Median Cleft Of The Lip: A Rare Facial Anomaly

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Citation

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Abstract

Median cleft lip is a midline vertical cleft through the upper lip in the absence of a prolabial ramnant. The incidence is about 1: 10,00,000 births. This may occur as a sporadic event or as a part of an inherited sequence of anomalies. It arises embryologically from incomplete fusion of the medial nasal prominences. The author presents a rare case of median cleft lip,broad nose, wide ala with hypertelorism. The embryology, presentation and surgical technique for treating these cases are discussed.

INTRODUCTION

Median cleft lip is defined as any congenital vertical cleft through the centre of the upper lip. Midline cleft lip defects appear to occur when medial nasal prominence derivatives are deficient or absent (Johnston and Sulik, 1979).

These rare anomalies occur with an incidence in the cleft patient population of 0.43 to 0.73% (Davis, 1935; Knox and raithwaite, 1958; Fogh-Anderson, 1965; Millard and Williams, 1968).

Two major categories of the frontonasal process dysplasias associated with cleft lip have been described:

1. DeMyer sequence: frontonasal deformity associated with hypotelorism, holoprosencephaly, and facial deformity which can range from cyclopia to midline facial cleft with premaxillary agenesis (DeMyer et al., 1964; DeMyer, 1967; Jaramillo et al., 1988)

2. Median Cleft Face syndrome: median cleft lip often associated with nasal deformity, hypertelonism, usually with no or little brain deformity (corpus callosum ageesis) (Millard and Williams, 1968; Weimer et al., 1978).

Surgical correction may be indicated in this second category which is generally quite feasible and should be done early.

CASE REPORT

A male weighing 6.25 kg at full term birth was seen at one month of age. He displayed a midline cleft of the upper lip and phitrum extending one third the distance to the base of the nose, and continuing as a depression and widening of the philtrum to the base of the columella. Hypertelorism as well as flattening of the nose with wide ala were also present. The head circumference was 40 cm. The child was first issue of full term normal delivery. There was no previous family history of any cranio-facial anomalies. The antenatal history was not significant with no exposure to any viral infection, drugs, alcohol, or radiation. There were no associated cardiovascular, gastrointestinal or genitourinary system anomalies.

On examination, the medial cleft was found to be extending to the alveolus with notching. The primary and secondary palate were intact but slightly high arched. The movements of the soft palate were normal.

SURGICAL TECHNIQUE

The cleft lip repair was performed at 4 months of age. The marking of the cleft edges was done in an inverted 'V' fashion. The philtral base was marked to be of 8mm and a columellar base 6mm. The incisions were marked to a point 2 mm above the vermilion border lateral to the defect, which were then carried medially. Incisions were made as per the markings.

The orbicularis oris was dissected anteriorly and posteriorly from the skin and mucosa to the lateral margin of alar base. The abnormal muscular insertions at the nostril sill and alar margins were released. Closure was then completed in straight line fashion beginning with the buccal sulcus. Sutures of 4-0 vicryl were used to repair the musle. Special

emphasis was given to evert the vermilion to create a midline pout. Skin closure was completed with 6-0 nylon. The post-operative period was uneventful. Sutures were removed on 5th day with good cosmetic result.

