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TESSIER'S 0-14 CLEFTS

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SUMMARY

Clefts on 0-14 axis of Tessier have two distinctly different presentations. With a hypoplastic presentation there is an involvement of the forebrain and as 'face foretells the brain' a severe hypoteloric appearance is always associated with severe brain damage and poor life expectancy. With hypertelorism however the spectrum of presentation may extend from the less severe hypertelorism alone to the most severe form of median cleft lip and palate, bifid nose and cranium bifidum occultum which are amenable to corrective craniofacial surgery.

Median facial clefts may have either hypoplasia or hyperplasia as their predominant theme. With hypoplasia, there is tissue deficiency and absence of parts in the midline of the face resulting in median cleft lip, premaxillary hypoplasia, cleft palate, absent or rudimentary columella, vestigeal nasal septum and in extreme cases, just a proboscis with hypotelorism. Congenital absence of skin on the vertex of scalp, congenital forebrain malformation also accompany such cases. On the other hand, there can be an excess of tissue in the midline of face, the lip, premaxilla and palate are all cleft in midline but of normal dimensions with intact central incisors on either side. A bifid nose with extra grooving and increased width of columella, thickened or duplicated nasal septum a broad and flattened nasal bridge and hypertelorism, are other associated features.

Case Reports

Case no. 1

A 4-month old male child, the first issue in the family with no significant prenatal history of the mother, presented with severe malformations of his face and trigonocephaly. Examination revealed a wide central deficiency of the entire height of the upper lip extending into the floor of the nose with an absent columella and a rudimentary nasal septum hanging freely. Intraoral examination showed an absent premaxilla and a complete cleft palate. The nose tip was depressed and the nasal bridge flattened caudally and absent cephalically. There was no palpable skeletal framework comprising the bony nose and a complete absence of projection of the nose in profile. There was severe hypotelorism with eyebrows of either side touching each other in the midline. The intermedial canthal distance was only $\frac{1}{4}$ of the total length of each palpebral aperture (Figs. 1 & 2). The patient died at the age of 9 months.

Case no. 2

A 5-year old girl, first among the 3 issues of normal healthy parents, presented with facial disfigurement and occlusal disharmony of anterior teeth. Examination revealed an absence of the entire vertical dimension of the upper lip in the philtral region, absence of columella, a rudimentary nasal septum hanging freely and a collapsed nose with no projection on profile. Intraoral examination showed a midline palatal cleft extending completely from the alveolar arch to the uvula (Fig. 3). There was absence of both central incisors, the lateral incisor and the canine of both sides were deviated towards the cleft (Fig. 4). There was Angle's class III malocclusion (Fig. 5) because of maxillary hypoplasia. The palate

and lip were operated at 5 years and a cantilever bone graft for nasal projection done at the age of 8 years. An orthognathic surgery is planned once she attains facial maturity.

Case no. 3

A 3-year old boy, the only issue of healthy parents presented with a very wide nose for aesthetic betterment. Examination revealed a

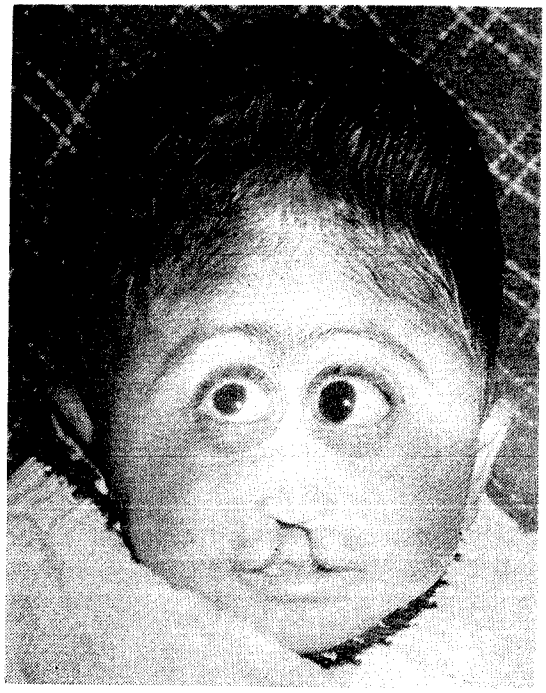

Fig. 1. Median cleft lip and hypotelorism with trigonocephaly front face.

