Oblique Facial Clefts: Pathology, Etiology, and Reconstruction

J. C. H. van der Meulen, M.D.
Rotterdam, The Netherlands
Oblique Facial Clefts: Pathology, Etiology, and Reconstruction

J. C. H. van der Meulen, M.D.
Rotterdam, The Netherlands

Modern views on embryology have increased our understanding of the nature of oblique facial clefts. The anomalies that have their origin at the junction of facial processes, such as the nasomaxillary dysplasias, may be named primary clefts or transformation. The maxillary clefts that are due to a developmental arrest of the skeleton are in fact secondary defects of differentiation defects. The teratology of these malformations is discussed, and attention is drawn to the amniotic rupture syndrome as a possible cause.

All these clefts are rare, their incidence ranging from 0.75 to 5.4 per 1000 common clefts. This author has been involved in the treatment of nine of these patients. Four had their malformation reconstructed with one of the conventional procedures described in the literature, but the results, although initially acceptable, soon deteriorated. A more aggressive approach was therefore chosen. Rotation and advancement of the cheek proved to be extremely effective and is now advocated as the procedure of choice. The transposition of a median forehead flap is considered an excellent alternative. Use of these procedures in five patients is reported. There were no complications.

Oblique clefts, excepting only the mildest forms, are a catastrophic facial anomaly. Usually multiple operations are required for reconstruction and the results in most cases are far from ideal.

Although oblique facial clefts had been reported before, in 1823 Laroche was the first to differentiate between ordinary cleft lip or harelip and clefts of the cheek. Further distinction was made in 1864 by Pelvet, who separated oblique clefts involving the nose from the other cheek clefts. Drawing on Ahlfeld's work, in 1887 Morian collected 29 cases from the literature, contributing 7 cases of his own. Morian recognized three different groups of oblique facial clefts. Since then, excellent reviews have been written by Grünberg in 1913, Boo-Chai in 1970, and Millard in 1977.

This paper is devoted to the morphology and morphogenesis of oblique facial clefts, to their etiology, and to the principles used in correcting some of these malformations.

MORPHOLOGY AND MORPHOGENESIS

Oblique facial clefts have been classified according to the following:

1. The direction they take, which may be oronasal-ocular (oculonasal) or oro-ocular (oculofacial). The latter type is being subdivided into oromedial canthal and orolateral canthal, depending on the relationship of the defect to the infraorbital foramen.

2. The period in which the development was disturbed, e.g., primary or secondary clefting.

3. The position of one cleft in relation to the other (Morian I, II, III; Tessier 3, 4, 5).

4. The areas in which the malformations have their origin: nasomaxillary and maxillary (medial and lateral) dysplasia.

Other craniofacial malformations, such as encephaloceles, hydrocephaly, widow's peak or cow's lick, hypertelorism, choanal atresia, anophthalmia and microphthalmia, colobomata of the iris, choroid and epibulbar dermoids, have also been reported, and associated deformities, such as constric-
tion rings of the limbs, aplasia cutis congenita, club feet, and ectopia vesicae have been described.

Nasomaxillary Dysplasia

Nasomaxillary dysplasias may be incomplete or complete. An incomplete or naso-ocular cleft (Fig. 1, above, left) runs from the alar base, which is drawn upward, to the inferiorly dislocated medial canthus. There may be a fistula of the lacrimal canal. The lip is intact. The alar malformations resemble those found in patients with the Johanson-Blizzard syndrome.

The complete or oronasal-ocular cleft (Fig. 1, above, right) starts in the upper lip as an ordinary cleft and passes through the nasal aperture, skirting the foot of the distorted and superiorly dislocated ala nasi. The cleft then continues to the inner canthus, which is always drawn inferiorly. There is a retrusion of the maxilla medial to the infraorbital foramen, and the frontal process may be deficient or absent.

Rintala et al. observed that the alveolar cleft was always situated between the medial incisor

---

Fig. 1. (Above, left) Incomplete nasomaxillary dysplasia (nasal-ocular cleft). (Above, right) Complete nasomaxillary dysplasia (oronasal-ocular cleft). (Below, left) Medial maxillary dysplasia. (Below, right) Lateral maxillary dysplasia.
and the canine tooth, while the lateral incisor was frequently missing. The complete cleft is frequently associated with a palatal cleft.\textsuperscript{5,16} The nasolacrimal apparatus is deficient in the majority of cases.

