Repair of the Median Microform Cleft Lip Using Z-Plasty

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ABSTRACT

A median cleft lip is a rare central midline deficiency of the upper lip. Multiple surgical techniques are described in the literature to address this defect, though there is little consensus on the preferred surgical technique. We describe an intra-oral approach for repair of the median upper lip cleft using mucosal Z-plasty. This technique provides excellent access to the attenuated orbicularis oris muscle and the frenulum fibrosed to the labial margin. The tethered lip can be mobilized and the notch converted with appropriate mucosal length, lip height, and vermilion fullness. The contour of the free labial border immediately improves, all while avoiding a cutaneous scar. The midline cleft lip notch can be effectively treated by adhering to 3 major principles: 1) excision of the tight, constrictive labial band; 2) achieving midline orbicularis oris muscle approximation; and 3) establishing mucosal lengthening using a Z-plasty.

Keywords: Median Cleft Lip; Midline Cleft Lip; Mucosal Z-Plasty

1. Background

A median or midline cleft lip is a vertical defect in the labial tissue. This physical anomaly is a rare variant that represents less than 1% of all cleft patients [1]. Phenotypically, median clefts can range from a small central vermilion notch to a wide midline cleft in combination with a bifid nose, bony defect, and hypertelorism. The median cleft lip defect has been found to occur both sporadically and as a part of a larger constellation of inherited anomalies.

Midline clefts can be grouped into two major categories: 1) false clefts, or those thought to be embryologically-related to tissue agenesis; and 2) true clefts, or those that result when fusion of the medial nasal processes fails. False clefts are further characterized by forebrain abnormalities and are thought to be a subtype of holoprosencephaly [2,3]. Flattening, or clefting, of the midline structures and widening of the lateral facial elements may also be appreciated in patients with false midline clefts. In the most severe cases, false clefts are not compatible with life, obviating the need for surgical correction.

Multiple terms have been used to describe the constellations of abnormalities observed with true median clefts, including median cleft face syndrome [4], frontonasal dysplasia [5], and Tessier #0 clefts [6]. True median clefts can be distinguished by a lack of forebrain abnormalities. However, hypertelorbitism, midline craniofacial osseous defects, hairline abnormalities, and a midline upper lip cleft may be present to varying degrees in these cases. The mildest form includes a small notch in the soft tissue of the upper lip that does not cross the vermilion border. The majority of these cases are sporadic, but familial aggregation has been documented [7, 8].

Multiple techniques have been described to repair the median cleft lip deformity. We describe a mucosal Z-plasty and midline orbicularis oris muscle unification to repair a median cleft lip notch.

2. Methods

2.1. Clinical Presentation

A 13-month-old male presented to the Yale Craniofacial Center with a midline cleft lip. The defect appeared as a notch in the upper lip wet and dry mucosa crossing the vermilion border (Figure 1(A)). Intraorally, the maxillary labial frenulum was comprised of several bands which were attached abnormally close to the free vermilion border. The maxillary alveolus was notched at the midline, but no osseous cleft was present (Figure 1(B)). Other pertinent facial findings included a shortened nasal length, maxillary hypoplasia, and delayed eruption of the maxillary anterior teeth. The anterior nasal spine was palpable and present.

2.2. Surgical Procedure and Outcome

A Z-plasty was designed on either side of the elongated frenulum and the constriction band was excised (Figure 1(C)). Raising the mucosal flaps exposed the orbicularis
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4. Conclusions

The midline cleft lip notch can be effectively treated by adhering to 3 major principles:

1) Excision of the tight, constrictive labial band;
2) Achieving midline orbicularis oris muscle approximation;
3) Establishing mucosal lengthening using a Z-plasty.

The technique described successfully releases the tethered lip, provides mucosal length and vermillion fullness, and avoids a cutaneous scar.

REFERENCES

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