Updated Surgical Techniques in Correction of Craniosynostosis

Thesis

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بسم الله الرحمن الرحيم

[وأنزل اللـه عليـك الكتــاب
والحكمة وعَلَّمَكَ مَا لَمْ تَكُن تَعْلَمُ
وكَانَ فَضْلُ اللَّهِ عَلَيْكَ عَظِيماً]

صدق الله العظيم
النساء .. آية ١١٣

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Content

List of figures

Introduction

Craniosynostosis is a defect in which one or more of the flexible and fibrous joints (cranial suture) between the skull bones close too soon; it occurs before birth or within a few months after birth. The skull can't 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.

Forms of craniosynostosis include:

- Coronal craniosynostosis (affecting the coronal suture between frontal and parietal bones)
- Sagittal craniosynostosis (affecting the sagittal that unites the two parietal bones)
- Lambdoid craniosynostosis (affecting the lambdoid suture between the occipital and parietal bones of the skull).
- Metopic craniosynostosis (affecting metopic sutue between tow frontal lobes)

Craniosynostosis is 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 1000 babies, typically males. The frequencies the various types of the types of craoinosynostosis 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 neurosurgeon, pediatric plastic 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 (sometime 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 endoscope 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:

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).***

Hippocrates described cranial deformations and their relationship to the cranial sutures. 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 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 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 ofacrocephalosyndactyly ***(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 ***(GraefeA, 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. In1892 ***(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)***

Khaled B. Aly described non endoscopic minimally invasive technique for anterior clavarial synostosis without use of post-operative helmet, published in Egyptian Journal of Neurology Psychiatry Neurosurgery 2010 and will be discussed in details in our thesis **(Khaled B. Aly 2010**).

SKULL ANATOMY

The skull consists of 8 cranial bones and 14 facial bones.
The cranial bones lined by meaning protecting the brain these bones are frontal bone, parietal bones [2bones], temporal bones [2bones], occipital bone, the sphenoid and ethmoid bone are part of the floor of the cranium ***[czorny a,et al,2003]***

Superior and Posterior Aspects of the Skull

Viewing the skull from above, the coronal suture is seen separating the frontal, bone from the parietal bones on each side of the midline and the sagittal suture which meets the lombdoid suture diverging on each side of .the occipital bone posteriorly.

The mid-point of the coronal suture, termed the bregma. The inter-parietal or sagittal suture runs from the bregma to meet the lambdoid suture at the lambda. In the infant at birth the bones do not meet at the bregma, but leave a membrane-covered diamond-shaped interval, termed the anterior fontanelle. In the posterior view the occipital bone lies below the lambdoid suture between the parietal bone on each side above and the mastoid region of the temporal bone on each side below. The protuberance on the midline of the occipital bone is termed the Inion. About two or three inches above the inion and usually felt as a slight depression is the lambda ***(Peter L, et al. 1989)***

Lateral Aspect of the Skull

This view is domed by the frontal, parietal and occipital bones; inferiorly the temporal bone, which presents the opening of the external acoustic meatus, rises in its squamous or scale-like art to overlap the inferior border of the parietal and descends in its mastoid part into the conical mastoid process.

Anteriorly, the squama meets the greater wing of the sphenoid, which in turn meets the parietal bone superiorly, and the frontal and zygomatic bones anteriorly. The spheno-parietal suture forms the crossbar of an H-shaped meeting of sutures, in an area termed the pterion. The zygomatic bone, felt prominently in the cheek, juts backwards from the maxilla and sends a process to form with the zygomatic process

of the" temporal bone the bar of the zygomatic arch. Upwards it meets the zygomatic process of the frontal bone.

