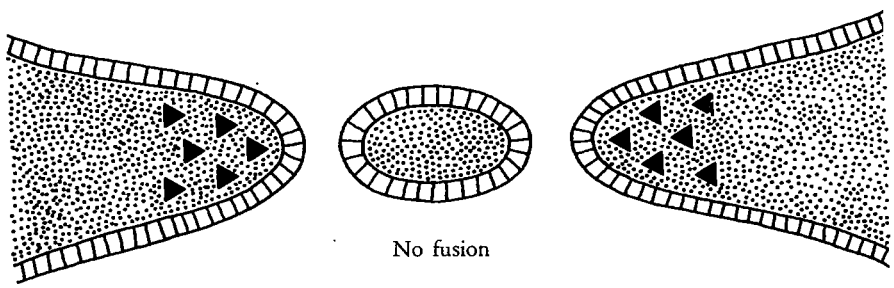
I. *Primary Deformity*

1. *Special Bilateral Cleft Embryology*

BRADLEY PATTEN of the University of Michigan, after a lifetime of research, in 1971 revealed an expert's acceptance of the unknown:

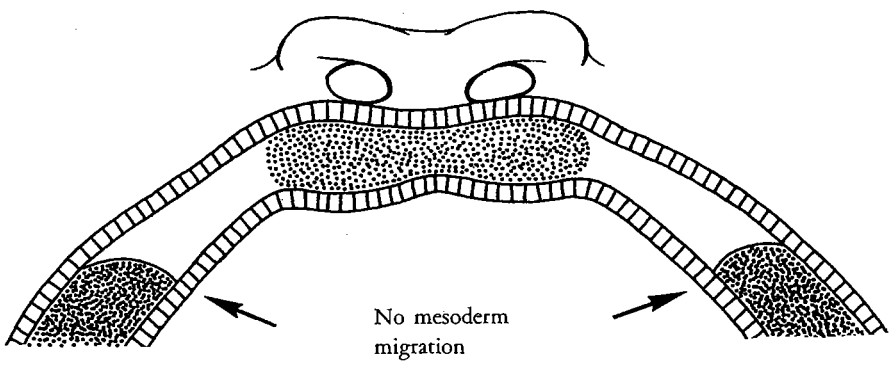
> Why clefts of the lip are sometimes unilateral and sometimes bilateral is not known. There seems no reason to suspect that their genesis differs in anything other than symmetrical or asymmetrical distribution of whatever the disturbing agent might have been.

The various embryological theories in vogue today have been discussed in some detail in Volume I. Whether it be failure in the fusion of Dursy-His, or failure of the mesodermal migration of

