Systemic Review and overview about Frontoorbital Advancement in Coronal and Metopic Craniosynostosis

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Conflict of interest: None declared

Funding: No funding sources

Abstract

Craniosynostosis is the premature closure of one or more of the sutures normally separating the infant's skull bony plates, causing abnormal growth of the cranial vault, which may affect brain growth and development. Craniosynostosis can happen as an isolated defect (non-syndromic) or as a part of a syndrome. Computerized tomography plays an important role in the diagnosis of craniosynostosis. Surgical treatment in the form of fronto-orbital advancement and forehead reshaping to correct the supraorbital rim recession and the abnormal form of the cranium. The goal of operative correction is to improve functional and aesthetic outcomes.

Keywords: Frontoorbital Advancement

TobRegul Sci. ™ 2023; 9(1): 5897 - 5905

DOI: doi.org/10.18001 /TRS.9.1.409

Introduction

Craniosynostosis is the premature closure of one or more of the sutures which normally separates the infant's skull bones, leading to abnormal growth of the cranial vault, which may affect growth and development of the brain. The skull growth is restricted perpendicular to the fused sutures while promoted parallel to it, in association with compensatory growth in the skull's unfused bony plates **(Nagaraja et al.,2013).**

The incidence of craniosynostosis is estimated to 1 in 2,000 live births, the sagittal suture is the most commonly affected. Epidemiological studies in Europe and some in the United States have noted a shift in the distribution of the different types of craniosynostosis with a considerable increase in the incidence of metopic suture synostosis **(Gonzalez et al.,2020).**

Craniosynostosis can occur as an isolated defect (non-syndromic) or as part of a syndrome. It is referred to as simple craniosynostosis, when only one suture is involved and as compound craniosynostosis when two or more sutures are involved **(Ferreira et al.,2006).**

The growth of skull bones occurs primarily by the expanding growth of the brain. The brain grows rapidly in utero and during the first 3 years of life. An infant born at term has nearly 40% of his or her adult brain volume. The infant's brain volume increases to 80% by the age of 3 years and by the age of 7 years this increases to 90% of adult brain size **(Kabbani and Raghuveer,2004).**

Craniosynostosis results in deformity of both cranial and facial skeleton. If untreated, it may lead to increase intracranial tension, visual impairment, restriction of brain growth and neurological disorders; with greater functional impairment in proportion to the number of sutures affected **(Zakhary et al.,2014).**

Bilateral coronal suture craniosynostosis Patients characterized by flattening of the forehead, recession and elevation of the superior orbital rim, shortening of anteroposterior diameter of the skull, temporal convexity, skull widening (brachycephaly), and elevation of the height of the skull (turricephaly) **(Adamo and Pollack,2009).**

Anterior plagiocephaly is a premature closure of one of the coronal sutures, characterized by flattening of the forehead and supraorbital ridge ipsilateral to the fused suture, with compensatory bulging of the contralateral forehead. The ipsilateral supraorbital ridge is elevated and recessed with the overlying eyebrow appearing higher than the contralateral side **(Jeyaraj,2012).**

Metopic suture craniosynostosis leads to an abnormal head shape termed trigonocephaly. The deformity is characterized by frontotemporal narrowing, midline forehead prominence, and increased biparietal width in early infancy **(Oi and Matsumoto,1987).**

There are two main indications for the surgical intervention in craniosynostosis; the first is to provide an adequate space for normal brain growth, and the second is to correct the skull shape for aesthetic and psychosocial considerations. The deformities which are associated with craniosynostosis are generally progressive for the first year of life **(Ozgur et al.,2006).**

Fronto-orbital advancement (FOA) is the gold standard corrective procedure for coronal craniosynostosis. The immediate goal of the FOA is to expand cranial volume thereby minimizing damage to the brain. FOA, the entire surgical procedure, aesthetic and functional outcome relies on the operator's experience and is a qualitative method **(Tan et al.,2013).**

Anatomy and development of the cranial vault

The skull is a composite structure made up of the neurocranium, which surrounds and protects the brain, and the viscerocranium, which forms the skeleton of the face. The neurocranium can be subdivided into the cartilaginous part, which form base of the skull , and the membranous part, which forms cranial vault. The cranial vault consists mainly of flat bones: paired frontal and parietal bones; the squamous parts of the temporal bone and occipital bone. All these bones are formed by intramembranous (IM) ossification **(Jin et al.,2016).**

