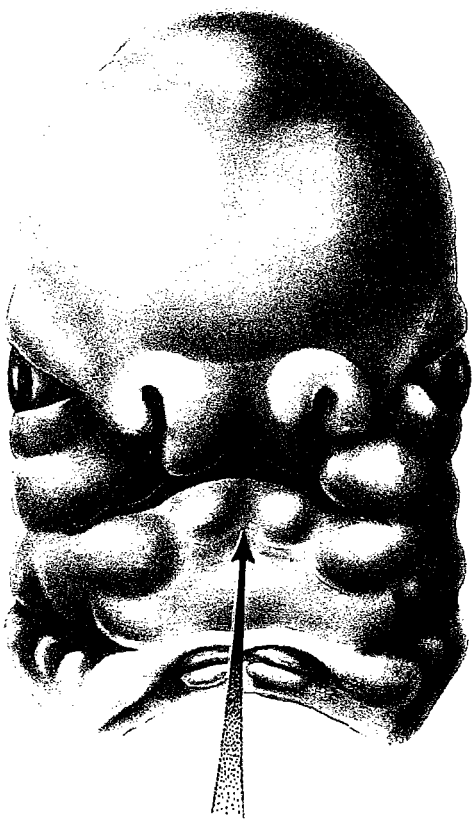
61. *Median Clefts of the Lower Lip and Mandible*



MIDLINE clefts of the lower lip and mandible are considered the result of failure of mesoderm migration or merging of the paired mandibular processes.

Midline clefts of the lower lip are exceedingly rare and can vary from a vermilion notch to a cleft involving the total lower lip, tongue and mandible extending to the root of the neck.

Couronné in 1819 was the first to mention this anomaly. Bouisson in 1840 mentioned some three or four earlier cases and recorded one that he had seen post mortem himself. According to Rose:

> The cleft extends in different cases to a variable extent. Thus Nicati, Couronné, F. Petit and Ammon saw clefts implicating the lower lip. Rikell operated [in 1870] on a cleft extending to the chin, through which the saliva was continuously dribbling. Faucon (1868) and Lannelongue (1879) recorded clefts of the lip and mandible conjoined, and in both cystic swellings (presumably of the dermoid type) were found between the segments. Parise's (1862) and Wölfler's [1890] cases were also associated with cleft of the tongue, through its whole thickness in the former, and only at its tip in the latter.

Wölfler's case is shown as a sketch. It is interesting that Parise's 14-day-old case had a median complete lower lip cleft with the free edges rounded as in "harelip" extending as a cicatricial band in the midline of the neck to the suprasternal notch. The median cleft of the mandible was separated several millimeters but bridged by connective tissue. The tongue was entirely divided, the cleft extending back to the glossoepiglotic ligament and downward between the geniohyoglossus muscles.

Sir Arthur Keith of the Royal College of Surgeons, London, noted:

Sir Arthur Keith

> Among the 250 specimens of malformations examined, only 4 showed this condition; a full-term child in the museum at St. George's Hospital and 3 specimens in the museum of this college; one from an ass, another from a cockatoo and a third from a sparrow.

In 1926 Brophy published this cast of a four-month-old girl with a midline complete cleft of the lower lip and mandible extending into the neck which had been presented to him by Keith from the Museum of the Royal College of Surgeons. He also reported an incomplete median cleft of the lower lip in an East Indian treated with a V excision and closure. In addition, he published an eagle with a cleft of the lower beak. When added to a cleft sparrow and cockatoo, the data does suggest that this anomaly is "for the birds"!

