Studies on populations of individuals with cleft lip or palate, or both, indicate that their facial morphology is markedly different from that of the "noncleft" population. Knowledge is incomplete, but this chapter attempts to describe the abnormal face and tentatively to explain the causes of the abnormality. A discussion of the effects and the management of abnormal growth may lead to new approaches to the solution of associated clinical problems.

Growth Potential

The formation of an orofacial cleft in the embryo not only interferes with the development and morphology of the affected areas but also causes alterations in neighboring structures which are developmentally normal. The anatomical arrangement of muscles, nerves, arteries, veins, skin, and mucous membranes is distorted. These are described in Chapters 40 and 41.

It would be helpful in treatment planning if one could distinguish between structures which are intrinsically abnormal and those which only appear to be abnormal because they have become distorted by secondary environmental disturbances. The former are resistant to correction; the latter may be self-correcting if the proper environment could be provided.

One major topic in any discussion of facial growth is the adequacy of the growth potential that is inherent in a child with a facial cleft. If this factor were known, there would be a baseline from which to determine environmentally induced deviations. In spite of inadequate data, there are many clues available which permit a reasonable assessment of the inherent capabilities for growth and development.

It is currently believed that cleft lip and palate have a multifactorial etiology. There is evidence that one of the predisposing factors is a deficiency of midface mesenchyme at the critical embryonic stages (Veau and Politzer, 1936; Stark, 1954; Avery, 1961). This does not appear to be a simple genetic trait, since parents of children with orofacial clefts do not have a small maxilla (Coccaro and coworkers, 1970; Karout and Ross, 1974). Parents of affected children tend to have less convex profiles (Dixon, 1966; Pashayan, 1968), but this is relative and due to a larger mandible. If a general maxillary complex deficiency occurs and persists into childhood, clinicians must strive to protect the existing growth potential and, equally important, to protect the compensatory mechanisms which nature has provided to overcome minor deficiencies. These mechanisms will be discussed later.

Before examining the maxillary complex in detail, it might be well to establish a frame of reference by considering the other components of the facial skeleton. With the occasional exception of the mandible and the orbits, there is no good evidence that any other bone is primarily affected to any significant degree by the presence of cleft lip or palate. In the vast majority of cases the mandible is normal in size and shape at birth. There are as many
variations as in the unaffected population, presumably because the mandible is not directly involved in the production of a cleft and normal genetic variation is expressed. The one notable exception to this statement is the group of children with Pierre Robin anomalad, in which the mandible is small or retruded (see Chapter 50). There appear to be several variants of the Pierre Robin syndrome, as regards etiology. A discussion of the mandible and its subsequent growth will be presented later in this chapter.

The orbital complex is affected in that there is a tendency toward greater interorbital width (Psaume, 1957; Graber, 1964; Moss, 1965; Ross and Coupe, 1965; Aduss and coworkers, 1971; Farkas and Lindsay, 1972b), although hardly sufficient to warrant the term **orbital hypertelorism**. This may be a secondary characteristic induced by environmental forces. The zygomatic bones (Harvold, 1954; Subtelny, 1955; Coupe and Subtelny, 1960), the pterygoid plates of the sphenoid (Van Limborgh, 1964; Atkinson, 1966), and the cranial base (Ross, 1965; Bishara and Iverson, 1974) are essentially normal. For all practical purposes we may conclude that we are dealing with an intrinsically normal facial framework to which is attached a mildly deficient and moderately malformed maxillary complex (Ross and Johnston, 1972).

**Prenatal Growth**

By the time of birth the face of the infant with cleft lip or palate has developed enormously from the embryonic condition. It has been subjected to many environmental forces in utero which influence the shape of the parts and their relation to each other. The reaction of the facial skeleton to external forces depends largely on the type of cleft which is present.

**Complete Unilateral Cleft Lip and Palate.** The infant with a complete unilateral cleft lip and palate has a maxilla which exhibits deficiencies of the alveolar and palatal bones (Peyton, 1934; Pruzansky, 1953; Stark and Ehrmann, 1958; Coupe and Subtelny, 1960; Huddard and coworkers, 1969) and perhaps the maxillary basal bone (Graber, 1950; Coupe, 1962; Kettle and Walther, 1966). Alveolar bone deficiencies may be related to the absence or abnormality of tooth development adjacent to the cleft, since growth of the alveolar bone is dependent on the presence of teeth (edentulous individuals have virtually no alveolar bone). The size of the palatal shelves at birth is difficult to ascertain, since many conflicting reports are available. One can assume that a deficiency in shelf width is not clinically significant in most cases, and the shelves grow at a normal rate thereafter. The tongue may intrude between the shelves and inhibit medial growth, causing a "clubbing" at the medial margin (Subtelny, 1955) and changing shelf angulation (Subtelny, 1955; Huddard and coworkers, 1969).

The most striking characteristic of the infant with complete unilateral cleft lip and palate is the severe deviation of the noncleft side of the maxilla away from the cleft. It carries with it the nasal structures, including the nasal septum. The explanation for this distortion is the sensitivity of poorly supported bone structures to pressure. In complete unilateral cleft lip and palate, the abnormal cheek muscle insertion on the maxilla at the base of the nose causes a rotating force on the larger segment during muscle contraction. This action is reinforced by tongue protrusion and perhaps by relatively unrestrained nasal septum growth (Latham and Burston, 1964). The smaller segment on the cleft side is exposed to less expanding force, and has a mild contracting force exerted by the base of the nasal ala on this side.
Since the nose is deviated toward the normal side, except for the alar base on the cleft side, the nostril on the cleft side is stretched and straightened. This configuration is established very early in the embryo (Atherton, 1967; Latham, 1969), and the alar cartilages develop and grow in a deformed matrix. The deformity is so well established by birth that the surgeon has great difficulty recontouring the nose to a satisfactory symmetry. It is quite possible that the stretching of the nostril on the cleft side is also sufficient to cause a retropositioning or growth inhibition of the smaller maxillary segment.

Measurements of dental models of the newborn (Peyton, 1931, 1934; Aduss and Pruzansky, 1968; Harding and Mazaheri, 1972) indicate that arch width is greater in cleft conditions. This finding confirms work (Subtelny, 1955; Ross and Coupe, 1965) on other facial widths (including the orbits as cited above) showing that the entire face is slightly wider in children with extensive clefts. This may be intrinsic but is more likely secondary to expanding forces as detailed above.

