I. Basics
1. Embryological Theories

The embryological development of the face is still somewhat of a mystery. From the scant evidence we have, it is impossible to determine exactly how clefts form. It is like trying to re-create an intricate and technical full-length movie from a few one-frame film scraps cut at random out of the reel. There seem to be a number of intriguing theories, each of which has some evidence in its favor.

FUSION OF PROCESSES

The theory that separate processes fuse to form the central face was first advanced by Meckel in 1808 and later supported by Baer in 1828, Rathke in 1832, Kölliker in 1860 and Kollmann in 1868. German anatomist Dursy in 1869 and German biologist Wilhelm His of Leipzig University in 1901, working on chick embryos, popularized the theory of embryological development of the mid-face by the fusion of five facial processes about the rim of the primitive oral cavity or stomodeum. Superiorly there is the frontonasal process; laterally there are the paired maxillary processes and inferiorly the paired mandibular processes. According to the classical theory, all of these processes grow forward as finger-like projections to fuse with each other to form the normal face between the fifth and eighth weeks. The frontonasal process gives rise to three processes, the frontal, nasomedial (globular) and nasolateral, responsible for the development of nose, prolabium and premaxilla. The maxillary process by fusing with the nasomedial process forms the lateral upper lip and
cheek. The mandibular processes meet to form the lower jaw, chin and lower lip. This hypothesis reigned some 30 years as the accepted basis of facial formation. The failure of fusion of these processes seemed to explain the formation of the various degrees of unilateral and bilateral clefts and even the rare midline upper and lower lip cleft.

Thomas Mullen of San Francisco in 1931, following the Dursy-His hypothesis, described it this way:

Embryologically the growth toward the median line of the processes going into the formation of the mouth and lips, progresses, until in the seventh week they have fused. The manner in which the processes unite is similar to the healing of wounds. The ectodermic coverings of the processes unite and the mesodermic elements spread across the line of epithelial union to give rise to the muscles and connective tissue of the adult structure. Epithelial ingrowths separate the lips from the alveolar portion of the jaw.

Thirty years later Patten, for Pruzansky’s 1961 book *Congenital Anomalies of the Face and Associated Structures*, presented this schematic design of the fusion hypothesis.

The fusion theory is no longer in vogue. The term “process” implies a finger-like projection of tissue, and “fusion” implies that the projections meet, their epithelial walls disappear and they then grow together. As shown as early as 1910 by Pohlmann, this is not the case. Inspired by Fleischmann, Pohlmann was the first to cast doubt on the classical theory, realizing it was not a question of separate processes but of localized prominences.

**MIGRATION OF MESODERM**

Zoology professor A. Fleischmann of Erlangen, Germany, in 1910 had a hypothesis that
cleft palate is the arrest of the disappearance of the epithelial membrane which remains intact, not penetrated by the adjacent mesoderm.

This mesodermal penetration theory appealed to Victor Veau, who admitted that until 1930, at the age of 60, he had never even looked at an embryo. As he wrote in 1935:

I was searching for an operative method for the treatment of cleft lip. . . . I ascertained the fact that the only productive methods were those which approximated normal development: surgery of malformations is experimental biology. . . . The theory of the coalescence of the processes led me to a method that I thought to be a good one because it had an embryological basis. I experienced a series of disasters.

In 1934 Veau, disenchanted with the old facial process theory which he now considered a “myth,” sent data obtained from Fleischmann to Professor Hochstetter of Vienna. Hochstetter had been the first to describe the oronasal membrane, an incomprehensible finding according to the theory of facial processes. Hochstetter answered Veau by sending him two embryo specimens that had been puzzling him for some time: a 22 mm. unilateral cleft and a 23.3 mm. bilateral cleft. Veau was ecstatic:

You can imagine how joyful I was. . . . I found the undisputable proof of the Fleischmann theory.

Veau, the surgeon, noted in 1935:

I have been the gardener who has been responsible for the growth of the small plant, once it was germinated. The embryologists ignored Fleischmann or only referred to his hypothesis with irony. I showed that his theory could be applied to all clinical varieties of the cleft lip malformation.

Thus Veau endorsed the theory that with the penetration of mesoderm across the groove (arrow) normal development ensued while failure of the mesodermal migration eventually led to breakdown and cleft formation.

It is of interest that, although Veau changed his embryological ideas, this switch did not greatly influence his treatment of clefts. His acceptance of the importance of mesodermal penetration might, however, explain his enthusiasm for wire approximation of the muscles across the cleft.
MORE EMBRYOS

Studious Richard B. Stark of St. Luke’s Hospital, New York, with access to several more embryos with clefts, probed the mysteries of cleft formation and, like Veau, came up enthusiastic about the mesodermal migration hypothesis. He has written much on this subject, beginning in 1954 and including his presentation at the 1971 Melbourne congress, but extracts from his personal correspondence in 1972 are most pertinent. He outlined the pathogenesis of facial clefts during the first four to seven weeks:

As the discoid embryo develops a head and tail fold, so too does it develop an oral dimple, a two-layered stomodeal plate, composed of oral cavity ectoderm and entoderm of the primitive gut. The bilamellar membrane that forms in the region of the upper lip consists of two layers of ectoderm called an “epithelial wall” by Hochstetter.

If these gossamer two-layered membranes are not bolstered early, rapid embryonic growth dooms them to rupture with the production of a cleft. . . .

In the head and neck regions, mesoderm migrates over as well as around the head. Migration over the head is necessary if the forebrain, nasal dorsum and the central lip are to develop normally. Migrating around the head bilaterally, mesoderm reinforces the “epithelial wall,” the branchial membrane of the lip, first posteriorly near the future incisive foramen. As more mesoderm migrates medially, it forms the nasal floor as far as the nostril sill and reinforces the alveolabial complex, then the lip and finally the vermilion.

Mesoderm does not always arrive in sufficient amount to prevent the unreinforced membrane from splitting apart. If reinforcement fails totally, the result is a complete cleft whereas if the mesoderm is only partially present there will be a partial cleft. If the mesoderm arrives too late for effective reinforcement the anomaly will exist as separated but fully developed parts.

While the primitive nose takes form as two nasal placodes, inverted
ectodermal horseshoes, it is reinforced with mesoderm in the region of the nostril floor. The ectodermal cells involved begin to proliferate carving furrows and sulci and digging cavities and tunnels to create nostrils and alveolabial sulci. Deeper burrowing extends the twin nasal fossae through the mesenchyme and mesoderm into the oral cavity with a double tunneling "break-through" which actually circumscribes the mesoderm of the prolabium, premaxilla, anterior nasal septum and columella of the primary palate (P.P.). All this occurs from the 4th to the 7th week. After the 7th week the dental lamina appears and late-arriving mesoderm migrates into the prolabium piling up on either side as philtrum ridges.

Richard Stark and Joshua Kaplan in 1973 attributed insufficient mesoderm migration into the lip and nasal floor as the basic cause of cleft formation and the absence of dental lamina and the philtrum in the area of the cleft. They also tag part of the blame on the cannibalistic sculpting of ectoderm burrowing into the wall, attenuating it, fraying it, finally severing it...

and the tension of rapid growth causing traction to the point of rupture. In an incomplete cleft it is their opinion that the epithelial plug has remained as Simonart's band.

The area of lip and palate clefting has been divided into two major parts with the incisive foramen as the demarcation point.

1. The primary palate, consisting of the anterior nasal spine, columella, medial portion of the upper lip and premaxilla, develops during the fourth to seventh week of intra-uterine life.

2. The secondary palate, consisting of the hard and soft palate, develops during the seventh to twelfth week of intra-uterine life.

Division of the primary and secondary palate is marked by the incisive foramen in the roof of the mouth and bilateral sutures which extend from this midline foramen to the space between the maxillary lateral incisor and first canine tooth.

A SIMPLIFICATION

Quite simply the mesodermal migration theory proposes that coincidental with invagination of the oral cavity and nasal pits
there is a "heaping up" of the adjacent tissue forming facial prominences. As the oral and nasal cavities deepen, there is an increase in the sizes of these prominences due to the penetration of mesoderm. As more mesoderm enters the area, the bulging effect is increased so that what used to be a wall of tissue with ectoderm on one side and endoderm on the other is transformed into a conglomerate of hills and valleys. The difference between a "hill" and a "valley," or a prominence and an apparent groove, is merely dependent on the amount of mesoderm between the two epithelial layers. As these prominences approach each other, their blending is at the expense of the "valleys" or grooves, and the normal facial contour evolves. Failure of sufficient mesoderm to migrate into a specific area would be responsible for the persistence of a groove. With consequent epithelial breakdown, the persistent groove gives way to an established cleft.

A M E R G I N G

Bradley M. Patten, Emeritus Professor of Anatomy at the University of Michigan, was the son of a professor of anthropology at Dartmouth College, and throughout his life there was always a bit of the Ivy League about him. He combined the Dursy-His and Fleischmann-Veau theories and was adamant about his interpretation. He proposed the theory that the original frontal area was submerged by tremendous forward and downward growth of the paired nasomedial processes, implying both mesodermal infiltration and fusion of parts. He diagramed his merging theory in a chapter on "Normal Development of the Facial Region" for Prautzansky's book *Congenital Anomalies of the Face and Associated Structures*. Then, for *Cleft Lip and Palate* by Grabb, Rosenstein and Bzoch, he discussed the embryology of clefts by comparing three unilateral clefts of the lip: minor, halfway and complete. He noted that the variable in this series is clearly that part of the nasomedial process which contributes to the formation of the median part of the upper lip. . . . On the medial side of the cleft there is a small portion of the prolabium which, by reason of its relation to the midline, could only have been derived from the left
nasomedial process. This process, however, was sufficiently feeble in its
growth so that it did not meet and unite with the maxillary process as
it normally should. . . . Close scrutiny of the mesenchyme adjacent to the
nasal fin of normal embryos reveals that on the nasomedial side the mesen­
chyme is more richly cellular and more highly vascular. I believe this
indicates that it is the prime mover in this important union and that when
its growth is inadequate a cleft will remain.

In reference to the accompanying nasal deformity, Patten noted
that even when the cleft of the lip is relatively small there is
a striking asymmetry in the configuration of the nose. He also
noted that

Avery has shown that in cases of unilateral cleft lip there is a marked
accompanying deficit in the growth of the nasal capsule on the side of
the cleft. This means that the disturbance of the potentialities of the
mesenchyme of the nasal processes, which is so obviously involved in
defective formation of the fibromuscular part of the lip, is manifested also
in its chondrogenic potentialities. It is not surprising, therefore, that the
extent of the nasal asymmetry is correlated with the extent of the defect
of the lip.

Hamilton, Boyd and Mossman summarized in 1962:

Current embryological opinion regards the elevations or "processes" of the
developing facial region as in the nature of surface swellings produced by
proliferation of the underlying mesoderm. The furrows between the eleva­
tions become smoothed out in subsequent development as growth and
fusion of the mesoderm centres proceed beneath the ectoderm.

They cited Streeter's 1948 stand:

Under the circumstances no ectoderm requires absorption; it is simply
flattened out in adaptation to the changed surface.

They admitted:

Nevertheless for descriptive purposes, it is convenient to retain the terms
maxillary, mandibular and fronto-nasal processes.

In the mesodermal migration and merging theories when the
mesoderm penetration is retarded, the groove persists and dis­
ruption along the line of a groove results in a cleft. This all
seems simple enough and quite logical. Yet, as Vaclav Karfik
of Prague noted in 1967, it is too simple, for there is no embryonic facial groove to explain the lateral oro-ocular cleft which, although rare, does occur.

OTHER THEORIES

There are other theories besides the failure of fusion, mesodermal penetration or the merging of prominences. There is the theory of failure of the epithelial wall to develop, as proposed by Tondury in 1950, and the rupture of previously formed cysts in the soft tissue bridges, as suggested by Steininger in 1939.

In Hamburg in 1966 Pfeifer offered still another theory. Clefts in which the vermilion ceases at a distance from the nostril with skin and mucosa meeting in the cleft in a scar line flanked on each side by ends of muscle were termed "secondary clefts." Pfeifer proposed the possibility that these were formed not by failure of fusion but by the breakdown of a lip once intact. Here again, to prove the theory we would have to rerun the movie, but preferably backward.

