THE FORMATION OF A CLEFT PALATE UNIT—A PRELIMINARY REPORT

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With cleft palates, as in other surgical fields, changing techniques and better management of the patient as a whole render inexcusable the operative procedures that merely drag together the edges of the defect and then leave a cosmetically and functionally crippled child, to adapt itself as best it may to the harsh competition of the outside world.

Ironically the problem of congenital defects increases as our competence to deal with them advances. In the past these cases struggled to take their place on the marriage market but the more skilled our rehabilitative procedures become, the more likely are they to perpetuate their disability, since a significant number show a hereditary tendency.

As in so much of modern surgery it is difficult for any one man to cover adequately a problem such as this and, following the example of several American and English Units, 3 years ago we established, at the Red Cross Children's Hospital, a Unit devoted entirely to the treatment of congenital clefts of the lip and palate. Since that date we have reviewed and treated approximately 300 cases and in another 2-3 years we should be able to judge our results dispassionately, and we hope then to draw conclusions that will enable us to improve further the treatment of these children.

It is fairly easy to recognize normal speech and, equally so, abnormal speech, but it is no longer sufficient to gaze raptly at a wide open mouth and decide whether to operate and what form of operation to use. Adequate surgery for these children depends to some extent on adequate investigations and follow-up. A plastic surgeon alone cannot do this; he needs the help of associates in the same field.

Composition of Unit

Our Unit is composed of the following:

(*a*) *Social worker*. The social worker assesses the history of the mother's pregnancy, judges extraneous factors which may have affected it, and investigates the hereditary background of the child. The parents are helped and encouraged to attend the clinic and provisions are made to help them at home if necessary.

(*b*) *Orthodontist*. This specialist sees the child shortly after birth and decides whether it is possible, by fitting acrylic prostheses, to lessen the alveolar deformity. When the operative procedures are completed he may assist the speech therapist by fitting temporary plates to improve the child's speech and later, when the child's permanent teeth have erupted, he is responsible for producing a normal occlusion and for remodelling the maxillary arch.

(*c*) *Speech therapist*. The speech therapist assesses the child's speech as soon as the child attempts to speak spontaneously and in sentences, and guides the parents in the establishment of the correct patterns of speech. Later, with the plastic surgeon, she assesses cases that present with speech problems or naso-pharyngeal incompetence.

(*d*) *Ear, nose and throat surgeon*. There is a high incidence of otitis media in children who present with a cleft palate and this may lead to partial deafness, which may prevent the child from speaking normally, although the anatomical mechanism for normal speech is perfect. Assessment of the degree of deafness in these children is, therefore, vital. These children, like any others, also present with the problems of enlarged tonsils and adenoidal pads, and it is for the otorhinolaryngologist to decide whether these should be removed and to perform the operation with the minimum of scarring.

(*e*) *Psychologist*. Where there is doubt as to the child's intelligence, estimation of the intelligence quotient is necessary to prevent unnecessary operative procedures in a vain drive for normal speech.

(*f*) *Radiologist*. Every child with a cleft palate has a palatogram taken as routine procedure, and in many this is followed by the use of an image intensifier to watch the movement of the palate during phonation or to make a record of these movements on cine film.

(*g*) *Photographer*. A complete visual record is kept of all cases.

(*h*) *Paediatrician*. We must not forget to treat the child as a whole since many of these children present with feeding problems or frank malnutrition. These children have to face a series of operations and hence need more care and protection than the ordinary infant.

(*i*) *Plastic surgeons* are responsible for the operative repair of these defects and for coordinating the group.

INCIDENCE AND CAUSATION OF CONGENITAL CLEFTS OF THE LIP AND PALATE

Most authorities agree that the incidence of these clefts ranges from 1 in 700 to 1 in 1,000 of all births. Inheritance plays a considerable part in the causation of these lesions and figures vary from authority to authority, but it appears that in at least one-quarter of all patients there is an incidence of lip or palate deformity in some other member of the family. The gene appears to be a dominant characteristic, but outside factors such as anoxia in the early weeks of pregnancy, rubella, and perhaps vitamin deficiency, may act as aggravating factors. With both complete unilateral clefts of the primary and secondary palates and clefts of the secondary palate alone there appears to be a sex linkage and the former deformity occurs 3 times more frequently in males while the latter is more common in females. We have on our records one family where every member for three generations has had some form of cleft lip or palate.

