Early Orthopaedic Treatment of Clefts of the Primary and Secondary Palates: A Surgeon's View

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An assessment of the value of any procedure or method of treatment is dependent upon a proper understanding of the underlying pathology, the natural history of the untreated condition, and an evaluation of what may be attributed to the procedure or treatment under examination. It would therefore seem essential to review briefly the development of clefts of the primary and secondary palates, the effects of these clefts upon subsequent growth and development of the middle third of the face, and the further effects of surgery in the absence of preliminary orthopaedic treatment upon these deformities. Only with such a background can one evaluate the early orthopaedic treatment of the clefts.

Formation of Clefts

The classical concept of the face developing as a result of fusion of various processes lying free in space (9) has, of course, been discredited. It is now well recognized that what occurs is the migration of mesodermal masses between two continuous sheets of ectoderm covering the face and roof of the oral cavity (23, 21) and that unless the ectoderm is supported and nourished by an intervening layer of mesoderm it will eventually break down and give rise to a cleft (20).

The maxillary processes appear during the fourth week as extensions or branches of the mesoderm of the mandibular or first arch, pass around the angles of the primitive mouth, and then across that region which lies below the olfactory placode to produce a solid bar of tissue which is the primitive or primary palate. This primary palate is formed by five weeks and from it develop the central lip and premaxillary complex as far back as the incisive foramen. Consequently, should there be a failure on the part of the mesodermal maxillary processes to penetrate between the layers of ectoderm, this ectoderm will break down and produce a cleft of the primary palate. Such a cleft will be present in the embryo by the fifth week.

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This paper was presented at the 1965 meeting of the Florida Cleft Palate Association in Tallahassee, Florida, and was supported in part by PHS Research Grant DE-01836, National Institute of Dental Research.

The severity of clefts of the primary palate will vary with the degree of failure of mesodermal penetration, and in the most severe cases will extend through the lip and alveolus and into the nostril floor (15).

It is only two to three weeks later that the secondary palate is formed by fusion of two maxillary palatal processes with each other and with the inferior border of the nasal septum. These palatal folds emerge at about eight weeks when the primary palate is already formed. Lying at first vertically at each side of the tongue, they rise above it at nine weeks consequent on the descent of the tongue, and then between 10 and 12 weeks fuse with each other and with the septum from before backwards to form most of the hard and all the soft palate. It should be emphasized that this is a true process of fusion as compared with the normal development of the primary palate, which is not.

This clear distinction between the development of the primary and secondary palates at different times and by different processes, which has long been recognized by embryologists, forms the basis of Kernahan and Stark's classification of clefts (10), thereby combining simplicity with logic.

The consideration of the aetiological factors giving rise to clefts is beyond the scope of this paper. Nevertheless, while it is conceivable that a cleft of the primary palate may in some circumstances promote a cleft of the secondary palate, the reverse does not hold true.

Burston has propounded what amounts to a mechanical theory for the cleft of the secondary palate seen in the Pierre Robin syndrome (2), though clearly no mechanical theory can apply to clefts of the primary palate.

Secondary Effects of the Clefts

By the end of the twelfth week of gestation, the cleft will be fully developed. It is during the remaining period of growth and development, which extends until adolescence, that the cleft exerts a secondary effect upon the whole of the middle third of the face and leads to the classical deformities.

To understand these secondary effects of the cleft, we must further examine the development of the middle third of the face, in particular the part played by the interorbital cartilaginous nasal septum. It is the growth of this relatively enormous structure which is responsible for the forward and downward growth of the maxillae (18, 19). Because the maxillae are attached to the nasal septum, they are therefore carried with it, the potential spaces at the surrounding sutures being 'filled in' by bone. This concept of sutural growth has recently been reinforced by the work of Latham and Burston (11) and the unequivocal experiments carried out by Sarnat (17). Subperiosteal appositional growth of the maxillae plays a minor role, certainly until the age of six or seven years, by which time ossification of the septal cartilage has deprived it of most of its growth potential.

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In a complete unilateral cleft of the primary and secondary palates, the maxilla on the cleft side is separated from the nasal septum. The lesser segment, being thus deprived of the growth impulse, lags in development and is small and retro-positioned. The premaxilla on the greater segment tends to grow forward and to rotate to the uncleft side taking the nose tip with it, bending the septum, and flattening the ala on the cleft side.

In the complete bilateral cleft, the unrestrained growth of the septum carries the premaxilla forward, leaving the lateral segments behind. In both uni- and bilateral clefts there is some slight lateral collapse of the maxillary segments which appears to increase even in the absence of surgery. This increasing collapse is shown by Burston (5) to be more apparent than real, because of the changing mandibulo-maxillary relationships.