Fontanels are the fibrous membranecovered gaps created when more than two cranial bones are juxtaposed, while sutures are fibrous connective tissue that separate the flat bones of the skull. A newborn has six fontanels as in figure 1 **(Kiesler and Ricer,2003).**

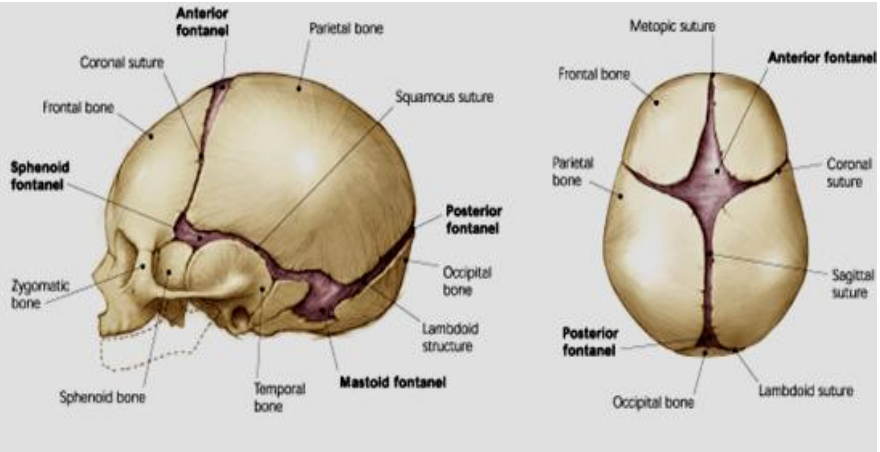


Figure (1) : (Left) Lateral view of the newborn skull. (Right) Superior view of the newborn skull (Kiesler and Ricer,2003).

Aetiopathology of Craniosynostosis

Craniosynostosis can be classified into primary and secondary according to their etiology. In primary craniosynostosis, the premature fusion of one or more sutures is supposed to be due to a developmental error during embryogenesis; the condition may be caused by various genetic mutations **(Apolito et al.,2015).**

In secondary craniosynostosis, the premature suture fusion is due to mechanical causes such as intrauterine compression of the foetal skull against the maternal pelvis, metabolic causes such as hyperthyroidism and the effect of teratogens (**Nagaraja et al.,2013).**

Genetic factors involved in the pathogenesis of syndromic and non-syndromic craniosynostosis include activating mutations in the fibroblast growth factor receptor (FGFR), associated with **Apert, Crouzon, Pfeiffer, Muenke** syndromes) (**Melville etal.,2010).**

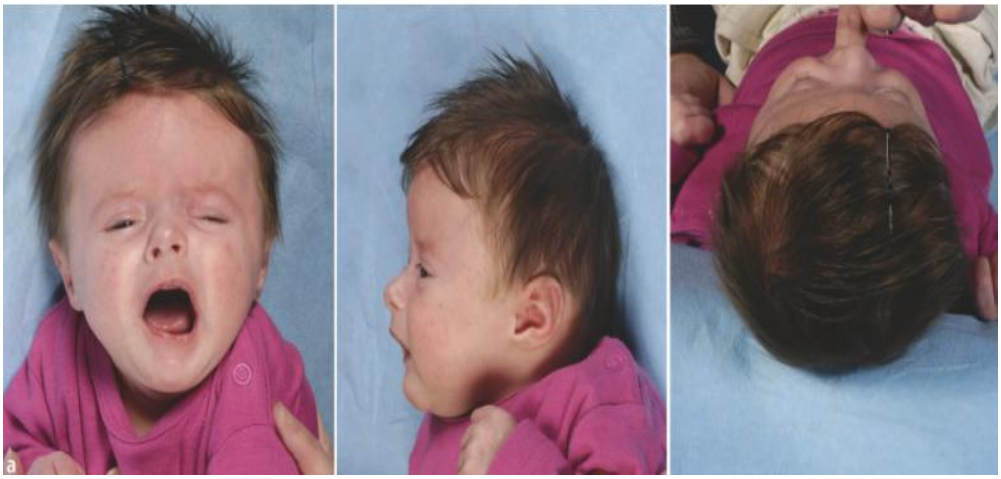
Physical examination is a very important tool in the diagnosis of craniosynostosis. Pre-operative imaging can be helpful to confirm this clinical diagnosis. Computed tomographic scans permit excellent visualization of the underlying bony architecture, helping surgeons appreciate bone deformities and plan surgical correction **(Engel et al.,2012).**