The morphology of these clefts can be simply explained\textsuperscript{15} on the basis of early embryonic development of the face up to and including the 17-mm C.R.L stage and that of the secondary palate from the 27-mm C.R.L stage on. An incomplete or naso-ocular cleft will result when the disturbance in fusion is restricted to the lateral nasal and maxillary processes. In these patients, the lip will develop normally. A complete or oronasal-ocular cleft will occur when fusion between the medial nasal and maxillary processes is also disturbed. A cleft between the medial and lateral incisors, if present,\textsuperscript{5} may be explained by one of the dental anomalies so common in cleft palate patients, such as agenesis or a supernumerary element.

The appearance of the incomplete as well as of the complete cleft will depend on the period in which the arrest occurred. If the arrest occurs before the different processes have merged at the 17-mm C.R.L stage, the lacrimal canal will fail to form and a so-called primary or transformation\textsuperscript{15} defect will be formed. If the disturbance takes place after closure of the ectoderm of the face has been completed and a canal has been produced, a secondary or differentiation defect\textsuperscript{15} will result.

**Maxillary Dysplasia (Medial)**

In this deformity, the cleft (Fig. 1, below, left) extends from the lip midway between the philtrum and the oral commissure, proceeding laterally from the intact but superiorly dislocated nasal aperture to a lateral coloboma. The maxilla, medial to the infraorbital foramen, is always hypoplastic, causing a severe retrusion of the rim of the piriform aperture and a funnel-shaped concavity (infundibulum) in the medial or anterior part of the orbital floor. There may be a cleft in the alveolus between the lateral incisor and the canine tooth or, rarely, between two incisors, as Morian\textsuperscript{6} reported. In his case, however, the incisor lateral to the cleft proved to be supernumerary. Not surprisingly, clefting of the palate, whether complete or partial, has also been reported, and malformations of the nasolacrimal apparatus have been observed in the majority of patients. The morphogenesis of this cleft can now be explained on the basis of known embryologic theories. The presence of a normal nasal aperture is proof that fusion of the maxillary processes has been normal and, consequently, the development arrest has occurred after the 17-mm C.R.L stage, when the ectoderm of the face has closed.\textsuperscript{15} The resulting secondary or differentiation defects\textsuperscript{15} may produce an hourglass deformity\textsuperscript{14} with the developmental arrest in the middle and a coloboma at both ends. The many defects of the nasolacrimal apparatus that are found in some of these patients have been extensively described by Ask and van der Hoeven.\textsuperscript{25} Their morphogenesis has recently been explained by Vermeij-Keers et al.\textsuperscript{26}

**Maxillary Dysplasia (Lateral)**

This cleft (Fig. 1, below, right) 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.\textsuperscript{8,11,27} Both unilateral and bilateral deformities have been described, but they are rare,\textsuperscript{6,8,11,19,21,27-33} and some are poorly documented. However, the malformation may in fact not be so unique considering the striking superficial resemblance\textsuperscript{11,34} with maxillozygomatic dysplasia, Tessier’s no. 6 cleft. In maxillozygomatic dysplasia the defect is found between the maxilla and the zygoma. The cleft extends into the inferior fissure and the alveolar process is not involved. In Chavane’s case,\textsuperscript{27} lateral maxillary dysplasia was observed on one side and maxillozygomatic dysplasia on the other. The symptoms observed in this malformation can be explained on the basis of a developmental arrest in the lateroposterior part of the maxillary ossification center(s).\textsuperscript{13,14}

**Incidence**

In their excellent reviews\textsuperscript{35,36} of rare facial clefts, Kawamoto et al. came to the conclusion that the occurrence rate of these malformations would range from 1.43 to 4.85 per 100,000 births and from 9.5 to 34.0 per 1000 common clefts. The best indication as to the frequency of oblique facial clefts can indeed be deduced from the figures produced by centers where large numbers of clefts are seen. These data show (Table I) that the incidence varies between 0.75 and 5.4 percent.

**Etiology**

\ldots infants become crippled in the following way: where in the womb there is a narrowness at the part where in fact
The foundations of the science of teratology were laid by Meckel and by father and son Geoffroy St. Hilaire. Meckel believed that oblique clefts were caused by a developmental arrest, while Geoffroy St. Hilaire (1832) felt that amniotic bands were responsible. Since then, the debate has never stopped.

Proponents of the first theory believe that focal fetal dysplasia is the primary defect. They have suggested a variety of mechanisms, such as linear necrosis along the watershed between developing vascular areas, the occurrence of a diminished arterial supply in a crucial period of development, a disturbance in the migration of neural crest cells, or a failure of mesodermal penetration and coalescence between facial processes. In theory, these mechanisms may be caused by genetic as well as nongenetic factors. Genetic factors do not seem to play a role, since familiarity has not been reported in the literature. Nongenetic factors have never been identified with certainty, but hemorrhage and fetal necrosis have been produced in experiments with a linoleic acid-deficient diet, maternal injection with epinephrine and vasopressin, and uterine ischemia caused by clamping of its vasculature.

