III. Rare Clefts

This world is very odd we see,
We do not comprehend it.
But in one fact most all agree,
It's worth a try to mend it.
An Interdisciplinary Workshop Conference on the Evaluation of Recent Advances in Cranio-Facial Surgery was held at the University of Illinois Medical Center, November 1974. Resident David Slepyan in attendance at the conference returned enthusiastic about Paul Tessier's classification of craniofacial clefts based on his lifetime collection of cases. Craniofacial and orbitomaxillary clefts are rare malformations, compared to cleft lip and cleft palate, and extend through constant lines or axes through the eyelids or eyebrows, nostrils, lips or maxillae. Bone and soft tissue are seldom involved to the same extent. Soft tissue defects are more common from the midline to the infraorbital foramen while bony defects are more severe lateral to the infraorbital foramen.

It is Tessier's theory that facial clefts have their origin on the cranial base and thus can be traced from the cranium through the orbit to the face regardless of main blood vessels or growth centers. Cleft O represents midline clefts from face to base of
cranium. Clefts 1, 2, 3 are standard cleft lip but traverse the nose at different positions, cleft 1 affecting the alar dome, cleft 2 passing through the mid-alar cartilage and cleft 3 being the more familiar nasolacrimal-ocular cleft. Cleft 4 spares the alar base as it extends to the medial orbit. Clefts 5 and 6 form curved patterns through the cheek, and, since they are lateral to the infraorbital foramen, the bony deficit is greater than the soft tissue deficiency. Cleft 7 is associated with hemifacial microsomia. Clefts 8 and 9 are not well documented. Clefts 10, 11, 12 and 13 are extensions of clefts 4, 3, 2 and 1 above the orbit at the cranial base.

A PRACTICAL APPLICATION

An adult Vietnamese male with a wide unilateral cleft of the lip involving the alveolus but with the columella intact and a unilateral cleft of the alar rim, was discussed by Jean Maes in Plastic and Reconstructive Surgery, September 1974. The patient had caused Maes much trouble for not only had he failed to return for scheduled surgery but he refused to fit into a standard classification. Richard Stark, after ruling out noma, interpreted the anomaly as a median cleft of the primary palate and cleft nose with imperfect naris formation. Actually, the case seems to fit quite simply into Tessier's classification of facial clefts as cleft 1, a standard unilateral cleft of the lip traversing the nose at the alar dome.
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 Debrasieux 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-Sanco Allet 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.

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 Apfelstaedt 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. Hypo-medialization 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.

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

I  Cyclopia  II  Ethmocephaly  III  Cebrocephaly  IV  Median cleft lip

DeMyer's group I is characterized by a median cleft of the lip and absence of the premaxillary bone, nasal septum, nasal bones and crista galli. The ethmoid bone, which sets the interorbital distance, is hypoplastic, resulting in orbital hypotelorism. The eyebrows join each other across the midline. Occasionally, the forehead reveals a median keel-like protrusion known as trigonocephaly. The secondary palate may or may not have a median cleft. This group has an unmistakable facies, and it is pathognomonic of a severe congenital brain defect with amentia.

This face predicts the lack of brain

DeMyer has a simplified delineation of the central nervous system as a tube sealed off at each end. Normally, the rostral end bulges out like an expanding balloon which undergoes diverticulization into paired cerebral hemispheres, into paired optic bulbs and into olfactory bulbs.
In this anomaly there is serious curtailment of diverticularization. Instead of dividing into hemispheres, the cerebrum remains holospheric like the original simple balloon, so the generic term for this process is *holoprosencephaly*.

The olfactory diverticula are almost always absent in such a brain, a condition which earns the term *arhinencephaly*. The arrest in diverticularization can occur at any stage. When there has been no hemispherization of the brain, the interhemispheric fissure is absent and there are no lobes, then the malformation is *alobar holoprosencephaly*. If the arrest occurs at a later stage and the face and brain are better developed—the interhemispheric fissure extending to the frontal poles, which still keep their continuity across the midline—the malformation is *lobar holoprosencephaly*.

Although median cleft lip with orbital hypotelorism is the commonest facies with holoprosencephaly, it is merely one stage in a spectrum of malformations. In 1967 C. J. Kurlander, W. DeMyer and J. A. Campbell diagramed a fascinating group of facies of holoprosencephaly.

Facies I is the severest degree of orbital hypotelorism, revealing *cyclopia* with a single median eye in a single median orbit. A subgroup showed a single globe in a single orbit but with two corneas.

Facies II is *ethmocephaly*, with the orbits completely separated and the tubed proboscis having migrated from the forehead to between the eyes.

In facies III or *cebocephaly*, the proboscis lies on its side but is still a fleshy tube lacking skeletal support. DeMyer calls the nasal shift from I to III the “march of the proboscis.”

In facies IV, the proboscis has been replaced by nares, but the nasal bones and septum are absent. In facies I through III, although the lip is not cleft, it has no true philtrum and the premaxillary bone is missing. Facies I through IV, according to DeMyer, are almost invariably associated with alobar holoprosencephaly, although one of his cases in IV had lobar holoprosencephaly.

In facies V there appears the intermaxillary segment consisting of rudimentary prolabium and premaxilla which removes it from
the median cleft grouping. Facies V has a variable expression of holoprosencephaly, sometimes arhinencephaly or even a brain that is generally normal but usually small and deficient in developmental potential. DeMyer states that the median cleft lip facies IV is pathognomonic of holoprosencephaly, for among 30 autopsied cases in the literature and 14 of his own there have been no exceptions. Occasionally, holoprosencephaly occurs without a warning facies so that DeMyer editorialized:

The face invariably predicts the brain, but the brain does not invariably predict the face.

**DIAGNOSTIC CONFIRMATION**

Confirmation of orbital hypotelorism is obtained by measurement of interorbital distance on posteroanterior x-ray films of the skull. Electroencephalography will show abnormal patterns. Transillumination is also possible.

**TREATMENT**

Treatment of median cleft lip with holoprosencephaly is usually not in the realm of plastic surgery. As pointed out by DeMyer, patients in facies I through III invariably die in the neonatal period, and those in facies IV usually die in the first year. Patients in facies V may survive to adulthood, but death in the early years of childhood is likely. Infants with alobar holoprosencephaly, facies I through IV, are amented, whereas infants with lobar holoprosencephaly, facies V, may show slight developmental progress but will be hopelessly retarded.

Patients with median cleft lip and holoprosencephaly have difficulty with feeding, temperature control and convulsions. DeMyer notes that a regular infant diet can be administered through a bottle or Asepto syringe, that oscillations in temperature need not be treated with antibiotics unless a specific infection is diagnosed and that phenobarbitol or Dilantin need be given only after the onset of clinical seizures.
GENETIC COUNSELING

From reports in the literature there is evidence that, once a child with median cleft lip and holoprosencephaly has been born in a family, the risk of another affected child is high. For this reason, DeMyer advises that the parents be warned of this reproductive risk so they can make a rational decision whether to procreate or adopt.

UTILITARIAN SURGERY

As I noted in 1968, all of these patients are without mentality, do not survive beyond infancy and thus do not require corrective surgery. Once the diagnosis of alobar holoprosencephaly has been made, the prognosis is sealed and the parents should know that only one in a hundred survives the first year. It is far better that the infant be placed in an institution. If the parents insist on keeping the child, and if it survives more than the usual period, closure of the median lip cleft is possible. Extensive facial reconstruction, of course, is not warranted, but any large median gap in the upper lip is ideal for a lower lip-switch flap. When the columella is missing, it can be partially constructed by extending the distal portion of the flap. The lack of nasal support will certainly prevent an aesthetic result, but closure of the cleft does facilitate feeding and improves appearance for the family’s relief during the indefinite existence of the patient.
This semilobar holoprosencephaly patient with small brain, hypertelorism, flat nose with absence of septum and columella and median cleft of the lip and alveolus had a primary Abbe flap with a columella extension to facilitate feeding. The patient lived to the age of 18 months and the case was published in *Plastic and Reconstructive Surgery*, July 1968.

C. R. Dehaan, for Stark in 1968, reported on a median cleft of the lip associated with microcephaly, arhinencephaly, orbital hypotelorism, bilateral exophthalmos, absence of the entire central portion of the lip (including the premaxilla but with a normal palate posterior to the incisive foramen) and a single central ostium leading into a nasal cavity devoid of vomer, septum and ethmoid plate. He had treated four "true" median clefts which were actually of the pseudomedian or "false" type. He described the surgical treatment:

The clefts were repaired by a simplified straight-line technique during the first week of life to facilitate feeding. The infants tolerated the surgery well, but none survived longer than 5 weeks.

C. J. T. Pinto and K. S. Goleria of Bombay, India, in 1971 at the Congress in Melbourne added three agenesis cases with median cleft lip and holoprosencephaly to the world records, stating that for these no surgical treatment had been indicated or administered.

**FOUR EXCEPTIONS**

The cases of B. Vilar-Sancho Altet, Jacob Longacre, Jack Fisher, and James Wells constitute four exceptions. B. Vilar-Sancho Altet of Madrid, Spain, reported a case in *Revista Española Cirugía Plastica* in 1968 that certainly confuses the issue and breaks DeMyer's rule that *the face invariably predicts the brain*. A nine-year-old male had agenesis of the right eye and hypotelorism, elongation of the middle third of the face with absence of the dorsum of the nose as well as absence of the premaxilla and prolabium. The patient also had tetralogy of Fallot but other-
wise was normal, even in intelligence. Vilar-Sancho Altet admitted that had he first seen the patient as a newborn he would have diagnosed his condition as arhinencephaly and postponed surgery.

Treatment: The median cleft of the lip was closed with an Abbe flap, the distal portion being used to construct the columella. Both Vilar-Sancho Altet and I reported the same approach on median clefts in the same year, 1968.

The generous Jacob J. Longacre of the University of Cincin­nati collected rare books and rare cases over the years until he accumulated the third largest private plastic surgery library in the world. He treated some very fascinating patients. Cognizant of his treasure chest of cases, I requested a median cleft and was rewarded with this rarity treated with split-rib grafts and this comment:

In review of the literature I have failed to pick up another case which following survival was so treated.

As crowned champion of the split-rib graft, Longacre first used the principle in 1952 and applied it to a variety of deformities, publishing about 20 papers on the subject. In December of 1974 he kindly forwarded this unpublished case report.
This child was born with a complete midline defect with absence of premaxilla, prolabium and columella with associated cleft lip and cleft palate deformity. The findings of midline cleft, hypotelorism, and microcephaly are most compatible with a diagnosis of arhinencephaly. The lip was repaired at five months and revision of the lip repair by Dr. O'Malley in Orlando at age one year. Motor and sensory development was slow; the patient was still crawling when first seen at age two. Frontal and lateral x-rays revealed the microcephalic skull with the grossly deformed and hypoplastic maxillae and maxillae with associated midline cleft and absence of the premaxilla. The interorbital distance measured 15 mm. due to hypoplasia of the ethmoids.

Following expansion and cleft palate repair by Dr. Longacre at age four, he developed a vocabulary of 250 words within six months and two of the words he used constantly were "no nose, no nose." A columella was reconstructed with local tissues and the defect of the middle third of the facial skeleton reconstructed in two stages with split-rib grafts to malar, maxillary and nasal regions improving patient's appearance at age four and one-half, and six.

Bilateral ptosis of eyelids still gave the patient a very stupid appearance. This was corrected with fascial slings tied into the frontalis muscle during the operation for insertion of the third set of onlay split-rib grafts. Following this, the patient showed a tremendous spurt of improvement in vocational, academic, and social adjustment. He is now ten years of age and attending regular school. Frontal and lateral x-rays reveal the autogenous bone support provided to the extensive midline defect of the facial skeleton by the staged split-rib grafts.
An odd coincidence

During the 1974 American Board of Plastic Surgery examinations in Portland, Oregon, one of my questions directed at Jack D. Fisher was on the subject of holoprosencephaly. Five months later, he repaid me by reporting that he now had a case of a related nature. Subsequently, photographs and the accompanying history and physical findings were forwarded from the University of Virginia Medical Center, Charlottesville, by resident James H. Wells, who with Jack Fisher will be publishing this case report.

This three-month-old female, born of unrelated parents but with a paternal aunt having unilateral cleft lip and palate, was the product of a pregnancy complicated with polyhydramnios and a question of aspirin overdosage at two to three months' gestation. Her birth weight was 5 pounds 5 ounces, her Apgar score 8 at one minute and 9 at five minutes.
Her deformities include a hypotelorism, cleft lip and palate, clinodactyly, and a microcephaly; head circumference was 32 cm. at two months. She had a neuroencephalogram performed, and this is a summary of the radiologist's report.

Impression: This examination shows evidence of severe focal superficial atrophy with an almost porencephalic area in the right parietal region. An absence of the septum pellucidum, slightly fuller ventricles than one would expect in this age, and midline cerebellar agenesis or atrophy.

It was reported by Wells:

To date, she appears to have a normal growth and development pattern. She is eating well, gaining weight, utilizing a Lamb's nipple.

Discussion with Fisher indicated his plan of bilateral rotation of the lateral lip elements with closure of the muscles in the midline. This seems a logical first step, and if the patient shows reasonable progress, then an Abbe flap with an extension could create a philtrum and columella. Craniofacial surgery in a reverse Tessier and split-rib grafts might be indicated in this case and possibly in that of Vilar-Sancho Altet.

SURGICAL TREATMENT OF HYPOTELORISM

Although most cases of orbital hypotelorism will not deserve surgical correction, as noted there are exceptions. In 1975 in Plastic and Reconstructive Surgery, Converse, McCarthy and Wood-Smith reported the first correction of an orbital hypotelorism. Although no cleft was involved, the case and the treatment are of interest.

The patient was a Caucasian girl born with orbital hypotelorism and hypoplasia of the nasomaxillary region. The nose was flattened, the columella shortened and the septum absent. She received grafts of autogenous cartilage, composite earlobe and preserved cartilage to her nose between the ages of two and nine years. At 14 years she revealed an intercanthal distance of 19 mm. and radiographic interorbital distance of 14 mm. (24 mm. expected for this age). A diagnosis of nasomaxillary dysostosis (Binder) was made.
In 1972 surgical correction was undertaken through a combined craniofacial route, similar to that for correction of orbital hypertelorism. The bony orbits were mobilized by superior horizontal osteotomies, vertical, inferior horizontal and paramedian osteotomies and a transverse cut of the orbital roof. Segments of bone were removed from the lateral aspect of each roof.

The orbits were then translocated laterally for a total distance of 8 mm. The resulting defects in the roof and in the nasofrontal areas were filled with iliac bone grafts [BG].

The patient subsequently had an inlay nasomaxillary skin graft, and the total surgery resulted in a definite improvement in her appearance.
58. Median Cleft Lip with Hypertelorism

In DeMyer's group II, the median cleft face syndrome, there is a median cleft lip, bifid nose, orbital hypertelorism and cranium bifidum occultum. The premaxilla is present except in the severest cases, but it may be cleft, with incisors erupting along the premaxillary cleft. The brain is usually normal or may show hypoplasia or absence of the corpus callosum. Intelligence is normal or only mildly impaired, and life expectancy is normal.

