# The Natural History of the Bilateral Cleft

J. D. Atherton, B.D.S., D.D.O., Ph.D.

## Introduction

The face of a baby with a complete bilateral cleft appears severely deformed at birth. Most noticeable is the characteristic protrusion of the premaxilla and midline lip tissue. This protrusion presents considerable clinical and social problems. The orthodontist in conjunction with the plastic surgeon and other disciplines can do much to alleviate these problems. However, before considering what may be done in the form of treatment it is important to know exactly how the face of the cleft baby differs from normal at the time of birth. It is also important to know the manner in which the cleft-affected face grows. There is a natural tendency, when faced with a newly born baby with a bilateral cleft, to think that the prominent premaxilla will become even more prominent with future growth. Few clinicians in these modern times have seen an older child with an unoperated cleft lip, never mind an adult, so that this attitude is understandable. One of the purposes of this paper will be, as far as is possible with the available material, to show how the bilateral cleft grows before, during and after birth. It is also of help in formulating a treatment for the prominent premaxilla to understand why the premaxilla is prominent. It is relevant to the understanding of this aspect of the problem to compare the situation in man with that in animals. As far as I am aware, all mammals suffer from clefts of the lip and palate of very similar types to those found in man because in embryonic life the face follows a common developmental pattern with regard to the formation of the primary and secondary palates.[1] There are in my collection examples of bilateral clefts in the dog, the pig, the sheep, the mouse and one chimpanzee. In demonstrating a possible reason for the prominent premaxilla in man with a bilateral cleft I hope to demonstrate important aspects relating to the morphology of the face of normal man.

The morphology, growth and comparative anatomy of the bilateral cleft which this paper is largely about might properly be called the "natural history" of the bilateral cleft, hence the title of this paper.

## The Morphology of the Bilateral Cleft

The face of the bilateral cleft at birth presents certain characteristic features. The small area of lip tissue in the midline (here referred to as the "prolabium") and the incisor bearing portion of the maxilla ("the premaxilla") are both protruding, often strikingly so (Fig. 1). The other feature which is remarkably constant is the absence of a columella, so that the premaxilla and prolabium lie beneath the tip of the nose. On a skull at Vrolik University (Fig. 2) it can be seen that there is a small anterior nasal spine and that the alveolus inclines forward from the spine rather than assuming a normal vertical contour. The teeth in this alveolus lie at a normal inclination so that the open pulp chambers have only a thin layer of bone overlying them. These features can also be seen on a good lateral cephalometric film. From the palatal aspect it can be seen that the vomerine process of the premaxilla and the vomer itself are elongated to form a stalk which carries the body of the premaxilla. At birth the maxillary

Fig. 1 Frontal, lateral and palatal views of a bilateral cleft baby aged 3 weeks showing a characteristic displacement of the prolabium and premaxilla and an absence of the columella.

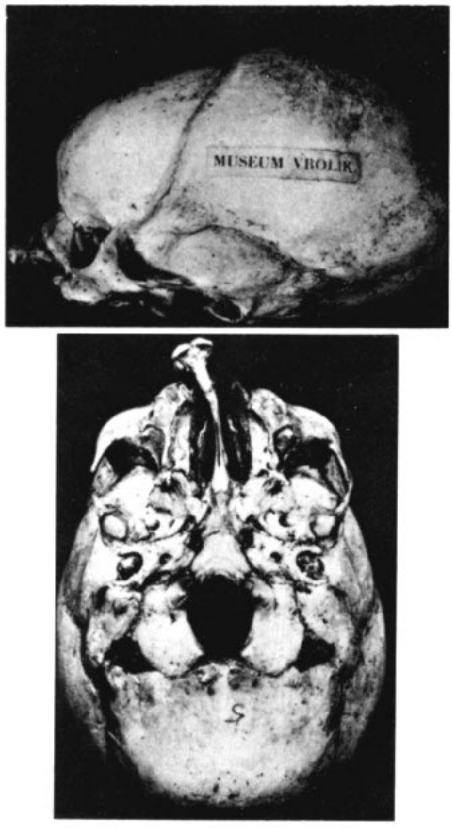


Fig. 2 Top, lateral view of the skull of a newborn baby with a bilateral cleft. The anterior nasal spine is small and the alveolar component of the premaxilla is inclined anteriorly. Below, palatal view of the skull shown above. The vomerine process of the premaxilla and the vomer are elongated with a displacement anteriorly of the premaxilla-vomer suture (Museum Vrolik).

