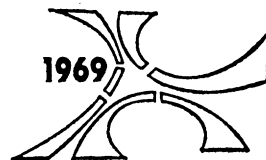


The Radical Repair of Cleft Palate Deformities



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In 1966, I suggested that, in complete unilateral and bilateral clefts of the primary and secondary palates, a radical one-stage repair might prove to be the treatment of choice (1). The concept of a one-stage repair has been discussed in a paper by Farina (3) and has undoubtedly been used by others before. However, one has the impression that these cases involved simple closure of the cleft with no attempt to lengthen the soft palate with a pushback procedure or to incorporate any of the finer points of multiple stage procedures. It also became apparent that the procedure was used mainly in the neglected case where the patient was an adult or an adolescent and that the rate of fistula formation was high (4). In the present series, 50 cases of complete unilateral clefts are presented and the first was begun in 1964. One-stage closure has been used for bilateral complete clefts and, to date, five cases have been repaired in this way. Technical problems have been overcome and the procedure is now routine at the Red Cross Children's Hospital in Cape Town.

In any discussion of this procedure, two questions spring immediately to mind: why close these defects in one operation, and what are the complications and contraindications of this procedure?

Probably the most valid indication for radical surgery is poor socio-economic conditions on the part of the patient's family, combined with a geographically widespread practice associated with poor follow-up conditions. When we feel that the child will not flourish at home, we admit the infant to a nursery where feeding can be supervised until the child is three months of age, weighs ten pounds, and has a hemoglobin level above 10 grams. The one-stage operation is then performed, and two weeks later the child is discharged, able to feed and presentable to the mother, who promptly takes more interest in her infant. Any possible risks attached to the operation must be weighed against a background of increased infant mortality in the first six months of life if the children are allowed to return home after birth without correction of the defect.

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Secondly, the operation is technically simple and, when performed under optimal conditions of intelligent anesthesia, has an operative time of two-and-a-half hours. The whole defect can be closed in layers, and one does not have the problem of scarred interfaces which is found in multiple procedures. Therefore, there is a lower incidence of fistula formation and the pushback achieved is more effective.

Possible contraindications are many. Is the mortality increased? It is too early to judge, but in a short series of 50 cases the mortality was nil. Is the morbidity increased? Are there complications which might prove serious to the health of the child? In this group we have had only one complication which we regard as serious and that was a tracheostomy early in the series. This was due to an error of judgment on our part in operating on a child with multiple congenital anomalies (in fact a case of Goldenhar's Syndrome) who had a bead of pus at the tracheal opening on intubation and where the endotracheal tube had to be changed halfway through the procedure. Laryngeal edema followed and twenty-four hours later it was necessary to do a tracheostomy. The child recovered well and there have been no untoward sequelae.

Regarding minor complications, we have had one hematoma of the chest wall, one child was returned to the theater to coagulate a small persistent bleeder, and one child was left with a two-millimeter fistula in the center of the hard palate.

It is too early to produce a satisfactory assessment of speech, but the palates appear long and mobile. Our speech therapist, Miss Whiting, feels that early results are very encouraging and that every child tested so far has the potential for normal speech.

But what of the maxillary growth? Study models of the alveolar arches and dentition have been taken by our orthodontists, Drs. Mellville and Miller, and a cephalometric survey has been started. Our first impressions are that successful primary bone grafting may well increase the incidence of Class III malocclusions. Where the bone graft has not taken or where there has not been a primary bone graft for some reason, growth does not seem to have been interfered with by the radical surgery alone. We face the added problem that clefts among the nonwhite races seem to be wider on an average than those seen among European children and this may also influence the future growth of the facial bones.

The primary bone grafting (and by this we mean bone grafting done at the time of the initial repair and not later) has shown an 80% take initially. This was encouraging as we felt that this bone graft would prevent further collapse of the minor segment and that by linking the two segments it would encourage growth generally. The opposite proved to be true. Much of the bone graft was gradually absorbed; there was no sign that tooth elements migrated into the graft; and a successful take seemed to limit development of the maxilla without preventing further collapse of the minor segment. This agrees with the conclusions of

recent experimental work (8) and later series (6, 7). As we now have forty cases in which the primary bone graft has taken, we will omit bone grafting in the next 50 cases and later compare the two series.

