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# Craniosynostosis: Surgical Management And Outcomes

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***Abstract:***
*Craniosynostosis is a condition resulting from premature fusion of the cranial sutures, leading to skull deformities and potentially increased intracranial pressure. We report our experience in managing 112 cases of craniosynostosis over a period from 2013 to 2019, with surgical techniques including open calvarial reconstruction for various disease presentations. Operability criteria include functional and cosmetic impairment, as well as increased intracranial pressure.*
*Postoperative outcomes are generally satisfactory, with marked aesthetic improvement and resolution of intracranial pressure in most cases. A multidisciplinary approach is crucial for evaluating and treating patients, involving specialists from various medical and surgical fields. Management depends on the type of craniosynostosis, with a variety of surgical techniques available. Close collaboration with patients' families is essential for making treatment decisions. In conclusion, craniosynostosis surgery aims to provide satisfactory aesthetic outcomes while preserving patients' functionality and neurological development.*
***Keywords:*** *Craniosynostosis, plagiocephaly, scaphocephaly, brachycephaly, cranial remodeling.*

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## I. Introduction:

Craniosynostosis is a condition characterized by premature fusion of one or more cranial sutures. The overall incidence for all forms of craniosynostosis is 1 in 2,000 to 1 in 2,500 live births. Sagittal synostosis is the most common, with an incidence of 1 in 5,250. According to various studies, the cumulative prevalence of craniosynostosis has significantly increased without obvious cause, resulting from abnormal and non-physiological suture fusion; it manifests as an abnormally shaped skull and, in severe cases, increased intracranial pressure (ICP), as well as sensory, respiratory, and neurological dysfunctions. We report our experience in managing 112 cases. All our patients underwent 3D CT scans. An MRI was performed for each syndromic craniosynostosis operated during the period (2013-2019). Cases with increased ICP who underwent decompressive craniectomy and syndromic craniosynostosis were excluded from our study.

## II. Methods:

A study involving 112 cases of craniosynostosis operated on during the period from 2013 to 2019. Among them, 102 cases (93%) were single suture synostoses of the cranial vault: unilateral coronal synostosis (31 cases), sagittal synostosis (52 cases), bilateral coronal synostosis (12 cases), metopic synostosis (07 cases). There were also 7 cases (4%) of Apert syndrome (acrocephalosyndactyly) and 03 cases (2%) of Crouzon syndrome. Cases with increased intracranial pressure (ICP) who underwent decompressive craniectomy were excluded from our study.

**Operability criteria:** Functional and preventive cosmetic impairment for infants diagnosed early. Cosmetic impairment (delayed diagnosis). Increased ICP (06 cases). All infants underwent remodeling technique. An anesthesiologist for all patients, 70% of infants were transfused intraoperatively (40 cases), 06 patients required additional postoperative transfusion. The average length of hospital stay was 4 days. Surgical treatment is generally undertaken shortly after diagnosis.
Current surgical methods include open calvarial reconstruction: sagittal synostosis (scaphocephaly).

Surgical objective: Anteroposterior shortening of the skull and remodeling of the parietal bones for a more rounded and convex shape. Correction of frontal protrusion or bulging of the occipital bone was not performed in children when parents accepted the deformation.(figure01)