bodies look small; however, measurements taken on the detached maxillary body of the unilateral cleft indicate that the canine to tuberosity distance is reduced by an average of only eight per cent.[2] The size of the maxillary body in the unoperated adult unilateral cleft confirms that there is a favourable growth potential for this region.[3]

To determine which structures were displaced in the bilateral cleft baby, measurements were taken from cephalometric films.

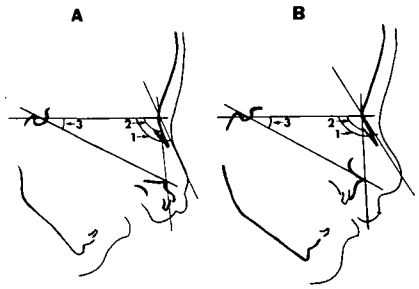


Fig. 3 Tracings of cephalometric films of newborn babies A) with a bilateral cleft and B) with a cleft of the secondary palate.

## Method

Fourteen babies with a complete bilateral cleft of the lip and palate were radiographed under light sedation and at a head-tube distance of five feet. All were within two months of birth. It was not possible to radiograph non-cleft babies for use as controls, therefore seventeen babies with a cleft of the secondary palate only were used as control cases.

Three angular measurements were used to test the normality or otherwise of the nasal septum and position of the anterior nasal spine (Fig. 3). They were (1) sella, nasion and tip of the nose (T), (2) sella, nasion and anterior nasal spine, and (3) nasion, sella and anterior nasal spine.

## Results

Angle 1 represents the prominence of the nose and hence is a measurement of the underlying nasal septum. Mean for the cleft group was 118.8 degrees and for the controls 121.2, not significant according to the student's 't' test. It is concluded, therefore, that the nasal septum is no more protrusive in the bilateral clefts than in the controls; if anything, there is a tendency for it to be less protrusive.

Angle 2 is larger for the cleft group (93.2) than for the controls (88.2). This difference is significant with a 't' value of 2.59. It must be assumed, therefore, that the anterior nasal spine lies in a more anterior position in the cleft group.

Angle 3 is smaller for the cleft group with a mean 25.7 degrees compared with a control mean of 29.1, a significant 't' value of 4.51. It must be assumed, therefore, that at birth the upper face height is reduced in the bilateral cleft group.

These findings indicate, as can be observed clinically, that in the bilateral cleft the nose and nasal septum are not overdeveloped, rather the reverse. The premaxilla is prominent because it lies in an abnormally forward position on the nasal septum. Clinically this is greatly exaggerated by the forward tilt of the alveolus.

## The Growth of the Bilateral Cleft

The clinician normally sees the unoperated cleft baby for the first few months of life. After this time, in modern society the cleft is operated and the basic morphology of the condition altered. Most clinicians do not have the opportunity to study the cleft face during fetal life or its growth during postnatal life without an operation. It is rather akin to treating a disease process without fully understanding the natural course of the disease. It is, however, possible to study the unoperated cleft in fetal life and to understand whether the peculiar morphology of the face at birth is the result of a progressive abnormality during fetal life or whether the condition with all its characteristics is established early in fetal life and persists unchanged to birth. Birth is simply a landmark in an evolving process and, as far as growth is concerned, is merely an incident in a long eventful history. After birth the cleft is usually operated which alters this experiment of nature.

