

Case Report: Duplication of Oral Structures with Cleft Palate

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Description

A female child with multiple congenital anomalies of oro-facial and cardiac structures was seen first at five and one half years of age in The University of Michigan Department of Dentistry and Oral Surgery, September 1967.

Records from Childrens Hospital, Detroit, Michigan, revealed that at full term birth, she was found to have a mass 3 cm in diameter protruding from the mouth from a stalk originating in the posterior area of the soft palate to the right of the midline. The child had a good cry but was in moderate respiratory distress and slightly cyanotic. Skull radiographs shortly after birth were negative. Chest film indicated cardiomegaly with dilation of pulmonary artery branches and other findings suggesting a left-to-right shunt. The baby had problems handling secretions and, after a trial in an oxygen croupette for 24 hours, surgery was scheduled. Under endotracheal anesthesia, the mass, measuring 5.5 x 2.3 cm, was excised by means of a snare from the area of an unusual cleft palate. It was later diagnosed as mucocutaneous tissue of lip and palate. The immediate postoperative course was uneventful but respiratory distress continued. After 24 hours, at two days of age, a tracheotomy was performed. Intravenous fluids were required for eleven days. Nasogastric tube passage caused bleeding and an oral feeding tube was not tolerated. Oral feedings were then accomplished by a dropper bulb and later with a cleft palate nipple.

The tracheotomy tube was removed on the 39th day and she was discharged at 43 days of age with the confirmed diagnosis of severe congenital heart disease after withstanding anesthesia and the removal of a teratoma from the mouth involved with the cleft palate condition.

Early development was slow and mental retardation was diagnosed at three years of age. She was able only to sit and was unable to walk at five and one half years of age. There was little effort at speech and no language development. She was unable to feed herself and was not toilet trained.

The patient's congenital heart disease was the basis for referral to

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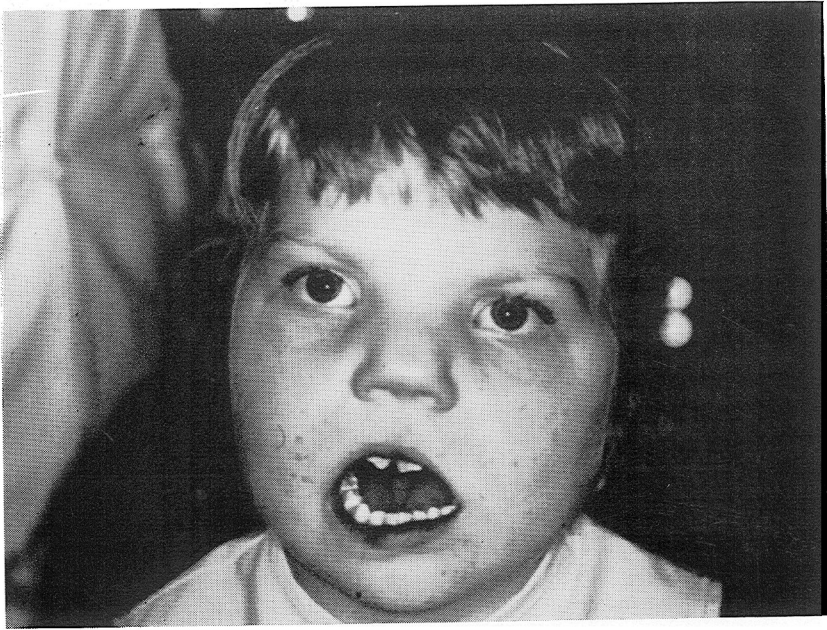


FIGURE 1. Patient at five and one half years of age exhibiting a broad flat nose and widely spaced eyes. She exhibits a short broad neck.

The University of Michigan Pediatric Department Cardiology Unit in August 1967. A severe tetralogy of Fallot or pseudo-truncus arteriosus was diagnosed. In addition, she was having increased feeding problems from abnormal dentoalveolar development and an enlarged tongue. The face exhibited a broad flat nose and the eyes were widely spaced. Cyanosis of the mucous membranes was noted (Figure 1). She had a short broad neck suggesting Klippel-Feil syndrome.