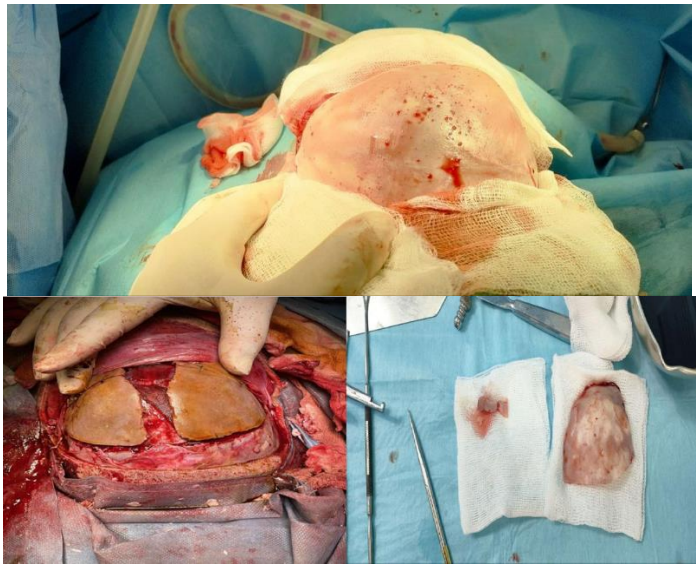

**Figure01 : Perioperative images**

**For cases of delayed diagnosis of sagittal synostosis:** If there is increased intracranial pressure (ICP), decompressive craniectomy is performed. In instances of significant deformation, when parents decline early correction of the deformity, follow-up includes clinical and ophthalmological evaluations.

The surgical technique involves performing an H-shaped osteotomy on the parietal bones and adjusting them with lateral barrel osteotomies in cases of large depressions. The midline sagittal ridge is then separated with two paramedian neosagittal cuts, and the sagittal ridge is divided into smaller segments, which are held in place using fibrin glue.**(figure02)**

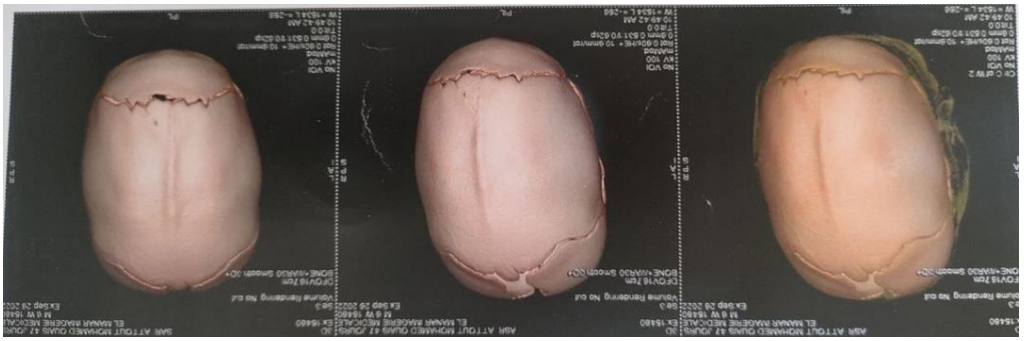

**Figure02 : Cerebral CT scan showing scaphocephaly**

**Brachycephaly:** Global fronto-orbital advancement. Reconstruction is secured on the nasal bone and lateral orbital rim (floating front). Posterior expansion was initially performed in some cases of significant cranial vertex elevation.

**Metopic synostosis (trigonocephaly):** Complete bilateral fronto-orbital reconstruction with remodeling of a neo-frontal bone; The supraorbital rim is opened medially before being reconstructed with the frontal bone; Reconstruction is secured on the nasal bone with Vicryl, the lateral part of the frontal bone is reconstructed to widen the bitemporal diameter.**(figure03)**

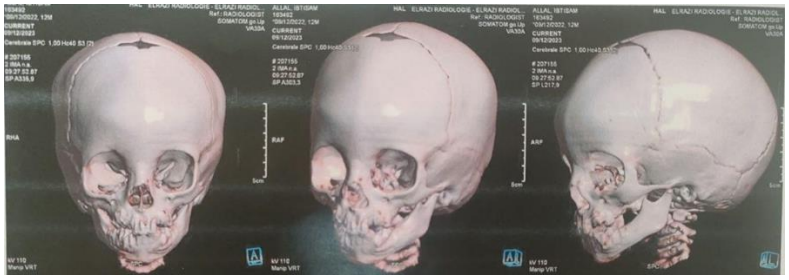

**Figure03 : Cerebral CT scan showing trigonocephaly.**

**Unilateral coronal synostosis (plagiocephaly):** We separately addressed the two components of the forehead (supraorbital rims and frontal craniotomy) using two remodeling techniques (unilateral, bilateral). Bilateral remodeling of the supraorbital rims (elevated, profiled, forward-oriented in an advanced position) yielded the best results; however, psychological issues were observed in infants operated on late +++ (2 cases).**(figure04)**

