**Craniosynostosis**

Craniosynostosis is describe as a pathologic condition associated with premature fusion of one or more of the cranial sutures. Typically, in developing infants, open sutures allow the skull to expand as the brain grows, producing relatively normal head shape. If one or more sutures are prematurely fused, however, there is restricted perpendicular growth to the fused suture(s) and compensatory growth in the skull’s unfused bony plates, producing abnormal head shape. Craniosynostosis occurs in 1 in 2100 to 1 in 2500 births and may be either nonsyndromic (isolated) or syndromic. In syndromic craniosynostosis, other birth defects are present with craniosynostosis and usually more than 1 cranial sutures have prematurely fused, typically involving both coronal sutures. Syndromic craniosynostosis are also normally genetic. The journal of craniofacial surgery states that, of the syndromic types, the Muenke syndrome is the most frequent, followed by Crouzon syndrome, and Pfeiffer syndrome. The Apert syndrome has the lowest prevalence.1

It is important to recognize and treat Craniosynostosis because it can be associated with many complications affecting sensory, respiratory and neurological function. The changes of head shape may be associated with increased intracranial pressure that may result in permanent brain injury. Besides the risks of brain injury, Craniosynostoses are associated with alteration of craniofacial growth leading to mid-facial hypoplasia; abnormalities in dental alignment; and orbital deformation. According to literature, no clear association has been seen between ethnicity and craniosynostosis. However, mothers of non-Hispanic white decent have been associated with an increased risk for having a child with craniosynostosis

The most common presentation of nonsyndromic Craniosynostosis are infants with an unusual head shape in the ﬁrst year of life. It typically affects males twice as often as females. Plagiocephaly is said to be the most common form of craniosynostosis and occurs in approximately one out of every 2,500 live births. Conversely, sagittal craniosynostosis is said to be the most common type of single suture craniosynostosis which is caused by the premature fusing of the sagittal suture. The second most common form of craniosynostosis is **unilateral or bilateral coronal** **synostosis**, which is caused by the fusing of the coronal suture. **Metopic synostosis which is** caused by the fusing of the metopic suture is less commonly seen, **while Lambdoid synostosis** the most **infrequent of all synostoses.**

According to many studies, the etiology of craniosynostosis remains unclear. However, it is thought to be associated with environmental and hormonal abnormalities. It is also thought be an abnormality of the suture itself, abnormal development of the cranial base, or have a genetic etiology in which activation of dominant mutations in the fibroblast growth factor receptors signal the mediating cranial suture fate. Studies revealed that numerous genes have been identified linking the occurrence of Craniosynostosis. Fibroblast growth factor receptors FGFR1, FGFR2 and FGFR3 (genes) which influence growth of the brain and connective tissue are essential in the development of the cranium and face. It appears that a mutation occurs in these genes that cause the sutures to fuse. Autosomal dominant inheritance with variable expression is seen with most common syndromic forms of craniosynostosis. A small percentage of nonsyndromic craniosynostoses have been found to be inherited.

Literature states that timing of surgery following specific categorization of cranial deformities is imperative because any delay in surgery beyond the first 9 to 12 months of life leads to progressive deformity of the cranial base, resulting in abnormal facial growth and asymmetry of the maxilla and mandible. Early intervention is ideal for infants 3 to 9 months old since the skull remains quite malleable making it easier to shape. Open craniotomy is a type surgical procedure used for the reconstruct the cranial vault. The objective of surgery is reconstructive, not cosmetic, allowing the brain to grow naturally. It is very invasive, with long anesthesia time as well as a high probability that blood products will be required. Conversely, minimally invasive endoscopic reconstruction is an alternative approach for sagittal craniosynostosis, in which the closed suture is resected.  This procedure is less invasive than traditional surgeries, which decrease the time spent under anesthesia, the need for transfusions, and lengths of hospital stay. This method of treatment relies on early surgery of less than 3 months of age because the correction requires rapid brain growth to help recontour the cranial bone. In addition, care of neonates and infants with severe multisuture synostosis is directed towards maintenance of the airway, support of feeding, eye protection, and treatment of raised ICP. Respiratory difﬁculty may require urgent sleep study assessment by specialist paediatric respiratory physicians and ear, nose and throat surgeons, necessitating either continuous positive airway pressure support and nasal stenting, depending on the anatomical cause.

The prognosis for craniosynostosis varies depending on whether single or multiple cranial sutures are involved, and whether other abnormalities are present. The prognosis is better for those with single suture involvement and no associated abnormalities. For the vast majority of children with craniosynostosis, growth and development proceed normally. Most of the children who undergo reconstructive surgery will require no further surgical procedures. Furthermore, procedure used to correct craniosynostosis have become safer during the last 3 decades. The once reported to be as high as 16.5 and 1.6% morbidity and mortality rates, respectively have declined to approximately 0.1% for each.2 Improvements in operative techniques and training as well as advancements in anesthesia care and patient blood management strategies have driven this trend.

In conclusion, the overall prevalence of Craniosynostosis has been estimated between one in 2,000 to 3,000 births worldwide and one out of 2,000 live births in the [United States](http://www.encyclopedia.com/topic/United_States.aspx). It can also be associated with more than 130 different syndromes. However, it most commonly presents as an isolated abnormality. The incidence of the nonsyndromic Craniosynostosis are as follow: Sagittal synostosis 1 in 5000 live births; Coronal synostosis 1 in 10,000 births and accounts for 20–30% of surgical cases. For Metopic synostosis 1 in 7000 to 1 in 15,000 births and incidence of metopic craniosynostosis is increasing. Lambdoidal synostosis less than 1 in 10,000 births and is rarely encountered as previously stated. It accounts for only 2% of all surgical cases.

**References**

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