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REVIEW ARTICLE

Craniosynostosis: Overview Update

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Craniosynostosis is a congenital condition when one or more of the baby's skull's sutures close earlier than usual. The premature result of a suture closing is an unnaturally formed head. Each year an estimated 84,665 and there are 72,857 non-syndromic craniosynostosis cases among the newborns with craniosynostosis. [1]. Further studies are needed to delineate prevalence in low & middle income countries (LMICs). This information would be needed to upscale training of personnel & augmenting necessary infrastructure [1].

The cranial vault sutures are distinctive, but other than that, the brain growth could be pathologically restricted with irreversible damage pathologically caused to it as the intracranial pressure increases the sutures were previously believed to be active growth sites that force the cranial plates apart, but further research has revealed that their function is passive. The neonatal brain doubles up in size in 6 months and quadruples by the 1st year of life, acquiring 80% of its adult size by 2nd years of age. Patent cranial sutures allow this rapid expansion with a minimal pressure of approximately 5 mm Hg provided by the growing brain. Bone is deposited at the borders of the sutures and the epicranium in response to the rapidly expanding brain, while resorption happens along the dural surface [2,3].

The cranial sutures are typically patent at birth and develop into a yielding fibrous union in the first year of life, allowing appositional bone growth to continue. The cranium reaches 90% of its adult size by the age of 6, although complete solid sutural bony union does not happen until at least the fifth decade. Virchow's law states that if a suture closes too soon, growth is stopped in a direction perpendicular to the fused suture, while compensatory expansion takes place in a plane parallel to the affected suture. Depending on which suture or sutures are closed, the subsequent deformity of the skull will occur [4].

There are significant hereditary and long-term growth implications for the two types of craniosynostosis. The non-

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syndromic fraction of malformations typically occurs infrequently, but the syndromic subset typically has a hereditary component. Additionally, normal craniofacial development is the norm following surgical correction for the non-syndromic patients, whereas those with syndromal attributes do not fare so well.

Epidemiology of craniosynostosis

With particular reference to low- and middle-income countries (LMICs), according to one estimate [5], 23,300 additional neurosurgeons are necessary to address about 5 million essential neurosurgical cases, hitherto, remaining attended. It needs to be recognized, that, neurocognitive impact of Craniosynostosis could be as significant while priority remains to be given to illnesses include hydrocephalus, tumor, and stroke [6-10].

There paucity of trained manpower with only a handful of centres offering the expertise of this surgery in LMICs [11]. There is a need for early recognition of this condition by pediatricians followed by early intervention by trained pediatric surgeons [12-14]. Surgical workforce in managing craniosynostosis could be expanded by creating centres of excellence, particularly in LMICs where the disease burden is profound.

Nonsyndromic Craniosynostosis

One in every 1000 live infants is reported to have nonsyndromic premature stenosis of the cranial sutures. These statistics cover all single suture fusion types, with unilateral coronal, metopic, and lambdoidal areas most frequently impacted and the sagittal being the least frequently affected. Multiple sutures are also involved, as well as bilateral fusion of the coronal.

Based upon the suture involved, the following deformities present clinically:

Scaphocephaly (Boat shaped)

The sagittal suture is either partially or completely fused; when fusion is complete, there is frequently an exterior bony overgrowth that extends from lambda to bregma in the midline. On the interior surface, overgrowth might appear occasionally (Fig. 1). X-ray of the skull in an anteroposterior (AP) view shows sagittal suture fusion (Fig. 2).

