Radical Reconstruction of Complex Cranio-Orbito-Facial Abnormalities*

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Introduction. The two possible justifications for surgery in craniosynostosis are first, to allow room for the growing brain, thus preventing damage to developing neural structures and second, to prevent or correct deformity. There is considerable doubt about the extent to which the first consideration is valid. Objective signs of increased intracranial pressure are rarely seen with a single closed suture. This fact, along with the difficulty in demonstrating either impairment or improvement in psychological functioning following single suture removal, has led to a generally negative view toward synostectomy with a closed single suture. On the other hand, there is very little question that multiple suture closure can lead to increased intracranial pressure, and here, early surgery is certainly justified. This ongoing argument about the benefits that might accrue with release of the fused sutures seems to have prevented growth of interest in the purely cosmetic aspects of the procedure. Although there is general agreement that early release gives better long-term results, there has been very little interest in the late effects of synostosis, such as marked frontal bossing or the associated facial deformities like those seen in Figure 1.

We have recently been attempting to reconstruct complex abnormalities involving the skull, orbit, and face at a progressively earlier age. Our cases include such conditions as Apert's and Crouzon's syndromes and single and multiple premature suture closure. We wish to present two illustrative cases, one of a combined unilateral coronal and sagittal synostosis and the other, a Crouzon's syndrome.

Case 1. T. D. was seen at the age of two weeks. He showed (Figs. 1, 2) the typical appearance of a right unilateral coronal combined with a sagittal synostosis. There was considerable frontal bossing on the left side opposite the coronal synostosis, which had resulted in depression of his left orbit. He was

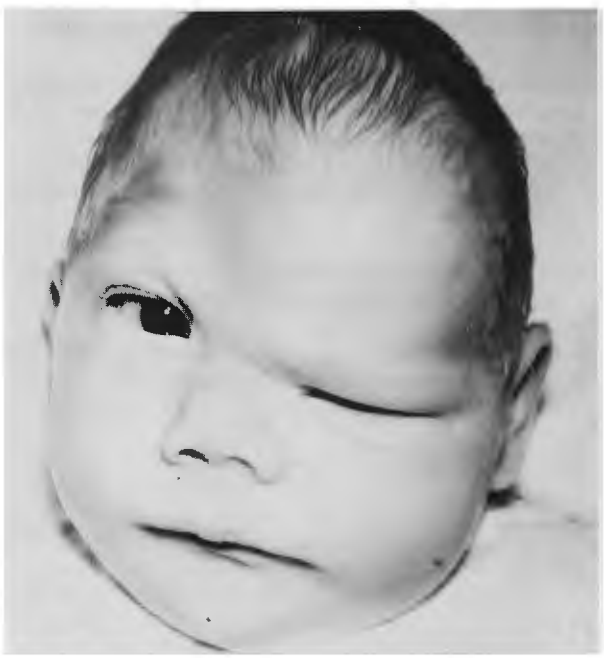


Fig. 1—Case 1 showing flattening of the right brow and depression of the left orbit in the instance of combined right coronal and sagittal synostosis.

* Presented by Dr. Jane at the 27th Annual Stoneburner Lecture Series, February 7, 1974, at the Medical College of Virginia, Richmond.

Fig. 2—View from above showing the extent of the left frontal bossing resulting in depression of the orbit.