STILL GUESSING

Obviously no theory enjoys universal acceptance, and as no one has been an eyewitness to the entire "in utero show," as yet it is impossible to propose and prove exactly what is happening. Embryologists continue to make educated guesses from 12 single frames, for as of Stark's latest publication only 12 known human embryos with cleft abnormalities had been studied. Of these only one, examined by Tondury, was less than 18 mm., and beyond 18 mm. the facial "clefts" are no longer normally present.

EMBRYOLOGICAL GEOGRAPHY OF FACIAL CLEFTS

With one exception, which is quite rare, congenital facial clefts have been correlated with grooves normally seen in the 6 to 12 mm. embryo. More specifically, a facial cleft has been considered to result from the failure of developing facial prominences
Cyclopia (B²)
Holoprosencephaly (B¹)
Hemicephalus (B³)

Median cleft (7)
Naso-ocular cleft (2)
Horizontal cleft (5)
Lower midline cleft (6)
Treacher Collins syndrome

Unilateral cleft (1)
Medial oro-ocular cleft (3)
Lateral oro-ocular cleft (4)
Holoprosencephaly (B₁)

Cyclopia (B²)

Hemicephalus (B³)

Median cleft (7)

Naso-ocular cleft (2)

Unilateral cleft (1)

Horizontal cleft (5)

Medial oro-ocular cleft (3)

Lower midline cleft (6)

Lateral oro-ocular cleft (4)

Treacher Collins syndrome
or processes to obliterate these grooves by mesodermal migration or merging. This failure then allows the abnormal persistence of a groove which, in turn, breaks down and produces a cleft. The relationship of facial clefts and embryonic facial geography can be charted in this manner:

1. **Cleft lip**—failure of both maxillary and nasolateral prominences to merge with the nasomedial prominence. The nasolateral prominence successfully unites with the maxillary.

2. **Oblique cleft, naso-ocular type**—failure of nasomedial, nasolateral and maxillary prominences to merge with each other (example from Bartels, O’Malley, Baker and Douglas).

3. **Oblique cleft, medial oro-ocular type**—failure of maxillary prominence to merge with either nasolateral or nasomedial prominences. There is successful merging of the nasomedial and nasolateral prominences (example is case of Thomas J. Zaydon, Miami).

4. **Oblique cleft, lateral oro-ocular type**—does not correspond to any embryonic facial geography. The cleft traverses the maxilla lateral to the infraorbital foramen (example from Pitanguy and Franco). As noted by L. Hovey, this anomaly bears some resemblance to examples of the Treacher Collins syndrome such as the one published by Rogers in Converse’s *Reconstructive Plastic Surgery*.

5. **Horizontal cleft**—failure of maxillary and mandibular prominences to merge (example from Millard and McNeill).

6. **Midline cleft of lower lip and mandible**—failure of merging of the paired mandibular processes (example from Stewart).

7. **Median cleft**—failure of merging of nasomedial prominences. These clefts can be divided into two main groups.

   A. Failure of mesodermal migration or merging of the nasomedial prominences resulting in a varying degree of a midline cleft (example from Millard and Williams).

   B. Complete agenesis of the frontonasal process resulting in arhinencephaly, cyclopia, etc.

   (1) Agenesis of nasomedial process—nasolateral processes and maxillary processes will merge. In this case there will be absence of the premaxilla, prolabium and nasal columella.
(2) Cyclopia—agenesis of frontonasal prominence including nasomedial and nasolateral processes. Maxillary processes may merge; hence the absence of the nose (example from Millard and Williams).

(3) Hemicephalus with median lip cleft (Darwin’s missing link?) (example from W. B. Davis).

This chart only scratches the surface
This classic chart of the geography of facial clefts does give a general surface outline of the position of most clefts but must not be considered the total picture. In 1957, Max Grob of Zurich diagramed the various paths of oblique and transverse facial clefts concluding,

Note inability to explain paths of clefting using classic embryonic concepts.

In 1970 Robert Gorlin of the Division of Oral Pathology, University of Minnesota School of Dentistry, questioned the validity of clefts running along embryologic grooves when he wrote,

Only rarely, however, does the oblique facial cleft follow the epithelial grooves and other explanations should be sought.

Facial grooves are superficial and do not represent actual through-and-through clefts. As pointed out by Otto Kriens, what is more important are the goings-on under the surface as a chain reaction of one fault inducing another. Then to this third dimensional factor must be added also the element of time. It is becoming more and more apparent that clefts occur not over a period of weeks or months but within a very short interval of hours or, at most, days.

MYSTERY VALLEY

Another area of mystery besides the various cleft formations is the method of creation of the philtrum. Ian W. Monie, a Scottish anatomist from the University of Glasgow, a past president of
the Teratology Society and a Guggenheim Fellow, has been Professor of Anatomy and Embryology at the University of California, San Francisco, since 1952. In 1972 he wrote:

The study of the philtrum was undertaken because the development of this region seemed to have received relatively little attention. I recall J. B. de C. M. Saunders, Professor Emeritus of Anatomy, drawing attention to this in the course of one of our chats.

Thus Monie with Cacciatore in their 1962 study of the development of the philtrum noted two previous theories of philtrum formation: (1) persistence of the groove between the globular elements of the frontonasal process (Waterston) and (2) a heaping up of the maxillary mesoderm on either side of the middle line (Boyd). They then proceeded to study transverse sections of the upper lips of embryos, ranging in age from nine weeks to term, along with comparable sections of the upper lip of adult humans. They found no evidence that the philtrum is related to the lines of fusion of facial processes in the embryo. On the contrary, the philtrum does not appear until several weeks after the union of such processes has been completed. They found the philtrum primarily in association with the increasing density of the connective tissue in the median portion of the upper lip and first apparent between the third and fourth months of fetal life. In the September 1962 issue of *Plastic and Reconstructive Surgery*, they diagramed three human embryos to show the timing of the philtrum. At three months (60 mm.) there was a transverse crease but no philtrum, at three and one-half months (85 mm.) a transverse bow-shaped crease but no philtrum and, finally, at four and one-half months (130 mm.) no transverse crease but at last a philtrum.

Monie and Cacciatore pointed out that the paramedian eminences which flank the philtrum were found closely associated with the development of the philtrum but were independent of the lines of "fusion" of the facial processes, and although they contain considerable muscle postnatally, their development and configuration seem independent of it.

R. A. Latham, oral biologist at the University of North
Carolina School of Dentistry, stated at the 1973 cleft symposium at Duke, and again wrote personally that

It is very likely that the philtral dimple is due to an attachment between the lip epidermis and the mid palatal suture; and that the philtral ridges are due to some extent to the flared out posterior ends of the medial crura of the alar cartilages.

He also promised:

If this is correct, it should be demonstrable and more information will be forthcoming.

In 1973 Latham discarded his previous theory, explaining that

Studies of the normal fetus show muscle fibers arising from the alveolus in the lateral incisor area coursing anteromedially to insert near the epidermis in the medial philtrum portion of the lip.

As this is contradictory to the findings of Monie and Cacciatore, it may or may not reduce the shadow hovering over "Mystery Valley."

Again, there are missing links, and again, the little embryological data available give us very minimal assistance in surgical correction. Creation of a philtrum still offers difficulty requiring relatively extensive surgical acrobatics.

ACADEMIC INTEREST

The embryological explanation of cleft formation is of great academic interest. No doubt if ever we are able to see an entire trimester movie of cleft formation from beginning to end, we may have better insight into the most appropriate surgical method of correction of the deformity. As today's fantasies are tomorrow's facts, such a movie may not be too far from reality. The fertilized ovum has already been developed and observed in vitro to the one-month stage. Were it not for moral and ethical dicta, space-age scientists could soon solve the mysteries of human development.
Yet, even after complete understanding of how a cleft happens, I am not certain the surgeon's task will be much enlightened. As with any large healed hole which has been created by gunshot, cancer ablation or congenital anomaly, the surgeon, to close it, still must take what he has available to make what he needs.

Anyway, by the time we know *how* a cleft occurs, we probably will know *why* it occurs and can devote less time to its repair and more to its prevention. Unquestionably, cleft causation and prevention is the goal of the future. Yet our knowledge of this subject today is so inadequate that no further lines will be wasted. The subject is now dismissed after the humble acknowledgment that it deserves greatly increased research and many books of its own and that its final solution along with associated findings will be important enough to usher in a new era in the history of man.
2. Anatomy in Multiple Dimensions

All anatomy and most surgical textbooks laboriously describe again and again the normal anatomy of the face. Most of us find that anatomy unrelated to surgery gets lost in its Latin. Yet when pertinent to the surgery, it becomes vital and exciting and will be called upon constantly throughout this book to influence the design of surgery.

Here in its own unrelated section, it will be reviewed in its multiple dimensions but in "bikini" briefness, just enough to cover the essentials but not so much as to put you to sleep. Certainly, a comparison of the anatomy of the cleft deformity and that of the normal in reference to muscles, blood supply and specific labial and nasal peculiarities merits our attention, as these elements should influence directly any plan of cleft lip surgery.

Remember, the anatomy of the cleft deformity reflects not only the varying extent of embryological failure but the ultimate result of growth and development in the absence of intact dynamic labial and palatal musculature as well as the lack of structural support of the bony arch and partition between the oral and nasal cavities. In fact, because of a unilateral cleft, growth and development exaggerate the asymmetry and with it the difficulty of correction.

Effect of the Septum on the Maxillae

Ralph A. Latham, trained at Queen's University, Belfast, and inspired in clefts by Burston in Liverpool, is now Associate
Professor of Oral Biology at the University of North Carolina School of Dentistry, Chapel Hill. In 1969, in *Cleft Palate Journal*, he proposed the hypothesis that the nasal septum is a key factor in the height and anteroposterior dimensions of the face and presented diagrams and microscopic studies for substantiation. He showed by photomicrograph a sagittal section from a 17-week fetus demonstrating the septopremaxillary ligament (SPL) in relation to the nasal septum (S) and the anterior nasal spine (ANS). Then at Georgiade’s 1973 Foundation Cleft Symposium at Duke University, he reproposed that in the embryonic period the nasal septum is the dominant growth structure and in the normal there is equal septal drag on both maxilla as diagramed. When a unilateral cleft occurs, one side is set free, but the forward drag is still present on the other side through the intact septopremaxillary ligament effecting deviation in growth. The broken line sketched by Latham shows that a bent septum must incur a deficiency in height of the premaxillary region. Then, in the latter half of the prenatal period and after birth when the maxillae begin to exert their own growth, the bent septum acts as a bridle impeding downward progress of one maxilla. Thus, Latham pins the blame on the septum for premaxillary rotation, first downward and later upward, pointing to the tethering restraint of the bent septum on both downward and forward growth of one maxilla. He describes the final stage as: “It is much like the way a fish pulling on the line bends the fishing rod.”

**ANATOMY OF THE UNILATERAL CLEFT LIP NOSE**

The typical nasal deformity associated with congenital unilateral cleft of the lip presents both a discrepancy and a displacement of parts which persists without great improvement during growth. The distortion, being confined to the cleft side only, is emphasized by the constant comparison with the normal opposite side.

1. **Platform.** When the actual platform of the nose is cleft, the projection and outward rotation of the premaxilla and the
retroposition of the lateral maxillary element certainly guarantee that the nose will sit *askew*. Even when there is no bony cleft, the discrepancies in maxillary contour are responsible for some degree of nasal asymmetry. This is an architectural fact, for any structure, with one of its key legs shortened or pulled out from under it, must tilt!

2. *Septum.* With the medial maxillary element forward and the lateral maxillary element backward the effect is reflected in the twist and slant of the septum. The anterior portion of the septum tilts over the cleft like a lean-to with its inferior edge dislocated out of the vomerine groove and presenting with the nasal spine in the floor of the normal nostril. This dislocation is responsible for a twist to the nasal tip.

3. *Nasal bones.* The asymmetry of the maxilla and premaxilla and the deviation of the septum assure some distortion of the nasal bones.

