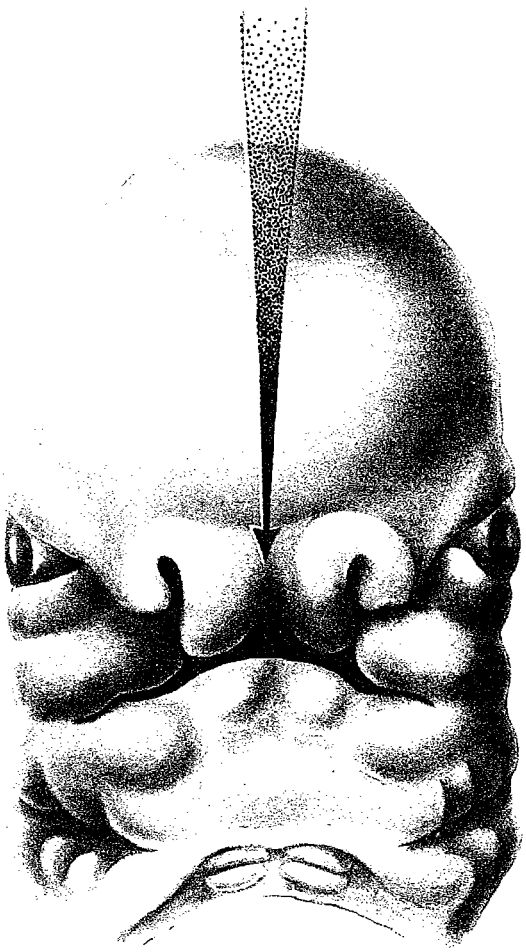
56. *Median Clefts of the Upper Lip*



MEDIAN clefts of the upper lip are considered the result of the failure of mesoderm migration or merging of the nasomedial prominences themselves.

INCIDENCE

Median clefts of the upper lip may be normal in such animals as the hare, the llama and the camel, but in the human they are rare. Belgian Debraisieux in 1904 reported a case and stated:

> Median harelip is one of the rarest occurrences in the list of congenital anomalies.

By 1923 Truman Brophy of Chicago had acknowledged 23 cases on record. Sir Arthur Keith had shown him 12 specimens in the museum of the Royal College of Surgeons, London, and to these he added the Belgian case, one reported by Burke, one by Dun, seven in his own practice and one in the clinic of his friend T. L. Gilmer. Von Bruntz reported no median in 555 clefts of the lip. In 1935 Warren B. Davis of Philadelphia found five median clefts in 688 cleft cases. In 1965 Fogh-Andersen of Denmark reported 15 in a total of 3,988 facial clefts, and in 1968 Vilar-Sancho Altet of Spain added six to the world literature and estimated the incidence at one in a million births.

It is difficult to evaluate these numbers and percentages because of the "true and false" controversy.

Friedrich Trendelenburg

CLASSIFICATION

Two varieties of median clefts were first delineated clearly in the latter part of the nineteenth century by the German surgeon Friedrich Trendelenburg, even more famous for his head-down position. He grouped midline clefts on this sound basis:

1. Double cleft of the upper lip with failure of development of the intermaxilla. . . .
2. True median cleft of the upper lip with development of the intermaxilla.

J. W. Ballantyne in 1904, E. J. Herbst and Apffelstaedt in 1930 and F. Braithwaite and J. Watson in 1949 grouped median clefts into "true" and "false" varieties. Thus, some recordings may be including both groups while others are confined to the so-called true median clefts. The only median cleft described in Brophy's series revealed absence of the lower portion of the nasal bones,

columella, prolabium and premaxilla and, in fact, must have been a "false" or pseudomedian type. Davis' series did not even record whether the cleft was complete or a notch. Out of Fogh-Andersen's series of 15, 7 were complete median (pseudomedian or "false") clefts, and in Vilar-Sancho's cases two were "true" and four were varieties of the "false" type. In addition, accurate recording of the "false" group is virtually impossible as the life expectancy is so short.