In the first fifty cases in our series, when one nine-year-old and two six-year-old children are excluded, the average age was 9.4 months and the average weight 16 pounds 5 ounces. Ten of the fifty children had moderate to severe malnutrition which had to be corrected before operation, and the average loss of blood, excluding the three older children, amounted to 79 milliliters for an average weight of 16 pounds 15 ounces, or 12.1 per cent of the blood volume. It is easier to perform the operation on a young child and we now regard the optimal age as three to four months.

We have all learned and believe implicitly that there is no substitute for team work as an approach to this problem. The representatives of the various disciplines must meet regularly and each must make an enthusiastic contribution to this problem if the project is to succeed. There is certainly no place for the prima donna surgeon. Nowhere is this shown more clearly than in the contribution made by the anesthesiologist to this problem and his part in the procedure should be discussed before going into the techniques of surgery. We are fortunate to have the services of several anesthetists who show a marked enthusiasm for pediatric anesthesia. The initial impetus came from Dr. T. Voss, Anesthetist-in-Charge at Red Cross Children's Hospital, and I am indeed grateful for his help and comments.

The apparatus used consists of a Boyle's machine to which is attached a circle absorber with an automatic ventilator. The Bird ventilator is used through an elephant tube and, attached to the expiratory line, is a Drager infant respirometer which is fundamental for measuring the ventilation on a small child. A humidifier is attached to the circuit so that inspired gases are passed over a warm water bath at 60°C; these enter the patient at 32° to 35°C. This apparatus does not have the power to overheat and the inspiratory tube is lagged so the heat is not lost. Normally, gas, oxygen, and Halothane are the main anesthetic agents and these are combined with a relaxant such as Curare or Alloperin.

The ventilation of the child is judged on the basis of weight in pounds $\times 6$ being equal to the tidal volume, and the frequency of respiration should be as follows: below ten pounds, 25; ten to twenty-five pounds, 20; up to fifty pounds, 18; and up to a hundred pounds, 15 (Figure 1). Ventilation according to this pattern should give a P_{CO_2} of ± 30 with the normal level being 40. We find that as the P_{CO_2} drops, the cardiac output drops slightly. The child is intubated with a McGill armored tube surrounded by a carefully placed throat pack. Anesthesia is maintained with nitrous oxide, oxygen, and 0.5% Halothane. Five to ten minutes before the administration of the adrenalin solution by the surgeon, the Halothane is turned off.

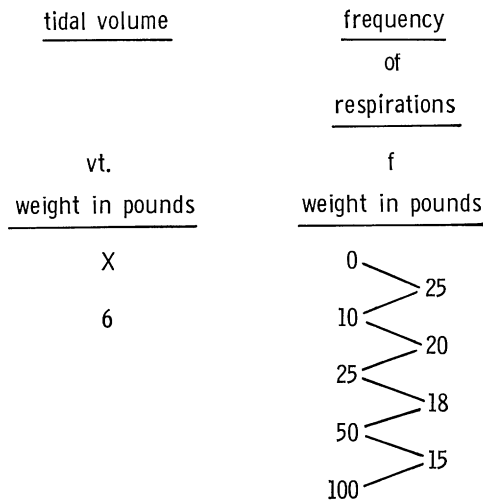


FIGURE 1. Ventilatory requirements for children undergoing surgery where a P-CO₂ of 30 mm Hg is the level intended.

The child is linked to an electrocardiograph and the heart beat is monitored both as sound and vision. A blood pressure cuff is attached to each upper arm, one to a mercury column and the other to a oscillometer. Each cuff must cover the entire upper arm from the elbow to the shoulder, as we find that if the cuff is too small the readings are misleadingly high. The normal rubber tubing from the cuffs is replaced by Tygon tubing with a one-eighth inch lumen which we find more rigid and which appears to give a more accurate reading. The oscillometer is used only when one is unable to get a reading from the mercury column and has proved to be hardly necessary in our series.