Figure 1

Figure 1: Preoperative

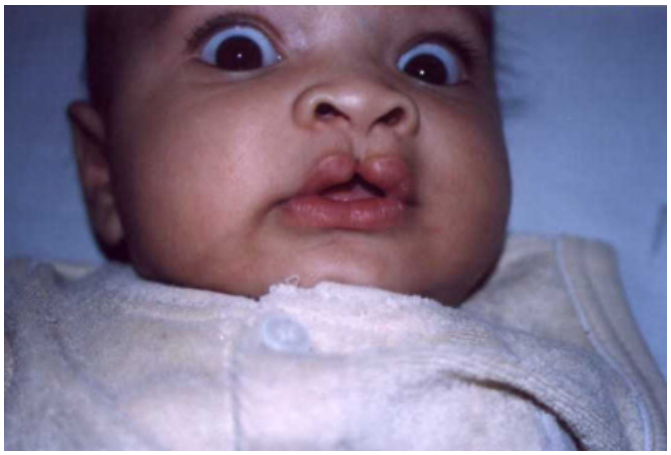
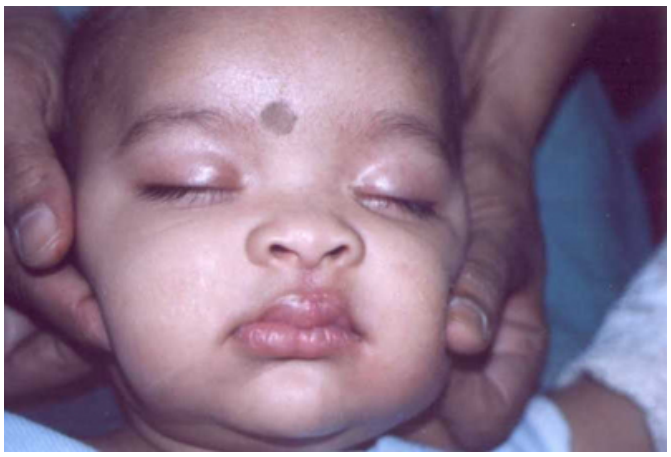


Figure 2

Figure 2: Postoperative



DISCUSSION

The rareness of the median cleft lip and the wide sprectrum of abnormalities observed result in the absence of a common surgical technique suitable for all patients. Various techniques like Francesconi triangular flap technique [1971], Pinto and Goleria's modified Z- Plasty [1971] and inverted 'V' excision technique depending upon the merit of the cases have been described. Repair of an existing shortened or widened columella may be incorporated into the repair (Millard and Williams, 1968; Millard, 1977; Weimer et al 1978). Lengthening of the upper lip is achieved by increasing the length of the incision from lip vermilion border to columellar base at the expense of transverse lip length (Weimber et al, 1978), alternatively, a Z-incision may be added. Creation of a proper midline lip tubercle is important and needs adequate eversion of the vermilion. Each case needs to be carefully assessed with individualization of surgical technique.

The developmental error that results in midline facial defects usually occurs during the third week in gestation. At this time bilateral thickening of the frontonasal process occurs. These thickenings of the surface ectoderm become the medial and lateral nasal prominences that contribute to the formation of the nasal philtrum and upper lip. Fusion of the globular processes is responsible for formation of the anterior portion of the hard palate, the central dentoalveolar ridge, and the central upper lip and philtrum. It is essential that fusion of these prominences occur for normal development of the upper lip. Furthermore, because of the interrelationship between the developing face and forebrain, midline cleft, especially when the premaxilla is absent, may indicate a sequence of associated facial and cerebral anomalies (the face predicts the brain - DeMyer et al., 1963). Since there is a close relationship between the cells of the nural crest and the closing neural tube, defects in these frontonsal process derivatives can lead to severe brain malformations.

Our case report represents the median facial cleft category of frontonasal dysplasias. The patients presented with typical features of median cleft lip, mild alveolar notching, flattening of nose, widened ala with hypertelorism.

Other deformities which may also be present with median facial cleft category are V-shaped frontal hairline (widow's peak), cranium bifidum occultum, telecanthus, and median cleft nose and palate (DeMyer, 1967). Despite the anomalies, these patients frequently have normal intelligence. Often agenesis of the corpus callosum may be seen which is some times associated with seizures, but is usually asymptomatic (Lynn et al., 1980). These patients usually have a normal life expectancy and reconstruction should be performed.

Patients falling into the first group (DeMyer sequence) generally have bad prognosis. These rare deformities may be genetically transmitted as autosomal dominant or recessive traits (Jaramillo et al., 1988). With the presence of family history of median facial dysmorphias the possibility of recurrence in a subsequent child is as high as 50% (DeMyer, 1967). Total agencies of the premaxilla is frequently associated with alobar holoprosencephaly or failure of

cleavage of the prosencephalon. These patients are severely mentally retarded, and their life expectancy is correspondingly short (DeMyer, 1975). Reconstruction is therefore not indicated in such patients.

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