If the maxillary complex in a complete unilateral cleft continued to grow and maintain its infantile relationship with the remainder of the face, the adult would have poor jaw relations, characterized by excessive maxillary arch width and a severe deviation of the maxillary midline toward the normal side. The anteroposterior relations, however, would be reasonably normal.

Complete Bilateral Cleft Lip and Palate. Although infants with complete bilateral cleft lip and palate have a very different maxillary complex, the differences would probably be minimal without the activity of attached muscles. The major morphologic characteristics are the result of an altered response of the skeletal element to muscular deformation tendencies. The premaxilla, relatively unsupported on the nasal septum, is not able to resist the force of the active tongue and tilts forward. The base (anterior nasal spine) is somewhat supported by the septum and by the nose, so that excessive protrusion of the base is limited. If the tongue habitually protrudes through one side, the premaxilla will be protruded and forced to the opposite side, giving the asymmetry. It must be pointed out that this explanation is not universally accepted. Some authors (Latham, 1969, 1973; Pruzansky, 1971) believe that an intrinsic excessive nasal septum growth is responsible for protrusion of the premaxilla (see also Chapter 40).

The smaller posterior segments of the maxilla respond to their environment in the same way as in the unilateral condition. That is, there is an increase in maxillary width and possibly a posterior displacement or growth inhibition from an intrinsic deficiency or pressure from the alar base.

The anterior bone of the maxilla in humans is not a premaxilla as in other species. We refer to the "premaxilla" for convenience in bilateral cleft lip and palate, but this area is abnormal in complete bilateral cleft lip and palate, because the clefts have prevented mesenchyme from the maxillary processes from migrating in during embryonic development (Frazer, 1931). The result is a premaxilla grossly deficient in basal bone. The bulk of the bone present is alveolar bone, which develops in response to the development of teeth. This is apparent when teeth are removed in later life and the premaxilla is soon reduced to a tiny mass of bone under the anterior nasal spine. The great variation in size of the premaxilla at
birth in bilateral cleft lip and palate is usually a reflection of the number and size of teeth present.

**Other Types of Clefts.** Facial morphology in other types of clefts varies, but the same principles of deficiency and distortion apply. Isolated cleft palate (CP) presents no gross disturbance in form, although there is greater width than normal in the area of the posterior tuberosity. Clefts of the lip and anterior maxilla may show distortions in the alveolus and nasal structures similar to those of unilateral cleft lip and palate, but these are localized and the overall morphology of the face is close to normal. The explanation is simple: if there is continuity of bone in any area, the resistance to distortion is sufficient to prevent major problems. This principle applies to incomplete unilateral or bilateral cleft lip and palate, where part of the palate or part of the anterior maxilla is intact and less distortion is possible. Even a bridge of lip tissue across the cleft (Simonart's band) will prevent many of the expected morphologic changes from occurring, particularly those of the nose. The maxilla in these cases, however, may be rotated, since the tongue and cheek muscles are exerting abnormal forces.

**General Growth**

Children with cleft lip or palate, or both, are usually found to be smaller than children without clefts (Snodgrass, 1954; Johnson, 1960; Drillien and coworkers, 1966) and they may have a retarded skeletal age (Menius and coworkers, 1966; Przézdziak, 1969). Shibisaki and Ross (1969) found evidence that the pubertal growth spurt of facial structures is delayed in isolated cleft palate cases. Ross (1965) found that the cranial base in children with cleft lip and palate was smaller, but the proportions were identical.

Initial growth retardation can be explained by the feeding difficulties preoperatively and the trauma associated with surgical procedures. One would expect that these adverse environmental influences would have only a temporary effect, yet the study of Dahl (1970) indicated that young adults with all types of cleft were shorter than a similar control group by several inches. This would suggest that an intrinsic deficiency is present and "catch-up" growth does not occur. Thus, at any age, smaller facial structures can be expected.

**Growth of the Normal Face**

Before proceeding to a discussion of postnasal facial growth in children with cleft lip or palate, it would be helpful to have some understanding of normal facial growth. This is a complex subject, not completely understood at present, but there is sufficient available evidence to permit some relevant conclusions which will aid in the analysis of abnormal facial growth. This chapter will limit discussion to three important skeletal elements: mandibular basal bone, maxillary basal bone, and the dentoalveolar component of each.

**Mandibular Basal Bone**

Many of the bones of the body are not single morphologic units. The mandible, for example, has many components which develop relative to specific functions, all somewhat independently of each other. There is a central core, termed the basal bone, which is under strong genetic control; a coronoid process, which is dependent for its size and shape on
temporal muscle function and the position of the mandibular basal bone; a gonial area, which is dependent on the masseter and medial pterygoid muscles; and a dentoalveolar process which will be discussed later. These are the major components of the mandible.

The central core or basal bone of the mandible appears to be genetically determined in size and shape. Growth occurs only at the posterior portion of the bone, primarily at the condyle (Symons, 1951; Björk, 1955; Enlow, 1968; Koski, 1968). Orthodontists have attempted to alter the size of the mandible by applying forces in a variety of ways, but there is no good evidence that this is successful, so that a disproportionately large or small mandible must be altered surgically.

The position of the mandible, however, is the result of all the muscular, soft tissue, and external forces which act on it, since the bone is not firmly attached to other bones, only indirectly attached through the temporomandibular joints. Thus a change in the surrounding soft tissues or an external appliance such as the Milwaukee brace (Weinstein, 1967) will easily induce rotation to a new position. Rotations over a long period of time are accompanied by alterations in the bony muscle attachments and dentoalveolar component, so that the shape of the mandible can be dramatically changed without affecting the shape or size of the central core. Alterations in mandibular position and shape occur in cleft lip and palate, as will be discussed later in this chapter.

Maxillary Basal Bone

The delineations between the various functional components are less obvious in the maxilla. There is an orbital process, a nasal process, a palatal process, a zygomatic process, and a dentoalveolar process, all of which are relatively independent in the normal child. The basal bone (that which remains when the other processes are removed) is probably the result of a genetic blueprint, but with some critical differences from the mandible.