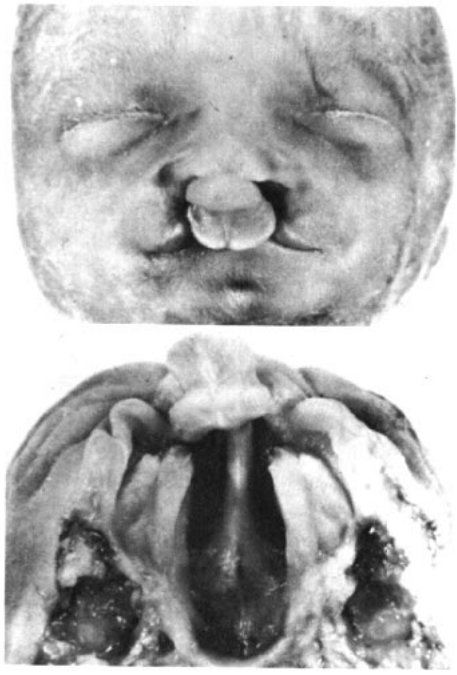

**Fig. 4 Frontal and palatal views of a 17 week-old fetus with a bilateral cleft of the primary and secondary palates. Reproduced from Latham.[4]**

Reports of unoperated cleft patients can be found in the literature and surveys of unoperated cleft patients have been made in parts of the world where modern surgery is not readily available. Even in modern society the occasional cleft is not operated for quite some time. The relative infrequency of the bilateral cleft, however, makes anything other than a gross study of the face impossible.

Latham[4] gives details of several bilateral cleft fetuses at different periods in fetal life. It is clear from his work that the condition arises early in fetal life and is apparent at the tenth week. It is probable that, like the unilateral cleft,[2] the characteristic morphology of the condition is established much earlier than this.

A typical midfetal specimen is shown in Figure 4 reproduced from Latham's thesis. This specimen at the age of 17 weeks is a miniature of the condition at birth. All these clefts, including one at 21 weeks personally available to the author, are similar in appearance to the specimen in Figure 4 and to birth. After the age of 24 weeks there are no reported specimens that I am aware of. However, the condition during midfetal is so similar to that at birth that it seems reasonable to assume that the cleft face between 24 weeks and birth grows in a constant manner. The growth pattern, therefore, through the greater part of the fetal life appears to be constant, the premaxilla being just as prominent at the tenth week of fetal life and onwards as at the end.

After birth (if the condition is untreated) the premaxilla maintains its position beneath the tip of the nose and does not become more prominent. If anything, due to the relatively greater vertical development of the face after birth, the premaxilla appears less noticeable. The nose itself grows in a normal manner, being neither unduly prominent nor recessive.

A large number of unoperated bilateral clefts have been reported in the literature. A typical and interesting example is the case shown in Angle's famous book[5] published in 1907 (Fig. 5-above). The age of this boy is not given but he appears to be about 8 years old. An older patient is a Nepalese boy aged 18 years (Fig. 5-below) treated in Nepal by Scott, a plastic surgeon at Alder Hey Children's Hospital, Liverpool. The features common to these cases are of a normal nasal structure, with a tendency to a widening of the interorbital distance and an increased width at the level of the alar base. The prolabium and premaxilla are variable in size but the position beneath the tip of the nose and the absence of a columella is common to all cases.