4. *Columella.* The columella is deflected by the deviation of the septum behind it. It also suffers a unilateral shortness in vertical height which can vary from three-fourths to two-thirds to even one-half that of the normal side.

5. *Nasal floor.* In complete clefts the nasal floor is cleft not only in skin and muscle but in bone, and the position of the maxillary elements can vary from overlap to abutment to gaps of millimeters or centimeters. In incomplete clefts there can be a variation from a thin skin bridge across a very wide nasal floor to a nasal floor within a millimeter of the normal width. I have never seen one the exact size of or smaller than the normal side.

6. *Lower lateral alar cartilage.* The alar cartilage on the non-cleft side should be normal but often seems to be overdeveloped when compared to the attenuated cartilage on the cleft side. The deformed alar cartilage arching the cleft is dislodged from its rightful balanced position beside its mate in the dome of the tip. Rather, its medial crus is lower in the columella, with the junction curve of the medial and lateral crus separated from the opposite alar cartilage and resting below it, being flattened, spread and stretched across the cleft at an obtuse angle.

7. *Alar crease.* The alar crease on the normal side runs parallel
to the upper border of the lower lateral cartilage and smoothes out as it approaches the bulge of the alar cartilage in the dome of the nasal tip. On the cleft side the alar crease has no alar cartilage bulge to give way to and consequently, unopposed by this structure, continues obliquely across the tip just lateral to the join of the columella and through the rim of the ala. This abnormal extension of the alar crease across the tip on the cleft side produces a disjointed effect to the tip and often is responsible for an actual kink in the alar margin itself.

8. Alar base. The alar base is invariably rotated outwardly in a flare. It can be wider in bulk than normal or grooved, everted or misformed in various ways to complicate the correction.

9. Alar rim. Invariably there is a skin curtain without cartilage which droops over the alar rim like a web further reducing the apparent length of the columella on the cleft side.

10. Vestibular lining. The lining of the nasal vestibule seems to be stretched over a greater area than on the normal side with actual eversion of the lining in the alar base region. Yet there is a paradoxical discrepancy in a shortness of lining along the axis from its lateral attachment to the pyriform opening to its join with the septum at the tip. A pull on the nasal tip will cause a band in the lining to rise like a web arching obliquely across the lateral vestibular wall. Gillies and I wrote that the bridling effect of the tissue shortage on the cleft side was responsible for dragging one entire alar cartilage from its normal riding position with its opposite fellow on the tip crest of the septum. In fact, this shortness in the vestibular lining inside coincides with the abnormal extension of the alar crease over the dorsum on the outside and may be partly responsible for the excessive grooving.
The total effect of these 10 unilateral nasal discrepancies produces a nose with the nostril aperture on the cleft side positioned along a horizontal axis, whereas the normal nostril aperture takes a vertical direction. This is accompanied by a flat nasal tip on the cleft side, along with a webbed nostril arch and a flaring ala which in the adult may be responsible for an increase of as much as one-half inch in total nasal length on the deformed side. In fact, as Gillies and I wrote in 1953 (published in 1957),

If the patient is approached from the left one may be presented with a prospective Hollywood profile, from the right a Fagan caricature.

The appearance is truly bizarre, eye-catching and pathognomonic of this congenital anomaly.

It would seem that the presence of a cleft in the nasal floor associated with generalized spreading, a cleft in the lip muscle allowing unopposed dragging of the nasal spine to one side and the alar base to the other and the inequality of the maxillary platform ensuring an asymmetrical nasal tilt could account for all the characteristics of the unilateral cleft lip nose. Yet minor and even moderate degrees of this same deformity can occur in the absence of any lip cleft at all. In fact, here is a somewhat faded photograph I took in the Lord Mayor Treloar Hospital courtyard at Alton in 1948 of one of Kilner's cases which revealed a cleft lip type of nasal deformity with no history of cleft lip except a slight congenital scar. At the time, this boy caused me much concern as he nullified the myth that the associated nasal deformity is directly dependent upon the actual cleft in the lip.

This microform is more common than is generally realized. Besides the English boy just mentioned, the American girl shown here, and even a super movie star who has mumbled his way to more than one Oscar, show a first-degree nasal asymmetry which, if not traumatic, could be congenital. Then there is the international array of published cases by R. Brown, Stenstrom and Thilander, Tuleenko, Boo-Chai and Tange and Kozin.

After H. Pashayan and F. C. Fraser wrote a paper entitled
"Nostril Asymmetry Not a Microform of Cleft Lip," Maria Tolarova of the Czechoslovak Academy of Sciences, Prague, challenged their stand, reporting that nostril asymmetry occurred frequently in the first-degree relatives of patients with clefts. That the excess of nostril asymmetries in her study was greatest in the relatives of patients with isolated cleft palate, hardly a microform embryologically, is puzzling. Yet Tolarova held her ground, noting that nostril asymmetry as a microform was accompanied by associated deformities such as: underlying bone deficiency or prenatal scar with vermilion deformity, alveolar ridge discrepancy at the lateral incisor, malpositioned teeth in this area, cleft uvula or osseous cleft or even bent alveolar arch in the lateral incisor region. At least all these examples of nostril asymmetry confirm that first there must be an interruption in the natural nasal development, probably associated with inadequate mesoderm migration in the nasal area.

To Stark's question, "Is the nasal deformity due to displacement of the cleft half as the alar base sinks into the crevasse, or is it due to an inherent tissue deficiency upon the side of the malformation?" there have been several answers. Huffman and Lierle blamed the malposition of the cleft half, and Stenstrom and Oberg pulled on cadavers to reproduce the deformity. Finally, Richard Stark and Joshua Kaplan turned to measurements of ectodermal volume on the two sides of the primitive nose in embryos with unilateral clefts and found a relative deficiency on the cleft side. In a 24.5 mm. embryo the ectodermal ratio of the normal versus the cleft side was 6.1 to 5.4 sq. cm. or a cleft deficiency of 7 sq. mm. In a 36 mm. embryo the ratio was 23.4 to 20.2 or a cleft deficiency of 32 sq. mm. In a 48 mm. embryo the ratio was 18.2 to 14.8 or a deficiency of 34 sq. mm. This last and largest (48 mm.) embryo had a smaller nasal ectodermal volume than the 36 mm. one, confirming the obvious fact that some individuals are destined to have larger noses than others.

So, the nasal deformity is probably the result of a combination of factors. Once the normal embryonic sequence of nasal events has been upset, all the other anatomical vectors act to exaggerate the distortion. In spite of the odd exception, there does exist
a vague correlation between the extent of the lip cleft and the
degree of nasal distortion. The nose in minor lip clefts, although
occasionally moderately deformed, usually has only minimal
distortion, whereas in complete clefts the nasal deformity is
consistently severe and often horrendous.

**ANATOMY OF THE UNILATERAL CLEFT LIP**

The upper lip, attached above to the nose, blending laterally into
the cheek and curving into the lower lip at the commissures,
is formed of muscles and glands covered in front with skin and
lined behind with mucous membrane. These layers are tightly
adherent to the muscles and are sealed along the free margin
with a vermilion border which is unique in man. Brescia of
Loyola University described the red of the lip vividly in *Cleft
Lip and Palate*, by Grabb, Rosenstein and Bzoch:

In this transitional zone the epithelium is thin and not keratinized; also
the connective tissue papillae are numerous, densely arranged, slender, and
extend close to the surface epithelial cell layers. The abundance of eleidin
in the epithelial cell layers, which increases translucency, and the num-
erous rich capillaries of the papillae, create the red color of this area.

Burkitt and Lightoller explained the development of this aspect
of the human lip:

When lips ceased to be prehensile organs and were being used in a modified
way for speech, the marginal portion became weaker and was dragged
upwards and forwards by the more powerful m. quadratus superior and
downward and forward by the m. quadratus inferior and the platysma.
Originally this action exposed some of the mucous membrane of the
mouth, which, in course of time, became modified to form the present
red lip area.

**MUSCLES**

The orbicularis oris muscle around the mouth with its
sphincter-like ability to contract and relax has influential muscle
associates that act as happy elevators and sad depressors. These oral muscles, which are involved in normal labial and nasal action, are divided into two groups, both supplied by the facial nerve. Sir Arthur Keith in 1923 noted that muscles supplied by the facial nerve are peculiar in that many mental states are reflected in them, and their development goes hand in hand with the development of the brain:

The more highly developed the brain of any primate, the more highly specialized are its facial muscles.

As we go up the scale of development of the mammalia, we find that the muscles about the mouth and in the lips become greatly specialized and are finer and more delicate in texture. For instance, in his study of the facial musculature of the Australian aborigine, Lightoller found muscles that were much thicker and more powerful with less apparent differentiation.

**Orbicularis oris**

Burkitt and Lightoller in 1928 described the orbicularis oris not as a true sphincter muscle but as eight muscle components with their origins in the small muscle mass, the modiolus, at each angle of the mouth. Arising from their respective modioli, the orbicularis fibers of one side end by decussating in the median line with fibers from the opposite side. The orbicularis is composed of four pars peripherales extending from the rima oris outward in an ever diminishing sheet reaching as far as the septum nasi above and the labiomental groove below on the right and left. It lies approximately in the center of the lip, and its fibers are pierced and interlaced by the fibers of the quadratus labii superioris and inferioris and labial portions of the platysma, the so-called labial tractors which pass through it to gain insertion in the fibrous tissue beneath the mucous membrane.

Intimately associated with the pars peripheralis is the pars marginalis with its two right and left components lying in a plane superficial to the pars peripheralis and confined to the area beneath the lip vermilion.
Tractor muscles

Greatly affecting the action of this circumoral musculature are the labial tractors. They are radially arranged as superficial and deep muscles, and most have as an attachment the modiolus at the angle of the mouth.

In the upper lip, these include the superficial zygomaticus major and minor, the quadratus labii superioris and the deeper levator anguli oris, which raise the lips and corner of the mouth and spread the nostrils. In the lower lip are the superficial depressor anguli oris and the deep depressor labii inferioris and mentalis muscles, which pull the lip down and the corner of the mouth outward. Then there is the superficial risorius, which pulls the corner of the mouth laterally, and the deep buccinator, which tenses the cheek. The fibers of these muscles insert into both the skin and mucous membrane by means of elastic tendon
extensions. According to Brescia, where the tendons insert in a concentrated area a dimple occurs, but where they insert in a linear fashion a crease is formed.

Cleft muscles

In the presence of a cleft, the orbicularis oris muscle fibers do not decussate transversely across the midline over the maxilla but tend to run up parallel to the cleft edges toward the base of the nose. With their integrity divided, they often contract into a disappointed, useless lump usually evident on the cleft side. With the orbicularis oris muscle sphincter crippled by the split and no longer a worthy opponent, the antagonist tractor muscles make the most of their advantage, exerting unnatural lateral lifting and distortion of the lip elements in both incomplete and complete clefts.
Muscle dissections

In 1965 Fara, Chlumska and Hrivnakova of Charles University, Prague, Czechoslovakia, dissected and described the orbicularis oris muscle in incomplete clefts. Then in 1968 introspective Miroslav Fara again reported his findings after dissecting the muscles and blood supply of three unilateral incomplete and four unilateral complete clefts of the lip out of 16 stillborns. His dissections revealed the muscle bundles running along the edges of the cleft turning upward toward the line of the nasal wing on the lateral side and to the base of the columella on the medial side. He found that the muscles on the medial philtrum side of the cleft were always more hypoplastic and did not extend to the very edge of the cleft as they did on the lateral side, suggesting limited ability of muscle fibers to grow across the midline. Rarely, but in two of his cases, he found muscle fibers
running horizontally to the cleft edge. In unilateral incomplete clefts the muscles did not, as a rule, cross the cleft unless the bridge was at least one-third of the height of the lip. These findings prompted Fara to exclaim:

They show what difficult tasks confront us, if we wish to bring the muscle fibers together "end-to-end" and not "side-to-side" or "end-to-side" in the primary suture of the lip—regardless of the method used.