The maxillary region revealed a pronounced intraoral ridge extending anteroposteriorly along the midline. This ridge contained teeth and could thus be termed an alveolar ridge. It appeared to separate the roof of the mouth into two palates positioned side by side, each with rugae anteriorly and a midline cleft in the more posterior aspects. Both clefts were totally open into the nasal chambers, the right being wider and longer, extending from the anterior aspects of the first primary molar posteriorly (Figures 2 and 4).

The right alveolar ridge of the right palate contained the normal number of two incisors, a cuspid and two molars. Several teeth observed in the midline alveolar ridge were oriented toward this palate. The left alveolar ridge had two incisors, a cuspid and two molars. In the medial alveolar ridge, a central incisor, a cuspid, and two molars were oriented toward one palate. This was clearly demonstrated in a cast of the maxillae (Figure 4).

The tongue was very wide and had an elongated mass in its center

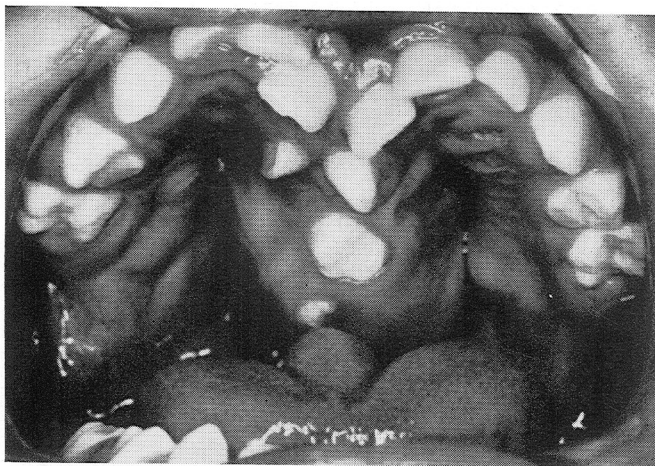


FIGURE 2. Intraoral view illustrating the two palates both of which are cleft. The medial alveolar ridge contains two accessory central incisors, cuspids and molars. Additional permanent teeth were embedded to this ridge.



FIGURE 3. Intraoral view of the dorsum of the tongue exhibiting the medial mass of tissue extending from the region just anterior to the base to near the tip of the tongue. Endotracheal tube on the left.

extending posteriorly from the base to near the tip. This mass in the medial dorsum consisted of tongue tissue and was freely movable although it was found firmly attached at its base to the underlying tongue (Figures 2 and 3).

The lower teeth were noted to be normal in number and position. It was of interest that the peripheral maxillary and mandibular teeth occluded to a functional degree (Figure 4).

Laboratory findings showed elevated hemoglobin to 23 grams per 100

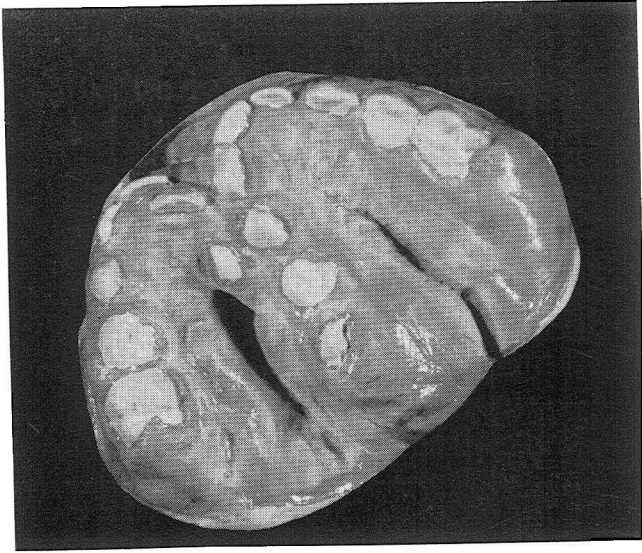


FIGURE 4. Cast of the palate illustrating the appearance of the cleft palates and the accessory medial located alveolar ridge and the incisor, cuspid and molar teeth.

ml; other blood studies were normal. Skull radiographs showed hypertelorism, foreshortening of the maxilla, and anterior cranial fossa. Films of the cervical spine showed incomplete segmentation of the second, third, and fourth vertebrae. Chest films revealed the cardiac abnormalities previously noted.