Embryologically, the median cleft face syndrome can be regarded as an arrest in facial development. The eyes begin on the sides of the face and, normally, migrate medially for stereoscopic vision. The nose begins relatively broad and flat, the nares being separated by a cleft formed by the medionasal processes. Somehow, the eyes stop their migration too far apart and the nose remains bifid, but whatever the process, it rarely interferes with forebrain morphogenesis and the brain develops normally.

**TREATMENT**

As patients with the median cleft face syndrome usually have normal mentality and longevity, they are candidates for corrective surgery.

In 1968 I attempted a total listing of all cases of this type recorded in the literature and added four that we had treated. This listing of 25 did not include any of Brophy's 23 as no
description of the type of cleft had been made, nor did it include Davis' series, for the same reason. To my list must be added two cases reported in 1934 by Ritchie and one of Vaughan's as well as a case published by DeMyer in 1967 with a median cleft of the lip, bifid nose, orbital hypertelorism and cranium bifidum occultum. Also in 1968, Vilar-Sancho Alter described two more of this type of median cleft.

Then in 1971 Pinto and Goleria added another eight cases and described their treatment. Others have been discovered to a total of 74, as will be noted.

In 1972 L. A. Krikun reported 64 median clefts of the nose, noting the typical deformities such as the groove, hypoplasia of the septal cartilage, cleft of its anterior margin, alar cartilage atrophy and deformity, abnormal position of the triangular cartilage and nasal bones and hypertrophy of the terminal part of the nose and columella. The most frequent abnormalities of the face were found to be hypertelorism, divergent strabismus, a wedge of hair on the forehead, deformation of the frontal bone, wide separation of the eyebrows, median cleft of the upper lip vermilion and abnormal development of the dentomaxillary system.

Then there are the great cases of Tessier and, undoubtedly, there are others. It is of interest that John Converse of New York University Medical Center, as of November 1973, had a series of 26 patients with ocular hypertelorism (Crouzon-Apert). Of these, two had bilateral cleft lip and palate and one a cleft of the hard and soft palate, but none had a midline upper lip cleft.

1. Median cleft of the lip musculature without cleft of skin or vermilion was reported by Pinto and Goleria of India. Treatment: Through midline vertical skin incision, muscles mobilized and approximated across the cleft with chromic catgut. Skin closed with nylon.

2. A vermilion notch was cited in a case of H. S. Vaughan of New York.

3. In a case reported by Braithwaite of England, a median
notch of the upper lip vermilion extended upward to the level of
the mucocutaneous junction, which continued upward as far as
the columellar base as a flattened shallow depression. The col­
umns of the philtrum were more separated than is normal, but
the nose was normal, except for a broad columella. A double
frenulum extended from the lip to the alveolus. Although
Braithwaite considered his “notch” case a separate entity, subse­
quent cases show it to be the least of varying degrees of the same
median cleft process involving the lip and nose.

4, 5. Similar vermilion notches were seen in two brothers
reported by Fogh-Andersen of Copenhagen.

6. A 25-year-old white female had a minimal midline notch of
the vermilion and a tiny dimple at its center with its tract
extending through the orbicularis oris muscle to a fibrous band
attached to the nasal spine. There was also hypertelorism (43 mm.
between medial canthi) in this case reported by Bartels and
Howard of Orlando, Florida. Treatment was confined to excision
of the total tract and closure.

7. A 19-year-old female with a minimal vermilion notch, lack
of muscle union, diastema and an ovoid swelling of the left part
of the lip was operated on once before being seen by Pinto and
Goleria. Treatment: Through midline incision, muscles approxi­
mated, fibrofatty mass excised and vermilion notch corrected.

8. Median notch of the vermilion and cleft of the soft palate
were reported by Vilar-Sancho Altet of Madrid. Treatment:
Z-plasty of the vermilion and Wardill-Veau-Kilner pushback of
the palate.

9. Midline cleft of lower quarter of the upper lip with an
apparently normal nose was noted by Gabka of Berlin.

10. Median cleft of the vermilion and lower quarter of the lip
with groove to the columella and no nasal deformity was re­
ported by Vilar-Sancho Altet. Treatment: Inverted V excision by
paring the edges and closure in V-Y fashion.

11. A one-year-old child with partial cleft lip associated with
notch of alveolus and bifid frenulum was reported by Pinto and
Goleria. Treatment: Excision of central vertical diastasis and use
of a double Z-plasty.
12. An 11-year-old male with a partial median lip cleft, a diastema, a vestige of a prolabium and a protuberant polypoid mass from the right nostril attached to the lower part of the septum was reported by Pinto and Goleria. Treatment: Excision of the polypoid mass and simple paring of the cleft edges in an inverted V with approximation in layers of the deeper tissues.

13. A five-year-old boy had a median cleft of the inferior quarter of the upper lip with a vertical submucous cleft extending to the columella associated with two pedunculated club-shaped skin masses, one projecting from the septum out of the left nostril and the other from the alveolus margin between the central incisors. There was also a double frenulum, crescent defect of the left ala, bony depression over the frontal bone and hypertelorism in this case reported by Laxman Sharma of Nagpur. Treatment: V excision of cleft margins closed lip defect. Septal mass used in reduced form to reconstruct alar margin.

14. A median cleft of the lip from vermilion to columella with a normal nose was reported by H. P. Ritchie of St. Paul.

15. A six-month-old boy with median cleft of half of upper lip, short broad columella with median groove, alar cartilages separated in the broad nasal tip and double frenulum was noted in India by Laxman Sharma. Treatment: Three-layer closure of lip cleft after paring of edges and dissection of muscles.

16. A 30-year-old male with a partial cleft of the vermilion, a double philtrum, irregularity of the teeth with a supernumerary incisor just below the cleft and cleft of the left nostril was reported by Pinto and Goleria. Treatment: V excision of middle of lip with approximation in layers and local rotation correction of notched nostril.

17. A midline vermilion cleft with shortened nasal alar cartilages, atypical cleft palate, malformed lobulated tongue and normal chromosome findings (oral-facial-digital syndrome) was reported by Brucker and others.

18. An oral-facial-digital syndrome with a slight median cleft lip, atypical cleft palate and 13–15 trisomy was reported by Fogh-Andersen.

19. An oral-facial-digital syndrome with a slight median cleft lip, atypical cleft palate and normal chromosome findings was also reported by Fogh-Andersen.
20. Oral-facial-digital syndrome was associated with cleft in the mid-portion of the tongue with long pedunculated mass of tissue, pseudocleft of mid-upper lip border, total absence of premaxilla, hypertelorism, atypical cleft of secondary palate and encephalocele in palate fissure. Treatment: Encephalocele operated on at six months and lip and tongue at one year. Tongue mass revealed flat epidermal-like epithelium without keratinization covered with nests of striated muscles in richly vascularized connective tissue stroma as reported by Poradowska and Jaworska.

21. Oral-facial-digital syndrome was associated with pseudocleft of the mid-upper lip extending to the alveolar process, short columella, gross mandibular hypoplasia, complete cleft of secondary palate with fibrous band running from buccal mucosa, trilobulated tongue attached to the floor of the mouth and microphthalmus. No treatment because of early death. Case reported by Poradowska and Jaworska of Warsaw.

22. A pseudo-median cleft of the upper lip was found in one patient with Treacher Collins syndrome by Poradowska and Jaworska.

23. A seven-year-old Haitian girl (operated on at Albert Schweitzer Hospital, Des Chapelles, Haiti, 1964) had a median upper lip cleft extending vertically through the vermilion and skin about 1 cm. and continuing as a flattened shallow depression to the base of a broad columella. The philtrum columns were widely separated, the columella was wide and it had a vertical elevated ridge running up its center. The left nostril was larger.
than the right. There was no alveolar notch or palate cleft. This is just a more advanced degree of notching, with the same but more exaggerated ancillary deformities. The case was published in *Plastic and Reconstructive Surgery*, July 1968. Treatment: Elliptical excision of the midline columellar prominence narrowed the column. Excision of a narrow inverted shield from the flattened central lip moved the philtrum column into better position, created a suggestion of a philtral concavity and produced the skin lengthening and the vermilion fullness of a cupid's bow and tubercle.

24. A nine-month-old male child with a wide median cleft of the lip, cleft of the alveolus and broadening and flattening of the nose, lip and columella with a fatty mass protruding from the base of the columella was reported by Pinto and Goleria. Treatment: The lip was closed by a vertical excision and a double Z-plasty. A vertical excision of the skin of the polypoid mass allowed use of the fatty tissue to round off the flat nasal tip.

25. A median fissure through the nasal tip, columella and lip was reported by H. P. Ritchie.

26. A case of bifid nose with minimal median cleft of the upper lip extending into the alveolus was reported by Francescon of Italy for Mustardé.

27. A 19-year-old female presented a partial median cleft of the lip, bifid frenulum, bifurcation of the nose, which was short but broad, and a diastema, as reported by Pinto and Goleria. Treatment: First step in treatment was a forked flap to elongate the columella and a V excision of the middle of the lip with closure in layers.

28. A one-year-old child with a median cleft of the lip and alveolus, a bifid nose with a projection of fibrofatty tissue from both nostrils and a coloboma of the right upper eyelid was presented by Pinto and Goleria. Parents refused surgical correction.

29. An incomplete median lip cleft, cleft palate and meningocele of the septum was reported by Baibak and Bromberg of New York.
30. A six-month-old Jamaican boy (operated on at the Kingston Public Hospital, 1963) had a median cleft of the vermilion and skin about one-half the vertical length of the upper lip. The philtrum columns were widely separated, with a groove extending to the columellar base. The columella was quite broad, contained a raised vertical ridge and was extremely short. There was a slight midline notch in the alveolus. This case was published in *Plastic and Reconstructive Surgery*, July 1968.

Treatment: A forked flap was marked on the medial flattened edges of the lip cleft. The midline ridge was excised from the columella; it was then narrowed and lengthened by advancement of the forked flap. The freshened lip was brought together effectively in the midline. Sutures were removed on the fourth postoperative day, and wounds were healing well. Patient never returned to the clinic for follow-up.
31, 32, 33. Median clefs, referred to as "true" and associated with malformations of the hands, were in Fogh-Andersen's series.

34. Stephanie in 1939 reported a median cleft of the upper lip with reduplication of the columella and nasal tip.

35, 36, 37, 38, 39. Esser in 1939, from Sanvenero-Rosselli's clinic in Milan, reported five such cases of median cleft lip and reduplication of the columella and nasal tip. These five were all in the same family!

40. Peet and Patterson of Oxford presented a bifid nose with a partial median cleft lip but no defect of the alveolus and palate. Treatment: After simple approximation of the lip elements, an inverted V to Y advancement of the excess skin from the wide upper nasal bridge moved tissue into the bifid tip. The alar cartilages were later approximated for an excellent result.

41. An adult with median cleft of the lower one-third of the upper lip and a true bifid nose with flattening and separation of the nasal bones and a cleft between the alar cartilages was reported by Francesconi of Italy in Mustardé's 1971 book, Plastic Surgery in Infancy and Childhood. Treatment: His management of this case is of interest. A midline vertical excision of the skin and subcutaneous tissue of the nose and lip was followed by approximation of the separated nasal structures and a modification of the Hagedorn-LeMesurier quadrilateral flap for the construction of the upper lip.
Of equal interest are his thoughts on the causes of the deformity and the timing of the surgery. Giuseppe Francesconi of the Universities of Pisa and Milan developed his early interest in median malformations while in training with Sanvenero-Rosselli. During that time he had followed 12 cases, and since then he has added another unpublished four, one of which with rhinencephalon has been studied in anatomical detail. He tends to accept Sanvenero-Rosselli's suggestion that bifid nose and median clefts of the lip are anomalies produced by disturbance in the fusion of the median raphe or median dysraphia of the face.

In his ancient villa in the Tuscan country near his Hospital of Lucca, Francesconi can escape these depressing malformations long enough to think and write about them. He expressed his leaning toward wise old Kazanjian's "late reconstruction," stating that extensive surgical procedures performed in childhood on delicate structures such as the nose may be dangerous. Although perhaps producing a satisfying result initially, the late result may be disastrous due to interference with normal tissue growth as the result of the trauma of the operation. Nevertheless, small operations . . . can be performed in childhood to improve nasal respiration and get rid of some of the deformity but a full plastic repair of the nose, which will involve cartilage and bone, must be deferred until late childhood or early adult life when the nasal structures are more developed.

42. A case was reported by Weaver and Bellinger of a median cleft of the lip extending to the base of the columella, with a split in the nasal septum, divarication of the alar cartilages and a bifid nose without skin division. Hypertelorism was present. Treat-
ment: This median cleft of the lip was closed by freshening the edges at two weeks of age. At two months the divided septum and alar cartilages were approximated, but no attempt was made to narrow the widely separated frontal processes of the maxilla. At three years the child still had a wide nasal bridge and tip.

43. A similar case (to 42) was reported by Lagos Garcia with a median lip cleft, bifid nose and hypertelorism.

44, 45. Similar cases to 43, and similar to each other except that one was Negro and the other Caucasian, were reported by G. C. Scrimshaw of Oakland. Both had median clefts of the lip and alveolus, bifid noses and hypertelorism.

46. DeMyer published a case with a median cleft of the lip, orbital hypertelorism, low V-shaped hairline, cranium bifidum occultum, bifid nose, median cleft palate, cleft premaxillary bone and normal life expectancy. No treatment was mentioned.

47. A three-month-old Jamaican girl was seen by us with an incomplete median cleft of the lip and bifid frenulum, but without cleft of the alveolus or palate. There was an asymmetrical bifurcation of the nose with a short wide columella and cleft of the right alar arch, and the right ala was high. Hypertelorism was present, as was a frontal bone defect with an encephalocele and convolutions of excess forehead skin.
This case was published in *Plastic and Reconstructive Surgery*, July 1968. Treatment: Still incomplete. A modified forked flap and midline closure of the lip cleft was carried out, along with alar rotation and transposition flaps by Sydney Williams. A neurosurgeon advised postponing any split-rib grafting to the frontal defect until the patient was older.

48. A 10-year-old boy from the island of Antigua with a median cleft of the lip vermillion was seen by us. He had a developmental confusion of the upper philtrum with divergence of the columns and a columella that was either absent or extremely wide. The nasal tip was wide and flat, the alae were notched bilaterally, the nasal bridge was wide and flat, and there was associated hypertelorism. A bifid frenulum was noted. His intelligence was within normal limits. This case was presented in *Plastic and Reconstructive Surgery*, July 1968. Treatment: A forked flap of the diverging philtrum columns was used to construct a columella. An L-shaped Silastic implant gave support to the tip and bridge. The alar notches were corrected by rotation into normal position. The patient’s return to Antigua postponed further nasal work.

49. A case was reported by Kazanjian in 1959 of a wide median cleft of the upper lip with absence of the prolabium and apparent absence of the premaxilla. Yet, at age three to six years
the patient presented upper incisors which had to be extracted, as their roots were not surrounded by solid bone. The palate appeared to be normal but x-ray films revealed separation of the two maxillae. There was a severely bifid nose with a split columella and small symmetrical functioning nostrils one inch apart. Hypertelorism was also present. Treatment: At four months of age the median cleft lip defect was approximated, and the nasal bifurcation was brought together. Subsequent multiple procedures included surgery of the cartilage and bone; later, bone and cartilage transplants were done, followed by a forehead flap rhinoplasty. At the age of 26 years, the patient had developed well mentally. In retrospect, the surgeon regretted having operated on the bony section of the nose before the age of 15 years.

