Recent Trends in the Management of Craniosynostosis

Thesis

Submitted in fulfillment of the MD Degree in Neurosurgery

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2008

بسم الله الرحمن الرحيم

(و قل رب زدني علماً)

صدق الله العظيم

Abstract

Craniosynostosis is abnormal early closure of cranial sutures by bony union. In literature the most commonly affected is the sagittal suture, however in Egypt oxycephaly is the commonest type. Less common is the syndromic type of craniosynostosis like Apert and Crouzon syndromes. Cases usually present with deformity and or symptoms and signs of increased intracranial pressure. Surgical intervention is recommended during the first year of life (3 to 9 months) to prevent serious complications resulting from long standing elevated intracranial pressure like optic atrophy, Blood loss during and after surgical correction is the most important risk of surgery.

Keywords

Craniosynostosis, Oxycephaly, brachycephaly, Scaphocephaly, Crouzon's syndrom, Apert's syndrom, Frontorbital advancement, Gradual Distraction Osteogenesis, Endoscopic Assisted Strip Craniectomies.

ACKNOWLEDGEMENT

Above all, deep thanks are to God, The Great, for everything, and for giving me health and strength to finish this Work .

I am greatly honored to express my deepest gratitude and respect to **Prof. DrAhmad Asaad,** Professor of Neurosurgery, Faculty of Medicine, Cairo University. No words can describe his stimulating supervision, continuous advice and encouragement.

I am deeply thankful to **Prof Dr. Ahmad Eissa** Professor of Neurosurgery, Faculty of Medicine, Cairo University. I am greatly honored and pleased to have had the opportunity to learn from his creative experience and wise counsel.

My deep gratitude goes to **Prof. Dr. Khaled Baseem,** Professor of Neurosurgery, Faculty of Medicine, Cairo University. He was the person who put the idea and plan of this work. He supervised closely its progress with great interest. His care has been the greatest help for me from day one till the finishing of this work.

My Special Thank to my family for their everlasting support and encouragement.

Contents

HISTORY OF CRANIOSYNOSTOSIS

Historical Perspectives

Unusual head shape has made a striking impression throughout history and across cultures. The ancient Chinese gods of good fortune and long life, Fukurokuju and Shou Lao, were described as having high, domed heads ***(Greig DM, 1926).*** In *The Iliad,* Homer speaks of Thersites, the ugliest man saying that his head was pointed at the top, based on this Homeric reference, the term *tete a la Thersite* for acrocephaly is found in the French literature. ***(Homer, 1948).***

Hippocrates described cranial deformations and their relationship to the cranial sutures as follows:

Men's heads are by no means all like to one another, nor are the sutures of the heads of all men constructed in the same form. Thus, whoever has a prominence in the anterior part of the head in him the sutures of the head take the form of a Greek letter tau, T. But whoever has the prominence in the back part of the head, in him the sutures are constructed in quite the opposite form to the former. But whoever has a prominence of the head both before and behind, in him the sutures resemble the Greek letter eta, H, although the statement can not be interpreted with certainty, when Hippocrates spoke of different forms of the cranial sutures, he may have been referring to premature synostosis of the coronal or lambdoid sutures in the first two instances, and to closure of all the sutures in the third instance ***(Fairman D, et al. 1949)***.

The significance of the cranial sutures was known to Galen, who described headache and exophthalmos in patients with too few sutures

(Batholin T, 1651). Aristophanes and, later, Galen used the term oxycephalus for tower head ***(Gunther H, 1931).***

Cornelius Celsus, a Roman physician who lived around the time of Christ, described skulls in which no sutures were present, although he made no mention of cranial deformity ***(Laitinen L,1956).*** Oribasius (A.D.325-403) observed in his writings that cranial deformities were associated with palatal defects and other abnormalities. He recognized that pointed and asymmetric skulls were manifestations of the same disorder ***(Bussemaker et al, 1858).*** In 1557, Lycosthenes described the birth of a child who had an unusual head with malformations of the hands and feet. This may represent a very early description of acrocephalosyndactyly ***(Valentin B, 1938).***

