Converse: Chapter 38

CLEFT LIP AND PALATE: INTRODUCTION

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History

There is an excellent historical review of the subject of cleft lip and palate by Dorrance (1933) and another historical review by Rogers (1971). This introduction will be concerned only with establishing historical trends in the treatment of this congenital anomaly.

The Age of Empiricism

In their approach to the problem of cleft lip and palate, surgeons through ages have attempted to correct the abnormal arrangement of the cleft lip and palatal tissues and achieve a "normal" appearance.

Boo-Chai (1966) reported a case of successful closure of a cleft lip at approximately 390 A.D. in China, although the surgeon's name is not mentioned. Yperman (1295-1351) was a Flemish surgeon who appears to have written the first fully documented description of cleft lip and its surgical repair. He closed the freshened borders of the cleft lip with a triangular needle armed with a twisted wax suture, a common method of suture at the time. In order to approximate the internal and external wound edges, he reinforced the closure with a long needle passed through the lip some distance from the edges of the cleft; the needle was held in place by a wraparound ligature-of-eight thread. A similar technique of lip closure was still being performed by Pancoast in 1844.

Palatal deformities caused by syphilis and gunshot wounds interested Jacques Houllier (cited by Gurlt, 1898), who appears to have been the first to propose direct suture of palatal perforations. However, the failure rate was high, and he suggested that, when surgery failed, the region could be occluded with wax or a sponge. Franco (1556) wrote: "... cleft lips are sometimes cleft without a cleft of the jaw or palate, sometimes the cleft is only slight, and at times the cleft is as long and as wide as the lip" (Rogers, 1967). In 1561 he wrote: "Those who have cleft palates are more difficult to cure; and they always speak through the nose. If the palate is only slightly cleft, and if it can be plugged with cotton, the patient will speak more clearly, or perhaps even as well as if there were no cleft; or better, a palate of silver or lead can be applied by some means and retained there". Palatal occlusion by plates of gold or silver was also described in 1564 by Pare, who designated such a plate as an "obturateur": Pare (1975) was also the first to use the term "bec-du-lievre" ("harelip").

Tagliacozzi (1597) described a lip closure utilizing mattress sutures passed through all layers of the lip tissue. This was a departure from the prevailing technique of needle closure and figure-of-eight suture material reinforcement. Thus, in the sixteenth century, closure of cleft lip to improve appearance was widely practiced, and the need for closure of the cleft palate to improve speech was appreciated in more limited surgical circles.

Treatment of the protruding premaxilla using a head bandage to achieve external compression of the premaxillary segment, thereby reducing it to a more favourable position for lip closure, was introduced by Desault and Bichat (1798). Over the years, various combinations of intraoral and extraoral devices were developed in order to reduce the protruding premaxillary segment and also to maintain the lateral arch segments in adequate anatomical relationship with the lower jaw. At the present time, these efforts appear to have reached their latest stage of development in the sophisticated work of Georgiade and Latham (1973).

The origins of the present techniques for successful closure of the secondary cleft palate are found in the early work of von Graefe and Roux, who in 1816 and 1819, respectively, closed the cleft of the *soft palate* with interrupted twine sutures. In Roux's patient, a dramatic change in the patient's voice was immediately noted and described.

Direct closure of the *hard palate* followed in 1826. Dieffenbach recommended that clefts of the hard palate could be closed by separating palatal mucosa from the bone. While he also recommended lateral relaxing osteotomies to close clefts of the secondary palate, he did not employ these until 1828. This technique is still employed in certain centers at the present time.

Early closure of the soft palate to induce a narrowing of a wide cleft of the hard palate was mentioned in 1828 by John C. Warren of Boston. This approach to wide clefts of the hard palate was repopularized by Schweckendiek in 1962 and is currently the subject of much debate. Langenbeck in 1859 and 1861 emphasized the need to elevate the periosteum with the palatal mucosa, thus forming bilateral mucoperiosteal flaps. This flap technique is still in use in some centers today. Veau drew attention to the fact that palatal lengthening was not achieved by this technique, launching a full-scale attack on the technique in the Deutsche Zeitschrift für Chirurgie in 1936 (Converse, 1962). He converted Langenbeck's bipedicle flaps into single pedicle flaps based on the descending palatine vessels. Modifications of Veau's basic techniques were made by Wardill (1937), Kilner (1937), and Peet (1961), resulting in a push-back technique for closure of clefts of the secondary palate that is widely used today. Simultaneous lengthening of the nasal surface of the velum can be accomplished by the Cronin modification (1957).