CLASSIFICATION OF DEFECTS

Analysis of these cases is valueless unless some classification is chosen on which to base the assessment of the results. As we have chosen to follow the classification of Starke and Kernahan,[1] I shall briefly review the embryology on which that classification is based (Fig. 1). It is generally agreed that there are no true facial processes as described by His, but rather a series of ectodermal grooves separating masses of mesoderm. These masses grow differentially, penetrate, and obliterate the grooves to form the primary nasal septum, prolabium and pre-maxilla. Normal development of this area 'the primary palate' depends upon the successful penetration of mesoderm from 3 masses; 2 lateral and 1 median. The primary palate extends as far posteriorly as the incisive foramen; and its development is completed by the 7th week of intra-uterine life. From this primary palate are subsequently formed the central portion of the upper

lip and the pre-maxilla. Failure of this mesodermal penetration leads to breakdown of the ectoderm and cleft formation.

From the 7th to the 12th week of intra-uterine life formation of the secondary palate (hard and soft) occurs through the growth medianward and fusion of 2 laterally placed shelves. At the stage when mesodermal penetration of the primary palate has occurred and the bucco-nasal membrane has ruptured to form the primitive choanae, the 2 lateral palatal shelves are rudimentary. Hanging downwards and lying laterally alongside the tongue at first, these 2 palatal processes subsequently assume a position above the tongue. They meet in the midline and fuse, a process which begins at the incisive foramen, and progresses posteriorly during the 8th–12th weeks of intrauterine life.

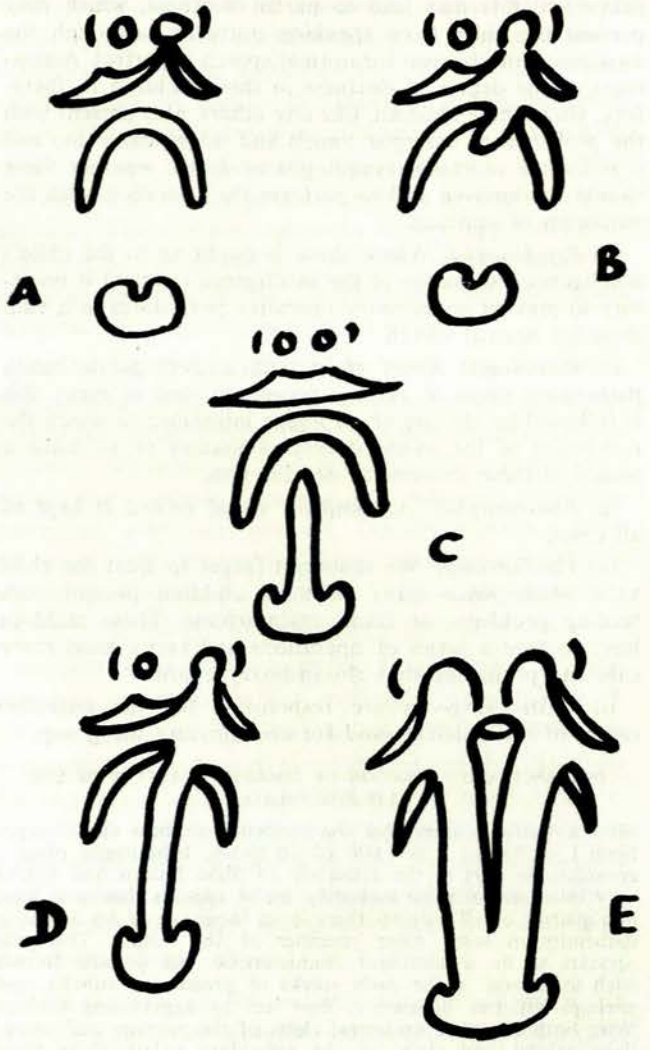

Fig. 1(A) Incomplete unilateral cleft of the primary palate. *(B)* Complete unilateral cleft of the primary palate. *(C)* Complete cleft of the secondary palate. *(D)* Complete unilateral cleft of the primary palate and complete cleft of the secondary palate. *(E)* Bilateral complete cleft of the primary and secondary palates