Temperature Control

The theater is maintained at 23°C and great care should be taken that the air-conditioning does not maintain the theater at too cold a temperature. This is pleasant for the surgeon but not suitable for the child. A humidity of 50 to 60% should be maintained and the child lies on a warming blanket warmed by circulating water set at a temperature of 37.5°C. The Humidifier in the anesthetic circuit is of greatest value in preventing heat loss. When this is not used, the dry anesthetic gases must be saturated by the child, and when the operation is a long one, this leads to heat and fluid loss. The child has a thermocouple in the rectum in order that a constant check can be kept on the body temperature.

Blood and Fluid Replacement

The fluid used for replacement is Ringer's Lactate with 5% invert sugar. The sugar in this formula is important as we find that these children sometimes become hypoglycemic due to preoperative starvation and

we have had isolated examples with slight ketosis. During the operation, 20% of the estimated blood volume should be given in Ringer's Lactate with 5% invert sugar. The blood volume is estimated in cubic millimeters by multiplying the body weight in pounds \times 40. During the operation all swabs used are collected and at intervals blood is cleared out of the suction apparatus. This is broken down in a measured volume of ammonia solution and the blood loss is read off on a colorimeter. Estimated blood loss is replaced plus 10% of the blood volume.

Management of the Child

Preoperatively, the anesthetist is responsible for checking on the state of the child's nutrition, the level of the hemoglobin, respiratory infections, and for prescribing the premedication according to the age and the weight of the child.

At operation the child is induced with 66% nitrous oxide, 33% oxygen and Halothane. The larynx is then sprayed with 2% Lignocaine and the child is intubated and the throat packed. The 0.5% Halothane is used until five to ten minutes before the adrenalin injection when the Halothane is discontinued. To tide the child over this period when adrenalin is being used, either a relaxant or methoxy fluorane (Penthrane) is used.

At the end of the operation, the relaxants are reversed with atropin 0.1 mg per ten pounds body weight and neostigmine 0.36 mg per ten pounds body weight. These are given routinely if a relaxant has been used at any stage in the operation, and they are given at the end of the operation while the patient is still being ventilated. The nitrous oxide is then turned off and the child is pumped on oxygen and given a measured dose of intravenous morphine, starting with 0.7 mg per ten pounds body weight. Then as the child is extubated and, in the immediate postoperative period, further doses are judged by the restlessness of the child. We find in our series that children in the fifteen- to twenty-pound range are the most resistant to sedation.

Throughout the operation the child has an intravenous drip in a foot vein which is inserted and running efficiently before the operation is started. A scalp vein drip set is used; we have not had to perform a cut-down on a vein in our series.

Postoperatively the child is kept in the recovery room until we are quite certain that he is breathing well, that there is no postoperative oozing of blood, and that the condition appears stable and the child adequately sedated. When the child is returned to the ward, he is placed in an oxygen-enriched, moisture-laden atmosphere and for this purpose we use the croupette routinely. The child is comfortable in this atmosphere and stays in the croupette for forty-eight hours. Care should be taken in a very hot climate that ice is placed in the water tank; otherwise the child may tend to show an excessive rise in body temperature while confined in the croupette.

Details of Operative Technique

When the anesthetist has completed his induction and the child is ready for operation, he is placed at the end of the operating table with a sandbag under his shoulders and the head thrown well back and resting in a ring to prevent side to side movement. The eyelids are firmly closed with two strips of zinc oxide strapping and the head is cleaned with a solution of PhisoHex followed by hibitane in water. The head is then lifted by the surgeon and replaced on a sterile towel. If a rib graft is to be used, the right side of the chest is prepared at the same time. The surgeon taking the rib graft and his assistant Sister have separate tables for their instruments and have no contact with the rest of the operation until the chest wall is closed and dressed.