Despite the apparently firm attachment of the maxilla to the skull, strong environmental forces can alter the position of the maxilla, and interference with the sutures or nasal septum can alter maxillary size. The maxilla is thus more easily influenced in its growth than the mandible (Wieslander, 1963; Jakobsson, 1967; Droschel, 1973).

Essentially all maxillary growth in a forward direction occurs by apposition of bone to the posterior surfaces (Björk, 1955, 1966; Enlow, 1968). Björk (1966) and Enlow (1968) have shown that during growth there is actually resorption of bone in the anterior basal area to permit remodeling of the surface.

Koski (1968) and others have concluded that the areas of growth in the face are not primary growth centers that would force the maxilla forward. The role of the nasal septum is not completely understood, although its importance in directing maxillary growth is currently held to be of minor significance (Moss and associates, 1968; Stenström and Thilander, 1970). Moss (1962) has demonstrated that the sutures are adjustment areas which provide bone to maintain skeletal continuity. He has theorized that the face is controlled by what he termed "functional matrix". This is the sum of the nonskeletal elements of the face, including the spaces as well as the muscles and soft tissues. He considers, for example, that respiration is one of the controls over maxillary growth.
Ross and Johnston (1972) have concluded that there is no major skeletal force driving the maxilla forward, that it seems rather to drift forward as part of the overall genetic and environmental pattern of growth, and that, as the maxilla moves forward, bone is laid down in convenient places (i.e., in the sutures and on the maxillary tuberosity).

Increase in width of the basal maxilla occurs in infancy in conjunction with activity in the midpalatine suture. This suture does not contribute further after the second year of life (Moyers, 1963; Sicher, 1965), and width in the anterior region of the maxilla remains constant from this time. The increase in maxillary width is due to greater posterior width concurrent with apposition on the tuberosity.

Dentoalveolar Processes

The dentoalveolar processes are truly remarkable adaptive mechanisms. Alveolar bone develops in response to the presence of teeth and disappears if the teeth are absent or lost in later life. Teeth do not erupt at predetermined distances along a predetermined path into occlusion. If they did, a precise meshing of maxillary and mandibular teeth would be extremely rare. Nature has provided a mechanism to compensate for minor disharmonies between the maxilla and mandible.

A tooth erupts until it meets resistance. In the normal course, this resistance is the tooth in the opposing arch, although the tongue, thumb, or lip may interfere with full eruption. Once contact is made, an equilibrium is established.

As the tooth erupts, a force exerted against it is transmitted to the alveolar bone through the fibers of the periodontal ligament (as pressure on one side of the tooth socket and tension on the other. The bone resorbs in response to pressure on the fibers and builds in response to tension on the fibers. Thus the tooth moves away from pressure. An erupting tooth is guided into position by the gentle pressure of the soft tissues, principally the tongue, lip, and cheek muscles. The orthodontist uses this normal biological mechanism in the correction of malocclusion by applying artificial forces to move teeth.

It is essential for the attainment of a good occlusion that the dentoalveolar mechanism is free to respond to the guiding forces in the mouth. This principle is important to the later discussion on the occlusion in the operated and nonoperated cleft lip and palate.

Facial Growth in Unrepaired Cleft Lip and Palate

There have been numerous reports in the literature of older children and adults with cleft lip and palate who have not received the benefits of surgical repair of the cleft. Their facial form is the result of the interaction of an intrinsic defect, subsequent normal and abnormal environmental influences, and the expression of the growth potential of the facial skeleton. Careful study of these individuals should provide answers to many of the questions which arise regarding the disturbed facial morphology frequently encountered in a cleft palate clinic.

In individuals with unrepaired cleft lip and palate there is evidence that the face is wider in all areas, probably as a result of environmental forces which cause a decrease in the
restraining factors (loss of lip continuity, loss of skeletal continuity) while maintaining the expansive forces (tongue pressure, facial growth processes). There is also the possibility that an increased width is inherent and may even be an etiologic factor in the formation of a cleft. In any case, the areas of the facial skeleton responsible for increasing width appear to function normally in the growing child with cleft lip and palate. Areas of bone apposition and suture growth can be neither deficient nor inhibited if the end result is as satisfactory as is found in all studies on unoperated clefts of the lip and palate.

The facial growth parameter which is of most concern in cleft lip and palate is undoubtedly midface depth. Studies by Graber (1950, 1954), Ortiz-Monasterio and coworkers (1959), Mestre and coworkers (1960), Hagerty and Hill (1963), and Boo-Chai (1971) indicate that the basal maxilla achieves normal relationships with the remainder of the face in the unoperated adult with cleft lip and palate. Again, it must be concluded that the areas of bone growth are neither intrinsically abnormal nor inhibited by the presence of a cleft.

Data on facial height are not adequate to permit firm conclusions. The available evidence indicates, however, that vertical height is normal, and there is no evidence that there is a deficiency of midface height. Most of the increase in the normal maxilla is related to dentoalveolar growth and the downward migration of the hard palate by means of an apposition-resorption mechanism (Enlow and Bang, 1965). Obviously the palate migrates in the cleft lip and palate condition, in spite of the gross disturbance in structure.

Finally, and most significantly, the dentoalveolar structures accommodate to the basal jaw relations almost exactly as they do in the normal child. The maxillary teeth are positioned in an essentially normal relation to the mandibular teeth, with local disturbances in the area of the alveolar cleft. This observation indicates that, even when a gross morphologic disturbance involves the maxilla and its alveolar ridge, teeth will erupt until they contact opposing teeth, and they can be guided into a satisfactory functional relationship through the influence of the adjacent soft tissue (ie, tongue, cheek, and lip muscle). The discrepancy of bone in the cleft area is sufficient to prevent complete dentoalveolar adaptation.

Many of the patients reported in the literature had had cleft lip repair. The reports indicate that surgery had little influence on facial growth and minimal influence on dental occlusion (Davies, 1951; Innis, 1962; Derhert, 1963; Ortiz-Monasterio and coworkers, 1966; Bernstein, 1968; Pitanguy and Franco, 1968; Boo-Chai, 1971). A repaired lip will exert a force on the anterior teeth which apparently is not usually sufficient to disrupt the tongue-lip guidance of the incisor teeth, although incisor crossbite can occur.

