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CRANIOSYNOSTOSIS

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SUMMARY

A 4 year old female child with Craniosynostosis (Crouzon's syndrome) was operated upon on 10.3.87, during the Ist National Workshop on Plastic Surgery at the King George's Medical College, Lucknow, in collaboration with the department of Neuro Surgery. The details of planning, management and the result of treatment in her is being presented.

Premature cranial synostosis can be divided into simple cranial synostosis and craniofacial dysostosis (Crouzon's and Apert's syndromes). Simple cranial synostosis is more common. Both Crouzon's and Apert's Syndrome are autosomal dominant. Apert's syndrome can be differentiated from Crouzon's syndrome by the presence of syndactyly of hands and feet in the former.

Depending upon the site of cranial suture synostosis, various patterns of cranial deformity (Oxycephaly, Plagiocephaly, Trigonocephaly and Scaphocephaly) may be present.

The maxilla and zygoma are hypoplastic. Due to vertical shortness of maxilla in the central area there may be an anterior open bite in addition to a class III malocclusion. The nose may be small and stubby and the palate may be high arched.

Due to increased intracranial tension secondary changes occur in cranial fossae. The anterior cranial fossa is displaced downwards. The middle cranial fossa is also displaced anteriorly and downwards. Besides hypoplasia of maxilla and zygoma, variable degree of exorbitism is seen in these cases.

Depending upon the severity of deformity, Lefort III advancement osteotomy, Lefort III and Lefort I osteotomy, Lefort III advancement osteotomy and Frontal bone advancement, Tripartite osteotomy, Lefort II osteotomy or other variations of maxillary osteotomy are required for correction of the deformities in these cases.

Case Report

E. T., a four years old female child (Fig. 1, 2) was admitted with the complaints of deformity of skull since birth and recurrent conjunctivitis for the last $3\frac{1}{2}$ years. For recurrent redness of both eyes she was regularly taking treatment from an ophthalmic surgeon. For the last two months, her parents noticed gradually increasing bulge of both eyes and prominent dilated veins around the orbit. For these complaints she was shown to a Paediatrician and was advised an x-ray skull. Anteroposterior and lateral views were done (Fig. 3, 4). She was diagnosed as a case of Craniosynostosis.

The child had normal intelligence. The abnormality of the skull in her was obvious. The head circumference was 55 cm. Both eyes were proptosed. The eyeball movements in all quadrants were normal. Veins around both orbits were dilated and prominent. Intercanthal distance was 28 mm. There was no syndactyly. The child had class III malocclusion.

X-ray skull showed a silver beaten appearance and fusion of the cranial sutures.

Operative Steps

The operation was performed under general anaesthesia using a standard coronal approach. Supraperiosteal dissection was done upto 3-4

cms. proximal to the supraorbital rim (Fig. 5). The pericranium was incised at this level. Pericranium was also incised in the midline and along the coronal incision line. The pericranium and temporal fascia were erased from the skull, laying it bare (Fig. 6). Subperiosteal dissection of superior and lateral orbital rim, temporal fossa and orbital roof was also done. The medial orbital wall was stripped down to the inferior orbital fissure. The dissection was continued over the medial half of the zygomatic arch. Medially the dissection was done upto the pyriform margin. The dissection was then carried posterior to the zygoma on the posterior surface of maxilla upto the region of the pterygomaxillary fissure.

Bifrontal craniotomy 2 cms above the superior orbital rim was performed (Fig. 7, 8). Tongue in groove cuts were performed on either side in the temporal bone and lateral orbital wall. Transverse cut of the frontonasal region and orbital roof cuts were performed. Then the superior orbital rim was removed (Fig. 9, 10). Osteotomy of the orbital wall and floor was then completed. Lateral orbital wall and medial orbital walls were cut. The floor of orbit was cut with an osteotome. The pterygoid osteotomy was done by a curved osteotome passed behind the zygoma. A finger placed intraorally in maxillary tuberosity area confirms the correct pterygoid osteotomy. The zygoma was split in line with the split of the lateral wall. Lefort III osteotomy was completed. The maxilla was brought forward by 1.5 cms using the Rowe's disimpaction forceps. Wires were placed in zygoma, fronto-zygomatic area and frontonasal area. A split rib graft was placed within the zygomatic gap (Fig. 11). Then the superior orbital rim was advanced by 2.5 cms and tongue in groove segment was fixed in place by interosseous wiring. The frontal bone and anterior portion of the parietal bone was placed anteriorly and wired firmly. In the bony deficit of the parietal region a layer of surgicel was laid and over it bone dust collected at the time of making burr holes was spread (Fig. 12). The gap in bone was bridged at three places by split rib grafts. The medial and lateral canthal ligaments were re-attached in place at a slightly higher level by wires. The bicoronal flap was reposited back and stitched in place (Fig. 13, 14). Two units of blood were transfused pre-operatively. Patient stood the operation well which took about 5 hours.