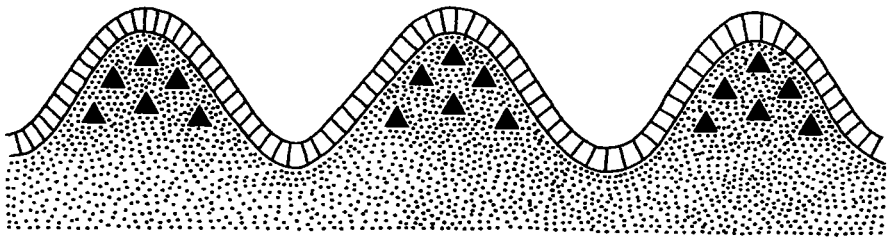


No fusion

Fleischmann-Veau-Stark, or failure of the merging of Patten, or a

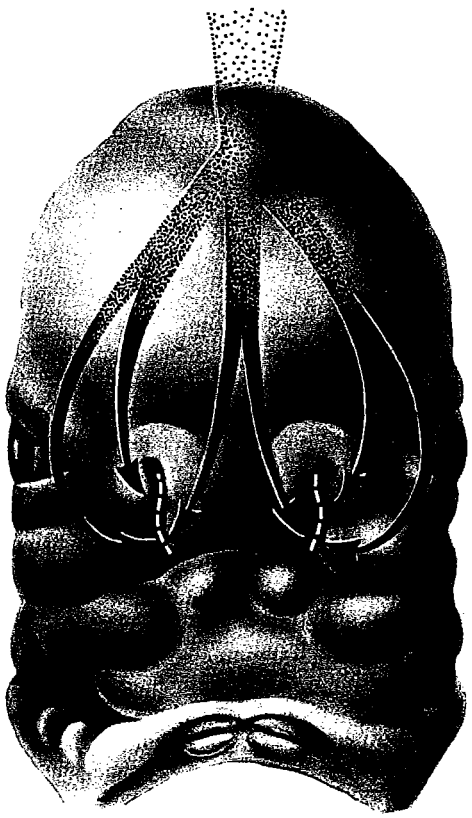


No mesoderm migration



No merging

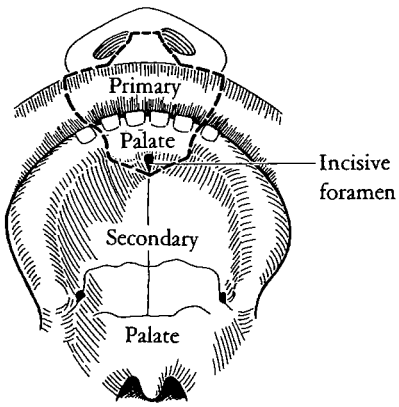
combination of these, whatever fails on one side in unilateral clefts fails on both sides in the bilateral deformity. At least in the standard bilateral cleft the site of the clefting is situated consistently along the embryonic grooves dividing the *maxillary* and *nasolateral* prominences from the *nasomedial* prominence. There can be varying but equal degrees of unsuccessful obliteration of these embryonic grooves on both sides or asymmetrical differences between the two sides.



The doubling of the fissures more than doubles the problem in the original deformity and its exaggeration during intrauterine growth and thereafter. The results of this will be discussed in Chapter 2 on anatomy.

PRIMARY AND SECONDARY PALATES

The primary palate and the secondary palate are delineated by the incisive foramen as the central landmark and sutures extending anterolaterally to the spaces between the maxillary lateral incisor and the first canine tooth on each side. This description was originally noted by Fogh-Andersen and later simplified and popularized by Kernahan and Stark.



As stated succinctly by Kernahan for Stark's book *Cleft Palate* in 1968:

> The classification was based on the evolution and mode of growth of tissues forming the primary and secondary palates. Briefly, the primary palate—comprising the central portion of the upper lip, premaxilla, upper incisors, and anterior nasal septum—forms between the fourth and seventh weeks of intrauterine life and extends to the nasopalatine canal site of the incisive foramen. The secondary palate—comprising the remainder of the hard palate and the soft palate posterior to the incisive foramen—forms between the seventh and twelfth weeks as a pair of shelves that grow toward the midline and fuse in a normally developing embryo.
>
> In view of these findings, the genesis of both rare and common lip and palate deformities becomes obvious. Either the primary or secondary palate or both may be involved, resulting in a degree of malformation from a submucous palate cleft or a vermilion notch to a bilateral cleft involving all dimensions of both palates.

BILATERAL PECULIARITIES

Certain aspects that are peculiar to bilateral clefts deserve special consideration. In the complete bilateral clefting the central frontonasal component is separated from both lateral elements. The middle segment seems thus to be released from any responsibility to the lateral elements, and they in turn, being somewhat passed by, withhold their usual contribution to the central element. Although the same cleft discrepancies occur as in the unilateral cleft, doubling the action compounds the effect on the central portion, including the prolabium, premaxilla, columella and septum.

