

CRANIOSYNOSTOSIS



ESSAY

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to
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ABSTRACT

The aim of our work is to highlight the importance of the surgery time as craniosynostosis should be operated on the first year of life as it is the period of maximal brain growth. A decision to operate must be made according to the importance of the dysmorphosis and possibilities of improvement of the functional impairment. Hypothermia and Blood loss during and after surgical correction is the most important risk of surgery. Accidental dural tearing , C.S.F leak and infection can also occurred. These complication could be reduced by a new surgical technique called endoscopic strip craniectomy.

Introduction

Craniosynostosis is a defect in which one or more of the flexible and fibrous joints (cranial sutures) between the skull bones closes too soon; it occurs before birth or within a few months after birth. The skull cannot expand normally with growth of the brain, and so assumes an abnormal shape. Craniosynostosis can occur alone or as part of a syndrome of craniofacial defects. (**Alderman BW, Zamudio S. 2003**)

This results in an abnormally shaped skull or face. The forehead may be very pronounced and inclined forward. Viewed from above, the skull may be more rectangular in shape rather than oval. Other forms of craniosynostosis include coronal craniosynostosis (affecting the coronal suture that crosses the top of the skull from temple to temple), metopic craniosynostosis (affecting the metopic suture of the forehead), sagittal craniosynostosis (affecting the sagittal suture that unites the two parietal bones), and lambdoidal craniosynostosis (affecting the lambdoid suture between the occipital and parietal bones of the skull). Craniosynostosis is a rare occurrence. The sagittal form of the disorder, in which the sagittal suture closes prematurely, is the most common form of craniosynostosis, occurring in three to five of every 1,000 babies, typically males. The frequencies of the various types of craniosynostosis are 50–60% sagittal, 20–30% coronal, 4–10% metopic, and 2–4% lambdoid (**Anderson FM, Geiger L . 2006**)

Diagnosis is made on the basis of a physical examination. Treatment involves medical specialists (pediatric neurosurgeons, pediatric plastic surgeons, craniofacial surgeons) and specialized nurses. Surgery is the common treatment for craniosynostosis. The traditional surgeries involve the exposure of the skull, physical breakage of the fused suture region, and the restoration of the scalp. Also, the surgeries produce much bleeding (sometimes a blood transfusion is necessary) and leave a large scar, and transient swelling and bruising can occur.

A new surgical technique called endoscopic strip craniectomy has been pioneered by two pediatric surgeons from the University of Missouri Health Care Center. This surgery is much less invasive, produces only a relatively small scar, and leaves little physical after effects such as swelling and bruising. In the procedure, an endo-scope is used to remove the closed suture through incisions that are only several inches in length (**Argenta LC, David LR, 2005**).

The outlook for a complete recovery for a child with craniosynostosis depends on whether just one suture is involved or whether multiple sutures have closed. Also, the presence of other abnormalities can lessen the confidence of a satisfactory outcome. Without surgical intervention, craniosynostosis can lead to increased brain pressure, delayed mental development, mental retardation, seizures, or blindness (**Arnaud E, Renier D, 2005**).

Historical view

Since antiquity, various cultures have focused on individuals with abnormal cranial contour. Allusions to aberrant skull shape have been noted in writings connected with the ancient Chinese gods of good fortune and long life (***Fukurokuju and Shou Lao,2006***).

Nearly 200 years ago, **Sommerring** reported the first scientific investigation of Cranial deformities .He discussed cranial sutures, recognized their primary importance in skull Growth and asserted that premature suture fusion produced cranial deformity.

Reports of craniosynostosis were increasingly disseminated and ophthalmological Perspectives were introduced. Authors also described craniosynostosis in association with other Anomalies and provided the impetus for future classification of syndromic craniosynostosis.

Apert and Crouzon ,among others, described those syndromes that continue to bear their Names. In the late nineteenth century, **Lane and Lannelongue** reported the first modern

Surgical corrections of skull deformity resulting from premature suture closure. From these Original pioneering experiences, subsequent advances in treatment have continued into Contemporary times.Virchow's hypotheses concerning craniosynostosis remained the standard For nearly a century.

However, in the mid-twentieth century, **Van der Klaauw and Moss** Questioned the primacy of the calvarial sutures as the antecedent mediator of skull deformities.Based on his original ideas, subsequent work and the efforts of others, **Moss** proposed That the primary anomaly in craniosynostosis arose in the cranial base. He hypothesized

that the Primary abnormality arose in the cranial base, and this resulted in the secondary fusion of the Cranial vault suture(s). His arguments were fourfold on occasion, suture patency was Found at surgery, despite pre-operative suspicion of premature suture fusion and characteristic Skull configuration characteristic anomalies of the cranial base were associated with Specific calvarial suture closures experimental removal of normal cranial vault sutures Resulted in no significant change in overall skull shape; and cranial base development and Maturation precedes those of the cranial vault.