Mirault introduced the modern cross-flap technique of lip closure in 1844, and since that time nearly every conceivable type of flap - triangular, rectangular, or curvilinear - has been tried. Mirault's technique remained popular and was advocated during the twentieth century by Blair and Brown (1930). Further modification of cleft lip closure was described in 1884 by Hagedorn, who devised a rectangular flap technique to prevent linear contracture. This procedure appears to have led to the operation of LeMesurier in 1949. During this period Z-plasty techniques were also used in various guises to relieve the tendency to linear scar contracture. This line of endeavor led to the Tennison (1952) low triangular flap technique and the high Z-plasty-rotation flap of Millard (1958).

Throughout the evolution of the techniques of treatment for cleft lip and palate, therapy for ancillary problems such as dentoalveolar arch deformities, nasal abnormalities, maxillary hypoplasia, and speech difficulties had also progressed to a point where, in modern

times, teams of specialists have been formed to manage the total problem, grown too complicated for one or two disciplines alone.

Management of the arch deformity in cleft palate by techniques of banding and prosthetic stabilization failed to achieve the goal of an adequate upper-lower dental arch relationship in early approaches to this problem. Orthodontic therapy proceeded during the period of the eruption of the permanent teeth, usually during the period of mixed dentition, and often after years of treatment, a class III malocclusion with significant crossbite remained.

This led dental inovators such as McNeil (1954) and Burston (1958) to advocate orthodontics in the first year of life in an attempt to establish proper arch relationships. They postulated that early alignment of arch segments would aid normal development of the maxilla. Arch position was maintained by appliances, initially a combination of internal and external appliances and finally a simple internal appliance. However, removal of the retaining appliance before puberty often resulted in recurrence of the original arch deformity. It was then thought that perhaps primary bone grafts might (1) stabilize the arch and (2) either grow or promote growth of the uninhibited maxilla. These speculations, however, had no scientific basis.

The effectiveness of primary bone grafting in the treatment of cleft palate arch deformities has not been satisfactorily determined. Initially, surgeons attempted bone graft in the region of the incisive foramen in an effort to improve their statistics on successful palatal closure (Lexer, 1908; Drachter, 1914). To produce adequate bony continuity between the premaxilla and lateral bony segments appeared to some surgeons such as Axhausen (1952) as the "final problem in the repair of complete clefts at the present time". The mere presence of the bony gap was enough to inspire a general surgical rush to fill it. However, as will become apparent in Chapter 48, filling the gap was not the end of the matter. Bone grafts appear to be unable on their own to "hold apart" any arch that has a tendency to collapse; the bone graft absorbs under pressure. Nor do primary bone grafts grow as was originally postulated, but rather they hinder growth with a significant limitation of maxillary development and a dramatic increase in crossbite malocclusion and pseudoprognathism (Kling, 1964).

Moreover, as the story of primary bone grafting in cleft palate surgery unfolds, it tends to confirm the prescience of Pruzansky, who in 1964 condemned the unscientific and unsubstantiated use of primary bone grafting when bone graft fever was sweeping many surgical circles.

In retrospect, however, we must marvel at the ingenuity of surgeons of the past who made major progress utilizing the trial and error method in an era when corollary scientific information was virtually nonexistent. There were, however, surgeons throughout history who attempted to apply their knowledge of anatomy and physiology and use scientific discipline in the design of their surgical procedures. The anatomical observations of Pancoast (1844) led him to design a specific operation, in which he divided the insertion of the palatal muscles "so as to prevent their straining the sutured edges of the palate assunder". Fergusson (1844-1845), noting that most palatal repairs disrupted, conducted a series of anatomical studies leading him to design an operation which divided the levator veli palatini muscles, the posterior tonsillar pillars, and, on occasion, the anterior tonsillar pillars. The incision provided

relaxation to the muscles and tissues of the palate in order to prevent lateral pull. The father of modern surgery of cleft lip and palate, Victor Veau, spent many long hours studying embryological specimens. His contribution to the study of cleft lip and palate in and outside of the operating room are significant.

The Scientific Approach

In surgery of the cleft lip, Veau (1931, 1938) pointed out the paucity of muscle fibres in the medial aspect of the unilateral cleft and also in the prolabial segment of the bilateral celft lip:

"The median border of the cleft lip is sterile. This anatomic fact, the inadequacy of the musculature of the median aspect, should provide us with a surgical directive: Demand nothing from the inner aspect which is sterile, utilize to the maximum the muscles of the lateral aspect which is fertile, sacrifice all of the mucosa of the inner aspect, but preserve carefully all of the mucosa of the lateral aspect.