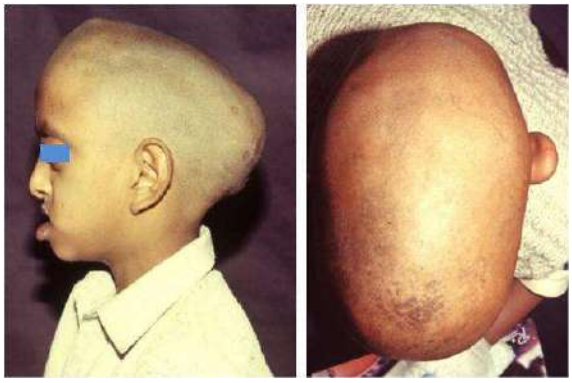

Fig. 1. Scaphocephaly – Boat Shaped

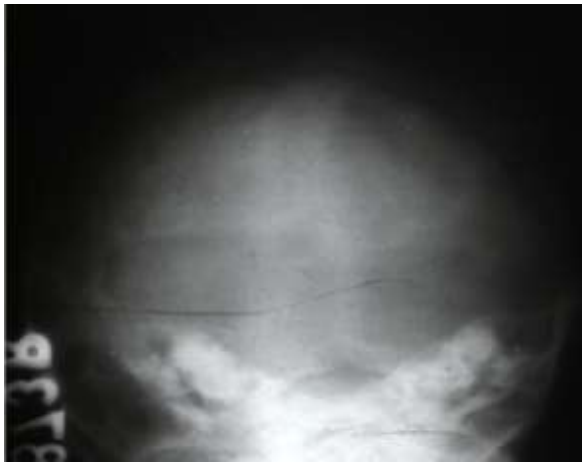

Fig. 2. X-ray – Sagittal suture ossification – Scaphocephaly

Plagiocephaly (Unilateral coronal suture fusion)

The lateral section of the damaged coronal suture exhibits synostosis. The

suture's medical end is typically patent in early newborns. The synostosis is typically entire and reaches the anterior fontanelle, which may be modest or even absent, in cases where the patient is older than 6 months. The suture may then leave no visible external traces or appear as a shallow groove in the bone that resembles a constriction ring (Fig. 3). Sutural fusion shall seen on a plain X-ray or a 3-D CAT scan of the skull (Fig. 4).

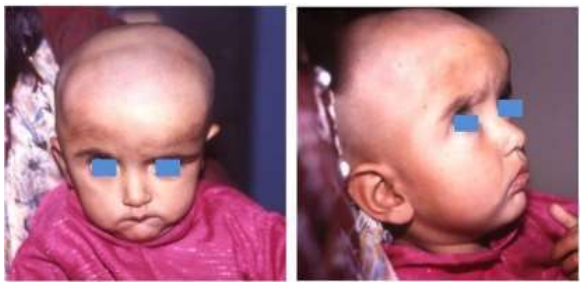

Fig. 3. Unilateral coronal suture ossification – Plagiocephaly

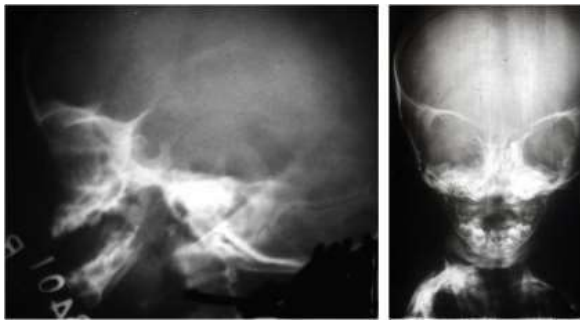

Fig. 4. X-ray – Unilateral coronal suture ossification — Plagiocephaly

Brachycephaly (Short-bilateral coronal suture fusion)

When both coronal sutures are involved the forehead becomes fore-shortened anterior-posteriorly (Fig. 5). The diagnosis can be confirmed by X-ray skull (Fig. 6) and 3-D CAT scan.