Because septal growth is maximal during the last six weeks of gestation, premature babies generally show less marked deformities than do full time babies. After a short neonatal pause, there is another growth spurt for about six months which accounts for the increasing deformity which may be seen in untreated babies during this period.

Effects of Surgery

Having considered the development of the cleft and the effect of the cleft on the subsequent growth and development of the face, it is important to examine the part played by surgery in the absence of preliminary orthopaedic treatment, in correcting, potentiating, or increasing the deformities. While repair of the cleft lip by establishing more correct muscle balance may correct some of the premaxillary deformity in both uni- and bilateral clefts, repair of the lip will perpetuate and possibly increase the malrelationship of the lateral segments and, therefore, the malocclusion. Furthermore, if repair of the anterior palate is combined with lip repair, the contraction of the band of scar tissue which must form across the site of the anterior palatal repair is felt to be a major factor in this lateral collapse. The repair of bilateral clefts may result in the premaxilla becoming locked in position anterior to collapsed lateral segments, and it was this particular difficulty which led to the now widely abhorred operations of premaxillary excision or surgical retropositioning. It must, however, be conceded that these procedures were occasionally necessary in gross cases if closure of the lip was to be obtained at all.

In the past, surgery was blamed for maxillary retrognathism and hypoplasia in patients with repaired clefts. This theory has been examined by Foster (8), Subtelny (22), and most recently by Coccaro and Pruzansky (6), all of whom conclude that surgery, at any rate as currently practiced, does not interfere with growth of the middle third of the face.

Early Orthopaedic Treatment

It was against this background that early orthopaedic treatment of clefts was undertaken by McNeil (12) and fully exploited by Burston (3) on the basis of his considerable embryological knowledge (4). The treatment, which should be started within a few days after birth, depends on plates so constructed that the chewing and sucking movements of the baby are converted to appropriate forces acting on the maxillary segments.

A close liaison has been established in Liverpool whereby Dr. Burston and his team are notified within a matter of hours of the birth of a baby with a cleft of the lip or palate in any maternity unit of the region. In this way, they really are able to commence treatment at the earliest possible time.

From an impression of the maxillary arch, an uncorrected feeding plate is constructed and used for about two weeks to accustom the child to wearing a plate and to assist with feeding.

Once the baby accepts this plate, and preferably before the end of the neonatal growth pause, a second, corrected plate is inserted. This corrected plate is constructed by making a model of a new impression, dividing and adjusting it to a corrected position; for example, in a unilateral cleft this entails moving the lateral segment slightly outwards and forwards, and then from this corrected model, making another plate.

Several important features of these plates should be stressed. They are all made of clear acrylic so that their pressure effects upon the palatal mucosa may be readily observed. They all have wings on them to facilitate insertion and to prevent the baby from swallowing them. A tape, which is often passed around the head and attached to the wings, is not to keep the plate in place, but only to prevent its extrusion by occasional excessive tongue movements. The pressure bearing points of the plates are kept off the main blood vessels to the palate and off the edges of the palatal shelves lest this hinder growth.

The real skill in constructing these appliances, and the critical factor in their function, is the height of the bite blocks at the sides. These heights are determined from a wax plate constructed from the corrected model, and are so judged as to be just in excess of the free way space. If the mouth is gagged open too widely by the bite blocks, the baby will not tolerate the plate and will extrude it. If, on the other hand, the blocks do not take up the free way space, reflex chewing will not be stimulated, and since it is this which provides the motive power for the plate's action, it will not have the desired effect.

Once the plate fits accurately, which will be apparent from the uniform blanching of the mucosa, a new plate incorporating further correction is indicated. On an average, three to four plates are required between birth and lip repair.

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This treatment quite obviously places heavy demands on the time and skill of the orthodontist and the parents. In the majority of cases, treatment is carried out on an outpatient basis, but there are exceptions to the rule. Quite often, the mother and baby must be admitted to a special unit for a week or so to instruct the mother in the care and handling of the appliance and to give her confidence. In other cases, either because of the poor home conditions or other special circumstances such as transportation difficulties in reaching clinics, the baby is admitted to the special unit for part or all of the treatment.

Correction of the deformity of the premaxilla is assisted in some unilateral clefts by extraoral traction; that is, gentle pressure exerted on the prolabial region (Figure 1). This extraoral traction is of paramount importance in the treatment of bilateral clefts, since only in this way will the unrestrained growth of the nasal septum be prevented from carrying the premaxilla out on a stalk, though it must be conceded that in some cases, excessive restraint has resulted in a degree of septal buckling.

