

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.



Galen

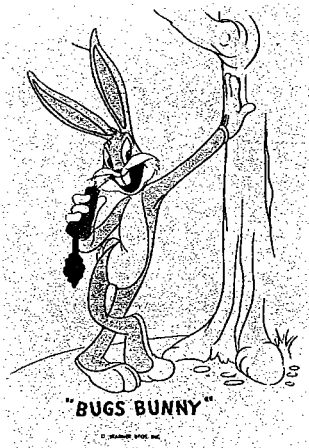




The third is also a red-brown and white stirrup spout vase depicting perfectly the unilateral lip cleft with exposure of the distorted maxilla and a malpositioned tooth, a philtrum dimple and the typical nasal deformity.

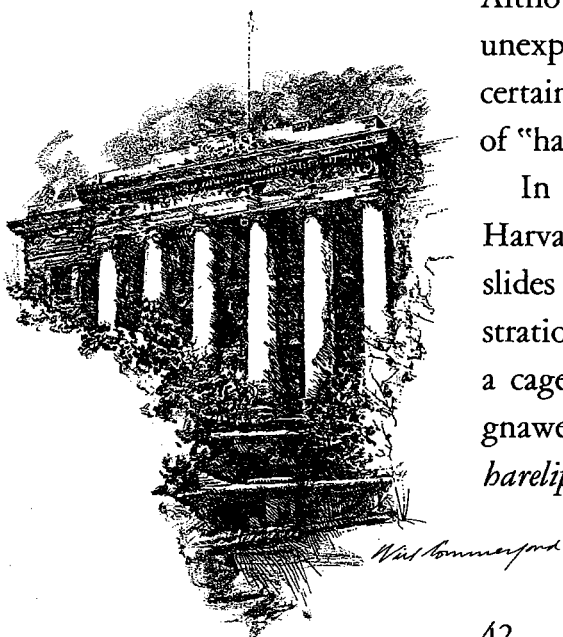
It seems that the earlier Peruvian cultures might have missed an opportunity. Had they capitalized on the midline lip cleft and flat nose of their beast of burden, the woolly-haired ruminant, this anomaly today might enjoy the melodious label of "llama lip."

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 *heirlip*. 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.

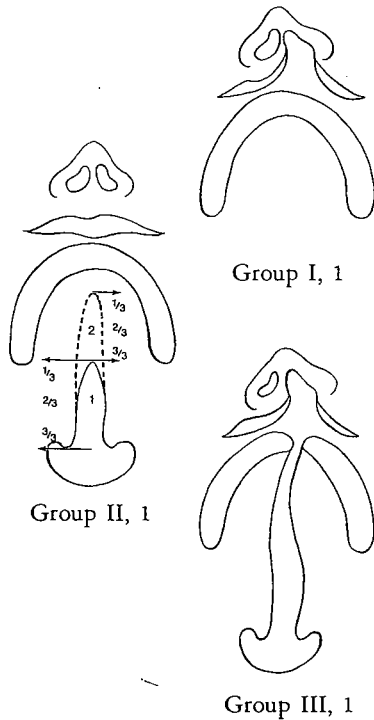


Staige Davis



Harry Ritchie

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.

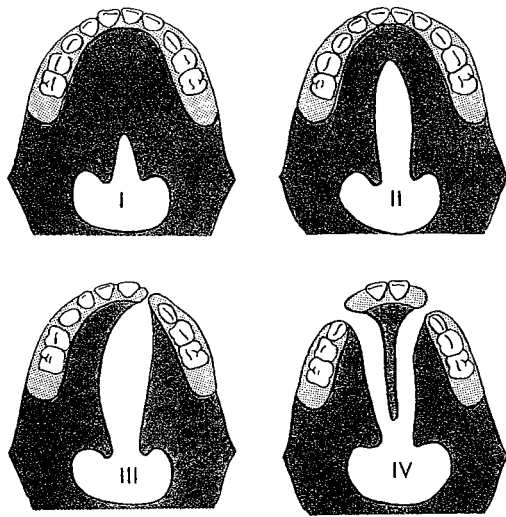
Ritchie had indicated that the alveolar border was the basis for all surgical groupings, and from a practical point of view he was not far off. Brophy gave his own grouping as 16 somewhat unrelated items which, as Ritchie pointed out, could be fitted into their classification. Blair approved of Ritchie's 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 prolabium 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



Poul Fogh-Andersen

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.

