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Case Report

# "Partial duplication of lower lip and hemimandible" A rare case

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## ABSTRACT

Duplication of mandible and lower lip is a very rare congenital entity. We report an extremely uncommon case of Congenital Duplication of Lower lip and Mandible in a 3 year old girl, who was treated surgically in a single stage for correction of both lip and mandible. This was a commissure preserving single staged procedure. The Surgical procedure, the problems related to this anomaly and the embryology are discussed.

## KEY WORDS

Duplication; lower lip; mandible

## INTRODUCTION

Even though duplication of various facial elements are described in literature only two cases of mandible with lower lip duplication have been described so far. Duplication of the mandible is an extremely rare birth defect, encompassed within the spectrum of duplication of facial elements. Duplication of mandible with lower lip is even rarer. McLaughlin was the first to describe it in 1948 and following that several other authors have attempted to illustrate this type of rare craniofacial anomaly.[1-6] Wu *et al*. reviewed the literature and identified the spectrum of complete or incomplete craniofacial duplication.[7] It was believed that this condition was a congenital anomaly and not a teratoma.[4] We report a case of a child with partial duplication of mandible and lower lip and discuss the surgical management in a single stage.

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## CASE REPORT

A 3 year old girl was referred to our department with complaints of double lower lip with drooling of saliva from angle of mouth [Figure 1]. There was no history of consanguity and no significant antenatal history. There was no history of other similar anomalies in the family. The child was born full term by normal vaginal delivery with no other detectable associated anomalies. Though the child was able to feed normally and achieved normal developmental milestones, there was constant drooling of saliva from the mouth. The child came for this drooling of saliva and correction of the deformity. The child underwent all routine investigations which were within normal limits. Lateral and Antero Posterior X- ray views of face showed duplication of right half of mandible with an extra row of tooth buds [Figures 2 and 3].

The child was planned for surgery with the goal to correct both the anomalies in a single stage. Patient was operated under general anaesthesia with nasal intubation. Plan was to preserve the right commissure with vermillion and excise the remaining tissues with the duplicated vermillion. Elliptical incision was given encircling the duplicated

odontogenic material extending to the left of chin along the relaxed skin tension line [Figure 4]. The incision was deepened to the anterior surface of mandible. The excess odontogenic tissues along with the inferior row of extra dentition was removed [The Hypoplastic vermillion was augmented by vermillion sharing and the lower lip was thus reconstructed [Figure 5]. The patient was discharged after seven days [Figure 6].

## DISCUSSION

Complete or partial facial duplication is an extremely rare malformation. Most cases appear to share a number of features, including cleft palate, duplication of the tongue, orbital hypertelorism and macrostomia.[8-13] Facial duplication may occur symmetrically or unilaterally, and it ranges from localized anomalies to complete doubling of all facial elements (diprosopus).[14] The variety of patterns of duplication suggests that the doubling of facial components arises in several different ways. These include forking of the notochord, duplication of the prosencephalon, duplication of the olfactory placodes, and duplication of maxillary and/or mandibular growth centres around the margins of the stomodeal plate.[9] Based on the structural abnormalities and presumed embryological mechanisms, facial duplication appears to represent both a clinically and pathogenetically heterogeneous malformation sequence in which overlapping findings are present.[13] Taruffi (1882) remarked on the preponderance of females among facial duplication with females outnumber males by 2:1.[15] McLaughlin first described a case of reduplication of mouth, tongue, mandible, stating that it is a rarity.[1] Fontaine reported a case in which there was a tooth containing bridge of bone extending from symphysis mentis to the medial aspect of right mandibular ramus.[2]

Only two cases of lower lip and mandibular duplication have been described in the literature.[3,6] This is in fact the third such extremely rare case. Since various spectrums of facial duplication have been described in literature this