The principal cause of the mediocre results obtained in bilateral cleft lip repair is the absence of muscle in the prolabial segment of the lip. One can hope for contour and shape approaching the normal only if the lip contains muscle. I have long emphasized this fact: The muscular sterility of the prolabial segment."

In the treatment of the bilateral cleft lip, Veau was one of the first to allow the pressure of the repaired lip to recess the premaxilla.

"We are operating on faces in full evolution. The profile of the face will be submitted to a dual transformation. In the nose, the vomer will grow on condition that it has not been altered and it will increase the projection of the nose. In the lip, the reconstituted muscular ring in front of the premaxilla will push it backward. The operation of the cleft lip in the newborn is not an ordinary definitive operation of the type one does in plastic surgery in the patient in whom growth is completed. Our role, in the newborn, is to create conditions of development as close to the normal as possible."

In condemning surgical intervention upon the vomer to recess the premaxilla, Veau wrote:

"In order that the face of the newborn becomes a normal adult face, a series of unknown factors must come into play. All of these factors have their role in the distribution of forces which create the definitive form. They are the instrumental contributions the assembly of which makes the harmony we know. Eliminate the violins and you will no longer recognize a Beethoven symphony. That is what we have done (by sectioning the vomer) in the treatment of bilateral cleft lip: We have done away with the axial beam supporting the evolution of the face."

It was in embryology, however, that Victor Veau made his greatest contribution. His career as an embryologist started when he was over 60 years of age.

"I am only a surgeon, yet circumstances have led me to play the role of an embryologist... Yesterday, everyone said "cleft lip is caused by the absence of coalescence of the processes of the face". Tomorrow, they will say, "cleft lip is caused by the persistence of the subnarial epithelial membrane".

This concept is not my own; it is the concept of Professor A. Fleishmann, who is still living in Erlangen, where he spent his entire academic career as Professor of Zoology. I have been, however, the gardner who has been responsible for the growth of the small plant, once it was germinated. The embryologists ignored Fleishmann, or only referred to his hypothesis with irony. I showed that Fleishmann's hypothesis could be applied to all clinical varieties of the cleft lip malformation and, in addition, I have supported the hypothesis by embryological findings outlined in drawings of the stages of development of the subnarial region.

The only productive methods were those which approximated normal development.

In 1935. I wrote a paper entitled: "Hypothesis of the initial malformation of the cleft lip". Staying on clinical grounds, I showed that the theory of the facial processes fitted poorly with what I observed in the cleft lip. The theory of Fleischmann, on the other hand, appeared to be the key to all the anatomical details and clinical varieties of the deformity. This was an indirect attack on the classical theory.

Professor Hochstetter was the first to describe the oronasal membrane, which is an incomprehensible finding according to the theory of the facial processes.

Embryologically, the oronasal membrane which plays a role in the cleft lip is constituted by two fundamentally different formations: (A) The floor of the nose between the integument and the nasopalatine canal. This region is formed by the primary palate, a very precocious embryonic structure (5 mm, 2 nd week) which appears when the mesoderm has invaded the epithelial wall (7 mm) and is definitively constituted when bone has commenced to differentiate into the undifferentiated mesenchyme (11 mm, 5th week). (B) The hard and soft palate. This long partition is constituted by the secondary palate, a relatively late embryonic formation, definitively constituted when the palatine processes have achieved their fusion (30 mm, 12th week). The malformation in the secondary palate is the congenital cleft of the palate. Most often (6 out of 10) the malformation of the primary palate, the true cleft lip, is associated with a malformation of the secondary palate and the two deformities form a tetralogic entity which is disociable because of its embryonic origins, but which forms, nevertheless, a clinical and surgical entity.

The gist of Fleischmann's hypothesis consists in the following: The cleft palate is the arrest of the disappearance of the epithelial membrane which remains intact, not penetrated by the adjacent mesoderm.

Contemporary Theories

Throughout the historical development of the treatment of cleft lip and cleft palate, different aspects of the problem have alternately received priority. At the time of publication of this book, there appears to be an emphasis on the problem of crossbite and malocclusion following cleft lip and palate surgery.