The conclusions that can be drawn from the foregoing are clear: the midface is not extensively damaged by the embryonic disturbance concomitant with cleft formation. Once the cleft has developed, further growth proceeds in a reasonably normal manner, with some environmentally induced distortions remaining. The maxilla, which must advance freely during growth to permit apposition at the posterior aspect and sutural adjustment growth, advances with no detectable inhibition. The teeth and alveolar bone, which must be free to adapt to the jaw relations and the soft tissue environment, adapt without inhibition except in the area of the cleft.
Several recent studies (Dahl, 1970; Bishara and Iverson, 1974) which included unoperated cases would appear to contradict some of these findings. The prosthetic management of these patients and the variable completeness of the clefting, plus a slightly different interpretation of the data, could affect the conclusions. It appears to this writer that the weight of evidence favors the conclusions that the unoperated child with a cleft lip and palate is capable of developing a functionally normal facial skeleton except for the presence of the local bony defect.

Facial Growth Following Surgical Repair

Surveys of children and adults with repaired cleft lip and palate indicate that a normally functioning occlusion is rare unless orthodontic treatment has been successfully completed. The dental irregularities adjacent to the cleft are expected, but the collapsed maxilla with overlapping segments, the generalized midface retrusion, and other features found in many cases are not observed in unoperated cases. One is forced to the conclusion that surgery has been responsible, in some way, for the malocclusions associated with the cleft lip and palate population. Occasional incidents such as those illustrated reinforce this conclusion.

There seems to be some difficulty in determining precisely what has happened as a result of surgery and why it had happened, since contradictory studies appear in the literature. The overwhelming mass of data, however, supports the conclusions enumerated in this chapter. A major cause of the confusion concerning this subject is the disproportionate number of published studies which are inadequate in design, sample, and interpretation of results. Conclusions are frequently presented which are unwarranted from the data available, yet which are accepted because they come from "reliable sources". Careful evaluation will reveal the flaws in many frequently quoted studies.

Effect of Lip Reconstruction on the Maxillary Complex

The effects of surgical reconstruction of the lip are generally beneficial. Continuity of the orbicularis oris and the buccinator muscles is established, providing a more normal functional matrix to influence maxillary growth. The influence of lip repair on the growth of the maxillary complex must be considered in terms of its effect on the two somewhat independent components of the maxilla. Reconstruction of the nasal floor and upper portion of the lip will initiate pressure on the maxillary basal bone and nasal septum. Reconstruction of the lower portion of the lip establishes pressure on the dentoalveolar structures.

In the observations made on unrepaired cleft lip and palate patients, it was noted that, even when the lip was repaired, midface growth and occlusion were essentially normal. This can be explained by the fact that the lip repair is undertaken over a maxilla which has widely separated segments. Thus the tension produced by repair serves to bring the segments together, and maxillary width becomes normal (Aduss and Pruzansky, 1968; Harding and Mazaheri, 1972). By the time the segments contact, the lip tension has stabilized and is usually close to normal. A modern, loose lip repair does not involve any growing area of the maxilla, nor does it interfere with the dentoalveolar adjustment mechanism. Thus, midface growth and adjustment proceed more or less normally. There is no evidence that the age at which surgery is done has any effect on the results.
There are occasions when lip repair can cause interference with maxillary development. An increased pressure on the basal maxilla, because of either tissue deficiency or the type of repair used, may inhibit forward drift of the maxilla, producing a slow, progressive retrusion of the midface. A tight lower lip will mold the dentoalveolar structures posteriorly to an excessive degree. If there is a severe bony deficiency in the cleft area, the segments may be brought together by lip pressure, resulting in a small maxilla.

**Lip Reconstruction in Unilateral Cleft Lip and Palate.** In complete unilateral cleft lip and palate, the major (noncleft) segment following surgery is subjected to lip muscle pull toward the midline of the face, reversing the previous pull away from the midline. The result is a rapid improvement in its position. The outward rotation of the premaxillary area is molded back, and a slight medial movement of the smaller segment occurs. These three factors result in reasonably normal maxillary arch shape, and apposition of the alveolar margins occurs in a large percentage of cases. The maxillary dental arch is usually well related to the mandibular dental arch as the teeth erupt.

In spite of the rotation of the noncleft segment, the midline of the maxilla (anterior nasal spine) invariably remains deviated from the midline of the face (Harvold, 1954b). The nasal septum, which was bowed toward the noncleft side, is brought back following surgery and assumes the shape which is so characteristic of unilateral cleft lip and palate.

The repair of the floor of the nose narrows the base of the cleft nostril to match the non-cleft nostril, and the columella returns to the midline of the face. Distortion of the nasal cartilages, however, persists to some extent. Subsequent improvement in alar form does not usually occur, and asymmetries persist throughout life unless reconstructive surgery is undertaken.

**Lip Reconstruction in Bilateral Cleft Lip and Palate.** The morphologic features of a bilateral cleft lip and palate are quite different from those of the unilateral, so that there are entirely different considerations in the sequelae of lip reconstruction. The basal component of the premaxilla is reduced in this condition, since the contributions from the maxillary processes are absent. Like all basal bone, it is resistant to environmental influences. Thus the position of the anterior nasal spine is only slightly forward at birth, and it is only slowly inhibited or displaced posteriorly by upper lip pressure. In most cases the basal bone of the premaxilla will respond by becoming progressively less protrusive than at birth (Harvold, 1954a; Holdsworth, 1963; Birch and coworkers, 1967; Friede and Pruzansky, 1972).

The dentoalveolar component attached to the basal bone is extremely sensitive to environmental forces. Lip repair which restores a soft tissue-muscle balance to this area will contain the protrusive alveolus and mold it back without directly affecting the basal bone. Where there is severe protrusion (anterior rotation) of the premaxilla, together with a deficiency of prolabium and lateral lip tissue, achievement of an ideal lip repair becomes a difficult problem. It is in such cases that presurgical orthopedics becomes such a valuable procedure in rotating the premaxilla posteriorly.