Post-operative Period

During the immediate post-operative period the pulse was 140/mt, blood pressure was 110/70 mm Hg and both pupils were normally reacting to light. One unit blood was transfused in the immediate post-operative period in view of the pallor which the child had. Intravenous fluids were continued for twenty four hours. From the 2nd post-operative day onwards the patient developed high grade fever which continued for about a week. Oedema over the face and eyelids subsided in about one week. Excessive mucoid discharge from the nose was noticed for about 10 days. Stitches were removed on the 11th day. On the left side of the scalp wound, minor gaping was noticed. It was about 1 cm in area with an exposed rib graft underneath. It required secondary suturing for its closure. partial proptosis noticed in immediate post-operative period persisted for about 3 weeks.

Post-operative Result

The forehead was advanced by 2.5 cms and midface was advanced by about 1.5 cm. Exorbitism was corrected (Fig. 15, 16). Movements of the eye ball in all quadrants and movements of the eyelids were normal. Both pupils were reacting normally to light. The prominent and dilated veins over the orbit disappeared completely.

A month later the patient developed redness of both the eyes followed by swelling, which spread to the forehead, and temples. Physical examination suggested sub-accute

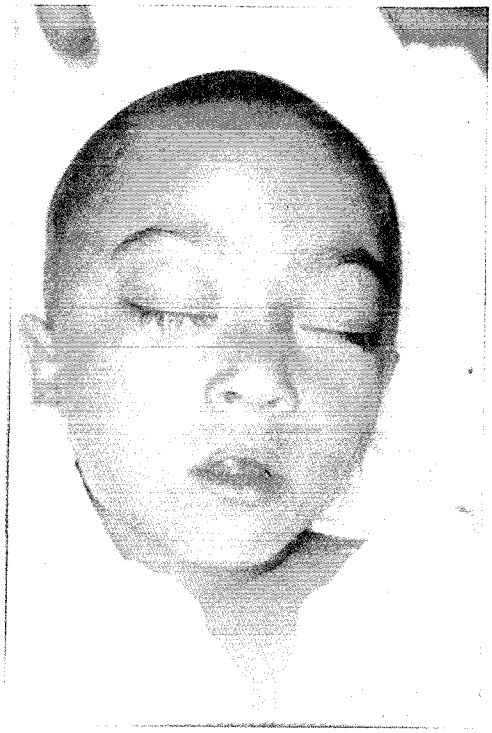


Fig. 1. Pre-operative photograph (Front view).

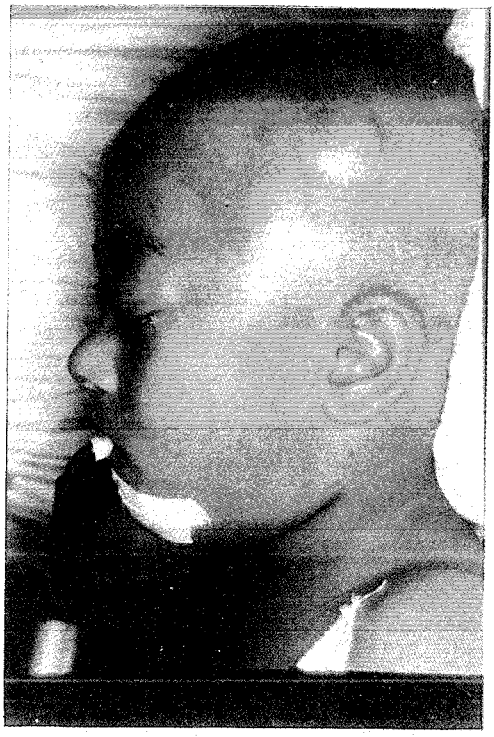


Fig. 2. Pre-operative photograph (Lateral view).