The temporal fossa is encircled by the temporal line curving backwards into the supramastoid crest of the temporal bone, the upper border reaching the frontal process of the zygomatic bone. This circle marks the attachment of the fascia covering the temporalis muscle that occupies the fossa. Laterally the fossa is bounded below by the zygomatic arch, and medially and below by the infra-temporal crest. The lateral surfaces of the greater sphenoidal wing and the squama turn on to the horizontal basal surface of the skull and form the roof of the infra-temporal fossa. From this infra-temporal surface of the greater wing of the sphenoid the lateral pterygoid plate descends as the medial wall of the infra-temporal fossa while the posterior surface of the maxilla forms the anterior boundary and the coronoid process and ramus of the mandible the lateral boundary. Between the upper border of the posterior surface of the maxilla and the greater wing of sphenoid the cleft of the inferior orbital fissure opens from the infra-temporal fossa into the orbital cavity. The asterion is the term applied to the point where the mastoid part of the temporal bone meets the parietal and occipital bones. The areas at the asterion and pterion are the sites in the infant of the small anterolateral and postero-lateral fontanelles. The supra-meatal triangle, sometimes palpable in the living subject as a slight depression below the supra-mastoid crest and behind the opening of the external acoustic meatus, marks on the surface the site of the mastoid antrum. The narrowest part of the skull anteriorly is at the waist just behind the beginning of the temporal line from the zygomatic process of the frontal bone. From this point a line backwards to the pterion marks the infero-lateral margin of the frontal lobe of the brain. A line along the upper border of the zygomatic arch, horizontal in the natural position of the head, prolonged backwards along the supra-mastoid crest marks the infero-lateral border of the temporal lobe of the hemisphere and the upper limit of the cerebellum ***(Peter L, et al. 1989).***

