Median Cleft of the Lower Lip, Mandible, and Tongue with Midline Cervical Cord: A Case Report.

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Inferior gnathoschisis is an extremely rare anomaly. In Table I are listed all descriptions we have found in the world literature of clefts of the lower lip, with cleft mandible and cleft tongue. Just prior to the publication of this paper, five additional cases were reported in Japan, and these are included.

In this paper we shall present the case of a newborn with midline cleft of the lower lip, mandible, and tongue; this case first came to our attention in January, 1967.

Case Report

A 17-day-old girl, M. O., was admitted to the Oral-Surgery service of the Kyushu University Hospital (Japan) on January 15, 1967. Chief complaints on admission were deformity of the lower lip and difficulty in feeding.

FAMILY HISTORY. The mother was 21 years of age and the father 27; both were in good health. There was no history of consanguinous marriage nor any record of congenital deformity in the family. Pregnancy was uneventful, with no surgery, medication, or known exposure to X ray.

PRESENT ILLNESS. The patient was delivered at term without difficulty and after an uncomplicated labor. Birth weight was 3100 grams. Examination in the immediate neonatal period revealed normal contour of the upper face and head; the upper lip and nose were intact. The lower lip was split in the midline, and the tongue presented a deep anteroposterior groove in the midline. Ankyloglossia was noted, and the tip of the tongue on each side of the cleft was bound with the lip to the loose edge of the symphysis of the mandible (Figure 1). Extending from the chin down to the manubrium of the sternum was a firm band over which the skin was freely movable. This band prevented the over-

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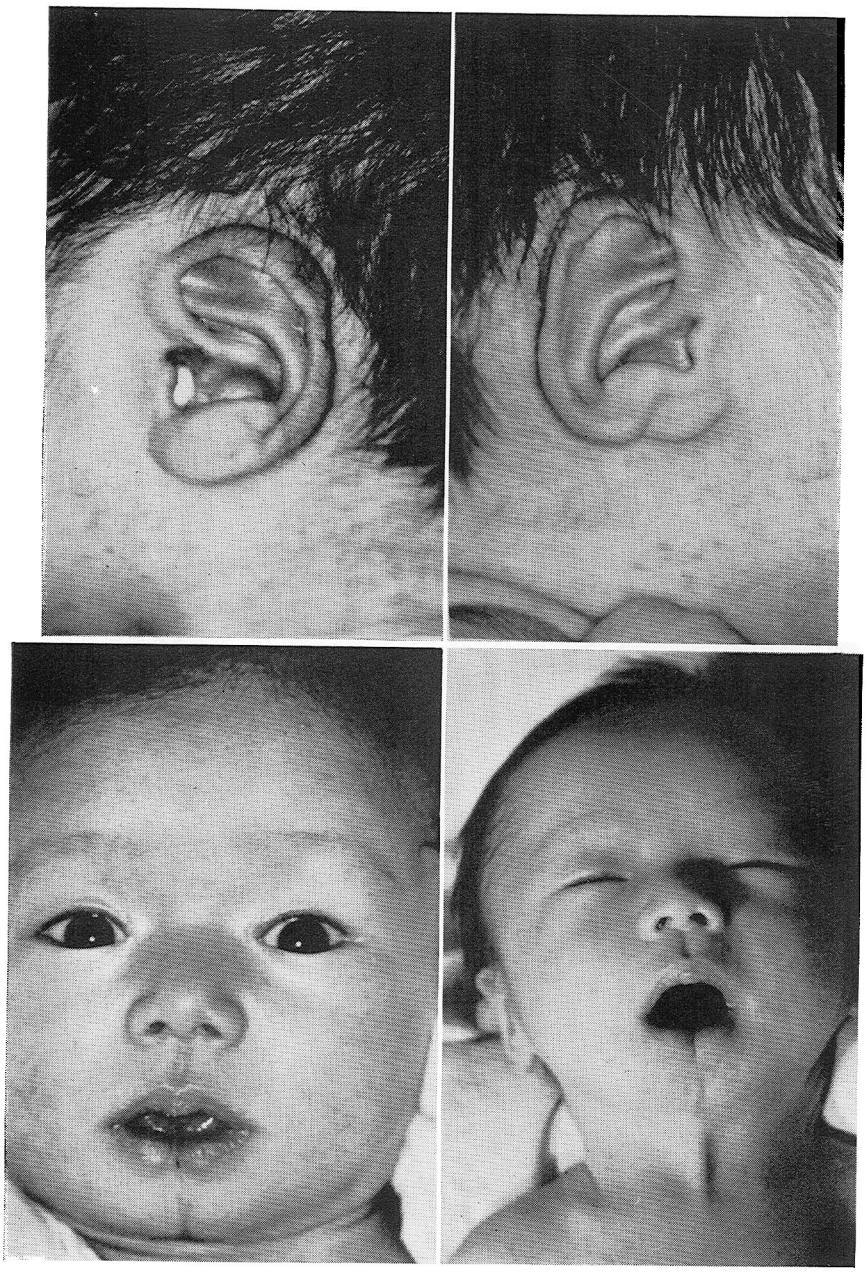