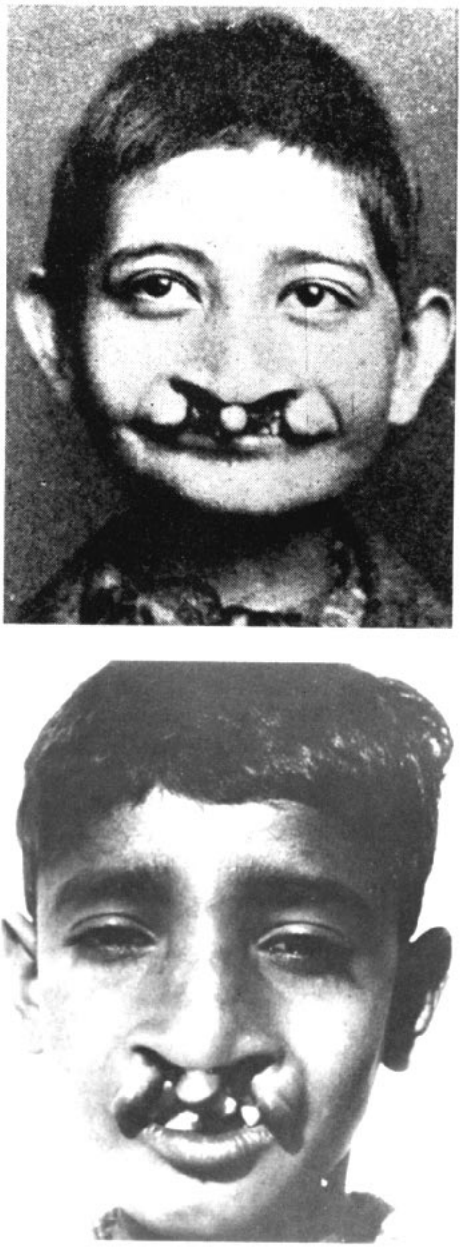

Fig. 5 Above, photograph of a youth with an unoperated bilateral cleft from Angle.[5] Below, an 18 year-old Nepalese male with an unoperated bilateral cleft.

It can, therefore, be seen in these cases which are typical of many reported in the literature that after birth there is no tendency for the premaxilla and prolabium to become more prominent than they are at birth. The growth pattern of the face is indeed remarkably good and must be regarded as favourable from the treatment point of view. Other reports in literature strongly support the concept of a favourable development of the unoperated cleft lip and palate face.[7,8,9]

## Comparative Anatomy of the Bilateral Cleft

Newborn animals, usually nonvital, were used for the anatomical study of bilateral clefts. The dog was the commonest animal received, (11 specimens) 4 were obtained of the pig, 3 of the sheep and 1 chimpanzee. In addition, a number of mouse specimens of the Ajax strain were available for study.

The specimens were prepared in various ways: 1) gross dissection; 2) maceration and preparation of the bony skeleton, the more delicate specimens such as the dog being stained with alizarin and embedded in clear plastic; and 3) embedding in low viscosity nitrocellulose and sectioning at 24$\mu$; sagittal, coronal and horizontal sections were prepared and stained with Masson's trichrome and also haematoxylin and eosin.

The morphology of the midline structures in the bilateral cleft lip and palate animal contrasts strongly with the condition in man. There is no protrusion of the premaxillary region. In the dog, for example, Figure 6, the premaxilla lies in an essentially normal position without distortion; this is confirmed by the study of the histological sections. The palatal view of alizarin stained plastic embedded specimens (Figure 7) shows a reduction in the size of the premaxilla (see Atherton)[10]

Fig. 6 Typical appearance of a newborn dog (beagle) with a bilateral cleft of the primary and secondary palate.

when compared with a control specimen but no increase in the length of the vomerine process of the premaxilla or alteration in the position of the premaxilla-vomer suture. As in the human, the size of the premaxilla may vary as may also the number of incisor teeth carried by the premaxilla. In the animals of which I have representative specimens, there is occasionally a striking reduction in the size of the premaxilla and in the number of teeth contained in it (Figure 8). In the pig the nasal septum may be affected and the central portion of the face be severely underdeveloped. There seems to be a species difference here, the deficiency of the midline tissues being more common in the pig than in the dog. Further information is needed, however, since the numbers are so small.

To study the further growth pattern of the bilateral cleft in an animal, a miniature Schnauzer was kept alive for three months. During this time lateral radiographs were taken of the head. Three of the serial tracings taken from the radiographs are shown in Figure 9. The general relationship of the premaxilla and nasal septum to the mandible was maintained and the overall development of the face was good. There was no tendency for the premaxilla to overdevelop.