In the classification of Stark and Kernahan the incisive foramen divides anomalies of the lip (or primary palate, which develops between the 4th and 7th weeks and consists of the anterior nasal septum, prolabium and pre-maxilla), and the palate (or secondary palate, which develops between the 7th and the 12th weeks and is formed by the elevation and fusion of the rudimentary palatal shelves). Description of the cases is therefore based on an embryological development rather than the arbitrary use of the alveolus as the dividing point. A complete cleft of the primary palate involves the structures as far back as the incisive foramen and a complete cleft of the secondary palate involves the structures as far forward as the incisive foramen. These defects may be incomplete or sub-total and may present in combination in the same patient.

Clefts of the Primary Palate

In these cases the alveolar deformity is usually minimal and we prefer to repair the defect when the child is approximately 4 months old. The child's general condition should be satisfactory, the haemoglobin level should be normal for its age, and the surgeon should look for associated congenital deformities, e.g. cardiac defects. Here in the Red Cross Unit, we use a Z-plasty repair developed at Red Cross which has proved both simple and satisfactory[2] (Figs. 2 and 3). The normal height of the lip is measured on the uncleft side and a Z-plasty is so planned on the cleft side that when the triangles are interdigitated the height on both sides of the lip will be the same, thus producing a normal, symmetrical lip with a pouting effect and a cupid's bow. The Z shape of the scar prevents alteration

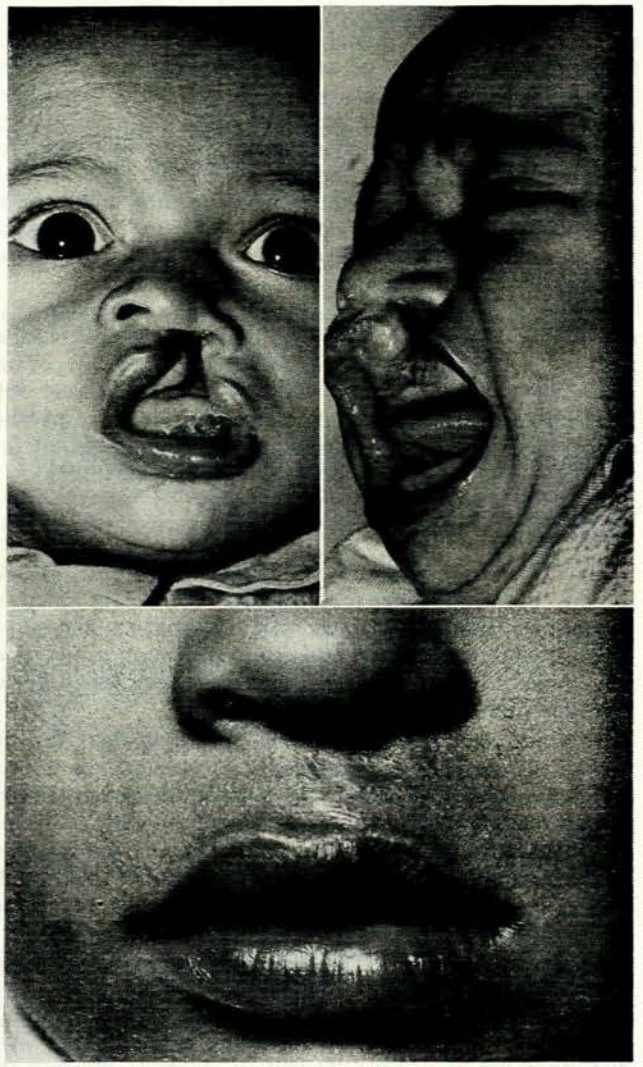

Fig. 2. Complete cleft of the primary and secondary palates before and 2 years after operation. Notice the normal appearance of the cupid's bow and the full pouting upper lip (by courtesy of Mr. B. Binnewald).

in shape of the lip postoperatively such as is found in the simple straight-line repair.