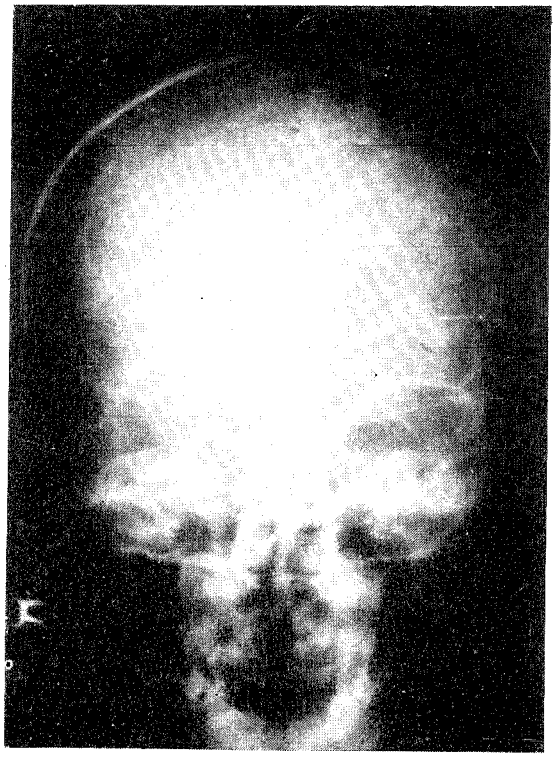


Fig. 3. X-ray skull of the patient.

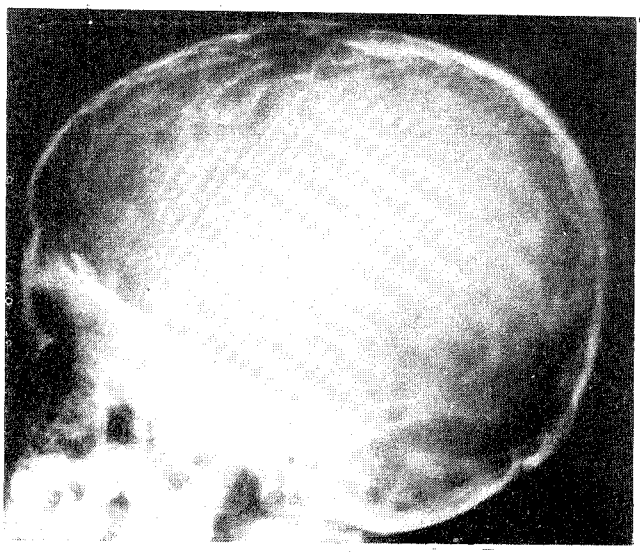


Fig. 4. X-ray skull of the patient.

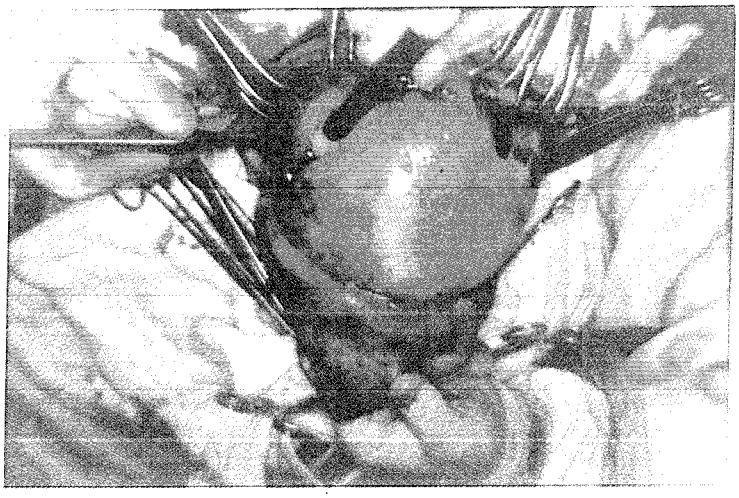


Fig. 5. Showing the elevated coronal flap.

