A Review on Craniosynostosis

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Abstract-

Craniosynostosis is a developmental craniofacial anomaly, resulting in impairment of brain development and abnormally shaped skull. The main cause of craniosynostosis is premature closure of one or more cranial sutures. It usually occurs as an isolated condition, but may also be associated with other malformations as part of complex syndromes. When left untreated, craniosynostosis can cause serious complications, such as developmental delay, facial abnormality, sensory, respiratory and neurological dysfunction, anomalies affecting the eye, and psychological disturbances. Thus, early diagnosis, expert surgical techniques, postoperative care, and adequate follow-up are of vital importance in treating craniosynostosis.

Keywords: Craniosynostosis, development, classification, diagnosis, treatment

INTRODUCTION

Craniosynostosis is premature fusion of one or more cranial sutures leading to abnormal shape of the skull. In a newborn, the brain develops at a rapid rate in the initial periods: Its size doubles during the first 6 months and quadruples by the 1st year of life. The brain acquires 80% of its adult size by 2nd year of life. This rapid brain growth is allowed by the patent cranial sutures in the growing child. The growing brain itself provides a stimulus for keeping the sutures patent. It has been found that a minimal pressure of approximately 5 mm Hg from the growing brain is required to stimulate new bone deposition at the suture margins.[1] Some people feel it is better to label craniosynostosis as 'absence of cranial sutures' rather than 'premature fusion' as most of the 'fusion' occurs during the intrauterine period.[1] The abnormal shape of the skull has been known since antiquity. Hippocrates was aware of the abnormal shape of the skull [2] and Sommerring,[3] observed that bone growth in the skull occurred primarily at suture lines and abnormal skull shape develop if the suture was replaced by bone. In 1851, Virchow hypothesised about the pattern of growth across a fused suture.[3] Observations of Crouzon[4] and Apert and Bigot.[5] from 1910 to 1920 established that in many cases, the facial deformities and abnormal skull shape are associated with a myriad of other features. These clinical entities are now named after them.

CLASSIFICATION OF CRANIOSYNOSTOSIS

Different classifications of craniosynostosis are used depending on the underlying mechanism, presence of other disorders, or number of fused sutures. For instance, if a craniosynostosis develops due to a primary defect of the ossification process it is called primary craniosynostosis. On the other hand, secondary craniosynostosis is the result of known systemic diseases with hematologic or metabolic dysfunction, such as rickets and hypothyroidism. Secondary craniosynostosis can also develop in newborns with microcephaly due to a failure of brain growth or following shunt placement in children with hydrocephalus. Furthermore, craniosynostosis can be classified into syndromic, e.g., as part of Apert, Crouzon, or Pfeiffer syndrome, and more commonly encountered, non-syndromic craniosynostosis, where it develops as an isolated disorder. Simple craniosynostosis is a term used when only one suture fuses prematurely, while complex craniosynostosis is used to describe a premature fusion of multiple sutures [6-11].

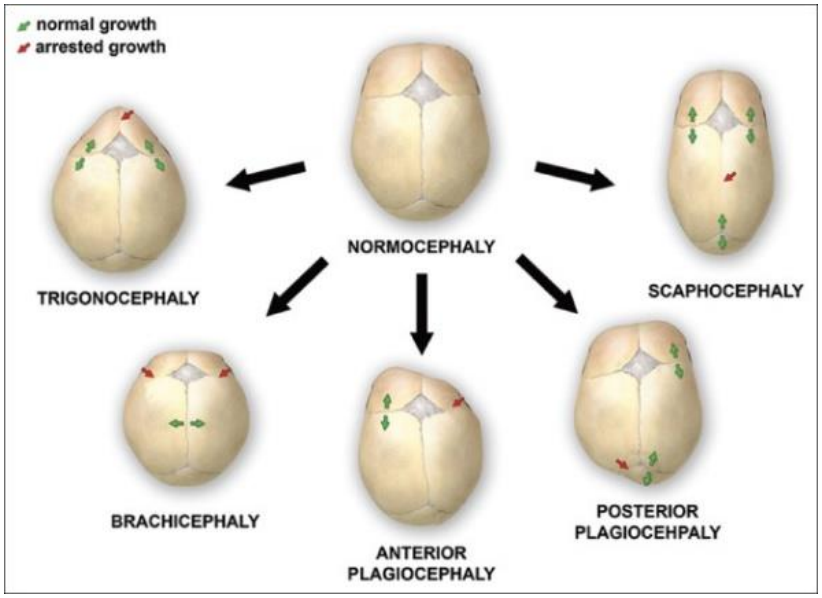


Figure. 1 Various deformations of the skull, associated with single-suture synostosis

Craniosynostosis is a condition in which fibrous sutures in an infant skull prematurely fuses thereby changing the growth pattern of the skull (Fig.2). They are various deformations of the skull, associated with single-suture synostosis.

- **Scaphocephaly** is an early closure of fusion of the sagittal suture. This is the most common type of synostosis. This suture runs front to back, down the middle of the top of the head. This fusion causes a long, narrow skull. The skull is long from front to back and narrow from ear to ear.
- **Plagiocephaly** involves fusion of either the right or left side of the coronal suture that runs from ear to ear. This is called coronal synostosis and it causes the normal forehead and the brow to stop growing.
- **Trigonocephaly** is a fusion of the metopic (forehead) suture. This suture runs from the top of the head down the middle of the forehead, toward the nose. Early closure of this suture may result in a prominent

ridge running down the forehead. Sometimes, the forehead looks quite pointed, like a triangle, with closely placed eyes

- **Brachycephaly** is a fusion of both coronal sutures which causes restriction of growth of the anterior fossa resulting in a shorter and wider than normal skull.