50. A similar case to 49, with median lip cleft and severe bifid nose, was reported by Baibak and Bromberg.

51. A case of median cleft lip and bifid nose was reported by Patten of Oakland.

52. A case of epignathus associated with a median lip cleft, palate cleft, bifid nose and severe hypertelorism was reported by Hirshowitz, Mahler and Heifetz of Haifa. Treatment: Surgical excision of tumor with postponement of surgical correction of the median lip cleft and bifid nose. Soft palate cleft was to have standard closure.

53. A 14-year-old Filipino boy had a bifid nose, encephalocele, microphthalmus, hypertelorism, median cleft of the lip, cleft of the primary palate and partial cleft of the secondary palate. Tomographic studies revealed an intact hard palate. Case reported by Converse, Horowitz and Becker of New York.

54. A case of marked bifid nose with a median furrow associated with a median cleft of the upper lip and hypertelorism was reported for Converse by Wang and Macomber of Albany.

55. Midline partial cleft of upper lip associated with broad nasal root in an infant girl with orofaciodigital syndrome was reported by Fuhrmann, Stahl and Schroeder.

56, 57, 58. Three siblings, a sister and two brothers, each with a median cleft of the upper lip and associated polydactyly in the girl and one boy, were reported to me in 1974 by Joya Chowdhury of Calcutta.
59. A median cleft of the lip with a normal nose but hypertelorism which was closed "simply" and was progressing well at two years of age was reported by R. Mladick, C. Horton, J. Adamson and J. Carraway of Norfolk, Virginia.

60. A median cleft of one-third of the upper lip with a groove extending to the columella and aptly entitled "The True Hare Lip" was reported in 1974 by James A. Lehman, Jr., and Suburaydu Cuddapah of Akron, Ohio. There was an associated double frenulum and tubercle and a flattened nasal tip and wide columella vaguely suggestive of a subliminal bifid nose. Treatment: Cleft marginal incisions, muscle approximation in the midline, and a "white roll" interdigitation at the mucocutaneous junction, which at one year revealed an excellent result.

61. Median cleft of the upper lip vermilion with hypodontia and occurrence of Ellis-van Creveld syndrome reported by Norbert Schwenzer of the University of Tübingen, West Germany. Treatment: Correction in one operation at age six years.

62. Unusual median cleft extending from the white portion of the lip to the lower third of the philtrum without complete fissure of the tissue. Treatment: Full thickness V excision correction at age two years by N. Schwenzer of West Germany.

63. A case with bilateral paramedial facial clefting, severe orbital hypertelorism, absence of nasal air passages and deformities of upper and lower extremities was reported in 1974 by Edgerton, Jane, Berry, and Fisher of the University of Virginia Medical Center, Charlottesville. Treatment: Glabella ostectomy, forehead flap for nasal lining and steel wiring at four months in an attempt at early shifting of the canthi. At eight months an abdominal flap on wrist vector was transported to create an adult-size nose. Further correction of orbital hypertelorism was planned prior to school age.

64-68. J. Chowdhury of Calcutta reported five siblings, three males and two females, with V- or quadrilateral-shaped median clefts of the vermilion of the upper lip. They each had postaxial polydactyly. No mention was made of the treatment as the surgery would be simple. It was found that these patients, their normal siblings and their parents all revealed no abnormalities in the chromosome studies.
69. A 57-year-old adult male with a median cleft of half the upper lip, a severely bifid nose and orbital hypertelorism was presented by David Frost on TV on January 18, 1975, from Gibsonton on the Gulf coast of Florida, where the freaks winter. This carnival performer, billing himself as the two-faced man, has been a popular attraction for years in the sideshow. He revealed remarkable adaptation to his untreated condition with not only a successful career in show business but a happy marriage to the lady two tents down exhibiting a peculiar skin condition.

Recently, Riccardo F. Mazzola of Milan University made a remarkable report of frontonasal malformations including five additional midline clefts belonging to this general category and one most unusual dyprosopia.

70, 71. Vermilion lip notch and bifid nose. No treatment reported.

72, 73. Midline cleft of the lip, bifid nose and hypertelorism. No treatment reported.

74. Median notch of lip, bifid nose and hypertelorism. No treatment reported.

Here is a very rare duplication of the face (dyprosopia) with a wide central cleft of the lip, alveolus and palate separating two well-formed, independent noses. Hypertelorism is present with an extra midline orbit, together with its eyebrow.

In the median cleft face syndrome, there are several general areas of surgery, which will be discussed in the sections that follow.

**MEDIAN CLEFT OF THE LIP**

If the cleft of the lip is minimal to moderate, the paring of the edges in an inverted V excision will allow a three-layer closure. A 90-degree angle in the excision is made 2 mm. above the mucocutaneous white roll on each side of the cleft. Approximation of these angles will give the lengthening of the skin in the specific area of the center of the cupid's bow. This will provide not only the skin "spear point" but the heaping of the vermilion of a midline tubercle to create the semblance of a natural cupid's bow.
In cases in which the columella is wide, the V excision should be extended to narrow the column.

![Diagram of nasal structures](image1)

If the lip cleft is minimal with a wide columella, then the columella reduction should be carried out separately.

When the cleft is extremely wide, a midline primary composite lip flap switched from the lower lip is available and can be used if direct closure of the cleft would tighten the upper lip relatively more than the lower.

**SHORT COLUMELLA**

If the columella is short, the sides of the lip cleft can be taken as a forked flap. This procedure, in turn, will freshen the edges of the cleft and allow closure in an inverted V fashion in layers. Again, if the edges of the cleft are incised 2 mm. from the mucocutaneous junction with 2 mm. transverse cuts, the skin point of the center of the cupid’s bow can be created along with the vermilion tubercle.

![Diagram of nasal structures](image2)
BIFID NOSE

This deformity varies greatly in depth of the clefting, extent of spread and amount of asymmetrical distortion. Surgical correction includes bisection with removal of the excess mid-portion of skin, subcutaneous tissue and bone combined with the shifting of the distorted elements into balance. Closure should bring alar cartilages side to side. Alar notches are usually corrected by local rotations.

BIFID NOSE AND HYPERTelorISM

Jerome P. Webster, New York plastic surgery tycoon of Presbyterian Hospital and College of Physicians and Surgeons, Columbia University, in his 1950 classic treatise with Deming in Plastic and Reconstructive Surgery on treatment of the bifid nose, pointed out the association with hypertelorism, either true or suggested, in 8 of his 10 cases. Yet true hypertelorism, or actual increase of the interpupillary distance, was present in only 4 of the 10. He pointed out with symbols that the illusion of hypertelorism was produced by wide spacing of component parts of the face adjacent to the eyes, such as increased intercanthal distance, flatness of a broad nasal bridge, presence of epicanthal folds and widely spaced eyebrows.

In that pre-Tessierian era Webster focused his surgical action on soft tissue and nasal bone reduction and shifting to reduce the illusion. He excised a wide vertical ellipse of forehead, glabella,
nasal bridge and tip skin from the columella all the way up into the hairline. He also resected the excess of the widened nasal bones, shifted them after in-fracture and fixed them with wiring. In addition he removed the subcutaneous fat at the sides of the nose and achieved medial shifting of the inner ends of the eyebrows by excision of the skin between or by V-Y advancement.

If, in addition, the nostrils were notched or retracted with the nasal tip flattened and the nose short, he advanced the entire skin of the nasal dorsum and glabella downward in a large V-Y, which not only narrowed the nasal bridge and corrected the tip but moved the eyebrows closer together.

As elevation of the nasal bridge is also effective in camouflaging the effect of wide eyes and improving epicanthal folds, onlay bone and cartilage grafts were advocated as a secondary procedure. In order to allow growth through infancy and early child-
hood, this aspect of surgery was postponed, but no longer than late childhood to reduce the inevitable psychic trauma.

Even these less extensive procedures can be time-consuming. J. P. Webster—"Webby" to his friends—was one of the founders of the American Board of Plastic Surgery and a great teacher, as is attested by the products of his renowned students. His meticulous and absolute attention to detail with no concern for time started a rumor that he operated at Presbyterian Hospital by the calendar rather than the clock. This is an appropriate time to repeat a Gillies principle: "Speed in surgery is never having to do the same thing twice." Get it right the first time without concern for the little extra time and beware jubilation over rapidity of action. Have your result rather than your speed of execution breathtaking!

**ORBITAL HYPERTELORISM**

As noted previously, median clefts of the lip are occasionally associated with orbital hypertelorism and can be treated simultaneously. The orbital hypertelorism with increased distance between the orbits, of course, is the major problem, involving surgical resection of the excess portions of bone and the shifting of the orbits together.

**THE TESSIERIAN ERA**

The imposing and dedicated Paul Tessier of Foch Hospital, Paris, a man of quiet poise, subtle charm and natural magnetism, has fathered this specialty, and others are following his inspiration and teaching. In the 1972 *Scandinavian Journal of Plastic and Reconstructive Surgery*, Tessier suggested the term *orbital hypertelorism* (OR.H.) and described it as a congenitally abnormally wide distance between the orbits and hence the eyes... [and]... is always a secondary syndrome, generally due to a facial or cranial cleft and sometimes to craniostenosis. Inter-orbital distance (I.O.D.) has to be measured on the skull [as the distance between the two "dacryons" (lacrimal crests) on a teleradiograph] or less accurately by measuring the distance between the medial canthi (I.C.D.).... Average normal I.O.D. is 25 mm.
in females, 28 mm. in males. Classification is based on the degree of inter-orbital widening. . . . 1st degree, I.O.D. 30 to 34 mm., is euryopia or telecanthus. 2nd degree, I.O.D. more than 34 mm., orientation and shape of orbits still are nearly normal. 3rd degree, I.O.D. usually 40 mm., orbits appear lateralized, the cribriform plate is often prolapsed, and the distance between the lateral canthus and auditory meatus is shortened.

Tessier concluded his opening abstract with this encouraging note:

We now have the proof that we can safely displace the whole of the "useful orbit" (i.e. within 8 to 10 mm. from its apex) in the three directions: transversely, as in OR.H.; sagitally, as in retrusion of the face or frontal bone . . .; vertically, as in orbital malpositions secondary to trauma or to some complete orbitofacial clefts, generally unilateral.

Post surgical review of efforts by Converse and Smith in 1959 and 1962, Schmid in 1966 and his own work with Guiot and Rougdrie in 1963 and 1965 showed that they were doomed to failure for, as Tessier noted:

They only moved a small portion of the orbital rim and practically nothing either of the orbital walls or periorbitum, and therefore had little effect in moving the globe itself.

This and other aspects pointed to the necessity for correction by the intracranial route, which was considered as early as 1960 but, as Tessier later wrote:

Nevertheless, it could not be carried out as long as there was a danger of meningeal infection; therefore, we decided to perform a preliminary operation to explore the anterior cranial fossa and to reinforce the dura by means of a dermal graft.

Infrabasal or cranial routes

If the medial orbital wall is vertical and if the olfactory grooves do not prolapse, an infrabasal osteotomy under the cribriform plate will be located above the eyeball equator and will promise success.

If the supraorbital rims diverge a good deal, if the nasal wall of the orbit is oblique and if the ethmoidal prolapse is noticeable, infrabasal osteotomy would be ineffective and the cranial approach is indicated.
Translocation of the orbits, including thin medial walls and hence the frontal process of the maxilla, which is the only strong structure, produces an atresia of the nasal airways, precipitating the necessity for full-thickness septal resection. Hypercorrection is suggested to counteract further growth in the child and to prevent possible intercranial hypertension.

**Tessier's general surgical indications**

For 1st degree cases with minimal hypertelorism, treatment consists of palliative operations. 2nd degree cases involve partial ethmoidectomy via infrabasal route when ethmoidal prolapse is not noticeable. 3rd degree cases involve orbital ethmoidectomy via the intracranial route.

It has been said of the results of this craniofacial surgery: "This is herculean effort to transform the hideous into the ugly."

Tessier responds to that vague retort:

It appears that OR.H. has to be taken in consideration much more from an aesthetic point of view than from a functional one [in these mentally normal patients]. The abnormally wide I.O.D. is ungraceful. However, ugliness arises from the primitive malformations which produce OR.H. Here is the stumbling block. We can easily move the orbits and eyes closer together and grossly correct OR.H. but that does not mean that binocular vision is now possible in every case nor that ugly facial deformities and defects are avoided.

In 1972 I asked my friend Tessier how many cases he had had of median cleft lip associated with hypertelorism and whether he would send examples with the outline of their surgery. He responded in June:

Many cases of ocular hypertelorism have a median cleft lip, but this cleft is more or less complete, sometimes rudimentary. Most of them also have a frontal encephalocele.

Some cases have been previously operated on probably because their cleft was complete, but I cannot assess.

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<td>Partial</td>
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Enclosed, please find two operated cases: complete and partial.

Further details were forwarded in 1973:

The same basic procedure has been used for these two patients.
— cranial and facial routes
— fronto-naso-ethmoidal resection
— "square like" osteotomies all around and inside the orbits
— bringing the orbits closer together
— bone grafts of the four orbital walls and of the malar bone
— bone graft of the nose.

The partial cleft case
The patient with encephalocele also had rib-grafts of the frontal defect, as figured on the drawing.

The case without encephalocele had a "crown-like" frontal bandeau used for locking the orbital frames in a backward position in order to prevent enophthalmia, as figured on preoperative pictures.

Paul Tessier, it seems, gravitates toward the "big ones" in both work and play. As a break from his 12-hour-a-day “skull cracking” surgery he chose to hunt bull elephants in French Equatorial Africa. In khaki jacket and bush helmet, accompanied by 40 black porters in white loincloths, he set off from Bangui into the
jungle on foot. Days later he reappeared with his line of black porters heavily laden with giant tusks of ivory.

There is a charming story of Tessier and Dingman in Paris after one of Tessier's operations for hypertelorism. Reed Dingman, not only an expert hard tissue surgeon himself but also a well-known big game hunter, in typical generous sincerity, congratulated Tessier on his great operation. Whereupon the quiet Tessier, somewhat out of character, said:

Reed, if you think _that_ was good, come let me show you something even better!

He led Dingman to his locker and brought out a giant elephant tusk exclaiming,

What do you think of that one? I have looked up your tusk record, Reed, and this is larger than any of yours!

and they all had a good laugh.

John Marquis Converse of the New York University Medical Center was asked what he considered to be his contribution to the operation for hypertelorism and he responded by recalling that Tessier, in his first stage, opened the cranial fossa and cut through the olfactory nerves placing a dermal graft over the frontal lobe and, in the second stage, proceeded with the necessary orbital osteotomies. He summarized his 1970 work with J. Ransohoff, E. S. Mathews, B. Smith and A. Molenar with:

In checking over the x-rays of all of our cases, I noticed that the cribriform was not enlarged and conceived the idea of preserving it, thus also preserving the sense of olfaction (and of taste). We were able to develop the one-stage procedure which is now generally employed.

