Abstract:
A median microform 0 cleft lip is a rare craniofacial anomaly representing less than 1 percent of all clefts. These can be true clefts or false clefts. We present this case of true median microform cleft with polydactyly of both upper and lower limbs as part of orofacial-digital syndrome. We have approached in the midline through vermillion and repaired the orbicularis oris muscle and the defect. The contour of the lip was improved. Lip height, vermillion fullness and mobility of the lip were achieved.

Keyword: Tessier microform 0 cleft with orofacial-digital syndrome, Median cleft, advancement flap

INTRODUCTION:
Craniofacial clefts are rare. These are most disfiguring of all facial anomalies. They exist from small faint cutaneous microform manifestations to more complex soft tissue and skeletal manifestations. Exact incidence of these clefts are not known, but ranges from 1.4 to 4.9 per 100,000 live birth [3]. Most of them occur along predictable embryologic lines. Causes of these clefts remain obscure but the theory of failure of fusion and failure of mesodermal penetration are applicable to craniofacial clefts [1]. These can occur sporadically or as part of inherited anomalies. Studies have demonstrated the influence of certain risk factors such as radiation, infection, maternal metabolic imbalance, drugs and chemicals. As per Tessier classification no 0 cleft is a median craniofacial dysraphia[3]. Developmental origin of Tessier no 0 cleft is not clearly understood. However midline clefts usually occur during the third week of gestation. During this time bilateral frontonasal processes appear on the primitive forebrain. Fusion of the inferior medial nasal prominence, lateral prominence, medial aspect of
maxillary process must take place for normal upper lip development [8]. No 0 cleft results from failure of the two medial nasal processes to fuse in midline. Midline clefts can be categorized in to 1) true clefts 2) false clefts. False cleft results because of tissue agenesis. True cleft occurs when fusion of two medial nasal processes fails [4]. True median cleft was first described by Bechard in 1823. These are characterized by a notch in the vermillion border, duplication of frenulum, and diastema between central incisors. True median cleft may occur as an isolated entity or as a part of syndrome such as orofacial-digital syndrome [3]. In our case patient presented with 0 cleft and polydactyly in both upper and lower limbs.

2. CASE REPORT
A 14 year adolescent boy came with complaints of midline notch in the upper lip since birth (fig 1). The notch was present in the wet and dry mucosa not crossing the white roll vermillion border. There was a thick short frenulum tethering the lip and restricting the lip movements. There was a central incisor diastema. There was no osseous cleft, no nasal asymmetry, no duplication of nasal spine, no angulation of teeth, no maxillary hypoplasia, and no delayed eruption of maxillary teeth. Patient presented with polydactyly in both hands and both feet. In both feet the sixth toe was present on the lateral border. In right hand ring finger was shorter compared to left side. There were two extra digits present on the ulnar side in the right hand. The sixth finger was longer than little finger. (fig2). In the left hand sixth finger was present on the ulnar side. Clinically there was no abnormality in the overall development relevant to the age in any other system. Baseline investigations were normal. X-ray of the face did not show any osseous abnormalities.

3. SURGICAL PROCEDURE
Patient was assessed and under oral ETT GA elliptical incision was made in the midline extending from the notch up to the frenulum and midline fibrous tissue was excised (fig3). The frenulum was cut and the lip was released. Mucosal flap and vermillion flap was raised from midline. Orbicularis oris was exposed which was discontinuous in the midline cleft region. Orbicularis oris was dissected from vermilion and mucosa both sides and sutured in the midline by horizontal mattress with 4.0 vicryl. (fig4). Dry vermilion mucosa was sutured with 6.0 prolene. Wet mucosa closed with 6.0 vicryl. The notch was corrected, lip height and fullness was achieved, contour of the free lip border was improved. (fig5). Patient on further follow up showed
DISCUSSION:
The goal of repair for median cleft is proper reconstruction of cupid bow, philtrum, vermillion and buccal mucosa [3]. Various methods have been described to repair the median cleft lip defect. We have done excision of scar tissue, release of the tight frenulum, midline vermilion muscle repair and bilateral vermilion mucosal advancement to correct the miline cleft notch without tension. Midline notch can be corrected by strictly following certain principles like excision of tight constricting band, midline orbicularis oris muscle approximation, and mucosal, vermilion lengthening for good aesthetic midline closure.
In the literature review Urata & Kawamoto described V-Y flap based in the maxillary vestibule and apex at the free lip border [2]. Weimer described the excision of diamond shaped piece of skin mucosa and fibrotic band and zig-zag closure of mucosa [7]. Samuwel O. Buonocore have done repair with Z plasty technique [4]. Francesconi described local triangular flap fashioned from laterally to fit in to triangular defect medially to reconstruct the central defect [5]. Pinto and Goleria have described modified Z plasty using two triangular flaps to fit in to two triangular defects for repair [6]. We have done correction of notch through midline approach raising flaps in the vermillion without interrupting the white roll, philtrum, and cupid bow. We were able to close the muscle, and mucosa without any tension. Post operatively the result was acceptable functionally and cosmetically. Lip movement was full and there was no unsightly scar.

CONCLUSION:
We have presented this case for its rarity and for the different –“Vermillion approach” we have used. Median cleft can be effectively corrected by releasing the constricting band, approximating the muscle in the midline and mobilizing the dry and wet mucosa to get good tensionless closure. By our technique lip was released and the notch was corrected giving a good functional and cosmetic results.

REFERENCES:
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