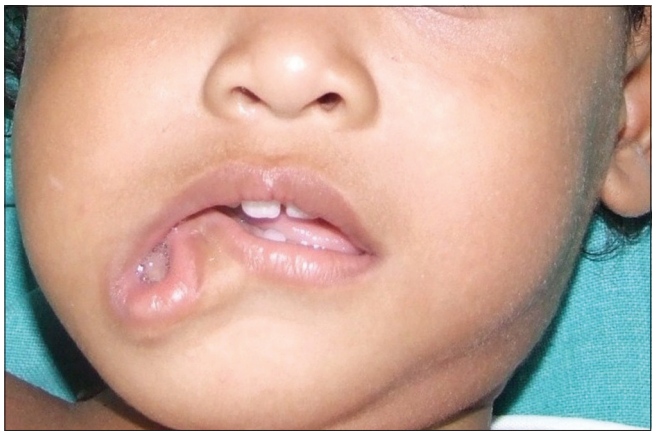

**Figure 1:** Frontal view showing duplication of right lower lip

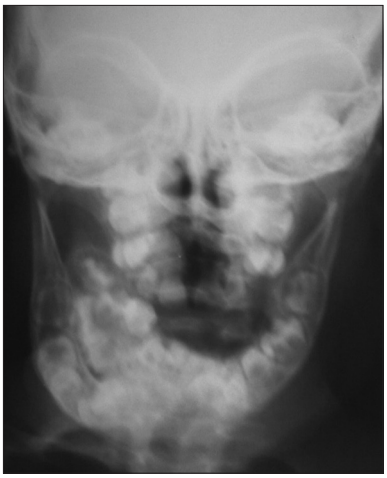

**Figure 2:** X-ray P-A view of mandible showing partial duplication of right half of mandible

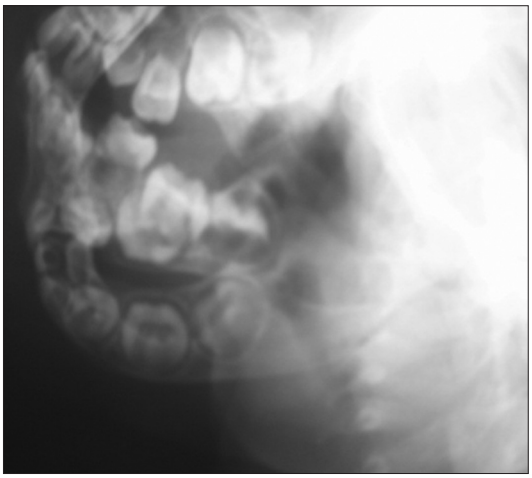

**Figure 3:** X-ray right lateral view of mandible showing duplication of right half of mandible with extra row of dentition

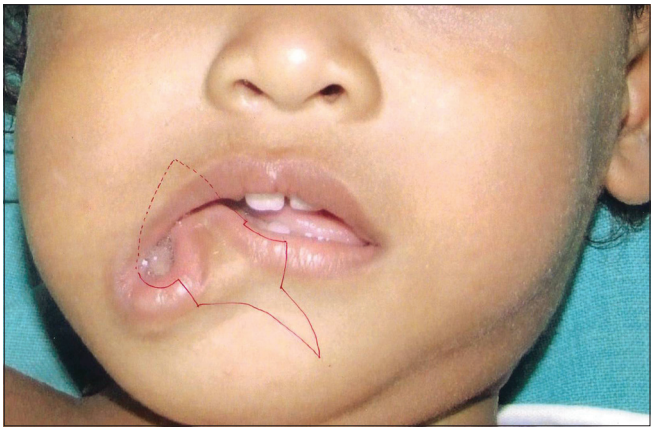

**Figure 4:** Planning of incision. Dotted lines indicative of intraoral incision

variety stands out for its rarity of occurrence. A duplicated mandible had been suggested to occur as a result of first branchial arch duplication[3] as a developmental anomaly arising from sequestrated totipotent cells,[4] as part of the split notochord syndrome[16] or secondary to duplication of the mandibular growth centre around the margins of stomatodeal plate.[9] Shaikh *et al*. reviewed the literature and showed that the duplication of the mid face was more commonly described than mandibular duplication.[5] Bhattacharya *et al*. reported a rare craniofacial cleft with duplication of alveolar arch.[17] Our case is similar to the case described by Suhaili *et al*. and Maisels *et al*.[3,6]