If the lower portion of the lip exerts excessive pressure, a dentoalveolar retraction will occur which may have almost no effect on the anterior nasal spine position. Lip repair which brings lateral lip tissue together under the prolabium may result in a tight lip. The use of the
prolabium for the entire midsection of the lip usually stimulates its growth, and an adequate lip results (Cronin, 1957; Bauer and coworkers, 1959; Skoog, 1965), although it may be short (Handelmann and Pruzansky, 1968). If the lip is so short that it is incapable of molding the alveolar process back, severe protrusion of the alveolus remains, and the teeth erupt without anterior support. In this situation the lower lip falls behind the maxillary teeth, encouraging further protrusion of the teeth. Since teeth erupt until they encounter resistance from the lower teeth or the lip and since this resistance is almost completely lacking, a great deal of excessive eruption of the teeth may occur in these cases.

Dentoalveolar distortions due to either inadequate or excessive lip pressures are extremely difficult to correct at a later age. It is incumbent upon the surgeon to provide the young child with a lip that will reverse the prenatal distorting influences. At the same time, the reconstructed lip should encourage a growth pattern which will result in the establishment of normal skeletal relationships at maturity and will guide the upper teeth into a satisfactory relationship with the lower teeth.

**Effect of Palate Reconstruction on the Maxillary Complex**

In the reconstruction of a cleft palate, the surgeon must obtain tissue to bridge the cleft. Many techniques are employed, but the most frequently adopted ones involve raising mucoperiosteal flaps from the palate and relocating them medially and posteriorly, leaving a denuded area of bone close to the alveolar process. The filling in of this denuded area produces scar tissue which exerts an initial contracting force on adjacent tissues (Jolleys, 1954; Lynch and Peil, 1966; Kremenak and coworkers, 1967). Following the early stages of contraction, however, there is only a mild reduction in arch width (Dixon, 1966; Aduss and Pruzansky, 1968; Kremenak and coworkers, 1970; Harding and Mazaheri, 1972). If maxillary growth could proceed normally from this point on (as it does in unoperated clefts), the dentoalveolar structures could easily cope with the mild discrepancy, and a normal occlusion would result.

The age at palate repair appears to influence maxillary growth directly: more inhibition results from early than from later surgery (Dorrance and Bransfield, 1947; Graber, 1949, 1950; Lewin and Ship, 1962; Herfert, 1963; Rosenthal, 1964; Bernstein, 1968; Longacre, 1970). There are so many other variables that it is not possible to evaluate how much interference occurs or to recommend an optimum age for surgery.

Another variable is the type of operative procedure used. Available evidence suggests that the least traumatic procedures encourage more favorable growth (Waldon, 1950; Rosenthal, 1951; Jolleys, 1954; Slaughter and coworkers, 1956; Bergland, 1963; Palmer and coworkers, 1969; Bariana and coworkers, 1972; Harding and Mazaheri, 1972). To encourage "mild" surgery, many authors have suggested that the soft palate be repaired early and the hard palate deferred for a later repair (Gillies and Fry, 1921; Coursin, 1950; Graber, 1950; McNeill, 1950; Rosenthal, 1951, 1964; Slaughter and Pruzansky, 1954; Trusler and coworkers, 1955; Webster and coworkers, 1958; Lewin and Ship, 1962; Lewin, 1964; Penkava, 1967). Multiple procedures to effect palate closure cause severe maxillary inhibition (Graber, 1949; Slaughter and Brodie, 1949; Kazanjian, 1950; Ponterio, 1952; Bill and coworkers, 1956; Chapman and Birch, 1965).
Frequently one sees repaired palates with irregular contours and a lowered vault. This appears to be the result of movement of the flaps after surgery because of poor fixation, hemorrhage and hematoma formation, or contraction during healing, which tends to pull the flaps away from the vault. It is likely that calluslike bone forms under the flaps in these areas (Hugg and Kremenak, 1968). The significance of this is that the lowered, irregular vault impinges on the space available for the tongue and interferes with tongue posture and function. There is the suspicion that analysis of maxillary morphology using the palatal plane may give a very misleading assessment of intraoral space (Ross and Johnston, 1972). This will be discussed more completely later in the chapter.

Before proceeding to a discussion of the effects of palate repair on the maxillary complex, it might be well to review the facial growth that occurs in the typical clinic population of children with cleft lip and palate following surgery.

Facial Growth Tendencies

The facial form of the older child or adult with a repaired cleft lip and palate could be characterized as being flat and long. While there is enormous variation related to the individual's racial and familial genetic background, the type of cleft, and the surgical and orthodontic management he has received, these two features are most striking in any examination of a group of affected individuals. Data from cephalometric studies invariably confirm this observation.

Maxillary Growth. Since the growth patterns are different with each cleft type, these will be considered separately.

Cleft Lip. Children with a cleft of only the lip and anterior maxilla have a minimal disturbance in facial form. There are dental irregularities in the region of the cleft, of course, and the anterior nasal spine shows some positional differences (Rosenthal, 1951; Harvold, 1954a; Levin, 1960; Coupe, 1962; Graber, 1964; Ross and Coupe, 1965; Ridley, 1966; Dahl, 1970; Zilberman, 1973). In all other regards, individuals with this type of cleft appear to have facial features more like normal persons than like the other cleft types. The exception to this is bilateral cleft lip and alveolus, particularly when the premaxilla is isolated from the remainder of the maxilla. In this type of cleft, many features of the bilateral cleft lip and palate are present, but to a lesser degree.

Cleft Palate (Isolated). Facial form in individuals with isolated clefts of the hard and soft palate is characterized by a deficiency in maxillary length (Osborne, 1966; Shibisaki and Ross, 1969; Dahl, 1970) and a retracted position of the anterior region of the maxilla relative to the cranial base (Graber, 1954; Jolleys, 1954; Levin, 1960; Foster, 1962; Hagerty and Hill, 1963; Ross and Coupe, 1965; Osborne, 1966; Shibisaki and Ross, 1969; Dahl, 1970; Bishara and Iversen, 1974), which becomes accentuated as growth continues to maturity (Osborne, 1966; Shibisaki and Ross, 1969). There have been some reports of normal maxillary length (Ponterio, 1952; Levin, 1960; Ross and Coupe, 1965). There may be posterior displacement of the entire maxillary complex, including the inferior orbital margin and the zygomatic process (Levin, 1960; Shibisaki and Ross, 1969; Dahl, 1970), although other studies report that the maxillary tuberosity is in a relatively normal anteroposterior position (Ross and

**Complete Unilateral Cleft Lip and Palate.** The majority of studies have noted that the anterior maxilla is retruded relative to the cranial base and becomes more retruded during growth. This is partly due to a decreased length of the maxilla and partly to a repositioning of the maxilla. Anterior vertical development is normal or slightly less, but posterior development is deficient. Studies in the literature include Ponterio (1952, Graber (1954), Jolley (1954), Brader (1957), Levin (1960), Foster (1962), Hagerty and Hill (1963), McNeill (1964), Ross and Coupe (1965), Osborne (1966), Chapman (1967), Ross and Johnston (1967), Dahl (1970), and Sadowsky and coworkers (1973).

