Facial Clefts

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Facial clefts, including those of the lip and palate, are extremely rare. Some arise at the junction of facial processes preventing fusion, others have their origin in or between ossification centers. Virtually all are associated with osseous deficiencies.

This article reviews the pathogenesis and morphology of these clefts. The principles of skeletal and soft tissue reconstruction are discussed and the experience of the authors with various new techniques is reported.

Pathomorphology

DeMyer [1] considers a cleft to be a defect in apposition of structures along a junction. This concept is easy to accept when it refers to a defect caused by failure of fusion of junctional structures or facial processes. Such primary or true clefts can only be produced at an early stage of facial development (the transformation phase) and then only in 4 different locations. These are: at the junction between the lateronasal or medianosal process and the maxillary processes, between the maxillary processes where the palate is formed, and between the maxillary and mandibular processes. Arising at an early stage (before 17-mm crown-rump length), the clefts become associated with secondary bony deficiencies [2].

The secondary or pseudoclefts have their origin at a later stage of development when the facial processes have fused and the differentiation phase has begun. They are obviously caused by abnormal ossification within or between the ossification centers. The appearance of these pseudoclefts can be understood when one visualizes what may happen when development is arrested in one skeletal area, while it continues in the normal adjacent tissues. An hour-glass deformity may be produced with the transition in the middle as the original site of the evolutionary disturbance [3]. This part, behaving as a scar, will then prevent the surrounding tissues from expanding normally. As a result, a series of opposed V-shaped anomalies will develop affecting the hairline, nose, maxilla, lips, and eyelids. The character of these anomalies will vary with the area of skeletal involvement and the nature of the adjacent tissues. The direction of these pseudoclefts, colobomas, and peaks is predominantly in a craniocaudal direction.

Normal facial growth is based on a harmonious development of the various differentiating ossification centers. Disruption of skeletal, muscular, and cutaneous continuity will interfere with this and, thus, create a 4-dimensional problem. The reason for this is obvious. Although the developmental arrest that is responsible for secondary clefts occurs after fusion of the facial processes has taken place, the final relationship between the skeletal parts has not yet been achieved and their position is, therefore, still subject to changes induced by the constant interaction between unrestricted growth on the one hand and failure or absence of differentiation on the other hand. The effect of a differentiation defect on skeletal development is best demonstrated by the phenomena observed in median and paramedian clefting of the face. In median clefting, a failure of differentiation involving the internasal area prevents narrowing and lengthening of the nasal septum. As a result, normal approximation of the facial and union of the nasal halves becomes blocked. In paramedian clefting due to nasal, nasomaxillary, and maxillary dysplasia, the situation is different. The forward projection of the nasal structures is normally controlled by the maxilla. Skeletal and muscular disruption in these areas will result in separation of the central and lateral parts of the skull and is, therefore, responsible for the cranial drift of the nose that is observed in these cases. The spectrum of facial dysostoses is dominated by the clefts, which have their origin in the middle third of the face, which is formed by the internasal, nasal, nasomaxillary, and maxillary structures. The facial clefts in this area have been named after the site of the developmental arrest and they will be the subject of our discussion. As shown by the following observations made in series of 1,000 facial clefts, they are all rare clefts: Internasal Dysplasia, 4 per thousand clefts [4]; Nasal Dysplasia (schisis), 4 per thousand clefts [4]; (Naso) Maxillary Dysplasia, 0.75-5.4 per thousand clefts [4, 5].

Internasal Dysplasia

The term internasal dysplasia refers to a developmental arrest in the groove separating both nasal halves before union has occurred (Fig. 1) [2, 3]. Depending on the stage of development that has been achieved before the insult strikes, a wide spectrum of abnormalities can be observed. In less severe cases, the anomaly is characterized by bifidity of the nasal tip, dorsum.

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Fig. 1. Spectrum of internasal dysplasia. A. Without cleft lip, B. With median cleft lip.

and distal part of the cartilaginous septum. Occasionally, it is also characterized by a median cleft of the lip, a median notch in the cupid’s bow, or a duplication of the labial frenulum. In more severe cases, the nasal halves are widely separated and teleorbitism is always present [6]. The premaxilla is never absent, but it can be retarded in development and bifid. The maxilla may show a keel-shaped deformity, with the incisors rotated upward in each half of the alveolar process. The wider the cleft, the greater the interorbital distance, the shorter the nose, the more arched the maxillary vault.