In 1938 Veau favored three median groupings: a notch, a median cleft extending to the columella and a median defect caused by atrophy of the whole median element. In 1963 Brucker, Hoyt and Trusler of Indianapolis, with three additional cases of agenesis of the frontonasal process associated with cerebral anomalies and with more focus on the facial anomalies, suggested a general descriptive term, "median cerebrofacial dysgenesis." In 1968, with Sidney Williams, I proposed

> that any congenital, vertical cleft through the center of the upper lip, no matter to what extent, be classified as a median cleft of the lip. If any vestige of prolabium is present, then it becomes bilateral.

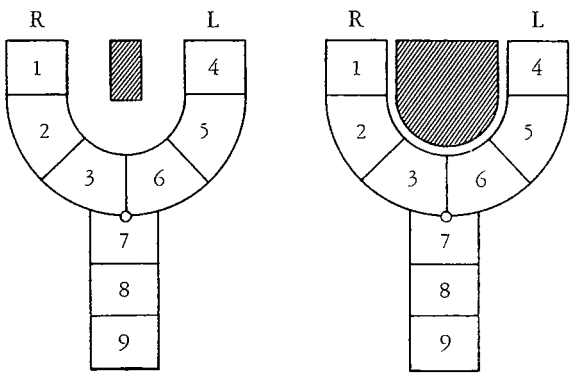
We further divided median upper lip clefts into two groups: (1) agenesis of the medial element and (2) clefts of the medial element.

In 1971 Pinto and Goleria of Bombay divided median clefts into two groups also and refer to group I as *agenesis* with gross deficiency of the tissues that would have developed from the median nasal process. They refer to group II as *failure of fusion,* which may vary from diastema to median lip muscle gaps to median clefts associated with broadening and duplication of philtrum, columella, nose tip and nasal septum. They suggested that faulty fusion accounts for midline sinuses, dermoid cysts and fistulae. They noted that the developing face presents a very broad, flat configuration with widely separated eyes and nasal pits. Reduction of this wide interorbital and interolfactory distance is accomplished by what Mehta Lopa and Kothari have termed medialization—the rapid growth of lateral mesoderm pushing the eyes and nose toward the midline. This process can vary from

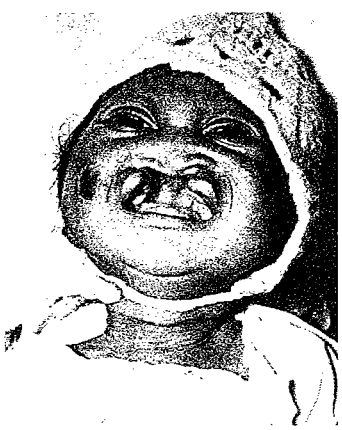
extreme hypertelorism to actual cyclopia. Medialization also brings about invagination of the median nasal process to form septum, columella, philtrum, frenulum and premaxilla. Hypomedialization will be responsible for hypertelorism, thick septum, broad or double philtrum, broad or double columella and double frenulum.

SYMBOLIC RECORDING OF MEDIAN CLEFTS

To facilitate the recording of the two main types of median clefts, Desmond Kernahan of the Chicago Children's Memorial Hospital in the May 1973 *Plastic and Reconstructive Surgery* added symbols in the center of the fork of his striped Y. A small midline block was used to represent a central cleft lip while a total filling of the fork signified a median cleft lip with absence of the primary palate.