The first scientific investigator of cranial deformities in modern times was Sommerring, who in 1800 described the structure of sutures and considered them of primary importance in skull growth. He noted that premature sutural fusion would result in head deformity and had observed it in children with synostosis of the lambdoid suture ***(Sommerrring ST,1800)***

In 1830, Otto observed premature cranial synostosis in both humans and animals and did not consider the abnormality particularly rare. He attributed the small head to either a defective brain (microcephaly) or to premature sutural fusion. He thought that the etiology was based on either fetal trauma or birth trauma; he specifically excluded rickets and hydrocephalus. In his report, Otto realized that premature suture closure resulted in cranial deformity with compensatory expansion elsewhere ***(Otto AW, 1830)***

In 1851, Virchow further developed Otto's ideas. What has generally become known as Virchow's law states that skull growth is arrested in a perpendicular direction to the closed suture and compensatory overexpansion takes place at patent sutural sites ***(Virchow R, 1851).*** In 1856, Minchin, a Scottish physician, reported two cases of sagittal synostosis ***(Minchin H, 1856).*** In 1866, Graefe reported an 8 year old boy with turricephaly, seizures, headaches, proptosis, and papilledema, giving a very thorough description of the visual disturbances ***(Graefe A, 1866).***

Although brachycephalic craniostenosis with syndactyly had been reported toward the end of the 19th century by several investigators, the French pediatrician Apert is generally credited with discovering the condition. In his publication, which appeared in 1906, he used the term acrocephalosyndactylia ***(Apert E, 1906).*** In 1912, Crouzon, a neurologist reported the condition that bears his name today ***(Crouzon O,1912).***

In 1890, the French surgeon Lannelongue reported an operation in which he cut channels along the margins of a fused sagittal suture. In 1892 ***(Lannelongue M, 1890)***. The inability of clinicians at that time to distinguish between primary craniosynostosis and primary microcephaly led to an uncritical acceptance of the surgical procedure.

In 1921, Mehner suggested extirpatiation of prematurely synostosed sutures ***(Mehner A, 1921)*** and in 1927, Faber and Towne advocated a more extensive craniectomy ***(Faber HK et al, 1927)***

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 khairy Samra in 1968 made a new modification and applied the bilateral flap operations in cases of scaphocephaly***. (Samra and Sorour. 1968).***

Then Gheita and Assaad in 1985 applied the new operations of Tessier in the treatment of craniosynostosis which was the forehead advancement by removing large segments cf the cranium and remodling the vault, the forehead and the midface***.(Assaad and Gheita, 1985).***Then they applied a modification to the forehead advancement flap by combining it with a strip craniectomy in 1987 and called it "The Three Flaps procedure" ***(Assaad and Gheita, 1987)***.

They also made a new operation for isolated sagittal synostosis which is called 'triple strip craniectomy ***(Assaad and Gheita, 1987).***In 1989 they applied a new modification for the previous "three flap procedure by breaking the intact strip of bone covering the sagittal sinus and called it type 2 operation and the primary one called type 1 ***(Assaad and Gheita, 1989)***

ANATOMY & DEVELOPMENT OF THE SKULL

Embryology of the skull

Unfortunately, many of the good descriptions of skull development in human embryos were published before the current staging system and therefore must be discussed in terms of crown-rump (CR) length.

General Landmarks of Skull Development

The early formation of the cranium begins in stage 11 (2.5 to 4.5 mm CR), at which time the position of the future temporal bone is marked by the optic plate. At this time, the anterior neuropore is closing and the first four somites are beginning to be incorporated into the occipital segmentation. Shortly thereafter (stage 12; 3 to 5 mm CR), the optic plate is transformed into a vesicle and the boundary of the future sphenoid and occipital bones is identifiable as the notochord separates from the rostral neural tube.

