THE OBLIQUE FACIAL CLEFT—A REPORT OF 4 CASES

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SUMMARY

The oblique facial clefts are quite rare. 4 such cases are being presented in this paper alongwith review of the literature.

The oblique facial cleft is an extremely rare congenital deformity. Von Kulmus (1732) recorded the first case of oblique facial cleft in Latin. The first case of oblique facial cleft in English medical literature was reported by Walter Dick (1837). Morian (1857) collected 34 cases from the literature and after making a thorough study divided these clefts into type I, type II and type III. Grunberg (1913) wrote a complete chapter on malformation of the face.

In 1962, the nomenclature committee of the American Association for cleft palate rehabilitation recognised two main forms of the oblique facial clefts i.e., the Naso-occular cleft and the Oro-occular cleft. The latter is further subdivided into Oro-medial canthus and Oro-lateral canthus cleft. Khoo Boo-Chai (1970) pointed that in all the cases of the second subgroup, the cleft does not end in the lateral canthus as will be shown in the cases of oblique clefts which are being reported here.

Naso-Occular Cleft

The nomenclature committee of the American Association for cleft palate rehabilitation has defined a Naso-occular cleft as "a fissure extending from the nostril region toward the medial angle of the palpebral fissures".

This may present in two forms, complete form in which the split may start in the region of the lip and involve the nose, the orbit and may go upto the temporal region (Sakurai et al., 1966). This form is very rare. The other form is incomplete form where split may end above the nostril, Khoo Boo-Chai (1970). In my case it started from the lip and extended upto the medial canthus of the eye involving the lateral wall of the nose. Burian (1963) and Ergin (1966) reported that when bone is involved the split lies between the medial and the lateral incisors with the disruption of the aperture pyriformis, and the cleft extends into the orbit. The maxilla is under developed on the affected side and only very seldom it is completely cleft. In these cases the Naso-lacrimal duct is absent or opened.

In the majority of cases the cleft is unilateral, involves the floor of the nostril, divides the ala and extends to the medial canthus (sometimes to the forehead). Cases reported by Gunter (1963) and Davis (1935) were unilateral. Case reported in this paper is also unilateral (Fig. I).

Oro-Occular Cleft

The Oro-occular clefts are further divided in two sub-groups:

Oro-occular cleft type I

Oro-occular cleft type II

In Oro-occular cleft type I, the cleft lies medial to infra-orbital foramen. The cleft instead of involving the nose runs upwards to end either in the inner canthus or in the lower

eyelid. This fissure may extend upto the forehead usually in the temporal region. In severe cases where bone is involved, the split lies between the lateral incisor and the canine. In case No. 2 the cleft started from the usual area of the cleft lip and extended upto the lower eyelid whereas in case No. 3, the cleft on the (L) side occurred lateral to the peak of the cupids bow and extended upto the lower eyelid. In the same patient there was a cleft of the lip only on the (R) side which was present in usual area of the cleft and this was associated with a bifid uvula (Fig. III & IV).

The Oro-occular cleft type II are very rare and in this type the cleft extends from the angle of the mouth upwards towards the orbit ending in the lateral canthus or in a coloboma in the mid portion of the lower eyelid. It's course lies lateral to the infra-orbital foramen and this distinguishes it from type I cleft. Khoo Boo-Chai (1970) reported one unilateral case of this type. One bilateral case of similar type was reported by Pitanguy (1968) whereas three cases of mixed group were seen by Skoog (1969), Greer Walker (1961).

In type II cleft, the point of origin is the same as that of the transverse cleft but it's direction is different. This type of cleft does not correspond to any of the known embryonic facial grooves, and Karfik (1969) calls it the "true oblique cleft". In my case (Fig. V) the cleft started from the angle of the mouth and extended upto the lateral canthus of the eye hence it can be taken as a case of true oblique cleft.

Seven cases of oblique facial clefts were reported by Bryon and Oliver (1981). Their cases include practically all types of oblique

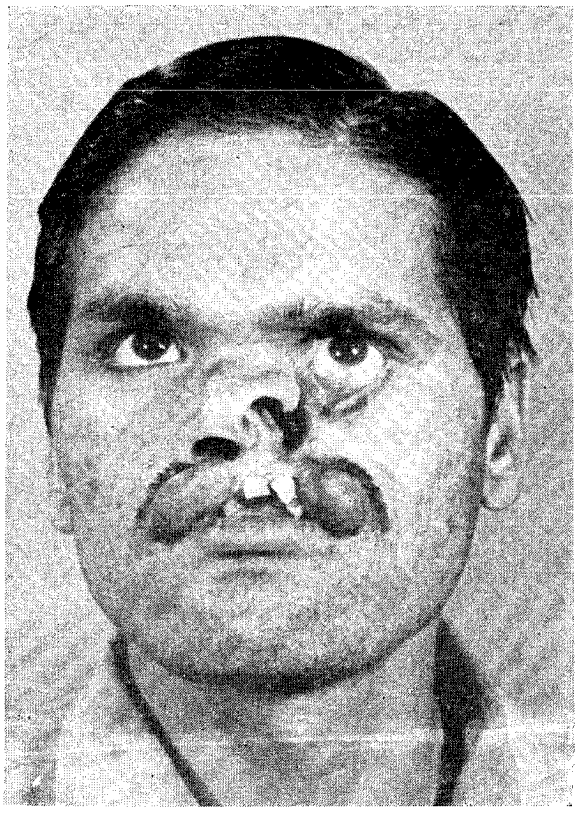

Fig. I. Showing Naso-occular cleft in a 24 year old patient.