**Bilateral Cleft Lip and Palate.** The maxillary complex as a whole in bilateral cleft lip and palate is retruded as in the other cleft types. The presence of a separate premaxillary area, however, results in a different growth pattern. As mentioned previously, the mild protrusion of the basal portion of the premaxilla and the severe protrusion of the dentoalveolar portion present at birth are reduced, rapidly at first following lip reconstruction, then more slowly as facial growth proceeds. Studies show that the protrusion of the premaxilla relative to the cranial base at ages 6, 8, 12, and 16 is greater than the normal value by 8, 4.4, 2, and 1.5 degrees, respectively (Swanson and coworkers, 1956; Ross and Johnston, 1967; Narula and Ross, 1970). In the adult the premaxillary position is normal (Birch and coworkers, 1967). Other studies (Ponterio, 1952; Swanson and coworkers, 1956; Levin, 1960; Friede and Pruzansky, 1972) give slightly different values, but the pattern is similar, modified by the type of surgery as well by individual variations. In many cases the growth inhibition is excessive, and at the completion of facial growth the premaxilla is retruded relative to the remainder of the face. In almost no instance does the early protrusion persist into adulthood.

Some clinicians feel that initial surgical repositioning of the basal premaxilla to a more retruded position is desirable in many cases. Surgical intervention at an early age seems unnecessary, since natural growth will accomplish the same result in time. The experience of most surgeons is that these procedures may interfere with growth (Peer, 1947; Slaughter and Brodie, 1949; Kazanjian, 1950; Pruzansky, 1954; Bauer and coworkers, 1959; Glover and Newcomb, 1961; Holdsworth, 1963; Fara and Hrivmakova, 1965; Birch and coworkers, 1967; Cronin and Penoff, 1971; Bishara and Olin, 1972), so that an apparently successful result in a young child may become a severe deformity by the time he has reached adulthood. Modern management of difficult cases by presurgical orthopedics offers a less dangerous alternative, since the initial reduction of premaxillary protrusion can be accomplished quickly and easily, with no long-term effect on sensitive growth areas.

Narula and Ross (1970) have shown that the posterior segments of the maxilla although retruded, are of normal size, so that the overall length of the maxilla is much greater than normal at age 6, but normal by 12 and 16 years (Ponterio, 1952; Levin, 1960; Narula and Ross, 1970).

**Growth of the Mandible.** The size of the basal bone of the mandible is determined by genetic factors. It appears that only a severe environmental influence can alter this genetic blueprint, but evidence of the subtle interplay of genetic and environmental forces is inadequate at present. The finding in studies on cleft lip and palate that the mandible is
essentially normal in length (Hagerty and Hill, 1963; Ross and Coupe, 1965; Chapman, 1967; Ross and Johnston, 1967; Dahl, 1970; Narula and Ross, 1970; Nakamura and coworkers, 1972) leads to the conclusion that the mandible is not directly involved in the cleft lip and palate anomaly.

Cephalometric studies constantly indicate, however, that there is a change in the position of the mandible. The chin is usually retracted and is lower; that is, there is greater facial height in spite of normal or decreased maxillary height. The change in vertical height is mild in younger children but especially noticeable in older children. The condyle is positioned slightly upward and forward. Some of the studies from which these conclusions were drawn are those of Ponterio (1952), Graber (1954), Jolleys (1954), Harvold (1960), Levin (1960), Coccaro and Pruzansky (1965), Ross and Coupe (1965), Chapman (1967), Ross and Johnston (1967), Shibisaki and Ross (1969), Dahl (1970), Narula and Ross (1970), Farkas and Lindsay (1971, 1972a), and Sadowsky and coworkers (1973).

The explanation for these differences is simple: the mandible is more open than in the normal individual. As the mandible opens, the chin point moves downward and backward and the condyle comes forward, since the center of rotation is close to the condyle. The mandibular plane becomes steeper. The gonial area should also move back and down, but generally the position of gonion is normal or higher. The reason for this is that the gonial area is not part of the basal bone but exists as a muscle attachment. Altering the position of the mandible will not permanently alter muscle length; instead, the bony attachment remodels to permit the muscles to maintain their correct length for optimum function. Thus the gonial angle (the angle formed by the "body" and the "ramus") is found to be more obtuse (open) in groups of individuals with cleft lip and palate (Harvold, 1960; Ross and Johnston, 1967; Shibisaki and Ross, 1969; Dahl, 1970; Narula and Ross, 1970) and increases with age (Narula and Ross, 1970), in contrast to the normal decrease with age (Munrose, 1966).

The observation that gonion is even higher in cleft lip and palate patients than in patients without this congenital anomaly (Ho Kim, 1958; Harvold, 1960; Levin, 1960; Chapman, 1967; Ross and Johnston, 1967; Shibisaki and Ross, 1969; Dahl, 1970; Narula and Ross, 1970) may be explained by the fact that most cephalometric assessments are made with the teeth in occlusion rather than with the mandible in rest position. In the latter state there is a greater freeway space in individuals with cleft lip and palate, which would decrease the differences in the gonion position. There may also be some intrinsic shortening of the muscles, an altered insertion on the pterygoid plates, or an interference with their growth as a result of palatal surgery (Ross and Johnston, 1972).

The Pierre Robin anomalad is a form of cleft palate in which the mandible is either micrognathic (small) or retrognathic (posteriorly positioned). The normal-sized tongue is positioned high in the cleft and back into the pharynx, resulting in glossoptosis and interference with breathing and swallowing. Acute periods of respiratory distress cause concern for the life of the infant. Randall and Hamilton (1971) have published an excellent review of this condition (see Chapter 50).

