

Median Cleft Of The Lip: A Rare Facial Anomaly

S Saraf

Citation

S Saraf. *Median Cleft Of The Lip: A Rare Facial Anomaly*. The Internet Journal of Plastic Surgery. 2005 Volume 2 Number 2.

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 andraithwaite, 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, hypertelorism, usually with no or little brain deformity (corpus callosum agenesis) (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 philtrum 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 muscle. Special

emphasis was given to evert the vermillion 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 spectrum 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 vermillion 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 vermillion. 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 neural crest and the closing neural tube, defects in these frontonasal 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 agenesis 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.

References

1. DAVIS WB. Congenital deformities of the face : types found in a series of one thousand cases. Surg Gynecol Obstet 1935; 61:201-209.
2. DEMYER W. The median cleft face syndrome. Neurology 1967; 17 : 961-971.
3. DEMYER W. Median facial malformations and their implications for brain malformations. Birth Defects 1975; 11:155-181.
4. DEMYER W., ZEMAN W., PALMAR CG. The face depicts the brain : diagnostic significance of median facial anomalies for holoprosencephaly (arhinencephaly) with median cleft lip and palate. Pediatrics 1964; 34 : 256-263.
5. FOGH -ANDERSON P. Rare clefts of the face, Acta Chirurgica Scand 1965; 129 : 275-281.
6. FRANCESCONI G., 'Bifid nose. Median Dysraphia of Face' Mustarde J.C., (Ed.), Plast. Surgery in Infancy and Childhood. Edinburgh, Livingstone; 1971.
7. JOHNSTON MC, SULIK K.K. Some abnormal patterns of development in the craniofacial region. Birth Defects 1979; 15:23-42.
8. LYNN RB. BUCHANAN DC, FENCHEL GM, FREEMON FR. Agenesis of the corpus callosum. Arch Neurol 1980; 37 : 444-445.
9. MILLARD DR. MEDIAN CLEFT LIP WITH HYPERTELORISM. In Millard DR. Cleft Craft. Vol. II Boston : Little, Brown, 1977 : 758-759.
10. MILLARD DR, WILLIAMS S. Median Cleft of the upper lip. Plast Reconstr Surg 1968; 42:4-14.
11. PINTO, C.J.T. AND GOLERIA K.S. The Median Cleft Lip, is Hueston, J.T. (Ed.) The Median Cleft Lip, in Hueston, J.T. (Ed.) Transactions of the 5th International Congress of plastic and reconstructive Surgery, Australia, Bulterwaths, 1971.
12. WEIMER DR, HARDY SB, SPIRA M. Anatomical findings in median cleft of the upper lip. Plast Reconstr. Surg 1978; 62: 866-869.

Author Information

Sanjay Saraf, MS, MCh. (Plastic Surgery), DNB Plastic Surgery, MNAMS
Specialist Plastic Surgeon, NMC Specialty Hospital