Treatment of the anterior palatal deformity has been modified following the publication by Walker, Collito, Mancusi-Ungaro, and Meijer (1966) of data indicating the deleterious effect of extensive lateral undermining to facilitate the lip repair. The authors suggested that utilizing the technique of lip adhesion, followed in several months by lip closure without lateral periosteal or soft tissue undermining, significantly reduces the incidence of crossbite and malocclusion.

In addition, early complete closure of the primary and secondary palates can also produce significant dental deformities. Ross and Johnston (1972) suggested that surgery should not be performed on the hard palate in areas adjacent to or abutting on teeth. As an alternative approach, Lindsay (1974) advocated a simple closure of the soft palate with obturation of the hard palate until age 2 or 3 years, when closure of the hard palate is completed by one of the "less" periosteally disturbing procedures, such as the von Langenbeck palatoplasty.

The effect of the later approach on speech development is not clear. In the past few decades out primary concern has been for speech, and we have been comfortable in the knowledge that if we gave something away in terms of initial dental management in order to obtain normal speech, it could be recouped later by advanced orthodontic techniques. However, it has now become evident that it is preferable *not* to yield on early dental management in order to decrease dental deformities. We must also be doubly alert to the effects of this "new" approach on speech. Speech compromises in early childhood might prove more difficult to correct later on than dental ones.

Classification

In the classification of Davis and Ritchie (1922), congenital clefts were divided into three groups according to the position of the cleft in relation to the alveolar process:

Group I, prealveolar clefts, unilateral, median, or bilateral.

Group II, postalveolar clefts involving the soft palate only, the soft and hard palates, or a submucous cleft.

Group III, alveolar clefts, unilateral, bilateral, or median.

Veau (1931) suggested a classification divided into four types:

1. Cleft of the soft palate only.

2. Cleft of the hard and soft palate extending no further than the incisive foramen, thus involving the secondary palate alone.

3. Complete unilateral cleft, extending from the uvula to the incisive foramen in the midline, then deviating to one side and usually extending through the alveolus at the position of the future lateral incisor tooth.

4. Complete bilateral cleft, resembling type 3 with two clefts extending forward from the incisive forament thorugh the alveolus. When both clefts involve the alveolus, the small anterior element of the palate, commonly referred to as the premaxilla, remains suspended from the nasal septum.

Kernahan and Stark (1958) recognized the need for a classification based on embryology rather than morphology. The roof of the mouth - from the incisive foramen or its vestige, the incisive papilla, to the uvula - is termed the secondary palate. It is formed after the primary palate (premaxilla, anterior septum, and lip). The incisive foramen is the dividing line between the primary and secondary palates.

A cleft of the secondary palate is further classified as incomplete or complete, depending upon its extent. An incomplete cleft is the common cleft of the velum, while a complete cleft includes both the velum and the hard palate as far as the incisive foramen. To this classification must be added the cleft of the mesoderm of the palate, or submucous cleft, which may be camouflaged unless the uvula is cleft. While it may not be easy to detect dehiscence of the velum musculature, the presence of velopharyngeal incompetence and palpation of a notching of the posterior nasal spine aid in the diagnosis.

Kernahan (1971) subsequently proposed a striped Y classification. As in the previous classification, the incisive foramen is the reference point. With stippling of the involved portion of the Y, the system provides rapid graphic presentation of the original pathology.

Harkins and associates (1962), at the instigation of the American Cleft Palate Association, presented a classification of facial clefts based on the same embrylogic principles used by Kernahan and Stark. A modified version follows:

I. Cleft of Primary Palate

A. Cleft Lip

(1) Unilateral: right, left

(a) Extent: one-third, two-thirds, complete

(2) Bilateral: right, left

(a) Extent: one-third, two-thirds, complete

(3) Median

(a) Extent: one-third, two-thirds, complete

(4) Prolabium: small, medium, large

(5) Congenital scar: right, left, median

(a) Extent: one-third, two-thirds, complete

B. Cleft of Alveolar Process

(1) Unilateral: right, left

(a) Extent: one-third, two-thirds, complete

(2) Bilateral: right, left

(a) Extent: one-third, two-thirds, complete

(3) Median

(a) Extent: one-third, two-thirds, complete

(4) Submucous: right, left, median

(5) Absent incisior tooth

2. Cleft of Palate

A. Soft Palate

(1) Posteroanterior: one-third, two-thirds, complete

(2) Width - maximum (mm)

(3) Palatal shortness: none, slight, moderate, marked

(4) Submucous cleft

(a) Extent: one-third, two-thirds, complete

B. Hard Palate

(1) Posteroanterior

(a) Extent: one-third, two-thirds, complete

(2) Width - maximum (mm)