Now that the child has been completely draped, his face is dried and the markings are made on the lip as for a Z-plasty repair (2). The lip and nasal area is now infiltrated with a solution of adrenalin which is mixed by adding 0.8 of a cc of 1/1,000 adrenalin to a small vacoliter of normal saline. These small vacoliters of normal saline contain approximately 180 cc, so the solution is in the neighborhood of 1/200,000. We are allowed to use 1 cc per pound body weight and usually on a fifteen-pound child it is only necessary to use 4 cc for the lip and 6 cc for the chest wall, leaving 5 cc which can be injected twenty minutes later for the palatal dissection. Seven minutes are allowed to elapse after the injection of the adrenalin and then the dissection and freeing of the lip is completed. Sulcal incisions are carried well down on each side but stop short of the opening of the parotid duct so that any relieving cross cut at the end of the incision has no chance of damaging the duct opening. When this dissection is complete the child's mouth is opened with a gag (at the present moment we use the Dingman gag) and the soft palate, hard palate, and vomerine mucosa are infiltrated with adrenalin solution. After seven minutes the edges of the defect are incised with a curved blade and then both mucoperiosteal flaps are completely raised and the dissection carried posteriorly to the pterygoid hamulus. The soft tissues are freed from the posterior edge of the bony palate, the nasal mucosa is undermined on the cleft side, and the vomerine mucosa is elevated on the uncleft side. The nasal mucosa is divided at the level of the posterior edge of the bony hard palate and this division allows a soft palate pushback of approximately one to one-and-a-half centimeters. The nasal mucosa and the muscle of the soft palate are then closed with 4/0 chromic catgut using everting sutures. This closure is continued on the oral surface of the soft palate with 4/0 black silk everting sutures. The freed nasal mucosa on the upper surface of the bony palate is then sutured to the vomerine mucosa with a continuous inverting suture of 5/0 chromic catgut, and this is carried on to the region of the alveolar defect. One is now left with an oval defect between the repaired soft palate and the

hard palate and this is filled by rotating an island flap of mucoperiosteum, as described by Millard (5), into the gap. Usually the mucoperiosteal flap from the smaller, or cleft side, is used for this and the larger mucoperiosteal flap from the normal side is then swung across to give a double layer closure. This flap is attached to the thin bone of the vomer and the bony palatal shelf of the opposite side (Figures 2 to 6).

The gag is now removed from the mouth, making sure beforehand that there is complete hemostasis, and the repair of the lip is commenced. The repair of the nasal floor is now completed and, if a bone graft is to be used, it is placed in the alveolar gap firmly wedged to the bone on each side. The oral surface of the defect is then closed with a flap from the mucosal lining of the lip. Finally, the lip itself is closed in layers using 4/0 chromic catgut for the muscle layer and 6/0 black silk for the skin and mucosa of the lip. A suitable soft portex nasopharyngeal