**CRANIUM BIFIDUM OCCULTUM**

Median cleft lip may be associated with midline clefts of the cranium often involving the scalp. According to J. J. Longacre in 1964:

In the newborn, the management is conservative, and the patient is placed under continuous observation. The necessity for operation on the larger
lesions overlying the superior sagittal sinus has been stressed by Peer and Van Duyn (1948) and Kahn and Olmedo (1950). These authors point out that if the lesion is more than 1 to 2 cm. in width it may become necrotic. Reconstruction with a scalp flap is recommended. In the larger lesions, the skull defect does not close spontaneously and grafting with bone (split-rib grafts) . . . is necessary at a later stage to provide an adequate protection to the brain.

**WAARDENBURG SYNDROME, OR MENDE’S SYNDROME**

In 1948 P. J. Waardenburg precisely described the syndrome of (1) congenital deafness, (2) lateral displacement of the medial canthi and lacrimal puncta with broad nasal root or telecanthus (pseudohypertelorism), (3) white forelock, or poliosis, (4) heterochromic irides and (5) hyperplasia of the medial portion of the eyebrows. In 1926 I. Mende also described the syndrome, as did Van der Hoeve in 1916 at least in part. The condition is inherited as an autosomal dominant characteristic and is seen in an estimated 2 percent of all congenitally deaf persons. R. J. Gorlin and J. J. Pindborg in 1964 noted that neither Mende nor Waardenburg had included lip clefts in the syndrome and mentioned that lack of clefts was also their experience. In 1961 A. Pirodda reported that cleft lip-palate and other palatal alterations were not uncommon in this syndrome. Then in 1965, independently, Feinberg, Hansen et al. and Puxeddu and in 1967 Reed et al. reported cases with cleft lip and/or palate.

According to Gorlin, Cervenka and Pruzansky, the syndrome described by Klein in 1950 was different from the Waardenburg syndrome. As they said,

Deafness, partial albinism, blepharophimosis and bony and muscle deformities of the shoulder girdle, in our opinion, represent another Syndrome.
59. Lateral Facial Clefts

**Horizontal or transverse clefts** are considered the result of failure of mesoderm migration or merging to obliterate the embryonic grooves between the maxillary and mandibular prominences.

**Transverse Clefts**

As these clefts are rare and almost everybody having one has reported it, it is possible to review most of the reported cases and
when described note the treatment. After specific case recordings in what may seem a helter-skelter arrangement, generalizations may be of value.

In 1891 Rose noted:

For long the very existence of this macrostomatous deformity was doubted, but cases have been recognized more or less since 1715, when Muralt pictured it for the first time.

One of the first cases reported was by Vrolik, who, in his 1849 work, gave several illustrations of commissural clefts as well as other deformities of the face. Other cases were reported by Reissmann in 1869 and Morgan in 1882.

Macrostomia or commissural harelip, according to Rose, is evidenced by an increased diameter of the mouth which may vary from a slight increase to a considerable distance; in a case reported by Rynd in 1862 the mouth opening extended as far as the first molar on the right side and to the last molar on the left. In 1887 Sutton published the drawing of a child with a very large cleft, the angles of which gradually passed into a red cicatrix. This scar ended in a gaping wound over the temporal region, extending to the dura mater.

Rose also pointed out:

Macrostoma is not only attended by great disfigurement, but is also troublesome from the impossibility of the child retaining its saliva and the food escaping during mastication. Suckling can be performed if the nurse’s nipple be long, but is difficult otherwise. This deformity is, perhaps, more frequently associated with defective cerebral power than any other of the facial clefts, a large proportion of the subjects having been idiots.

In 1862 M. Debout first noted the association of macrostomia with abnormal conditions of the external ear—either defective development or the production of accessory auricles. In 1875 Ahfield reported a transverse cleft of the mouth which included the ear. In 1886 Roulland published an account of a double macrostomia with accessory auricular appendages, absence of middle ear and eustachian tube and absence of the temporomaxillary joint on the left side. Macrostomia with auricular appendages, shown as a sketch, was presented by one of Fergus-
son’s patients. Then, in 1895 Ballantyne enumerated 16 cases of macrostomia with preauricular appendages. In 1909 Edington reported a transverse oral cleft that stopped in front of the tragus but with a fissure extending to the external auditory meatus. Sir Arthur Keith in 1920 recorded a case of transverse facial cleft extending to the tragus and external auditory canal, which he reported in 1940. As did many plastic surgery textbooks, Padgett and Stephensen in 1948 reported a transverse facial cleft associated with anomalous pretragal tags. Then in 1937 McEnery and Brennemann presented a case of macrostomia which coexisted with a nasomaxillary cleft. This is a real mix-up, with disturbance of the first branchial cleft and arch and persistence of the naso-optic groove, and enough to give any embryologist a "splitting" headache.

In 1950 Blackfield and Wilde of the University of California reported five cases. One was a three-month-old male with bilateral clefts extending from the corners of the mouth backward and above the ears. Absence of the terminal phalanges of the left hand, syndactyly of the left foot and absence of the right great toe were additional deformities. In the other four transverse facial clefts, one had a sinus of the dorsum of the nose, and two were associated with ear deformities. Treatment: Clefts closed and grooves excised and subsequently correction of syndactyly and excision of sinus or appendages.

Others reporting transverse facial clefts were Sanvenero-Rosselli in 1958, Phoner in 1958 and Piotti in 1958. In 1964 Gorlin and Pindborg surmised that transverse facial clefts seem to appear more commonly in males and when unilateral were most often on the left. In 1965 Fogh-Andersen of Copenhagen reported 13 transverse facial clefts out of his 3,988 clefts.

In 1968 Powell and Jenkins of Chicago reported three cases. A white male had a transverse facial cleft, preauricular skin tags, dermoid cyst of the conjunctiva and thoracic hemivertebrae, a condition they diagnosed as oculoauriculovertebral dysplasia or Goldenhar’s syndrome. The other two patients were white females, one with bilateral transverse clefts and the other with unilateral clefts associated with preauricular skin tag and retroauricular dermoid.
Treatment: Early closure was carried out on the transverse clefts. The bilateral clefts were closed with reconstruction of the oral angles with small flaps of vermilion in a modified Estlander technique. Subsequently the cysts, dermoid and skin tags were excised.

In 1970 Eiseman, Walden, Platzer and Hoppe reported a 13-year-old Negro female with bilateral transverse facial clefts associated with maxillary protrusion. Treatment: Closure of the clefts with Z-plasties followed by extraction of strategic teeth for aid in the osteotomy and setback of the maxilla.

Blackfield and Wilde admitted in 1950 that the etiology of these clefts is unestablished and then devoted pages to the possible causes. The most generally accepted explanation was the failure of fusion between mandibular and maxillary processes. Powell and Jenkins noted:

One theory on the genesis of lateral clefts is failure of the mesoderm to penetrate completely the regions of the epithelial fusion at the oral commissures, or they may be the result of absolute deficiencies in quantities of mesoderm.

According to Streeter:

There is no evidence of ectodermal resorption between the meeting surfaces, but rather the surface is simply flattened out by the proliferation of the growth centers beneath.

Blackfield and Wilde also leveled a truly wild “shotgun blast” at all possible etiological factors involved in cleft formation. Such causes were mentioned as Finley and Keith’s ideas of placental infarction, Von Winckel’s amniotic bands, Streeter’s amniotic adhesions, faulty implantation with poor nutrition, diabetic mothers, vitamin deficiencies (vitamin E), rubella infection during pregnancy and radiation. They concluded that the etiology of transverse facial clefts is probably a combination of several of the above-mentioned factors:

It would be well to stop thinking of heredity as a cause of disease and to consider it merely the pattern of incidence of a condition brought about by one or more factors.
MANDIBLE INCLUDED

In addition to transverse facial clefts and ear deformities, there is often an associated mandibular malformation. This grouping was first reported by Kirmisson in 1902. In 1938 British oral surgeon Martin Rushton reported a transverse facial cleft associated with maldevelopment of the mandible. T. P. Kilner suggested for this case closure of the cleft and at the same time a bone graft onlay to the mandible.

In 1939 Kazanjian of Boston reported five cases of congenital absence of the mandible and noted that two of them had macrostomia and four had microtia. He observed that, although this anomaly seemed to be the expression of improper development of the first branchial arch and cleft, as other facial and cranial bone anomalies appeared, they may have been secondary to the absence of the mandibular segment, which deprived the surrounding tissue of normal growth.

Treatment: He advised closure of the oral fissure and reconstruction of the ear only. Surgery of the mandibular defect, however, he postponed until the permanent teeth were available to allow fixation during tibial bone grafting.

In 1950 Michael Lewin reported a case of mandibular malformation associated with an anomaly of the ear and an associated groove-like thinning of the cheek extending from the corner of the mouth to an auricular appendage. In 1955 Hunt and Smith also reported a case of oral-mandibular and auricular deformities.

Lateral facial clefts can be associated with what Gorlin and Pindborg referred to as hemifacial microstomia (microtia, macrostomia and failure of formation of the mandibular ramus and condyle). They suggested that oculoauriculo-vertebral dysplasia or the Goldenhar syndrome is a variant of this complex characterized by vertebral anomalies, most often hemivertebrae and epibulbar dermoids of the eye.

In April 1961 Ken McNeill of Kingston, Jamaica, and I set off on a West Indian plastic peregrination, having cabled ahead for the local doctors to collect all of their worst faces. Among our "touchdowns" was the island of Barbados, "a little England"
with more sunshine, more British color and charm and more people and sugar cane per square mile (21 × 14 mi.) than any other isle of the Antilles. Here, Jack Leacock was waiting for us with a rare macrostomia extending across the cheek with a groove passing above the slightly deformed ear. There were an associated seventh nerve paralysis and a gelatinous cyst in the cleft between the maxilla and the hypoplastic mandible. First the cyst was excised. Then the edges of the cleft were freed to facilitate a two-layer closure interrupted by a Z-plasty of the skin and a vermilion flap from the cleft to turn the corner of the new commissure, as published in the *British Journal of Plastic Surgery* in 1962. Because of the happy atmosphere on this sunny island, both the main wing of the Z flap and the mucosal commissure flap were turned upward in an attempt to suggest a smile in the presence of facial paralysis.

In 1961 J. J. Longacre of Cincinnati, with DeStefano and Holmstrand, also reported a case of oral, mandibular and auricular deformities. They advised Z-plasty of the cleft closure, and Longacre emphasized the importance of early split-rib autografts to the mandible to minimize the psychological trauma.

In 1962 Hans May of Philadelphia reported three cases of transverse facial clefts associated with ear and mandibular anomalies along with other defects. His treatment was of interest. One case of transverse facial cleft was accompanied by a rudimentary left ear in the middle of the cheek, coloboma of the left upper eyelid, dermoid cyst of the conjunctiva, coloboma of the iris, absence of the malar arch and maldevelopment of the mandible. This is getting close to a description of the so-called Treacher Collins syndrome, which, not having clefts, will be bypassed. May closed the transverse cleft directly, and when the scar developed hypertrophy he excised it and used a Z-plasty with what he reported was a success.

Another of his patients was a male with a transverse facial cleft with underdeveloped mandible and forward displacement of the tragus. He closed the cleft and shifted the tragus posteriorly.

His third patient was a female with a transverse facial cleft and underdevelopment of the mandible and ear. In addition to ear
revision and mandibular lengthening, May closed the cleft and reconstructed the commissure.

His summary was important:

Transverse facial clefts seldom constitute an isolated deformity. As a rule, they are accompanied by deformities of the mandible, the ear, and perhaps various other facial deformities, all centering around the first (mandibular) branchial arch. . . . These clefts require closure since the orbicularis oris muscle is divided. Closure by simple fusion of the separated structures overcomes only part of the deformity. The key point in the repair must center upon the reconstruction of a muscular commissure, the closure of the orbicularis ring at the cleft side. This can be done effectively by rotation of a full-thickness vermilion-lined flap from the lower into the upper lip after Estlander.

In 1963 Nagai and Weinstein also advocated the use of a modified Estlander-type flap for reconstruction of the oral commissure in transverse clefts.

Side by side with Hans May in the March 1962 Plastic and Reconstructive Surgery was a paper by Richard Stark and David Saunders officially grouping the combined oral-mandibular-auricular anomalies into the first branchial syndrome. They reported five cases of this combination and stated:

Macrostomia, hemignathia and auricular deformities are intimately related congenital anomalies. All are rare but they may coexist, representing a syndrome.

They explained the association:

Embryologically, the mandible develops from the mesoderm of the first branchial (mandibular) arch. A portion of the auricle (tragus and helical crus) also is derived from this mesoderm. Normal development of the mouth depends upon the amount of mesoderm of this arch and the extent of its migration.

Treatment: Their cheiloplasty included paring of the cleft edges and closure in layers. Insurance toward preservation of the corner of the mouth was achieved by mucosal flaps: either a triangular one that fits directly into the commissure or a mucosal flap taken from the upper or lower vermilion and used as a "wraparound" at the corner which shifts the scar out of the
commissure itself. Excision of preauricular tags, ear reconstruction and mandibular bone grafting were mentioned briefly.

**LANDMARKS FOR THE COMMISSURE**

In the *British Journal of Plastic Surgery* in 1968 Khoo Boo-Chai of Singapore reported four cases of transverse facial cleft in which the cleft was the presenting complaint in a constellation of other deformities seen in the first and second branchial arch syndrome. William Grabb in 1965 published a classical review of the clinical aspects of this syndrome. For the treatment of his four cases Boo-Chai gave an excellent outline of the essential points in the surgical correction (one case is shown). He wisely noted:

The rim of the cleft is lined by vermilion which is of a slightly lighter shade than that of the normal vermilion. The line of demarcation which we were able to detect in all our four cases marks the beginning of the cleft. Pressure with the examining finger in this region will help to bring out a muscular ridge which corresponds with the line of demarcation in the vermilion. We place importance on this landmark because we believe it to be the correct position of the new commissure. . . . Once this is accurately identified and marked, the vermilion of the rim is excised together with the triangular piece of skin on the buccal surface of the cheek. Beginning intra-orally at the apex of the triangular raw area, the edges of the mucosa are closed with 4-0 plain interrupted catgut working outward toward the new commissure . . . the cut through the vermilion at the site of the new commissure is not vertical but it is made slightly oblique from within out . . . The oral musculature is then apposed on the outside . . . It is important to get the muscle together as close to the commissure as possible. Otherwise, we will get a "goldfish mouth" appearance. A "Z" plasty for the skin completes the operation.

**DOUBLE Z**

O. T. Mansfield and D. C. Herbert of England noted in 1972 that unilateral transverse facial clefts are associated with facial hypoplasia and proposed introduction of additional tissue into the plane of the cleft to make up the deficiency. For two cases they designed a double Z-plasty along the line of the cleft from the
commissure to the tragus: a small Z at the commissure and a large Z with thick flaps in the cheek area which they claim not only increased the length of the scar but improved the contour.

According to Tord Skoog of Sweden in his 1974 book *Plastic Surgery*:

Macrostomia should be recognized as a malformation involving several tissue layers. The external defect is always combined with a more extensive separation of the deep tissues of mesodermal origin, i.e., the oral and buccal musculature.