Before Veau got Fleischmann and Hochstetter together on the mesodermal migration hypothesis, Thomas F. Mullen of San Francisco in 1932 made some interesting observations:

I have never been able to demonstrate any muscle in the parings from the margins of the prolabium in complete bilateral clefts of the lip. . . . A study of the reported cases of cyclopia in which the nasofrontal process fails to descend and enter into the formation of the mouth but remains above the single eye in the form of a proboscis reveals the fact in many instances that while there was an absence of the premaxillary bones, the lips were normal. Evidently in these cases the maxillary processes have united in the mid-line and the lips have been formed without any elements from the nasofrontal process (Craig, Malherke, Priano). I am of the opinion that the orbicularis oris never receives any fibers from the nasofrontal process but that the muscle elements grow into this area from the sides and there is never any muscle in the isolated prolabium of complete bilateral clefts. . . . The fact that the entire muscle bed from which this whole system is developed is confined to the area of the hyoid arch until the seventh week, whereas the lips are formed by the union of the maxillary processes with the median processes *before* this time, may account for the fact that there is no muscle in the tissues of the prolabium in complete bilateral clefts.

Miroslav Fara with histologist J. Smahel of the Plastic Surgery Clinic created by Burian at Charles University, Prague, finding no striated muscle fibers in the prolabium of complete bilateral clefts, concurred in 1967 with the earlier deductions by F. Burian, Albert D. Davis and I. Stanek:

The muscle fibers of the musculus orbicularis oris are not formed as a result of the transformation of the mesenchymal cells in situ, but . . . as a result of the growth of said muscles from the side toward the center of the philtrum. The formation of a complete cleft, which in embryogenesis precedes the growth of the muscular fibers, thus causes the sterility of the prolabium.

Union of the orbicularis oris muscle in the upper lip can occur independently of the frontonasal process participation. In cyclopia, the primitive proboscis projects above the single central eye, and the intact upper lip has muscle union below it. Yet this lip is not normal and presents no philtrum columns, associated vertical groove or cupid's bow. Thus I would like to hypothesize

that although the frontonasal component contributes no muscle to the upper lip its participation in the "philtrum party" is responsible for partial midline interruption of what would otherwise be a flat, uninteresting end-on union of the lateral muscle bundles. Instead, when the frontonasal component *normally* enters from above, descending vertically between the muscled maxillary elements, it causes ripples expressed in columns, dimples and the curves of Cupid.

Just how this process takes place is still open to argument. In 1962 Monie and Cacciatore of the University of California, San Francisco, after the first real research on this subject, reported no evidence that the philtrum is related to the lines of fusion of facial processes in the embryo. Rather, they said, it is associated with increasing median density of connective tissue of the upper lip apparent between the third and fourth months of fetal life.

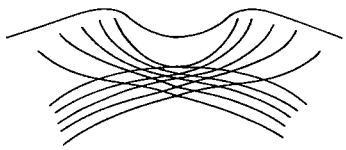
One bilateral cleft closure in which I was able, without tension, to bring three-quarters of a circle of muscle around a tiny tethered mid-philtrum inverted into a dimple and persisted as such. This serendipitous dimple led me to conjecture further on the etiology of the philtrum configuration. Monie was requested to review his bilateral cleft specimens to determine the time of upper labial sulcus formation. The question was whether the prolabium-philtrum is still totally tethered to the premaxilla while the groove and eminences are being formed. Monie explained that his specimens had been cut on the transverse plane to give the best view of the philtrum but unfortunately did not show the labial sulcus.

Latham's research does not seem to confirm the findings of Monie. He wrote in July 1973:

> A look at some horizontal sections through the upper lip region of a normal fetus made it clear that the philtral groove is not caused by fibers emanating from the mid-palatal suture. The flared out posterior ends of the medial crura of the alar cartilages do lie in the superior part of the philtral ridge; however, a much more striking and obvious mechanism causing the philtral groove was seen in the arrangement of the labial muscles. The sections show muscle fibers arising from the alveolar bone over the lateral incisor, then coursing anteromedially to an insertion in the medial philtral part of the lip, just beneath the epidermis. This has been confirmed in another specimen sectioned in the sagittal plane.