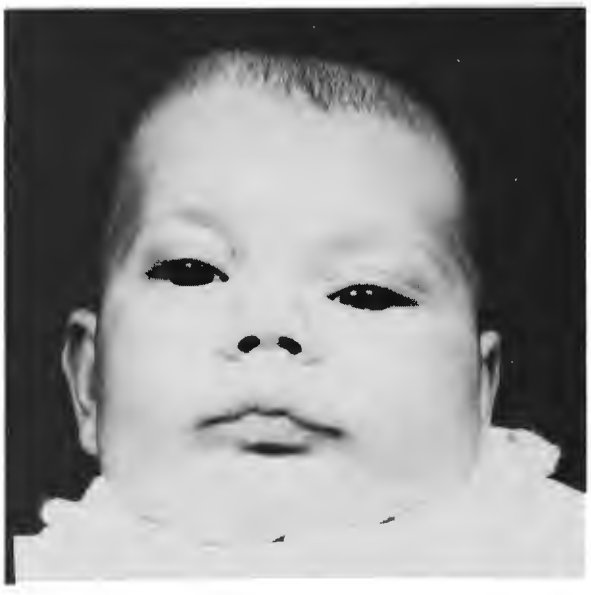


Fig. 3—A postoperative view showing improvement in the skull shape with some residual depression of the orbit.

taken to the operating room where the fused sagittal suture was removed and used to reconstruct the deficient orbital rim on the side of the coronal synostosis. A dural graft also was placed on this side, and the dura was plicated over the side of the frontal bossing. This resulted in shifting the intracranial contents to the right side with a more normal appearance of the skull. We consider this maneuver of remoulding the dura to be an important step in the long-term results. Figure 3 is a postoperative

Fig. 4*A*, *B*—The typical appearance of Crouzon's deformity with proptosis, underdevelopment of the maxilla, and relative overdevelopment of the mandible.

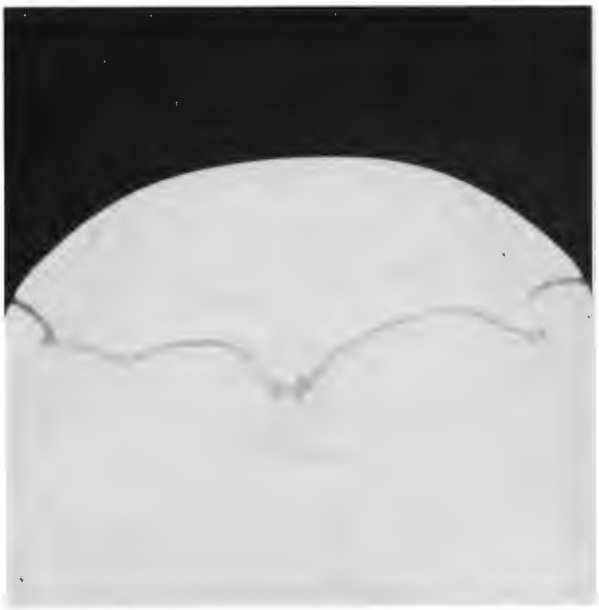


Fig. 5—Cadaver skull showing the point at which the osteotomy is carried out.

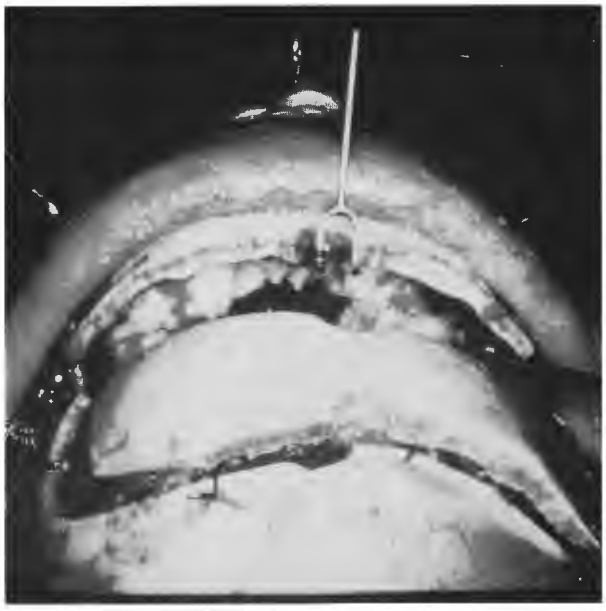
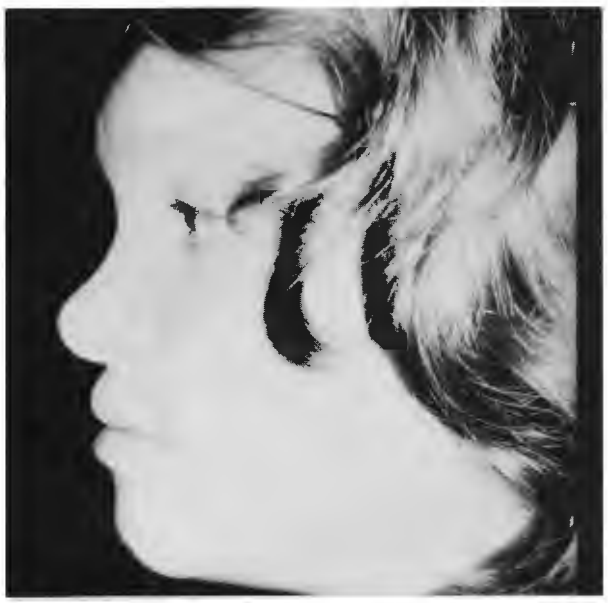