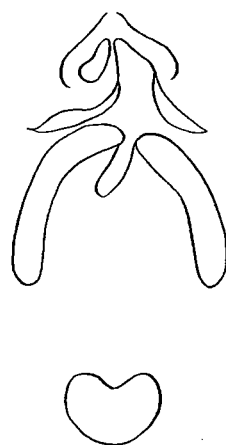
4. A group of rare atypical clefts was optional.

As Fogh-Andersen wrote in 1965:

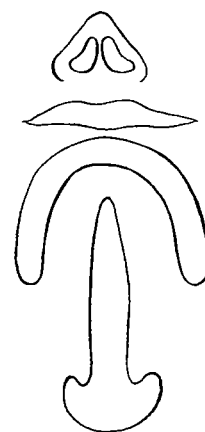
This classification has been adopted later by a series of writers, and Kernahan and Stark's classification in 1958 is in reality identical with it.

KERNAHAN AND STARK REDUCE IT TO TWO GROUPS

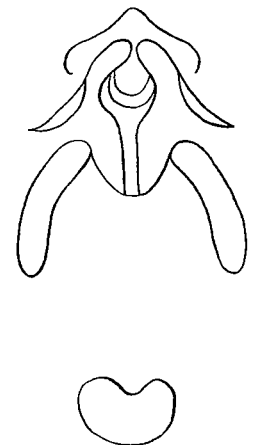
Kernahan and Stark's 1958 classification emphasized the embryological basis of the incisive foramen's being set as the boundary marker. Clefts of the lip and premaxilla, occurring at four to seven weeks of embryonic life, were termed *clefts of the primary palate*. Clefts of the hard and soft palate 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.



Unilateral cleft-primary palate



Cleft of secondary palate

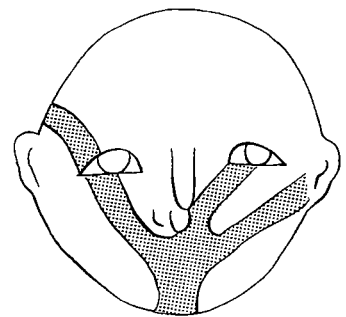


Bilateral cleft-primary palate

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 sub-categories.

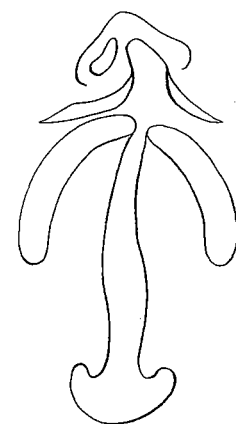
H A R K I N S

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.

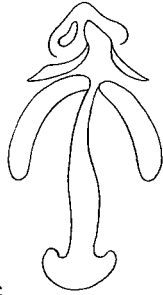
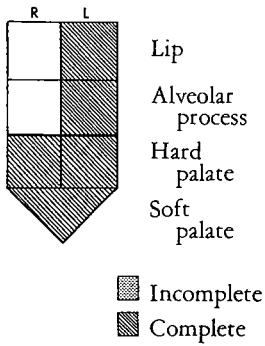


V I L A R - S A N C H O

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.



KI GI UIS SK



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"

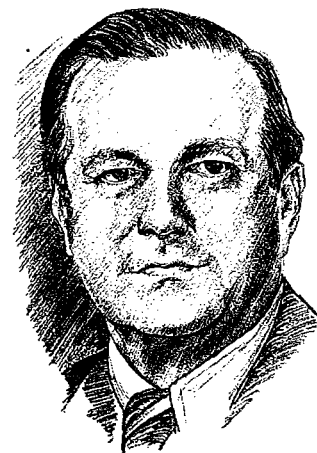
Desmond Kernahan had his early training in cleft surgery with Kilner at Oxford and later with Osborne and Burston in Liverpool. Then he joined Stark in New York to create their embryologically based classification. Kernahan finally landed at the University of Manitoba in Winnipeg, where in 1971 he noted

... 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

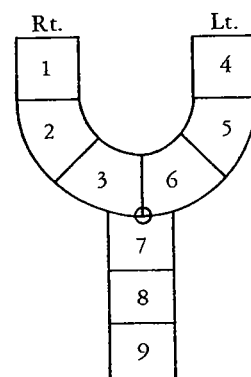
can be represented as a Y. The dividing point between the primary and secondary palates—namely, the incisive foramen—can be represented symbolically at the junctions of the limbs of the Y by a small circle.