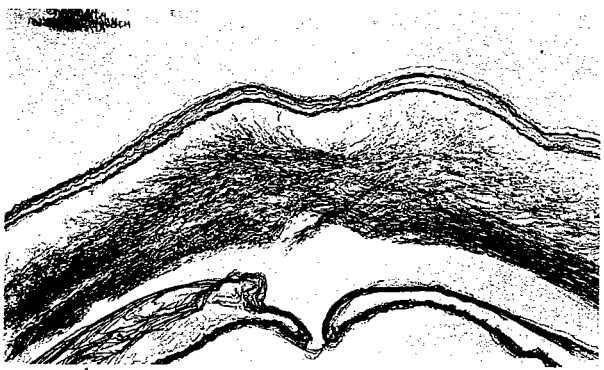
This readily explains why the lip of a bilateral cleft infant does not retract much when the underlying premaxillary bone is retracted—because there are no muscles in the bilateral cleft lip! This, incidentally, would also appear to support the recent view that muscle should be pulled into the midline. It would be nice if the right muscles could be brought in!

Finally in 1976 Latham, with T. G. Deaton, clarified his latest anatomical findings on the philtrum. What follows is a summary of these findings with some of the illustrations published in the *Journal of Anatomy.*

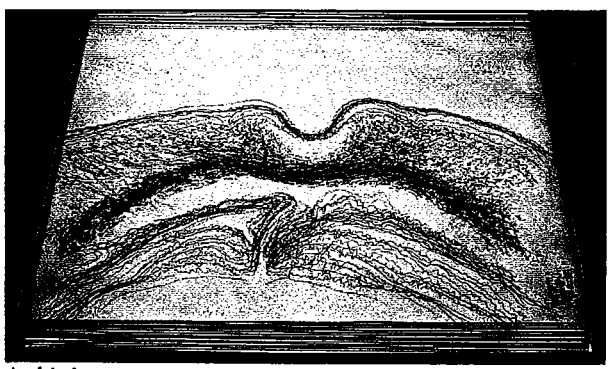


The course and insertion pattern of muscle fibres in the philtrum of the human upper lip were studied in seven postmortem specimens using serial histological sections and a reconstruction method using sheets of Plexiglas. The fibres of the *musculus orbicularis oris* entering the upper lip from one side were observed to decussate in the midline and proceed to insert into the skin of the opposite side lateral to the philtral groove. The philtral ridges appeared to represent the medial borders of bilateral muscle insertion zones of the lip in which the fibres of the *m. orbicularis oris* raised the level of the skin by splaying out and inserting into it.

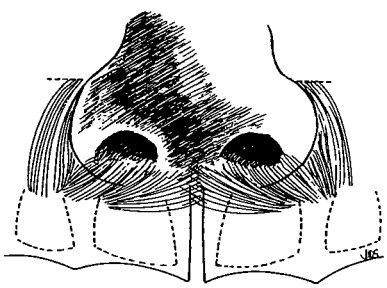
Here are horizontal reconstructions of the upper lip musculature of an 18½ week fetus and of a newborn infant.



18½ weeks

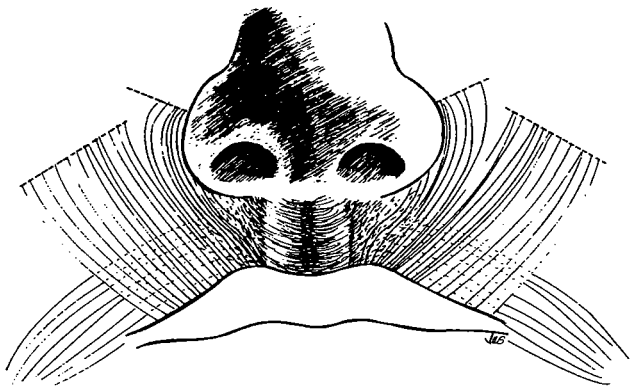


At birth



Contributions to philtral form also came from the *musculus levator labii superioris* as it descended as far medially as the philtral ridge to insert into the vermilion border lateral to the median groove, and from fibres of the *musculus nasalis* which inserted into the philtral ridges superiorly. The philtral groove corresponded to the more compact median decussation of the *orbicularis oris* where lip thickness was also reduced by the relative absence of muscle fibre insertions into the skin. The displacement of the vermilion border giving rise to Cupid's bow appeared to result from the

lifting action of the *m. levator labii superioris* lateral to the median groove in conjunction with a depressor action by the *m. orbicularis oris* on the median tubercle.



