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CONGENITAL MEDIAN CLEFT LIP: A VERY RARE PAEDIATRIC FACIAL ANOMALY

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ABSTRACT
The median cleft lip is a midline vertical cleft through the upper lip. This is a very rare anomaly and very few cases have been reported in the literature. This is also known as Tessier number 0 cleft. It arises embryologically from incomplete fusion of the medial nasal prominences. The author presents a rare case with median cleft of the upper lip. The patient has no other anomalies of the nose or alveolus.

INTRODUCTION

The median cleft of the upper lip is defined as any congenital vertical cleft through the centre of the upper lip [1]. These are very rare facial anomalies with an incidence of 0.43 to 0.73% out of the craniofacial cleft population [2,3]. The midline cleft lip defects appear to occur when medial nasal prominence derivatives are either deficient or absent [4,5].

The developmental origin of a median cleft of the upper lip is still unclear. It is suggested that the defect of the midline face can be traced to the third week of gestation. During this period bilateral thickenings of the frontonasal process appear. The facial embryology shows that the upper lip forms from the fusion of the inferior medial nasal prominence, lateral prominence and the medial aspect of the maxillary process. The median cleft results from the failure of the two medial nasal processes to fuse in the midline [6].

Millard and Williams [4] suggested that any congenital cleft through the centre of the upper lip constitutes a median cleft of the lip. They classified the median clefts of the upper lip into three categories (1) A notch confined to the vermilion. (2) The cleft extends to involve the columella. (3) A defect caused by lack of development of the whole median element.

The authors here describe a rare case of a median cleft of the upper lip with normotelorism and no other malformations. This is the first case of an isolated median cleft of the upper lip from our institution and authors wished to add this to the present literature.

CASE REPORT

A 7 month old male child was presented to the Department of Pedodontics and Preventive Dentistry, Post Graduate Institute of Dental Sciences, Rohtak for dental evaluation of midline gap in relation to upper lip since birth. The mother reported that the child was born with normal full term pregnancy with no complications. The mother did not have any diseases during the pregnancy. History did not reveal any habit of smoking or alcohol. Further, 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. The child birth weight was normal (3.24 kilogram) and presented with no other anomalies. There was no past family history of clefts or any other cranio-facial anomalies. The patient has normal intelligence. Extraoral examination revealed a midline cleft of the upper lip since birth. The nose as well as the inter-

canthal distance was normal (Figure 1). The total length of the vermillion and the vertical height were normal. The vermillion was separated in the midline (Figure 2). On intra oral examination, there was a median cleft of the upper lip with a shortened bifid frenulum (Figure 3). The primary and secondary palates were intact but slightly high arched. The movements of the soft palate were normal. There were no bony abnormalities seen and the alveolus was normal. All routine laboratory investigations have been normal.

CT scan of the head confirmed normal intracranial structures and no evidence of intracranial abnormalities. MRI scan of the spinal cord was normal. The patient was put under regular dental follow-ups and had been referred to higher medical centre for further pediatrician and neurological evaluation.

Figure 1. Extra-Oral View of the patient showing medial cleft of the upper lip

Figure 2. Magnified View showing fissured vermillion border of upper lip

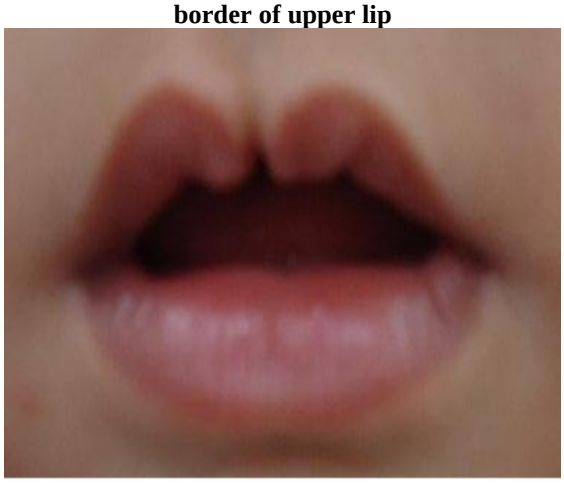
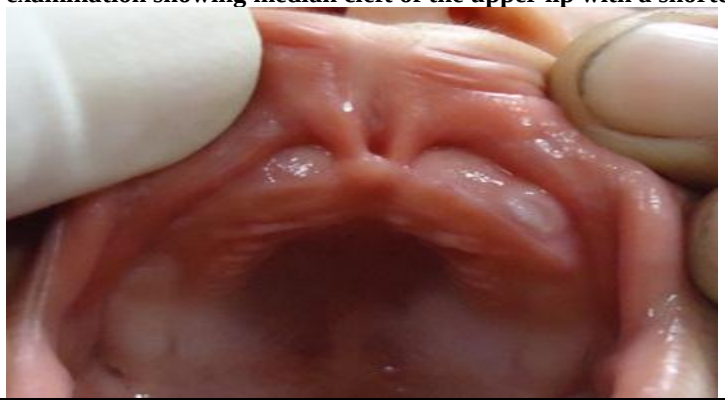


Figure 3. Intra oral examination showing median cleft of the upper lip with a shortened bifid frenulum



DISCUSSION

A median cleft of the lip is defined as a vertical cleft through the center of the upper lip. This is a rare facial anomaly with an incidence of 0.43 to 0.73%. The exact etiology is not known. There have been a few reports in the literature describing different variations of the median cleft lip [3,4].

The recent advances in the embryological study revealed that the developmental error results in midline facial defects usually occur during the third week in gestation. This is the time when bilateral thickening of the frontonasal process occurs. These thickenings of the surface ectoderm divided to form medial and lateral nasal prominences that contribute to the formation of the nasal philtrum and upper lip. The 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. 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. However, the present case reported no signs of any neurological involvement [7].

The Veau in 1937 [8] classified three varieties of median clefts: 1) notch of the lip, 2) median cleft extending to the columella, 3) a defect due to atrophy of midline facial structures. DeMyer [7] described two groups of

syndrome associated with the median cleft lip. The first is associated with orbital hypotelorism and the second with hypertelorism. Furthermore, Millard in 1968 [4] classified a median cleft of the lip as any congenital or vertical cleft through the center of the upper lip, regardless of the extent. Median clefts have been divided into two groups by Millard. The first group involves agenesis of the frontonasal process, and the second group is described as a cleft of the median element. The latter is associated with various degrees of bifurcation of the nose, hypertelorism, and cranial malformations. In this case report, the patient described had an isolated median cleft of the upper lip without any deformities of the nose, philtrum, or alveolus. There was a short bifid frenulum present. The patient also had normotelorism in contrast to the prior descriptions by Millard and Veau. These patients usually have a normal life expectancy and reconstruction should be performed with expert opinion.

CONCLUSION

An isolated median cleft of the upper lip was presented with detailed patient history and examination. This is a very rare facial deformity.

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CONFLICT OF INTEREST:
The authors declare that they have no conflict of interest.

STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

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