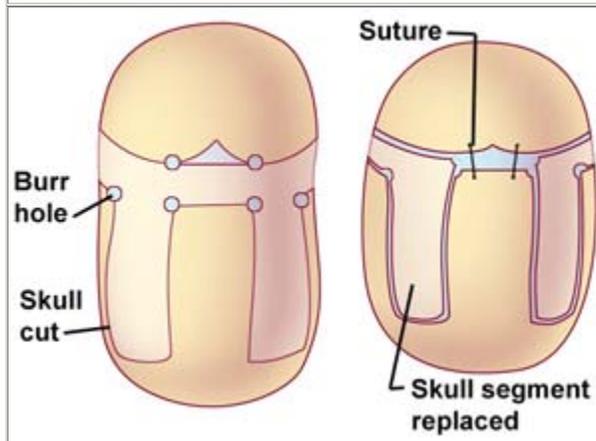


Figure 10 - Pi procedure for sagittal synostosis. Large sections of bone are removed on both sides of the sagittal suture. Once removed the skull is remodeled by drawing the center section of bone

forward. The results in decreasing the front to back diameter of the head and spreads the head in the side to side direction. © T. Graves

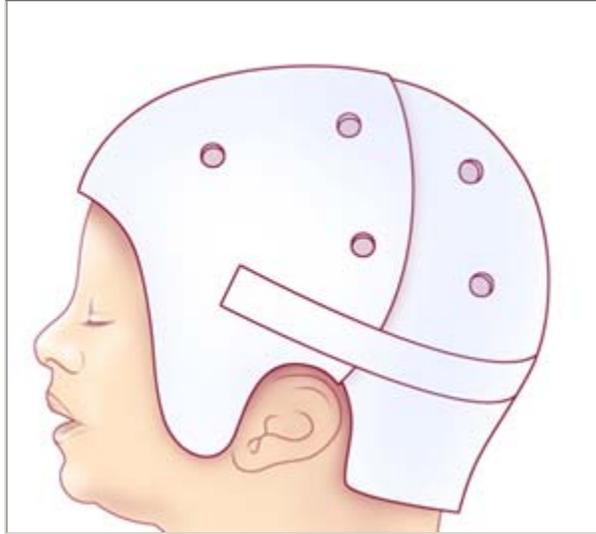


Figure 11 - Helmet for remolding the skull. The helmet is form fitted and readjusted as the head grows. © T. Graves

- Coronal synostosis
 1. Strip craniectomy for coronal synostosis (coronal syndectomy). In the very young patient the fused suture alone may be removed. Ensuing brain growth allows the skull to be remodeled
 2. Fronto-orbital advancement for coronal synostosis. The bones of the forehead are separated from the skull as well as those around the eye socket and are advanced forward (Figure 12). Depending on the age and degree of deformity one or both side can be reconstructed
 3. If there is associated hypo or hypertelorism (eyes closer together or farther apart), the eye sockets may need to be moved farther apart or closer together (Figure13)

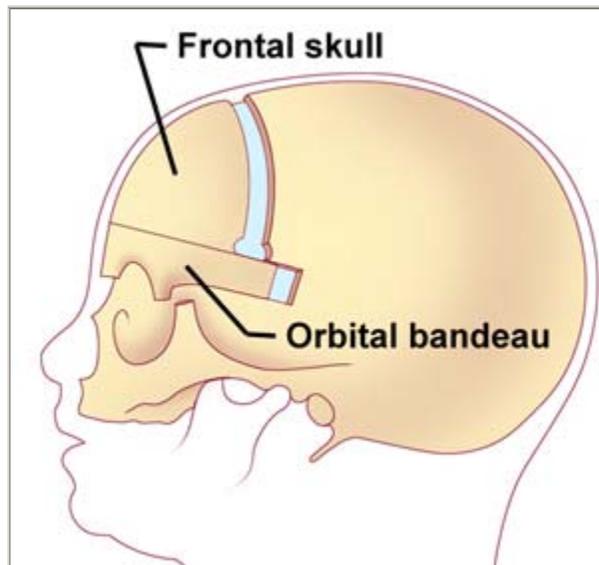


Figure 12 - Fronto-orbital advancement for coronal synostosis. The bones of the forehead are separated from the skull as well as a band of bone that includes the upper orbit (orbital bandeau) and are advanced forward. © T. Graves