A SUBDIVISION

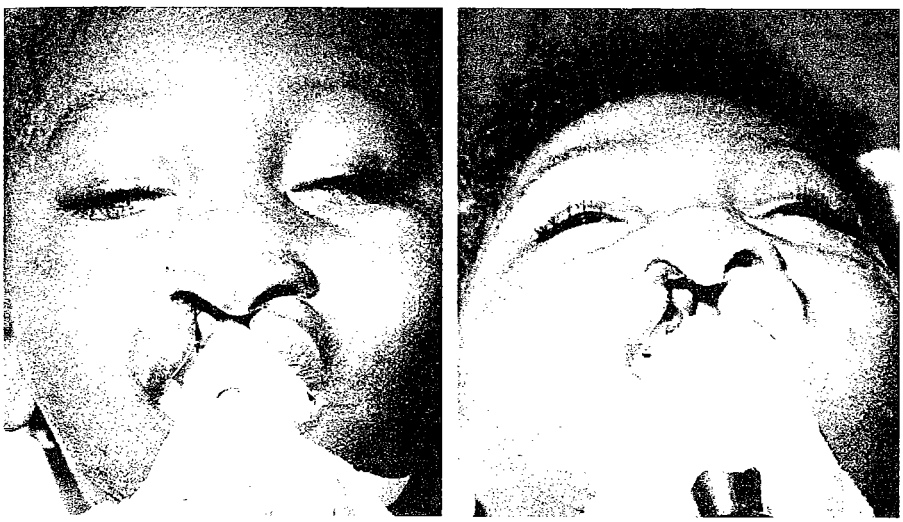


In 1973 Ingolf Koblin of the University of Düsseldorf proposed the classification of median and pseudomedian clefts of the primary palate. He acknowledged the true median clefts (deficient development and penetration of the central mesoderm) and his special type of pseudomedian clefts (absence of the central mesoderm causing a defect of the related structures) as seen in unilateral and bilateral clefts associated with hemiplasia and aplasia of the premaxilla.

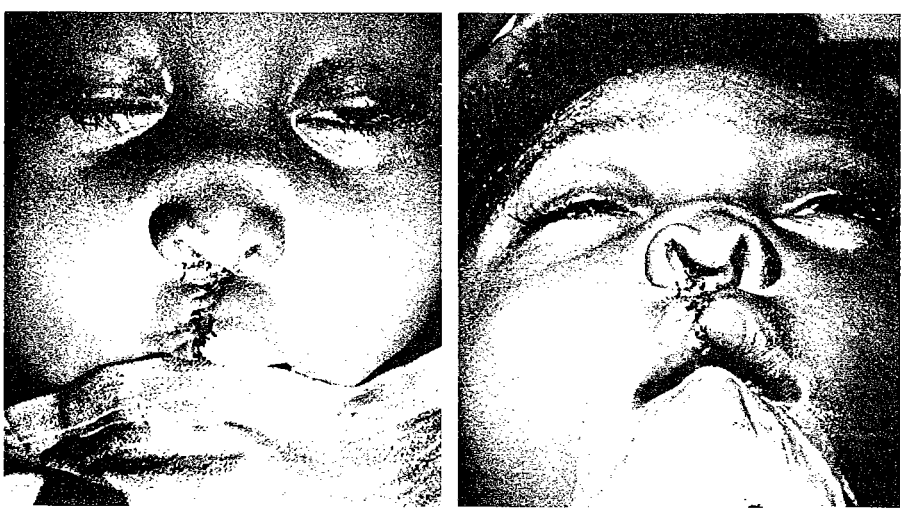
This case, with no family history of anomalies, might be

considered a case of pseudo-pseudomedian cleft. There was a unilateral cleft lip with absence of the anterior septum and premaxilla but a severe median cleft of the secondary palate. There were also several associated anomalies: cryptorchidism, hypospadias, heart defect, webbed neck, aplastic anemia and cephalic abnormalities possibly including mongolism. The patient died in infancy.

Here is what I would interpret as a Koblin pseudomedian cleft with a unilateral cleft of the lip, absence of the septum and hemiplasia of the premaxilla.



The temptation was to treat this anomaly like a median cleft, but the presence of two-thirds of a Cupid's bow on the left stimulated use of the rotation-advancement principle, which at least achieved lip balance with normal landmarks. The alar bases were placed in symmetry, and the skin bridge, which was all that was present of a columella, was centralized.



If the patient reappears at about 15 years of age, a Gillies hinge graft will raise the nasal tip and create a bridge at the same time.