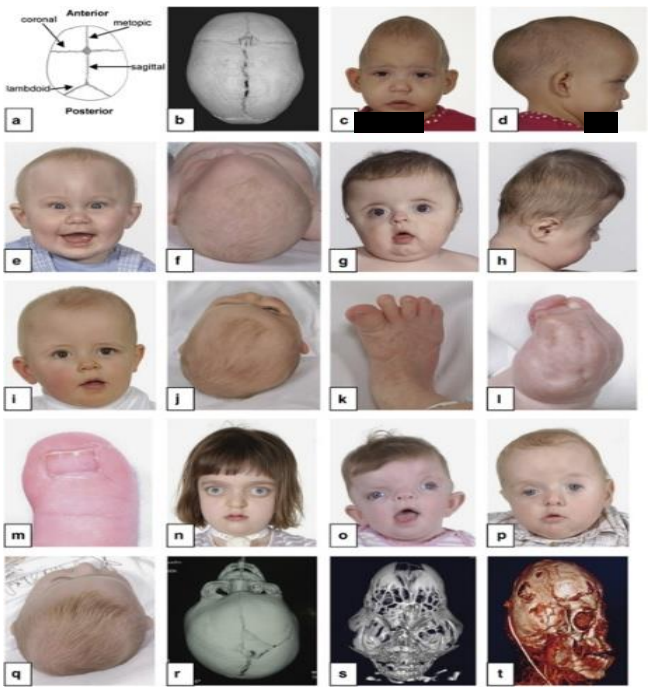
SYMPTOMS

The signs of craniosynostosis are usually noticeable at birth, but they'll become more apparent during the first few months of your baby's life. Signs and severity depend on how many sutures are fused and when in brain development the fusion occurs. These can include:

- A misshapen skull, with the shape depending on which of the sutures are affected
- An abnormal feeling or disappearing fontanel on your baby's skull
- Development of a raised, hard ridge along affected sutures
- Slow or no growth of the head as your baby grows

DIAGNOSIS:

Diagnostic features of craniosynostosis (Fig 2). (a) Schematic diagram showing positions of the major cranial sutures. (b) CT scan (vertex view of skull) showing major sutures; anterior is at top. (c,d) Sagittal synostosis: note long, narrow head. (e,f) Metopic synostosis: note hypotelorism and triangular profile of forehead. (g,h). Bicoronal synostosis: broad, flattened head. (i,j) Right unicoronal synostosis: note flattened brow and anterior position of ear on affected side, deviation of nasal tip and prominent brow on unaffected side. (k–m), Congenital anomalies of feet or hands characteristic of Pfeiffer syndrome (k), Apert syndrome [(l)] and craniofrontonasal syndrome (m). (n) Crouzonoid facial appearance. (o) Severe hypertelorism, grooved nasal tip and left unicoronal synostosis in craniofrontonasal syndrome. (p) Ptosis and left unicoronal synostosis in Saethre-Chotzen syndrome. (q) Positional plagiocephaly: prominence on right anteriorly and left posteriorly, with right ear anterior and parallelogram shape to skull. (r) CT reconstruction showing left unicoronal synostosis. (s) CT reconstruction showing cloverleaf skull. (t) CT venogram showing abnormal venous drainage in multisuture syndromic craniosynostosis. [13]



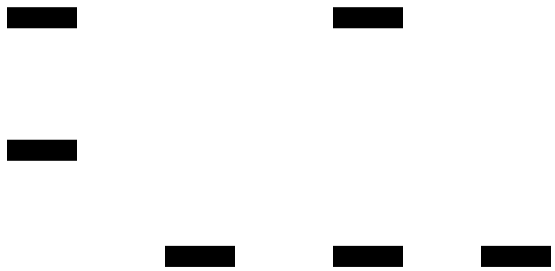


Figure 2 Image showing various types of synostosis

TREATMENT

The primary goal of surgical intervention(Fig. 3) is to allow normal cranial vault development to occur.[14] This can be achieved by excision of the prematurely fused suture and correction of the associated skull deformities[14] If the synostosis goes uncorrected, the deformity will progressively worsen not only threatening the aesthetic aspect, but also the functional aspect. This is especially prevalent with asymmetric conditions, such as unilateral coronal synostosis, with compromised function of the eyes and the jaw. In addition, signs of compromised neurodevelopment have been seen amongst all the synostosis, although this may also be caused by primary maldevelopment of the brain and can thus not be prevented by surgical intervention [15]The most important factors in determining the extent of surgery and surgical modality are the patient age and presentation [12]. Although the surgical treatment of craniosynostosis is most commonly used, the conservative approach may be adopted first, especially in patients with positional plagiocephaly and in cases in which unilateral synostosis is not very pronounced.

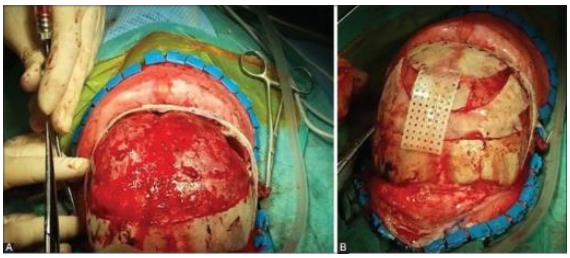


Figure .3 The surgical reconstruction of anterior plagiocephaly.

CONCLUSION

Craniosynostosis is a rare craniofacial anomaly which may lead to various complications, deformations, and neurological impairment during the child's development. Early identification and appropriate treatment are therefore vital. The aim of the surgical treatment is to enable the normal brain development and to achieve an acceptable cosmetic effect.

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