Clefts of the Secondary Palate

These are more commonly found in girls and where the cleft of the secondary palate is complete there is often a poorer functional result postoperatively than with complete clefts of

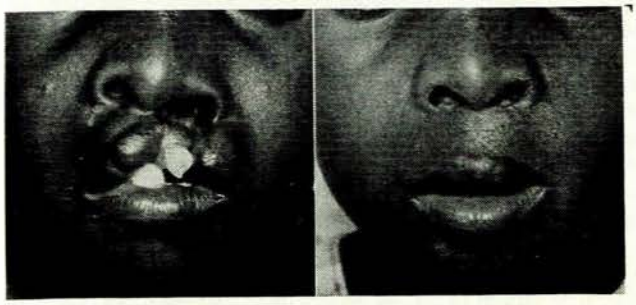

Fig. 3. Complete cleft of the primary palate, before and after repair. The child was 6 years old when first seen. It is interesting to note how invisible a zig-zag scar can become. The post-operative picture was taken 6 months later.

the primary and secondary palates. The object in these cases is to reconstitute the palate before the child starts to speak and yet at the same time to defer the operation for as long as possible in order not to interfere with the normal growth of the maxilla by producing early and unnecessary scarring. We find that the optimal age lies between 15 and 18 months. These defects are repaired by closing and pushing back the palate, using either a modification of the method described by Dorrance or, more commonly, the V Y retro-position described by Wardill and Kilner.

TREATMENT OF CLEFTS OF THE PRIMARY AND SECONDARY PALATES

Unilateral

These cases usually present with a fairly gross deformity of the alveolar arch and are seen shortly after birth by the orthodontist, who attempts some pre-operative modelling of the alveolar arch with acrylic moulds, if he finds the case suitable for this procedure[3, 4] (Figs. 4 and 5). It is this

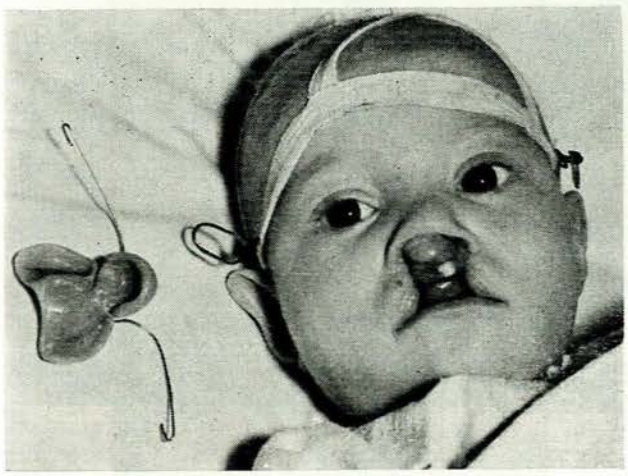

Fig. 4. Child with scrum cap and acrylic prosthesis. In this case a very prominent pre-maxilla is being moulded into a more normal position.

particular category which may present as an early feeding problem, but there is never any indication for early or heroic surgical interference. Gentle persistence with a normal teat which has an enlarged aperture, a teat with an obturator attached, or feeding with a teaspoon, succeeds in all these children.

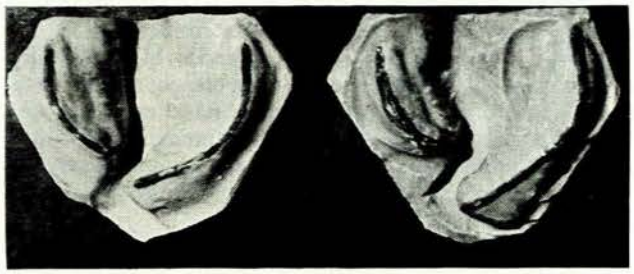

Fig. 5. Plaster models of the alveolar arch in a child with a complete unilateral cleft of the primary and secondary palates, showing how orthodontic treatment in the pre-operative phase has improved the shape of the arch.