He presented an example of a minimal lateral facial cleft possessing preauricular tabs characteristic of congenital malformations involving the first and second branchial arches. William Grabb noted in 1965 that a distinctive tag just anterior to the junction of the tragus and antitragus was almost always associated with macrostomia.

Skoog's reconstructive design was basic, using mucocutaneous incisions around the commissure defect for exposure, dissecting both ends of the orbicularis oris muscle, freeing the buccinator and then closing in three layers. The oral mucosa was sutured in a straight line and the orbicularis oris muscle stumps were approximated with 4-0 Dexon sutures to construct the sling around the commissure.

The buccinator was sutured with 5-0 catgut and the skin closed in a transverse lateral line except for a small skin interdigation near the commissure. This technique indeed seems to be a sound and effective approach to this type of cleft.
TRANSVERSE CLEFTS IN BILATERAL FACIAL MACROSTOMIA

In October 1974 John M. Converse, Donald Wood-Smith, J. G. McCarthy, P. J. Coccaro and M. H. Becker of New York University Medical Center presented their 15-case experience with bilateral facial macrostomia. They divided this congenital syndrome into four groups, the fourth being

macrostomia, transverse facial clefts, absent parotid ducts and abnormalities of the auricles and facial skeleton.

They elaborated in Plastic and Reconstructive Surgery:

This group included 4 patients, whose auricles were small and low-set bilaterally. A full complement of auricular elements was present in each... One patient had preauricular pits and appendages, another showed atresia of both external auditory canals on temporal bone tomography. The remaining 3 patients had no hearing abnormalities.

Varying degrees of macrostomia and transverse facial clefting were the hallmark of this group. In two patients, subcutaneous cleft-like mesodermal deficiencies extended across the buccal region in a transverse direction toward the external auditory meatus. In two other patients, the cleft involved full thickness of the cheek—in one patient extending approximately two cm. from the oral commissure—in another patient involving the full width of the cheeks as far as the tragus. ... The patient with the most pronounced transverse facial clefts also had a wide cleft of the hard and soft palate. Two patients, studied radiographically, also showed extramaxillary foci of bone and ectopic dentition in the region of the pterygoid processes and maxillary tuberosities. In both of these patients the mandibular excursions were restricted, but they improved after intraoral excision of the extra-alveolar segment of ectopic dentition. All 4 patients had pronounced micrognathia and some degree of apertognathia (open bite), with class II dental occlusal relationships.

Tomographic studies showed abnormalities of the mandibular condyles, ranging from absence to surface irregularities. Each ramus tended to be shortened, the ramus and mandibular body assuming a straight-line relationship. Two patients in this group had cervical spine abnormalities.

Their plan of treatment for this group involved

(1) restoration of mandibular size and form and (2) correction of soft tissue deficiency.
In the worse of the two cases shown, there had been multiple Z-plasties of the transverse facial clefts in infancy. Further surgery included the following:

(1) An increase in the anteroposterior dimensions of the right mandibular ramus was obtained by vertical section and iliac bone grafting; a sagittal osteotomy was done on the left ramus. (2) An intraoral skin graft was inlaid to remedy the soft tissue deficiency and distend the premental region. (3) A step osteotomy and iliac bone grafting was used to elongate the body of the mandible [as diagramed].

In spite of the zigzagging cheek scars, the result shown was excellent.

**SUMMARY ON CLEFT CLOSURE**

Transverse facial clefts are associated with multiple anomalies, and the actual cleft is often the least of the problems encountered. Early closure of the cleft in layers is important to the patient’s ability to feed. As the cleft transgresses the natural lines of the face, the scar of its closures must also. For this reason and for the added precaution against straight-line contracture by interruption, some form of a Z-plasty can be valuable. It is always happier to turn the main flap upward when possible, as seen on the Barbadian baby.

The other important aspect of transverse clefts, of course, is the reconstruction of the commissure. Various mucosal flap interruptions of the scar at the commissure are useful, but May’s use of a small Estlander flap to reconstitute the muscle at the corner of the mouth and, even better, Skoog’s reconstruction of the orbicularis oris musculature at the commissure seem sound.
The first report of an oblique facial cleft was made in 1732 by Von Kulmus, and it is not surprising that he recorded it in Latin. Then in 1828 Delpech presented such a case in French, and in 1832 Walter Dick of Glasgow reported one in English.

William Rose of London in 1891 acknowledged his teacher, Sir William Fergusson, as the only English surgeon, as far as he knew, who had observed the rare facial clefts, the majority of recorded cases hailing from Germany and France.

Rose noted that several cases occurred with red cicatrices rather than actual clefts and cited this case (Tillmanns after Kraske) of an oblique cicatricular deformity of the face along the line usually transversed by such a cleft.

He also cited a facial cleft in a child, involving the lower lid and eye and with a development of accessory teeth along the cleft margin (Tillmanns after Haselmann).

Rose wrote of these cicatrices and clefts:

The defect begins at the free margin of the upper lip, and usually at the spot whence starts the ordinary harelip cleft; but occasionally from the angle of the mouth. It then trends upwards and outwards, leaving the nose entire, and skirts round the ala nasi to reach its upper limit at the middle of the lower eyelid which is cleft, or at the inner canthus. The eye itself may show a coloboma iridis, usually downwards and inwards. The facial skeleton may be divided or not; sometimes a large opening into the antrum exists.

Rose noted that the deformity could be bilateral and cited the case of Guersant. He also acknowledged:
Albrecht recorded a most interesting case in a newly born pup of double cleft extending from the lip margin upwards not only into the nostril but also towards the eye on either side, i.e. a double associated harelip and facial cleft. The specimen is taken from the Royal Veterinary College of Brussels.

M. Guersant, in the middle of the nineteenth century, reported a case of bilateral oro-ocular clefts of the face and illustrated the condition. Although the sketch portrays the cleft involving the entire lower eyelid, this probably was a medial type of oro-ocular cleft.

In 1887 Morian collected 34 cases in the literature, 77 percent of which were stillborn. In 1970 Khoo Boo-Chai, reviewing the literature since Morian, collected 43 cases of live-born oblique facial clefts including two of his own. Tessier in 1969 reported three; Bartels, O'Malley, Baker and Douglas in 1971, two; and Wilson, Musgrave, Garrett and Conklin in 1972, seven. According to Wilson's calculations, this rare anomaly represents approximately 0.25 percent of all facial clefts. Then in 1973 Dey of Sydney reported another five oblique facial clefts, and Miller, Wood and Hag reported one case from Nairobi. Also added is a case of Tom Zaydon's, who was my patient for a short time but is now in Bethesda. And, of course, there are an unknown number of patients around the world, like this Jamaican, who are operated on by the local surgeon and never get included in any published statistics.

According to the Nomenclature Committee of the American Association for Cleft Palate Rehabilitation, there are two main forms of oblique facial clefts: the naso-ocular and the oro-ocular. The oro-ocular type is subdivided into medial and lateral, depending on the relationship of the cleft to the infraorbital foramen. Adding to the complexity is the mixed occurrence of these various clefts in the same patient.

Numerous fascinating combinations can occur. Burian had a case with a naso-ocular cleft on one side and an oro-ocular on the other. Dey of Australia also had one case of a boy with both a
naso-ocular and an oro-ocular cleft. Greer-Walker and Skoog each had cases with both subgroups (medial and lateral) of the oro-ocular cleft. The oblique facial cleft has been associated with a transverse facial cleft in three reported cases—two on the contralateral side, one recorded by Lexer and one by Burian, and one on the ipsilateral side, recorded by McEnery and Brennemann.

Persistence of ectodermal grooves due to retardation in normal mesodermal migration could explain all oblique clefts except that, as Karfik pointed out, the lateral oro-ocular cleft corresponds to no embryonic facial groove. These clefts do not seem to be familial. Wilson, Musgrave, Garrett and Conklin put the blame vaguely on “some environmental insult to the developing embryo,” backing this proposal with:

It is not surprising therefore to find multiple anomalies in many of these patients and a high incidence of still birth.

David Dey favors Ida Mann’s thesis that the nasolacrimal ducts and apparatus are derived from ectoderm cut off from the surface by a forward growth of the maxillary process, which reaches the inner and outer nasal folds. The naso-ocular group of clefts follows the line of this epithelial inclusion, which at all times maintains connections with the nasal pits, extending any cleft of the lip that occurs in the usual position. The oro-ocular cleft, however, has no connections with the nasal pit, has no correspondence with the site of the ordinary cleft of the lip and may pass medial to the inferior punctum. Dey suggests that Sir Arthur Keith’s theory of linear intrauterine necrosis along the watershed between developing vascular areas seems apt. Sanvenero-Rosselli in 1953 blamed the arterial system for congenital anomalies and quoted Kundrat in ancient writings as having found arhinencephaly to be due to a disturbance of the whole region supplied by the anterior cerebral artery.
Naso-ocular clefts are considered the result of failure of mesoderm migration or merging to obliterate the embryonic grooves between the nasomedial, nasolateral and maxillary prominences each with the other.

The naso-ocular cleft extends from the pyriform aperture to the medial canthal area along the approximate course of the nasolacrimal duct. As the nasolacrimal duct is intact only in the mild cases, it is usually absent or opened.

Daniel Marchac of Paris forwarded photographs of a naso-ocular cleft from an 1828 book by Professor Delpech of Montpellier. A short excerpt translated from the original French text is of interest.

In the month of April, 1820, we met in the streets of Montpellier, a young boy 12 years old with a singularly severe congenital deformity.

With what is considered to be the first frontal flap used in France, a hemirhinoplasty was accomplished. It was impressive, to say the
least, that an early nineteenth-century surgeon had the sophistication to call upon the Indian forehead rhinoplasty to correct both nasal and ocular defects of an oblique facial cleft.

In 1962 C. S. Harkins, with A. Berlin, R. L. Harding, J. J. Longacre and R. M. Snodgrasse, defined a naso-ocular cleft as a fissure extending from the narial region toward the medial angle of the palpebral fissure, differentiating it, as Bartels noted, from the oro-ocular cleft, which extends from the mouth to the orbit without involving the nose.

In 1966 Sakurai, Mitchell and Holmes noted that the fissure need not involve the entire line from the lip through the nose to the eyelid orbit with extension to the temporal region. Boo-Chai distinguished between complete and incomplete forms, noting that complete clefts are rare, usually seen in stillborn monsters, often associated with cleft lip and palate. When the bone is involved, there is hypoplasia of the body of the maxilla and only very seldom a complete cleft. When there is a split in the bone, it occurs between the medial and lateral incisors with disruption of the pyriform aperture and extension of the cleft into the orbit, as described by Burian in 1963 and Ergin in 1966.

In 1963 G. S. Gunter of the Royal Children's Hospital, Melbourne, referred to this anomaly as the nasomaxillary cleft and attributed it to the persistence of the naso-optic groove between the maxillary and frontonasal processes. He made several observations:
When the cleft appears to occupy the same portion of the primary palate as does the usual cleft lip, it then involves the nostril floor and passes on both sides of the displaced alar base. At the upper end there is involvement of the inner canthus with an upward extension across the brow and forehead.

Gunter was the first to note the various strange disruptions and extensions of the hairline and the hairs of the eyebrows.

Treatment is difficult and calls upon every facet of reconstructive surgery. Gunter did make some specific suggestions. In his experience, blocked nasolacrimal duct and repeated infection of the sac occurred in all cases, prompting him to advocate total extirpation of the sac and duct. He also noted that the normal eye must have early and adequate lid reconstruction. When additional tissue is required in the cleft of the lip and face, forehead and Abbe flaps can be used. The nose on the cleft side is always short between the alar base and the inner canthus; if the discrepancy is great enough, new soft tissue should be introduced. Gunter acknowledged the difficulty of reconstruction in asymmetries of the skull and face. He stressed the importance of having the afflicted children accepted within their family circle and not allowing them to become lost socially and educationally in the multiplicity of surgical procedures.

Gunter, who has added to his interests the breeding of Aberdeen Angus cows and Holsteiner horses, was asked for his current thoughts on naso-ocular clefts. He answered in December 1973:

I used to call this cleft “naso-maxillary” because it appears to lie between those structures. . . . However this is not always strictly true . . . nor does the cleft always lie in the naso-optic (naso-ocular) groove, to use the other terminology, and I wonder if the much older concept of some mechanical band, possibly amniotic, being responsible might not still be worthy of consideration. I've had personal dealings with one more case since my paper appeared in 1963 . . . This child also had anomalies of the hands of the "congenital ring stricture" type. . . .
This child has had three operations to date. 1. Repair of lip and anterior palate. 2. Z-plasty to left side of nose and inner canthus to lengthen nose on that side and bring the canthus up to a correct level, and 3. Repair of the cleft palate proper.

The complexity of these deformities seems to require many operations, as reported by all involved surgeons.

In April 1971 Roger Bartels, Joseph O’Malley, James Baker and William Douglas, in a paper “Naso-Ocular Clefts” in Plastic and Reconstructive Surgery, presented two interesting cases. In 1974 Bartels was questioned about these cases and he kindly sent recent details of their follow-up.

Case 1 was a six-pound Caucasian female with no history of clefts in the family but a record of maternal virus cold in the first trimester. Patient had left naso-ocular cleft, coloboma of the lower eyelid, left anophthalmia and bilateral complete cleft lip and palate. Treatment by O’Malley had included the following staged procedures: (1) right cheilorrhaphy, (2) closure of the naso-ocular cleft with left cheilorrhaphy and a vomer resection, (3) staphylorrhaphy with vomer flaps, (4) Wardill pushback palatoplasty, (5) left alarplasty, (6) left orbitoplasty, (7) composite graft from the right ear to the columella.
At 10 years of age she was reported of normal intelligence, doing well in school, a happy well-adjusted child.

Bartels' recent communication noted:

Since the article, Dr. O'Malley performed two additional surgical procedures on this patient using a midline forehead flap to construct a lower lid for this unfortunate girl. As you can see from her photographs, the left eye prosthesis was displaced downward to a considerable degree and this lower lid reconstructive procedure was done in an attempt to give her a more normal appearance. I have not seen her since her last photograph but obviously she needs additional work.

Case 2 was a seven-pound Caucasian female with no history of clefts in the family but a record of maternal viral infection during the first trimester of pregnancy. The patient had a right incomplete naso-ocular cleft with a coloboma of the right lower eyelid, coloboma of the right iris, left anophthalmia, slightly enlarged cranial vault with separation of the suture lines and tense frontanelles, hemangioma of the forehead, bilateral cleft lip and palate, diastasis of the rectus abdominis, abnormal hand lines and hypertelorism. Other congenital anomalies included an extra cervical vertebra and a grossly abnormal brain as revealed by encephalography. A ventricular-peritoneal shunt had to be performed to decrease intracranial pressure.

Bartels carried out two surgical procedures, in the first closing the naso-ocular cleft and the bilateral lip clefts and in the second closing the coloboma of the right lower eyelid and releasing the short ala on the right with a large auricular composite graft.
Bartels' recent report follows:

The child's postoperative care was provided by the physicians at Sunland Hospital here in Orlando and when I returned at a later date for postoperative photographs, I was informed that the child had died of meningitis, apparently related to her ventriculo-peritoneal shunt.