Vascularization occurs in the mesoderm surrounding the rostral neural tube during stages 12 and 13 (4 to 6 mm CR). Also, the four occipital somites are now more clearly delineated, and a "sclerotomic fissure" is present in the most caudal of these segments . This fissure contributes to the formation of the atlas, but the significance of sclerotomic fissures is possibly more applicable to vertebral than to cranial segmentation. These fissures divide somites into rostral and caudal portions, the caudal portion of one somite eventually fuses with the rostral portion of the next somite to form a vertebral segment ***(Sensing EC, 1957)***

During stage 14 (5 to 7 mm CR), the meninx primitiva is first seen, and in stage 15 (7 to 9 mm CR), the skull is clearly demarcated from the vertebral column by the first cervical segmental artery. Pia mater is present around the brain during stages 15 to 17 (11 to 14 mm CR). The skull has a membranous roof present during stage 16 (8 to 11 mm CR), while the first evidence of chondrification is not found until the next stage, when it is present in both the body of the sphenoid and the basiocciput. Chondrification continues until stage 20 (18 to 22 mm CR), which is also when the first indication of dura mater is found in the skull. Unfortunately, the changes in skull development become too complex to discuss collectively after this early period because ossification begins. ***.(Sensing EC, 1957)***

Frontal Bone

The frontal bone develops as bilateral structures separated by the metopic suture and fontanel. Inman and Saunders studied the frontal bone in 98 specimens from the sixth gestational week to the tenth postnatal month. The membranous bone first shows finely reticulated areas of ossification at 32 to 36 mm CR. These areas are 2 to 3 mm in length and are found in the position of the superaciliary arch, which is on the lower portion of the frontal squama. ***(Inman VT, and Saunders JB deC m, 1937)***

The ossification pattern then spreads rapidly over the pars frontalis and pars orbitalis. At 42 mm CR, in the lateral two-thirds of the orbital margin, a linear thickening of osseus material is present. Ossification spreads to the zygomatic process (49 to 59 mm CR), which is separated from the zygomatic bone until 250 mm CR. The orbital plate begins to fuse with the, medial angular process at 80 mm CR. The frontal bones remain separated until after birth, and ossification of the metopic suture takes place during the

second postnatal year. Frontal sinuses do not develop until the first postnatal year ***(Barden CR, 1910).***