Pediatric cardiology consultation expressed the view that oral surgery to remove the central dentoalveolar structures could be tolerated. With appropriate antibiotic coverage, the dental extractions, total alveolectomy and reconstruction of the palatal vault was accomplished under endotracheal anesthesia on September 3, 1967 (by JRH). The midline bony mass was representative of alveolar process containing two primary cuspids, two primary molars, and crowns of premolars (Figures 5 and 6). The child tolerated the procedure well. She was discharged on September 14, 1967. The tolerance of this anesthetic and procedure prompted additional studies for correction of the cardiac deformity.

Three months later, after catheterization and cardiac diagnostic studies, the septal defect was corrected with reduction in the degree of cyanosis. Improved general status followed. When seen in September 1968, she was able to walk, was completely toilet trained, was able to feed herself, asked and answered simple questions, and was considered in an educatable range of limited mental development.

Discussion

References describing facial duplication are few and no two cases are identical in regard to duplication of these structures. In 1907, Schwalbe



FIGURE 5. The medial alveolar ridge after removal. The mass measured about two centimeters in length.

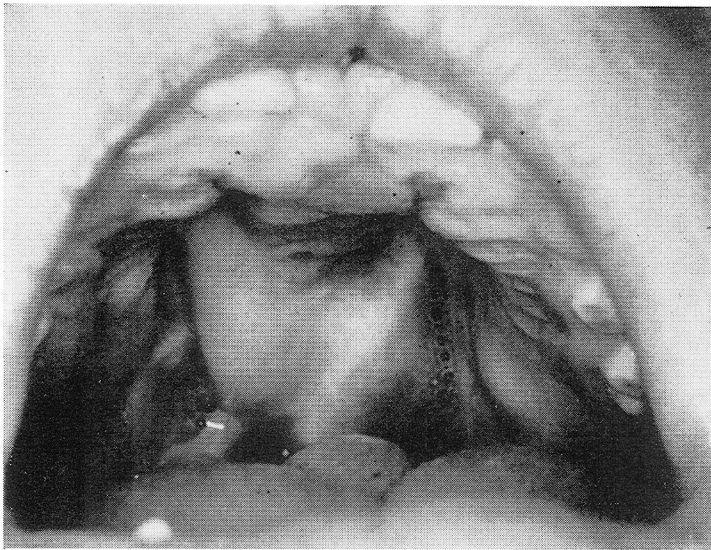


FIGURE 6. Healing of palate after removal of central alveolar process.

(11) described a case with a globular mass which was a partial duplication of the face.

McLaughlin (8) in 1944, reported a case of duplication in which a small funnel-shaped mouth protruded from the lower right portion of the

mandibular area facing laterally rather than anteriorly. This and the normal mouth demonstrated a coordinated orbicularis oris sphincter mechanism. Both had lips and the accessory mouth contained a small tongue which was found to be an extension of the normal tongue. A rudimentary mandible with eight accessory teeth appeared fused with the normal mandible near the symphysis.

A case of diprosopus was found by Latteier and Anderson (7), who described this monster as having a large head and two faces fused together with four eyes, two noses, and two mouths, each having a cleft lip and palate. Both of the oral stomata opened into a common pharynx where the two tongues joined at a common base.

Beatty (2), in 1956, described a case of an accessory mouth containing a tongue and a mandible. In this case, there was an anophthalmia and a polycystic kidney. It is of interest that the tongue in the accessory mouth was not attached to the true tongue. The accessory mandible contained a few teeth.