Nasoschisis

This condition (Fig. 2) is caused by a developmental arrest of the lateronasal process, resulting in clefting of one nasal half. Nasal septum and cavity are usually not involved. The severity of this anomaly ranges from a small notch or coloboma in one or both alae to a more or less complete absence of nostril and nasal bone exposing a wide mucosa-lined cavity at the piriform aperture. Complete absence of the nostril is inevitably associated with abnormalities in the frontal, fronto/nasoethmoidal, and internasal areas. Teleorbitism is present in more severe cases. Paramedian clefting of the alveolar arch and a keel-shaped deformity of the maxilla may also be observed. Clefting of the lip is occasionally seen.

Nasomaxillary Dysplasia

This condition (Fig. 3) is caused by a developmental arrest at the junction of the lateronasal and maxillary processes [6]. Nasomaxillary clefting may be incomplete or complete. The anatomy of these clefts can be explained on the basis of early embryonic development of the face up to and including the 17-mm crown-rump length stage and that of the secondary palate from the 27-mm crown-rump length stage on [2].

An incomplete or nasoocular cleft will result when the disturbance in fusion is restricted to the lateronasal and maxil-
There may be a cleft in the alveolus between the lateral incisor and the canine tooth or, rarely, between 2 incisors, as Marian of the nasolacrimal apparatus have been observed in the majority of patients. Not surprisingly, clefting of the palate, whether complete or partial, has also been reported and malformations of the nasoalveolar apparatus have been observed in the majority of patients.

Maxillary Dysplasia (Lateral)

This malformation is caused by a developmental arrest in the lateroposterior part of the maxillary ossification center(s) which results in secondary clefting. The cleft starts in the lip, in or near the oral commissure, and arches upward to a coloboma in the lateral part of the lower eyelid. There are skeletal defects lateral to the infraorbital nerve. One frequently finds a cleft in the alveolus between the canine and the first molar.

Treatment

Tessier’s pioneering work on the restoration of the cranio-orbita-nasal skeleton as a basis for adequate repair widened the horizon for many children afflicted with facial clefting [8-13]. The results of correction improved dramatically and it was, therefore, only natural that the importance of soft tissue repair in internasal, nasal, and maxillary dysplasia was overshadowed by priorities imposed by the restoration of the cranio-orbita-nasal skeleton. In addition, the significance of associated maxillary malformations was somehow neglected.

Reestablishment of facial integrity in patients with median or paramedian clefts involves all disrupted structures: the skeleton, muscles, and skin. It is based on the following principles:

1. Reconstruction of the skeleton by the transposition of skeletal parts and the apposition of bone grafts.
2. Reinsertion of the muscles by transposition and fixation of dystopic remnants. An intact muscular layer serves to establish and maintain form, to animate the face, and to stimulate growth.
3. Restoration of the skin by transposition and apposition of flaps. The cutaneous layer provides protection for the underlying structures and preserves facial contour by its fixation to the skeleton at strategic points.

Application of these principles is usually rewarded with a good result, but failures are still observed. The explanation for this can be found in the eternal conflict between surgical correction and scar formation, which is the reason that much of the initial benefit of the operation may be erased with growth. A discrepancy in growth between the scarred area and the adjacent normal tissues is a natural phenomenon which should be expected. The detrimental effect of scarring can, however, be minimized or avoided by better timing of the correction, better planning of the incisions, and better anchoring of the tissues.

1. **Timing of the correction.** The desire to correct a malformation soon after birth is understandable from several points of view but not always wise. In selected cases, extensive rearrangement of skin should be avoided if there is any chance that the condition will improve spontaneously or can be improved by skin expansion. In nasal reconstruction, this objective may be reached by the implantation of a T- or L-shaped bone graft, of a temporary prosthesis made of alloplastic material, or a skin expander to increase the nasal surface.

2. **Planning of the incision.** If possible, incisions should be: (a) away from the midline of conspicuous areas, such as the forehead, nasal dorsum, or philtrum; (b) hidden in creases or folds whenever possible; (c) parallel to the lines of minimal tension.

3. **Anchoring of the tissues.** Even when parallel to the lines of minimal tension, 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, and careful re- and appositioning of muscles whenever this is required will help to improve scar formation.

Surgery

Internasal and Nasal Dysplasia

The Skeleton. Teleorbitism is frequently observed in both internasal and nasal dysplasia or nasoschisis. The type of correction is dictated by the degree of teleorbitism and maxillary involvement. When the maxillary deformity is of little importance, reduction of the interorbital distance by medialization of the orbits alone can be considered.

