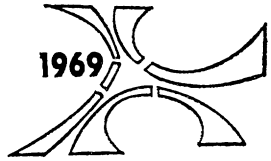
Early Management of the Pierre-Robin Syndrome



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The baby with cleft palate, micrognathia, and respiratory obstruction is in a potentially lethal situation and the management of this neonatal emergency has exercised the minds of all clinicians charged with solving the problem. In various clinics throughout the world, treatment has followed one of two fundamentally different approaches: either conservative management, with posturing in the prone position aided by pharyngeal suction until the baby is able to control his tongue, or some method of fixing the tongue in a forward position by operative means.

At the Royal Children's Hospital in Melbourne, a surgical technique has been evolved for forward fixation of the tongue to the lip. This paper will describe the operative and anesthetic technique employed and give an indication of the results. During the period under review, from 1952 to 1969, 27 cases were judged to fall into this group of cases of Pierre-Robin syndrome.

In some cases, the respiratory obstruction is mild and transitory; in others, the condition is severe and prolonged with a gradation from one extreme to the other. It is sometimes difficult to exactly distinguish the milder case from the much larger groups of babies with isolated palate cleft and micrognathia, but without respiratory obstruction; and this difficulty should be taken into account when comparing methods and results from different centers.

Case Material

During the review period, 289 babies with isolated cleft palate, most with some degree of micrognathia, were admitted to the hospital; it would seem, therefore, that approximately 10% of all babies with Group 3 cleft may be at risk.

Figure 1 demonstrates that the outcome in these cases prior to 1956 was discouraging. That was during the period when conservative meas-

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	CASES	DIED	SURVIVED	
			WITHOUT OPERATION	WITH OPERATION
1952 - 56	IIIIII	IIIII	I	
1957 - 61	IIIII	I	III	I
1962 - 66	IIIIIIIIIII	II	III	IIIIII
1967 - 69	IIIII			IIIII

FIGURE 1. Total number of cases with deaths, and survivals with and without the operation of tongue-lip anastomosis.

ures were followed, and the techniques of posturing and pharyngeal suction then in use were inadequate. In the late 1950s some infants survived without operation, but only after many weeks in the hospital, under constant and exacting nursing care. The babies were always a cause of great concern and the prolonged morbidity and convalescence kept mother and baby apart for long periods.

With the introduction of lip-tongue anastomosis, a dramatic reduction in mortality, morbidity and hospitalization occurred. In the past eight years, no case has been lost from respiratory obstruction after this operation, the morbidity has been markedly reduced, and the stay in the hospital shortened so that most babies are back with the mother within two or three weeks. There have been two deaths from associated congenital abnormalities.

Emergency Management

Respiratory obstruction is a neonatal emergency; it requires early and positive treatment and rapid transferral to a center with the personnel and experience to cope with the problem. If posturing the baby in the prone position is not immediately effective, emergency treatment should consist of a stout silk suture placed transversely through the dorsum of the tongue as far back as possible to pull the posterior half of the tongue out of the pharynx and palate cleft. Failing this, a towel clip or safety pin further forward, but avoiding the tip, is effective.

Assessment

In assessing the condition, one must take into account the range of severity, from the child with an isolated cleft palate (so commonly associated with micrognathia) who has no sign of respiratory difficulty

at any time, to the child with severe obstruction at all times even when lying prone. Somewhere between these extremes is the baby who has respiratory difficulty for a day or so, but who rapidly gains control of his tongue within a few days of birth, and the baby who manages well for two or three weeks and then develops obstruction, as the tongue enlarges.

Once the child is in the hospital, the problem must be assessed as completely and as accurately as possible. Here the trained nurse in charge of the neonatal ward, with several years of experience behind her, is an invaluable ally. Several babies have entered the hospital after one or two attacks of cyanosis and rib retraction, usually associated with feeding, but settle down quickly and have no further trouble no matter in which position they lie, after pharyngeal mucus has been cleared with careful and efficient aspiration.

In some cases, there may be good reasons for postponing surgical fixation of the tongue. Perhaps a misguided and clumsy attempt has been made elsewhere to perform an anastomosis and has failed; perhaps a crushing clamp has been applied to the tongue tip; or perhaps the baby is premature or otherwise small and any operation would be considered hazardous.

To overcome this difficulty, invaluable help has come from an expert team of anesthesiologists with the passage of a nasotracheal plastic tube in the conscious baby (Figure 2). This technique, first introduced by Brandstater (*2*), and developed by Allen and Steven (*1*) and McDonald and Stocks (*5, 8*), immediately provides an adequate airway; the tube can be left in situ for several weeks if necessary but the aim in these cases is to use it as an interim measure for a short period, until the anastomosis can be performed at leisure and in safety.