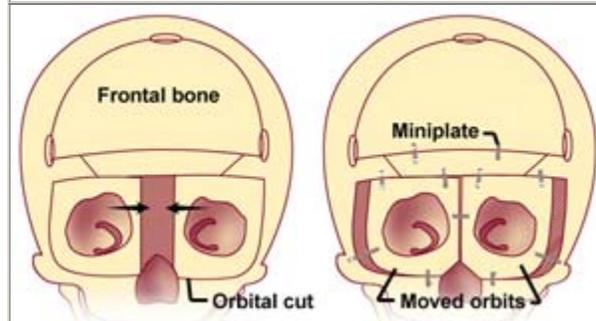
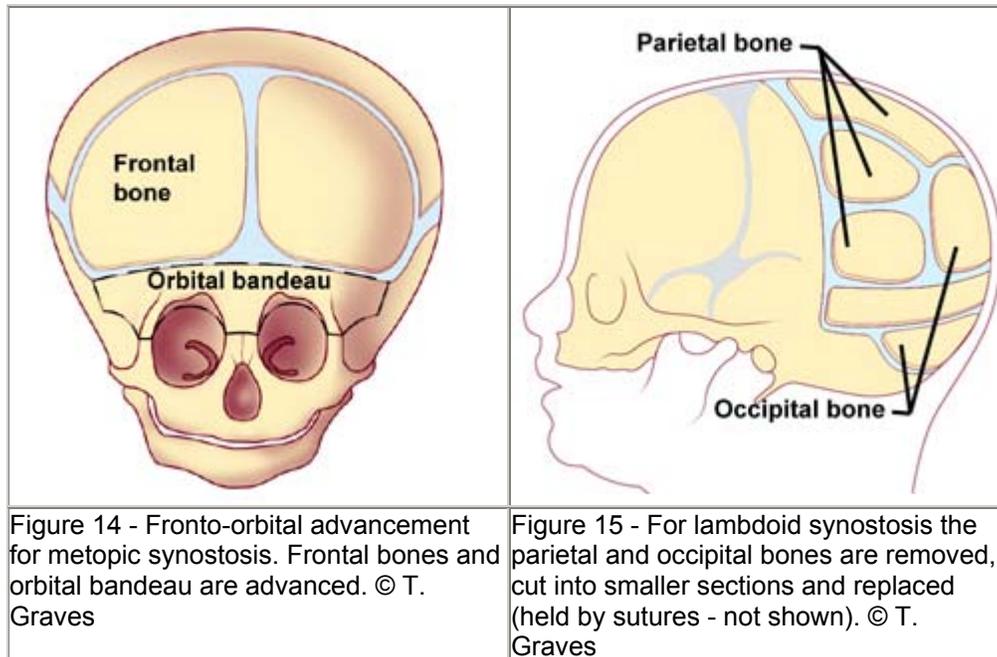


Figure 13 - Left. Case of hypertelorism (orbits too far apart). Orbital bone cuts are made to free the orbits from the skull and face. Right. The orbits are moved closer together and the construct held with miniplates. © T. Graves

- Metopic synostosis
 1. Strip craniectomy for metopic synostosis (metopic syndectomy). In the very young patient, the fused suture is removed and the ensuing brain growth allowed to remodel the skull
 2. Fronto-orbital advancement for metopic synostosis. The bones of the forehead as well as those about the eye are cut and advanced forward (Figure 14)
- Lambdoid synostosis
 1. Craniotomy for parieto-occipital advancement for lambdoid synostosis. The bones of the back of the skull are removed and repositioned (Figure 15)
 2. The procedure may be performed on both sides if necessary
- Deformational plagiocephaly
 1. This is usually treated without surgery with repositioning the infant so as not to lie on the flattened area of the skull or by helmet therapy in which the infant's head is placed in a remolding helmet (Figure 11)
 2. A similar operation used for lambdoid synostosis may be performed but is reserved for those cases failing conservative management and only for the most severe cases
- Helmets (cranial orthosis) can be used for deformational plagiocephaly as well as to help remolding following strip craniectomies. Several varieties of helmets are available and need to be worn for extended periods of time (months)
- Multiple Suture Synostosis
 1. Multiple craniectomies may be performed in which the affected sutures as well as deformed areas of the skull are removed. The skull is allowed to remodel as the brain grows
 2. This is usually performed in younger patients as intracranial pressure is often elevated
- Syndromic Synostosis:
 1. Surgery for these cases is more involved and complex and frequently requires removal of multiple fused sutures and reconstruction of the skull
 2. If there is associated hypo or hypertelorism the eye sockets may need to be moved farther apart or closer together (Figure 15)
 3. If hydrocephalus is present a shunt may be necessary

- 4. If a Chiari malformation is present then removal of some of the bone from the back part of the skull may be needed
- Cranioplasty (See Craniotomy) is a procedure that may be necessary for the repair of defects in the skull with either bone grafts or bone replacement prosthesis



Complications

- Overall, craniofacial surgery is relatively safe but certain risks need to be recognized. The overall occurrence of these complications is low
- All the cranial procedures carry the same risks, which include:
 1. Anesthesia
 2. Infection
 3. Bleeding
 4. Brain swelling
 5. Stroke
 6. Leak of cerebrospinal fluid
 7. Injury to the brain or eyes with temporary or permanent neurologic dysfunction or blindness
 8. Seizures
 9. Death
- Craniofacial surgery carries certain unique risks, which include:
 1. Unsatisfactory cosmetic result
 2. Leptomeningeal cyst (bulging out of the meninges under a bone defect) with need for reoperation
 3. Cranial defect requiring second procedure
 4. Recurrence of synostosis requiring additional procedures
 5. Need for possible removal of hardware

Postoperative Care

- Postoperative care begins in the recovery room and continues on in the Pediatric Intensive Care Unit

- Vital signs (pulse, blood pressure and respirations) are monitored
- A catheter may be inserted in an artery to monitor blood pressure and obtain blood for red blood cell counts and other blood tests
- A transfusion may be necessary
- A catheter is placed in the bladder to measure urine
- The endotracheal (tube in the windpipe) may be left in place after surgery to control respirations
- Within a short period of time most patients get facial swelling, which will go away in a few days
- Medications are given which include antibiotics to prevent infection and steroids to decrease swelling
- A drain may be placed under the scalp to drain fluid and is removed in a few days
- The infant is checked to be sure that brain function is intact
- A CAT scan may be done to view the brain and remodeled skull

After Care

- The infant is discharged when the blood count is stable, head swelling decreased and the infant is eating well. This usually takes several days
- A helmet may be necessary for protection or remodeling or both depending on the age and surgery performed
- Outpatient follow-up is necessary to ensure continued satisfactory growth and to monitor for the potential need for additional procedures (repair of defects, recurrence of suture closure, etc.)