FIGURE 1. Photographs of M. O., illustrating the cleft of the lower lip, malformation of the right ear lobe and cervical cord.

extension of the head. There was also a slit at the right lobe of the ear. The remainder of the physical examination was entirely within normal limits.

X-RAY FINDINGS. Roentgenograms confirmed a mandibular separation of

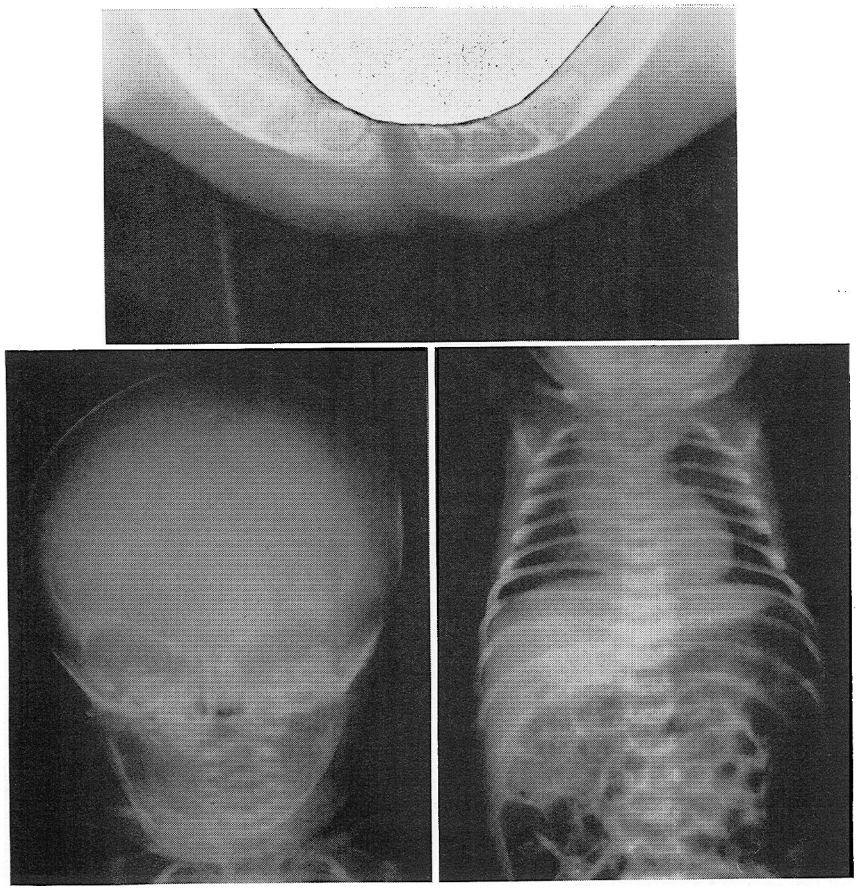

FIGURE 2. Roentgenograms of M. O., illustrating the missing deciduous lower right central incisor and separation of the mandible (upper), the skull (lower, left), and the chest (lower, right).

approximately 6 mm at the symphysis (Figure 2). The deciduous lower right central incisor was absent. No other abnormality was noted in views of the face, trunk, or extremities.

HOSPITAL COURSE. The infant was given careful nursing attention and the weight gradually increased. The patient had an otherwise normal neonatal course.

COMMENT AND SUMMARY. We have found 34 cases of median cleft of the lower lip reported in the world literature. It is of interest that only two of these cases were adults, and perhaps indicates a high coincidental occurrence of other congenital defects leading to early mortality. It is also possible that the deformity per se results in an increased incidence of pneumonia with consequent higher neonatal morbidity. Over-all, there is no significant preponderance of either sex, although the Japanese reports show a higher incidence among females.