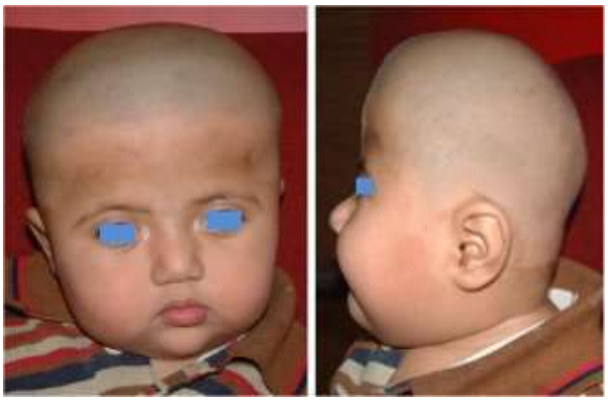

Fig. 5. Bilateral coronal suture ossification – Brachycephaly

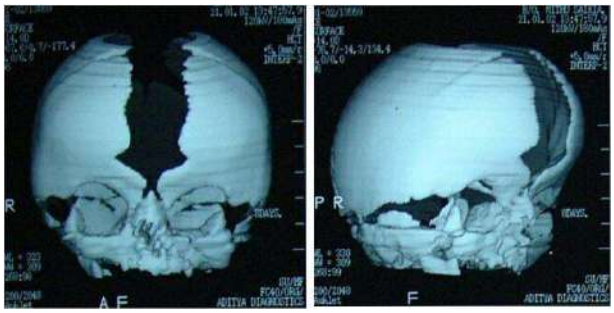

Fig. 6. Bilateral coronal suture ossification – Brachycephaly – 3-D CAT scan of the skull

Trigonocephaly (Triangular-metopic sutural fusion)

Premautre The metopic suture can fuse completely or not at all in the area around the anterior fontanelle, where it is typically most noticeable above the glabella. On the inner surface of the frontal bone near the lower end of the synostosis, there is frequently a noticeable bony outgrowth. (Fig. 7). X-ray of the skull (Fig. 8) or 3-D CAT scan is unmistakable.

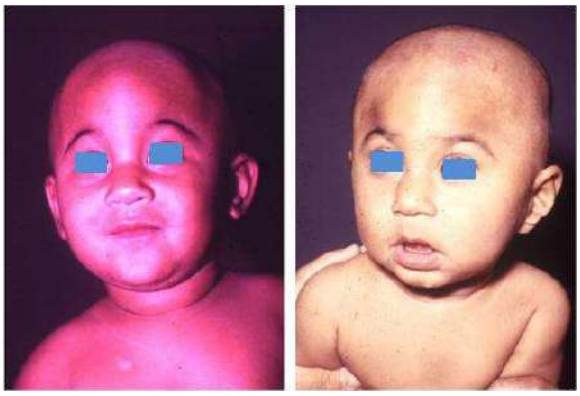

Fig. 7. 3D Cat Scan — Trigonocephaly (Triangular-metopic suture ossification — Trigonocephaly

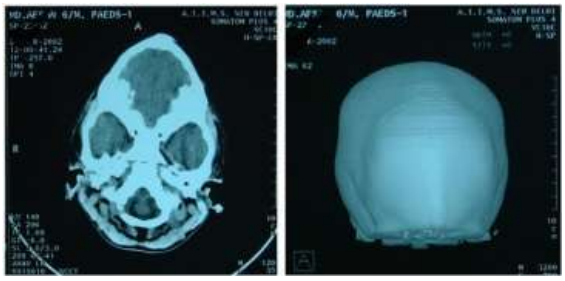

Fig. 8. 3D Cat Scan — Trigonocephaly (Triangular-metopic suture ossification— Trigonocephaly

Syndromal Craniosynostosis

In contrast to nonsyndromic fusions, syndromal craniosynostosis behave to these deformities differ from one other, have genetic ramifications, and an established pattern of inheritance. The most prevalent syndromal craniosynostosis anomaly, Crouzon's syndrome, with an estimated prevalence of up to 1 per 10,000 live births. The autosomal dominant inheritance pattern has almost full penetrance. [15].

