MIDWEST NEUROSURGERY & SPINE
Adult & Pediatric SPECIALISTS

# Craniosynostosis and Craniofacial Disorders

Craniosynostosis is a congenital deformity of the infant skull that occurs when the fibrous joints between the bones of the skull (called cranial sutures) close prematurely. Due to this closure, the baby develops an abnormally shaped skull because the bones do not expand normally with the growth of the brain. The condition is usually apparent in infancy as an abnormal but characteristic head shape, and in some patients, abnormal facial features. In some cases, the growth of the skull is restricted enough to cause increased pressure in the head and can lead to headaches, visual problems, or developmental delays.

The severity and type of deformity depends on which sutures close, the point in the development process that the closure occurred, and the success or failure of the other sutures to allow for brain expansion. Synostosis of a particular suture alters the skull shape in a recognizable manner. An abnormal skull shape at birth is not always craniosynostosis, and may be related to fetal head position or birth trauma. The difference is that those abnormalities usually self correct, while craniosynostosis worsens if it is left untreated.

## Skull Anatomy

The human cranium, which houses and protects the brain, is composed of six major bones: the ethmoid, frontal, occipital, parietal, sphenoid, and temporal. In normal development, the cranial bones remain separate until about age 2. Then the separate cranial bones fuse together and remain that way throughout adulthood.

The **ethmoid** forms part of the eye cavity.

The **frontal** forms the top front of the head, the forehead, the brow ridges and the nasal cavity.

The occipital is located at the lower rear of the head, and forms the back and base of the skull. This is the point of articulation with the neck.

The **parietal** forms a major part of the cranium, covering large portions of the top, sides and back of the head.

The **sphenoid** is located by the temple of the head, and forms part of the eye cavity.

The temporal is located at the side of the head above the ear, and extends down behind the ear towards the jaw.

These bones are held together by strong, fibrous tissues called cranial sutures. In an adult, these sutures are fused together and the skull is rigid to protect the brain, but in an infant, these sutures are flexible.

The **coronal suture** is located on the side of the head extending from the soft spot to the area in front of the ear.

The **lambdoidal suture** is located at the back of the head between the occipital and parietal bones. The metopic suture is located between the soft spot and the root of the nose, allowing the forehead to grow normally and the eye sockets to separate correctly.

The **sagittal** suture is located on top of the head, extending from the soft spot to the back of the head.

The spaces between the bones within the fibrous tissues are called fontanels. The anterior, posterior, sphenoid, and mastoid fontanels are openings which close on their own as a part of normal growth. The fontanel most commonly referred to as the baby's soft spot is the anterior fontanel on the top of the head.

## Types of deformities

**Coronal synostosis** begins at the ear and goes back to the sagittal suture. Premature closure leads to a condition called anterior plagiocephaly. This may cause the baby's forehead to flatten on the affected side. Elevation of the eye socket, (vertical dystopia) on the involved side, a deviated nose and a slanted skull may also occur. If untreated, this may lead to amblyopia - loss of vision on the affected side.

**Lamboidal synostosis** is the rarest form of craniosynostosis. Premature closure of this suture leads to a condition called posterior plagiocephaly. This may cause flattening of the back of the head on the affected side, protrusion of the mastoid bone and posterior positioning of the affected ear. It may also cause the skull to tilt sideways. This condition can be misdiagnosed as occipital flattening (positional plagiocephaly).

**Bicoronal synostosis** occurs when both the left and right coronal sutures are involved. Premature closure of these two sutures leads to a condition called brachycephaly. This may cause the baby to have a flat, elevated and recessed forehead and brow.

**Metopic synostosis** begins at the nose and goes back to the sagittal suture. Premature closure of this suture leads to a condition called trigonocephaly. This may cause the baby to have a pointed forehead, midline ridge, triangularly shaped skull, and eyes that appear too close together.

**Sagittal synostosis**, the most common type of craniosynostosis, affects three to five infants in every 1,000 live births and is more common in males. Premature closure of this suture leads to a condition called scaphocephaly. Because the skull cannot expand sideways, it is forced to grow forward and backward. This may cause the baby to have a protruding forehead, narrowed temples and an elongated head.