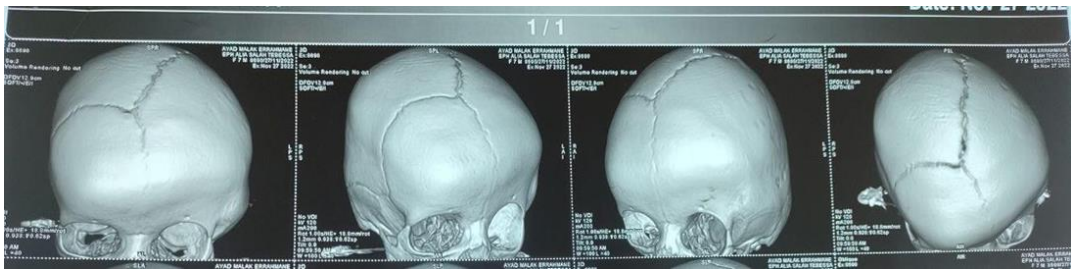


**Figure04 : Cerebral CT scan showing plagiocephaly**

## III. Disscusion :

Craniosynostosis results from the premature fusion of cranial sutures. These sutures exist to facilitate the passage of the baby through the birth canal and subsequently allow for the expansion and growth of the brain. When one or more sutures close prematurely, the structure of the skull is altered, developing along the path of least resistance (perpendicular to the closed suture) and resulting in an atypical skull shape leading to increased intracranial pressure (ICP) and affecting the respiratory and neurological systems, as well as the child's development.**(04)**

Craniosynostosis can be classified as simple when it involves a single suture and complex when it involves multiple sutures. Another popular classification is syndromic (Apert, Crouzon, Pfeiffer) and non-syndromic with isolated discovery.(06) The prevalence of craniosynostosis is 1 in 2000 to 1 in 2500 live births. It has increased over time, with predisposing factors being either environmental (maternal smoking, in utero exposure to teratogens, intrauterine constraint, fetal position) or genetic (mutations). Close to 20% of all craniosynostoses are due to genetic causes, with most being inherited in an autosomal dominant manner, although new mutations occur in 50% of cases. Non-syndromic craniosynostosis occurs in 75% of cases, and 25% are syndromic craniosynostoses.**(16)**

As with any other condition, a thorough history and physical examination are most useful in determining the diagnosis. When taking the history, it is vital to know if there is a family history of abnormal head shapes, in utero exposure to teratogenic drugs, intrauterine constraints, or abnormal fetal position, as well as any complications during pregnancy and any delays in developmental milestones. The physical examination allows the clinician to assess whether suture fusion is present and if concomitant features would make craniosynostosis an integral part of a syndrome,**(08)** such as any congenital anomalies and dysmorphic features. Although the diagnosis is clinical, many professionals will require radiological imaging to further evaluate and confirm the diagnosis. The most accurate means is computed tomography with 3D reconstruction, where all sutures are assessable, but due to the risk of radiation, this option requires careful consideration. Plain radiographs are inexpensive; therefore, they are useful for evaluating infants at low risk of craniosynostosis, but they are not sufficiently accurate. MRI is less accurate compared to CT, but it is still an excellent method to use, usually reserved for children in whom CT has revealed brain anomalies.

