

## Review Article

# True Median Cleft Of Upper Lip: Review Of Anatomic Changes And Surgical Management

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### ABSTRACT

The median clefts of the facial skeleton and soft tissue are least in occurrences. The relative low incidence of such type of clefts especially those involving just the lip the detailed anatomy and reason for incidence of such clefts are not reviewed or discussed extensively in recent past. Various clinical innovations in the cleft surgery has drastically improved the results with the clear understanding of the anatomic changes. This article would discuss the detailed changes in the anatomy of mid-line upper lip cleft.

### INTRODUCTION

The unilateral clefting of the lip (soft tissue) clefting is the most common cleft which is commonly associated with cleft of palate and alveolus<sup>[1-5]</sup>. Among the incidence of the clefting of lip, the incidence of the median clefting of the upper lip is very rare followed to that of the incomplete cleft lip.<sup>[3,5]</sup>

This article would bring in notice the anatomic description of the midline cleft and would depict its difference from that of other clefts. We have attempted to represent the changes

diagrammatically to clearly distinguish the changes that occur

### Morphopathogenesis:

The exact etiology and pathogenesis of the median cleft of facial skeleton is not known. Various descriptions have been given by different authors with the changing time but none of the theories have been able to describe the exact reasoning.<sup>[6]</sup>

The proposed morpho-pathogenesis would be clearer after understanding about the various

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process involved in the development of face. The individual structures that develop from

Summarizing the whole detail regarding the development of various facial structures, the upper lip is formed from the ectodermal placodes of the frontonasal process and the elevated mesodermal placodes that would fuse in the midline to form the complete upper lip. Any form of mechanical or inductive disturbances would lead to failure of fusion of the process causing an upper lip midline cleft. Even the origin of the tesseris 0 is not clearly understood. The tesseris 0 cleft occurs in the 3<sup>rd</sup> week of gestation. The most probable reasons which is widely accepted now a days by most authors is the failure of medial nasal processes to fuse.<sup>[8]</sup>

### **Review of Anatomy:**

The anatomic variation of the median cleft lips are associated with the fusion that fails to occur and the location of the placodes where the fusion does not occur.

In the median clefts of the upper lip the fibers of the orbicularis oris do not fuse with each other in the mid-line. Clefts are often associated with excess soft tissue bulk in the line of the cleft and this remains the same in the

each process have been mentioned in the table given below<sup>[7]</sup>

mid-line cleft of upper lip. The fibers are positioned obliquely along the cleft margins.

### **Associated features:**

The median cleft are often associated with various deformities in the brain along with the facial structures. This was widely studied in depth by De- Myer. He published various articles on this aspect of the median cleft. He in 1963, in his article mentioned that “there is inter- relationship between the face and the brain which may initiate a sequence of associated facial and cranial anomalies.”<sup>[9]</sup>

The authors in the recent past has classified the median clefts mainly into two types i.e the true median cleft lip and the false median cleft lip.<sup>[10]</sup>

True median cleft lip has following associated feature that differentiates it from false median cleft

1. Median cleft upper lip
2. Duplicated labial frenulum.
3. The nasal septum is duplicated.
4. A wide diastema of the upper central incisor

S. No.	Process involved	Structures derived
1.	Lateral nasal process	Ala of nose Lateral crus Lower lateral cartilage Upper lateral cartilage External bones of nose Lateral wall of nose
2.	Median nasal process  Caudal portion	One half of the nasal septum Medial crus of lower lateral cartilage Nasal tip Columella Prolabium Frenulum Palate ( primary )
3.	Naso-frontal process	Midline of upper lip

teeth may be present.

Whereas, the false median cleft lip is characterized by absence of philtrum and columella, premaxillary bone, nasal septum, and crista galli.

#### **Review on description of various clefts:**

The median cleft are often associated with various deformities in the brain along with the facial structures. Veau was the first to observe these deformities and had classified into three varieties as clefts causing notching of the upper lip, midline cleft up to columella and defect causing atrophy of midline structures in 1937.