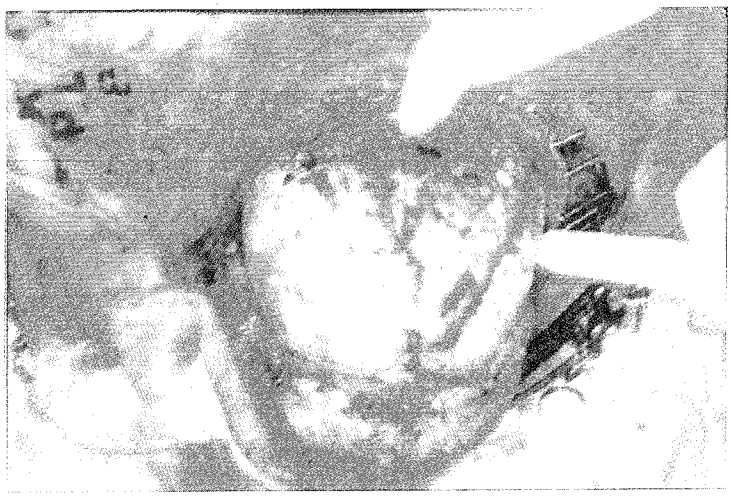


Fig. 6. The galea-aponeurosis has been erased from the skull. Markings show the frontal bone to be elevated.

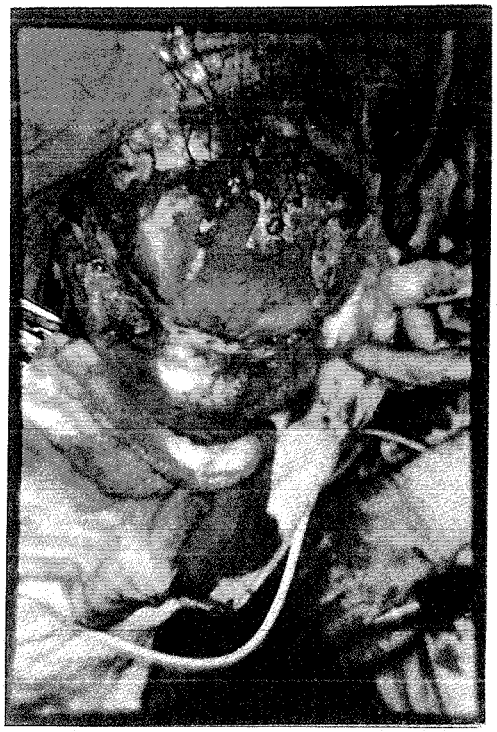


Fig. 7. The frontal bone has been removed. Moist cotton pads protect the underlying brain.

Fig. 8. Dissecting the orbital roof.

Fig. 9. The orbital roof too has been removed.

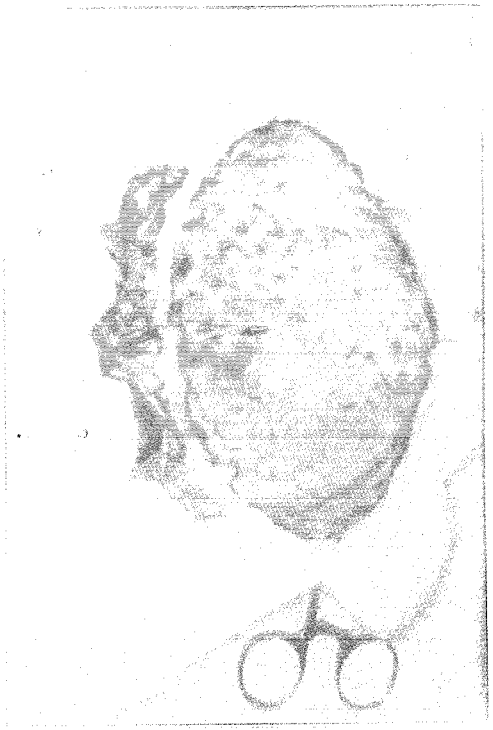


Fig. 10. Showing the bones removed. Inner surface of the bones show evidence of raised intracranial tension.