In 1971 Bartels had noted 11 cases of congenital naso-ocular clefts in the world literature, to which he added two. In 1974 he stated:

I personally feel that a naso-ocular cleft is a result of failure of fusion between the lateral nasal process and the maxillary process. To the best of my recollection every case of naso-ocular cleft reported also had a cleft of the primary palate, the only exception being the case report by Julio Ortega, appearing in Volume 43 of Plastic and Reconstructive Surgery.

In June 1969 in Plastic and Reconstructive Surgery Julio Ortega and Enzo Flor of Luis Vernaza Hospital, Guayaquil, Ecuador, reported a rare case of a 16-year-old country girl with an incomplete naso-ocular cleft with no history of clefts in the family and no incidents during the first trimester of her mother's pregnancy. Ortega considered the deformity an underdevelopment rather than an absence of elements, which was consistent with the absence or abnormal development of a specific portion of the mesodermal mass in the naso-optic groove. The deformity included the following conditions: moderate hypoplasia of the orbital and maxillary region, inner canthus of the left eye 15 mm. lower than the right, lacrimal apparatus present but
underdeveloped and abnormally located, lacrimal sac on the floor of the orbit, epiphora present, incomplete bony nasal process of the maxilla consisting mostly of cartilage with a 2 mm. cleft between it and the nasal bone, partial lack of left nasal ala, underdeveloped lateral crura and especially the superior lateral cartilage and hypoplasia of the chin with defective dental occlusion. This was the sequence of surgical procedures:

1. Rhinoplasty to reduce the size of the nose and the size of the alar defect.
2. Silicone implant to chin.
3. Reconstruction of alar defect (two stages).
4. Z-plasty to the left canthus.
5. Dental prosthesis.

The alar correction by logical local flaps in two stages was well executed.

The complex surgical problem offered by these clefts is exemplified again in the four cases reported in 1972 by Wilson, Musgrave, Garrett and Conklin, who stated:

Tissue defects present may be so extensive as to preclude truly satisfying aesthetic results . . . and can be expected to include multiple staged operations. . . . The only generalization possible is that an exposed eye requires immediate treatment.

One of their cases had had nine operations elsewhere and received another nine operations. Their other three cases averaged nine
operations each, including the usual cleft lip and palate procedures and eyelid, cheek and nasal reconstruction. In spite of all this surgery, the cases were considered still unfinished, indicating the severity of the problem.

**DEY ADDS FOUR MORE TO THE LITERATURE**

Dey reported four naso-ocular cleft cases. One was a bilateral naso-ocular cleft, far worse on the left side, where a small skin tube connected the nostril margin to the eyebrow region. There were associated anomalies such as cleft palate, absence of great toes, syndactyly of the second and third toes on the left side, constricting ring on the left ring finger and left little finger and congenital amputations of the right index and middle fingers. The intelligence was good and social adjustment satisfactory.

The second was a boy with almost complete right-sided oro-ocular cleft and complete left-sided naso-ocular cleft, left cleft alveolus but intact palate. There was also a bilateral posterior choanal atresia. The patient developed normally and has done well in school with a pleasant personality.

The third case had complete naso-ocular clefts on both sides with hypertelorism and blindness (right anophthalmia, left microphthalmia).

The fourth patient was a baby boy with bilateral cleft lip, plus cleft palate. On the left side the cleft was naso-ocular, and on both sides well-marked grooves extended the cleft high onto the forehead with the hair showing associated “cowlick” on the left side.

The first three of these had been treated prior to their coming to Dey, and the last is a patient of George Gunter.

**ORO-OCULAR CLEFTS**

In oro-ocular clefts the fissure extends from the mouth to either the medial or the lateral canthus leaving the pyriform aperture intact. The subgroup, medial or lateral, of the oro-ocular cleft is
Medial oro-ocular clefts are considered the result of failure of mesoderm migration or merging to obliterate the embryonic grooves between the nasolateral or nasomedial prominences and the maxillary prominences, the nasomedial and nasolateral prominences having merged with each other successfully.

determined by the cleft’s position in relation to the infraorbital foramen. These clefts can occur in complete and incomplete forms. Mild incomplete oro-ocular clefts can be confused with mild incomplete lip clefts and can be distinguished from them by two main characteristics:

1. The cleft lies lateral to the peak of the cupid’s bow rather than through it, as in the standard cleft.
2. Because of shortening of the soft tissue element on the affected side there is an upward tilt of the alar base instead of the usual downward flare.

THE MEDIAL ORO-OCULAR CLEFT

Medial oro-ocular clefts are considered the result of failure of mesoderm migration or merging to obliterate the embryonic grooves between the nasolateral or nasomedial prominences and the maxillary prominences, the nasomedial and nasolateral prominences having merged with each other successfully.
The cleft lies medial to the infraorbital foramen and, instead of involving the nose, bypasses it, running upward in the region of the nasolabial (cheek) groove to terminate in the inner canthus of the lower eyelid. This fissure may extend up into the forehead, usually in the temporal region, and when the bone is cleft, the split lies between the lateral incisor and the canine.

Although the nose is well formed, in unilateral cases it is usually rotated around its long axis; in bilateral cases the bony and cartilaginous nose is detached from its lateral bony segments, drawn upward with forward protrusion. The orbit is sometimes shifted downward and is capacious because of the irregular and deficient growth of its walls. According to Rogalski, the eyeball may be deformed.

In 1935 Warren B. Davis found, out of a series of 1,000 clefts, four oblique clefts with absent nasolacrimal duct and five with coloboma extending into the facial cleft. He published a bilateral example of the medial group of the oro-ocular cleft and described the deformity:

The clefts involve the lips, cheeks, lower eyelids, alveolar processes, anteromedial portions of the maxillae and the orbital floors. Posterior to the premaxilla, the palate is intact. Atresia of the posterior one-third of the nasal passages was from a thick mass of tissue, partly osseous. Note the rotation of premaxilla, and the elevation of all anterior nasal structures, which, in association with the prolapse of the eyeballs, placed the anterior nares and the pupils of the eyes on the same horizontal plane.

Harry P. Ritchie spied this strange cleft of Davis' and included it in his surgical interpretation of embryology in 1934:

This case is particularly important for my purpose, as it shows the nose, prolabium and premaxilla normally formed in the frontonasal process. This process is shown to be an embryonal entity, separate from the lateral maxillary processes.

In 1950 John Potter of Newcastle reported a bilateral oblique facial cleft extending from the medial end of the lower eyelid to the lateral side of the premaxilla. There was also a complete cleft of the lip and alveolar margin, but this passed lateral to the nose. The central part of the face protruded at a level much higher than
normal with the nasal tip on a line with the eye. The nasal airways were normal and remained so. The nose was shorter than normal. There was a complete bilateral cleft of the palate. The lacrimal system was grossly abnormal, the inner canthus being unformed and caruncle and puncta absent, with notching of the upper lid. There was no lacrimal sac, and the nasolacrimal duct was represented by an open cleft covered by pink epithelium. The eyes were normal and moved normally.

Potter noted:

On each side the cleft involved superficial parts of the soft tissues only at its upper end, deepening as it descends so as to involve the full thickness of the lip, and being complete in the alveolus and palate.

This case seems to illustrate a view expressed by Frazer in 1939:

It must be understood that the cleft is only present where the maxillary process applies itself, in its growth to the surfaces of the nasal folds. Further back it is never present, the maxillary mesoderm being applied directly to the paraxial mesoderm without any intervention of ectoderm.

Treatment: W. E. M. Wardill closed the left cleft at six weeks of age. He used Veau's palate procedure, suturing the mobilized hard palate flap to the vomerine mucosa flap. He pared the edges of the lip cleft and closed with sutures after wide undermining.

Six weeks later John Potter closed the right palate cleft in the same manner but used a modified Blair-Brown-Mirault procedure for the lip. Six weeks later he revised Wardill's lip on the left with the Blair-Brown method. The palate was closed at age 12 months by Potter, using Wardill's V-Y pushback and pharyngoplasty. Then, at age two years, Potter freed the depressed inner end of each lower eyelid and transposed a flap from each upper lid into the defect to correct the inner droop.

This interesting case was published in the *British Journal of Plastic Surgery* in October 1950 with records of the patient up to two years of age.

Recently I wrote my friend Potter for more up-to-date records and he obtained photographs from Newcastle of the patient in 1967 at the age of about 20 years. Although further surgery has been carried out since, these photographs are revealing. The
eyelid construction had been quite satisfactory. Potter, true to his old chief Wardill’s dictum “Follow up a few cases well and carefully and keep trying,” wrote almost in anguish:

You can see the problem—the premaxilla has grown again in his teens.

It seems that the frontonasal component, being more or less detached from the lateral segments in relation to mesoderm and consequently muscle and bone, has failed in its vertical descent and has continued in its forward growth. Thus this final result has occurred without the benefit of the downward pull of normal maxillary attachments or the constricting restraints of the intact orbicularis oris muscle. At least here is an honest and true follow-up on this rare type of cleft that shows what will happen under certain conditions; it should give some direction toward treatment of the future.

Postponement of early closure of the alveolar and hard palate clefts should prevent any reduction in growth that is caused by surgery but certainly will avoid locking in the short frontonasal component at its undescended position, which evidently is destined to be exaggerated by future growth. Possibly a controlled device as described by Georgiade and Latham could exert the necessary prolonged downward traction to encourage growth of this stunted segment and at the same time position better what-
ever is already present. Then, of course, joining the orbicularis oris muscles across the cleft will give further molding benefits.

Few clinics in the world ever see a bilateral medial oro-ocular cleft patient. For instance, Joachim Gabka with his great volume of cases in Berlin borrowed for his book one of these bilateral oblique clefts from the even larger collection of Rosenthal.

In 1964 Gabka, in his book Hasenscharten und Wolfsrachen, diagramed his plan for treating a unilateral medial oro-ocular cleft. His design was simple inturning of the edges of the cleft for lining and rotation of a cheek flap for cover while aligning the lateral and medial vermilion of the lip.

Fogh-Andersen in 1965 reported three oblique facial clefts out of 3,988 clefts. One was a severe oblique cleft combined with bilateral cleft lip and palate, nasal defect and preauricular appendages. He also published an account of a less severe incomplete oblique cleft of the lip involving the medial portion of the lower eyelid. His surgery corrected the lip and cheek with a Z-plasty.

Paul Tessier of Paris, who seems to gravitate to facial bone pathology, especially in the orbital area, in 1969 reported on 16 coloboma patients with 22 facial clefts. He differentiated between two types of medial oro-ocular clefts, vertical and oblique.

He noted differential features. In the eyelids, localization of the cleft seems to be outside the punctum lacrimale in vertical clefts and inside in the oblique cleft. The medial canthal ligament is almost normal in direction and insertion in vertical clefts but atrophic, obliquely directed and associated with ectopia in oblique clefts.
The lacrimal sac and canal are absent in the oblique clefts—but in only three vertical clefts were they salvageable. There is an osseous cleft involving the floor of the orbit and the maxilla with deeper skeletal upheaval in the oblique clefts. The lower edge and the floor of the orbit are always separated by a gap which may vary in both depth and width but is situated inside the infraorbital foramen. The contents of the orbit sink into this fissure and at times reach the palate, causing prolapse of the eyeball. Vertical clefts pass into the maxilla via the sinus. In oblique clefts the internal wall of the maxillary sinus is absent. Skeletal clefts are usually located between the canine and the lateral incisor, although Tessier has also observed an accessory cleft between the central incisors.

In the nose the ala is normal but tilted up in vertical clefts with the distance between the internal canthus and the foot of the ala short. The same distance is extremely short in oblique clefts since the cleft itself occupies this general site. Then, too, the ala is unsupported with the bone cleft behind it, and the internal wall and the frontal process of the sinus are absent. In the vertical cleft the labial cleft lies outside the ala, not extending to form the usual cleft lip. The vertical-type cleft extends onto the lip as a standard cleft lip and may be accompanied by alar and labial fissures.

Treatment by Tessier is a combined operation consistent with his grand style. The design has a similar format for incomplete and complete oblique cleft correction but is carefully planned for the specific need of the case:

A multi-stage operation is replaced by simultaneous management of the eyelid, inner canthus, floor of orbit, cheek, lip and ala nasi. The point of novelty is this and one which I consider to be of the maximum importance. The treatment of coloboma must be carried out in a single stage because it is much easier to combine the different skin flaps, extensive cleavages, external cantholysis, inner canthopexy and multiple bone grafts.

Palpebral elongation demands total disinsertion of the lower septum and external cantholysis. As correct placement of the eyelid is opposed by the connection of the septum with the periosteum, this must be broken. Ectopia of the inner canthus,
more marked in oblique clefts, deserves transnasal inner canthoplasty in most cases. The maxillary cleft through the floor of the orbit allows the contents of the orbit to sink out of position, requiring correction and maintenance with bone grafts to establish continuity of the floor and edge of the orbit, plug the gap and improve the contour of the aplasia. As noted by Tessier, in oblique clefts the orbit, nose, sinus and mouth are in communication, not facilitating the success of bone grafts. He explains:

The grafts must be made to rest on the palatine plate and the outer edge of the alveolar arch (externally) and on the inner face of the orbit and the remains of the frontal apophysis (internally). Overhang is important and must be sustained above and externally by floor grafts. In spite of this, however, resorption may take place and secondary bone grafts may be required.

The ala nasi is always atrophied, and for the more severe hypoplasia a composite graft may be of value later. For the cleft lip Tessier suggests rotation and advancement:

We have obtained good results from Millard's mark-out for harelip in external labial cleft accompanying vertical cleft.

The striking shortness of the vertical distance between the canthus and the lip requires at least two interdigitations of skin flaps to reposition the eyelid and drop the ala nasi. Then, with the rotation-advancement, the cleft lip is corrected.

Six of Tessier's 16 cases had a bilateral coloboma in which there was considerable protrusion of the "central massif" exaggerated by gross hypoplasia of the malar bones and the cheeks. He accused this deformity of being a "devourer of bone," and the greater the malformation, the more pressing the need for bone grafts to reconstruct the normal contours of the face.

David Dey of Sydney reported a case of bilateral medial oro-ocular clefting, complete on the left, somewhat less than complete on the right. The eyes were exposed by proptosis and downward displacement of eyelids. Medial to the point of entry of the cleft no real lid margin was evident. As usual, the central upper lip, nose and nostrils appeared basically normal except shortened and displaced upward. A groove in the maxilla on both
sides extended from the alveolus at the canine tooth to the orbital margin. Bilateral choanal atresia was also present.

Dey's outline of treatment for this case included penetration of the atresia and establishment of airways by otolaryngologist B. Benjamin, rotation of the lower lids as flaps upward and medially to the inner canthi and inrolling of the edges of the complete cleft to form an artificial lacrimal duct. He described his lip closure:

Three months later, the cleft was repaired bilaterally—using a lateral cheek flap (somewhat reminiscent of the Millard advancement-rotation operation) combined with a triangular lip flap used Z-fashion. The flaps of excess vermilion tissue on the lateral margins of the clefts were used in the central lower border, where the vermilion was very narrow.