Medialization of orbit and maxilla by means of a medial faciostomy (Fig. 5) is, however, indicated in cases with a more pronounced maxillary deformation. The idea of transposing orbit and maxillary segment as one unit was conceived when the association of hypertelorism and maxillary malformations was observed and the possibility of correcting these skeletal anomalies in one procedure was considered [14-18]. It was, however, first put into practice in patients with severe hypertelorism and median nasal clefing. The technique permits a rotatory movement in each facial half, reducing the interorbital distance and lowering the height of the palate.

The Nose. Formation of the nose may involve the cartilaginous skeleton as well as the mucosal lining and skin. Nasal reconstruction in internasal dysplasia requires:

1. Removal of the anomalous cartilaginous tissue.
2. Restoration of a nasal framework. A costal, calvarial, or iliac bone graft is used, bearing in mind that this framework never grows and that some degree of resorption is bound to occur.
It is for this reason that the cover of the graft should consist of well-vascularized tissue.

3. Elongation of the mucosal lining of the roof of each nasal half by transposition of flaps if retraction exists.

4. Medialization and fixation of the alar cartilages over the new skeleton.

5. Correction of the skin deficiency in the midline. This can sometimes be achieved by linear closure of the dorsal defect following resection of redundant tissues and redistribution of skin over the depressed area. Advancement of the contracted skin toward the tip of the nose in a V-Y manner and the transposition of flaps over the dorsum carry the risk of conspicuous scarring and are, therefore, not advocated.

Skin expansion procedures prior to a more definitive repair may help to minimize the extent of surgery and to improve the quality of scar formation. Rotation of a forehead flap is rarely indicated (Fig. 6). The anomaly can usually be corrected by local rearrangement of skin as shown in Fig. 7.

Nasal reconstruction in nasochoisis requires:

1. Restoration of a nasal framework in the more severe cases.
2. Reconstruction of the nasal lining by mobilization and apposition of the mucosal edges. This procedure is normally sufficient to create a passage of adequate dimensions. It may, however, be necessary to lengthen the roof of the mucosal cavity by transposition of flaps.

3. Resurfacing of the nose by transposition procedures. This step is attended by the familiar problems related to scar formation and different color match. In less severe anomalies, transposition of V-shaped flaps in a Z-fashion is usually sufficient. In the more severe anomalies, it is advisable, first, to provide cover by rotation of the available surplus of skin in the nasal dorsum and glabella, a procedure that is facilitated by a backward cut in the medio-fronto-nasal region; and second, to remove skin from the ala on the affected side up to its rim and close the resulting defects by judicious and economic use of the rotation flap (Fig. 8) [19].

This approach provides a new covering, with the final scars on the lateral aspect of the nose and in the alar rim where they are least visible (Figs. 9, 10); however, scarring over the nasal dorsum cannot always be avoided (Fig. 11).

Nasomaxillary and Maxillary Dysplasia

From a surgical point of view, the treatment of nasomaxillary and maxillary dysplasia have much in common. The cranially-directed drift of the nasal structures is always associated with a significant shortening of the distance between the lower eyelid or medial canthus and the alar base. In both malformations, there may also be a cleft of the upper lip. The eyeballs are usually unprotected and the anomaly must, therefore, be treated as an emergency—preferably in a center where one has experience with the management of these cases.

The Skeleton. Management of the skeletal anomalies is first determined by the condition of the maxilla that is characterized by caudalization of the roof of the maxillary sinus, by retrusion of the anterior wall of this cavity, and by lateralization of its medial wall. Correction of the first and the second of these abnormalities can be achieved by application of bone grafts. Obliteration of a defect in the alveolar arch with bone grafts is never indicated because this procedure would interfere with the anticipated repositioning of the dislocated nasal structures. This repositioning is effected by the forces that will be produced after closure of the soft tissue defects. These forces are generated by the contraction of scar tissue and by the restoration of the muscular activity in the original areas of disruption.

The management of skeletal anomalies is also dependent on the presence of teleorbitism and orbital dystopia that are occasionally seen in combination with nasomaxillary dysplasia.
Fig. 6. Nasal reconstruction in internasal dysplasia by transposition of forehead flap. A. Preoperative views (Reprinted with permission of publisher [16]). B. Postoperative views.
Fig. 7. Nasal reconstruction in internasal dysplasia by local rearrangement of skin. A. Preoperative views. B. Postoperative views.
Fig. 8. Technique of nasal reconstruction in nasal dysplasia (nasoschisis). A. Unilateral. B. Bilateral.