In 1957, Morton (10) described a case in which a very large wide mouth appeared with a double lower lip, two frenums, two supernumerary central incisors, and two mandibles, each with a tongue, which were joined together posteriorly. In the roof of the mouth was a bilateral cleft palate. This child died three days after birth and, upon autopsy, was found to have two pituitary glands, each composed of neural and adenohypophysis. The brain was not otherwise abnormal except that the olfactory tracts were absent.

In 1964, Bacsich, Dennison, and MacDonald (1) reported a case of a full term infant who died 11 days after birth from bronchial pneumonia. The child had two mouths, side by side, each of them surrounded by lips and showing a well formed alveolar process and unerupted teeth. The larger of the two mouths had a cleft palate and in the floor of the mouth was a normal tongue. The smaller mouth was a blind pouch with no tongue and only a rudimentary mandible. Again, in this case there was the presence of a double pituitary gland situated in a broad sella turcica. The post-mortem examination revealed the brain to be otherwise normal, although upon radiographic examination of the vertebral column, there appeared a partial duplication of the cervical vertebra.

Goulian and Conway (5), in 1964, reported a case of a child with multiple congenital abnormalities including a moderate hydrocephalus and a wide separation of the fontanelles, hypertelorism, a broad wide nasal dorsum giving rise to three nostrils and three nasal chambers. The central chamber was widely cleft with a cleft in the upper lip to the left of the midline. In addition, there was a marked macrostomia in which two oral stomata appeared separated by a large soft tissue mass which connected the floor and the roof of the mouth. This extended from the anterior part of the mouth to the posterior limits of the hard palate. This mass thus represented an incomplete separation of the oral cavity into two parts. On each side of this mass there appeared two pairs of fused

alveolar ridges, two palates, and two tongues. The posterior one-third of the two tongues were fused together into a common structure in the pharynx.

It is of interest that no two cases of facial and palatal duplication are closely similar. Of the nine cases described, only that reported by McLaughlin (8), Smylski (12), Beatty (2), and the present described case have lived. Fortunately for these, surgical repair was possible.

Facial duplication has also been reported in animals. Jenkins and Hardy (6) described a calf in which two faces, two mouths, two tongues, and two palates appeared. Thus, the entire anterior part of the face and the associated oral structures were paired. He noted two tongues, of which the roots did not appear to be fused posteriorly although both oral cavities led jointly into a common medial pharynx. In this case, both of the palates were cleft similar to the human case, which we have described. The two oral cavities joined in the region of the pharynx. Jenkins noted that 25 teeth appeared in the two oral cavities, whereas the normal number of deciduous teeth in the ox is 20. These teeth represented 11 incisors, four in the right oral cavity and six in the left, with only one located in the median part of the bilateral fused mandibles. Both the right and left oral cavities had seven premolars each. The cleft in the left palate was wider than the one in the right and extended from the second primary molar posteriorly, whereas the cleft of the right palate appeared only in the posterior third of the palate.

The possible embryologic series of events that led to the defects described in the present case can be postulated. Although no cases of developmental malformations were apparent in two older siblings or other members of the immediate family or grandparents, more subtle genetic factors may be involved.

As was previously pointed out, duplication of structures on a limited area of the body are even more rare than more completely formed conjoined twins. From a developmental viewpoint, the brain, heart, and facial area are organizing and differentiating early in the first trimester at a similar period of time. During this period, the forebrain and heart are growing rapidly and differentiating as are the structures arising from the first branchial arch. Therefore, any factors disturbing the normal development of the heart, brain, and orofacial structures would most probably have had their effect prior to the fourth prenatal week.

Morton (10) has an interesting theory in regard to duplication of the tissues of this developmental period. He theorized that the duplication of the cells of the interior end of the notochord process initiate duplication of the connective tissue and overlying epithelial structures resulting in duplication of the facial-oral structures. More recently, Bacsich and associates (1), 1964, noted duplication of the pituitary in the case of oral duplication they described. Bacsich agrees with Morton's theory that facial duplication is part of a split notochordal complex.