This was then widely studied in depth by De-Myer. He published various articles on this aspect of the median cleft. He in 1963, in his article mentioned that “there is inter-relationship between the face and the brain

which may initiate a sequence of associated facial and cranial anomalies.”<sup>[9]</sup>

Millard however told that not all midline clefts are associated with the brain anomalies. The defects due to the dysplasia of the frontonasal process were divided into two categories by Millard in which one was midline cleft face syndrome and other according to him was the De-Myer sequence. In the midline cleft face syndrome no or little brain deformity was noted which was commonly associated with the corpus callosum agenesis. Such defects and deformity showed association with nasal deformities also. However the patient who were affected by cleft face syndrome had normal intelligence and life expectancy with no risk or life threatening anomalies associated. The De-Mayer sequence on the other hand is associated with hypotelorism and holoprosencephaly. The patients affected with

this syndrome have a shorter duration of life along with severe mental retardation. This is generally autosomal dominant or recessive in pattern of inheritance.<sup>[11]</sup>

Millard and Williams in 1968 however modifies the classification and divided the median cleft of the lip into two groups. The first group showed agenesis of the frontonasal process and the other that is described to be the cleft of the median element.<sup>[11]</sup>

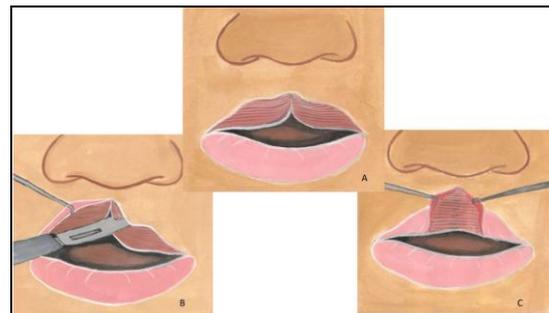
After 1968 various authors extensively studied the median cleft and classified them into true and false median clefts. The true median cleft according to various authors had no brain malformations or nose split or other features. The false median cleft on the other hand is considered to be subtype of holoprosencephaly.<sup>[12]</sup>

In the surgical point of view even the slightest change in the symmetry or the contour of face. In the median clefts reconstruction is more challenging in terms of reconstruction of properly contoured cupid's bow and vermillion border. The closure of the midline cleft is relatively simple but complicated by the vast diversity of presentation of the medial clefts. The lengthening of the upper lip is achieved by triangular flaps or increasing length of the incision and compensate for the hypoplastic lip which is due to the disturbance in the normal

development of the lip.<sup>[6]</sup>

The anatomic orientation of the orbicularis oris muscle obliquely along the cleft (Figure 1A) and the muscle dissection and reorientation of the muscle is clearly demonstrated in Figure 1B. The muscular repositioning of the fibers and closure in the midline remain same in all the techniques with the modification along the skin which remains the anatomic basis for treatment of the midline clefts (Figure 1C).

**Figure 1: Anatomic Basis of treatment of Midline Cleft. A. Orientation of the muscle fibers B. Dissection along the cleft margins C. Repositioning of the Muscle in the midline**



The detailed understanding of the midline cleft concludes that the usage of “z plasty” and modified incision on the skin has provided adequate lengthening of the lip. The oblique insertion of the fibers of the orbicularis oris is

directed to more straight line insertion.

### Review of the various surgical techniques:

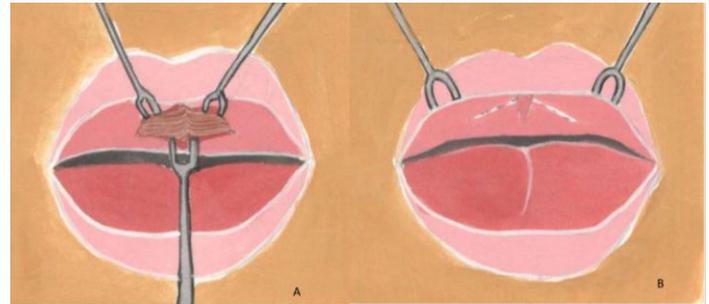
The surgical technique which is involved for the correction of the median cleft lip is mainly aimed at correction of the orbicularis oris muscle orientation and to achieve equal lip length bilaterally with adequate symmetry and function. The orbicular oris muscle in median cleft is mainly by transverse repair except in Z Incision pattern or paired v incision.

#### The inverted V incision: (Intraoral technique)<sup>[6]</sup>

This incision was explained by miller in which he used a inverted v incision. The two incisions on the left and right side of mid-line formed 90 degree to each other.(Figure 2A) The excision of tissue was done 2mm above the mucocutaneous roll on each side of the cleft. This was followed by dissection and release of muscle bilaterally till the nasolabial folds bilaterally and then the muscle was then approximated in the center. (Figure 2B)

The noted advantage of this technique was increase in the length of skin in the cupid's bow. This however was reviewed and found to have visible scar.