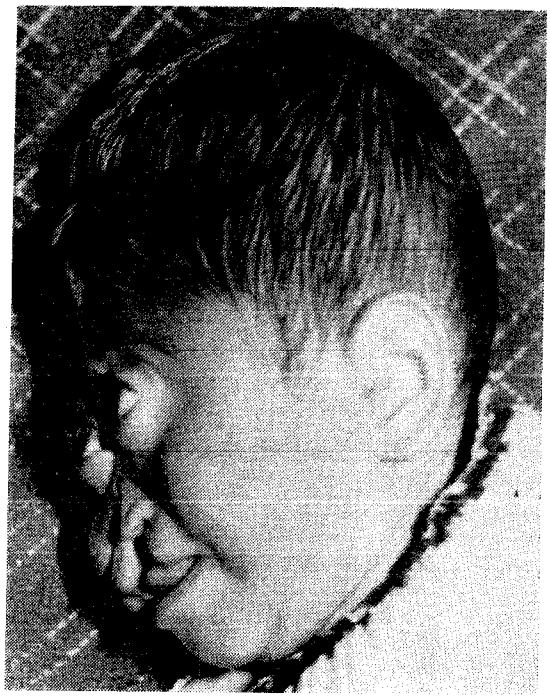

Fig. 2. Median cleft lip and hypotelorism with trigonocephaly profile.

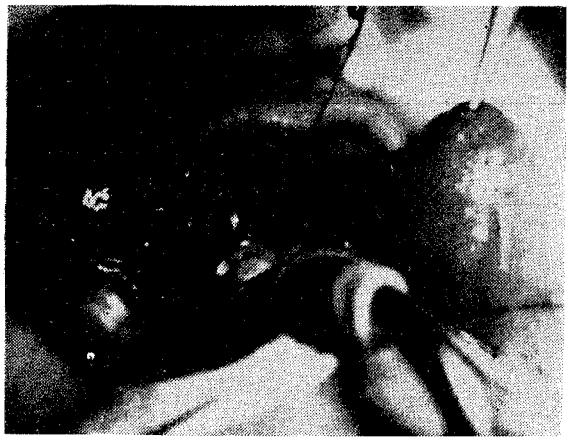

Fig. 3. Median cleft lip alveolus and palate with an absence of columella and vestigeal nasal septum hanging free.

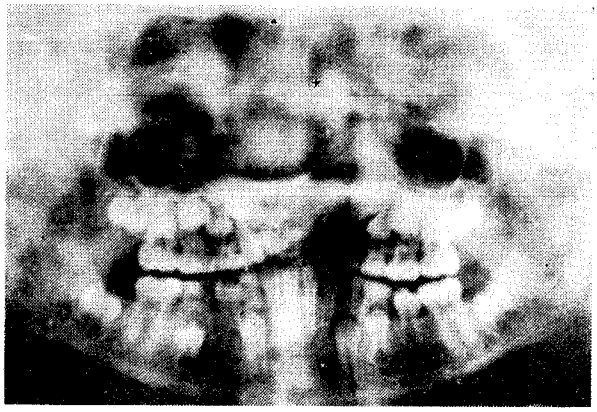

Fig. 4. Orthopantogram of a patient with median cleft lip alveolus and palate showing the midline defect in the alveolar arch extending upto the pyriform aperture, absence of central incisors and lateral incisors deviated into the cleft.