It is possible that in these cases of facial duplication, the pituitary duplication is another characteristic of this syndrome. It is pertinent to note that in several other cases, if autopsies had been performed, evidence of pituitary gland pairing might have been seen. Radiographic evidence of wide cranial base bony outlines were reported in several cases and may also be true in the present case. It would have been of interest to know more about the mass that was removed at birth. It was said to have protruded through the cleft in the palate. There are some 100 cases of Rathke's pouch or nasopharyngeal tumors in newborns in the literature which, as in this case, are sufficiently large to cause asphyxia (4). In the present case, this tumor was probably tissue duplicating some part of the naso-oral area.

Partial facial duplication may be even more limited as in the case of nasal duplication noted by Erich (3), 1962. This condition seems to be even more rare where two distinct noses, each with two nostrils and nasal cavities, appear. In this case, it is possible that the mechanism causing duplication results in accessory nasal placodes differentiating from the epithelium of the frontal area overlying the forebrain. As the mesenchyme around the nasal placodes develop, the nasal pit is molded and then further development of nasal capsule and bony skeleton of these nasal cavities occurs. If on the other hand no nasal placodes arise, no nasal pits nor nasal capsule may possibly then develop.

The interventricular defect and the pulmonary stenosis would be less important prenatally than postnatally. At birth, when the lungs begin to function, the foramen ovale and ductus arteriosus bypass of the lungs normally closes. If this does not occur, then hypoxia could result and brain damage could occur as well, as the various compensatory changes take place. On the other hand, prenatal oxygen deficiency has been considered a possible cause of cleft palate. Embryos of experimental animals subjected to hypoxia may develop cleft palate if the episode occurs just prior to palatal shelf elevation (9).

The rotation, elevation and fusion of the palatal shelves is a complex embryologic process. Three masses of tissue, the medial and two lateral palatine processes, take part in this process to form the roof of the oral cavity. No simple explanation of malposition of the palatal anlage could give rise to paired shelves and associated dental lamina and alveolar ridges such as appear in the present case. The complexity of the duplication can best be seen in the midline of the palate where the teeth appeared (Figure 4). The dental lamina must have been paired since two rows of teeth are present. It is of interest that an anterior-posterior tooth differentiation occurred incisor to molar as did the orientation of the erupted and non-erupted teeth in this medial bony ridge to their respective palates. Again, evidence of a primary and secondary dentition appeared. It is probable that the bony alveolar ridge developed in response to the appearance of the dental lamina and the resulting tooth primordia

that appeared in that location. Why then does this ridge divide anteriorly and follow the arches of the maxilla?

It is possible that crowding could be a factor in the clefts or lack of closure of the palatal shelves. If the shelves were forced superiorly after their elevation to the horizontal plane due to lack of space for proper development, then it is possible that the shelves were unable to approximate and fuse. In this respect, it is of interest that the combined width of the two palates and maxillary arches was approximately the same as that of the single mandible, so that the outer row of the maxillary and the mandibular teeth occluded.

The most common defects of the tongue are an enlarged tuberculum impar and a bifid tongue. The present case looks like neither of these, but more like an attempt at tongue duplication. The developing tongue musculature begins its anterior growth surge from the region of the occipital myotomes during the fifth and sixth week. The medial mass seen in Figure 3 rises well above the surface of the tongue and seems to be fairly independent of it except for its attachment along the midline. This mass arises just anterior to the base and extends to near the tip of the tongue. It appears to be an attempt at formation of a second tongue or tongue duplication rather than enlargement of the tuberculum impar of the tongue. Many of the face duplication cases described tongues with two bodies and a single base. The present case shows no evidence of duplication of the base, which thus supports this concept.

Summary

A report is made of a patient who had two palates, both of which were cleft; a midline alveolar ridge with incisors, cuspids, and molars; and a partially duplicated tongue. A severe tetralogy of Fallot or pseudo-truncus arteriosus and mental retardation were also diagnosed. The case was described and some of the etiologic factors related to the development of such defects were discussed.

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