Crouzon's Syndrome

Mid-face hypoplasia, parrot beak nose, lateral canthal dystopia, hypertelorism (Broad set orbits) and relative mandibular prognathism characterize the defect. High arched palate is also an associated finding. Although other head forms are seen, brachycephaly is the predominant skull malformation. Exorbitism is a serious warning. It may endanger sight and is frequently extremely prominent.

Apert's Syndrome

Also termed as acrocephalo syndactyly, the incidence of this anomaly has been estimated to be 1 in 1,60,000 live births. As with Crouzon's syndrome, exorbitism, midfacial retrusion with maxillary constriction, and pseudomandibular prognathism are present. Exorbitism is more asymmetrical and modest. The face also is transversely flattened. The distance between the orbits is greater. The cranial Premature fusion of the coronal sutures is nearly always linked to deformity, however the base of the skull is also affected. The severity of the syndactyly, which entails bone fusion of the phalanges of at least the index, middle, and ring fingers, separates the Apert's syndrome from other acrocephalosyndactylies. (Fig. 9). Autosomal dominant is the method of inheritance, albeit sporadic incidence is more common. Similarly to Crouzon's condition, a hereditary flaw has been identified [2].

Fig. 9. Apert's syndrome: The differentiating feature from other acro-cephalo-syndactyliesis highlighted by the severity of the syndactyly. There is bony fusion of the phalanges of the index, middle, and ring fingers

Others

More than 50 syndromes addition to the most prevalent Crouzon's and Apert's syndromes, there have also been described conditions connected with craniosynostosis. They are rare in number, therefore it is difficult to determine how frequently they occur. These syndromes include Pfeiffer's, Carpenter's, and Saethre-Chotzen Syndromes. Pfeiffer's is characterised by large toes and thumbs, Carpenter's by preaxial polysyndactyly and soft tissue syndactyly with shortened fingers, and Saethre-Chotzen by soft tissue webs between the second and third digits. These malformations are also with the exception of Carpenter's syndrome, which is autosomal recessive, all diseases are autosomal dominant. It is difficult to forget the unusual Kleeblattsschadel (cloverleaf skull) abnormality. The trilobular form of the skull cap features a projecting vertex and enlarged temporal regions (see Figs. 10 and Fig. 11 X-ray imaging).

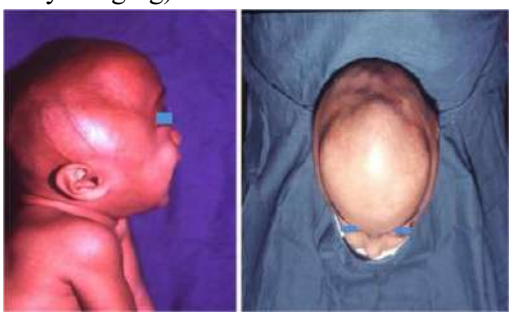

Fig. 10. Cloverleaf skull — The skull cap assumes a tri-lobular configuration with a protruding vertex and bulging temporal regions.

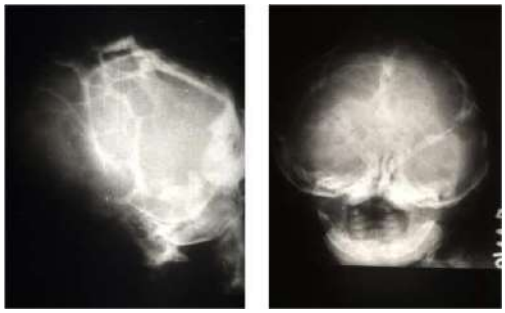

Fig. 11. X-ray — Cranio-lacunae & Cloverleaf skull

Multiple Suture Involvement

Oxycephaly or Turricephaly

The term "oxycephaly" refers to a condition in which the head is excessively high and conical. Whether seen from the front or the side, the vertex seems pointy in a case of typical oxycephaly. The coronal suture may also be involved in the deformation process, depending on which area of the cerebral capsule is initially affected.