In 1973 Miller, Wood and Hag reported a case of bilateral medial oro-ocular clefting seen in Nairobi. The patient had a left complete medial oro-ocular cleft with coloboma and patent nasolacrimal duct. The left globe, covered by an epithelial membrane with "inadequate visual structures present for future sight," had complete range of motion. On the right there was a medial incomplete oro-ocular cleft with skin grooving up to the lower eyelid, anophthalmia with no nasolacrimal system, oblique cleft of the secondary palate and hypoplasia of the left malar eminence and maxillary sinus.

Treatment: Bilateral straight-line closure of each oro-ocular cleft was performed along with attempted closure of the coloboma, which had to be repeated.

In 1973 Poradowska, Jaworska, Dudkiewicz and Reszke of Warsaw, Poland, reported a case of complete medial oro-ocular cleft.

Treatment: The blind hypoplastic eyeball was retained to carry a prosthesis. The surgical construction was carried out in multiple stages rather than in the one grand slam of Tessier. First, the cleft was closed superficially by skin advancement. Then, both lacrimal sac and fistula were excised, and a lower lid was partially constructed. Maxillary mucosa was turned for lining so that the lower orbital rim could be grafted with split rib. Elongation of the vertical shortness of the cheek was treated with multiple
In this case, a superiorly based muscle flap turned off the rotation edge of the medial element and was inserted into the upper lateral advancement flap to fill out its deficiency. The result still left much to be desired since insufficient new tissue had been moved into the area of this horrendous defect.

PERSONAL CASES

In 1957 Max Grob noted the various paths of oblique facial clefts as those associated with cleft lip and involving the nostril and those lateral to the philtrum that skirt the nostril. This case shows nasal ala notches that correspond to Tessier's cleft 1 and 2 and lateral to the alar base in his cleft 3.

It has not been my fortune to treat many oblique clefts. I have had several incomplete cleffts, however, that seemed to show an obliqueness of direction, slanting not into the nasal floor but toward the alar base or lateral to it. These clefts are well corrected by the rotation-advancement principle. The direction of the cleft tends to shear off the point of the advancement flap, but extension of the lateral incision around the alar base releases enough lateral lip element to fill the rotation gap and complete the lip construction.

In this case, a superiorly based muscle flap turned off the rotation edge of the medial element and was inserted into the upper lateral advancement flap to fill out its deficiency. The
denuded end of the freed alar base flap was sutured to the septum for permanent fixation of the alar base and nostril sill.

Here is another oblique incomplete cleft threatening to skirt the ala.

And another!

This little boy had an interesting family history in that his mother's cousin had a cleft of the palate and his father had a microform including a unilateral congenital ridge and groove of the lip with slight elevation of the vermillion of the bow peak and notching of the free border along with a first-degree cleft lip nose. There was no difficulty with rotation in the child's lip as it required only a 5 mm. drop. A small amount of the nostril bridge was used in the advancement flap.
BILATERAL ASYMMETRICAL OBLIQUE CLEFTS

Here is a case not previously published as it is still unfinished. This patient (born February 4, 1963) of Thomas J. Zaydon of Miami had a bilateral medial oro-ocular cleft, complete on the left and incomplete on the right, with extension through the hard and soft palate.

As soon as the patient was one month of age, Zaydon operated on the left facial cleft with emphasis on achieving coverage of the left eye. He turned local mucosal and conjunctival flaps for lower lid lining and then, after extensive undermining, advanced a large facial-cheek flap medially for cover. Two months later a similar procedure was carried out on the right side using local conjunctival flaps and advancement of the cheek.

During the next two years five more operations were aimed at creating more conjunctival lining and skin in the area of the eyelids. An upper buccal sulcus was partially constructed, and on December 14, 1965, a Langenbeck-type procedure effectively closed the palate cleft. An operation in 1966 and another in 1967 continued to try for more eyeball coverage and the construction of a philtrum. After four years of Zaydon's heroic effort to correct this horrendous deformity, the patient revealed good progress in function and appearance. On his retirement from the Florida Crippled Children's Commission, Zaydon referred this patient to me.
At age eight, on September 1, 1971, bilateral fistulae were closed through the upper buccal sulcus, and during the process the antrum was opened on the left, allowing the escape of thin, dark, non-odorous fluid. This was suctioned, the antrum irrigated with neomycin-bacitracin solution and the opening closed with muscle. Split-rib grafts and chips were used to fill the bony gap on the left and to overlay the hypoplastic maxilla on the right. The skin of the prolabium was undermined and the unnatural corded subcutaneous vertical ridge split as two flaps based inferiorly. These flaps were shifted laterally into philtrum column positions, and a central dimple stitch was placed in an attempt to create a philtrum concavity out of an abnormal convexity. The straight-line oblique skin scars of the cheeks were interrupted by small interdigitations. Then the transverse ridge of the inferior vermilion of the prolabium was reduced and the adjacent horizontal groove filled with a dermal graft to improve the contour fullness of the vermilion border, particularly in the area of the tubercle.

Obviously, the next surgical procedure planned was the transport of distant tissue to make up the bilateral lower lid deficiency. During this period a change of homes was evidently in progress, and eventually the patient was taken by a remarkable and talented new parent who adopts only handicapped children. This course of events necessitated a move to Maryland, and her rehabilitation is being continued expertly by Alfred J. Suraci of Washington, D.C. This is part of his 1975 report.

When I first saw her in the latter part of 1972, it was quite evident that the sclera of the eyes bilaterally were becoming damaged due to her inability to close her eyelids. Hence, the first operative procedure on March 27, 1973, concentrated on this pathology and the severe ectropion was corrected on both sides by utilization of a full thickness right clavicular skin graft. In addition, adhesions of the lower eyelids to the sclera bilaterally were removed surgically and the upper and lower eyelids then sutured to each other for support. Fortunately, there was a 100% take of the skin graft and her eyes took on a much improved appearance, the severe hemorrhagic conjunctivitis disappearing with the ability to close her eyelids and protect the sclera.

The next surgical procedure was performed on July 13, 1973, at which time the severe scarring of the cheeks bilaterally, infraorbital regions bilat-
eraly and particularly on the left side was excised in their superficial portion, utilizing the underlying dermal scars to build up the cheeks. A "V-Y" advancement of the vermilion border on each side was accomplished. . . .

On January 29, 1974, a pharyngeal flap based superiorly with attachment to the nasal surface of the severely scarred soft palate was accomplished and this operative procedure has turned out quite well, her speech having improved remarkably.

In retrospect, techniques to "pull" growth and encourage downward positioning of the frontonasal component, as with the most modern Georgiade-Latham Mark III apparatus, and early side-to-side union of the orbicularis oris muscle might have been beneficial. This would be an effort to avoid a final result with a short central segment only moderately apparent at present but probably destined to increase proportionately in the late teens, as observed by Potter in 1974.
Lateral oro-ocular clefts do not correspond to any embryonic grooving.

The lateral oro-ocular cleft extends from the angle of the mouth upward to the orbit terminating in the lateral canthus or in a coloboma in the mid-portion of the lower lid lateral to the infraorbital foramen. More often than in the naso-ocular type, incomplete forms occur in which the central portion of the cleft in the region of the cheek is replaced by a scar-like groove. In mild cases the nasolacrimal duct is intact but in severe cases it is defective or absent. This is a mysterious cleft. It has the same origin as a transverse cleft, but its direction turns oblique, not corresponding to any of the known embryonic facial grooves. Karfik in 1969 called it the "true oblique cleft."

This is the rarest of all clefts; six such clefts are reported in the world literature. Three examples have occurred on one side in cases of the mixed group reported by Skoog and also by Greer-Walker. One unilateral incomplete cleft reported by Boo-Chai in
1970 was treated by paring and approximation of the edges of the coloboma. The tissue of the cheek along the line of the cleft and scar from the level of the alar base to the eyelid was lengthened vertically by a double Z-plasty, which also interrupted any tendency toward contraction of the oblique line.

Always a little more grandiose than the rest of us, Ivo Pitanguy of Rio, with Franco, reported a bilateral lateral oro-ocular cleft. In fact, he bragged that Brazil's 83 million people produced a higher percentage of rare clefts than reported by Fogh-Andersen in Denmark or Burian in Czechoslovakia. In 1967 he presented this table:

<table>
<thead>
<tr>
<th>Source</th>
<th>Total Clefts</th>
<th>Rare Clefts</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fogh-Andersen</td>
<td>3,988</td>
<td>48</td>
<td>1.20 ± 0.77</td>
</tr>
<tr>
<td>Burian</td>
<td>4,000</td>
<td>97</td>
<td>2.42 ± 2.44</td>
</tr>
<tr>
<td>Pitanguy</td>
<td>736</td>
<td>25</td>
<td>3.39 ± 0.66</td>
</tr>
</tbody>
</table>

Pitanguy's bilateral lateral oro-ocular cleft is the only one ever recorded in the world. In his patient the clefts ran obliquely in the lip from just medial to the commissure through the cheeks and into the lateral aspect of the lower eyelids. There was no cleft in the palate.
Treatment: Pitanguy approximated the soft tissue of the cheeks to the central component on each side with simultaneous positioning of the lower eyelids and construction of the oral commissures aided with Z-plasties.

This case with its early postoperative photographs was published in *Plastic and Reconstructive Surgery* in 1967. Here is a later follow-up record.
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 Wöllfer'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.

Wöllfer'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 glossoepiglottic ligament and downward between the geniohyoglossus muscles.

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

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"!
<table>
<thead>
<tr>
<th>Author and Year</th>
<th>Subject</th>
<th>Lower Lip</th>
<th>Mandible</th>
<th>Tongue</th>
<th>Ankyloglossia</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>Couronné, 1819</td>
<td>Adult F.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Mocckel</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>Perit, 1826</td>
<td>15 d.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Neck contracture</td>
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<tr>
<td>Bousson, 1840</td>
<td>1 1/2 yr.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Neck contracture, bulging of neck</td>
</tr>
<tr>
<td>Parise, 1862</td>
<td>2 1/2 yr. M.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>Dermoid of nose, neck contracture</td>
</tr>
<tr>
<td>Faucon, 1874</td>
<td>Child</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
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<tr>
<td>Lannelongue, 1879</td>
<td>21 d. M.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Hamilton, 1881</td>
<td>8 mo. M.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Wölffer, 1890</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
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<tr>
<td>Redard et al., 1891</td>
<td>3 mo.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Salzer, 1902</td>
<td>10% yr.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Debraisieux, 1904</td>
<td>3 d. F.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
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<tr>
<td>Keith, 1909</td>
<td>44 yr. M.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
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<tr>
<td>Brophy, 1923</td>
<td>2 yr. M.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Miyata, 1926</td>
<td>4 yr. F.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Morton et al., 1935</td>
<td>6 yr. F.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Stewart, 1935</td>
<td>1 yr.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Wassmund, 1935</td>
<td>15 yr. M.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
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<tr>
<td>Ashley et al., 1943</td>
<td>15 d. F.</td>
<td>+</td>
<td>+</td>
<td>+</td>
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<tr>
<td>Braithwaite et al., 1949</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Davis, 1950</td>
<td>4 yr.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
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<tr>
<td>Abramson, 1952</td>
<td>Newborn F.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Haym, 1952</td>
<td>5-6 yr.?</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Weyer, 1953</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
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<tr>
<td>Vigil-Lorenzo, 1955</td>
<td>Newborn</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Kawai, 1955</td>
<td>13 yr. F.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Torres et al., 1956</td>
<td>2 yr. M.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Recamier et al., 1957</td>
<td>1 yr.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td></td>
</tr>
<tr>
<td>Russell et al., 1961</td>
<td>15 yr. M.</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
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Comprehensive data on cleft types and associated conditions.
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.

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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.

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 vermillion 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.
62. Congenital Sinuses

LOWER LIP

CONGENITAL lip sinuses, equally well known as mucous pits, were first described in the lower lip by DeMarquay in 1845. In 1951 Watanabe could find only 100 cases reported in the literature, but by 1967 Coccia and Bixler had found 200 and the numbers are mounting, as evidenced by Hoffman’s 13 cases in 1971 from Mt. Sinai Hospital Cleft Palate Clinic, New York. Of course, many cases are not specifically reported. For instance, Viale-Gonzales, Barreto and Ortiz-Monasterio have so many clefts they can afford to include a case with mucous sinuses in association with a bilateral cleft presentation and not bother to report it separately.

INCI DENTS OF ASSOCIATION WITH CLEFTS

In 1964 Gorlin and Pindborg found this anomaly associated with cleft lip or palate in approximately 70 percent, but a review of the world literature reveals that most authors find the association closer to 80 percent. In 1954 Van der Woude estimated that 0.5 percent of patients with cleft lip and palate also have mucous pits while Rintala, Lahti and Gylling estimated the association at 0.9 percent.
ETIOLOGY

Theories of the etiology of these sinuses range from intriguing to ridiculous. Sir Arthur Keith suggested that the phylogenetic origin might be found in the mucous canals of the lower lip of sharks, but Ludy and Shirozy failed to find any sharks with mucous canals. Other theories lay the blame on an attempt by the lower lip to close a cleft of the upper lip, amniotic adhesions, abnormal invagination of lip mucosa, faulty union of the mandibular processes and presence of epithelial pearls. Wang and Macomber considered each in detail and then struck them all off.

Evidently Steida in 1906 was the first to suspect the "sulci laterales labii inferioris." In 1912 Huber noticed in the human embryo a secondary notch on either side of the median groove of the lower lip after complete fusion of its two halves. In 1934 Sicher and Pohl found the presence of lateral sulci in 6.5 mm. embryos and, more clearly defined, in 9.2 mm. embryos; in later embryos these had disappeared. In 1952 Warbrick, McIntyre and Ferguson studied serial sections of human embryos and proposed that these sinuses were due to failure of obliteration of the cephalic end of the lateral sulcus of the developing mandibular arch.

Wang and Macomber in 1956 and S. Hoffman in 1971 favored this lateral sulcus theory. These congenital sinuses most often occur as a pair of dimples on each side of the midline. Yet they can occur unilaterally, usually on the left but occasionally on the right and very rarely in the midline, which condition seems
consistent with the theory of the persistence of the lateral sulci and, when in the middle, the median groove.

**Heredity**

Wang and Macomber considered the anomaly due to a single defective gene. Van der Woude did also, and in her series of five families, including 94 persons, these interesting findings appeared which are of value for genetic counseling:

1. Autosomal dominant inheritance with a 50 percent chance of inheriting the gene was shown.
2. The affected individual could have pits alone, cleft lip or palate alone, all three or any combination (and a few had no abnormality but could transmit the gene).
3. Of her 94 persons, 55 had abnormalities (pits and/or cleft lips and/or cleft palates) and 30 of the 55 had cleft lips and/or palate.
4. Persons most severely affected (i.e., having pits and cleft lips and cleft palates) transmitted more severe defects to their offspring: 25 percent had cleft lips and palate, 8 percent had cleft lip, 8 percent had cleft palate.

Yet as cleft lip and palate and cleft palate alone are believed to be different entities by Fogh-Andersen and congenital lip sinuses have been found in both these conditions, it may be more likely that two genes are involved. Test and Falls found lip sinuses in five generations of the same family. Others have reported families with various combinations of sinuses and cleft lip and/or palate which can be traced back for several generations. In 1943 Straith and Patton from Detroit reported a family of 13 persons which, over a span of three generations, produced six persons (or 50 percent) with bilateral cleft lip and/or palate. Each member with a cleft also had bilateral mucous pits of the lower lip secreting tenacious mucus from each of its pair of ducts opening just above the mucocutaneous line.