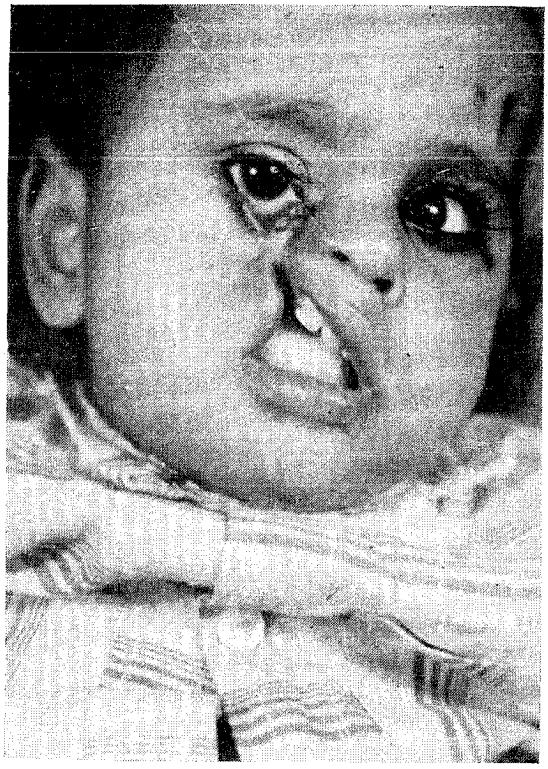

Fig. II. Showing Oro-occular cleft in a 1 year old child.

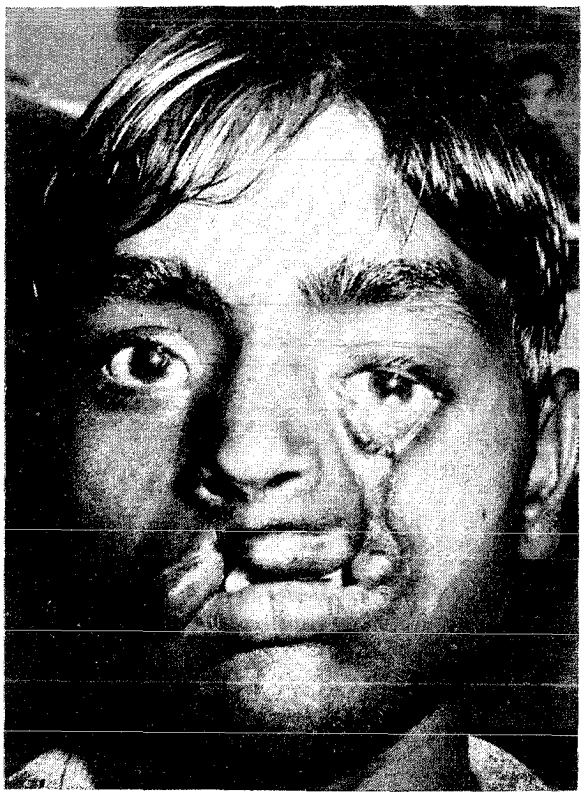

Fig. III. Showing Oro-occular cleft in a 16 year old patient.

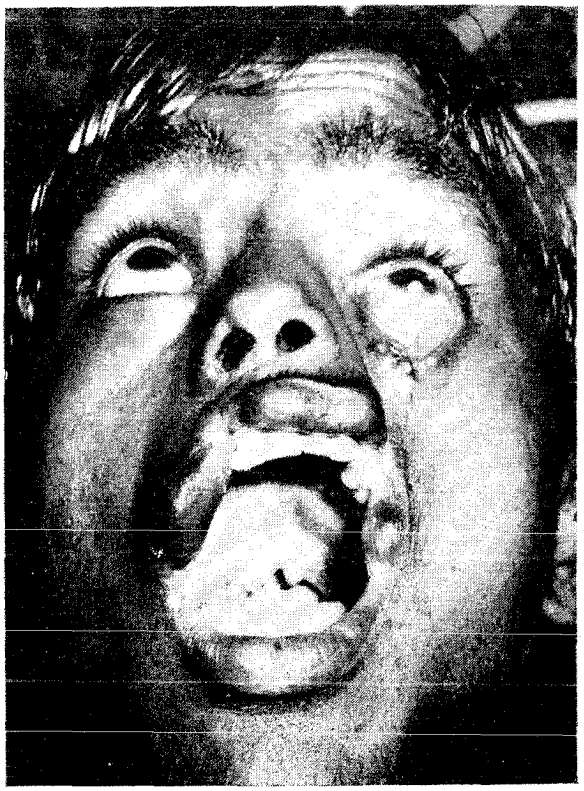

Fig. IV. Showing bifid uvula in case No. 3.