(3) Vomer attachment: right, left, absent

(4) Submucous cleft

(a) Extent: one-third, two-thirds, complete

3. Mandibular Process Clefts

A. Lip

(a) Extent: one-third, two-thirds, complete

B. Mandible

(a) Extent: one-third, two-thirds, complete

C. Lip Pits: Congenital lip sinuses

4. Naso-ocular: Extending from the narial region toward the medial canthal region.

5. Oro-ocular: Extending from the angle of the mouth toward the palpebral fissure.

6. Oro-aural: Extending from the angle of the mouth toward the auricle.

Spina (1974) modified and simplified the above classification as follows:

Group I. Preincisive foramen clefts (clefts lying anterior to the incisive foramen). Clefts of the lip with or without an alveolar cleft.

A. Unilateral

B. Bilateral

C. Median

Group II. Transincisive foramen clefts (clefts of the lip, alveolus, and palate).

A. Unilateral.

B. Bilateral.

Group III. Postincisive foramen clefts.

Group IV. Rare facial clefts.

Epidemiology

Clefts of the lip, with or without clefts of the palate (CL/P), must be distinguished from isolated clefts of the hard and soft palates (CP) because of different embryologic, etiologic, and epidemiologic backgrounds (Fogh-Andersen, 1942; Fraser and Calnan, 1961).

Clefts of the secondary palate can be induced in the mouse by teratogens after the primary palate has formed; moreover, clefting of the secondary palate in association with clefts of the primary palate probably represent a secondary (tongue positioning) rather than a primary defect (Trasler and Fraser, 1963).

Family studies have also shown that siblings of patients with CL/P have an increased incidence of CL/P but not of isolated CP; conversely, siblings of patients with CP have an increased frequency of CP but not of CL/P (Fogh, Andersen, 1942; Woolf, Woolf and Broadbent, 1963a, b).

The collection of epidemiologic data is associated with many problems. The surveys are conducted from three sources: birth certificates, hospital records, and habilitation or surgical records. The most accurate data are collected from the records of better hospitals. Birth certificates are often hastily completed and lacking in detail. Habilitative or surgical records tend to be slanted toward a certain segment of the cleft lip and palate population. Other factors which must be considered in evaluating the data include the percentage of ascertainment, the racial and socioeconomic make-up of the population segment under study, the quality of the records, and the absence of detail such as degree of clefting and presence of associated anomalies.

The reader is referred to the publications of Fraser (1970) and Ross and Johnston (1972) for a complete discussion of the epidemiology of cleft lip and palate.

Incidence. In the classic studies of Fogh-Andersen (1942), the overall frequency of cleft lip and palate in Denmark was reported as 1.47 per 1000 live births; the incidence of CL/P was 1.16 and that of CP was 0.34 per 1000 births. A similar overall incidence was reported by Woolf, Woolf, and Broadbent (1963a) for a section of the United States and by Wilson (1972) for a region in Great Britain. Racial differences will be discussed later in the chapter.

In most series cleft lip with palate involvement is reported as 1.5 to 3.0 times as frequently as isolated cleft lip. Isolated cleft palate (CP) occurs in a frequency of 0.39 to 0.50 per 1000 births (Drillien and associates, 1966).

In a follow-up study of the Danish statistics, Fogh-Andersen (1961) noted an increase in the number of patients operated upon from 1.31 per 1000 live births (period from 1938 to 1942) to 1.64 per 1000 live births (period from 1953 to 1957). In a German study (Tünte, 1969) confined to CL/P, an increased incidence of approximately 50 per cent over a 50-year interval was observed, and it was felt the rise could not be attributed to underreporting alone. Fára (1975) in a Czech study has also noted an increased incidence of cleft lip and palate.

Many factors could account for the observed increased frequency of cleft lip and palate. A fall in postnatal mortality, and/or a decrease in operative mortality associated with improvements in anesthesia. In addition, contemporary rehabilitative and surgical efforts yield results that are so favorable that many more of the patients marry and transmit their genetic potential for clefting.

Fogh-Andersen (1942) noted a distribution according to type of cleft of 25 per cent cleft lip alone, 50 per cent cleft lip and palate, and 25 per cent isolated cleft palate. Ingalls and associates (1964) reported a respective frequency of 16 per cent, 30 per cent, and 54 per cent. Fraser and Calnan (1961) reported 21 per cent, 46 per cent and 33 per cent. Unilateral left-sided clefting, unilateral right-sided clefting, and bilateral clefting occur in a 6:3:1 relationship (Wilson, 1972).