Coronal suture synostosis

- **Bicoronal suture synostosis (Brachycephaly)**

Brachycephaly (bilateral coronal synostosis) is the result of premature fusion of the bilateral coronal sutures and includes typical clinical features such as; a sagittally short and transversely wide skull shape, and elevation of the height of the forehead **(Kronig et al.,2021).**

Figure (2): Picture of patient with bicoronal synostosis. (van Veelen-Vincent et al,2010).



- **Unilateral coronal suture synostosis (anterior plagiocephaly)**

Plagiocephaly means twisted head and is used to describe the skull shape of unilateral coronal synostosis. Unilateral coronal synostosis is accompanied by significant progressive facial deformity if left untreated **(Veelen-Vincent et al.,2010).**

Anterior plagiocephalic skull is characterized by ipsilateral frontal and superior orbital rim retrusion , contralateral bossing of the forehead. The nasal root generally points in the direction of the fused suture; whereas, the nasal tip points in the direction of the unfused suture **(Wu ,2020).**

Figure (3): Picture of patient with unicoronal synostosis (Veelen-Vincent et al., 2010).

Metopic Synostosis (Trigonocephaly)

Trigonocephaly is the skull deformity due to premature closure of metopic suture where, there is palpable midline ridge at the fused suture **(Chandler et al.,2020).**

Metopic synostosis is easily recognized by the triangular shape of the forehead (trigonocephaly) when viewed from above as shown in Figure 4 **(Veelen-Vincent et al, 2010).**

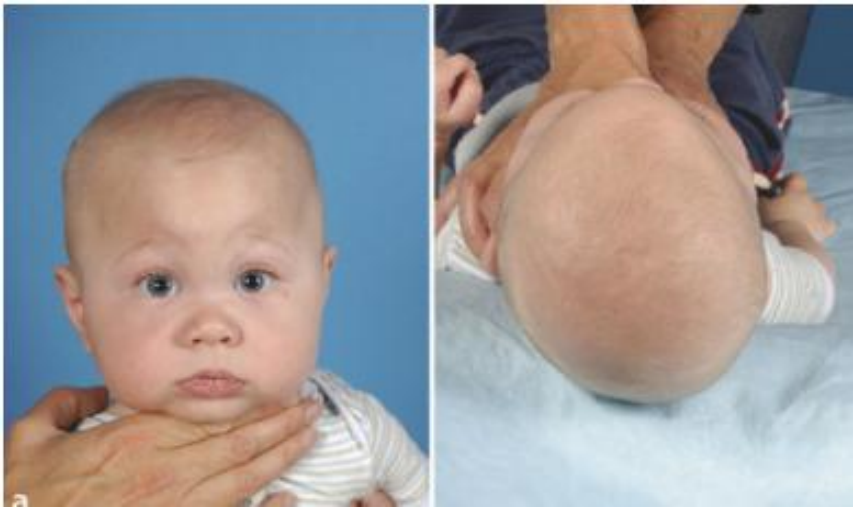


Figure (4): Picture of patient with metopic synostosis (van Veelen-Vincent et al,2010).

The Role of Imaging in the Diagnosis of Craniosynostosis

The newer generation of CT scanners allows reconstruction of images in coronal, sagittal, and oblique planes from a single set of axial scans. These computer-generated images are described as reformatted. **Marsh and Gado** described an oblique image reformatted along the plane

connecting the apex of the orbit and the center of the globe and have named this image as longitudinal orbital projection. Normally, the corneal surface is tangent to a line extending between the midpoint of the superior and inferior orbital rims. The longitudinal orbital projection can demonstrate the relationship of the eyes to the orbital rims **(Marsh and Gado,1983).**