Fig. 9. Nasal reconstruction in bilateral nasoschisis. A. Preoperative view. B. Postoperative view. Reprinted with permission of publisher [17].
A patient with complete nasomaxillary clefting is obviously a candidate for a hemifacial bipartition procedure [14-17] because the position of the displaced orbitomaxillary unit in relation to the remaining part of the face can, thus, be improved. The hypoplastic dimensions of the maxilla would, however, not be altered by this technique, requiring secondary surgery at a later stage.

The Soft Tissues. Correction of the soft tissue defects requires: (a) repositioning of the medial canthus, alar base, and orbicularis oculi and oris muscles; (b) repair of buccal, nasal, and conjunctival lining; and (c) restoration of the skin. It is the latter part of these steps that offers the most difficulties. Reconstruction of the lower eyelid and of the nose requires a significant amount of skin, which can be found in one or more
of the available donor areas: the upper eyelid, forehead, glabella, and cheek.

1. Skin of the upper eyelid may be used for reconstruction of the lower eyelid with the restriction that only small flaps can be transposed.

2. Skin of the forehead is available in sufficient quantity, and although its texture and color are not the most desirable, it may be used for reconstruction of the lower eyelid and the medial canthus region. The surplus of tissue provided by a forehead flap may be of particular value in bilateral cases if reconstruction of the medial canthus and alar base are not sufficient to produce adequate lengthening of the nasal dorsum (Fig. 12). In these cases, the distal half of the flap may serve to close the defect on one side of the face in the first stage, while the contralateral defect and the defect created by the transection of the nasal dorsum may be resurfaced with the pedicle of this flap in a second stage. The scars running across the nasal dorsum are, however, a distinct disadvantage (Fig. 13). This problem does not exist, however, in the correction of unilateral anomalies (Fig. 14).

3. Skin of the glabella can be used in unilateral cases when less skin is needed. Downward rotation of a nasolabial flap in conjunction with lengthening of the nasal lining by transposition of mucosal flaps will reposition the dystopic and distorted alar base. This technique has been used for the correction of incomplete nasomaxillary dysplasia.

4. Skin of the cheek may be used in a variety of ways, and its texture and color are perfect. The only question is how to
take the best advantage of these qualities. Interdigitation of the cutaneous edges of the cleft has been reported by many authors, but the shortage of skin may be extreme and an initially good result may deteriorate with time since the scarred area does not keep up with growth.

Rotation and advancement of the cheek has recently been recorded and the results obtained with this technique in our patients (Fig. 15) show that it permits maximal correction at the expense of minimal scarring [20]. The technique has one drawback. Following rotation of the flap, a contracture may
Fig. 16. Technique of reconstruction in maxillary dysplasia by rotation and advancement of the cheek.

tend to develop along its medial edge. Straight forward interdigitation of the flap and the nasal skin offers one solution to this problem. Even more length can be gained, however, by a back cut at the lower end of the medial edge of the rotation flap near its pivot point [21]. The second flap which is, thus, formed can be rotated over more than 90°, thus bringing skin to the area where it is most needed (Fig. 16). Even more skin can be produced by expansion of the cheek as was suggested by Argenta [22].

Résumé
Les fentes faciales—les fentes labiales et palatines—sont très rares. Certaines se voient à la jonction des os de la face, empêchant la fusion, d’autres se trouvent entre ou dans les points d’ossification. La plupart sont associées à des déficits osseux.

Cet article passe en revue la pathogénesis et la morphologie de ces fentes. Les principes de reconstruction squelettique et des parties molles sont discutées et l’expérience des auteurs avec ces nouvelles techniques est rapportée.

Resumen
Las hendiduras faciales, incluso aquellas de labio y paladar, son extremadamente raras. Algunas se originan en la unión de los procesos faciales, lo cual impide su fusión, y otras tienen origen en los centros de osificación o en medio de ellos. Virtualmente la totalidad de las hendiduras se presentan asociadas con deficiencias óseas.

El presente artículo revisa la patogénesis y morfología de estas hendiduras. Se discuten los principios de la reconstrucción ósea y de tejidos blandos y se informa la experiencia de los autores con diversas técnicas operatorias.

References