very wide philtrum and columella (Fig. 6). The nasal tip was wide and saddled. The alar and upper lateral cartilages appeared to be displaced laterally and the nostrils were wide apart. The bony dorsum of the nose was wide and the intermedial canthal distance was 4 mm more than the length of individual palpebral fissures. The nasolacrimal ducts were normal on either side and there was no epiphora. The anterior nasal spine was broadened and the

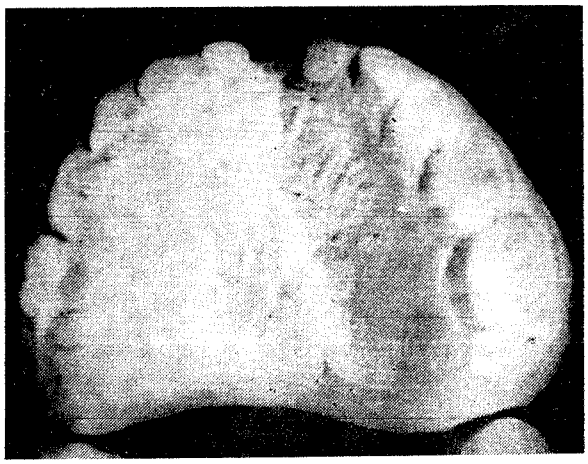

Fig. 5. The dental model of the upper jaw of the same patient.

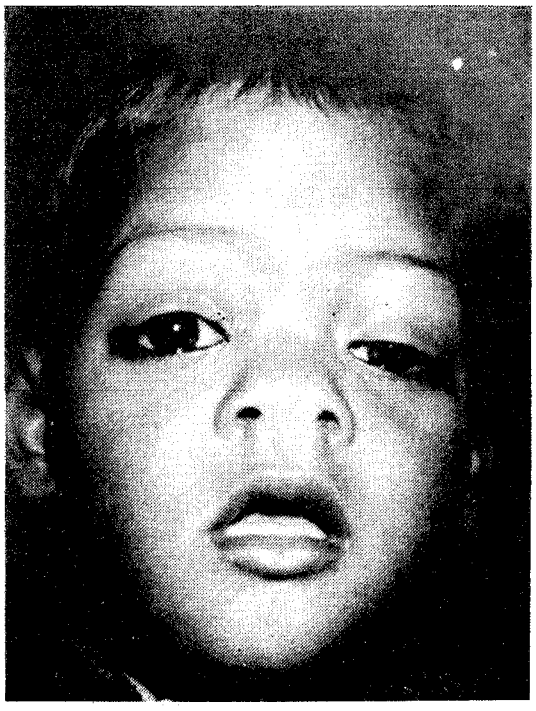

Fig. 6. Mild hypertelorism with a widening of labial philtrum and columella and saddling of the nasal tip.

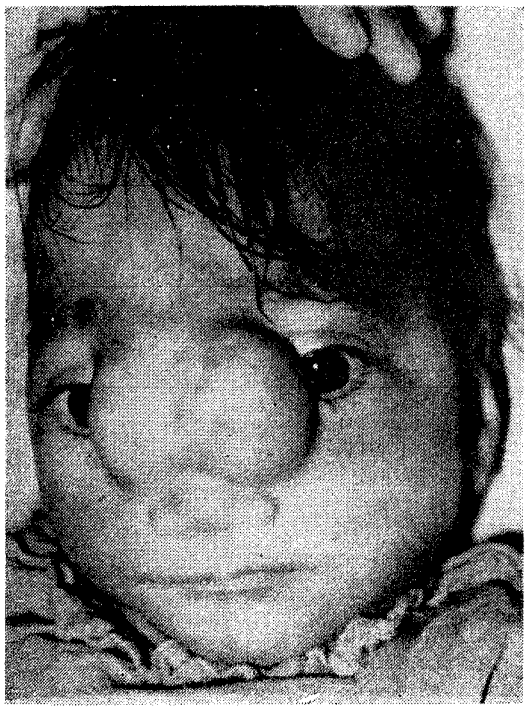

Fig. 7. Frontonasal meningocele with hypertelorism and telecanthus.

nasal septum was grossly thickened. Intraoral examination suggested a wide diastema between 2 central incisors and a large muscular frenulum of the upper lip.