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)



Desmond Kernahan

I wish that were our main problem

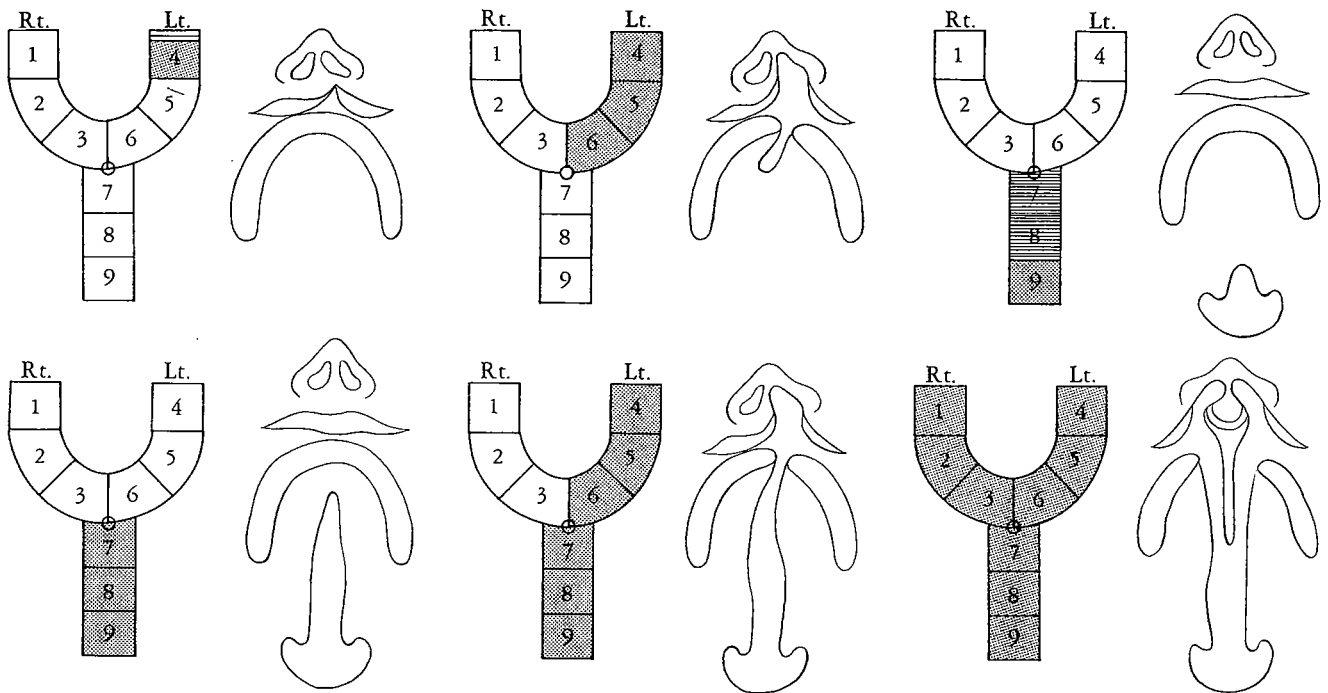


and soft (9) palate are also divided into three segments. This segmented Y can be reproduced by a rubber stamp.

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".

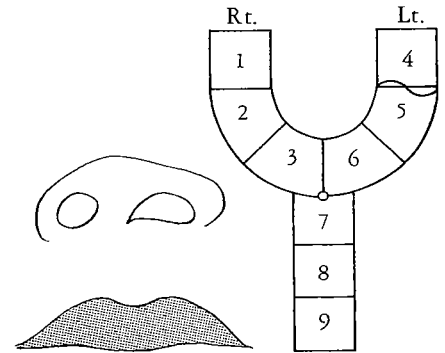


When asked how he charted median clefts of the lip, Kerna-
han explained in 1972:

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.

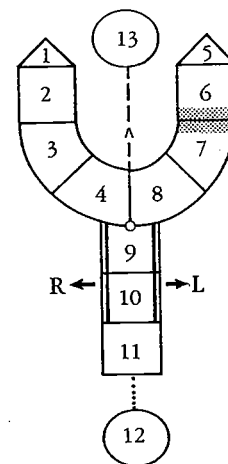


STRETCHING THE "Y"

Then in 1972, just as those interested in clefts were settling down to this sound classification with its nice, neat, symbolic representation, Egyptian Nabil Elsayh, 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.