In the management of craniosynostoses, a holistic and multidisciplinary approach is imperative to provide patients with comprehensive care tailored to their needs. Preoperative assessment is crucial as it must address a multitude of questions from various domains,**(11)** including pediatric, aesthetic, neurological, psychological, anesthetic, genetic, and social. To begin with, it is essential to fully understand cranial dysmorphia by carefully analyzing clinical and paraclinical morphological elements. This allows for an accurate diagnosis and determination of the best treatment strategies. In parallel, evaluating the functional impact on the child's overall development is crucial to ensure appropriate surgical intervention and optimal long-term outcomes. Additionally, screening for potential associated malformations, particularly cerebral ones, is an essential step to anticipate complications and adjust management accordingly. This approach also helps classify the malformation syndrome, guiding treatment planning andcommunication with the patient's family.**(14)**

A comprehensive multidisciplinary team is therefore necessary to effectively address all aspects of craniosynostosis management. This team may include various healthcare professionals, including craniofacial specialists such as plastic surgeons specializing in cranial reconstruction, neurosurgeons experienced in neurological interventions, and anesthesiologists skilled in pain management and anesthesia. Furthermore, an assessment group composed of geneticists, pediatricians, psychologists, and neuroradiologists is needed to thoroughly evaluate the medical, genetic, and psychological aspects of the condition**(03)**. Facial specialists, such

as ophthalmologists to assess vision, maxillofacial surgeons to treat facial and jaw anomalies, otolaryngologists to manage auditory and respiratory problems, and orthodontists to correct dental issues, complement the team for comprehensive and specialized care.

Additionally, other professionals, such as photographers to document treatment progress, anthropologists to study morphological aspects, and illustrators to assist in surgical planning and communication with patients and their families, can also contribute to comprehensive and personalized care.**(02)**

Craniofacial surgery to correct craniofacial dysmorphologies in children with craniosynostoses requires intracranial exposure of the bone structures. This surgical intervention, often bilateral, presents particular challenges, especially in infants**(04)**. Furthermore, secondary surgery in children already operated on for their craniosynostosis poses additional challenges. The bone flaps, usually located on each side of the midline, are cut from trephine holes. Their location must be carefully chosen to allow optimal detachment of the dura mater before osteotomies and to respect the areas of the vault that will be used during subsequent reconstruction. The selection of drilling sites depends on the type of malformation and the planned intervention. However, several difficulties may arise, especially in infants.

In infants, particular care is needed during the dissection of the bregmatic fontanelle, where the periosteum is continuous with the dura mater**(01)**. Additionally, detaching the dura mater can be difficult due to adhesions to functional sutures. In some complex cases of craniosynostoses, the internal table of the vault may be deeply altered, increasing the risk of dura mater tear. At all ages, it is important to consider the anatomical variation of the dural sinuses and to plan preoperative exploration by CT or MRI. Any accidental meningeal opening must be carefully sutured to prevent postoperative complications such as infections and to promote reossification.**(19)**

As for dura mater suspension to prevent postoperative extradural blood collections, this rule is not always applicable due to the frontal advancement performed during the intervention, which moves the bone away from the meningeal plane. This creates a potential dead space prone to hematomas and infections, although such complications are rare. In summary, craniofacial surgery for craniosynostoses requires careful attention to anatomical details and meticulous planning to minimize risks and ensure good postoperative outcomes.**(06)**

A collaborative and multidisciplinary approach is essential to provide optimal care for craniosynostoses, considering all medical, surgical, genetic, psychological, and social aspects of the condition. This ensures the best possible outcomes for patients and their families, providing comprehensive support throughoutthe treatment process. The management of these patients depends on the type of craniosynostosis.

Uncomplicated and non-syndromic types can be surgically treated but electively compared to some syndromic forms that require urgent surgery due to involvement of the respiratory, ophthalmologic, and neurological systems.**(09)** A conservative approach with remodeling helmets may be attempted initially in cases where unilateral craniosynostosis is not too severe. Differential Diagnostics Positional plagiocephaly requires differentiation from craniosynostosis**(18)**. It does not involve premature suture fusion. It presents with a parallelogram-shaped head, ipsilateral anterior displacement of the ear and head, ipsilateral occipital flattening with contralateral occipital bulge. The prevalence has increased over

## IV. Conclusion:

Surgery for single-suture craniosynostosis yields satisfactory morphological outcomes compared to syndromic craniosynostosis, attributed to its less complex craniofacial involvement. However, late diagnosis remains a challenge, affecting a small number of cases.

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