It is interesting that almost simultaneously, in 1966, Pennisi, Shadish and Klabunde in San Francisco compared microscopic sections of the philtrum and the skin bridge in incomplete clefts with normal lips. Their work was finally published in 1969 when they noted that the muscle fibers in the normal were orderly and transverse whereas in the cleft lip the muscle fibers swept up vertically toward the nose, running parallel to the edges of the cleft.

These general findings were further corroborated by two Muscovites in 1969 when R. D. Novoselov and A. A. Lavrentiev studied the surgical anatomy of the mimetic muscles in the oral region in three cadavers of newborns with congenital unilateral clefts of the upper lip.

On the side of the cleft they have a number of morphological peculiarities: they are less differentiated (more densely adhere to each other and have common muscle fibers), the beginning on the bone is displaced 2-3 mm. anteriorly and posteriorly. The muscles of the surface layer are somewhat longer and wider, approach the angle of the mouth lower by 2-3 mm. Muscles of the deep layers are shorter and narrower in the cross section and are situated to the oral cleft in similar or blunter angles. Splitting of the musculus orbicularis oris is attended by changed direction and attachment of the main muscular bundles of the deep layer. The most powerful of them, the superior muscular bundle on the side of the cleft, is attached to the base of the nasal ala, and on the healthy side in the region of the nasal base. They play an important role in the mechanism of displacement of base of the nasal ala on the side of the cleft and base of the nasal septum on the healthy side. This should be taken into consideration when restoring the continuity of the musculus orbicularis oris. In connection with splitting of the musculus orbicularis oris, the function of the latter is lost. This leads to incoordinated contraction of mimetic
muscles in the oral region. The non-counterbalanced muscles-antagonists at the moment of contraction exert a faulty effect on the nasal cartilages and fragments of the alveolar process. This effect should be considered as one of the active factors in the mechanism of deformations of the nose and facial skeleton.

VESSELS

The main blood supply to the lip and nose area comes from the facial arterial branch of the external carotid artery. Auxiliary sources come from the ophthalmic and the infraorbital arteries. The facial artery gives off inferior and superior labial branches which arise near the corner of the mouth and course as the coronary vessels close to the free border of the upper and lower lips deep to the muscle and close to the mucous membrane. In the upper and lower lip, the right and left labial arteries freely anastomose to form a circle surrounding the oral aperture. The facial artery then proceeds upward along the nasolabial fold and at the ala gives off the lateral nasal branch and then becomes the angular artery proceeding up to anastomose with the dorsal nasal artery, a branch of the ophthalmic. Meanwhile, the posterior septal artery arising from the sphenopalatine artery in the roof of the nasal cavity courses down the vomerine groove to the incisive foramen, anastomosing with the major palatine and ascending septal branches of the superior labial arteries.

Near the inferior lateral attachment of the ala, the lateral nasal artery splits to run one branch along the lower border and another along the upper border of the lower lateral cartilage. These branches anastomose in the midline with the terminal branches of the anterior ethmoidal arterial extension of the internal carotid artery. The anterior ethmoidal artery comes through the cribiform plate of the ethmoid bone, enters the nose and passes along the undersurface of the nasal bone arch, continuing distally over the upper lateral cartilages to the tip of the nose. Here it joins the lateral nasal branches to continue into the columella, anastomosing with the ascending septal branches of the superior labial artery.
Fara of Charles University, Prague, in his dissections of three incomplete and four complete unilateral clefts found the arterial networks generally coursing along the edge of the cleft upward parallel with the muscle fibers and stronger on the lateral cleft side than on the medial philtrum side. In incomplete clefts the vessels crossed the bridge always from the lateral side.

Slaughter, Henry and Berger dissected out the blood vessels in clefts to demonstrate the variation from the normal, and their 1960 vascular patterns, with slight corrections, have been used as a guide for these anatomical drawings. Although there is an interruption in the usual arcade in the upper lip in unilateral clefts, there is sufficient blood supply to both lip elements and the nose to allow surgery without slough and with the expectation of adequate healing.
NERVE SUPPLY

The sensory nerve supply to the involved areas of the lips and nose comes from branches of the fifth or trigeminal nerve surfacing through the infraorbital foramen as the infraorbital nerve and through the mental foramen as the mental nerve.

The motor nerve supply to the muscles of the lips and nose comes from the seventh or facial nerve through its zygomatic, buccal and mandibular branches.

Of course, the presence of a cleft through the lip musculature interrupts the normal course of the nerve fibers, but they do extend to or influence the muscles up to the edges of the cleft. As the field of action is involved mostly with the terminal branches of these nerves, their significance in the surgery is limited.
SURFACE ANATOMY

The surface anatomy of the lip and nose is also important in the planning of surgery. It was Leonardo DaVinci, artist, anatomist, sculptor, architect, biologist and engineer, who during the Italian Renaissance, among other things, divided the face into three equal parts—the forehead from hairline to brow, the nose from its root to its base and the lips and chin from nasal base to inferior border of the mentum. These proportions with slight variations are essential for a normal face.

In the normal, the columella stands as a graceful central column, straight and narrow right up to the proud nasal tip. At its base, the columella flows as a nostril sill across in front of the nasal floors toward non-flaring alar bases. The arches of the alae are symmetrical, with equal bulges of the alar cartilages in the nasal tip.

The ideal length of the upper lip at rest places its inferior edge at the lower one-third of the upper incisor teeth. As the upper lip rises, more of the incisors are revealed until with smiling there is three-fourths to total incisor exposure.

The eversion of the upper lip places it slightly out in front of the lower. At the mucocutaneous junction of the upper lip, there is an uninterrupted 1 to 2 mm. rounded roll from com-
misure to commissure which tops the vermilion and picks up a white light. It coincides in its curves with the undulations of the cupid's bow of the vermilion, which has a central free border tubercle flanked by slight indentations. From the height of each arch of the bow, the philtrum columns curve upward toward the base of the columella. Between these columns is a philtrum hollow or dimple which accentuates the effect of the eminences.

Human noses vary according to race, sex and circumstance as to the straightness of the septum, height and width of the bridge, position and shape of the tip, shape and size of the nostrils, position and thickness of the alar bases and length and width of the columella.

Human lips also vary with race, sex and circumstance in length and width, muscle strength, amount of expression, degree of hair bearing, curve of the cupid's bow, width of the muco-
cutaneous "white roll," depth of the philtrum dimple, height and direction of the philtrum columns, fullness of the vermilion, strength of the tubercle and amount of eversion.

MEASURING THE NORMAL

Farkas and Lindsay measured 100 normal Canadians, 50 boys and 50 girls from the ages of 16 to 20 years. They found:

<table>
<thead>
<tr>
<th>Measurement</th>
<th>Male</th>
<th>Female</th>
<th>Mean</th>
</tr>
</thead>
<tbody>
<tr>
<td>Length of columella</td>
<td>10-16 mm.</td>
<td>9-15 mm.</td>
<td>12.3 mm.</td>
</tr>
<tr>
<td>Width of columella</td>
<td>7-10 mm.</td>
<td>6-9 mm.</td>
<td>8.2 mm.</td>
</tr>
<tr>
<td>Lateral vertical length of lip</td>
<td>11-21 mm.</td>
<td>10-20 mm.</td>
<td>16.4 mm.</td>
</tr>
<tr>
<td>Medial vertical length of lip</td>
<td>18-26 mm.</td>
<td>16-24 mm.</td>
<td>22.0 mm.</td>
</tr>
</tbody>
</table>

The normal Canadian measurements for medial vertical length of lip were similar both to those of central European norms, which Hajnisova found to average 21.2 mm. in the male and 19.2 mm. in the female, and to those of west European norms, which Hajnis found to average 21.5 mm. in the male and 20.5 mm. in the female.

Michael Franz and Anthony Sokol of Columbus, Ohio, measured 40 normal philtrums from peak to peak of the cupid's bow and the total distance from commissure to commissure. They then computed the philtrum-commissure ratio to be the commisural distance divided by a factor of 3.75 to provide the measurement of the proposed philtrum. More simply, the width of the philtrum should be slightly more than one-fourth of the width of the mouth.
Fully aware of the significance of *knowing the normal*, my residents, with Gaston Schwarz, a Molson Foundation Fellow in training from Montreal, as principal investigator, have taken calipers into their homes, kindergartens, hospital nurseries and wards to compare the key distances in the lip-nose complex at various ages. They have used the calipers on each other and have shown exceptional diligence in measuring the attractive secretaries, nurses and patients of our plastic surgical division. The numbers may not be of great statistical significance, but they follow a trend that might be expected.

**Racial Comparisons**

Of course, there are normal differences in races other than skin color, and in the nasolabial area these can be important in the design of cleft correction. A comparison of nose and lip measurements in Caucasian and Negro males and females at various ages reveals, in spite of specific variations, some general consistent differences. For simplicity, all measurements have been rounded off to the nearest tenth of a centimeter.
Averages in cm.

<table>
<thead>
<tr>
<th>Age</th>
<th>Newborns</th>
<th>5 Years</th>
<th>Adults</th>
</tr>
</thead>
<tbody>
<tr>
<td>Race</td>
<td>Caucasian</td>
<td>Negro</td>
<td>Caucasian</td>
</tr>
<tr>
<td>Sex</td>
<td>M</td>
<td>F</td>
<td>M</td>
</tr>
<tr>
<td>Nose</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1. Columella height</td>
<td>0.4</td>
<td>0.4</td>
<td>0.3</td>
</tr>
<tr>
<td>2. Columella width</td>
<td>0.4</td>
<td>0.5</td>
<td>0.4</td>
</tr>
<tr>
<td>3. Nasal width</td>
<td>2.2</td>
<td>2.3</td>
<td>2.3</td>
</tr>
<tr>
<td>Lip</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. Vertical height (base of columella to tubercle)</td>
<td>1.5</td>
<td>1.2</td>
<td>1.4</td>
</tr>
<tr>
<td>5. Vertical height (nasal base to cupid's bow peak)</td>
<td>1.1</td>
<td>1.0</td>
<td>1.1</td>
</tr>
<tr>
<td>6. Width (cupid's bow to commissure)</td>
<td>1.5</td>
<td>1.5</td>
<td>1.6</td>
</tr>
<tr>
<td>7. Width (philtrum, peak to peak)</td>
<td>0.7</td>
<td>0.7</td>
<td>0.8</td>
</tr>
<tr>
<td>8. Width (entire mouth, commissure to commissure)</td>
<td>3.3</td>
<td>3.5</td>
<td>3.8</td>
</tr>
<tr>
<td>Total number of cases</td>
<td>12</td>
<td>12</td>
<td>10</td>
</tr>
</tbody>
</table>

GENERALIZATIONS ON CAUCASIAN AND NEGRO MEASUREMENTS

*Columella:* There is no real difference in height at birth, but with growth there is a greater increase in height and width in Caucasians.

*Nasal width:* In the Negro the nose is only slightly wider at birth but with growth becomes considerably wider.

*Lip length:* Negro and Caucasian are close in lip length at birth, but with growth there is more elongation in the Negro, especially in females. It is of interest that our Caucasians resemble the Canadian and European Caucasian in vertical lip measurements.
**Mouth width:** Sexes and races are close, but total mouth width is greater in the male Negro at birth and as an adult.

**Philtrum:** The races and sexes are close at birth, but Caucasian adult males have wider philtrums than Negro males.

**Comparisons with Orientals**

Curiosity about comparison of the Caucasian and Negro with the Oriental prompted me to ask my friend Khoo Boo-Chai in Singapore to measure some Chinese lips and noses. His measurements unfortunately were carried out on postoperative clefts, but as the normal side was measured there probably can be some correlation.