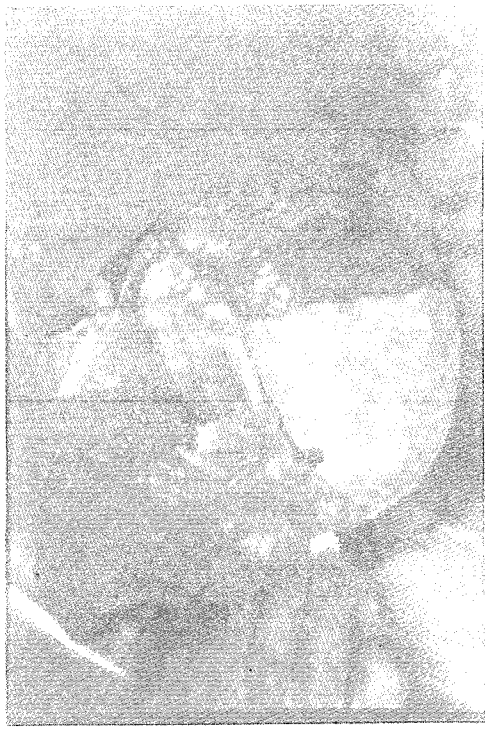


Fig. 11. The bones replaced in an advanced position. Note the graft in the region of the zygoma and the gap in the region of the frontal bone.

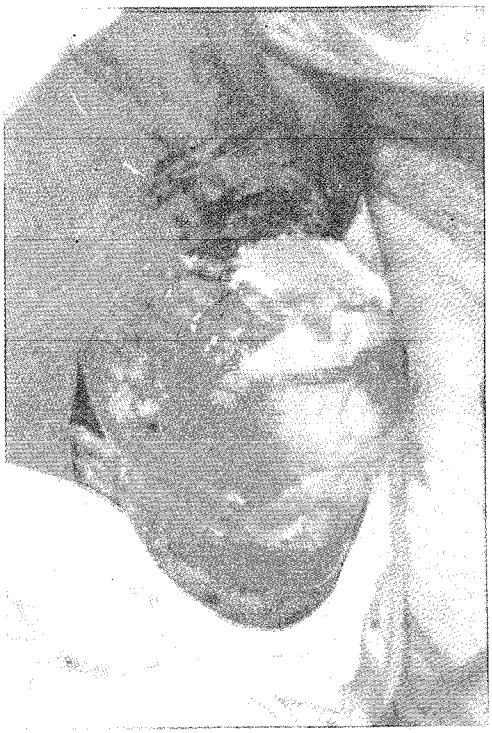


Fig. 12. Rib graft and surgicel being used to bridge the gap in the frontal bone.

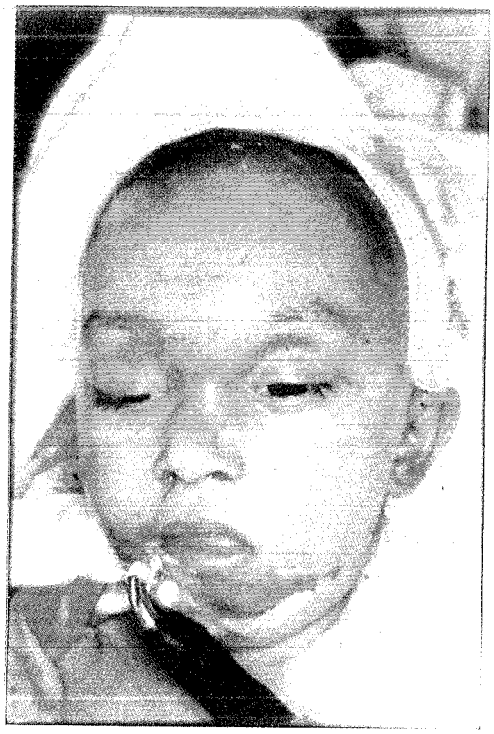


Fig. 13. Immediate post-operative photograph.