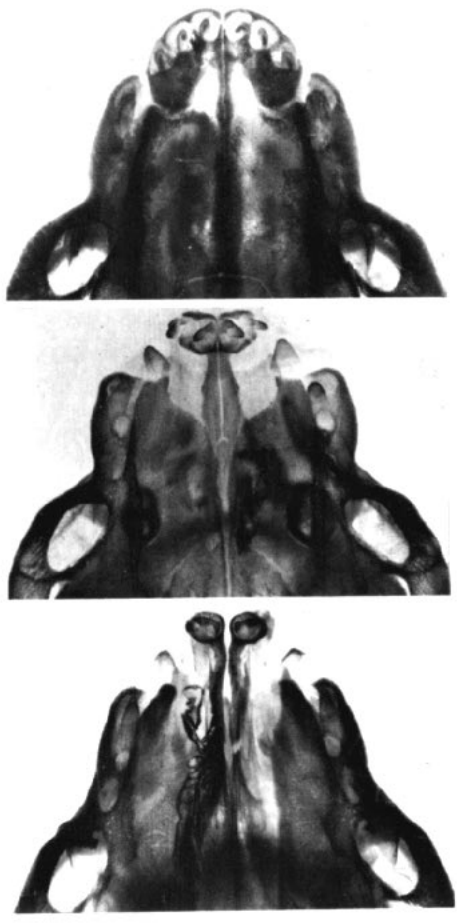

Fig. 7 Skulls of newborn dogs prepared by maceration, alizarin staining and embedding in Ward's Bioplast. Top, noncleft control beagle specimen. Middle, beagle with a cleft of the primary palate only. Bottom, beagle with a cleft of the primary and secondary palates.

It is concluded that in animals with a bilateral cleft the premaxilla does not differ in its relation to the nasal septum from that in the normal animal and that the growth potential is good.

## Hypothesis for the Cause of the Prominent Premaxilla in Man

At first acquaintance it would seem that man differs from other mammals

Fig. 8 Palatal view of newborn pig specimens. Left, noncleft control specimen. Middle, specimen with a bilateral cleft of the primary palate only showing a well-formed and substantial premaxilla carrying central and middle incisors. Right, specimen with a bilateral cleft of the primary palate only showing a small premaxilla and a congeminated central incisor. The nasal septum is thinner than normal and the whole anterior part of the face reduced in size.

in that the premaxilla is prominent in man with a bilateral cleft whilst in other mammals it retains its normal position. The hypothesis of this paper is that the true situation is that the position of the cleft premaxilla in man is similar in position to the premaxilla in animals, cleft or normal. What is *different* is the face of normal man. The prominent premaxilla arises in man because the cleft allows the face to assume a normal mammalian growth pattern. The cleft acts as a premaxilla-maxilla suture. What is peculiar about man is not the growth of the cleft face which is typically mammalian, but the growth of the normal face.

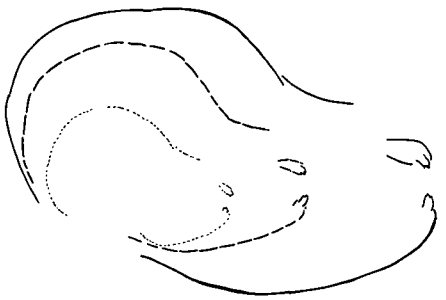

Fig. 9 Tracings from lateral radiographs of the head of a Schnauzer pup. Inner dotted line, birth. Inner broken line, 24 days old. Outer complete line, 96 days old.

The normal face of man differs from other animals in the absence of a premaxilla-maxilla suture. The consequence is that there is a restriction in the forward growth of the anterior part of the maxilla, the superior part of which becomes attenuated to form the anterior nasal spine by its attachment to the nasal septum through the septopremaxillary ligament described by Latham.[11] A further consequence of restricted forward growth is the production of the columella. Both the columella and the anterior nasal spine are unique to and characteristic of man.

The "experiment" of nature which produces an artificial suture in the form of a cleft allows the premaxilla to be carried forward in the "normal" mammalian growth pattern. Because of the absence of restriction on growth there is no columella and the anterior nasal spine is diminuitive. The premaxilla-vomer suture responds by bone apposition producing an elongation of the vomer and vomerine process of the premaxilla in the same manner as the

cranial bones respond to intracranial pressure.

It should also be observed that the forward inclination of the alveolus with vertical incisors resembles such mammals as the dog. Further supportive evidence is shown by the growth pattern of the cleft face. The prominent premaxilla does not get more prominent with age; it is in a prominent position as early as specimens permit examination and maintains this position through fetal life and through to maturity.

## Clinical Relevance

It is not the purpose of this paper to discuss in detail the case management of the bilateral cleft but to suggest certain principles which should be followed in the treatment of the cleft. Whilst early treatment should be designed to fit the baby into the norm of society, it should also be borne in mind that the condition will not deteriorate. No drastic action in my opinion should be taken which may lead to a reduction in this favourable growth process.