<table>
<thead>
<tr>
<th>Age</th>
<th>3 Months</th>
<th>5 Years</th>
<th>Adults</th>
</tr>
</thead>
<tbody>
<tr>
<td>Race</td>
<td>Chinese</td>
<td>Chinese</td>
<td>Chinese</td>
</tr>
<tr>
<td>Sex</td>
<td>M</td>
<td>F</td>
<td>M</td>
</tr>
<tr>
<td>Nose</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1. Columella height</td>
<td>0.4</td>
<td>0.4</td>
<td>0.5</td>
</tr>
<tr>
<td>2. Columella width</td>
<td>0.5</td>
<td>0.5</td>
<td>0.5</td>
</tr>
<tr>
<td>3. Nasal width</td>
<td>3.1</td>
<td>3.0</td>
<td>3.5</td>
</tr>
<tr>
<td>Lip</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. Vertical height (base of columella to tubercle)</td>
<td>1.2</td>
<td>1.1</td>
<td>1.3</td>
</tr>
<tr>
<td>5. Vertical height (nasal base to cupid's bow peak)</td>
<td>1.2</td>
<td>1.2</td>
<td>1.2</td>
</tr>
<tr>
<td>6. Width (cupid's bow to commissure)</td>
<td>1.7</td>
<td>1.7</td>
<td>2.5</td>
</tr>
<tr>
<td>7. Width (Philtrum, peak to peak)</td>
<td>0.7</td>
<td>0.7</td>
<td>0.8</td>
</tr>
<tr>
<td>8. Width (entire mouth, commissure to commissure)</td>
<td>3.3</td>
<td>3.4</td>
<td>4.1</td>
</tr>
<tr>
<td>Total number of cases</td>
<td>7</td>
<td>3</td>
<td>7</td>
</tr>
</tbody>
</table>

Averages in cm.
Columella length and nasal width of the Chinese at five years are closer to those of the Negro and in the adult fall between those of the Caucasian and Negro.

In the Chinese lip length is shorter and mouth width is smaller than in either Caucasian or Negro, but the philtrum width is about equal to that of both Caucasian and Negro.

It is possible that these slight but consistent differences may explain, in part, the rash of variations appearing in the Oriental cleft lip literature.

A BUILT-IN NORMAL

In the unilateral cleft at least the surgeon has the non-cleft side as a guide to this individual norm and should use it as such.
3. The Naming and Classifying of Clefts

A CLEFT BY ANY OTHER NAME

The cleft anomaly may be more ancient than man himself, but the early evidence is sparse. Ortiz-Monasterio with Serrano found a ceramic sculpture of a chief with a cleft of the lip from Nayarit on the west coast of Mexico, which seems to date from approximately A.D. 12.

The Greek physician Galen, near the end of the second century, about A.D. 170, mentioned cleft lip, applying to it the term "colobomata."

One thousand years before the Inca Indian empire of the Andes, the Mochica culture flourished on the northern coast of Peru. Almost 2,000 years ago its people portrayed realism in their ceramic art, specializing in the human face, its expressions and deformities and the human genitalia, sexual positions and "perversions." Alberto Carrion Vergara, a plastic surgeon who does many of the cleft lips that go down to Lima from the Andes, forwarded pictures of ceramic sculptures of clefts from the Museo Arqueologico of Lima dating from A.D. 200 to 400.

One specimen is a small red-brown and white figure with a central lip and nose deformity which appears to be a median cleft but could represent the ravages of leishmaniasis or punishment for adultery by mutilation.

Another is a black stirrup spout vessel portraying with remarkable accuracy a bilateral cleft lip with a small prolabium.
The third is also a red-brown and white stirrup spout vase depicting perfectly the unilateral lip cleft with exposure of the distorted maxilla and a malpositioned tooth, a philtrum dimple and the typical nasal deformity.

It seems that the earlier Peruvian cultures might have missed an opportunity. Had they capitalized on the midline lip cleft and flat nose of their beast of burden, the woolly-haired ruminant, this anomaly today might enjoy the melodious label of "llama lip."

Instead, by A.D. 390 cleft lip was being referred to as "harelip" or T’u Ch’ueh as written in the original annals of the Chin Dynasty, compiled during the early Tang Dynasty and in 1966 translated by Khoo Boo-Chai. Through the centuries, the cleft anomaly consistently has been called T’u Ch’ueh, lagocheilos, bec-de-lièvre, hasenscharte, labis leporino or harelip. Indeed, there is a striking resemblance between the lip of the hare with its midline fissure exposing two prominent upper incisors and the congenital cleft of the lip, rarely midline but unilateral and bilateral, revealing incisor teeth. This haunting similarity is not limited to the lip alone as the flat nasal tip, short columella and flaring nasal alae blend into the unfused lip without a nasolabial angle to present an actual harelook.

Somewhere along the line a translator missed the point by a hair for by A.D. 950 in the Leech-Book the cleft anomaly was being referred to as hairlip. This spelling evidently was retained at least in Anglo-Saxon Britain until the fourteenth century. Although there is still a controversy raging as to how this unexplained and illogical reference ever occurred, it can most certainly be dismissed as a translator’s error. The same misspelling of "hare" as "hair" is seen even today on cleft lip hospital charts.

In fact, my introduction to plastic surgery as a student at Harvard Medical School was begun by a fascinating lecture with slides by Donald W. MacCollum. During his entire demonstration a distracting display remained at the front of the class: a cage containing a pair of large white rabbits. Each dutifully gnawed on a carrot with its exposed incisors to emphasize that harelip, not hairlip, was the present synonym for cleft of the lip.
The resemblance of the anomaly to the hare has now been noted but is not considered sufficient justification for continuing the comparison of a cleft lip infant and a burrowing rodent. Out of deference to patients and parents, the label of harelip, except in quotations, will not hereafter be perpetuated in these pages.

The influence of inheritance on the incidence of these congenital clefts might better justify the title heredit. Fogh-Andersen stated in 1967 and confirmed in 1971:

With our present knowledge, we still must consider heredity the most important etiologic factor in typical cleft deformities, for CL(P) probably in 40 to 50% and for CP in 20 to 25%.

The more severe deformity of bilateral cleft lip and palate with a projecting premaxilla is often referred to as "wolf's snout." Here again, it is mentioned only to be dismissed.

THE CLASSIFICATION DILEMMA

There is a place for an anatomical and embryological classification of cleft lip, alveolus and palate deformity. Many systems have been offered but none has been universally accepted because of language differences, inaccuracies, omissions and lack of simplicity.

At the American Medical Association meeting in St. Louis in 1922, John Staige Davis of Baltimore and Harry P. Ritchie of St. Paul, after years of collaboration, proposed a new classification. This must have been a hotly contested presentation as in the assembly were other experts on lip and palate work such as J. E. Thompson of Galveston, T. Brophy of Chicago and, of course, V. P. Blair, each with his own ideas of classification. For the sake of uniformity and to facilitate communication and comparison, Davis and Ritchie submitted a grouping that proved to be the best of its time and was used for many years.
THE DAVIS AND RITCHIE
THREE GROUPS

This classification recognized three major types of deformity: cleft lip = group I, cleft palate = group II and cleft of both lip and palate = group III. Unilateral, bilateral and median variants of these three main groups were indicated by further numbers so that a cleft of the lip unilateral would be group I, 1, a cleft of the lip median would be group I, 2 and a cleft of the lip bilateral would be group I, 3.

Clefts of the palate alone, or group II, were subdivided into uvula and soft palate as 1 and hard palate as 2, and each of these two subgroups was further divided into $\frac{1}{3}$, $\frac{2}{3}$ and $\frac{3}{3}$ cleft.

Clefts of lip, alveolus, hard palate and soft palate, or group III, were subdivided into unilateral = group III, 1, median = group III, 2 and bilateral = group III, 3.

At the time there was heated discussion of the Davis-Ritchie grouping. James Thompson said:

If a classification is to be accepted, it must not alone have an anatomic or embryologic foundation but it must be of practical surgical value.

Ritchie had indicated that the alveolar border was the basis for all surgical groupings, and from a practical point of view he was not far off. Brophy gave his own grouping as 16 somewhat unrelated items which, as Ritchie pointed out, could be fitted into their classification. Blair approved of Ritchie’s plan but astutely suggested that surgery of the palate possibly had not progressed far enough to make didactic classification. He also made a plea that the classification finally chosen have universal acceptance. Actually it did not for 45 years.

The Davis-Ritchie classification is no longer in vogue. It did not subdivide the various degrees of incomplete clefts. As explained by Stark,

If the demarcation point were moved backward from the alveolus to the incisive foramen, the Davis and Ritchie classification would be comprehensive, embryologically correct and surgically acceptable.
VEAU’S FOUR GROUPS

In 1931 Veau classified degrees of deformity by a simple numerical scale: clefts of the soft palate = group I, clefts of the soft and hard palate = group II, unilateral complete clefts of the alveolus, hard and soft palate = group III and bilateral complete clefts of the alveolus, hard and soft palate = group IV. Although he ignored clefts of the lip and alveolus completely, his classification had many ardent followers such as Kilner and still is referred to often even today.

FOGH-ANDERSEN’S THREE GROUPS

In his 1942 monograph, Inheritance of Harelip and Cleft Palate, the sagacious Poul Fogh-Andersen of Copenhagen described his morphological classification of cleft lip and palate based on embryology and genetics. He divided clefts into three main groups:

1. Harelip (single or double) including all degrees from a small notch in the probium to a complete cleft of the lip extending “as far as the incisor foramen.” When the cleft was bilateral through lip and alveolus, he noted, “There is prominence of the premaxilla.”

2. Harelip and cleft palate, which is the largest group. He noted complete clefts from nostril to uvula and others broken
by osseous and skin bridges. This group included single and double clefts.

3. *Cleft palate.* This group included isolated cleft palate which he noted may involve the soft palate or the soft and hard palate and "is always median and it never reaches further than the incisor foramen." Fogh-Andersen also included the *submucous cleft* presenting a cleft in the soft palate but only a bony cleft of the hard palate with intact oral and nasal mucous membrane.

4. A group of rare atypical clefts was optional.

As Fogh-Andersen wrote in 1965:

This classification has been adopted later by a series of writers, and Kernahan and Stark's classification in 1958 is in reality identical with it.

**Kernahan and Stark Reduce It to Two Groups**

Kernahan and Stark's 1958 classification emphasized the embryological basis of the incisive foramen's being set as the boundary marker. Clefts of the lip and premaxilla, occurring at four to seven weeks of embryonic life, were termed *clefts of the primary palate.* Clefts of the hard and soft palate posterior to the incisive foramen, occurring at 7 to 12 weeks, were termed *clefts of the secondary palate.* Then further description, such as left and right, complete and incomplete, was added.
It is significant that the International Confederation of Plastic Surgeons accepted this classification in 1967, and many use it today. The fact that lip is not mentioned in the terminology reduces its popularity. Then, too, Conway, McKinney, Climo, Hugo, Cole and Goulian in 1968 used the Kernahan-Stark Classification on 850 clefts and found they had to add subcategories.

**HARKINS**

In 1962 Harkins, Berlin, Harding, Longacre and Snodgrass, for the American Cleft Palate Association, proposed three main cleft groups: (1) *prepalate* included unilateral, bilateral and medial lip clefts as well as congenital scars and all variations of alveolar clefts; (2) *palate* included all forms of palate cleft forward as far as the incisive foramen; (3) *prepalate and palate* included unilateral and bilateral complete clefts but also the combination of incomplete clefts of lip and palate. They then added a group (4) of rare clefts other than prepalate or palate types. These included the various facial clefts and lower lip clefts, which were charted in a diagram.

**VILAR-SANCHO**

Another interesting approach is that of Spanish plastic surgeon Vilar-Sancho, who in 1962 classified all clefts (SK) as incomplete (small letter) or complete (capital) using the appropriate letter of the Greek word for the area involved: K for *kilos* (lip), G for *gnato* (maxilla), U for *urano* (hard palate) and S for *stafilos* (velum). After the letter of the location and cleft extent, the side affected is indicated with d for right, l for left and s for bilateral. As pointed out by Stark, the declining popularity of classical languages in the Western world makes this impractical. It would, in fact, be “greek” to most of us today.
SCHUCHARDT

Professor Karl Schuchardt contrived a visual symbol to facilitate indexing cleft lip and palate cases for his Northwest German Jaw Clinic in Hamburg in 1964. One cannot but admit it has appeal, and certainly a symbol has possibilities.

INTERNATIONALLY APPROVED CLASSIFICATION

The subcommittee on cleft lip and palate nomenclature of the International Confederation for Plastic and Reconstructive Surgery announced in the newsletter following the 1967 Rome Congress the official Confederation cleft classification.