Indications for Operation

In the average case, the decision to operate is made if the baby has one attack of cyanosis and rib retraction while under expert care and observation. Some years ago several babies were lost, usually in the early hours of the morning, before we were sure enough of the technique to recommend operation without hesitation.

Technique

The child is anesthetized *after* the passage of an endotracheal tube. The bulky middle third of the tongue is transfixed transversely from the dorsum with a stout silk suture for control and traction.

On the undersurface (Figure 3a), a transverse incision is made just distal to the submandibular ducts between the plica fimbriata, and an incision is made at right angles to each end of this first incision, coming forward almost to the tip. Thus a rectangular flap is outlined, deepened into muscle, and raised.

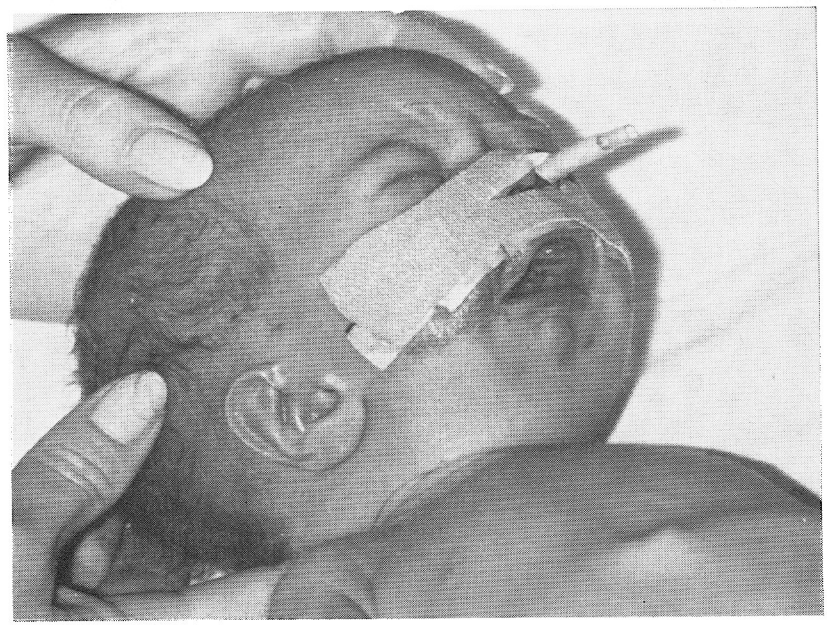

FIGURE 2. A severe case with gross micrognathia and indwelling nasotracheal tube as an emergency measure.

It is noteworthy that as the flap is raised and the tight frenum and sublingual mucosa are brought forward, the tip of the tongue becomes more mobile and can now be drawn out of the mouth to a greater extent. We believe this mobilization of the tongue tip is an important reason why we have had no experience of the tongue-hump described by Routledge (*7*); the operation of Douglas (*3, 4*) more closely parallels our technique in this regard.

A rectangular flap of lip mucosa and muscle (Figure 3b), based in the labial sulcus and coming forward almost to the vermilion border, is then raised. The free end of this lip flap is then sutured to the free margin of tongue mucosa close to the submandibular ducts, one or two buried catgut sutures unite tongue muscle to lip muscle and the flap sides are sutured together, lip flap to tongue flap. The repair is completed by suturing the free end of the tongue flap to the free margin of lip mucosa close to the vermilion border (Figures 3c, 3d, and 4).

This is a meticulous three-layered repair, carried out on a quietly anesthetized baby. In no case has the repair come apart.

In some cases, the traction suture has been left in place for a day or so postoperatively, but with increasing confidence it has been removed as soon as the baby is awake. More recently, if there has been any doubt about the postoperative airway, a nasotracheal tube has been left in place for a day or so. It is then removed by the expert anesthesiologist,

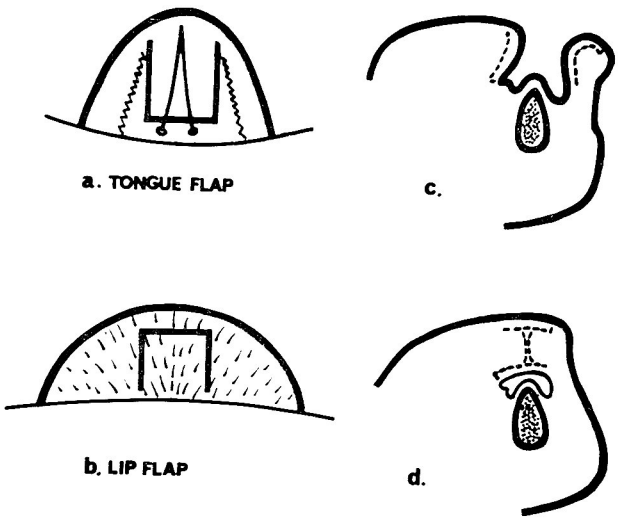

FIGURE 3. Diagram to illustrate the principles of the operative technique: a) the tongue flap; b) the lip flap; c) outline (dotted) of the flaps; and d) outline (dotted) of raw surfaces sutured in apposition.