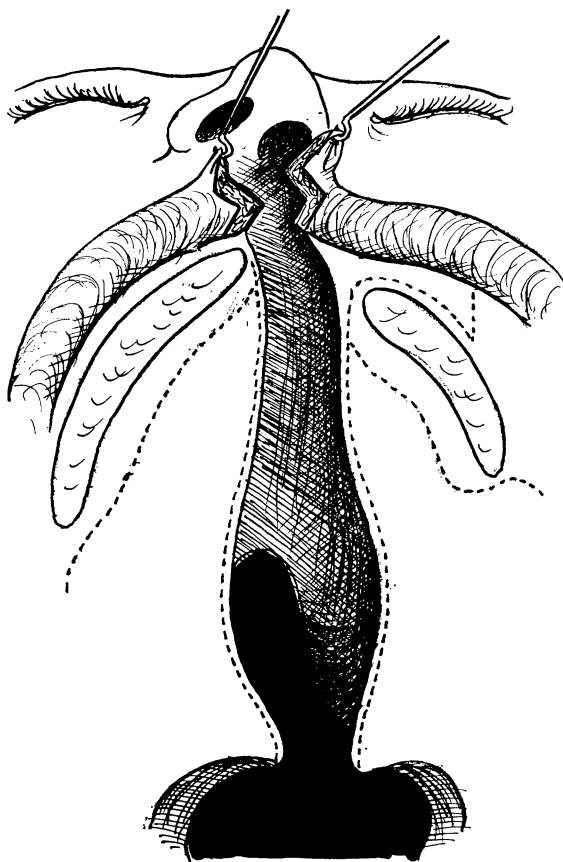


FIGURE 2. Sketch of a complete unilateral cleft of the primary palate and complete cleft of the secondary palate. The lip has been incised as for a Davies Z-plasty repair and the dotted lines on the diagram show the palatal and sulcal incisions.

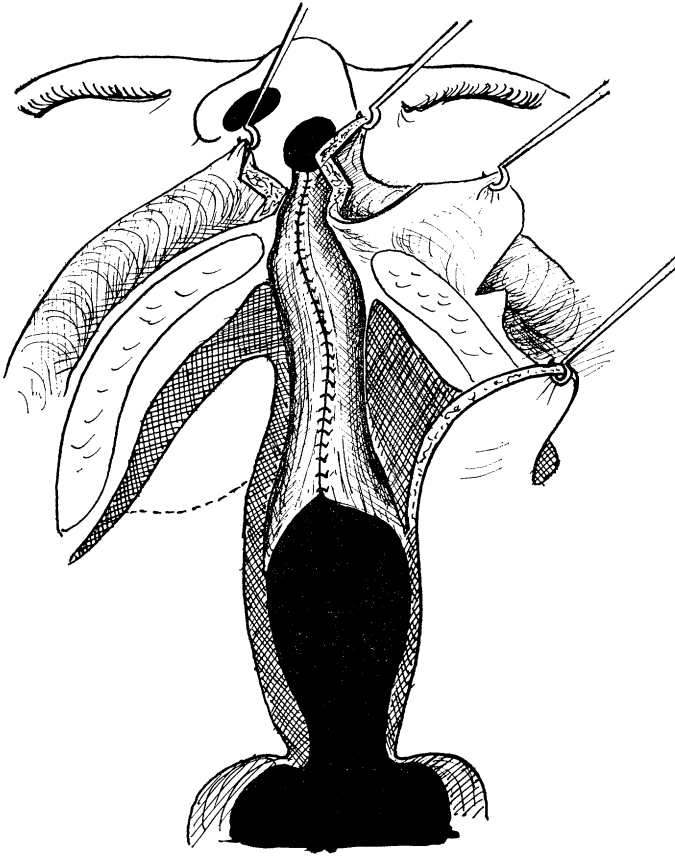


FIGURE 3. Vomerine and septal mucosa has been reflected and sutured with fine everting sutures to the nasal mucosa of the cleft side. A mucosal flap has been raised from the sulcus on the cleft side.

tube of the correct diameter is now selected and passed into the nostril on the normal, or uncleft, side; a lighted retractor is used to make sure that the flanged end of this tube projects just beyond the posterior border of the repaired soft palate with the open part of the flange facing towards the oral cavity. A small safety pin is then inserted through the margin of this tube at the level of the nose and zinc oxide strapping is placed across the cheeks fixing the safety pin firmly in position. The purpose of this fixation is to prevent the tube from entering further into the nasopharynx, from sliding out with the movement of the child's head, or from rotating and causing the open flange to face the posterior wall of the nasopharynx, thus occluding itself. A final inspection of the repaired palate should confirm hemostasis and the anesthesiologist now extubates the child. A fine flexible polyethylene catheter can now be passed down the nasopharyngeal tube and the nasopharynx and oro-