Classification of Clefts of the Lip, Alveolus and Palate
(classification based on embryological principles)

Group 1: Clefts of anterior (primary) palate:
(a) Lip: right and/or left.
(b) Alveolus: right and/or left.

Group 2: Clefts of anterior and posterior (primary and secondary) palate:
(a) Lip: right and/or left.
(b) Alveolus: right and/or left.
(c) Hard palate: right and/or left.

Group 3: Clefts of posterior (secondary) palate:
(a) Hard palate: right and/or left.
(b) Soft palate: medial.
(For further subdivisions the terms "total" and "partial" should be used.)

Rare Facial Clefts
(classification based on topographical findings)

(a) Median clefts of upper lip with or without hypoplasia or aplasia of premaxilla.
(b) Oblique clefts (oro-orbital).
(c) Transverse clefts (oro-auricular).
(d) Clefts of lower lip, nose and other very rare clefts.

This was, in fact, the 1942 classification of Fogh-Andersen
but also confirmed the 1958 primary and secondary palate grouping of Kernahan and Stark.

In 1972 V. Spina, J. M. Psillakis, F. S. Lapa and M. C. Ferreira of São Paulo concurred with the Fogh-Andersen, Kernahan and Stark classification of clefts centered on the incisive foramen. In fact, they suggested the identical classification but went one step farther to include the incisive foramen in the actual terminology, grouping clefts as I, pre-incisive foramen clefts; II, trans-incisive foramen clefts; III, post-incisive foramen clefts; IV, rare facial clefts.

THE ‘‘Y’’

Desmond Kernahan had his early training in cleft surgery with Kilner at Oxford and later with Osborne and Burston in Liverpool. Then he joined Stark in New York to create their embryologically based classification. Kernahan finally landed at the University of Manitoba in Winnipeg, where in 1971 he noted . . . that one recurring problem in a cleft palate clinic is the size of the charts of these patients. . . .

He ingeniously simplified the cleft record-taking, from the least to the greatest defects, in what seems to me to be the best and most practical method yet. It is interesting that Kernahan, a clock collector, has switched the principle and set his dials on top of three interconnecting hands. As he explained, the bilateral total cleft of the primary and secondary palates

can be represented as a Y. The dividing point between the primary and secondary palates—namely, the incisive foramen—can be represented symbolically at the junctions of the limbs of the Y by a small circle.

The right and left limbs of the “Y” are divided into three sections: the anterior portion = lip (1 and 4), the middle = alveolus (2 and 5) and the posterior = the area of the hard palate from the alveolus back to the incisive foramen (3 and 6). Posterior to the incisive foramen, the hard (7 and 8)
and soft (9) palate are also divided into three segments. This segmented Y can be reproduced by a rubber stamp.

Kernahan elaborated:

To facilitate data processing in the cleft palate clinic, we have assigned a number to each of these subdivisions. This gives us a striped Y . . . .

The method is adaptable. Cleft areas are indicated by stippling the respective segments. Submucous clefts of the palate are indicated by horizontal lines where a true cleft is not present. A Simonart's band at the threshold of the nostril is indicated by horizontal lines across the most anterior portion of the respective limb of the “Y”.

When asked how he charted median clefts of the lip, Kernahan explained in 1972:

So far as median clefts are concerned, we have simply drawn in a straight line centrally between one and four on the striped “Y” for true central lip clefts and in cases where the whole primitive palate is absent as in trisomy and arhinencephaly we have blocked in the entire area between the two limbs of the “Y.”

Lest complacency set in, Khoo Boo-Chai of Singapore, aided and abetted by Ichiro Tange of Tokyo, conjured up a new clinical subgroup of the cleft lip deformity, that of the isolated cleft
lip nose, which they suggested must be included in any comprehensive classification along with the usual minimal cleft lip with congenital scar. Kernahan was quizzed on this point and he suggested that the isolated cleft lip nose be indicated on the Y as a lazy S at the junction of the lip and alveolar segments.

**STRETCHING THE "Y"**

Then in 1972, just as those interested in clefts were settling down to this sound classification with its nice, neat, symbolic representation, Egyptian Nabil Elsahy, also from Winnipeg, Manitoba, dared to offer extensions which deserve consideration. He added triangular peaks (1 and 5) to the ends of the prongs to represent the nasal floor in case of incomplete clefts of the lip. This modification caused a shifting of the numbers in the squares with the lip represented by squares 2 and 6, alveolus by 3 and 7, hard palate anterior to the incisive foramen by 4 and 8, hard palate by 9 and 10 and soft palate by square 11. To indicate a vermilion notch on the left a narrow band of stippling is placed in the lower portion of square 6 while an alveolar notch on the same side has a band of stippling in the upper portion of square 7.

Collapse of the maxillary segments is indicated by filling in or stippling 3 and 4 or 7 and 8. Elsahy double-lined the squares (9 and 10) in the hard palate area and used arrows to indicate direction of deflection in complete clefts. He placed a circle (12) under the stem of the Y to represent the pharynx. Then with a dotted line from the Y to the O reflecting velopharyngeal competence, any break in continuity of this dotted line could be interpreted as the amount of incompetence. Elsahy also added circle 13 to represent the premaxilla, and the amount of its protrusion is indicated by the dotted line with an arrow and the position of circle 13.

An added value of this modified striped Y is that its symbolic representation, embryologically, clinically and physiologically, of
the cleft lip and palate deformity allows simple charting of the progress of the patient, not only before but during and after treatment, by mere comparison of the consecutive Y’s.

Then it became apparent that there is no reason not to top Elsahy’s triangular tips to Kernahan’s prongs with inverted tips to be marked with horizontal lines indicating the amount of nasal deformity, ad infinitum.

In fact, a modified Y is being used in this volume as a simple preoperative case record. The Y will be capped with triangular peaks for the nasal floor, and these will be topped with similar triangular peaks turned upside down to represent the nasal arch. To indicate a cleft the area will be stippled, to indicate submucosal muscle and bony clefts, the area will be marked with horizontal lines, and to indicate the degree of nasal deformity the top triangle will be lined horizontally in density proportionate to the severity of the distortion.

THE DANGERS OF BUREAUCRACY

After full consideration of all efforts to group and number these anomalies, it is thought that the Y, striped, stippled, peaked or with a ball up and a ball down, in basic accuracy and endearing simplicity, offers the best plan yet for charting clefts in medical records.

It is further suggested that neither llama, hare, hair, heir, group I, 1, cleft of the primary palate nor KL, SK describes this portion of the anomaly as accurately and unemotionally as the simple term “cleft lip.” Let us pursue Ivy’s plea for universal acceptance of cleft lip, complete and incomplete, unilateral and
bilateral, left and right. It is even possible that the simple blanket term “cleft lip,” further described as unilateral, complete and right, is still too bureaucratic in its classification. As operating plastic surgeons, we must look at each case not as one of a group or part of a series but as an individual with its own minutely varied detail.

A SUGGESTION OF REPAIR

Closure of lip clefts is most commonly referred to as repair of cleft lip, but the word “repair” suggests that the lip was once intact, has separated and must be repaired. Webster defines the verb “repair”: 1. To restore to a sound or good state after decay, injury. . . . 3. To remedy, heal, . . . or mend; as, to repair a break, a wound. . . .” If we consider that the embryological processes become denuded of their epithelium, fuse and later split asunder, then our surgery could be spelled repair or better repare. As there is no proof of this event and as the cleft appears without evidence of previous soundness, I have avoided the word “repair” whenever possible except in others’ quotations.

A NOTE ON THE PHOTOGRAPHIC RECORDING OF CLEFTS

Sir Harold Gillies opened the First International Congress of Plastic Surgery in Stockholm in 1955, touching lightly on the development of this specialty through the years and reminiscing on what “the ancients and the not so ancients” had achieved in their plastic surgery. He concluded puckishly by whispering that the one most important factor responsible for modern improvements in results was “photography.”

It is true that photography can “make” or “break” a plastic surgeon. Even though plastic surgeons are knowledgeable enough not to be fooled consistently by photographic tricks, final results continue to be presented with the benefit of favorable effects. Bright flat lighting “burns out” the scars to invisibility and flattens unnatural contours while the position and angle of recording hide asymmetries. Although photographs can flatter
and deceive, they can also nullify a result by flattening normal contour, highlighting scars and exaggerating distortion. For example, here are three unretouched photographs of the same patient, evidently treated with a modified LeMesurier procedure, taken consecutively within a few minutes of each other with the same camera, with the same lens and by the same photographer, Jim Fletcher.

The first (A) is an honest record of the actual appearance of the patient as seen and as we have tried to portray the cases in this book. The second (B) exaggerates the surgical scars, and the third (C) wipes them out to such a degree that one might think remarkable surgery has been performed. According to Fletcher, the most deceiving of all photographic recording is that following reproductions from overexposed color transparencies which show no scars at all.

In the early days I took my own pictures. Then in Korea, fortunately the services of Marine photographer Brusseau became available. In 1960 John Madge, originally a baby photographer, joined my staff, and finally, in 1971 Jim Fletcher took over and has been responsible for most of this volume's final photography. Unfortunately, one photographer has not been recording from the beginning to the end, but great effort has been made to photograph accurately, and with babies this is no easy matter.

Consistent front, profile and subnasal views, although ideal,
have not always been available. Yet even these do not tell the complete story for it is impossible to judge a result from one still shot. A true evaluation must be live and in color, observing the combined actions of lips and nose in various positions from absolute stillness up and down the entire expression gamut from laughing to crying. Nevertheless, it is hoped that the photographic records presented will at least provide a clue or confirm a claim.
4. Incidence of Clefts in the World

<table>
<thead>
<tr>
<th>Date</th>
<th>Source</th>
<th>Location</th>
<th>Incidence</th>
<th>Ratio to Normal Births</th>
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<td>Mummies</td>
<td>Egypt</td>
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<td>Hamburg, Germany</td>
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<td>Finland</td>
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<td>1960</td>
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<td>1961</td>
<td>Traverso</td>
<td>Montana</td>
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<td>1961</td>
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<td>Africa</td>
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<tr>
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<td>Curtis</td>
<td>Canada</td>
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<td>Trinidad</td>
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<td>1972</td>
<td>Carlisle</td>
<td>Phoenix</td>
<td>1:515</td>
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</tbody>
</table>

If all the microforms could be included, then undoubtedly these percentages would be altered.
METHODS OF RECORDING

The discrepancies in the incidence of clefts in the different countries may be due in some part to the methods of recording. In Denmark, the fact that all of these defects must be reported to the National Institute of Speech Defects accounts for the accuracy of Danish statistics. In Finland, 99 percent of clefts are operated on at the Finnish Red Cross Hospital. In Pennsylvania, U.S.A., all defects must be recorded on the birth certificate, and yet Ivy showed that in spite of this regulation only 83.3 percent of cleft lip and palate deformity were recorded. Imagine the discrepancies elsewhere!

INCIDENCE

In all the ancient tombs of Egypt only one mummy with a cleft palate has been dug up, suggesting an incidence of about 1:1,000 during the years 4000 to 2000 B.C. But not just anybody could get pickled and swathed, so this probably eliminates the lower income population and reduces the recorded incidence. If, on the other hand, the ingenious Egyptians made a special effort to mummify clefts, the apparent incidence may be too high.

THERE IS A DIFFERENCE IN RACIAL INCIDENCE

Caucasian

<table>
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<th>Date</th>
<th>Author</th>
<th>Incidence</th>
<th>Ratio to Normal Births</th>
</tr>
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<td>Woolf</td>
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<tr>
<td>1965</td>
<td>Longenecker</td>
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<td>1:433</td>
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<tr>
<td>1967</td>
<td>Chung</td>
<td>30:16,385</td>
<td>1:546</td>
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<tr>
<td>1971</td>
<td>Fogh-Andersen</td>
<td>150:75,000</td>
<td>1:500</td>
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</tbody>
</table>

In countries where the population is of Caucasian origin, the incidence is now generally between 1:500 and 1:600.
Although there are very few studies from Africa in those series reported, the incidence of cleft lip and palate seems less common. The practice of infanticide in many uncivilized tribes of Africa would certainly tend to reduce the incidence. Oluwasanmi reported 128 cases from Ibadan in Nigeria, but in general it is difficult to get records with infanticide in progress and with the unwillingness of the family to give a history of a cleft because of the stigma attached thereto.

