CRANIOSYNOSTOSIS

What is Craniosynostosis?

Craniosynostosis affects 1 out of every 2000 live births. It represents the premature closure of the sutures or fibrous joints that connect the skull bones. Cranial sutures represent important growth centers for the skull. The size of the brain increases greatly from approximately 30-40% of adult size at birth to approximately 80% of adult size by age 3. It is this brain growth that drives the growth of the skull. As the brain grows, it signals bone growth at the sutures. The resulting increase in skull size accommodates the growing brain. When a suture closes prematurely, growth of the skull is restricted. This will lead to an abnormal skull shape and may cause an increase in intracranial pressure.

When sutures close early... This will lead to an abnormal skull shape and may cause an increase in intracranial pressure.

- Craniosynostosis represents the early closure of a skull suture.
When do sutures normally close?

The skull is made up of 6 main sutures; metopic, sagittal, left and right coronal, and left and right lambdoid. When one of these sutures closes prematurely, the remaining sutures try to compensate for the growth restriction at the fused suture. The metopic suture normally fuses by age 2; while the sagittal, coronal, and lambdoid sutures remain open until the 2nd decade of life. In general terms, growth is restricted perpendicular to the fused suture and compensatory growth occurs parallel to the fused suture. This phenomenon, called Virchow’s Law, is an oversimplification, but it helps explain the skull shapes seen in different types of craniosynostosis.

Are there other causes of abnormal head shape?

While craniosynostosis causes abnormal head shape, most cases of abnormal head shape are caused by positional plagiocephaly rather than craniosynostosis. Positional plagiocephaly is caused by molding of the head by external forces, e.g. consistently sleeping in the same position. The back of the head is flattened on the side of the deforming force and the ear and forehead on the same side may also be pushed forward. While the treatment of positional plagiocephaly is non-surgical (e.g. positional therapy and molding helmets), treatment of craniosynostosis often requires an operation. As missing a diagnosis of craniosynostosis could be detrimental, any suspicion should prompt a referral to a craniofacial surgeon.
Treatment of Craniosynostosis

As the growth of the skull and brain are intertwined, surgical correction is best performed by a team comprised of a pediatric craniofacial surgeon and a pediatric neurosurgeon. The pediatric neurosurgeon, who specializes in surgery on the brain and nervous system, evaluates for any related brain abnormalities (e.g. a Chiari malformation) and safely removes the skull bone while protecting the brain. The craniofacial surgeon, who specializes in surgery of the skull and facial bones, designs the skull bone cuts and expands and rearranges the skull bones to a more normal position while allowing for future skull growth. A team approach, involving a pediatric craniofacial surgeon and pediatric neurosurgeon, is essential in ensuring optimal treatment of these children.