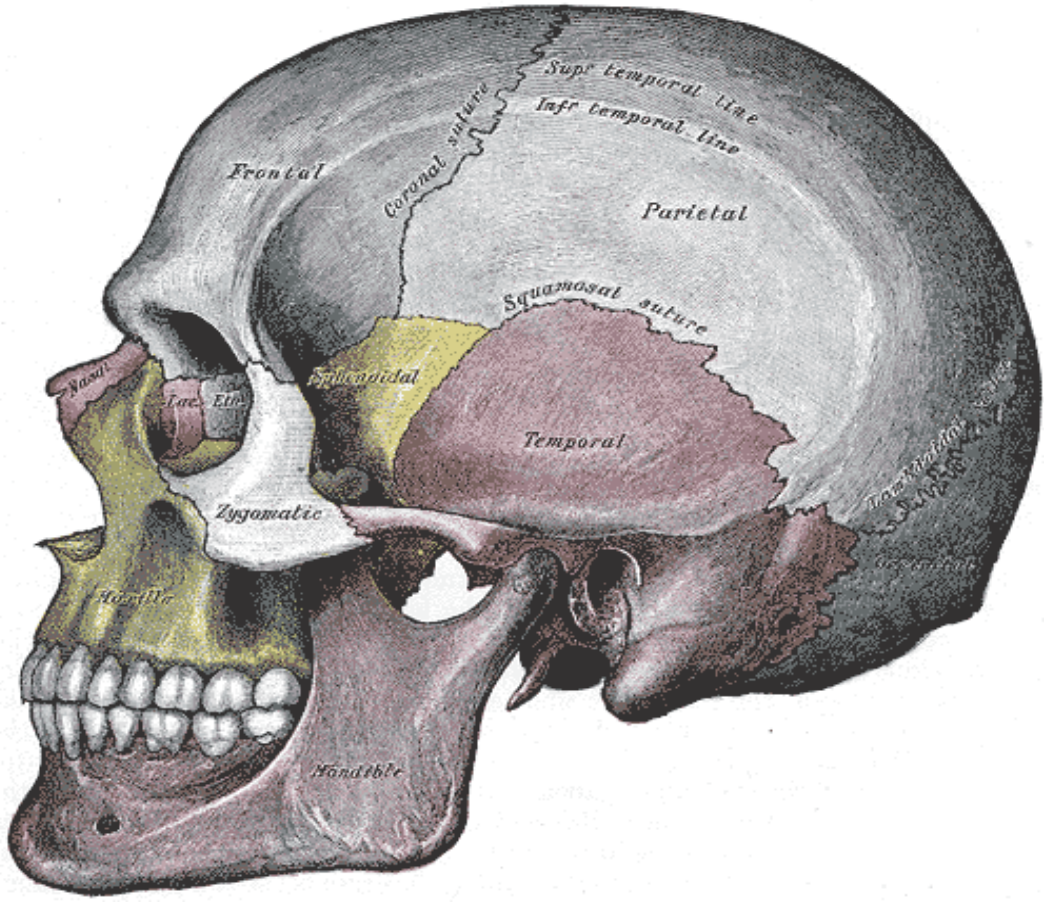

Fig.1: External surface of the lateral aspect of the skull ***(Peter L, et al. 1989)***

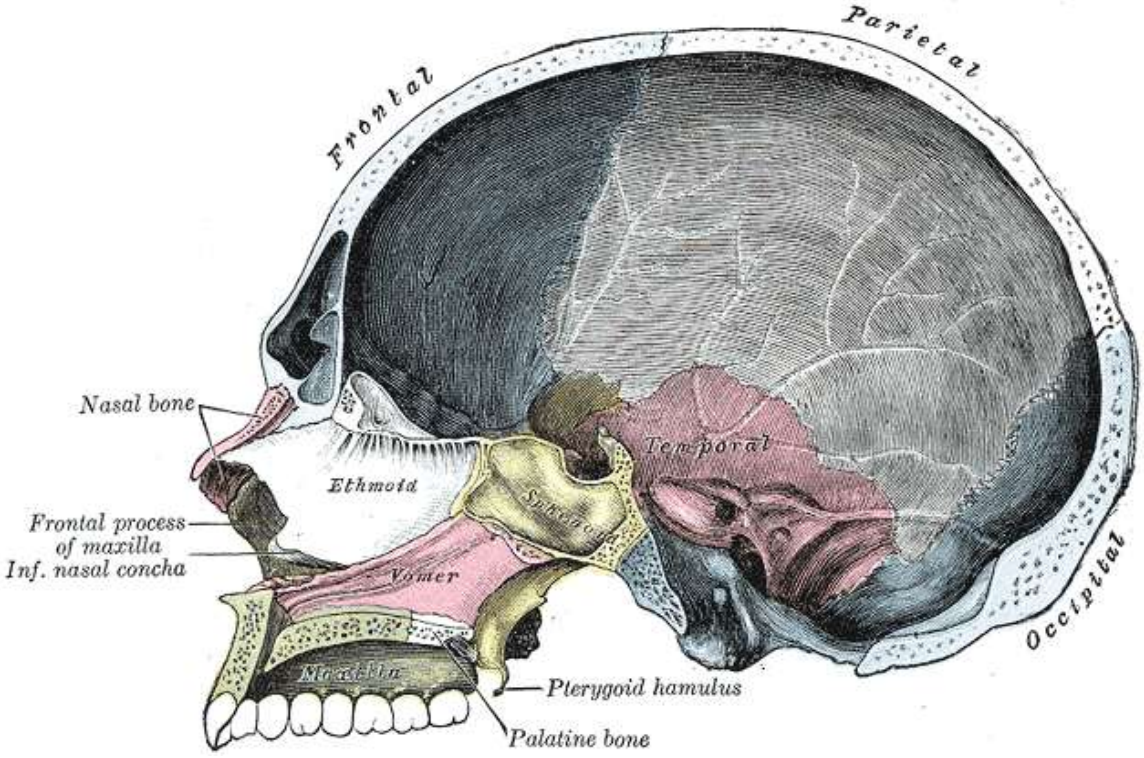

Fig.2: Internal surface of the lateral aspect of the skull . ***(Peter L, et al. 1989)***