**New World Negroes**

The low incidence of cleft lip and palate in the American Negro has often been noted. In 1924 in Baltimore, John Staige Davis reported an overall incidence of 1:1,170, crediting the low incidence on the large number of Negroes in his area. He showed a much lower incidence in the Negro (1:1,790) than in the Caucasian (1:915). In 1965 Longenecker in New Orleans corroborated Davis’ findings with an incidence of 1:1,553 in Negroes compared with 1:692 in Caucasians. Altemus reported 1:2,218 in Negroes.

**Caribbean islands**
Robinson's Trinidad is composed of 54 percent Negro-mixed and 36 percent East Asian Indian. Although the total incidence of cleft lip and palate was 1:857, the East Asian Indian incidence was 1:500 and the Negro-mixed was 1:1,888.

McNeill and I studied the percentage of clefts in the 56,256 births at Victoria Jubilee Maternity Hospital in Kingston, Jamaica, from 1960 through 1963. At least 90 percent of these births were Negro or Negro-mixed, and the incidence over these four years is of interest: lip only, 1:6,250; palate only, 1:9,091; lip and palate, 1:3,704; all types combined, 1:1,875.

**Logical reasons for low incidence in New World Negroes**

INFanticide. It has been suggested that in the uncivilized tribes of Africa the practice of destroying all deformed newborns would have a discouraging effect on the propagation of clefts.

SELECTIVE SPECIMENS. The reduced incidence of cleft lip and palate has been demonstrated only in the Negroes of North America, Trinidad and Jamaica. These are a selective group, having been chosen originally as excellent physical specimens to bring a good price in the slave market. It is unlikely that the avaricious slave traders would have taken up the space in their ships to transport cleft lip and palate slaves who would be certain to demand a lower sales value.

SURVIVAL OF THE FITTEST. Infants with the more severe cleft deformities, particularly as seen in the bilateral type, have great difficulty with breast feeding. In uncivilized areas or even in underdeveloped countries where breast feeding is the only source of food for the newborn, the chance of survival for infants with such clefts is slim. Even in Jamaica, when a child with a severe cleft is seen at the initial visit at Kingston Public Hospital, he is extremely malnourished. This infant is usually one of a large poor family, and if the cleft is not closed early the child often does not survive.

**Confirmation in reverse**

As will be noted later, in the New World Negro the entity cleft palate alone is relatively more common than in the Caucasian. This finding can be explained by the same logic in reverse. A
simple palate cleft unnoticed at birth would spare the infant's life, unnoticed in the slave mart would not affect the sale and if not severe would allow enough breast feeding for survival.

**Japanese**

<table>
<thead>
<tr>
<th>Date</th>
<th>Author</th>
<th>Incidence</th>
<th>Ratio to Normal Births</th>
</tr>
</thead>
<tbody>
<tr>
<td>1958</td>
<td>Neel</td>
<td>171:63,796</td>
<td>1:373</td>
</tr>
</tbody>
</table>

In 1958 Neel reported the high incidence of 1:373 in Japanese.

**American Indian**

<table>
<thead>
<tr>
<th>Date</th>
<th>Author</th>
<th>Tribe</th>
<th>Incidence</th>
<th>Ratio to Normal Births</th>
</tr>
</thead>
<tbody>
<tr>
<td>1963</td>
<td>Miller</td>
<td></td>
<td>31:12,337</td>
<td>1:398</td>
</tr>
<tr>
<td>1963</td>
<td>Tretsven</td>
<td>Flathead</td>
<td>27:7,461</td>
<td>1:276</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Montana</td>
<td>14:4,499</td>
<td>1:321</td>
</tr>
<tr>
<td>1966</td>
<td>Niswander</td>
<td></td>
<td>50:25,340</td>
<td>1:507</td>
</tr>
<tr>
<td>1972</td>
<td>Carlisle</td>
<td>Navajo</td>
<td>32:16,495</td>
<td>1:515</td>
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</tbody>
</table>

In 1963 Miller reported that the incidence in American Indians in British Columbia is nearly as high as among the Japanese. Tretsven reported an even higher incidence of 1:276 in Montana Indians.

The high incidence among American Indians has been attributed by Jaffe to the traditional life style of these poor people, who suffer from infectious diseases and widespread malnutrition. There is also considerable inbreeding as marriage outside the tribe is discouraged.

Carlisle in Phoenix noted that the incidence of clefts varied greatly in the different Indian tribes in the southwestern U.S.A.
In the Flathead tribe of Montana there is a very high—1:154—in
cidence of clefts. Any group bearing such a name and producing 
clefts at that rate naturally stimulated our curiosity. H. Wolfgang 
Losken of Pietermaritzburg, South Africa, during his Maytag–McCahill 
Fellowship in Miami, assisted in compiling these data on cleft incidence 
and on his way home stopped off at the Museum of North Arizona in Flagstaff to trace the origin of the name Flathead. According to Catlin, it was the custom of the Chinook to flatten the head of the papoose with a wicker cradle head-board, and the squashed result was considered a mark of distinction and superiority. It was disappointing, however, to find that the people listed in the official reports as Flatheads never practice artificial head flattening. It would be too much anyway to have both a flat head and such a good chance for a cleft!

**Split uvula.** Jaffe examined 944 Navajo school children and found an incredible 1:9 (106 cases) incidence of split uvula. He reported a 65 percent one-fourth cleft, 25 percent one-half cleft and 10 percent three-quarter and total clefts of the uvula.

Shapiro and Cervenka reported a 1:10 bifid uvula in American Indian school children having at least five-sixteenths Chippewa ancestry. A striking positive relationship between prevalence of bifid uvula and percentage of Indian ancestry was observed. In children having thirteen-sixteenths to full-blooded Chippewa ancestry, a remarkable 1:5 incidence of bifid uvula was recorded.

### Hawaii

<table>
<thead>
<tr>
<th>Date</th>
<th>Author</th>
<th>Nationality</th>
<th>Incidence</th>
<th>Ratio to Normal Births</th>
</tr>
</thead>
<tbody>
<tr>
<td>1940</td>
<td>Henderson</td>
<td>Hawaiian</td>
<td>35:18,024</td>
<td>1:515</td>
</tr>
<tr>
<td>1946</td>
<td>Krantz</td>
<td>Chinese</td>
<td>1:1,000</td>
<td></td>
</tr>
<tr>
<td>1958</td>
<td>Kung and Chu</td>
<td>Chinese</td>
<td>1:1,000</td>
<td></td>
</tr>
<tr>
<td>1963</td>
<td>Robinson</td>
<td>East Indian</td>
<td>1:500</td>
<td></td>
</tr>
<tr>
<td>1966</td>
<td>Krantz</td>
<td>Filipino</td>
<td>15:4,249</td>
<td>1:283</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Caucasian blends</td>
<td>16:3,834</td>
<td>1:240</td>
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<tr>
<td></td>
<td></td>
<td>Hawaiian blends</td>
<td>27:7,748</td>
<td>1:287</td>
</tr>
</tbody>
</table>

An incidence of 1:1,000 was reported in the Chinese. Krantz and Henderson in Hawaii found a high incidence of clefts in
Filipinos, Caucasian blends and Hawaiian blends. It was felt that mixed races have a higher incidence than pure races.

Indonesia

Anthony Pelly of Sydney, Australia, passed by Miami in 1973 after a plastic surgical sojourn with the Javanese of Indonesia. He had some interesting statistics. Six babies with clefts are born every hour in Indonesia or 360 clefts in 2½ days. This is the total number of clefts produced in Australia in one year where the ratio of clefts to normal births is 1:600. At Tjipto Hospital, Jakarta, he found the ratio of clefts to be 85 percent CL, 2 percent CLP and 13 percent CP. This again suggests high perinatal mortality in total clefts causing selection by survival of the fittest.

Racial Incidence of Cleft Lip With or Without Cleft Palate

There is considerable evidence to support the genetic independence of cleft lip, with or without cleft palate, as a distinct entity from cleft palate alone (Fogh-Andersen, 1943; Woolf and Broadbent, 1963; Clark Fraser of Canada, 1955; Metrakos, 1943; as well as Tor Goran and Henriksson of Sweden and Palmi Moller of Iceland).

Ivy and Stark feel that cleft lip, cleft palate and cleft lip and palate should be studied separately.

Cleft lip with or without cleft palate is very rare in New World Negroes (Millard, 1:2,344; Altemus, 1:4,696). Robert Ivy stated that cleft lip is six times more common in whites than non-whites.

The percentage of cleft lip in most Caucasian series of clefts is 25 percent, CLP 50 percent and CP 25 percent.

Cleft Palate

Fogh-Andersen reported that 25 percent of clefts in Denmark were cleft palate alone. A similar figure is found in most series consisting of only Caucasians. In the American Negro cleft palate alone is relatively more common. Fifty percent was reported by
Chung and Myriantopoulos and 47 percent by Altemus. Fogh-Andersen reported a 76 percent cleft palate incidence among clefts on the Faroe Islands and 82 percent among Eskimos in Greenland.

GENERALIZATION OF INCIDENCE

A concise guide to the pertinent and approximate facts on the incidence of clefts has been compiled by Lazarus and Ryan of Tulane University School of Medicine in their programmed instruction text on cleft lip and cleft palate.

Cleft lip and cleft palate together occur 1:1,289 live births.
Cleft lip alone occurs 1:1,000 live births, more commonly in the male sex and three times as frequently as cleft palate alone.
Cleft palate alone occurs 1:2,500 live births, with a higher incidence in the female sex.

According to Sesgin and Stark in 1961, the incidence of cleft lip–cleft palate as compared to the frequency of the 10 most common congenital anomalies (and least wanted) is low on the list.

1. Foot deformities
2. Hydrocele
3. Hypospadias
4. Mongolism
5. Cryptorchidism
6. Congenital heart disease
7. Polydactyly
8. Hemangioma
9. Cleft lip and cleft palate
10. Hydrocephalus

CLEFTS ARE ON THE INCREASE

Recent studies show an increasing incidence, particularly over the last century, to 1:500. Poul Fogh-Andersen in 1963 stated that the frequency in the population had doubled in the past 50 years and trebled in the last 100 years. His accurate records in the kingdom of Denmark over the last 30 years have shown a definite rising trend. In 1941 the incidence was 1:770 and in 1971 it was 1:500.
### REASONS FOR INCREASE

1. **Falling perinatal mortality.**

2. **Decreased operative mortality.** Peron quoted a 13 percent mortality rate in the first 10 days of life, and Fogh-Andersen quoted a similar figure. In 1954 Ivy reported that 10 percent died within the first year of life—nearly all had multiple congenital anomalies. In 1962 Lewin reported that of the 5,000 infants with clefts born in Russia in one year, one-third die. Fogh-Andersen reported a reduction to 0.4 percent mortality (3 deaths in 900).

3. **Attendant increase in fertility.** Molsted Pedersen (1964) reported a 1:170 (5 in 853) incidence of clefts in children born of diabetic mothers. Many who not so long ago would have died of grave illnesses like diabetes now have children.

4. **Importance of intermarriage.** Consanguineous marriages may account for rising incidence in small countries such as Denmark, Finland, Greenland and Tasmania. Small communities where marriage outside the tribe is frowned upon may account for the high incidence in American Indians.

5. **Steadily improving operative results.** Books like this, devoted to the study of the evolution of corrective surgery of these anomalies, suggest definite improvement of the methods and their results. This, of course, continues more and more to prevent a cleft from interfering with marriage and possible cleft propagation. As 30 to 40 percent of patients demonstrate heredity for the defect, it is not surprising that the incidence is increasing.
It is probable that the contraceptive pill may take part in the reduction of cleft incidence. Yet, until the geneticists find the true causes of cleft, prevention will remain out of reach. In the meantime, we must accept our part of the blame for the increase and get on with improvements until having a child born with a cleft is of no more concern to a family than having a child with an inguinal hernia.