Cloverleaf Skull

This is a constriction ring forms in the lambdoid-squamosal zone as a result of multiple sutural fusion, which permits disproportionate bulging in the frontal and temporal bones, resulting in deformity (triphyllocephaly). From the front, the head appears to trilobular (Fig. 10). There is almost always accompanying hydrocephalus Less severe variations of this syndrome may look pointedly oxycephalic.

Diagnosis

Diagnosis of craniosynostosis clinical examination in the majority of cases due of the distinctive deformity of the skull cap and indications of a ridge generated by the fused suture on palpation. Plain X-rays of the skull in different views should be performed in order to support the clinical impression. A radiographic linear opacity takes the place of a patent suture's wormian lucency. On a postero-anterior skull film, a characteristic "Harlequin" eye sign for unilateral coronal synostosis is created by the elevated ipsilateral lesser wing of the sphenoid. Typically, computed tomography (CT) scans are not diagnostically helpful unless reformatted in three-dimensionally. Detailed psychological assessment, ophthalmic evaluation for evidence of raised

ICT and dental examination should be carried out.

Differential Diagnosis

Positional distortions can be caused by muscular torticollis, cervical spinal abnormalities, and preferred sleeping postures. In these circumstances, the sutures are visible. Positional dyskinesia should be treated primarily for its underlying deformities. (Table 1)

Table 1: Differential diagnosis of craniosynostosis

- Primary microcephaly
- Hydrocephalus-sutural fusion secondary to decompression and overlapping of bones
- Postural Plagiocephaly

Table 2: Investigations

- Skull radiograph-Antero-posterior, lateral, Towne's and basal views
- CAT scan-3 dimensional
- Ophthalmological examination
- Psychological assessment
- Genetic evaluation
- Dental evaluation
- Clinical photograph

Posterior position when viewed from the vertex, a plagiocephaly features an obliquely oriented skull cap and an affected side ear that is pushed anteriorly and away from the flattened posterior side. True unilateral lambdoidal synostosis, on the other hand, results in a trapezoid deformity and draws the ipsilateral ear toward the damaged suture. It is critically important to distinguish positional deformations from those caused by true craniosynostosis.

Investigative workup has been summarized in Table 2.

Computer-aided design and manufacturing (CAD/CAM) technology

To accomplish a consistent, objective, and accurate correction of CS, Computer-aided design and manufacturing can be utilized. It helps in detailing and adds precision in realizing osteotomies which enable accurate positioning of bone segments, thus, eliminating subjectivity & allowing more reproducible surgical results [17,18]. This technology helps in identifying the precise location of dural venous sinuses during the surgical steps for Cranial surgery, such as appropriately plan burr holes and cuts& enhancing superior outcomes [19,20, 21].

Virtually Surgery Planning (VSP)

The surgical team is able to use VSP to achieve more predictable results and prepares the families for anticipating the postoperative results[17]. Devising VSP based, pre-surgical planning offers a near realistic visualization of the anatomy in a controlled environment.

3D printing/CNC machining

Adding further precision during surgical planning & its subsequent execution, 3D printing/CNC machining further help in eliminating anypresumption [17 21,22].

Resorbable plating system

Providing stable osteosynthesis, ultrasound-assisted, pinned resorbable plating system also saves time.[23]Resorbable plates are safe& stable with superior aesthetics & allow unhindered ongoing ossification which continues in the frontal area [24,25].

3D photogrammetry

3D photogrammetry/photography avoids radiation and is reproducible, thus,

minimizing operator dependent variation & errors [26,27,28].

3D simulation

Computer-aided simulated surgery& intraoperative navigation reduces operative time and facilitates surgery.[29]

Surgical Management

Non-syndromic craniosynostosis should be corrected because of the severe distortion the face is reflected. Other explanations discuss potential functional issues that the illness may bring about, all of which are related to increased intracranial pressure. Thirteen percent increased pressures in neonates with single suture synostosis, and 42% of those have multiple sutures involved. Visual impairment is possible but unusual when only one suture is united too soon.