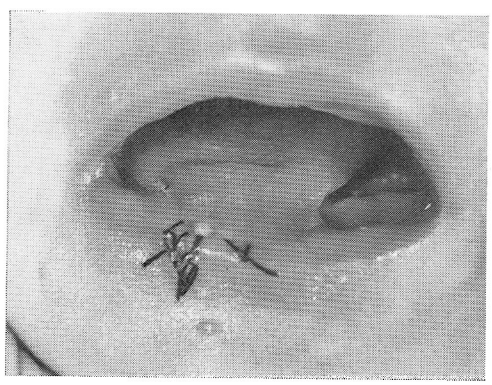

FIGURE 4. An anastomosis four days after operation, showing the early stage of sound union.

who can reinsert it if necessary with no more difficulty than in the pre-operative period.

Experience has shown that, following the anastomosis, there is a dramatic improvement in the airway, and the early resumption of normal feeding and normal weight gain. This relieves the nursing staff of a tremendous responsibility, and allows the baby to leave the hospital within two or three weeks of the operation, in the care of its own mother.

The anastomosis is maintained until after the palate is repaired at twelve months (Figure 5). This has not prevented the normal mandibular growth described by Pruzansky and Richmond (*6*), nor interfered with

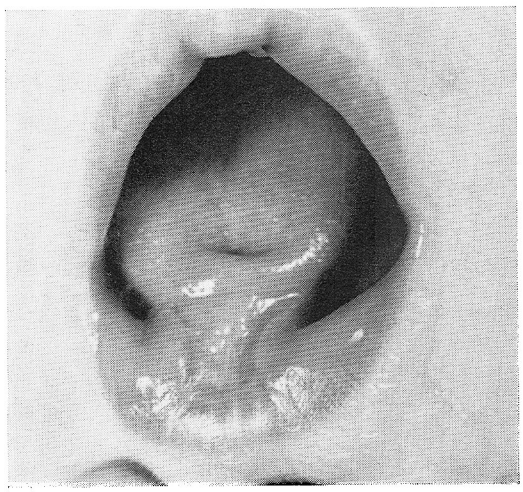

FIGURE 5. An anastomosis twelve months later.

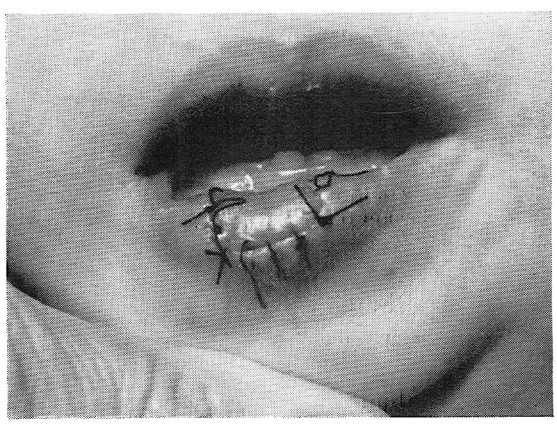

FIGURE 6. After separation of the anastomosis, showing eruption of lower central incisors and replacement of lip flap (tongue flap also replaced to its original position but not illustrated). The lip flap would have been in contact with the incisors; it shows no ill effects.

tooth eruption. Nor have the erupting teeth caused any trouble to the undersurface of the flap (Figure 6).

Summary

A series of 27 cases of the Pierre-Robin syndrome admitted to the Royal Children's Hospital in Melbourne during a 17-year period is presented. Mortality in the earlier years was very high. During an interim period, mortality was reduced by intensive nursing care, but morbidity remained high until tongue-lip anastomosis was developed to a stage where it could be recommended with confidence. An operation to fix tongue to lip has been performed in twelve cases without mortality and

with a dramatic reduction in morbidity and length of stay in the hospital. Details of the operation have been given. The value of nasotracheal intubation as an emergency measure has been discussed. Anesthesia is induced after an endotracheal tube has been passed. There is no evidence that this operation has an adverse effect on mandibular development or on tooth eruption.

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