Each of these theories could, I suppose, represent the residual effect of the muscleless frontonasal component's becoming overwhelmed, dimpled and ridged by the inflow of lateral musculature.

PREMAXILLA

Woo found from embryological studies that the premaxilla develops from two pairs of ossification centers. The principal pair forms the primordia of the lateral incisors, extends upward and with the maxilla proceeds forward to embrace the premaxilla on either side. This union is complete by the end of the third month.

Time of protrusion

In 1954 Stark presented a 60-day-old embryo of 46 mm. with bilateral clefts revealing advanced protrusion of the premaxilla. In 1966 Kraus, Kitamura and Latham presented three interesting embryos that seem to tag the time of protrusion. Their 41- and 43-day-old embryos showed no signs of premaxillary projection, but in their 47-day-old specimen protrusion was beginning to appear. Thus Latham in 1973 concluded that protrusion in bilateral clefts begins at about 45 days (10 days after the original clefting, which occurs at 35 days) and then develops rapidly for

25 days to reach proportions at 70 days (10 weeks) comparable to those seen at birth. Recently he has been working on a 13-week-old bilateral cleft fetus which reveals premaxillary protrusion equivalent to that of the 10-week fetus and in fact similar to that seen at birth. This timing is similar to what he noted in unilateral clefts in his 1969 *Cleft Palate Journal* study.

Cause of protrusion

BONE GROWTH. There have been numerous theories as to the cause of premaxillary protrusion in bilateral clefts. In 1934 Victor Veau proposed a bone-centered concept of a forward growth force within the developing vomeropremaxillary stem thrusting the premaxillary segment into a protrusion position. Some surgeons have upheld fellow surgeon Veau's belief in the excessive growth between the vomer and the premaxilla. In fact, in 1949 Denis Browne of London demarcated this zone from the quiescent vomer by "a cartilage-filled suture line" and designed his excision for "set-back" in this area of so-called overgrowth.

MUSCLE RUPTURE. Many authorities point to the disrupted facial musculature as an adequate explanation of the cause of this skeletal deformity and thus advocate early restoration of the muscle continuity of the lip as a logical treatment. Pruzansky in 1954 and again in 1964 took this stand. Slaughter, Henry and Berger in 1960 noted the position of the philtrum and premaxilla displaced far anterior to the normal plane and accused:

> This is due to the lack of restraining action of the normally constituted orbicularis oris and its accessory muscles. The unopposed, powerful action of the tongue is then free to exert itself, accentuating the deformity.

In 1967 Fara and Smahel stated:

> The anatomical incompleteness and functional inefficiency of the musculus orbicularis oris in complete bilateral clefts contributes most probably to the formation of a marked protrusion of the premaxilla, manifesting its functional discordance in all the muscles of expression and the disturbance of the harmonious development of the whole middle facial region.

CARTILAGE GROWTH. Others have placed a great part of the responsibility for premaxillary projection on the associated cartilaginous structures.

Patten queried as late as 1971:

The forward growth of the embryonic nose is a relatively late process. If the intermaxillary segment is not properly anchored by the time the nasal growth accelerates, might that be a factor in the way it is carried far out of its normal relations?

Yet it was James H. Scott, Professor of Dental Anatomy, Queen's University, Belfast, who in 1953 redirected attention to the role of the nasal septum in facial growth. An indefatigable research worker with devastating logic and a dental liberal with a dry Ulster wit, he opened his lectures with

James Scott

I'm going to speak to you about bone growth but there's not going to be any newfangled histochemistry in what I say.