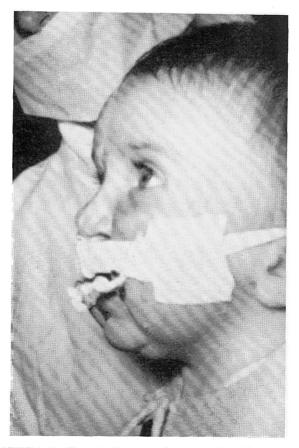


FIGURE 1. To illustrate the type of extra-oral traction employed.

Surgical Benefits of Orthopaedic Treatment

Having had the advantage of spending some three years in Liverpool, perhaps it is justifiable to make a surgical assessment of early othopaedic treatment as practiced by Dr. Burston and his colleagues. His experience with this treatment is now very extensive indeed, and he would be the first to admit that this is due in no small measure to the spirit of willing and friendly cooperation which has long existed between the orthodontists and Mr. Osborne and the other surgeons of the area (16). It should be stressed at this stage that no harmful or dangerous results of this treatment have emerged, and no contraindications to its employment.

Two by-products or side effects are perhaps worthy of mention. The first, which is of no practical consequence, is that the mucoperiosteum of the palate seems to hypertrophy and thicken. The second, which is of very material benefit is that, as noted by others too (7), the plate assists the baby in its feeding. This it does, both by its mere mechanical presence, and also by stimulating the sucking reflex and thereby improving the co-ordination and power of the suck and swallow mechanisms.

It was McNeil's original view that this treatment would stimulate growth of the palatal bony shelves and result in their union without either soft tissue surgery or bone grafting. Although this bony union does not seem to occur, in very many cases the width of the palatal cleft narrows appreciably during the period of presurgical treatment. It has been suggested (5) that this narrowing is the result of the appliances stimulating the blood supply and keeping the tongue out of the cleft. Whatever its cause, it is a real and obivous benefit to the surgeon.

That the dental occlusion is improved by this treatment is so obvious as not to merit mention, and in any case is not strictly within the surgeon's province.

A very definite surgical advantage of orthopaedic treatment is the marked narrowing of the lip cleft. Not only is the cleft reduced in its lateral dimension but, also, excessive rotation of the premaxilla is correctable by external traction (Figure 2).

While it may be argued that any surgeon worth his salt will close a lip no matter how wide the cleft, it is nevertheless also true that the easier the closure, the better will be the final result. This is particularly true of any method of closure which relies, as does the Millard technique (13), on a 'cut as you go' principle in which the lip is tailored by eye and craftmanship until it looks right. Quite obviously if the underlying alveolar segments are grossly displaced at the time of lip surgery, their movement consequent on the moulding effects of the repaired lip will alter the appearance of that lip and will to varying degrees detract from the final cosmetic result.

At the time of lip surgery, the alveolar segments are separated in many instances by a gap which represents an amount of missing tissue.

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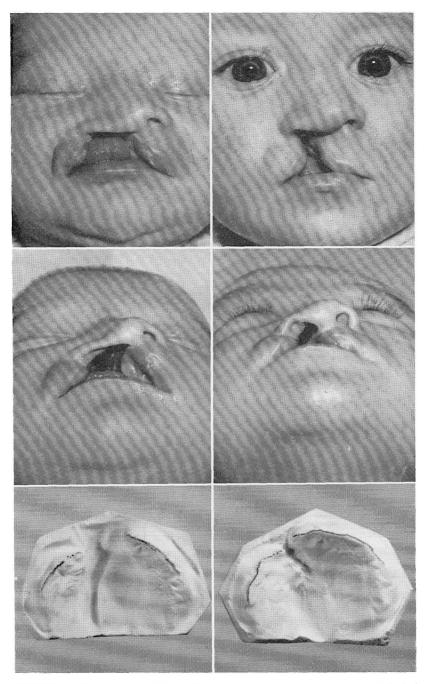


FIGURE 2. Upper left, middle left, and lower left show unilateral cleft before orthopaedic treatment. Upper right, middle right, and lower right show patient prior to lip surgery and after early orthopaedic treatment.

Lip repair, and especially a simultaneous closure of the anterior palate, will tend to close this gap though it may be maintained in some cases by continued orthopaedic treatment. Where this gap represents missing tissue, it may be that it is best filled by a bone graft, but experience in this field is still so limited that a firm opinion at this stage seems premature.