Proponents of the second theory, who believe that clefts are caused by bands, are now in the minority, since, logically, the origin of these clefts cannot be band-related. First, the pattern of facial clefts, although subject to slight variation, is always consistent and reflects a cranio-oral direction. It is furthermore inconceivable that perfectly symmetrical clefts can be caused by haphazardly produced bands. Second, several malformations that may be associated with an oblique cleft cannot be explained by amniotic bands, such as encephaloceles, widow’s peak, microphthalmia, choanal atresia, and cleft palate. Third, amniotic bands consist of embryonic tissue that is not known to possess contractile qualities. Finally, the existence of bands between different parts of the face cannot be attributed to an amniotic process. The origin of these bands may, however, be readily explained by the healing of fetal defects with adhesion formation.

Is it perhaps possible to reconcile these different schools of thought? Proponents of a third theory believe that craniofacial clefts and amniotic bands may be part of the amnion rupture syndrome together with visceral and extremity defects. The incidence of this syndrome has been reported to vary from 1:5000 to 1:15,000. Familial incidence has not been reported, and the fact that facial clefts have been observed in one of two monozygotic twins clearly demonstrates that nongenetic factors may be involved in their production. Drug ingestion has been implicated in some patients, as well as in animal models, but an amniogenic mechanism seems to be the predominant cause.

Torpin has stressed the role of maternal trauma and premature amnion rupture in the production of the syndrome, and his observations are consistent with those made in experiments. Amnion rupture or amniocentesis is known to produce compression-related malformations of the craniofacial complex (encephaloceles, hydrocephalus, palatal clefts), of the vertebrae (spina bifida), and of the limb (constriction ring anomalies, acrosyndactyly, etc.). Similar effects have been obtained by injection of amnion with glucose.

The spectrum of abnormalities produced by these experiments only rarely includes oblique facial clefts, but despite this, the possibility should be considered that oblique facial clefts and other anomalies have their origin in compression-related focal fetal dysplasia. Oblique facial clefts and constriction ring defects of the extremities are frequently observed in the amnion rupture syndrome. Differences in nature and incidence of the several defects can be explained by a variation in susceptibility of the developing area and by the timing and severity of the causative insult.

**TREATMENT**

An attempted correction of a facial cleft may easily result in a surgical catastrophe, for the deficiencies of skin and skeleton are frequently underestimated. Scar tissue does not keep pace.
with growth of normal tissues. Secondary bone deformities may be produced by the contraction of skin and mucosa, and what initially appears to be an excellent result may gradually turn into a monstrous disfigurement.

In the past 20 years, I have been involved in the treatment of nine patients with oblique facial clefts, three caused by nasomaxillary dysplasia and six by medial maxillary dysplasia. Four of my patients with oblique clefts were either lost to follow-up or their history is of little interest to the reader.

**Case Reports**

**Case 1**

This patient (Fig. 2) was born on May 9, 1968, with the following severe craniofacial malformations: plagiocephaly, hypertelorism, unilateral microorbitism, complete bilateral nasomaxillary dysplasia, and palatoschisis. Constriction ring anomalies of the extremities were also observed. The clefts were closed in a series of successive stages by transposition and interdigitation of glabellar nasal flaps and by a lip adhesion procedure. The palate was closed at a later stage. Since the boy had been deserted by his unmarried mother, he was adopted by foster parents and later raised in an institute for backward children. Further corrections, although urgently needed, were refused on several grounds. However, despite his handicap, the boy has developed surprisingly well and now has himself asked for further correction. The increased interorbital distance has now been reduced and his nasal deformities have been corrected by rotation and advancement of both cheeks and by transposition of a median forehead flap.

**Case 2**

This patient (Fig. 3) was born on November 22, 1969, with a right-sided medial maxillary dysplasia. Exploration after primary closure elsewhere revealed severe hypoplasia of the body of the maxilla. No cleft was found in the bone, and the alveolar process was normal. On September 20, 1976, the following corrections were carried out: repositioning of the ala nasi and the medial canthus, reconstruction of the lacrimal canal with a flap from the nasal mucosa, augmentation of the skeleton with an iliac graft, and supplementation of the skin shortage in the lower eyelid and medial canthal regions with a median forehead flap. The result was satisfactory, except for the difference in color match between the skin of the forehead flap and that of the cheek. Further augmentation of the maxilla will be needed in the future.