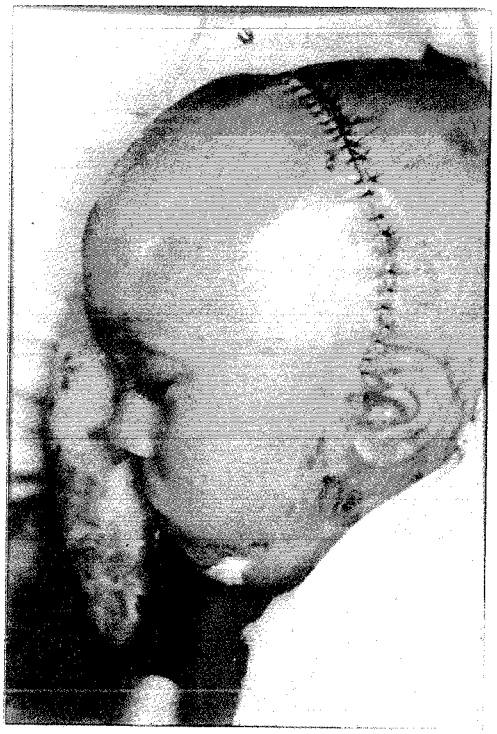
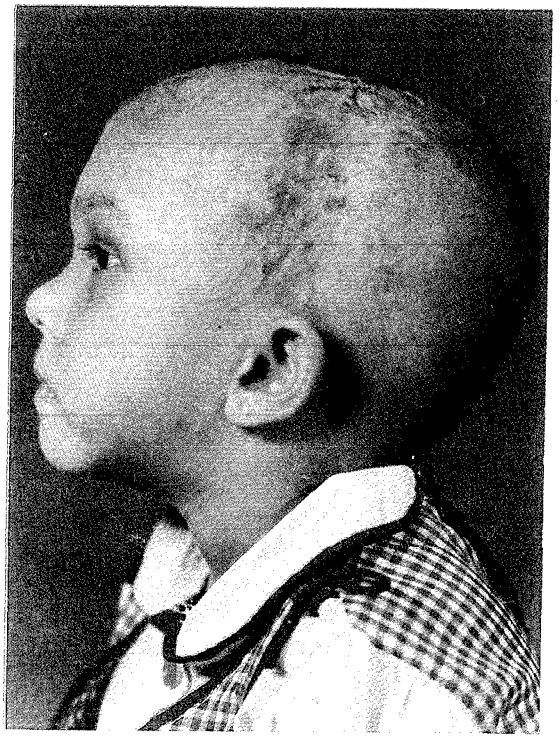


Fig. 14. Lateral view of the same.



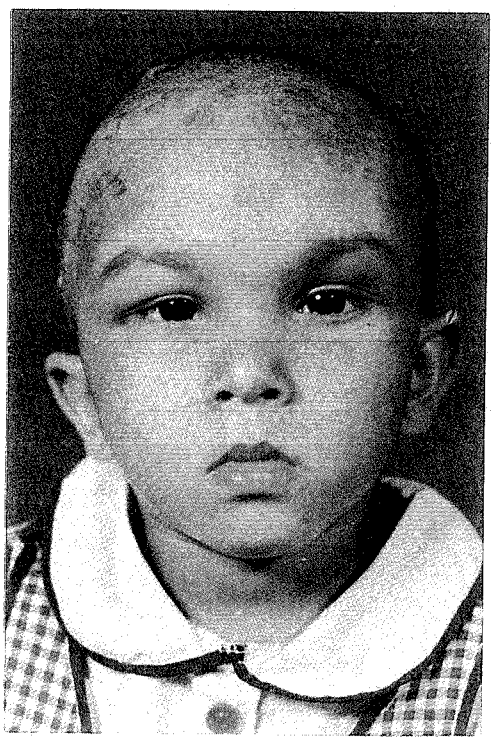


Fig. 15. and 16. Post-operative photographs two months later.

inflammation. Wide bore needle aspiration revealed the presence of pus from both the temporal regions. The abscess was drained through incisions on both the sides. The pus culture was positive for staphylococcus aureus. With antibiotic therapy (Gentamycin, Cloxacillin and Metronidazole) the inflammatory process was controlled. The patient is now well and there is no discharge from the drainage sites. She is progressing normally and has been allowed to go home.

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