THE INFLUENCE OF GENETICS

F. Clarke Fraser, Professor of Human Genetics, McGill University, and Director, Department of Medical Genetics, The Montreal Children’s Hospital, always liked mathematics and then, during the first genetics lecture he attended, something clicked. He was halfway through medical school when the argument on the relative importance of heredity and environment grew hot. As the pendulum seemed to be swinging too far toward environment and he felt it was actually a combination of both, he leapt to the defense of genetics. His hypothesis was that environmental agents fired at animals of different constitutions would produce different frequencies of malformations. Hamilton Baxter got him some cortisone with which he produced cleft palates in mice and confirmed his theory when he did, indeed, produce strain differences!

In the Grabb, Rosenstein, and Bzoch’s 1971 book *Cleft Lip and Palate*, Fraser outlined in a chart the chances of parents having children with clefts, assuming that known genetic and chromosomal syndromes had been excluded. Noting the frequency of the defect in the general population to be 0.1% for CL ± CP and 0.04% for CP, he correlated various situations with estimated percentages.

If both parents are unaffected and they have an affected child, the probability that their next baby will have the same condition if,

- they have no affected relatives: 4% in CL ± CP, 2% in CP
- they have an affected relative: 4% in CL ± CP, 7% in CP
- they are related to each other: same as general population
- the affected child has another malformation: 2% in CL ± CP, 2% in CP
If unaffected parents have two affected children, the probability that their next baby will have the same condition is 9% in CL ± CP and 1% in CP.

If one parent is affected and they have no affected children, the probability of the next baby being affected is 4% in CL ± CP and 6% in CP.

If one parent is affected and they have an affected child, the probability that their next baby will be affected is 17% in CL ± CP and 15% in CP.

If both parents are affected, Fraser estimated, assuming a heritability of 80%, the risk for the offspring would be about 60%. Their having one or two affected children increases the risk only slightly above this, presumably because the two affected parents contribute about all the "susceptibility" genes there are.

Distant relatives
It is interesting that affected relatives; outside the parents and siblings, have been found unrelated by Fraser. My cases of unilateral CL ± CP revealed a 20% positive family history. Fraser explains:

20 percent positive family histories is about par for the course and means that you did not go out into the fourteenth cousins (in which case it might have been a 100%) or stopped at first degree relatives (when it might have been about 7 or 8%).

Other malformations
It is puzzling that when there is another major malformation which is not part of a genetic syndrome, the risk becomes smaller for recurrence of the cleft anomaly.

The degree of cleft
According to Carter, the more severe the patient's defect, the higher the recurrence risk with 2.5 percent for unilateral cleft lip to 5-7 percent for bilateral cleft lip and palate.

A feminist trend
Both Carter and Woolf concur that the rate of recurrence is a little higher for females than males.
SURGEONS have disagreed as to the best time to close the lip cleft through the centuries, and the controversy continues in most clinics throughout the world today.

The cleft surgeon of the Chin Dynasty had such strict and prolonged postoperative orders that it is unlikely he would have attempted closure on any but mature and responsible adults.

What illustrations have survived from the works of the sixteenth-century French surgeons such as Paré and Guillemeau suggest that they operated on their patients in later childhood or early adult life.

About 1666 James Cooke of Warwick, England, noted:

'Tis more dangerous to perform upon a grown than young person, though happily perform'd on some of 28 years of age. The younger children are when cut, 'tis the better, yea while Infants, unless they be sick or weak. It's more fitly done in Summer than Winter, in Spring than Fall . . .

To operate in, choose a very clean place, and put the Child in the Lap of a discreet person, and let one stand behind to hold the Head, the Child's Hands being ty'd down, and if possible keep it from Sleep for ten or twelve hours before the Operation, that it may be disposed to Sleep presently after . . . cut both sides of the Hair-Lip with Scissors, so much is needful; after pass through a Needle or two, . . . leaving them in, winding the Thread about, as Taylors do when they stick them on their skirts.

The skillful Dutch surgeon Henrik van Roonhuyzi, in Amsterdam in 1674, recommended surgery on cleft lips at three to four months, warning that if performed prior to this the chances of success were markedly reduced.

Consistent with the general French attitude that early cleft
operations were dangerous and unnecessary, LeClerc in 1701 advised that the operation should not be practiced
upon old nor scorbutic Persons, nor upon young Children by reason that their continual Crying would hinder the Re-union. But if any are desirous that it should be done to these last, they are to be kept from taking Rest for a long time; to the end that they may fall asleep after the Operation, which is thus effected.

As Heister, the founder of scientific surgery in Germany, wrote in 1739:

It has been the opinion of the ancients that it is not safe to perform the operation for the harelip upon infants before they are two years of age or even four or five. The contrary of which is taught by experience from whence we are furnished with instances of infants happily cured of a harelip when they have not been above five or six months old, if they are well in other respects and the operation rightly performed . . .

Even the Boston Evening Post in colonial 1770 put in its two cents’ worth with an editorial comment after reporting two cases, a young man and a child, treated for cleft of the lip by Mr. Charles Hall:

The Impressions these unhappy Sights are apt to make on married Women, should be an Inducement to have this Defect in Nature rectified early in Life, as there are numerous Instances of the Mother’s Affection having impressed her Offspring with the like Deformity.

Dupuytren preferred the second or third month, Sir William Fergusson the end of the first month, while Dieffenbach advised postponement until dentition was accomplished.

Surgeons like Malgaigne and Giraldes approved a very early operation: immediately or soon after birth. Guersant in 1826 reasoned that children can do without the breast for four days and noted that out of seven operations performed immediately after birth he had failed only once, whereas out of seven performed at one month he failed five times. Mason cited the examples of Dawson of Dungannon, who operated on an infant at seven hours, and Douglas of Shatford (1854), who operated successfully at two hours. Blair at Washington University, St. Louis, in 1930 advocated cleft lip closure early:
During the first few days of life, there probably remains some of the immunity to surgical shock which is necessarily present during the process of birth. Operation may be done in the first 24 hours. In our series no deaths have occurred from operations on 24 hour old babies. During the period of jaundice, usually from the fourth to the tenth day, the clotting time may be prolonged and operation is not done in this period. The technique of the operation at this early age is difficult but the advantages to the baby and its mother outweigh the disadvantages to the surgeon.

Robert H. Ivy of the University of Pennsylvania has often spoken forth with sense backed by experience. At school he was a long-distance runner, then a dentist, a 1905 missionary in China and finally a plastic surgeon who continued to run the long race of life, and even after 92 years still with a spry but steady stride. Although a longtime friend and admirer of Vilray Blair, he did not let personal feelings influence his common sense. In 1955 he wrote:

Regarding early treatment, it should be stated that newborn cleft lip and cleft palate do not constitute a surgical emergency. Frequently the physician who has delivered a baby with one or both of these anomalies is under the impression that immediate closure of the cleft is imperative to allow the baby to nurse and without surgical closure, starvation is imminent. Nothing is more erroneous. The surest way to kill a baby in a poor condition of nutrition is to operate on it. . . . By the use of a little ingenuity and patience, feeding can be carried out with a medicine dropper, spoon or special feeders. Swallowing of the food is facilitated by holding the baby in the upright position. Some surgeons advocate operation a few days after birth. We do not subscribe to this, as we feel that accurate coadaptation of the cleft edges is more difficult when the parts are so small and much better end results are obtained by waiting until the child is six weeks to three months old or has reached ten pounds in weight.

In 1954 MacCollum and Richardson of Boston Children’s Hospital answered the parents’ question “When will the operation be done?” with “We usually operate when your child is around six pounds, must be gaining in weight steadily, and must be in good health.” Evidently, this dropping of the limit by four pounds was not a hazard at Boston Children’s Hospital as they reported in 1958 no operative deaths in a series of 2,635 cases.

Clarkson, of Guy’s Hospital, London, said in 1955:
Once the baby is gaining weight and is considered by the surgeon fit to stand his operation, the lip should be closed. Apart from the psychological upsets which the open cleft causes to the parents, and apart from the feeding difficulties associated with it, there is the fact that the infant by sucking its thumb and using it as a lever in the cleft will enlarge the size of the palatal cleft and increase the difficulties of its successful closure. The rule which holds generally in this country [England] that the baby should be at least 10 lb. in weight was possibly a reasonable one when this work was done in general services. Its effect in practice to-day is to delay the primary repair of most cleft lips in England until the baby is between 3 and 6 months of age. I believe this to be quite unnecessarily late, and indeed undesirable, when the work is done at plastic centres.

Claire Straith in Detroit operated at a very early age and used local anesthesia to reduce the dangers. But Straith was an unswerving Scotsman and a fast surgeon not easily disturbed by minor details such as a twisting head or distances measured in millimeters. I have heard him defend his stand on early surgery with

If I don't operate, someone else will and it is important that the lip be done correctly.

A peek behind the Iron Curtain in 1959 revealed to me, and was confirmed by Michael Lewin in 1962, that most Russian surgeons operate on clefts of the lip between six months and one year. The old master, Limberg of Leningrad, withholds surgery for at least an entire year.


The operation for cleft lip should be done in the first few months of life; it may be done within the first twenty-four hours if the baby is healthy in every other respect. . . . The mother need never see the deformed baby. Another period of hospitalization for the baby will be unnecessary. The problem of feeding will be simplified . . . If early operation is impossible, closure of the lip should be postponed at least until after the birth weight has been regained—usually in two or four weeks.

Holdsworth of London noted in 1970 that in Great Britain 4.5 kg. weight or three months age is the usual criterion for primary lip operation. He wisely suggested:
Parents will be more reconciled to their child, and his surgery, if they have lived a few weeks with the untreated deformity. As a gesture of kindness to over-wrought parents, the early operation is a mistake.

After one month of age, the patient has better cardiovascular-pulmonary adjustment, nutritional transition and ability to combat infection. This combination emphasizes Wilhelmsen and Musgrave's 1966 preoperative requirements in the "rule of 10":

- Weight—10 pounds
- Hemoglobin—10 grams
- White count under 10,000 per cubic millimeter

Most modern surgeons follow this general rule.

As I wrote in 1965, the cleft of the lip can be closed any time from the day of birth to old age. There is no need to rush from the womb to the operating room. The best final results are being achieved when the first operation is carried out at about three months, which is after the nose and lip components have had a chance to increase in size along with the patient, who should weigh 10 to 12 pounds. In 1967 the general "rule of over 10" was proposed as a criterion for lip surgery:

- Weight—over 10 pounds
- Hemoglobin—over 10 grams
- Age—over 10 weeks

The addition of the simple adhesion procedure in the wide clefts has enabled us to move up the initial surgery to a few weeks after birth and to postpone the final lip closure to six months.

Denis Glass lamented in 1970:

It is a pity that the work by Straith (1955) and McCash (1957) has not received more attention. Lip closure in the first week of neonatal life under local anaesthesia would seem to have many beneficial effects.

This was spoken like a true orthodontist, interested in alveolar molding with little insight into the details of actual lip surgery. He would probably settle for a "lip adhesion" at three weeks which will start early molding of the maxillary elements. This might be, indeed, the best compromise for all sides, leaving the
detailed surgery until the baby is six months old, when he will be stronger and offer more and better tissue toward the lip and nose construction. As the time for surgery is not a life-and-death matter today, surely if the surgeon can do a better operation later with a more nearly perfect result, this advantage can never be outweighed by either comfort or expediency to patient and parents.

THE BEST OF BOTH TIMES

Today I postpone an incomplete unilateral cleft to at least three months of age and preferably to four or five months when possible. Of course, at the time of surgery a hemoglobin level of 10 gm. and freedom from infection are required. The same timetable is used for complete clefts without alveolar distortion or palate cleft. For complete clefts with alveolar distortion and cleft of the palate, a lip adhesion procedure and a soft palate closure are carried out at about two to three weeks of age with the same general blood requirements even though these preliminary operations are quick and bloodless. Then, at approximately six months of age the definitive closure of the nasal floor and lip and correction of the nose is accomplished.