Reconstruction of Oblique Facial Clefts

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The oblique facial cleft is a rare congenital deformity believed to represent only about 0.25% of all facial clefts (*1*). In the twenty year period from 1948 to 1968, seven previously unreported patients with this anomaly have been treated at the Health Center Hospitals of Pittsburgh. Of this group, five patients have remained available to us for long term follow up.

Embryogenesis

The embryogenesis of oblique facial clefts remains disputed. A current concept visualizes the developing face of a five week embryo as a continuous sheet of ectodermal tissue lying over the bulging forebrain. This sheet is divided into three areas by the appearance of a series of ectodermal grooves. These grooves separate the central, or frontonasal region, from the maxillary areas on the right and left. Three main masses of mesodermal cells migrate into these regions and their coalescence produces obliteration of the grooves. If this mesodermal migration is retarded, the grooves persist. Disruption along the line of a groove results in cleft formation. This theory despite its attractiveness probably represents an oversimplification. Karfik has pointed out that the lateral oro-ocular cleft corresponds to no embryonic facial groove (*2*).

Oblique clefts apparently are not familial. Probably most are the result of some environmental insult to the developing embryo. It is not surprising therefore to find multiple anomalies in many of these patients and a high incidence of still birth.

Classification

Two distinct groups of oblique facial cleft exist, naso-ocular and oro-ocular. In the naso-ocular type the cleft extends from the pyriform aperture to the medial canthal area along the approximate course of the nasolacrimal duct. Often there is an associated cleft of the lip and palate.

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In the oro-ocular type the pyriform aperture is intact and the cleft extends from the mouth to either the medial or lateral canthal areas. The position of the oro-ocular cleft in relationship to the infraorbital foramen determines the subclassification of medial or lateral type.

Oblique clefts of all types can be present in incomplete forms. Mild incomplete oro-ocular clefts are particularly interesting because they can be confused with mild incomplete lip clefts. It is helpful to remember that the oro-ocular cleft lies lateral to the peak of the cupid's bow and is associated with an upward tilt of the alar base or inset.

Treatment

Oblique facial clefts, especially the more severe ones, present real challenges in reconstruction. The tissue defects present may be so extensive as to preclude truly satisfying aesthetic results. Surgery must be individualized and, except in the mildest cases, can be expected to include multiple staged operations carefully selected from the surgical armamentarium of lip, nose and eyelid reconstructions. The only generalization possible is that an exposed eye requires immediate treatment.

Case Report

I. R.H. (Figure 1) was born in 1946 with a right naso-ocular cleft associated with cleft lip and palate. He was first seen at the University of Pittsburgh at the

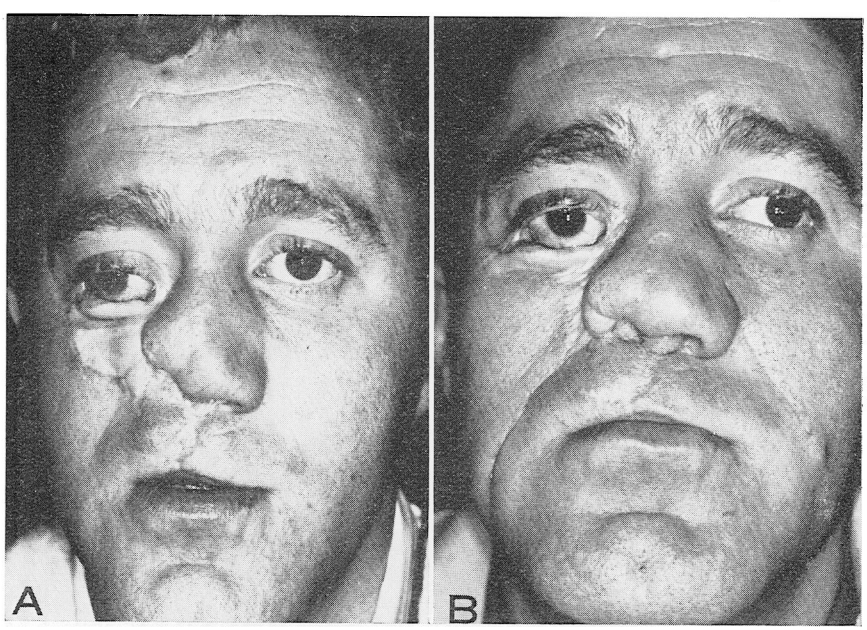

FIGURE 1. (A) Patient R.H. (Case I) as he appeared when first seen on our service after 9 operations elsewhere. (B) Appearance following 9 more operations to relieve nostril stenosis, reposition the ala, lengthen the columella, resurface the cheek and eyelid, and revise scars.

age of 20 having had 9 operations elsewhere (2 operations to repair the lip and palate in infancy followed by 3 skin graftings of the lip, lower lid, and nose and 4 unspecified nasal operations in adolescence). From 1967 to 1969 at the University of Pittsburgh he underwent 9 more operations to relieve nostril stenosis, resurface the cheek and eyelid, reposition the ala, lengthen the columella, and revise scars.