Author and Year	Subject	Cleft Type				
		Lower Lip	Mandible	Tongue	Ankylo-glossia	Other
Couronné, 1819		+				
Moeckel		+				
Petit, 1826	Adult F.	+			+	
Bouisson, 1840		+			+	
Parise, 1862	15 d.	+	+		+	Neck contracture.
Faucon, 1874	1½ yr.	+	+		+	Neck contracture, bulging of neck
Lannelongue, 1879	2½ yr. M.	+	+		+	
Hamilton, 1881	Child	+				
Wölfler, 1890	21 d. M.	+	+	+	+	Dermoid of nose, neck contracture
Redard et al., 1891 (cited by Monroe)	8 mo. M.	+	+		+	
Salzer, 1902	3 mo.	+	+		+	
Debraisieux, 1904		+				
Keith, 1909	Child?	+	+			
Brophy, 1923	Baby F.	+				
Miyata, 1926	19 yr. M.	+	+	+		Polyp
Morton et al., 1935	13 d. F.	+	+	−	+	Thyroid gland absent, neck contracture
Stewart, 1935	3 d. F.	+	+	+	+	Congenital heart lesion, neck contracture
Wassmund, 1935	44 yr. M.	+	+		+	
Ashley et al., 1943	Stillborn F.	+	+		+	Cleft upper lip and palate, anencephaly, clubfoot
Braithwaite et al., 1949	4 yr. F.	+		+	+	Micrognathia, microtia, congenital cystic eye
Davis, 1950	4 yr.	+	+		+	Hyoid and manubrium absent, congenital heart lesion, neck contracture
Abramson, 1952	Newborn F.	+	+		+	Growth from palate, bifid uvula
Haym, 1952	5–6 yr.?	+		+	+	Iris coloboma
Weyers, 1953	4 w. F.	+	+			Polydactyly, oligodontia, cleft palate
Vigil-Lorenzo, 1955	Newborn	+	+	+	+	Absence of skin in midline of neck
Kawai, 1955	13 yr. F.	+	+		+	
Torres et al., 1956	2 yr. M.	+			+	
	2 yr. M.	+	+		+	
Recamier et al., 1957	Few days	+	+		+	Tumor of tongue, neck contracture
	1 yr.	+	+		+	Neck contracture
Russell et al., 1961	15 yr. M.	+	+	−	+	
Oota et al., 1965	15 d. F.	+	+		+	Accessory tongue
Nolens, 1964		+	+			
		+	+			
		+	+			
		+	+			
		+	+			
Watanabe et al., 1964	6 yr. F.	+	+	−	+	Congenital heart lesion
Tange, 1965	4 mo. M.	+	−	−	−	Deformity of external ear
Monroe et al., 1966	6 hr. M.	+	+	+	+	Congenital heart lesion, tumor of lower lip
Fujino et al. (present study), 1967	17 d. F.	+	+	+	+	Deformity of external ear, oligodontia, neck contracture
Chouard, 1967		+				
Rea, 1967		+				
Knowles et al., 1969		+	+			
Lauro and Verga, 1969		+				
Fitzgibbon (unpublished		+				
Millard et al., 1971	Newborn M.	+	+		+	

Reviews of the world literature by Monroe of the United States in 1966 and Fujino, Yasuko and Takeshi of Japan in 1970 list a total of 35 cases. This did not include Brophy's case in 1923, five others reported by Nolens in 1964, and one each by Chouard in 1967, Rea in 1967, Knowles, Littlewood and Bush in 1969 and Lauro and Verga in 1969. To this have been added other known human cases to a total of 47. It is certain that there have been many unreported cases, like the incomplete median cleft of the lower lip associated with mucous pits seen in Fitzgibbon's clinic in Bristol.

EMBRYOLOGICAL ASPECTS

Fundamentally, the anomaly seems to be the result of failure of mesodermal penetration into the midline structures of the mandibular portions of the first branchial arch. There is a broad variation in the severity of this failure, which ranges from minor clefts to complete clefts with loss of the supporting structures of the neck and sternum noted by Davis in 1950. Morton and Jordon in 1935 proposed that failure of the mandibular processes to fuse probably prevents the ventral ends of the succeeding arches from uniting inasmuch as fusion proceeds from above. Possibly this explains the absence of the hyoid bone, thyroid cartilage, strap muscles and manubrium in some of the more severe cases.

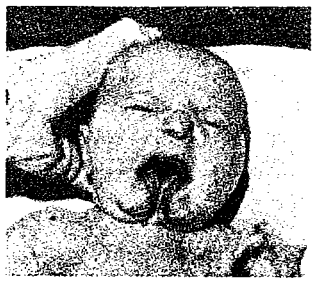
TREATMENT

Treatment of this anomaly has not varied greatly. Some of the early cases were museum specimens and some died before surgery could be accomplished. The cases of Braithwaite and Watson and Fujino, Yasuko and Takeshi were published prior to surgery. Later, Fujino with Yasuko and Katsuki reported their plan of treatment of the patient at age three years.

> Free tongue as soon as possible, then repair the lower lip in infancy, Z-plasty [neck] in childhood and mandibular wiring or bone grafting in later stages of life.

Most reports show a simple V excision of the lip with direct closure and the use of a Z-plasty if the cleft extended into the neck. In 1935 William Stewart in the *Archives of Surgery* reported this infant with a cleft extending through the lower lip, tongue and mandible. The two sides of the bifid tongue were attached to the floor of the mouth and vermilion. Stewart approximated the soft tissues and released the tongue without an attempt at closure of the cleft in the mandible in the initial procedure.

The charming Albert D. Davis of Stanford University School of Medicine, San Francisco, who published a forthright paper on median clefts of the lower lip and mandible in *Plastic and Reconstructive Surgery* in 1950, was probably the first young American student to study with Gillies in London. This was in the early 20's when it was common for plastic surgeons in Europe to demand remuneration for their teaching. Davis recalled to me with a twinkle how he would "slave" under Gillies as long as his money held out and then cross the Channel to work in a Paris hospital to make enough money to return to England for further months of study.