clefts associated with other types of clefts and various other congenital abnormalities e.g. Acrosyndactyly, constriction bands, aplasia cutis of temporal and occipital region and talipes equinovarus. They stated that oblique facial clefts are rare and represent only 0.25% of all facial clefts. In cases of more common cleft lip and palate, the aetiology is generally explained in terms of failure of fusion of the mesoderm of the embryonic facial processes which forms the basis of the international classification. The lateral Oro-occular cleft which extends from the corner of the mouth to the lateral canthus of the eye cannot be explained embryologically. They suggested that these atypical clefts might be the result of constriction due to amniotic bands.

Mixed Type

Apart from the above mentioned types of oblique clefts, Khoo Boo-Chai described a mixed group of four cases in which the various types of clefts occur together one on each side. Thus, the Naso-occular cleft and Oro-occular cleft may occur together (Burian, 1963). Skoog (1969), Greer-Walker (1961) reported that type I and type II Oro-occular clefts existed in the same patient. The oblique facial cleft is also associated with a transverse facial cleft in three cases—two on the contralateral side (Lexer, 1897; Burian, 1963) and one on the ipsilateral side (McEnery & Brennermann, 1937).

Case Reports

Case No. 1: F. C., a male, aged 24 yrs. was seen in plastic surgery O. P. D. He presented with cleft of the upper lip on left side starting at the usual site of cleft lip going upto the medial canthus of the left eye. The lateral wall of the nose was also cleft and there was a colo-

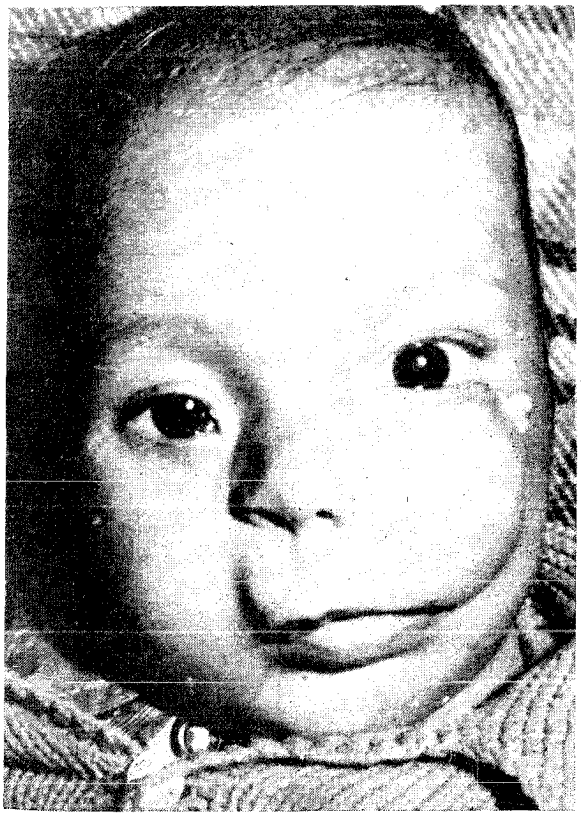

Fig. V. Showing true-oblique cleft in a 3 month old baby.

boma of the left lower eyelid. He was the third child in a family of five. He had a full term normal delivery. All the siblings were normal. He did not have any other congenital abnormality (Fig. I).

Case No. 2: R., a male aged 1 year was brought by his parents for repair of his facial deformity. He had a cleft of upper lip on the (R) side starting from the usual cleft site going upwards upto the lower lid. There was a coloboma in the lower eyelid. The Naso-lacrimal duct was normal. The maxilla on the (R) side was under-developed. Besides this the child had ventricular septal defect. He was the first child of his parents (Fig. II).

Case No. 3: A 16 yrs. old male came to plastic surgery O. P. D. for the repair of his facial deformities. In this patient there was a cleft of upper lip on the left side starting in the region of lateral incisor and canine and going upto the middle of the left lower eyelid. The nostril on the left side was smaller. There was a dense white corneal opacity in the left eye at 6 o'clock position. On the (R) side there was a cleft involving half of the upper lip. Intraoral examination revealed that his uvula was bifid (Fig. III & IV). His speech was very good and he did not have any other congenital abnormality.

Case No. 4: A male child aged 3 mths. had an oblique cleft starting from the left angle of the mouth and reaching the lateral canthus of the left eye and making the cheek area more prominent on the left side. There was a small mole like soft tissue mass present near the lateral canthus (Fig. V). Besides this he had a cleft of hard and soft palate also.

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