II. T.V. (Figure 2) was born in 1946 with a left oro-ocular cleft of the medial type associated with an underlying notched alveolus and a notched vermilion on the opposite side and a thin central prolabium. The palate was intact. From 1948 to 1958 she underwent 10 operations. Reconstruction of the eyelids and medial canthal area proved troublesome and accounted for the major portion of the operations. This patient considers her reconstruction complete.

III. C.N. (Figure 3) was born in 1958 with a right naso-ocular cleft associated with cleft lip and palate. From 1958 to 1971 she underwent 12 operations for her facial anomalies. The first procedure was designed to save the eye and included full thickness skin grafting, tarsorrhaphy, and introduction of a cheek flap. Subsequent surgery has focused about equally on the orbital area, the lip-nose complex, and the palate. Palatal surgery has included a pharyngeal flap. Reconstruction is not complete and at a minimum will require rhinoplasty.

IV. L.B. (Figure 4) was born in 1965 with a right naso-ocular cleft associated with cleft lip and palate and incomplete left oro-ocular cleft of the medial type. Associated anomalies included syndactyly and a congenital band of the right ankle. From 1965 to 1971 she had 9 operations for her facial anomalies beginning with a cheek flap to the right lower eyelid and a Z-plasty of the left lower eyelid.

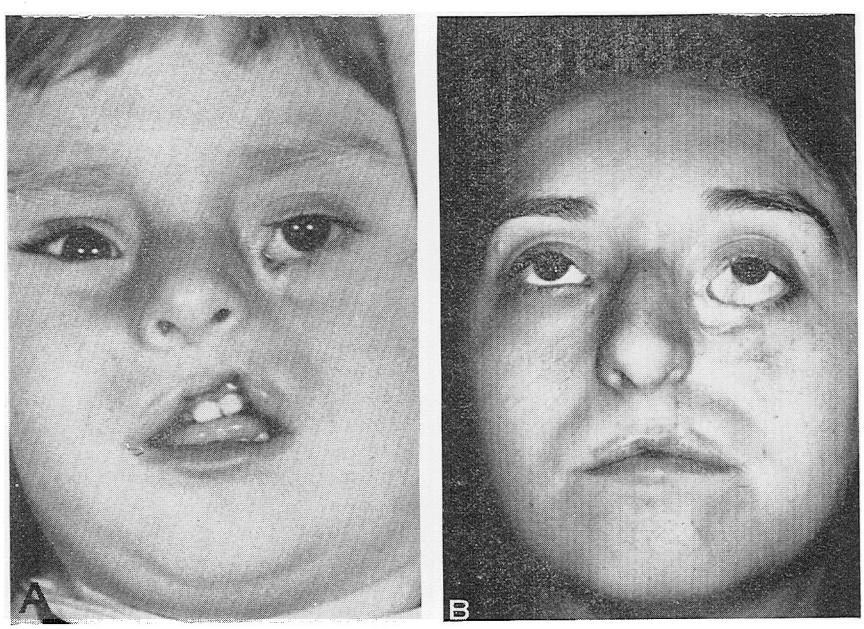

FIGURE 2. (A) Patient T.V. (Case II) prior to treatment. (B) Appearance after 10 operations. Reconstruction of the eyelids and medial canthus proved especially difficult.

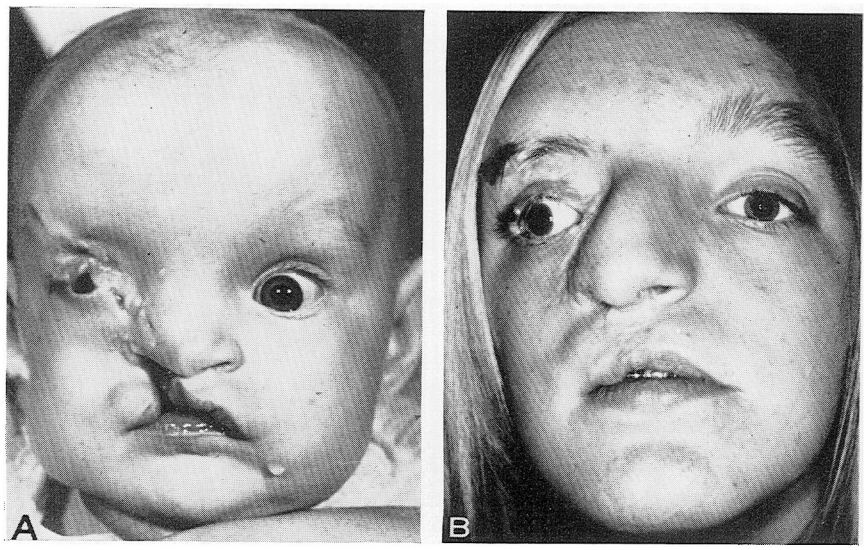

FIGURE 3. (A) Patient C.N. (Case III) prior to treatment. (B) Appearance after 12 operations. Rhinoplasty remains to be performed.