Short, stocky, with a big amiable face, he was crippled with rheumatoid arthritis which forced him to teach from a wheelchair. Even up to the end he bubbled with vitality and cheerfulness, stressing the role of the cranial sutures and pointing to

the cartilaginous parts of the cranial base and nasal septum as the pace setter in skull growth.

According to Scott, the growth of the cartilage of the nasal septum acts as the force for growth of the upper facial skeleton in anteroposterior and vertical dimensions. The growing septal cartilage separates the suture lines, permitting new bone growth. It has been said to act as a kind of "epiphyseal plate" for the whole of the upper facial skeleton. This is the mechanism of growth from the latter part of fetal life through the first three years of life. Then for the rest of the first decade sutural growth diminishes as surface apposition and resorption gradually take over.

Another of Scott's endowments to clefts was the inspiration of ardent students such as Burston and Latham. Burston actually went to Scott for a thesis topic and was given the chondrocranium of the sheep. His work on this subject was later applied to cleft palate. In 1958 he reported the abnormal development following clefting and since then he has created a world-renowned cleft center in Liverpool, England.

Ralph Latham

LATHAM'S HYPOTHESIS. Under normal circumstances, the bony premaxilla is kept in place by its early fusion with the maxilla to form one bone and by the continuity of mucogingival tissues around the developing dental arch. The development of bilateral clefts creates havoc. Irish oral biologist Ralph A. Latham, originally of Belfast, trained by Scott, stimulated by Burston and now in research at the University of North Carolina School of Dentistry, has put it all together in a logical hypothesis. In 1973, both in the *British Journal of Plastic Surgery* and at the Foundation Cleft Palate Symposium at Duke University, he proposed:

> In the bilateral cleft the premaxillary segment is under no restraint laterally either from bone or gingival fibrous tissue; consequently its attachment to the nasal septum by the septopremaxillary ligament becomes a dominant factor.

He also noted that in normal growth, as the nasal septum grows forward, it draws the upper jaw with it, but not at the same rate. In the bilateral cleft, the premaxillary segment is carried forward at the same rate as that of the growing septum to which it is firmly held.

For bone formation to occur at a skull suture, there must be tension between bone edges (Scott, 1948, 1953, 1954; Moss, 1954; Selman and Sarnat, 1957; Pritchard, Scott and Girgis, 1956). The premaxilla united to the forward-growing nasal septum but restrained by the vomer sets up a tension at the vomeropremaxillary suture creating a condition for bone formation. Indeed, the vomeropremaxillary suture is the major site of rigid hard tissue formation and elongation of this segment. Pruzansky confirmed this finding in 1971 with serial radiography following placement of metallic implants.

Yet, as pointed out by Latham, premaxillary protrusion is not driven forward by excessive vomeropremaxillary suture growth. Rather, the premaxilla is carried forward by its attachment to the nasal septum through the septopremaxillary ligament. To complete the picture Latham points out:

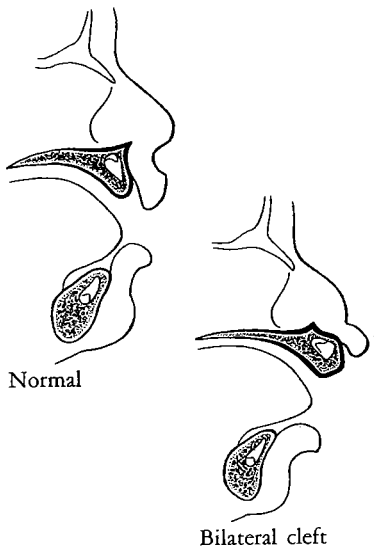
> If bone may not push against bone it appears bone may push against soft tissue! Growth of the maxilla from late foetal life into the early years of

childhood is characterized by bone formation on the upper and posterior surfaces with progressive downward and forward progress of the upper jaw.