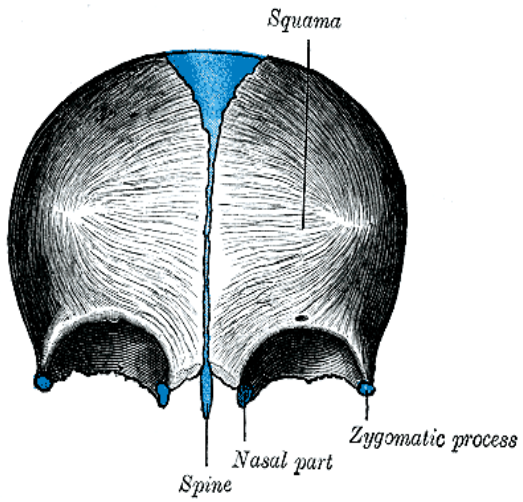

Fig (2-1)**:** Frontal bone at birth ***(Peter L, et al. 1989)***

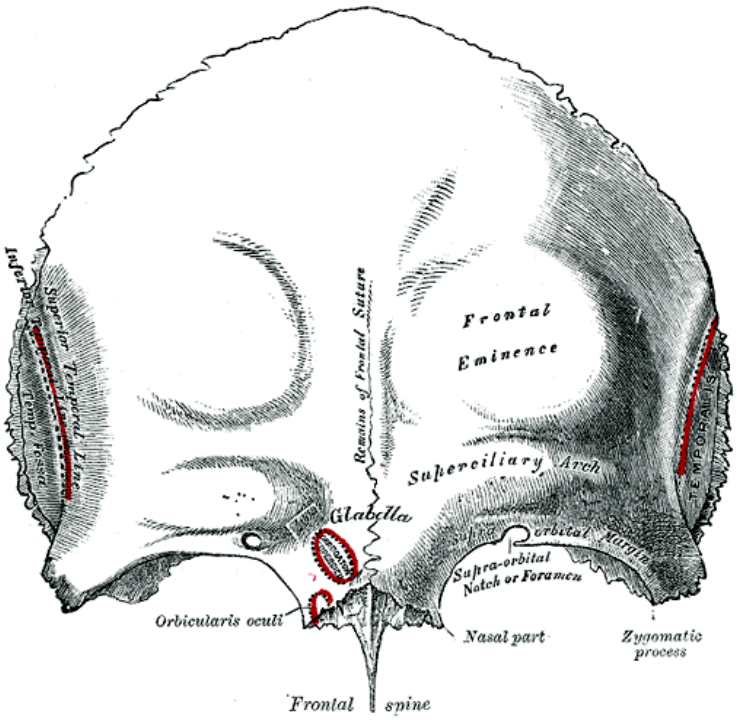

Fig (2-2): External surface of the frontal bone ***(Peter L, et al. 1989)***

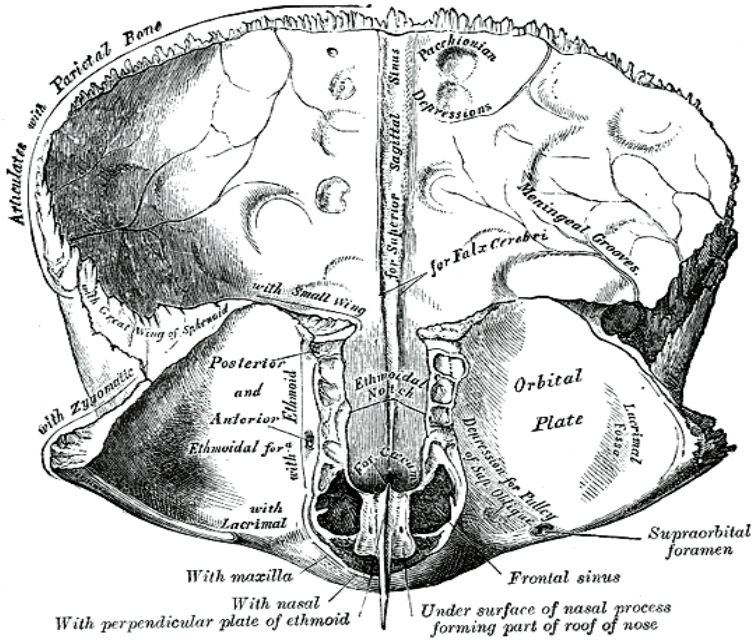

Fig (2-3): Internal surface of the frontal bone ***(Peter L, et al. 1989)***

Parietal Bone

Information about specific features of parietal development is scarce, probably because the process is relatively uncomplicated. The membranous parietal bone arises from a superior and an inferior center. Ossification, first seen at 37 mm CR, radiates progressively in all directions.

