

Case Report

Median cleft lip: an uncommon and unique anomaly

Vivek Parameswara Sarma*

Department of Paediatric Surgery, S.A.T. Hospital, Government Medical College, Thiruvananthapuram, Kerala, India

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*Correspondence:

Dr. Vivek Parameswara Sarma,
E-mail: vivsarma@gmail.com

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ABSTRACT

Median or midline cleft lip [MCL] is an uncommon anomaly characterized by a midline vertical cleft through the upper lip and are either isolated or part of multiple anomalies. It can involve the pre-maxilla, the nasal septum, and the central nervous system. MCL includes Complete (42%), Incomplete (49%), and Minor forms (9%). The three main groups distinguished were: 1. Isolated MCL; 2. MCL with craniofacial malformations; and 3. MCL with extra-facial malformations. To analyze two operated cases of median cleft lip and review the relevant literature. The details of two cases of median cleft lip that were operated in 2017 were analysed. Both cases underwent wedge excision with the classical inverted V incision and muscle reconstruction with satisfactory result. Both the patients had no syndromic association or associated anomaly. All cases of MCL require evaluation for associated abnormalities. Isolated MCL can be repaired surgically with a good outcome.

Keywords: Median cleft lip, DeMyer sequence, Median cleft face syndrome

INTRODUCTION

Median or midline cleft lip [MCL] is an uncommon anomaly characterized by a midline vertical cleft through the upper lip and are either isolated or part of multiple anomalies.¹ It can involve the pre-maxilla, the nasal septum, and the central nervous system.^{1,2} In a review of 209 patients reported in literature, MCL included complete (42%), incomplete (49%), and minor forms (9%). The three main groups distinguished were: 1. Isolated MCL; 2. MCL with craniofacial malformations; and 3. MCL with extra-facial malformations.¹⁻³ To analyze two operated cases of median cleft lip and review the relevant literature.

CASE REPORT

The pre-operative, operative and post-operative details of two cases of median cleft lip that were operated in 2017 were analyzed.



Figure 1: Pre-operative picture of the appearance of classical midline cleft lip.

The first patient was a 4 month old female (Figure 1) and the second one was 5 month old male. Both the patients had no syndromic association or associated anomaly.

Both cases underwent wedge excision with the classical inverted V incision and muscle reconstruction with satisfactory cosmetic result (Figure 2).



Figure 2: Post-operative appearance of operated midline cleft lip.

DISCUSSION

The incidence of MCL has been reported to be 0.4% to 0.7% in the cleft lip population.¹ MCL can occur as a sporadic event, or as a part of an inherited sequence of anomalies. Fusion of the inferior medial nasal prominence, the lateral prominence, and the medial aspect of the maxillary process of the first visceral arch results in normal development of the upper lip. MCL occurs due to incomplete merging of the median nasal prominences which form the inter-maxillary segment.^{2,3} Two major categories of MCL are described:

- Demyer sequence: frontonasal deformity associated with hypotelorism, holoprosencephaly and facial deformity which ranges from cyclopia to midline facial cleft with pre-maxillary agenesis.
- Median cleft face syndrome: characterised by nasal deformity, hypertelorism and less chance of brain deformity (corpus callosum agenesis).²⁻⁴

Radiographic imaging for the diagnosis of the associated alveolar defect and accompanying bone defects should be performed by preoperative/postoperative evaluation using 3-dimensionally reconstructed CT imaging.^{3,4} Long-term follow-up of the bone defect for treatments including alveolar bone grafting is required. When indicated, iliac bone grafting with a conventional method can manage the bone defect.²⁻⁴

Various surgical techniques have been described for MCL including wedge excision with inverted-V incision, inverted-U incision and forked flap Z plasty repair.⁴⁻⁶ The surgical technique here adopted was Inverted-V excision of the upper lip and repair of the orbicularis oris muscle. As recommended by Millard, a combination of an inverted-V excision and a 90-degree angle in the excision 2 mm above the muco-cutaneous white roll on each side of the cleft results in lengthening of the skin on the midline of the Cupid's bow.^{5,6}

This technique also reduces the philtrum transversely; therefore, a natural appearance of the Cupid's bow can be achieved.⁵⁻⁷ Lengthening of the midline skin can be achieved with inverted-V incision right above the muco-cutaneous white roll.⁸ Excessive excision of the skin can lead to hypertrophic scarring, whereas insufficient excision can lead to an unnatural depression on the midline of the philtrum.⁷⁻⁹

CONCLUSION

All cases of MCL require evaluation for associated abnormalities. Isolated MCL can be repaired surgically with a good outcome.

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