There appear to be at least two forms of this condition with regard to the mandible. The first is true microcephalia, in which the mandible is extremely small and remains small throughout life. There is a second situation, however, in which the mandible appears to be
equally small but grows rapidly after birth and soon is within the normal size range. It is often impossible to identify an older child as having had the Pierre Robin anomad.

It is difficult to explain the second type of mandible. A likely possibility is that the potentially normal-sized mandible in the embryo is compressed between the head and the pericardial region and is either deformed in shape or prevent from normal embryonic development. This would force the tongue to remain between the palatal shelves and prevent normal palate formation. The posterior position of the chin makes it impossible for the genioglossus to advance the tongue out of the airway after birth, and glossoptosis occurs (Randall and Hamilton, 1971). Another possibility is that the tongue for some reason is positioned (not forced by the mandible) in the nasopharynx during embryonic and fetal life. The potentially normal mandible is thus deprived of a necessary growth stimulus during this period. After birth, when the tongue is brought down into the oral cavity, the mandible is able to express its growth potential and "catches up". A third possibility is that there is a retrognathia (the posterior displacement of a normal mandible), perhaps due to immature or abnormal musculature (Bosma, 1963; Ross and Johnston, 1972), which prevents the normal forward adjustment of the mandible at birth.

In any event, there is a strong likelihood that the cleft palate associated with the Pierre Robin anomad is secondary to the mandible and tongue relationship and is caused by a mechanical blockage of palatal shelf elevation and fusion. The incompleteness of the cleft in most cases and the disproportionately high incidence support this concept of an etiology different from those for other cleft palates (Randall and Hamilton, 1971). A mechanical explanation has also been advanced for the high incidence of cleft palate in Klipper-Feil syndrome (Ross and Lindsay, 1965).

Jaw and Tooth Relations. The retrusion of the maxillary basal bone in cleft palate and unilateral cleft lip and palate is somewhat balanced by the rotation of the mandible and consequent retrusion of the chin. Anteroposterior jaw relations are found to be either satisfactory (Ponterio, 1952; Bill and coworkers, 1956; Swanson and coworkers, 1956; Shibisaki and Ross, 1969) or more frequently associated with a slightly excessive maxillary retrusion (Levin, 1960; Hagerty and Hill, 1963; Osborne, 1966; Ridley, 1966; Iyer, 1967; Ross and Johnston, 1967; Chapman, 1967; Bishara and Iversen, 1974), particularly in older children and in patients who had more traumatic surgical procedures (Grabert, 1949; Slaughter and Brodie, 1949; Graberb, 1950; Kazanjian, 1950; Ponterio, 1952; Jolleys, 1954; Bill and coworkers, 1956; Chapman and Birch, 1965).

The occlusal relationship is quite satisfactory in the primary dentition, although there is a high incidence of incisor crossbite, but there is a deterioration in the permanent dentition, with incisor crossbite and a flattening back of the incisors almost invariably in both arches (Ponterio, 1952; Graber, 1954; Swanson and coworkers, 1956; Harvold, 1960; Foster, 1962; Hagerty and Hill, 1963; Derichsweiler, 1964; Graber, 1964; Shibisaki and Ross, 1969; Narula and Ross, 1970; Pickrell and coworkers, 1972). In bilateral conditions the maxillary incisors are frequently inclined palatally, so that incisor crossbite may occur even when the premaxilla is protractive.

Maxillary width varies, but in general there is a narrowing of the dental arch following surgery, and posterior crossbites are frequent (Swoisken, 1957; Herfer, 1958; Forter, 1962;

Increased vertical face height due to the more open mandible is compensated for by extra eruption of the teeth, particularly the maxillary posterior teeth and the mandibular anterior teeth.

**Causes of Abnormal Facial Growth**

Discussion of the actual causes of abnormal facial growth remains in the realm of speculation. The author has presented in detail elsewhere (Ross and Johnston, 1972) one hypothesis which appears to fit the available data. Studies can be found which appear to contradict parts of the hypothesis, but until further evidence is available, it stands as a plausible explanation of the abnormal growth and development of the face. A resumé of this hypothesis follows.

Failure of adequate growth of the maxilla in length must be considered partly a basal bone problem, partly a dentoalveolar problem. The basal bone probably has a mild intrinsic defect caused by a general deficiency of mesenchyme in the embryonic maxillary region. The maxillary deficiency observed in any population of operated older children or adults, however, is more than could be expected and more than that observed in individuals with unoperated cleft lip and palate. The evidence is almost overwhelming that surgical repair of the lip and palate has induced a maxillary basal underdevelopment.

There are several ways in which this could occur: the possible inhibiting effect of reconstruction of the nasal floor and upper lip has been discussed earlier. Growth in length of the maxilla requires that the entire maxilla move forward freely so that bone can be deposited on the tuberosity. If during palate repair there is undermining of tissues and hamulus fracture in the area of the pterygoid plate - palatine process - tuberosity junction, scar tissue may form across this sensitive growth area and inhibit the forward movement of the maxilla. It is not necessary that the inhibition of growth be severe, because even a fraction of a millimeter of inhibition each year would create a severe problem by the time of facial maturity. Evidence cited indicates that maxillary underdevelopment is progressive.

Vertical growth of the posterior maxilla may be inadequate owing to an intrinsic deficiency or surgical interference. Comparisons based on the palatal plane give the appearance of posterior underdevelopment, but this may indicate merely that the palatal plane is altered by shelf angulation or inhibition, while the basal maxilla is less affected.

The width of the basal maxilla is probably not appreciably reduced in cleft lip and palate patients (Coupe and Subtelny, 1960; Dahl, 1970; Nakamura and coworkers, 1072). The apparent narrowing is mainly in dental arch width and may also be related to the decreased length of the maxilla, which places a narrower portion of the dental arch in contact with a wider portion of the mandibular dental arch. There are, of course, some patients with a true deficiency in maxillary width.