Albert Davis

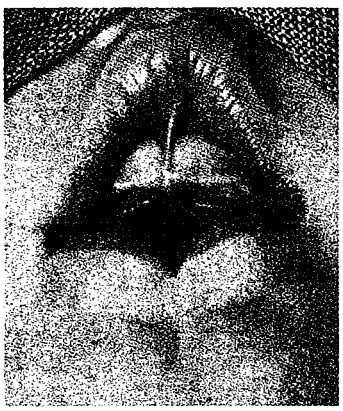
In 1948 a 4-year-old girl who had had a simple closure of a complete cleft of the lower lip at one month of age came under the care of Davis. At this time she had a scar extending from the midline of the lower lip downward into the anterior neck region with the chin held in such marked flexion that there seemed to be no chin. Crying caused the entire anterior neck region from sternum to chin to balloon as a distended pouch, which with inspiration collapsed in retraction. The chin was plastered to the suprasternal region by cord-like strands, and there was a 2 cm. gap in the midline of the mandible with a full complement of deciduous teeth present on each side. Davis described his 1949 operation:

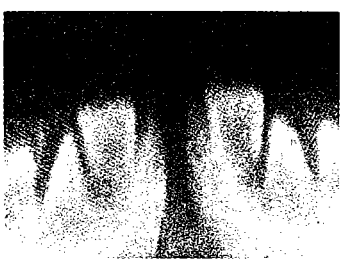
> A low collar incision was made above and between the clavicles. The skin overlying the cords was mobilized. These cords were seen to be dense scarred bands replacing the normal ribbon muscles in the anterior neck. No remnants of the hyoid bone or thyroid cartilages were felt. The anterior hypopharynx and laryngeal wall appeared to be intrinsic with these bands. The scar tissue was released and excised as much as possible. . . . The bone ends were cut back to healthy bone and the two edges of the mandible wired together.

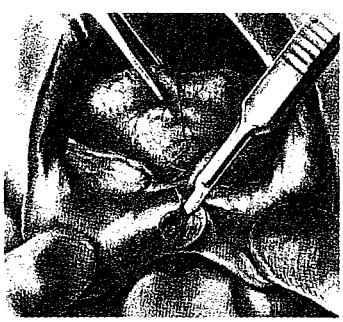
Davis further reported candidly:

> Following this procedure, the chin could be lifted higher without stretching and there seemed to be less contracture of the vertical cords. After several weeks, however, it became evident that contractures had again occurred, and that the edges of the mandibular cleft, while more nearly in approximation, were being pulled downward. . . . Several procedures will be necessary to obtain further correction.

When there is severe absence of tissue, particularly in the concavity of the neck as in such cases, flap tissue shifted locally when available or from a distance when necessary will probably offer the best final solution to the problem.





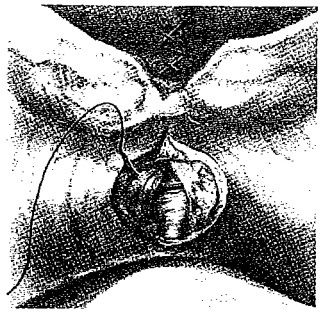


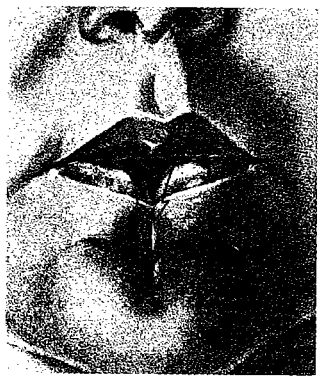
In 1971, in the *British Journal of Plastic Surgery* with J. A. Lehman, Jr., M. Deane and W. P. Garst, I presented the forty-sixth case to be reported in the literature. The patient was a newborn with an incomplete midline cleft of the lower lip and a bifid mandible. The mother had been diagnosed as having Stein-Leventhal syndrome, and a bilateral ovarian wedge resection had been performed several years prior to the conception. There was no familial history of congenital anomalies. The infant, seen 22 days after birth, revealed an incomplete cleft of the lower lip with a submucosal cleft of the orbicularis oris and a midline furrow to the chin. There was hypertrophy of the upper lip frenulum and a tight frenulum tethering the tongue to the groove in the alveolus. There was bifurcation of the mandible which was confirmed by x-ray film.