As noted by Fogh-Andersen (1942) and confirmed by other studies (Fraser and Calnan, 1961; Ingalls and coworkers, 1964; Fraser, 1970; Wilson, 1972), there is a left-sided preponderance of cleft lip. In addition, there is a male excess in CL/P and a female excess in isolated CP. Cleft palate is more often associated with bilateral (86 per cent) than with unilateral (68 per cent) clefts of the lip (Fraser, 1970), and this finding is consistent with the concept that cleft palate is seen in the more severe type of lip deformities (Fogh-Andersen, 1942).

Racial Influences. While the Caucasian race has been the most extensively studied, there are also statistics available for the incidence of cleft lip and cleft palate in the Negro and Oriental races.

The mean incidence of CL/P in Caucasians is approximately 1 per 1000 population (Fraser, 1970). A higher frequency of CL/P among Japanese infants was reported as approximately 2.1 per 1000; the incidence rate for CP was 0.00055 (Neel, 1958). The data of Fujino and associates (1963) also support the findings of an increased frequency among Orientals.

The Negro race in the United States has been extensively studied, and it was noted that Negroes have a considerably lower risk for cleft lip and palate (CL/P) than do Caucasians. In a large collaborative survey of births in several university hospitals, the frequency of CL/P per 1000 births was 1.34 for whites and 0.41 for Blacks (Chung and Myrianthopoulos, 1968). Review of birth records in two Washington hospitals serving mainly Black patients (Altemus and Ferguson, 1965) and in a similar hospital in New Orleans (Longenecker and coworkers, 1965) showed a decreased frequency for CL/P in Black individuals. Ivy (1962) reported similar data from Philadelphia.

Unlike the data for CL/P, there is less evidence to show racial variation in the incidence of isolated cleft palate (CP) between Blacks and whites (Altemus and Ferguson, 1965; Chung and Myrianthopoulos, 1968).

Thus, there is supporting evidence that racial heterogeneity exists in the frequency of CL/P in a descending order of frequency amont Orientals, Caucasians, and Blacks. There appears to be no such heterogeneity in the incidence of isolated cleft palate (CP) among the three races; it is approximately 0.5 per 1000 births.

Sex Ratio. There is an excess of males in CL/P, the proportion ranging from 60 to 80 per cent (Drillien, Ingram and Walkinson, 1966). Fogh-Andersen (1942) noted that male preponderance is more marked in the more severe or complete CL/P defects and in bilateral rather than unilateral clefts. Male excess in CL/P is less pronounced among the Japanese (Fujino and coworkers, 1963).

Female excess has been reported in isolated cleft palate (CP) (Fogh-Andersen, 1942; Fraser and Calnan, 1961). In addition, those clefts extending more anteriorly toward the incisive foramen are far more frequent in females.

Parental Age. Birth order appears to play no role in the development of either CL/P or CP.

There is some evidence indicating that the risk of producing a child with CL/P is decreased in younger parents and increased in older parents (Woolf, 1963); Fraser and Calnan (1961) felt the most important factor was elevated parental and not maternal age.

A significant positive relationship between parental age and isolated cleft palate (CP) could not be demonstrated (Woolf, Woolf and Broadbent, 1963b).

Genetic Factors. As discussed earlier, there is a significant increase of CL/P among relatives of CL/P propositi, but isolated cleft palate (CP) occurs in a frequency expected in the general population. Conversely, there is an increased frequency of CP among relatives of patients with CP without an increased incidence of CL/P (Fogh-Andersen, 1942; Woolf, Woolf and Broadbent, 1963a).

Fogh-Andersen (1942) described the inheritance of CL/P as of "variable expressivity", and Roberts (1964) suggested it had a "multifactorial etiology" dependent on multiple genes and environmental factors. Affected females with CL/P have a higher frequency of affected offspring than do affected males with CL/P (Woolf and coworkers, 1964).

The following table of risk was established (Ross and Johnston, 1972) based on the data of various authors:

Affected Relatives	Predicted Recurrence	
	CL/P	CP
One sibling	4.4	2.5
One parent	3.2	6.8
One sibling, one parent	15.8	14.9

The risk to siblings being born of unaffected parents rises from 4.4 per cent to approximately 9 per cent after two affected childern are born (Curtis and associates, 1961).