**Case 3**

This patient (Fig. 4) was born on May 27, 1963, and was seen in 1980 with severe deformities following the closure of a left-sided medial maxillary dysplasia and right-sided cleft lip. Examination revealed clefting of the left eyebrow with underlying skeletal retrusion, lagophthalmos due to scarring of the upper and lower eyelids, ophthalmic dysplasia, canthal dystopia, upward displacement of the left ala, massive scarring of the cheek and upper lip, retrusion of the hypoplastic maxilla, and dental abnormalities. Treatment
was performed in several stages and consisted of the following steps: Le Fort II osteotomy, augmentation of the rim of the orbital roof and of the maxillary body with bone grafts, repositioning of the attachment of the left medial canthal ligament, resection of the scars in the left cheek and lower eyelid, downward displacement of the left ala, and closure of the resulting defects using a cheek rotation procedure. The left eye was camouflaged with an episcleral prosthesis, and the patient's appearance has greatly improved. However, further corrections are indicated.

Case 5

This patient (Fig. 6) was born on September 4, 1967, with minor left-sided medial maxillary dysplasia. Following partial treatment in his "home town," definite improvement was obtained on March 17, 1982, by correction of the skeletal deformities, repositioning of the medial canthus and the ala, closure of the skin defect by rotation and advancement of a cheek flap, and transposition of a flap from the upper lip.

Technique

From a surgical point of view there is little difference in the treatment of nasomaxillary and medial maxillary dysplasia. In both malformations there is shortening of the distance between the lower eyelid or medial canthus and the alar base, and in both malformations there may be a cleft of the upper lip. Since the severity of these soft-tissue defects and of the underlying skeletal abnormalities are intimately related, corrective surgery of skin and mucosa should be combined with that of the bony structures in one major procedure and, occasionally, by orthodontics as well. To avoid unnecessary scarring and loss of time, it is essential that the principles of cleft surgery so well outlined by Tessier should be strictly adhered to.
The Skeleton

Dissection and mobilization of the edges of the cleft are followed by inspection of the maxilla. Here, anomalies are characterized by caudalization of the anterior part of the orbital floor, by lateralization of the medial wall of the maxillary sinus, and by retrusion of the anterior wall of this structure. Correction is obtained by apposition of bone grafts on the orbital floor and on the anterior surface of the sinus. The cleft should be left open in order not to prevent repositioning of the central structures by the forces that will be generated by closure of the soft-tissue defects.
The Soft Parts

Correction of the soft-tissue defect requires repositioning of the medial canthus, of the alar base, and of the orbicularis oculi and oris muscles, repair of buccal, nasal, and conjunctival lining, and finally, restoration of the skin. It is particularly the latter part of these steps that offers 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...
Fig. 6. Case 5. Moderate medial maxillary dysplasia. (Left) Preoperative view. (Right) Postoperative view following rotation and advancement of cheek flap.
donor areas: the upper eyelid, the forehead, and the cheek. Skin of the upper eyelid may be used for reconstruction of the lower eyelid, but with the restriction that only small flaps can be transposed. Skin of the forehead is available in sufficient quantity, and although its texture and color leave somewhat to be desired, it may be used for reconstruction of the lower eyelid and the medial canthal region. The surplus of tissue provided by a forehead flap\(^7\) may be of particular value in bilateral cases if reconstruction of the medial canthus and alar bone are not sufficient to pro-

**Fig. 7.** The use of a median forehead flap to lengthen the nose. *(Above, left)* Marking the incisions. *(Above, right)* Mobilization of the flaps on both sides of one cleft. *(Below, left)* Transposition of the forehead flap and interdigitation of the raised flaps in the cheek and nose to correct one-half the malformation. *(Below, right)* Final result with symmetrical scars after mobilization and interdigitation of the flaps in the contralateral half of the face using the pedicle of the forehead flap.
duce adequate lengthening of the nasal dorsum (Fig. 7). 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.

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 so extreme that the result obtained is far from optimal. Rotation and advancement of the cheek 79 (Fig. 8) has to our knowledge not been recorded, but the results obtained with this technique in our patients indicate that it permits maximal correction at the expense of minimal scarring and should therefore be advocated as the procedure of choice in the majority of cases.

J. C. H. van der Meulen, M.D.
Acad. Hospital Rotterdam-Dijkzigt
Department of Plastic and Reconstructive Surgery
Dr. Molewaterplein 40
3015 GD Rotterdam
The Netherlands

REFERENCES
2. Von Kulmus, Vide R. Morian.
Vol. 76, No. 2 / OBLIQUE FACIAL CLEFTS

30. Hydro Vide Walker.
34. Hovey, J. Vide Millard.
40. Hippocrates Vide Ballantyne.
41. Meckel, J. Meckel’s Archiv, 1828.
64. Carakushansky, G., Neu, R. L., and Gardiner, L.