These anomalies, as noted, can be transmitted to approximately one-half of the offspring. Even an unaffected member of an involved family may have severely affected offspring. Cases
reported in the literature suggest simple dominant inheritance
with variable expressivity. In 1943 Fogh-Andersen first pointed
out that the inheritance of clefts in the families with congenital
lip sinuses is of a different character from that in families where
no pits occur.

Robert J. Gorlin, Chairman of the Oral Pathology Division of
the University of Minnesota, who also has an M.S. degree in
chemistry, with J. Cervenka and S. Pruzansky, compiled a learned
treatise for Birth Defects in 1971 entitled "Facial Clefting and Its
Syndromes." They noted, in reference to lower lip pits and cleft
lip and palate:

The syndrome is transmitted as an autosomal dominant trait with 80%
penetrance of any component of the syndrome but there is a possibility that
the type of cleft present is influenced by modifying genes. The syndrome is
seen with a frequency of about 1:75,000 to 1:100,000 live births and affects
both sexes equally. On the basis of an analysis of 39 pedigrees, Cervenka,
et al showed that an affected individual has a 22 to 39% chance of having an
affected child with a cleft with or without lip pits. . . . Congenital lip pits
of the same type have also been seen in association with the orofaciiodigital
syndrome and with the syndrome of popliteal pterygia.

A MUCOUS VESUVIUS

These dimples, which may appear as a circular depression or a
transverse slit, are often situated at the apex of a nipple-like
elevation. Each dimple is the orifice of a blind sinus extending
downward and backward to penetrate the orbicularis oris muscle.
The pit can vary in diameter from pin caliber to 2 mm., and its
tract may extend in depth from 5 mm. to 2.5 cm. Lined by
keratinized squamous epithelium like the vermilion with nu¬
merous mucous glands in the depth of the blind end, the pit may
secrete a copious amount of mucus requiring wiping. Sir
Arbuthnot Lane reported a case in which the secretion increased
at mealtime.

MEDIAN LOWER LIP SINUSES

Although these sinuses most commonly occur bilaterally and
anteriorly near the mucocutaneous junction, four median lower
lip sinuses have been reported—by Ruppe and Magdelaine in
1927, Sato in 1938, Wang and Macomber in 1956 and Rintala and Lahti in 1973. Then there is a case reported by Rintala, Lahti and Gylling in 1970 and presented again in 1973 which was unusual in that the sinus was midline, relatively large and opening more posteriorly on the mucosal side of the lip. This sinus was found to be bipartite in its deeper part, with its septum covered by intact stratified epithelium. There was an associated bilateral cleft of the lip and palate. Oberst in 1910 reported a similar case. Miller in 1896 and Rose in 1868 also reported somewhat similar cases, in which the middle part of the lower lip resembled a nose with two nostrils from which the fistulous canals ran 2 cm. deep, each terminating in a separate cul-de-sac close to the mucosa of the labiogingival sulcus.

The occurrence of lower lip sinuses is often associated with minor facial developmental anomalies such as a short frenulum (found by Holbrook), sinus of the frenulum and fistula of the nasal bridge (MacKenzie) and preauricular tragal hillocks and occular dermolipoma (Parisien and Berken).

**TREATMENT**

In 1868 E. Rose suggested the indirect approach of an intraoral window to open the fistula into the oral cavity. In 1939 Baxter advocated more direct electrocoagulation of the entire tract. Mark Wang and Brandon Macomber, from experience with 15 cases, noted that the modern approach was complete surgical excision of the mucosal tract together with the surrounding glandular tissue. They warned that incomplete excision of the mucous glands would result in a mucoid cyst which in turn requires excision. They also advocated careful closure of the defect with special attention to the muscles to avoid undue looseness of the lower lip following orbicularis oris stretching from the presence of the sinus. Hoffman mentioned vertical elliptical excisions of the pair of sinuses in one of his cases.

Bill Lindsay in Mustarde’s *Plastic Surgery in Infancy and Childhood* warned about mandibular lip pits:

The pits may be single or double, superficial dimples or deep sinuses. The
treatment is transverse elliptical excision including sinus opening and tracts. All the involved mucous membrane must be excised or the sinus will recur.

T. D. Rees and D. Wood-Smith in their elegant red and gold 1973 book, *Cosmetic Facial Surgery*, presented a case of upper and lower lip vermilion hypertrophy which also had a small midline pit surrounded by a tubercle in the vermilion of the lower lip. Paring of the excess vermilion and excision of the pit improved appearance but still enabled the patient to achieve a normal lip seal.

The most fascinating description of lip sinus surgery was reported by A. Rintala and A. Lahti of the Finnish Red Cross Hospital in Helsinki in 1973 in the *Scandinavian Journal of Plastic and Reconstructive Surgery*,

After making an elliptical incision parallel to the lip around the lower lip sinus and elevating the edges of the sinus in order to facilitate dissection, it appeared that the sinus, extensive in its upper part, was divided into two at a depth of about 7–10 mm. Division was caused by a septum in the mid-line, like a pair of trousers. . . . Each “leg” continued separately almost to the bottom of the labiogingival sulcus for a distance of over 1.5 cm. The “legs” ran relatively close to the oral mucosa and converged toward the fundus, terminating close to one another in separate blind sacs from the bottom of which a fibrotic strand passed on to the anterior surface of the mandible.

**PERSONAL CASES**

I have had several congenital mucous sinuses of the lower lip in cleft lip and palate cases. There has been nothing of special importance to note except that complete excision is not always easy. Of those I have treated, four are of varied interest. In two I treated the clefts and the sinuses primarily, in another the clefts primarily but the sinuses secondarily and in the third both the clefts and the sinuses secondarily.

One case of bilateral lower lip sinuses, first seen in 1963, was associated with a severe complete right unilateral cleft of the lip and palate. There was no family history of clefts or pits, but it can be predicted that there will be. The mucous sinuses were in the vermilion of the lower lip near the mucocutaneous junction on
either side of the midline. They were excised individually and transversely and the scars revised several years later.

Another case of bilateral lower lip sinuses was seen in a girl with associated complete bilateral cleft of the lip and alveolus with protruding premaxilla but no cleft of the hard or soft palate. There was no history of clefts or pits in the family.

At 2½ months the premaxilla was set back and fixed in the notch in the hard palate after subperiosteal resection of a portion of the vomer. Lateral mucosa and muscles were joined behind the prolabium and a forked flap was banked as “praying hands” with the alar bases.
Eight months later, methylene blue was painted into the depths of the two pits and a transverse elliptical excision was extended 2 cm. deep into the lower lip dissecting out the sinuses like a pair of closed end trouser legs. The methylene blue facilitated total excision. The wound was closed in layers.

A boy was born in 1970 with a right complete cleft of the lip and alveolus associated with mucous sinuses of the lower lip. The mother had had a cleft of the lip and had a history of others in her family with clefts, but no mucous sinuses were reported. Rotation-advancement closure of lip was carried out at four months of age but sinus excisions were postponed. An attempt elsewhere resulted in right transverse and left oblique scars but persistent secretion of mucus.

At age four years both scars were included in one large transverse excision carried deep to remove all mucosa of the sinuses. Wounds healed without difficulty, but the patient returned to South Africa. Although the father reported that all is well, no photographs have been forwarded as yet.

A baby girl was born in 1968 with right complete cleft of lip and alveolus with bilateral mucous sinuses of the lower lip. Lip closure and sinus excisions were carried out elsewhere. It is of particular interest that the great-grandfather, the grandfather and the father all have had mucous sinuses of the lower lip. It is also of importance that the patient has a normal fraternal twin.
In 1972 the lip and nose were revised and the mucous sinuses reexcised. A large mucous cyst formed postoperatively which required another, more extensive transverse excision resulting, finally, in complete removal of the intact mucocele and excellent healing.

Total excision, of course, is the answer. If the sinus appears as a transverse slit, it may be easier to excise it horizontally, particularly if there is a pair. Occasionally better scar and contour may be achieved by excision of a vertical ellipse. Fortunately this anomaly, if handled correctly, leaves little to no residual deformity, and the importance of this outcome is multiplied as the defect has a 50-50 chance of reappearing in future generations.

**Midline Sinus of the Upper Lip**

The earliest cases were reported by the French, Lannelongue and Menard in 1891 followed by Clavet in 1899. There are eight cases in the world literature; seven of the sinuses opened in the center of the philtrum and one at the base of the frenulum. MacKenzie’s case had a 1 cm. blind tract not piercing the orbicularis oris ending near the nasal spine. Holbrook’s case had a 2 cm. wide opening in the center of the philtrum surrounded by red epithelium. Microscopically these tracts have been seen to be lined with squamous epithelium. Clavet’s case also had areas of columnar and polyhedral epithelium. In Holbrook’s case the tract was surrounded by hyaline cartilage and, unlike the other cases, had
no hair follicles or sebaceous glands. Most cases had occasional mucus drainage, and two had a history of infection. There have been associated anomalies reported: Kriens' case had a double frenulum; MacKenzie's case had a cyst and fistula on the dorsum of the nose. Treatment, when recorded, has been excision of the entire tract.

A VERMILION SINUS

Roger Bartels and Robert Howard of Orlando in December 1973 reported in *Plastic and Reconstructive Surgery* a case with mild hypertelorism and a minimal midline cleft of the vermilion of the upper lip with a tiny dimple at its center, the squamous epithelialized tract extending superiorly through the orbicularis oris muscle for 1 cm., then submucously for 2.4 cm., ending in a blind sac attached by a small fibrous band to the nasal spine. Total excision of the tract was carried out and followed by an uneventful recovery.
Bartels and Howard summarized upper lip sinus embryology in 1973:

Whether a midline sinus of the upper lip is a variation of (1) failure of downgrowth of the nasofrontal process, (2) breakdown of the mesenchyme-poor, fused, maxillary processes, or (3) failure of complete fusion of the maxillary processes growing together over the nasofrontal process, is not known. The association of hypertelorism with a midline sinus and cleft of the upper lip in our case supports the theories of Boyd and Frazer, that this anomaly is related to an abnormality of the nasofrontal process.

A Dimple in the Dimple

In 1960 I reported the incidental finding of a midline dimple in the skin of the medial element of the upper lip just above the mucocutaneous junction in a right complete unilateral cleft of the lip and palate. The tract extended several millimeters. It was observed as the medial component was rotated down into balanced position and then later simply excised.
Many articles have been written in minute detail to encourage surgeons to explore the primary and secondary possibilities of rotation and advancement, the forked flap, the midline Abbe flap and many other procedures. Even after faithfully reading all published material, visitors to Miami often say:

I've read every paper, but seeing the operation makes all the difference; and incidentally, it's encouraging to see that you too have to struggle to get the result.

It is true that words and diagrams are an aid, but nothing teaches like the actual surgical procedure, when the method is adapted to the peculiar aspects of a specific case. It has been my privilege to rotate and advance, to fork-flap and to lip-switch in numerous places, varying from a tiny hospital on the volcanic island of Grenada to Reed Dingman and Bill Grabb's great center at the University of Michigan, Ann Arbor; from the famous Massachusetts General Hospital, Boston, to TV from Hermann Hospital, University of Texas, Houston. The circumstances have sometimes been a bit distracting.

A demonstration at the military hospital in Bogotá, during a Latin American Congress for the northern zone, had my dear friend Hector Marino moderating the surgery and spicing the show like a sportscaster at a bullfight:

Now he is ready to finish it off. The needle is poised. Wait! No! No! He is taking out all the stitches, ladies and gentlemen, and starting all over again.
Then there was the cleft lip demonstration at New York University Hospital when I was competing with the great John Marquis Converse doing his first hypertelorism operation in the adjoining room; or the time behind the Iron Curtain at Comenius University in Bratislava with such luminaries as Professor Demjen assisting and Peet and Schmid standing by, not to mention a likely Communist or two.

These have all been exciting times. I appreciate the kind invitations of my colleagues and cherish the memories.

The most I ever did in one visit was in 1964 at Mellon’s Albert Schweitzer Hospital on then Papa Doc’s island of Haiti. No plastic surgeon had been over for two years, and in three days, with two anesthetists, I did my best to handle the more severe problems such as extensive burn contractures, huge hemangiomas, a congenital hand deformity, an orange-sized keloid, adding up to 23 operations. They included 10 unilateral cleft lips, one bilateral and one median, with the rotation-advancement cases averaging 30 minutes apiece. As many preoperative clefts as could be found had been collected in front of the hospital the first morning, and a photographic record was taken.
Then, in the dawn's dim light on the morning of my departure, all cleft surgery patients who could stand, sit, lean or lie were herded together again, stitches and all, for the final photograph.

This series enjoyed a comforting influence not ordinarily offered rotation-advancement and forked flaps. At the entrance of this little hospital was the witch doctor's endorsement as indicated by his specially arranged voodoo pile of sticks, strings, stones and bones.

Eight years later I received a note from Frank Lepreau, Medical Director of Albert Schweitzer Hospital:

I have in hand a reprint of your 1964 article about rotation-advancement cleft lip technique which you autographed and left here for Dr. Mellon some years ago. I used it a few days ago and you are quite right. This approach definitely does help Haitian lip repairs. As a very general surgeon, I need this kind of help to get me through the day's schedule.

After thousands of words, hundreds of illustrations, and thousands of miles, it becomes a bit tiring to watch some surgeons take a "free flying" principle and unconsciously, with numbers and measurements, clip its wings, placing a ceiling on
its potential height of flight. Any old seagull on Miami beach, because of the natural width of its wingspread, can glide with grace into the sun, but it is not supposed to achieve great speed and certainly not in the dark of night. Yet Jonathan Livingston Seagull, using the same basic principles of flight but with persistence and adaptability, trimmed his wings to falcon proportions and exceeded the exploits of other gulls.

He climbed two thousand feet above the black sea... brought his forewings tightly to his body... and fell into a vertical dive... The wing-strain now at a hundred and forty miles per hour wasn't nearly as hard as it had been before at seventy and with the faintest twist of his wingtips, he eased out of the dive and shot above the waves, a gray cannonball under the moon.

The repetition by great and established surgeons of such ceiling-restricted clichés as "not suitable for wide clefts," "more than 3 mm. is the cutoff," "a banked fork vanishes," "only the author can make it work" almost sends me up the wall and without the aid of wings! At such times it is tempting to call upon the caustic words of Johann Friedrich Dieffenbach, who after an absorbing section on principles of reconstruction in his 1831 book concluded:

Should the surgeon find my description not sufficiently circumstantial, and be unable to supply anything from his own knowledge of general principles, that he may find wanting, he had better altogether abstain from operating.

This would indeed be the easy way out. Yet no teacher should turn his back on even one willing student, and I, therefore, once more offer this challenge. Our only true ceiling is the ideal normal, which in turn demands our eternal dedication to the study of knowing this beautiful normal. It is then that we can see clearly both what is misplaced or missing and what is present but superfluous. Finally, with a little imagination in the use of our basic plastic surgical techniques of rotation, advancement, transposition and free grafting, we can quite effectively execute the shift, taking what is expendable to create what is desirable. From this simple format the present procedures can be not only understood and mastered but, far more important, transcended!