



# Median Cleft Lip: A Rare Malformation, Best Prognosis for the Isolated Form. Case Report

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## Summary

The median cleft lip is a very rare form of cleft lip. It can be isolated or to be part of a complex malformation association which can involve the premaxillary bone, the nasal septum, or even the brain. In some cases, it is part of a syndrome. For the management of the median cleft lip, the excision with inverted-V incision or inverted-U incision is the most used. Muscle repair is the main step of surgery. We report the case of an incomplete and isolated median cleft lip in a 14-years-old boy. The inverted-V excision technique was used. The aesthetic result was satisfying. The isolated form has generally a good prognosis.

**Keywords:** Median cleft lip; Median cleft face syndrome

## Introduction

The medial cleft lip is a very rare form of cleft lip [1]. It can be isolated, characterized by a midline vertical cleft of the upper lip. It can also involve the premaxillary bone, the nasal septum or even the central nervous system, entering into the framework of a syndrome in which the cleft is only one element [2]. The age and chronology of care must take into account any associated malformations. We report a case of isolated medial cleft lip diagnosed and treated at the age of 14 years old.

## Case Report

Our observation concerns a 14-year-old boy from a landlocked area sent by a missionary priest for a medial cleft lip. He presents an incomplete medial cleft lip with bifidity of the labial brake (Figures 1 & 2). The facial x-ray and the cranio-cerebral CT scan were normal. It was thus an isolated form. No similar family case was noted. The surgery was done under bilateral suprazygomatic maxillary nerve block supplemented by a labial infiltration of xylocaine-epinephrine 1%. The inverted V technique was adopted. The incision extended to the tops of each hemi-tubercle, cutting the crests of each labial hemi-brake before joining at the base of it (Figures 3 & 4). A complete disjunction of the orbicularis muscle

has been observed. This muscle was dissected and sutured with Vicryl® 4/0. The aesthetic result was satisfying. We obtained an ad integrum reconstruction of the philtrum, cupid's arch, lobule and respect for the alignment of the red lip-white lip junction.



Figure 1: Incomplete Medial cleft lip.



Figure 2: Bifidity of the labial brake.



Figure 3: Traces of the incision.



Figure 4: Traces of the incision.

## Discussion

The medial cleft lip is an extremely rare malformation [1]. It represents 0.4% to 0.7% of all cleft lips and affects around 1 case per 1,000,000 births [2,3]. Embryologically, during the fourth week of pregnancy, the fusion at the midline of the distal part of the internal nasal bud with the lateral nasal buds and the maxillary buds insures the normal formation of the upper lip. The failure of this fusion is responsible of the medial cleft lip [4,5]. There are three groups of medial cleft lip: group I for the isolated form, group II for clefts with craniofacial malformations and group III for forms associated with extrafacial malformations [1]. This malformation can be part of a syndrome. Medial facial cleft syndrome associates medial cleft lip, nasal deformity, hypertelorism with or Pithou malformation of the central nervous system [6]. Thus, the diagnosis must include the systematic search for associated malformations. Facial x-ray and craniofacial scan are of classic indication [3,6].

For the treatment, the age and the chronology of the surgery depend on the possible associated malformations [7]. In case of alveolar bone defect, an iliac graft is conventionally used [6,8]. For the management of the medial cleft lip, several operating techniques have been described. The most commonly used are inverted V and U excision [4,6]. According to Millard, the use of inverted V excision and 90° angle in the excision, 2 mm above the mucocutaneous white roll on each side of the cleft which lengthened the skin in the center of Cupid's bow [4,9]. Muscle repair is the main step of treatment. Even for incomplete forms, a defect in the orbicularis muscle is almost constant [10,11]. Skin excision must be well calculated. Excessive excision can lead to hypertrophic scarring, whereas insufficient excision can lead to an unnatural depression on the midline of the philtrum [7]. For the isolated form, the aesthetic outcome is generally satisfying.

## Conclusion

In case of a median cleft lip, the treatment must take into account any associated malformations. The inverted V excision gives an excellent aesthetic result for the reconstruction of the lip.

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