The lip is repaired at the age of 4 months using the Z-plasty method, but we feel that it is absolutely vital to close the anterior palate at the same time in order, firstly, to reduce the incidence of oronasal fistulae and, secondly, to provide adequate cover for any subsequent bone grafts for the reformation of the maxilla on that side. Later, at 18 months of age, the posterior palatal deformity is closed by a Wardill V Y retro-position.

Bilateral

Here we face originally the problem of the projecting pre-maxilla which may be very marked in some of these cases. However tempting it may be to push this back surgically by excising a segment of the vomer or prevomerine bone, this cannot be too strongly condemned, since in a large proportion of cases it leads to gross underdevelopment of the middle-third of the face and a subsequent dish-face deformity. Reconstruction of the normal lip across the projecting pre-maxilla usually exerts enough dynamic force to help with the normal remodelling of the alveolar arch. Here again the orthodontist is able to help us pre-operatively in the first 4 months of life. To date we have been repairing these lips with a straight-line repair closing both clefts at the same time, but we find that we are achieving more satisfactory results by closing one side at a time by the Z-plasty technique and repairing the anterior palatal defect at the same time.

The prolabium is essentially the central structure of the lip and must not be sacrificed to form a columella nor must it remain an isolated island in the centre of the lip while the lateral elements are crossed beneath it, giving a long, flat expressionless lip as is seen in too many of lips repaired with the bilateral Le Mesurier technique.

At 18 months of age the posterior palate is repaired and at 2 years the columella, which is usually very short in these cases, is lengthened, using triangular flaps from the upper lip.

POSTOPERATIVE SPEECH ASSESSMENT AND ANALYSIS OF CASES

As soon as the child is speaking, the speech therapist takes a tape-recording of its speech, and decides whether the child has normal speech, is in need of speech therapy, or is unlikely to respond to speech therapy because of incompetence of the naso-pharyngeal sphincter. To aid this decision the child now has a palatogram and movements of the palate are followed on an image intensifier.

The child is then subjected to a series of tests which attempt to gauge the degree of competence of the naso-pharyngeal sphincter and the extent of nasal escape of air.[5] The initial tape-recording helps in the elimination of articulatory errors. These may be caused by an abnormal anterior arch and such factors as dentition or an unsatisfactory pattern of learning.

This is then followed by a simple test in which the child is persuaded to blow and to suck against resistance while the pressure achieved is measured. There is a constant leak in the testing system so that the child cannot cheat by blocking off the naso-pharynx with the tongue pressed against the palate, thus maintaining a fixed pressure.

Finally, a vital capacity machine is used to produce a graphic record of expulsion of a deep breath with the nose open and closed. Vital capacity studies are really a measure of the patient's ability to achieve palato-pharyngeal competence against a minimal resistance and the extent to which the 2 lines on the vital capacity graph diverge gives one some idea of the degree of nasal escape and thus of naso-pharyngeal incompetence. Therefore, as the patient responds to therapy and learns to compensate, this is the first test to move towards normal.

The pressure and suction studies require performance against an increased resistance and therefore demand still better control of the naso-pharyngeal sphincter. These studies do not measure speech efficiency, but we can certainly assume that if palato-pharyngeal competence is high, then speech training should be of great value.