The human face develops from the frontonasal process, pair of maxillary processes and a pair of mandibular arches centered around the stomodeum [Figure 7a]. The differentiation of the facial processes appears in the early part of fifth week by migration of cells of the neural crest of the forebrain vesicle. The nose, the upper lip, the palate, cheek and upper jaw develop from the frontonasal and maxillary processes. The lower lip, mandible and integument covering it develop from the paired mandibular arches. The maxillary processes develop as triangular projections from the cephalic side of dorsal part of the mandibular arches. Neural crest cells arise along the entire edges of the neural folds, under the signaling of BMP proteins. In the hind brain the crest cells migrate in the eight segments of rhombomeres to populate specific pharyngeal arches which form the skeletal components characteristic of each arch. Endoderm is responsible for patterning of skeletal derivatives and this process represents epithelial-mesenchymal interaction. The crest cells contain HOX genes and their different transcription factors emanated from pouch endoderm are expressed in specific patterns.[18] The duplication in our case is likely to be due to the duplication of right first branchial arch or an accessory branchial arch after the development of the maxillary process [Figure 7b]. This assumption is based on the fact that the right maxilla and its embryological derivatives are absolutely normal. The elements of duplication are so well differentiated that it rules out the teratoma theory.

The facial disfigurement and drooling of saliva are detrimental to the social development and surgery is the only option to achieve better cosmetic and functional result. We conserved the right oral commissure and excised the extra vermillion and lower lip along with the extra inferior row of dentition and odontogenic tissues and skin in a single stage unlike Suhaili[6] who managed a similar deformity in two stages.

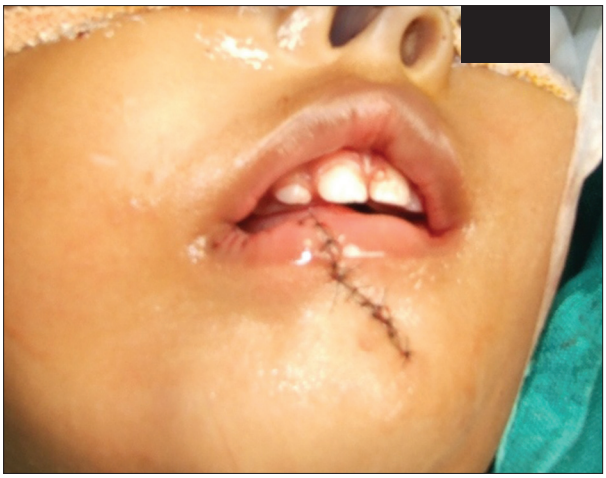

**Figure 5:** Immediate post op photograph

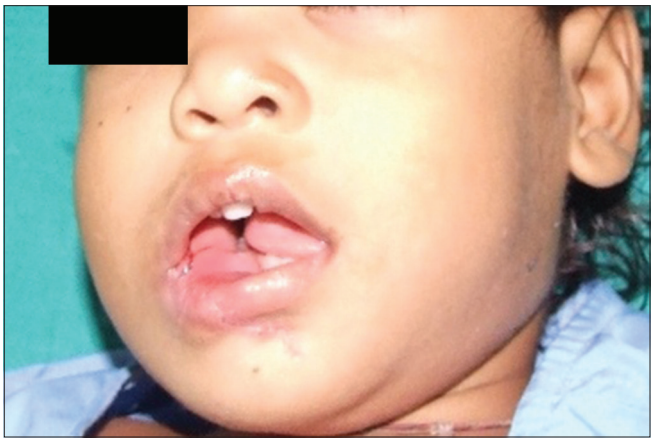

**Figure 6:** One week post surgery after stitch removal

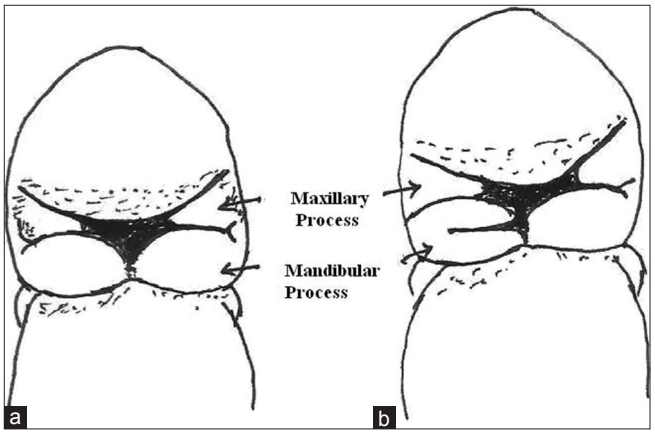


**Figure 7:** (a) Hypothetical configuration of the embryological structures around the stomodeum (b) Hypothetical duplication of the right mandibular process

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