Thus far, little mention has been made of the nose, apart from the initial deformity resulting from the cleft. One of the great problems in the surgery of clefts has been the persistence of the nasal deformity even in the presence of a good result on the lip. A major element of this nasal deformity is of course the alar distortion, and it was felt that this could be corrected if the alar base was sitting upon a correctly positioned lateral segment. Unfortunately, our hopes in this respect are unfulfilled, for even with an almost perfect arch the ala in many cases is far from perfect.

I have recently had the very great fortune and privilege to examine a number of Dr. Millard's earlier cases who had not had early orthopaedic treatment, and in whom, while the arch alignment was somewhat less than pefect, the alar bases gave the appearance of almost perfect symmetry. Although the Millard technique will not overcome all the deficiencies of poor arch alignment in every case (14), nevertheless insofar as the appearance of the nose is concerned, soft tissue surgery would seem to be more important than orthopaedic treatment in determining the result. Quite obviously the ideal must lie in a combination of a sound, correctly aligned foundation on which to build a Millard repair. Only in this way can the best of both worlds be obtained, and this in fact is now



FIGURE 3. Left, severe unilateral cleft. Right, aged $2\frac{1}{2}$ years following repair by Dr. Millard without the use of orthopaedics.

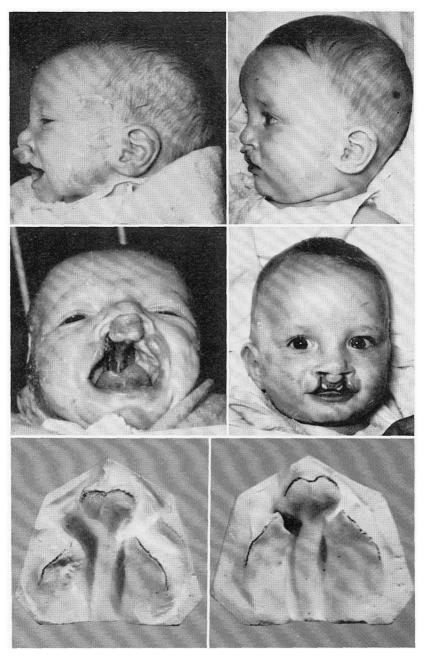


FIGURE 4. Upper left, middle left, and lower left illustrate bilateral cleft. Upper right, middle right, and lower right illustrate patient 14 weeks later prior to lip surgery, showing effects of early orthopaedic treatment. Note slight septal bucking in lower right photograph.

being attempted. Resulting in large measure from Burston's visit to Miami in 1964, Dr. Balber is applying early orthopaedic treatment to Dr. Millard's cases, while there is an increasing acceptance of the Millard repair in Liverpool (Figure 3).

There remains for consideration the problem of the protruding premaxilla in bilateral clefts. In the absence of early orthopaedic treatment the only way to achieve closure in the very worst cases is to excise the premaxilla or to employ one of the 'pushback' operations (1), both of which give results which are almost equally disastrous. In the less severe case, while lip closure may be obtained without interference with the premaxillary or septal areas, it results frequently in the premaxilla becoming locked forward anterior to collapsed lateral segments. This produces a tight upper lip and unsightly, protuberant philtral region. The assistance afforded the surgeon by early orthopaedic treatment is probably greatest in these cases. The unrestrained growth of the septal cartilage which carries the premaxilla out on a stalk is effectively controlled by external traction and some of the forward impetus of septal growth is transmitted via the plate to the lateral segments, which at the same time are prevented from collapsing. Consequently the baby comes to operation with a more correctly aligned arch which, following greatly facilitated surgery, keys into almost perfect position (Figure 4).

Conclusions

In considering all the above, what firm surgical indications for the early orthopaedic treatment of clefts of primary and secondary palates have emerged? First, there can be little doubt of the value of this treatment in complete bilateral clefts. The same applies to unilateral clefts with marked deformity. Second, although the indications for bone grafting would seem still to be inadequately defined, clearly, if one intends to undertake either primary or early secondary bone grafting, it would seem desirable to do this in the presence of the best possible arch alignment, and this may most readily be achieved by early orthopaedic treatment. There are, as has been mentioned, other advantages to the surgeon resulting from this treatment but they might best be regarded not so much as firm indications for treatment but as fringe benefits.

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Acknowledgment: It gives me pleasure to acknowledge my great debt to Dr. Burston and Dr. Millard for their teaching and inspiration which I trust is evident in the text of this paper. My thanks are also due to them for allowing me to use their illustrations.

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