If it is decided that the child has naso-pharyngeal incompetence which is due to shortness of the palate or abnormal depth of the naso-pharynx, these tests will give us some idea of the severity of the nasal escape and we can then judge whether this can be improved by a further push-back procedure to the palate, although we are reluctant to do these since they usually increase scarring in what is already perhaps a mobile but slightly short palate. If the palate is too short to reach the posterior wall of the naso-pharynx, the most physiological operation would seem to be the implantation of cartilage in the posterior naso-pharyngeal wall in order to allow the short palate to occlude against the augmented posterior wall (Figs. 6 and 7). We reserve the use of pharyngeal flaps of the Rosenthal type based either inferiorly or superiorly for those cases where the defect on the palatogram appears to be more than 0.75 cm. in depth, or where there has been no response to other forms of treatment. This operation certainly narrows the naso-pharyngeal opening, but the resultant action is a poor imitation of the normal functioning sphincter—thus we feel that it is wrong to use this procedure incorporated in the primary repair.

In rare cases where the child is a complete cleft palate cripple it may be necessary to rely entirely on a permanent prosthesis.

CORRECTION OF SECONDARY DEFECTS OF THE LIP

In cases of clefts of the primary palate which present with the initial repair already completed, the result can be assessed by considering 3 main factors.

Is the form of the lip satisfactory? The shape of the lip should be considered and one should judge whether it is too long, too tight, or badly notched. The most common complication of form found in older repairs of clefts of the primary palate is that of the tight upper lip. It is usually necessary in these cases to raise a small central triangular

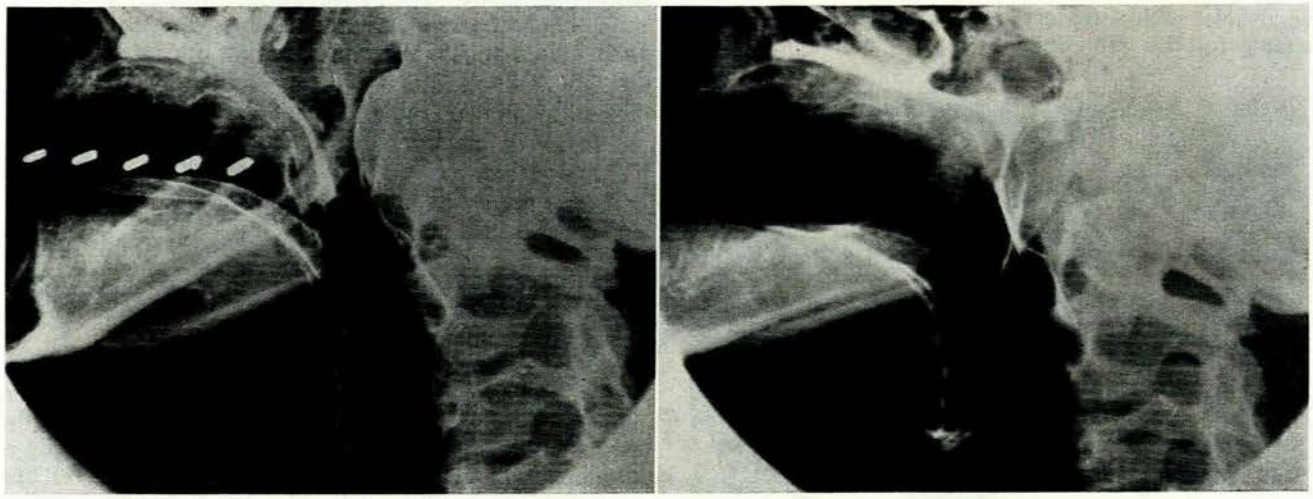

Fig. 6a *Fig. 6b*

Figs. 6a, b. The patient had a complete cleft of the secondary palate repaired at 16 years of age. The naso-pharynx was cavernous and the palate very short of tissue. Palatograms show that movement is good but the palate is too short to reach the posterior wall. The markings in the mouth in Fig. 6(*a*) are due to streaks of lead in a plastic rod and aid in the measurement of the defect. In (*a*) the palate is at rest; in (*b*) the patient is saying 'EE'.