The dentoalveolar component of the maxilla is more certainly affected by surgery and is probably an even more important factor in the abnormal development of the face, at least
from the orthodontist’s viewpoint. Surgical procedures on the palate result in scar tissue adjacent to the alveolus. The initial scar tissue contraction during healing causes a movement of the maxillary segments until, in most cases, contact between the segments is achieved. There is probably some constriction of the dental arch as well, since the suspensory mechanism of the teeth, the periodontal ligament, sends fibers into the surrounding mucosa which are caught up in the scar tissue. More important, the scarred palatal mucosa resists further growth to some extent, and tension on the periodontal fibers during subsequent tooth eruption causes a posterior and medial deflection of the teeth by the continuum of scar tissue from the pterygoid plates to the incisor region. Thus the satisfactory relationship of the primary teeth progressively worsens into the full permanent dentition.

It has been reported (Rosenthal, 1964; Neumann, 1967; Hotz, 1973) that, if only the soft palate is repaired at the initial phase of palate surgery, the palatal shelves will grow and the cleft will narrow almost to obliteration. The second phase of repair thus becomes a simple midline closure at a later date. The advantage of this method (the Schweckendiek procedure) is the avoidance of surgery in the periphery of the palate. Since there is no scar tissue near the teeth, there is no interference with the dentoalveolar adjustment, and deformation of the dental arch should not occur.

Several other factors contribute to the dentoalveolar constriction and posterior deflection. The lower portion of the upper lip exerts a moderate pressure which can be excessive when added to the other factors. These are primary factors which inhibit the free expression of the ability of the teeth and alveolar bone to compensate for minor skeletal disharmonies.

There are many secondary factors acting to alter facial growth. Normal tongue posture in the palatal vault is particularly important in resisting abnormal growth tendencies. The frequent occurrence of mouth breathing in children with cleft lip and palate is related to the deviated nasal septum and the high incidence of upper respiratory infections which interfere with breathing and cause enlarged tonsils and adenoids.

Mouth breathing causes a lowered tongue posture, out of the palatal vault. This is compounded by the decreased size of the palatal vault (constricted dental arch and postsurgical vault shape), which inhibits proper tongue placement. Enlarged tonsils mechanically induce a forward tongue position. The altered tongue posture with or without mouth breathing does two things. First, it removes some of the essential tongue support of the maxillary arch, encouraging further constriction (Linder-Aronson, 1970; Paul and Nanda, 1973). Second, it forces the mandible to open and accounts for the altered position of the mandible discussed previously (Linder-Aronson, 1970; Dunn and associates, 1973; Paul and Nanda, 1973). If the mandible is held excessively open at rest, the mandibular incisors do not support the maxillary teeth adequately, and they are less able to resist the constricting tendency.

As mentioned earlier, vertical development of the dentoalveolar process is not inhibited in the early years. Even those cases with an excess freeway space are characterized by normal molar eruption; the excess freeway is caused by an excessive amount of mandibular opening. A vertical problem does occur frequently in the incisor region during the final stages of mandibular growth. Growth normally ceases in the maxilla before it does in the mandible, but
compensatory dentoalveolar development maintains the occlusion. In cleft lip and palate the maxillary incisors frequently are unable to make this compensation, and an open bite results.

**Effect of Preoperative Orthopedics on Facial Growth**

Despite the enormous volume of literature in the past 20 years on the subject of infant orthopedics, there is no evidence available that these procedures influence long-term facial growth. It is the author's opinion that the changes induced are minor and have no effect on the many factors discussed in this chapter which influence the eventual facial form in the adult.

**Effect of Bone Grafting on Facial Growth**

There are grounds for theorizing that bone grafting in the anterior region of the maxilla in infancy should not affect facial growth. The clinical evidence to date is inconclusive and contradictory but appears to indicate that maxillary growth is somewhat inhibited by these procedures (Derichsweiler, 1964; Kling, 1964; Thilander and Stenström, 1967; Lynch and associates, 1970; Hogemen and coworkers, 1972; Jolleys and Robertson, 1972; Pickrell and coworkers, 1972; Robertson and Fish, 1972) (see also Chapter 48).

**Effect of Orthodontics on Facial Growth**

Orthodontic treatment has an effect on the dentoalveolar structures but appears to have little effect on the basal bone of either the maxilla or the mandible (Ross and Johnston, 1967). The potential exists for altering jaw relations much more than we presently do by standard clinical methods.

**Discussion**

The surgeon who undertakes the habilitation of an infant with cleft lip and palate has three major aims: to reconstruct a functional speech mechanism, to reconstruct the features of the lip and nose for maximum esthetics, and to encourage adequate growth of the facial skeleton. While the last is perhaps the least important of the three, providing for facial growth and preventing dental malocclusion in these children are real problems.

The available evidence has been examined and a logical explanation developed for the abnormal growth and development of the facial skeleton and dentition. To recapitulate the major points covered:

1. The intrinsic defect in an individual with cleft lip and palate is mild, mostly limited to the immediate area of the cleft.

2. The potential for growth of the maxillary complex is adequate to produce harmonious skeletal relationships.

3. The teeth and alveolar bone will develop to overcome minor deficiencies in the maxillary complex and produce an excellent occlusion.
4. Surgery produces scar tissue in many areas which may interfere with maxillary forward growth. It is not necessary that this be a severe restriction; any reduction in maxillary growth can be significant for children with cleft lip and palate.

5. Surgery produces scar tissue in the palate which prevents the free adjustment of the teeth and causes distortion of the dental arch by deflecting the eruption of the teeth.

6. Secondary changes in tongue position cause displacement and deformation of the mandible.

7. The surgeon should examine current procedures and devise new ones which do the following:

   (a) Protect the growth sites, specifically the maxillary tuberosity-palatine-pterygoid junction, by avoiding this areas as much as possible, by operating at as late an age as possible, and by avoiding operations subsequent to initial one.

   (b) Protect the dentoalveolar adjustment by avoiding surgery in the area adjacent to the alveolus.

   (c) Establish normal tongue posture by achieving a high, smooth palatal vault, eliminating arch constriction, and maintaining nasal breathing. The latter requires constant monitoring of respiratory infections, correction of the obstructing nasal septum, and removal of tonsils and adenoids if indicated (and if speech will not be affected by their removal).

   (d) Overcome severe secondary deficiencies by the use of maxillary and mandibular osteotomies.