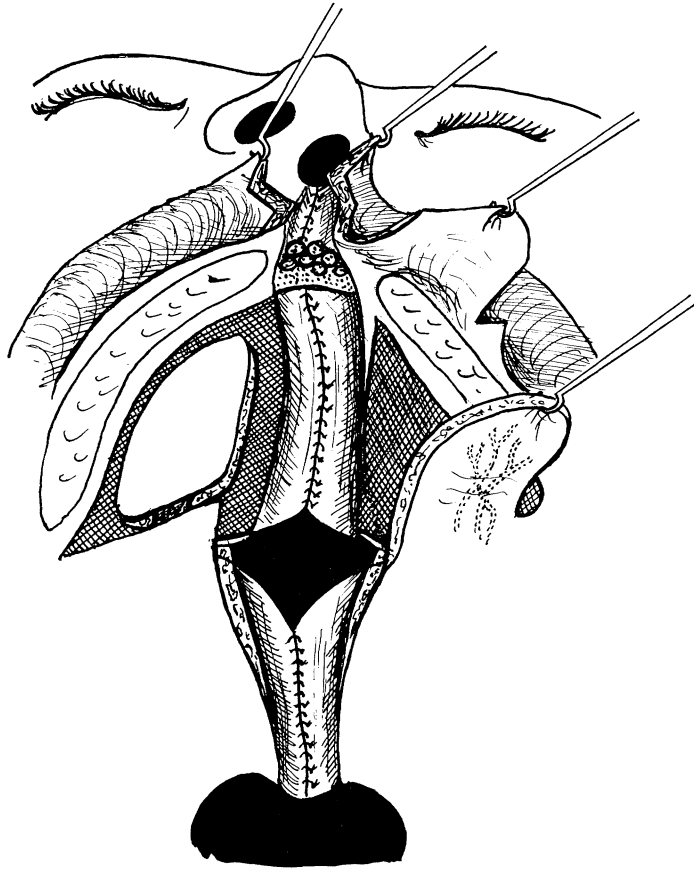


FIGURE 4. The nasal mucosa of the soft palate has been closed with fine everting sutures; the rib bone graft has been laid across the alveolar gap; and the island flap has been isolated on its vascular pedicle. This diagram shows the island flap as coming from the uncleft side, but it is usually routine to use the smaller area on the cleft side. The nasal mucosa is divided from the posterior edge of the hard palate allowing the soft palate to fall backward leaving a large diamond-shaped defect.

pharynx sucked out by this means without damaging the palate. This method has proved so convenient that it is now used routinely even where the repair has been confined to a cleft of the secondary palate.

As soon as the anesthesiologist is satisfied that the child has recovered from the anesthetic and is breathing well by himself, the child is moved to the recovery room where he is watched for the next hour. If at the end of this period there are no signs of excessive blood loss and the child appears to be breathing well and adequately sedated, he is moved to the ward where he is nursed in a croupette in a moist oxygen-enriched atmosphere. Precaution is taken at all stages after the operation that the