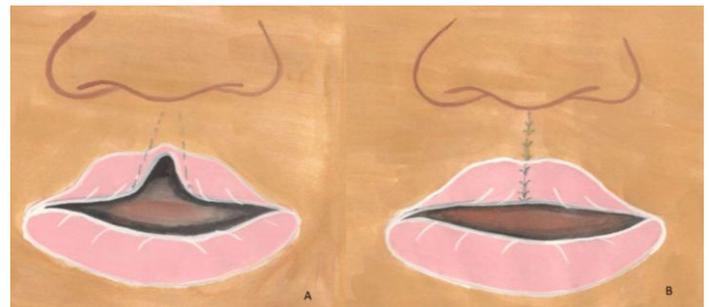
#### Figure 2: Inverted V incision – Intraoral technique



#### Straight line closure:<sup>[6]</sup>

The earliest attempt was given to close the midline defect in a straight line by just excision of the hyperplastic tissue.(Figure 3A) The complete muscle dissection was done to release the muscle from their abnormal position and then the closure was done in the midline to achieve esthetics. (Figure 3B) The disadvantage was the decrease in the total lip length which was observed in this technique.

#### Figure 3: Straight line Closure

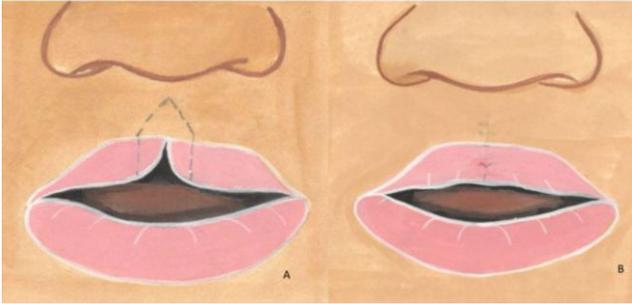


#### The V-Y advancement flap:<sup>[6]</sup>

The use of this technique for the midline cleft is done in patients along with abnormal frenula attachments. The dissection is done in the midline. In cases with abnormal frenum

attachment it is observed that the muscle is attached abnormally into nasal sill. Abnormal muscle attachments are separated from the base of the nose. Medial bands are then excised and muscles are then approximated in the midline.

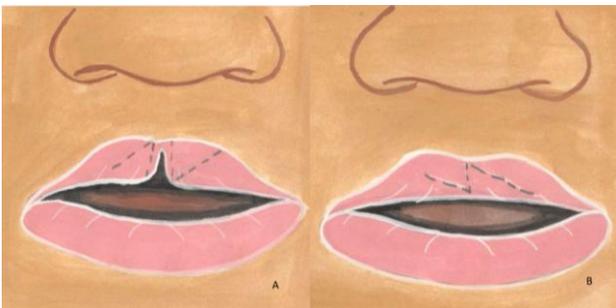
**Figure 4: V-Y advancement flap**



**Paired inverted 'V' pattern:<sup>[13]</sup>**

Forked flaps were marked and inverted 'V' incisions were used. (Figure 5A) The abnormal muscular incisions were released and suturing of the orbicularis muscle was done in 'z' pattern instead of transverse repair. (Figure 5B) Two triangular flaps were marked on skin followed by skin closure.

**Figure 5: Paired inverted V**

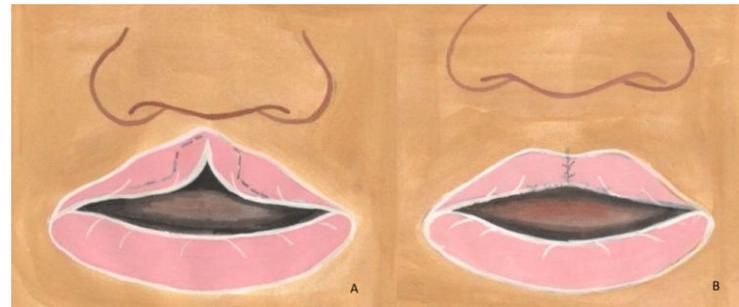


**Modification of Z plasty:<sup>[14]</sup>**

In the traditional "Z plasty" the markings were done on the lip to mark the

midpoint of the future cupid's bow. The modified z incision had marking on cleft margins and involve the midpoint of future cupid's bow. (Figure 6A) The incision continued perpendicular to reach the red line and then followed the wet line on the upper lip posteriorly to form modified 'Z'. (Figure 6B) This modification was employed to limit the increase in the total lip length following cheiloplasty.

**Figure 6: Modified 'Z' Plasty**



**Conclusion:**

The median clefts are the rarest of the rare cleft in the maxillo-facial region. The incident of the median cleft ranges between 0.4 to 0.7 % even among the clefts <sup>[7]</sup>. As this defect is symmetrical most of the patients do not undergo any treatment for the same as they do not affect the adjacent structures. The deformity is subtle and seldom reported to various centers for treatment.

There is need for proper understanding of median clefts of upper lip and detail review of surgical methods for treating such rare clefts

further in depth. The median clefts are not widely described in literature but has been taken note every now and then owing to such low incidence of the cleft.

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