The first object of treatment is, of course, to close the lip. To give the plastic surgeon the best chance for a successful operation several methods have been devised to bring back the premaxilla. The technique used by Burston[12] and others of restraining the forward growth of the premaxilla from birth fits in well with the theme of this paper. It is doubtful whether the premaxilla as a whole is restrained and the position of the anterior nasal spine in relation to the anterior nasal septum altered; the improved position of the premaxilla is largely due to the alteration in angle of the alveolar component of the premaxilla.

After lip closure at approximately four months and the subsequent closure of the palate at about twelve months, the next stage is the restoration of a more normal columella. The full development of a columella does not follow the postnatal restriction of the forward development of the premaxilla so that it must be accepted that the columella can only be completed by surgery. The question arises at what age should this operation be done. It seems logical that the columella should be restored as early as possible so that the nose can assume its normal shape. The age of 3-4 years is recommended by Maisels[13] using the Millard fork flap technique. This is a balance between an early operation date and having the benefit of an operation site which has grown sufficiently well to perform a precise operation.

Unfortunately, no hard and fast rules can be applied to this biologically variable problem and there are cases to which this outline of treatment cannot be applied. Medical, social and completely unforeseen factors may make presurgical orthopaedic treatment impossible or not as adequate as it might be. Then there seems nothing inherently wrong with surgically repositioning the premaxilla. Two considerations must be borne in mind. An operation which impairs the vomer-premaxilla suture by removal of the suture and subsequent scarring may lead eventually to an anterior crossbite by restricting the forward movement of the premaxilla. Damage to the nasal septum may also be a hazard to normal forward development. However, the type of operation which slides the premaxilla over the vomer would appear to have few drawbacks and permit the subsequent growth at the bone surfaces of the premaxilla-maxilla suture.

Whether the premaxilla is retracted by presurgical orthopaedics or by premaxilla slide-back operation, there is a tendency for "relapse" to take place, a process familiar to orthodontists. At

the time of lip operation, therefore, the position of the premaxilla should be secured by a closure of the anterior part of the palate which is thereby locked in position.


## Summary

The face of a newborn baby with a bilateral cleft of the lip and palate presents certain characteristic features, the most obvious of which are the prominence of the premaxilla and prolabium, and the absence of a columella. A comparison of angular measurements from cephalometric radiographs of bilateral cleft and of secondary palate cleft babies shows that the nasal septum is not overdeveloped antero posteriorly although it is reduced in height. The anterior nasal spine in the bilateral cleft group is more anteriorly placed on the nasal septum than that of the secondary palate cleft group. The characteristic deformities associated with the bilateral cleft arise early in fetal life and persist through birth and on to maturity without becoming more severe. In bilateral cleft animals (dog, pig, sheep and chimpanzee) the premaxilla is not displaced forward. It is hypothesised that in man the cleft acts as a suture between the maxilla and premaxilla and allows the premaxilla and prolabium to be carried forward with growth so that the situation in cleft-affected man resembles the general growth pattern of the mammalian face. When the human face is growing normally, the absence of a premaxilla-maxilla suture restricts the forward growth of the premaxillary region leading to the production of the typical human columella and anterior nasal spine. Treatment should not inhibit the favourable growth pattern of the cleft face and, as an initial treatment, presurgical orthopaedic treatment would seem the most suitable means of repositioning the alveolar component of the premaxilla.



*School of Dental Surgery and*
*Alder Hey Children's Hospital*
*University of Liverpool*
*Liverpool, 3, England*



## Acknowledgments

I am greatly indebted to Dr. W. R. Burston for the use of the lateral cephalometric films of cleft and control babies, to Professor J. Van Limborgh for his permission to study and photograph the skull shown in Figure 2, to Dr. R. A. Latham for his permission to publish Figure 4, and to Mr. C. B. Scott for Figure 5, below. I wish to thank Mr. Ken Walters of the photographic department of the Institute of Child Health, Alder Hey Children's Hospital for the photographs.


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