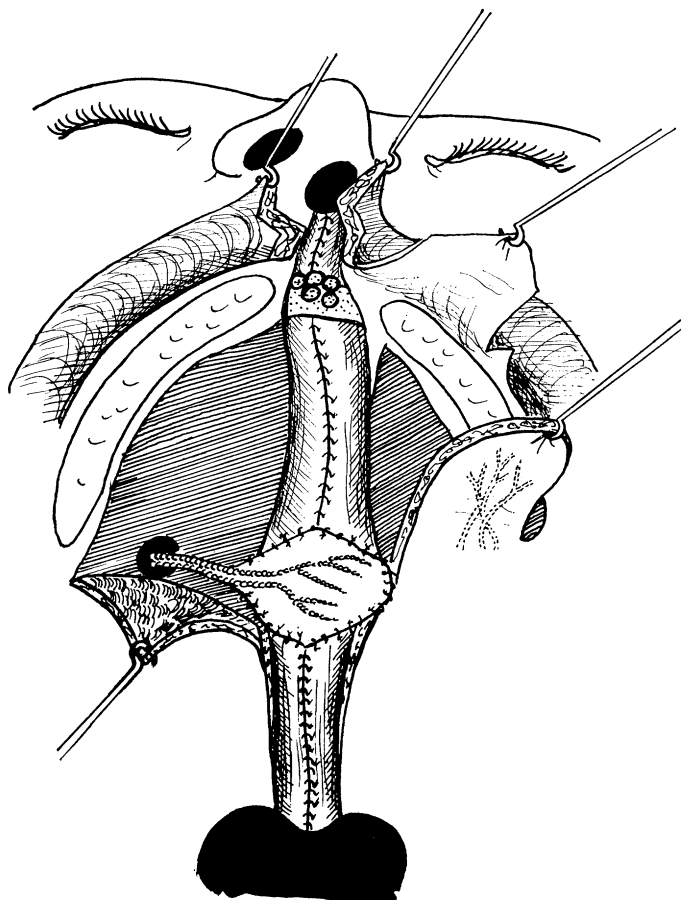


FIGURE 5. The island flap has been swung into the defect in the nasal mucosa thus maintaining the pushback achieved.

child's hands are tied down or fixed in some way in order that they cannot disrupt the repair by insertion into the mouth.

Postoperatively the child is nursed in the croupette for forty-eight hours. Twenty-four hours after the operation the nasopharyngeal tube and the outer dressing on the lip are removed. The sutures on the lip are covered with an antibiotic cream, and application is repeated twice daily for the first four days. For the first twenty-four hours the child is fed on clear fluids and then proceeds to a semi-soft diet. One week after the operation the sutures are removed from the lip repair using Halothane analgesia, and finally, two weeks after the operation, the stitches are removed from the palatal repair by the same method.

In the past, authorities have disagreed violently on the age at which one should undertake surgery and the staging of these surgical repairs.

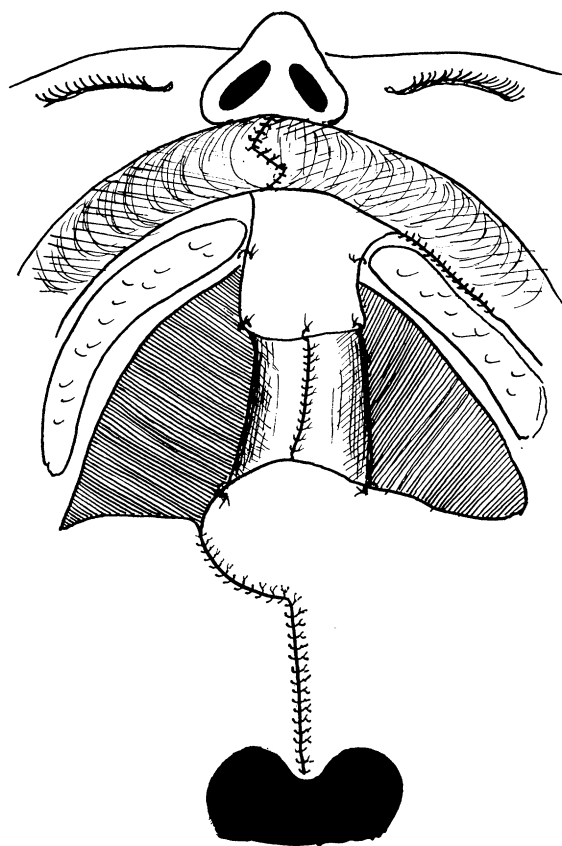


FIGURE 6. Mucomuscular sutures have closed the oral surface of the soft palate. A palatal flap is swung across to cover the island flap and fastened by sutures to the bony hard palate. The mucosal flap from the sulcus has been used to provide an oral surface to the bone grafted area, and the Z-plasty repair of the lip is finally completed.

We do not wish to be unnecessarily optimistic or enthusiastic, but we feel that a case can be made for the radical repair of these defects at an early age.

Summary

Since 1964, the author has repaired complete unilateral clefts of the primary and secondary palates as a one-stage procedure. Immediate results are satisfactory, but we must defer final evaluation of the effects on growth until the patients are at least 10 years old.

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