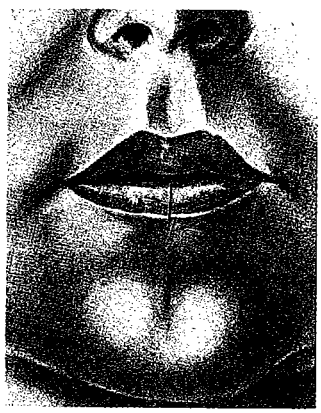
Release of the tongue and correction of the lip defect were performed at age five months. First the upper lip was released by a Z-plasty of the hypertrophied frenulum. The normal-sized tongue was freed by release of the lingual frenulum's attachment to the alveolar notch and closure of the defect on the ventral aspect of the tongue in a straight line except for a Z-plasty interruption at its inferior extremity.

The lower lip cleft, being a submucosal type, notched in the vermilion but with the skin only grooved by a depression without an actual fissure, called for surgery designed to avoid unnecessary scarring of the skin and even maintenance of that part of the congenital groove that lay in the normal chin dimple posi-

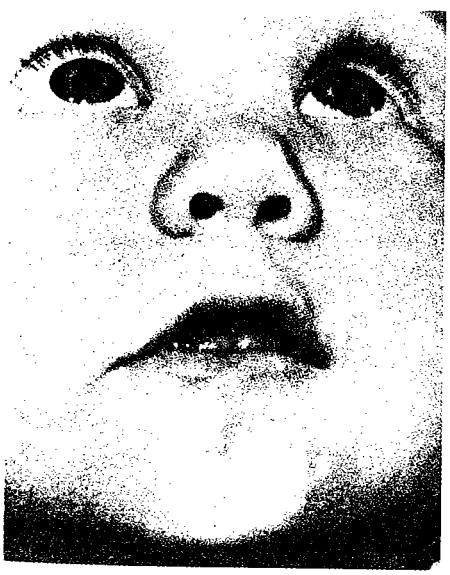
tion. Thus, a midline incision was made in the posterior mucosa and continued down vertically to the labial sulcus. The mucosa was undermined on each side of the defect and the orbicularis oris muscle dissected free. The fibrotic union of the muscle in the midline was excised vertically, except at the normal chin dimple position, and the muscles were sutured together across the cleft. Excess mucosa and vermilion were trimmed, and the posterior wound was closed with fine catgut.

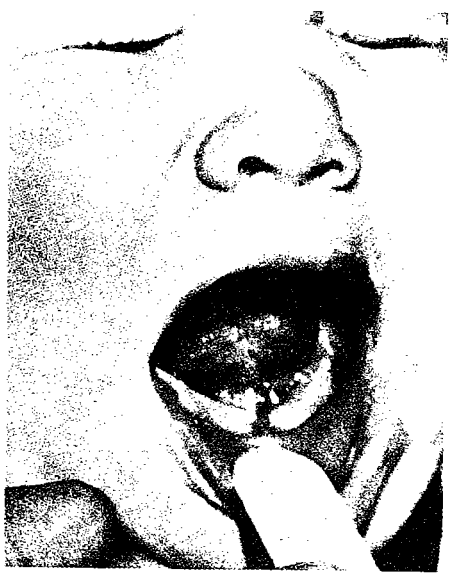






This closure of the muscle gap advanced the sides of the lower lip cleft medially enough to allow excision of the notched free border vermilion. A step closure was designed which carried the mucocutaneous junction line in an overlapping vermilion flap. Then only a 3 mm. vertical V skin excision was necessary to facilitate the alignment of the mucocutaneous "white roll" and still achieve a full-bodied vertical closure of the vermilion free border without tendency toward notching. The ends of the mandible were left undisturbed with the plan to complete bony continuity at a later age. This will probably require a curved iliac onlay graft for added contour as well as union of the two fragments. The tongue maintained its freedom, and the lip healed with a satisfactory aesthetic and functional result.





A complete cleft of the lower lip would be treated with the same general design but would require an inverted V paring of the cleft edges to allow a three-layer closure with emphasis on the muscle approximation. When the cleft extends into the neck, a

Z-plasty may be of value, but if there is a marked lack of tissue in the area, well-planned local flaps may be necessary to achieve adequate chin-neck construction.

EARLY BONE GRAFT

At the 1973 Cleft Palate Congress in Copenhagen, a lower lip cleft was presented by Jan Grochowski, Puk Erwin and Gallas Zofia of Krakow, Poland. This was a complete cleft of the lower lip and mandible 2 cm. in width with a bifid anterior tongue, fistula in the mental region, absence of the hyoid bone and 1 cm. wide connective tissue bands extending from the free mandibular margins to the sternum. At about three months of age, two tibial bone grafts were used to bridge the mandibular gap. Three months later, the fistula was excised and the lower lip closed by the LeMesurier quadrilateral flap principle. Two years after surgery growth and development were reported to be progressing normally.