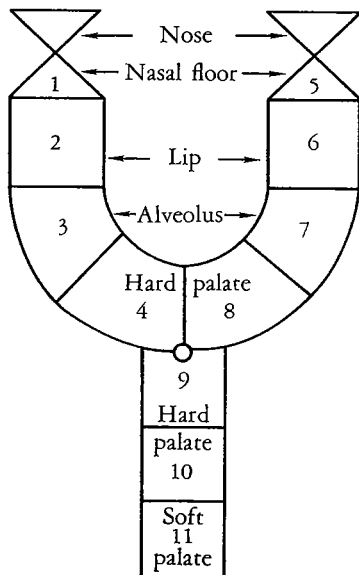
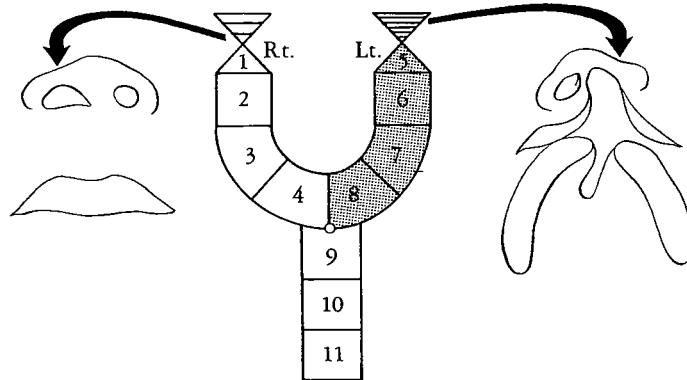
Collapse of the maxillary segments is indicated by filling in or stippling 3 and 4 or 7 and 8. Elsayh double-lined the squares (9 and 10) in the hard palate area and used arrows to indicate direction of deflection in complete clefts. He placed a circle (12) under the stem of the Y to represent the pharynx. 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. Elsayh 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 Elsahey'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

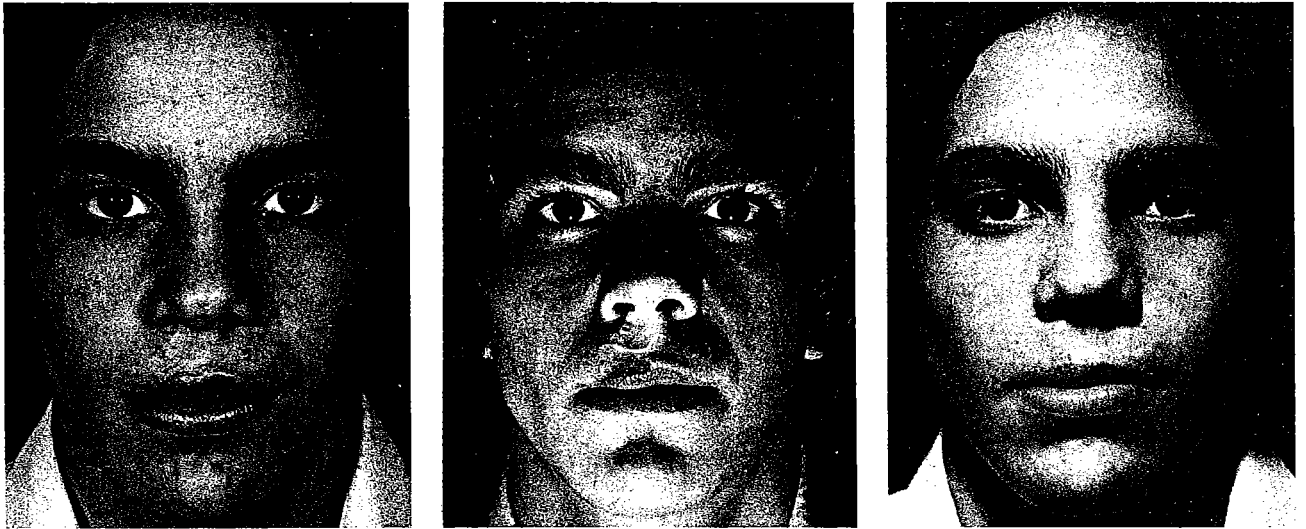
Closure of lip clefts is most commonly referred to as *repair of cleft lip*, but the word "repair" suggests that the lip was once intact, has separated and must be repaired. Webster defines the verb "repair": "1. To restore to a sound or good state after decay, injury. . . . 3. To remedy, heal, . . . 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.