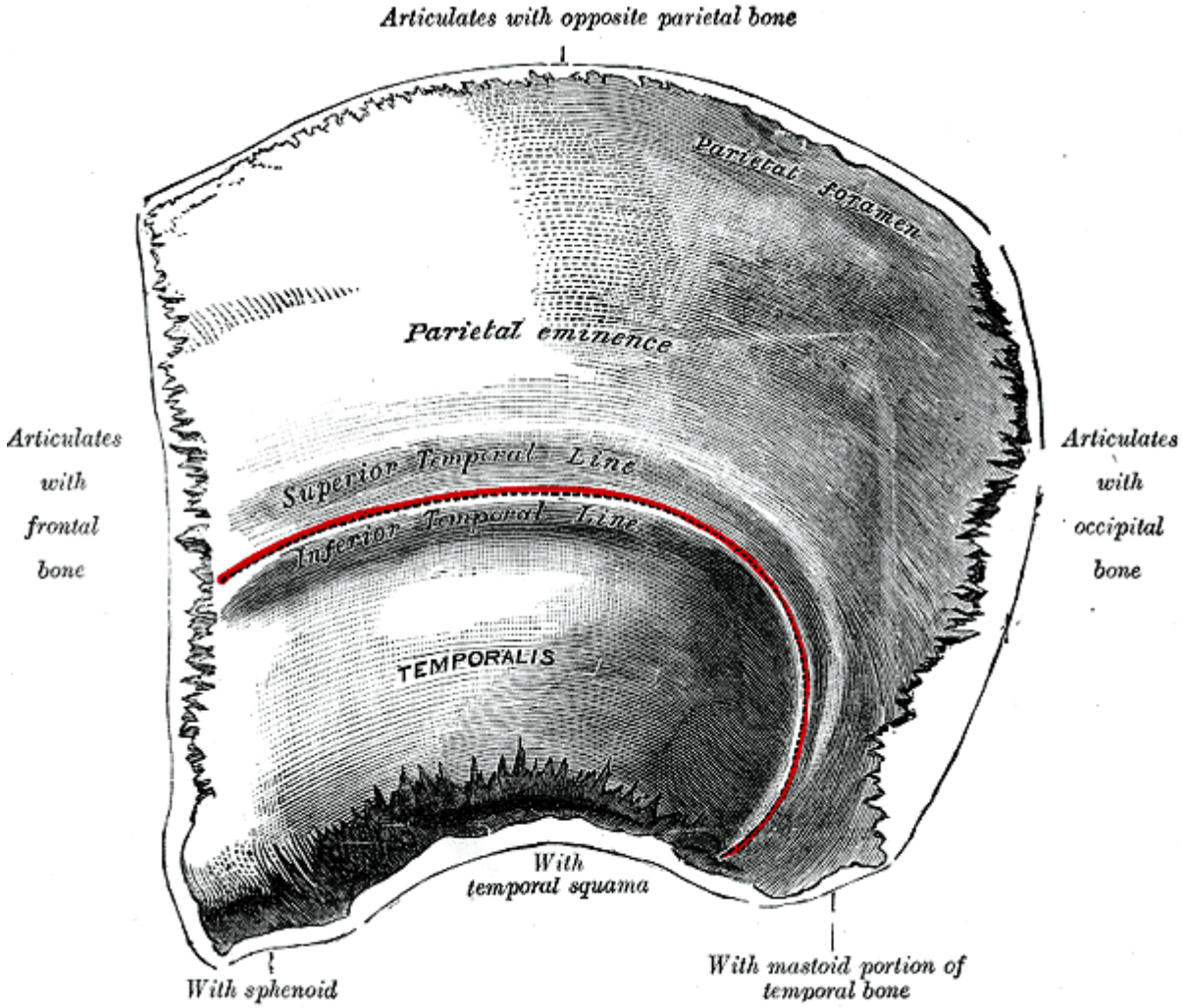

Fig (2-4): External surface of the parietal bone ***(Peter L, et al. 1989)***

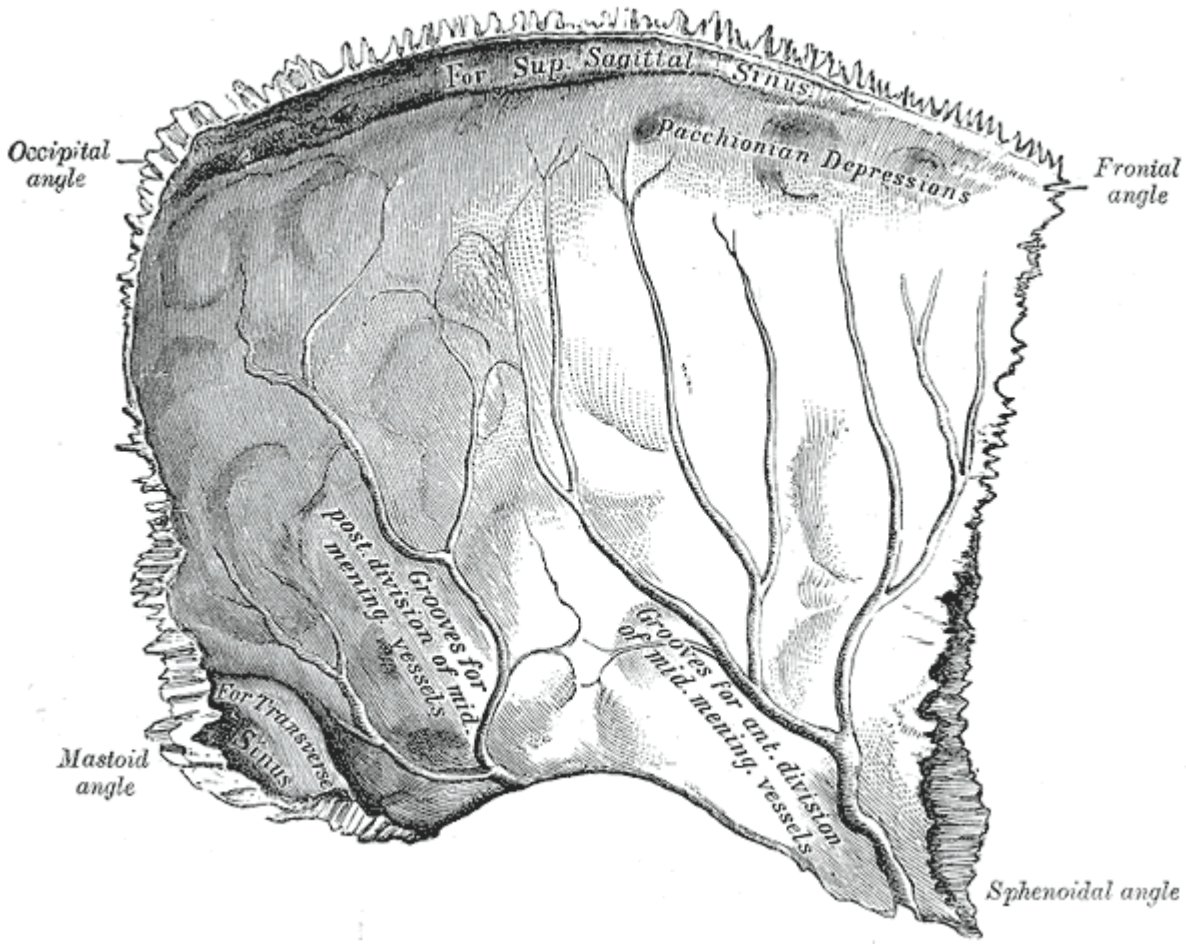

Fig (2-5): Internal surface of the parietal bone ***(Peter L, et al. 1989)***

Sphenoid Bone

The sphenoid is one of the most complex bones in the body. Because of its position in the base of the skull, the sphenoid participates in the formation of all three cranial fossae as well as the orbit, nasal fossae, and temporal and infratemporal regions. Changing patterns of morphology characterize the sphenoid.

At birth, it consists of three pieces of bone, a midline body (presphenoid, basipostsphenoid. and lingulae) and two lateral pieces (alisphenoid with pterygoid processes). The body contains paired sphenoid sinuses separated by a thin septum. The first portion to ossify (around 30 to 32 mm CR) is the pterygoid plate and perhaps the hamulus. The latter becomes the very tip of the internal pterygoid process, and it is remarkable