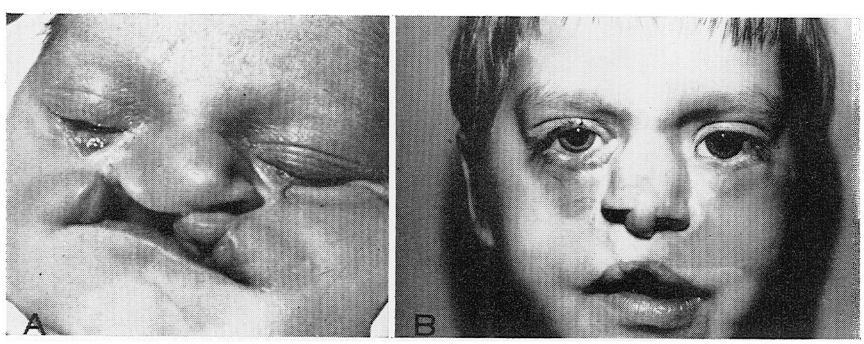

FIGURE 4. (A) Patient L.B. (Case IV) prior to treatment. (B) Appearance after 9 operations for facial anomalies.

Subsequent operations have focused about equally on the eyelids, lip-nose complex, and palate. This patient is still under active treatment.

V. J.N. (Figure 5) was born in 1968 with bilateral oro-ocular clefts of the medial type associated with cleft palate and an independent complete cleft of the left lip. From 1968 to 1971 this patient underwent 6 operations of which 4 have dealt with the protruding premaxilla and repair of an unusually wide double cleft of the palate. Only 2 operations (the first and third) have dealt with facial soft tissue repairs. The rudimentary right eye was removed by the consulting ophthalmologist as part of the first operation. This patient remains under active treatment.

VI. L.S. was born in 1954 with bilateral oro-ocular clefts of the medial type associated with cleft palate and mental retardation. This patient has been lost to follow up.

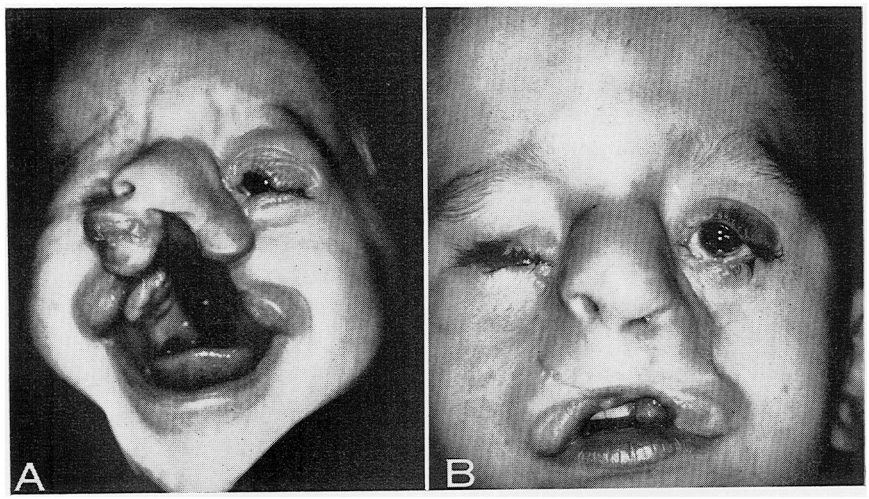

FIGURE 5. (A) Patient J.N. (Case V) prior to treatment. (B) Appearance after 6 operations.

VII. T.H. was born in 1958 with a right naso-ocular cleft extending through the lip, an incomplete left oro-ocular cleft, and cleft palate. This patient also has been lost to follow up.

Summary

Seven additional cases of oblique facial cleft are added to the literature to bring the total reported number to 52.

Oblique Facial Clefts Reported Since 1887

(Live—Born Patients)

	naso-ocular	*oro-ocular*		*mixed*
		medial	*lacteral*	
Boo-Chai (*3*)	13	23	3	4
Bartels et al (*4*)	2			
Wilson et al	3	2		2
total	18	25	3	6

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.

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