TABLE 1. Summary table of reports of cleft lower lip, cleft mandible, and cleft tongue. Data author, year of report, age and sex of subject, presence or absence of the three types of clefts and of ankyloglossia, and listing of additional anomalies.

author and year	*subject*	*cleft type*				
		lower lip	*mandible*	*tongue*	*ankyloglossia*	*other*
Couronné (*5*) 1819		+				
Moeckel (*15*)		+				
Petit (*20*) 1826	adult f	+			+	
Boisson (*3*) 1841		+			+	
Parise (*19*) 1862	15 d	+	+		+	neck contract.
Faucon (*8*) 1874	1½ yr	+	+		+	neck contract., bulging of neck
Lannelongue (*13*) 1879	2½ yr m	+	+		+	
Hamilton (*9*) 1881	child	+				
Wölfler (*31*) 1890	21 d m	+	+	+	+	dermoid of nose, neck contract.
Redard, et al. 1891 cited by Monroe (*16*)	8 mo m	+	+		+	
Salzer (*23*) 1902	3 mo	+	+		+	
Debraisieux (*7*) 1904		+				
Keith (*12*) 1909	child ?	+	+			
Miyata (*14*) 1926	19 y m	+	+	+		polyp
Morton, et al. (*17*) 1935	13 d f	+	+	−	+	thyroid gl. absent, neck contract.
Stewart (*24*) 1935	3 d f	+	+		+	congenital heart lesion neck contract.
Wassmund (*28*) 1935	44 y m	+	+		+	
Ashley, et al. (*2*) 1943	stillborn f	+	+		+	cleft upper lip & palate, anencephaly, clubfoot
Braithwaite, et al. (*4*) 1949	4 y f	+		+	+	micrognathia, microtia, congenital cystic eye
Davis (*6*) 1950	4 y	+	+		+	hyoid & manubrium absent, congenital heart lesion, neck contract.
Abramson (*1*) 1952	newborn f	+	+		+	growth from palate, uvula bifida
Haym (*10*) 1952	5–6 y ?	+		+	+	iris coloboma
Weyers (*30*) 1953	4 w f	+	+			polydactyly, oligodontia, cleft palate
Vigil Lorenzo (*27*) 1955	newborn	+	+	+	+	absence of skin in midline of neck
Kawai (*11*) 1955	13 y f	+	+		+	
Torres, et al. (*26*) 1956	2 y m	+			+	
	2 y m	+	+		+	
Recamiel, et al. (*21*) 1957	few days	+	+		+	tumor of tongue, neck contract.
	1 y	+	+		+	neck contract.
Russell, et al. (*22*) 1961	15 y m	+	+	−	+	
Oota, et al. (*18*) 1962	15 d f	+	+		+	accessory tongue
Watanabe, et al. (*29*) 1964	6 y f	+	+	−	+	congenital heart lesion
Tange (*25*) 1965	4 mo m	+	−	−	−	deformity of external ear
Monroe, et al. (*16*) 1966	6 h m	+	+	+	+	congenital heart lesion, tumor of lower lip
Fujino, et al. (present study) 1967	17 d f	+	+	+	+	deformity of external ear, oligodontia and neck contract.

Most of the previously reported cases showed only median cleft of the lower lip and mandible, with the remainder having various associated anomalies, including the cleft of the tongue, ankyloglossia, oligodontia, heart anomalies and malformation of the sternum and extremities (Table 1). With regard to treatment, most reports favor lip repair with tumor excision and freeing of the tongue in the neonatal period. In infancy, z-plasty of the neck contracture is performed and

in later stages of life mandibular wiring is carried out. Some fatalities were noted following bone grafting.

We are planning to free the tongue as soon as possible, then repair the lower lip in infancy, z-plasty in childhood and mandibular wiring or bone grafting in later stages of life.

Concerning etiology, the most reasonable theory at present is that there is an incomplete mesodermal penetration into the mandibular process. Considering the developmental stage of the organs involved, it would appear that the disturbing influence occurs at the end of the fifth or at the beginning of the sixth week of gestation.

Summary

A case report is presented of median cleft of the lower lip, mandible, and tongue, along with a bibliography and summary table of report of arch defects from the world literature.

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