Additionally, **Moss** believed that the primary force Driving the sutures' deposition of bone (with consequent expansion and modeling of the skull) was growth of the underlying brain. This was termed the "functional matrix theory" .Further work by **Persson** and others endeavored to clarify the primary locus of craniosynostosis.**Persson et al**, demonstrated that experimental restriction of a suture's growth Produced skull deformities that mimicked craniosynostosis in humans. In addition, Cranial base and facial abnormalities appeared to occur in response to the cranial suture Restriction. This suggested that craniofacial anomalies were primarily the result of suture Fusion – not the cranial base, as **Moss** had proposed.

Marsh and Vannier, reported that preexisting cranial base abnormalities resolved After surgery in which only cranial vault alteration was undertaken .Collectively, Considerable data have accrued against Moss's stance that suggest, at least in most cases of non-Syndromic craniosynostosis, that the cranial vault sutures assume a major inciting role in the Pathogenesis of craniosynostosis. In states of syndromic craniosynostosis (*e.g. Apert or Crouzonsyndromes*), however,

a more generalized pathologic process involving the cranial vault sutures And cranial base may exist.

Work by **Opperman and colleagues** emphasized the critical influences of Mesenchymal tissues, including the dura mater and periosteum, at the suture site in regulating And maintaining suture patency during development .Recognition of this dynamic Interaction and the existence of factors, including matrix and cytokine influences (fibroblast Growth factors (FGF), fibroblast growth factor receptors (FGFR) and transforming growth Factor beta (TGF- β), have been instrumental in the refinement of our contemporary Molecular understanding of craniosynostosis (*Burke MJ,et,al.1999*).

Historical Perspectives in the Treatment of CranioSynostosis:

In the early century; the aim of surgery in craniosynostosis is countering at the functional Problem of the disease (*Marchae and Renier,1982*) .

Classic Craniectomy:

The technique is divided into two basic operative approaches:

1-The linear craniotomy whose aim is the creation of a new cranial suture at the site of the the synostosed cranial suture.

2-The fragmentation of the cranial vault where several pieces of flat bone are used as grafts or Pedicle flaps to refashion a vault.These classic techniques may result in a satisfactory cerebral decompression, but

create two Problems which are rapid reossification of the cranial vault and morphological disturbances (*Marchae and Reiner, 1982*).

Total Craniectomy:

Total craniectomy of the vault extending to the roof of the orbits was advocated in (1965) by **Hieroium Powiertowski**, a polish neurosurgeon. Relapse of brain is radical and reossification is supposed to proceed rapidly as long as the Dura is intact. The child wears a protective helmet Until the cranial vault is reformed and the shape of the reossified vault should be satisfactory.

Application of craniofacial technique:

Craniofacial principles applied to the treatment of craniosynostosis have proven by **Tessier** Was the first in 1971, to publish the result of radical correction of the recessed forehead of facio-Craniosynostosis of Crouzon or Apert syndromes. **Tessier** made a horizontal advancement, with A tongue and groove lateral fixation, this operation was performed on teenagers or adult.

Rougerie, Derome and Anques working with Tessier proposed in 1972 an early treatment Of caniosynostosis by mobilization of the free bony segment of the cranial vault, at the same Time decompressing and remodeling the vault. They rearranged the free bony pieces and Maintain them in proper position. The results were good in cases involving only the sagittal Suture, but often inadequate in cases of coronal suture affection.

Sricker and Montaut in 1972 proposed the rocking the supraorbital bar in children with Oxycephaly with fixation with laterally based bone grafts, and a transposition of frontal bone Flaps pedicle on the temporal muscle. The muscular pedicles complicated the adjustment of The bony segments and prevented full rotation of theses pieces.

In 1977, **Marchae** described an early bilateral frontocranial remodeling for Trionocephalies, plagiocephalies and brachycephalies introducing the float in forehead.

Craniosynostosis in Egypt:

In Egypt **Osman Sorour** begun the surgical treatment of craniosynostosis by Application the four flaps operation in cases of early infancy in 1961 (**Sorour,1961**). Then **Osman Sorour and Khiary Samra** in 1968 made a new modification and applied The bilateral flap operations in cases of scaphocephalies (**Samra and Sorour, 1968**). **Gheita and Assaad** in 1985 applied the new operations of **Tessier** in the treatment of Craniosynostosis which was the forehead advancement by involving large segments of Cranium and remodeling the vault, the forehead and the midface (**Assaad and Gheita, 1985**). And they applied a modification to the forehead advancement flap by combined it with a strip Craniectomy in 1987 and called it "The Three Flap Procedure" (**Assaad and Gheita, 1987**). They also made a new operation for isolated sagittal synostosis which is called Tripple Strip Crainectomy" (**Assaad and Gheita, 1987**).