Discussion

Rare Craniofacial Clefts: Tessier No. 4 Clefts

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The authors state that a major difficulty in understanding rare craniofacial clefts arises from the fact that previous reports have focused on a single case or have grouped together different types of clefts. Although it can be very useful to focus on a single case, and although many of the advances in science are made by studying the similarities and difficulties between several groups, it is undoubtedly true that our knowledge of this anomaly could be enriched by the observations made in this group of eight patients. The question is: Did we learn from this study?

In their discussion of the morphology of these clefts, the authors mention extrophy of the maxillary sinus in severe cases, thus making a comparison with extrophy of the bladder and suggesting that the anomaly is caused by a developmental abnormality of the anterior wall of the sinus. Such an abnormality, unknown to this discussor and difficult to explain from an embryologic point of view, is not consistent with the presence of a closed hypoplastic sinus that was repeatedly observed by this author.

The pathology of oblique facial clefts has in fact been described by Vermey-Keers et al., who concluded from their study that this abnormality has its origin in the differentiation phase that follows the fusion of the facial processes. The anomaly seems to be caused by abnormal ossification in or between the ossification centers. Its typical morphology can be explained by the four-dimensional process that will occur when differentiation of a skeletal part is disturbed. This part, behaving as scar, will prevent the adjacent normal tissues from developing normally. As a result, an hourglass deformity characterized by opposed V-shaped anomalies will develop, affecting the hairline (widow’s peak), the nose, the maxilla, the eyelids and the upper lip. The illustration of this process (Fig. 1) resembles the four-dimensional space-time diagram in A Brief History of Time by S. Hawking. The authors advocate early repair of skin and muscles in combination with early bone grafting. They conclude, however, that the result of this treatment may be disappointing, because of progressive aggravation of residual defects such as canthal dystopia, because of scar contraction and poor cosmesis of interdigitated flaps, and because of differences in color, bone-graft resorption, and so on. Additional operations are therefore needed.

If we want to improve our results, we must first establish the cause of the gradual deterioration of our patients’ appearances. I feel that this is due to a conflict between two opposing sets of forces. The first of these sets is formed by the anteriorly directed forces produced by the forward thrust of the centrofacial segment with its enormous growth potential. This thrust is normally controlled by the maxilla. In the presence of skeletal and muscular disruption, it will result in separation of the central and lateral parts. The second set of forces is formed by the posteriorly directed forces that are generated by the shortage of tissues in the lateral segments of the face and by the contraction of scar tissue once healing has occurred.

The goal of plastic surgery is to obtain a maximum of result with a minimum of scarring in a minimum of time. How can we achieve these objectives in the repair of this type of clefting? My answer to this question is

1. By reduction of surface tension in the lateral facial segment. Skin expansion as a first
step in the more severe cases may thus be indicated.
2. By reinsertion of the orbicularis oculi et oris muscle if possible in conjunction with the periosteum. This step will reposition

the dislocated central segment and control its forward thrust by restoration of skeletal muscular continuity.

3. By reconstruction of the orbital floor to allow for elevation of the orbital contents. Calvarial bone grafts have become very popular in recent years, but a carefully tailored cartilage graft is probably as good or better.

I doubt the value of bone grafts on the maxilla at an early stage and condemn the insertion of grafts in the cleft because they may prevent natural remodeling of the facial skeleton by the muscular forces.

4. By redistribution of skin over the area of disruption. Early resurfacing by rotation of a cheek flap seems to me the procedure of choice.\(^5\) Elongation of the superior edge of this flap can be achieved by a Z-plasty in the temporal area.\(^6,7\) Elongation of the medial edge is done by the formation of a second flap within the limits of the first flap\(^8\) (Fig. 2). In order to anticipate and avoid the detrimental effect of scarring, incisions must be made parallel to the lines of minimal tension. However, even when parallel, a scar may be subject to tangential or shearing forces. The effect of these forces is particularly harmful in the early postoperative period when scar tissue is still young. Fixation of the skin to the skeleton at strategic points, such as the medial and lateral canthal area, following reinsertion of the orbicularis muscles will help to improve scar formation and preserve facial contour.

"hourglass deformity"

Fig. 1. Four-dimensional space-time diagram showing sites of colobomata produced by developmental arrest.

Fig. 2. Rotation flap of the cheek incorporating additional flaps for lengthening of superior and medial margins.
Scars are best camouflaged by natural folds and creases. Ideally, therefore, the final scar should resemble that of a linear Ferguson Weber incision. Every effort should be made to avoid the patchwork that is so common in these types of repairs.

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