Case no. 4

A $1\frac{1}{2}$-year old girl presented with a large swelling in the root of nose since birth (Fig. 7). The intermedial canthal distance was increased many times by the presence of this fluctuant and transilluminant swelling which had replaced the cephalic 2/3 of her nasal dorsum. The caudal 1/3 of the nose, the lip and palate were normal. The nasolacrimal ducts on either sides were normal. At operation through frontal craniotomy, the cribriform plate was found at an abnormally low level and a bony gap was seen at the root of the nose and the overlying frontal bone through which the meningoencephalocele was seen herniating out.

Discussion

Till Tessier (1973) propounded his classification of craniofacial clefts based on 8 time zones and numbered them 0 to 14, the midline clefts of the face were "either completely omitted (AACPR endorsed classification) (Harkins, 1962) or variously described, precluding a wider understanding. A list of confusing names viz. frontonasal dysplasia (Sedano, 1970), median facial dysraphia, holoprosencephaly (De Myer, 1963), cranium bifidum, median clefts face syndrome (De Myer, 1967) median cerebrofacial dysgenesis (Brucker et al., 1963), gave no indication of the clinical nature of the disease and neither its morbid anatomy. Tessier's classification however provides both with reasonable clarity.

Clefts of midline craniofacial structures as a result of development failure may range from a median cleft in the upper lip caused by imperfect union of paired globular process, to a bifid frenulum, a median notch in alveolus, a midline cleft of palate or a bifid nose, a bony cleft in the intermedial canthal space and resulting median frontal meningoencephalocele and orbital hypertelorism. Developmentally the frontonasal process remains in its embryonic location. The forebrain thus is low and interferes with the normal convergence of optic placodes towards the midline and so the eyes retain their lateralised hyperteloric setting.

At the other end of the spectrum the morphokinetic arrest of the frontonasal prominence produces monsterous hypoteloric malformations viz. cyclopia, ethmocephaly and cebocephaly. Mildest expression of this syndrome is absence of the philtral region of the lip and agenesis of primary palate. Cohen et al. (1971) suggested that the abnormality lay in the floored interaction between the notocordal plate and the neuroectoderm of brain plate and oral plate. Because of the intimate association of the frontonasal prominence with the development of forebrain, the severity of craniofacial malformations appear to parallel that of forebrain (De Myer, Zeman and Palmer, 1964). Yakolev (1959) in a study of 10 cases of arrhinencephaly showed the common denominator of these malformations in a failure of evagination of secondary telencephalic vesicles and of the cleavage of prosencephalon. Prechordal mesoderm situated around the dorsal lip of the foregut induces forebrain differentiation and is also an analouge of medial facial bones. Thus defects of the later as seen in hypotelorism predict a poor forebrain development.

Tessier (1972) emphasized that orbital hypertelorism is not a syndrome as described by Greig (1924), but a physical finding secondary to cranial and facial clefts nos. 11 to 14 and premature synostosis of cranial and facial sutures. Clefts involving frontal bone, particularly those occurring at the junction of desmocranium and chondrocranium favour a downward and forward protrusion of brain and lowering of cribriform plate. Frontonasal and nasoethmoidal meningoceles, dermoid cysts and glial tumours of the root of nose result in cranial clefts which can be associated with orbital hypertelorism.

One should refrain from using the term 'hypoplasia' in place of cleft which is the basic malformation. Hypoplasia does not evolve into clefting, however clefts, whether complete or incomplete, always have hypoplastic edges. An interruption of soft tissue (hairline, eyelid, eyebrow, nostrils, lips and ears) or skeleton differentiates rudimentary clefts from hypoplasia.

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