ROSS AND JOHNSTON. Orthodontist R. B. Ross, of the Hospital for Sick Children, Toronto, and geneticist M. C. Johnston, of the National Institute for Dental Research, Bethesda, Maryland, expressed a different opinion about the position of the premaxilla in complete bilateral clefts in their 1972 book, *Cleft Lip and Palate:*

The premaxillary segment appears to be tremendously advanced, protruding beyond the tip of the nose. This has often been cited as an illustration of the vigorous growth of the nasal septum and as confirmation of the theory that the cartilaginous nasal septum provides much of the stimulus for maxillary growth [Scott]. . . .

However, the premaxilla is tipped forward . . . and most of the protrusion is due to the forward rotation of the alveolus. The anterior nasal spine (which is a fairly accurate indicator of premaxillary position) is only mildly advanced, although pointing superiorly because of the rotation. Protrusion of the premaxillary segment is probably the result of normal tongue action against a flexible and relatively unsupported bone, without the opposing force of circumoral musculature to establish an equilibrium of forces. The tip of the nose is invariably tilted slightly at birth as a result of the position of the premaxillary segment. The base of each nostril is pulled laterally by the cheek musculature, giving the typical appearance.



Other aspects of protrusion: actual and relative

Much of the premaxillary protrusion is due to the abnormally anterior position of the alveolar process, which proceeds as forward alveolar growth for seven to eight months of intrauterine life. The fact that the teeth continue in a normal upright position suggests that the alveolar segment does not actually rotate but by expansion of the dental alveoli reaches a maximum at about four to five months. After that, alveolar protrusion persists as a static feature.

The degree of premaxillary protrusion may be exaggerated by the relative underdevelopment of the unattached maxillary segments, as normal forward development of the early maxillae is to some extent dependent on a forward pull from the nasal septum.

Underdevelopment of the maxilla

Intrigued by maxillary underdevelopment associated with the absence of the cartilaginous nasal septum, J. V. Harvey Kemble studied all such cases under age 20 years seen at Queen Mary's Hospital, Roehampton, London, in the past 10 years. The total was eight—three congenital, four traumatic and one unknown.

Kemble acknowledged three theories on the development and growth of the maxilla:

1. Scott's implication of the nasal septum as the determinator of mid-facial growth (1953, 1956, 1959), confirmed by Weinmann and Sicher (1955).
2. Latham's theory that the maxillae have inherent potential for growth which does not depend on outside influences (1968).
3. Sarnat's experimental findings that the normal nasal septal cartilage growth is necessary for full maxillary development (1963).

As Kemble concluded that his clinical findings confirmed theory 3, it is of interest to review his abstract of Sarnat's work:

Sarnat (1963, 1966) . . . deduced from experiments of extirpation of circum-maxillary sutures in monkeys and in rabbits, that because little or no deformity resulted when compared with controls, growth which occurred at sutures was of a secondary accommodating nature. Furthermore, he showed that extirpation of the cartilaginous nasal septum from growing rabbits produced smaller nasal and maxillary bones than in controls. The extent and severity of the deformities varied approximately in proportion to the amount of septum removed. Removal of cartilaginous septum from full-grown rabbits did not result in collapse of the snout, so that it seemed unlikely that it was lack of support that produced underdevelopment of the snout in growing rabbits (Sarnat 1967). From these experiments it would seem likely that the cartilaginous nasal septum is primarily responsible for forward and downward growth of the maxilla, and that growth at sutures maintains and consolidates its position, at any rate in experimental animals. The fact that Stenstrom (1970) found only minor deformities resulting from removal of septal cartilage from young guinea-pigs can only be explained on the basis of a species difference.

Latham's Irish reaction, in November 1973, to this stand is rather provocative:

There is good evidence that the nasal septum is an important determinator

of upper facial growth in the embryonic and fetal periods. Individuals with impairment of the nasal septum usually show underdevelopment of the upper jaw at *birth*. . . . It is obvious that the human face is capable of growth *after birth* in the absence of the nasal septum. . . . There must be a considerable period before and after birth into childhood when nasal septum stimulus and intrinsic maxillary growth are complementary and working together. . . . There is a tendency for writers on this subject to want one mechanism to do the whole job. Kemble appears to think of three mutually exclusive mechanisms, each making their main contribution at a different time in a sequence. . . . Kemble's conclusion, however, is fair enough. It seems that the septum does make an important contribution both in growth and in structural support to the upper jaw and in its absence *full* maxillary growth will not be realized.