Fig. 6—Operative photograph showing the frontal bone being pulled forward over the orbit.

photograph showing excellent skull shape, although the orbit is still slightly depressed. Facial appearance has continued to improve.

Case 2. Figure 4*A*, *B* shows a 13-year-old girl with the typical appearance of Crouzon's disease. There is relative underdevelopment of the maxilla and overdevelopment of the mandible with proptosis. Our surgical procedure involved a combined intracranial and extracranial approach; one of the procedures used is illustrated in Figure 5. The line across the frontal fossa indicates the cut that is made in the bone. Figure 6 shows the actual operative



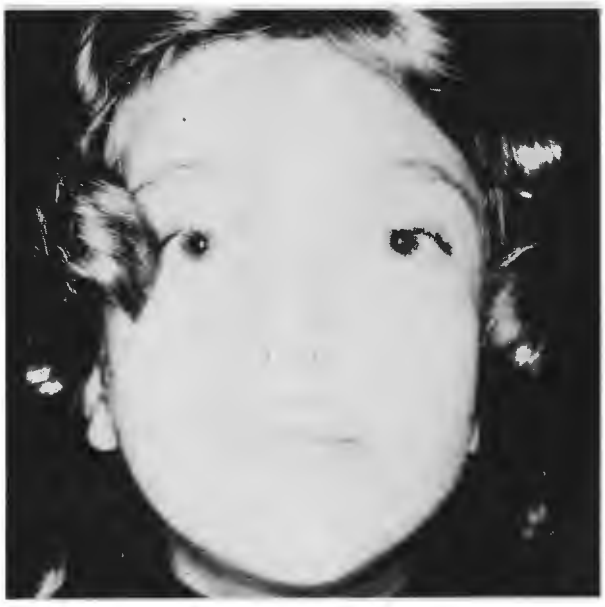


Fig. 7*A*, *B*—Postoperative photographs.

procedure, the frontal bone being moved forward over the orbits to relieve the proptosis. Figure 7*A*, *B* shows the postoperative results.

Discussion. These procedures are generally long and, particularly when performed on young children, considerable care must be taken in the maintenance of blood volume and humidification of the respiratory system. In the postoperative period, chronic measurement of intracranial pressure has also proven to be most helpful.

The first case illustrates what we think may be an important aspect of the early repair of craniofacial malformation, that is, the question as to whether remodeling of the cranial vault alone, without concomitant remodeling of at least the dura, is sufficient to obtain good late results. It may well be that the underlying brain and dura are the prime factors responsible for the ultimate shape of the cranial vault as has been suggested (1). Experiments in our own laboratory also suggest that this is the case. We have performed a series of experiments on neonatal hooded rats, in which we have systematically altered the brain or the dura alone or we have altered both together. The findings suggest the importance of the dura and brain in determining skull shape (2).

Summary. Excellent cosmetic results can be obtained by early intervention in severe cranial-orbital-facial deformity. This intervention should take the form of a combined intracranial and extracranial approach. While the procedure is of great magnitude, the benefits to the patient and to the family seem worth the risk.

REFERENCES

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