## Diagnosis

Diagnosis can usually be made by feeling the skull for suture ridges and soft spots, as well as checking neck position and facial deformities. A radiological examination is usually necessary to confirm the problem, characterize the deformity, and guide the corrective surgical procedure. Plain x-rays of the skull may show the deformity, but computerized tomography (CT or CAT scans) provide more precise information about the fused sutures and the status of the underlying brain. Three-dimensional (3-D) CT scans may provide additional information to guide surgical correction.

## Surgery

Significant advancements have been made in the surgical treatment of cranial deformities. In more complex cases, there is a team approach utilizing the expertise of a pediatric neurosurgeon and craniofacial surgeon. Surgery involves releasing the fused suture and reshaping the brow, eye orbits and skull as needed. The goal of surgery is to correct cosmetic deformities and allow for normal expansion of the brain within the cranium. 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.

Traditionally, surgery is performed by making a scalp incision from ear-to-ear, mobilizing the scalp to expose the skull, and removing/reshaping the affected portion of the skull. In some cases, tiny plates and screws are used to fix the bones into proper position. These are frequently made of material that absorbs over time rather than metal. Surgery usually takes between three to seven hours depending on the case, may require a blood transfusion, and involves a hospital stay of three to seven days.

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. Using the aid of endoscopes, the surgical correction is performed through one or two small scalp incisions of about an inch each. The point of incision depends on which sutures are affected. The affected suture is opened and the brain is allowed to grow normally. There is less swelling and blood loss with this method of surgery. This process may be aided through postoperative helmet therapy. Length of surgery is usually about an hour, and most patients can be discharged from the hospital the second day after surgery.

## Other cranial deformities

### Occipital Flattening (Positional Plagiocephaly)

One of the most common reasons for a malformed head shape is plagiocephaly, a condition which is frequently confused with lambdoidal synostosis. In infants with plagiocephaly, the head may be flattened in the back (the occiput) because the infant lies persistently on the back of the head (often with the head turned primarily to one side). In addition, the ear on that side is often pushed forward, and even the forehead may look a bit prominent compared with the other side. This produces a parallelogram shaped head. Plagiocephaly is not caused by craniosynostosis and usually does not need to be treated surgically. It may be treated with a custom fitted helmet, which helps mold the baby's head back into a normal position.

### Apert's Syndrome

Apert's syndrome is a rare condition, affecting only one infant in every 100,000 to 160,000 live births. Patients with Apert's syndrome have very distinct facial and extremity features, including an abnormally shaped skull from craniosynostosis. This may cause the skull to be shortened, excessively tall or abnormally wide. The face may have a sunken-in appearance with a thick or beaked nose and bulging eyes. The upper jaw often has a narrow arch with an open bite and dental crowding. Other possible clinical problems include hydrocephalus, moderate hearing loss, speech impairment, and acne. There may be developmental disabilities, although some patients with Apert's syndrome have normal intelligence. All patients with Apert's syndrome demonstrate a unique hand malformation. This is characterized by a complex fusion of the skin, soft tissue, and bones of the fingers. Both hands are affected equally, as are the feet. This unusual variation is helpful in distinguishing Apert's syndrome from other like conditions.

The treatment of patients with Apert's syndrome differs due to significant variations in facial deformities, the age of diagnosis, and previous operations performed. Primary concerns are compression of the brain, breathing problems, protruding eyes with corneal exposure, and lack of facial growth. Surgery to repair the craniosynostois is preferable between the ages of 3 to 8 months. However, subsequent operations at different ages may be necessary.

### Crouzon's Syndrome

This syndrome affects about one person in 25,000. It is an inherited syndrome, although 25 percent of reported cases claim no family history. Patients with Crouzon's syndrome have distinct facial features similar to Apert's syndrome, although developmental disabilities are less prevalent. Premature fusion of both coronal sutures (bicoronal) leads to craniosynostosis in a majority of people with this condition. Aside from facial deformities, other possible clinical problems include hearing loss, dental crowding, nasal airway obstruction, a v-shaped palate, and a condition of the cornea called keratitis.

Surgery to repair the skull deformity is usually performed between the ages of 4 to 6 months. Subsequent operations to reshape the face and correct dental crowding are necessary; the first stage usually performed between the ages of 4 to 6.