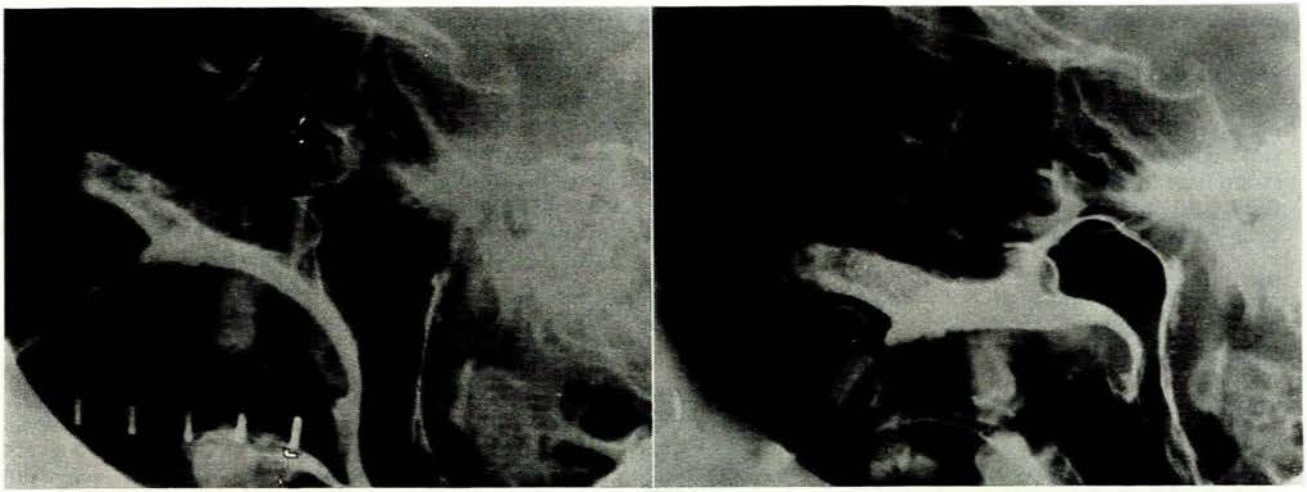

Fig. 7a *Fig. 7b*

Figs. 7a, b. Postoperative palatograms after a cartilage implant had been placed under the posterior naso-pharyngeal wall. In (*a*) the palate is shown at rest. In (*b*) it should be noted how the palate meets the posterior wall, and contrast medium accumulates above the now competent palato-pharyngeal sphincter. Postoperative speech is normal.

flap from the lower lip based on the marginal artery, and inset this into the centre of the upper lip. This flap should usually not exceed 0.75−1 cm. in width. Secondly, the degree of scarring on the lip should be considered and plans made to revise very unsightly or prominent scars.

Secondary revision of these scars can often be combined with a Z-plasty procedure, which also improves the form of the lip. Thirdly, the degree of associated nasal deformity should be considered. Minor variations in the symmetry of the nostrils can be corrected before the child goes to school, but we feel that it is a mistake to undertake extensive reparative procedures on a young child's nose since this must inevitably grow to adult size, and if too much scarring is produced one may find that the nose that looks satisfactory at the age of 6, after an extensive procedure, is completely asymmetrical at a later age when the child is fully developed. A more extensive corrective rhinoplasty with adjustment to the septum can be done between 17 and 21 years of age.

CONCLUSION

I have tried in this brief survey to show what is done in the recently formed Cleft Palate Unit at the Red Cross War Memorial Children's Hospital and to sketch our hopes for the future. We expect normal speech in 75% of our patients with clefts of the secondary palate and a near normal appearance of the repaired cleft lip by the time that the child goes to school.

One conclusion stands out above all others, and that is that the first man in the field has the best chance. Secondary corrective procedures, however extensive, are a poor substitute for a well-executed first operation in which gentle handling of the tissues is the keynote.

I wish to thank my associates in the Cleft Palate Unit for their unremitting interest and advice: to them belongs the credit for anything we have accomplished. I also wish to thank Mr. B. Todt for the photographs and illustrations; Dr. J. F. W. Mostert, Superintendent of the Red Cross War Memorial Children's Hospital, for kind permission to use this material; and finally, but by no means least, Prof. J. H. Louw, without whose encouragement and help the Unit would never have flourished.

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