Latham, in conclusion, warned of the danger of evaluating maxillary underdevelopment as measured by the sella turcica-nasion-anterior portion of maxilla angle.

The septum, premaxillary bone and teeth all appear to come from the neural crest cells in the young embryo and if the septum is absent, the incisor teeth and bone are likely to be absent too. Such is the case in cyclopia and arhinencephaly. Then the SNA angle could be low because of the absence of these structures suggesting more underdevelopment of the middle third of the face than is actually present.

A combination of factors

Actually the premaxillary projection deformity must be the result not of one factor but of the interrelation of many factors—lack of bony continuity, growth at the suture, cleft of the orbicularis oris muscle, forward growth of the cartilaginous septum and expansion of the alveolar process.

THE NOSE

With the bilateral cleft deformity, the entire nose is deficient. In 1973 Stark and Kaplan reported tracing two bilateral cleft embryos to measure the ectodermal volume ratio of the primitive nose. They found extremely small measurements of 8.1 : 7.9 cubic cm. for the 40.5 mm. embryo and 12.1 : 10.8 cubic cm. for the 46 mm. one. These findings exhibit the great deficiency of ectoderm in the bilateral cleft lip nose, a horrendous lack of nasal germ plasm.

WHATEVER HAPPENED TO THE BABY COLUMELLA!

In 1974 R. A. Latham and C. Workman noted, in Georgiade and Hagerty's volume on the transactions of the Cleft Lip and Palate Symposium held at Duke University:

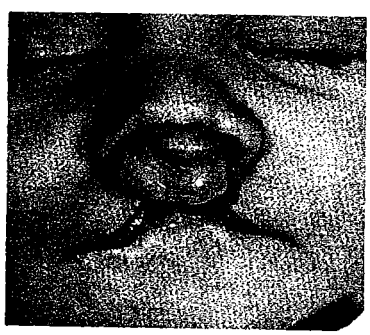
In the bilateral cleft condition the anterior nasal spine nestles between the flared out ends of the medial crura. The present interpretation is that the spine is too far forward, not that the crura are too far back.

The premaxillary bones are clearly set farther forward on the nasal septum than normally. However, a large part of the problem with the columella is due to the forward expansion of the alveolar process beneath the medial crura of the alar cartilages.

Latham blames the total clinical absence of the columella primarily on failure of differential growth and secondarily on the invasion of the columella area by the forward growing and expanding alveolar process.

Normal development of the columella is dependent upon the fact that the anteroposterior dimension of the cartilaginous septum increases at a faster rate than the vomer and premaxilla, thus pulling an elongation of the columella and shoving a projection to the nasal tip. Latham explains it this way:

When bilateral clefts divide the primary palate, the counterbalance on the septopremaxillary ligament is vastly reduced. The septopremaxillary ligament then exercises a dominant influence on the premaxillary bones, which are held tightly to the anteroinferior border of the nasal septum. This would account for the forwardly placed basal part of the premaxillae. The forward growth differential between the nasal septum and premaxillary bone fails.



Thus, the loss of columella is due directly to the lack of this differential growth between the septum and the premaxilla. The nasal tip and its cartilages also must suffer the consequences. Like a tent without the insertion of its front pole, the nasal tip never rises, leaving the angle of its alar cartilages flattened and the feet of its medial crura splayed.

As further proof that the premaxillary segment persists in an embryonic relationship to the nasal septum in bilateral clefts, Latham found Jacobson's organ as a pit in the septal mucous

membrane not in the full-term normal position *anterior* to the alveolar process but rather *above* and *behind* it.

When, in addition, the developing alveolus bulges forward during gradual anterior expansion of the incisor teeth and alveolar process to encroach upon the columella base, there is further obliteration of this nasolabial angle. The swindled columella is left relatively as well as actually destitute!