Before the era of a linear strip craniectomy of the pathologic suture was the historically established form of treatment in craniofacial surgery. However, the rate of refusion and ongoing deformation was regrettably high.

Early intervention is encouraged to benefit from the infant brain's rapidly increasing development. Most centres advocate the window of time between 3 and 6 months old; some wait until after the sixth postnatal month. Early correction produces superior results than late intervention in the long term.

The concepts of treatment have constantly changed as a result of failures with strip craniectomy procedures. The focus has been on releasing the afflicted suture and promptly restoring the normal architecture by realigning and recontouring the malformed bones, regardless of the type of craniosynostosis. The prevalent surgical management globally and AIIMS technique (Table 3).

Table 3. Surgical management

Surgical Management	
Primary	**AIIMS Technique**
Linear Craniectomy Morcellation Cranioplasty Holfmann's Tongue–in–Groove Technique	Partial Calvariectomy with Onlay Periosteoplasty
Supplementary	
CSF Shunts Mid–Face Advancement	

Partial Calvariectomy and Onlay Pariosteoplasty

Partial calvariectomy and harvesting the periosteum is an alternative technique. in this procedure not only the involved sutures are removed but the adjacent deformed bones are also excised after separating and preserving the periosteum . Towards the end of the procedure this harvesting periosteum is laid down as an onlay over the exposed dure. Subsequently neocalvarium is formed by the periosteum according to the contour of growing brain. (Figs. 13 and 14).

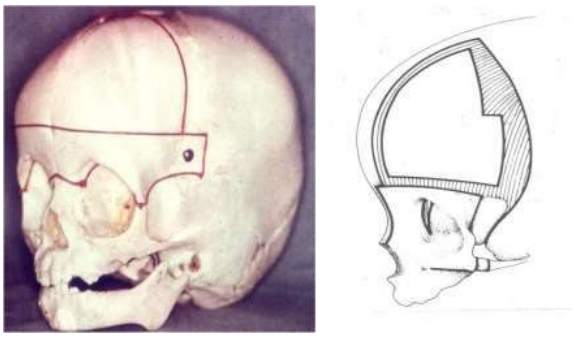

Fig. 13. Partial Calvariectomy, Onlayperiosteoplasty&Fronto-Orbital advancement.

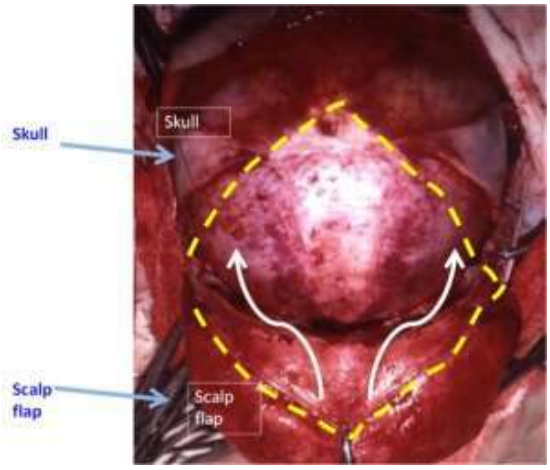

Fig. 14. Author's Technique: Periosteum harvested and placed as 'onlay' on the exposed DURA.