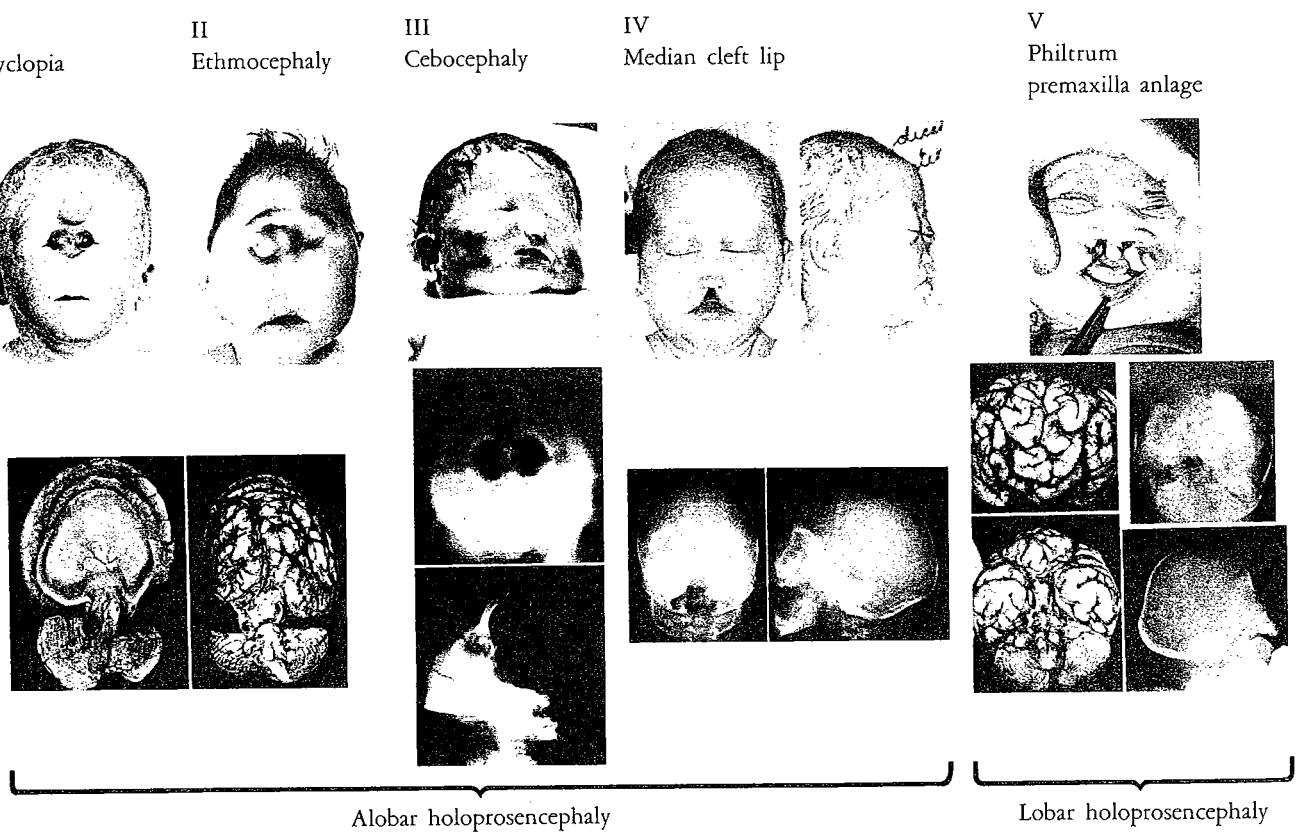
William DeMyer

DEMYER

The present undisputed champion of median cleft lip, its classification, diagnosis and medical management, is William DeMyer, Professor of Neurology, Indiana University School of Medicine. He sees three or four cases a year. In previous articles with W. Zeman and C. G. Palmer in 1963 and 1964 and unaided in 1967, DeMyer set the stage for Grabb to invite him to write an excellent section in 1971 on median cleft lip. He noted the distinct and separate syndromes of facial anomalies associated with complete median cleft lip:

1. The syndrome of median cleft lip with orbital hypotelorism.
2. The median cleft face syndrome with hypertelorism.

Diagnostic Facies of Holoprosencephaly and Parallelism with the Brain



Reproduced from Kurlander, C. H., DeMyer, C. J., and Campbell, J. A., *Radiology* 88:473, 1967.