Different methods can be applied for prenatal diagnosis of craniosynostosis, such as two-dimensional (2D) and three-dimensional (3D) ultrasound, magnetic resonance imaging (MRI), computed tomography (CT) scan. The most important consideration in the prenatal ultrasound assessment of craniosynostosis is the distinction between isolated and syndromic craniosynostosis. Therefore, it is extremely relevant to detail fetal anatomy as a whole. Special attention should be paid to fetal hands and feet, central nervous system and heart **(Helfer et al.,2016).**

It is generally accepted that computed tomography with three-dimensional reconstruction optimally evaluates the presence of sutural involvement and its degree. Also, it is useful in assessing associated facial and intracranial abnormalities **(Apolito et al., 2015).**

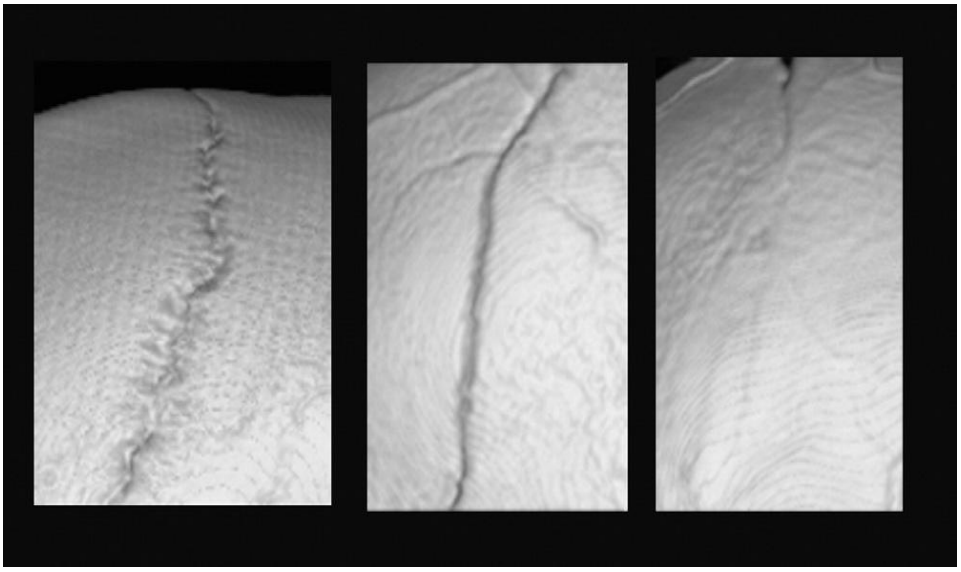


Figure (5): Fusing sutures. 3D-CT: a normal suture (left), a fusing suture (middle) which has lost its architecture, and finally bone bridging of a fused suture (right) (Branson and Shroff,2011).

Treatment of craniosynostosis

Non syndromic craniosynostosis can present in varying severity of the deformity of the skull. Surgical treatment depends on the basis of: the associated risk of increase intracranial tension , the prevention or restriction of associated neurologic and morphologic abnormalities, with both aesthetic and functional consequences **(Mathijssen,2015).**

The ideal time frame of 4 to 13 months has been suggested to take advantage of the infant skull's regenerative capabilities **(Bruce et al., 2018).**

Treatment of coronal and metopic craniosynostosis includes the use of fronto-orbital advancement and forehead reshaping to increase the cranial volume, improve forehead aesthetics, andnormalize the relationship of the supraorbitalrim to the eye **(Alex et al.,2019).**

Recent trend in craniofacial surgery has been the acceptance of endoscopic suturectomy techniques for single suture synostoses. Early release of the affected suture in the first 3 to 4 months of life when combined with either spring-mediated suture expansion or postoperative helmet therapy has resulted in morphologic improvement and has also decreased transfusion requirements, intensive care unit admissions, and hospital length stay **(Alex et al.,2019).**

In open cranial vault reconstruction abicoronal incision was made and bifrontal craniotomies were turned with the assistance of a neurosurgeon. An orbital bandeau was fashioned. Barrel stave osteotomies were carried out in the parietal and temporal regions. The forehead and superior orbits were then reconstructed using the craniotomy bone flaps and orbital bandeau, then fixated with resorbable plates and suture **(Badiee et al., 2022).**

No Conflict of interest.

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