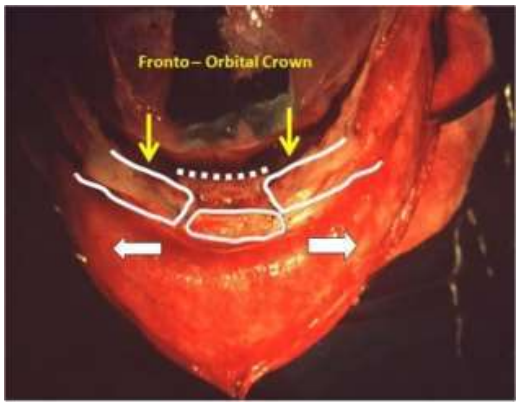

Fig. 15. Author's (MB) Technique: Fronto – orbital crown

All syndromic craniosynostosis abnormalities are surgically treated in one of three chronologic ways. In contrast to the non-syndromic variant in this group, surgical intervention does not result in the normalisation of facial growth. The urgent issue is the focus of treatment. A stepwise method is therefore utilised. Release of increased intracranial pressure brought on by the premature suture closure, airway impairment brought on by significant midfacial retrusion, and corneal protection due to exorbitism are all issues that need to be addressed. Additionally, the synostotic suture is loosened during the first year of life as well as fronto-orbital advancement (Fig 14 and 15). This results in more regular calvarial outlines, more protection for the canines, and a release of the quickly swelling brain.

Depending on the patient's preferences and experience, treatment schedules may change. Around the sixth year of life, when the orbits and skull cap have grown to 90% of their adult size, a second procedure is carried out. The traditional LeFort III midfacial advancement is utilised to advance the entire midface IF the forehead is of normal shape. The maxilla is purposefully positioned in an overcorrected anterior position because there is limited room for further midfacial expansion. This position corrects the exorbitism and unclogs the congested nasopharynx.

Two alternate routes can be taken if the forehead is seen to be sunken, as is frequently the case. A fronto-orbital advancement is done as the first procedure. An additional LeFort III is added around six months later. By separating a sterile intracranial operation from the contamination brought on by the subsequent extracranial midfacial advancement, this method maintains sterility, making it safer.

The other approach is the monobloc frontofacial advancement. The advantage of this technique is that only one operation is required to reposition the forehead.

When orbital hypertelorism is present, a facial bipartition either the frontofacial monobloc advancement or the LeFort III technique can be supplemented. This treatment is very useful for treating people with Apert's syndrome's distinctive flattened forehead and midface.

This last phase needs to wait till facial development is finished. A LeFort I (maxillary) advancement is used to treat the

pseudomandibular prognathism following presurgical orthodontic tooth alignment. Oftentimes, a simultaneous advancement osseous genioplasty correct the retruded chin. The result of pre and early post-operative technique of fronto-orbital advancement author's technique (MB): (Fig. 16a to Fig. 16c).

Figure 16 shows early post-operative result of fronto-orbital advancement.

Fig. 16a. Brachycephaly: Pre & Postop.

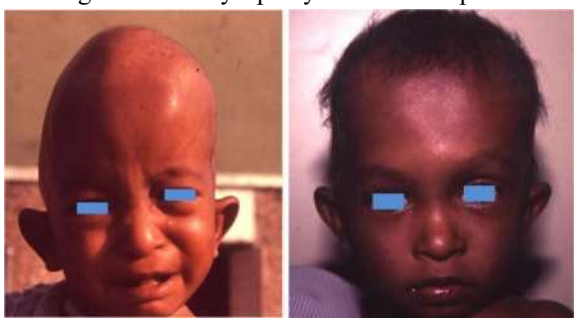

Fig. 16b. Scaphocephaly: Pre. & Postop-front views.

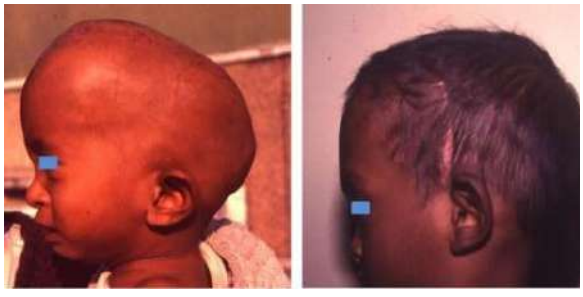

Fig. 16c. Scaphocephaly: Pre. & Postop- lateral views.

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