TREATMENT OF CLEFT PALATE ASSOCIATED WITH MICROCYNATHIA

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In spite of the present-day technique and medical research no entirely satisfactory method has been developed to relieve the distressing symptoms of the baby born with a cleft palate and an associated micrognathia with glossoptosis. This syndrome—known as the Pierre Robin syndrome—is relatively rare, and a plastic surgeon may see only one to two cases a year. Because of the limited number of cases available the writer is putting forward the following observations, appreciating that two cases are insufficient to form any definite scientific opinion.

The cleft palate usually involves two-thirds of the hard palate and is the most difficult type of palate to repair. The mandible is unusually small but is proportionally developed. The lower alveolar ridge is well behind the upper alveolar ridge. There is difficulty in breathing and feeding. Other congenital abnormalities, such as congenital heart, may be present and may be the cause of early death. This subject has been well reviewed by Kiskadden and Dietrich (1953), who discussed the treatment and formulated the recent research into mandibular growth. It is interesting to note that normal mandibular growth is expected to take place in the early years of life with infants suffering from micrognathia.

In addition to the micrognathia the tongue appears smaller than normal and the tip is not well developed. A tongue tie may be present. The small mandible causes the tongue to be in a more posterior position than normal and it is displaced backwards into the hypopharynx. In addition the tongue tends to fill in the cleft, particularly the soft palate cleft, and cause obstruction of the nasal airway. Swallowing is difficult, probably due to the fact that the tongue falls back and acts as a ball-and-socket joint in the pharynx. The child is unable to suck and has to be fed with a spoon or gavaged. The interference with function of the mandible causes loss of tone and subsequent undevelopment of the muscles of mastication.

The principle of the treatment is to alleviate the obstructed breathing and to aid feeding and sucking. If the child can be maintained for six to twelve months it will be found that normal breathing and feeding will return and the palate repair may be performed about twelve to eighteen months. The treatment advised falls mainly into six groups:

1. Posture.—Quite a number of these cases respond well to posture. The baby is placed on the side or on the front and this allows the tongue to come forwards and relieve the obstruction of the airway. There are, however, babies with micrognathia who do not respond adequately to this method of treatment and have to be aided with their oxygen intake by an oxygen tent or intranasal oxygen.

2. Surgical Fixation of the Tongue.—Douglas (1950) had advised that the tongue be fixed surgically to the lip. This is done by creating a raw surface
on the underneath part of the tongue and suturing it to a raw surface made on the lip and the alveolus. In a limited number of cases this method is very effective, but it does fail to bring forward the posterior part of the tongue and completely relieve the obstruction.

3. Apparatus attached to Feeding Bottle.—Davis and Dunn (1933) have advised the use of a special adapter fitted to a feeding bottle, which presses on the upper lip whilst the baby is feeding. The apparatus can be adjusted till the teat is satisfactorily engaged between the upper and lower jaws. By gradually moving the position of the teat outwards it has been suggested that the lower jaw will be brought forward and growth of the mandible stimulated. This method has been used, but the babies have not sucked satisfactorily and have had to return to spoon or gavage feeding. It is doubtful if the apparatus helps mandibular growth and Kiskadden in his review states that normal growth takes place in early life.

4. Traction of the Mandible.—Many ingenious methods have been devised to maintain the mandible in a forward position and thereby relieve the obstructed airway. Longmire and Sanford (1949) obtained traction with a wire attached to a metal plate placed on the lingual side of the symphisis of the mandible. There is no doubt that this method would bring the mandible and the tongue forward but it would be difficult to maintain this apparatus effectively over a long period of time.

5. Tracheostomy.—This operation is difficult to perform on a child so young and is only a temporary expedient. Aspiration of vomit may occur with fatal termination, especially after a feed. It is felt that this line of treatment is undesirable.

6. Early Repair of the Palate.—Early repair of the palate has been advised by Mitchell-Nelson (1950), but there is no suggestion as to how early this operation should be undertaken. Should the severe symptoms of obstructed breathing and interference with feeding persist, and not respond to postural treatment, the writer feels that repair of the palate should be undertaken without delay during the first few months or even during the first forty-eight hours of life. At this age an infant stands operative treatment well. It is better to undertake palate repair early than to delay until there is rib retraction and chronic chest complications. If there are no distressing symptoms, the palate may be repaired at a later date.

Before discussing treatment it is interesting to compare the X-ray of a baby with a normal mandible with the X-ray of a baby with micrognathia and a cleft palate.

An X-ray of a normal baby (Fig. 1) shows a tongue lying well forward in front of the soft palate, giving adequate airway between the anterior surface of the soft palate and the posterior part of the tongue.

The X-ray of a baby with a soft palate cleft but no micrognathia shows more or less a similar picture (Fig. 2, A and B).

A baby with micrognathia has the tongue in a much more posterior position, and with the associated cleft palate the tongue tends to fill the cleft and lie on the posterior pharyngeal wall, completely obstructing the airway. Repair of the palate
allows the tongue to rest against the soft palate and prevents it from completely obstructing the airway.

Fig. 1.—Showing tongue and soft palate in normal 6-month-old baby.

Fig. 2 shows position of tongue and soft palate in baby with a normal mandible but with a cleft of the soft palate.

Fig. 1

Fig. 2

From the clinical course of the following two cases described there is evidence to support the view that early palate repair is beneficial to a baby with micrognathia.

CASE REPORTS

Case 1.—M. C., male; date of birth, 1st October 1953; weight, 7 lb. This child was born with micrognathia associated with cleft palate (Figs. 3 and 4, A and B). For
the first few months of his life he was a constant nursing problem. He had to be placed on his side to relieve breathing, but there was no need for an oxygen tent. All feeds had to be gavaged and bottle-feeding was impossible. At 5 months of age (Fig. 5) he was just 10 lb. in weight, having gained only 3 lb., and the paediatrician suggested that

Fig. 3.—Case M. C. One day old.

Fig. 4.—Case M. C. Ten days old, showing micrognathia and glossoptosis. Note tongue filling hypopharynx and also the nasopharynx through the palate cleft.

the palate repair might be helpful. In the first instance this treatment did not appear logical, but on consideration it was thought that it might be helpful. Soon afterwards, on 12th March 1954, the palate was repaired and the tongue sutured forward according to Douglas.
For the first twenty-four hours post-operatively the child fed well with a spoon, and the following day bottle-feeding began. This was the first time the child had been able to take feeds in this manner, and within forty-eight hours routine bottle feeds were instituted. The tongue suture pulled out within forty-eight hours, with a result that the tongue was free again and sutures were then removed. About the third day the

Fig. 5.—Case M. C. Five months old and 10 lb. in weight just before palate repair.

Fig. 6.—Case M. C. Aged 21 months (seventeen months after palate repair). Note position of tongue and soft palate and development of mandible.
nurses observed that the child was putting the tongue out in anticipation of his feed, as is often seen in a normal baby. The child made a quick recovery, and within a few days breathing was normal and he was sleeping lying on his back. He was discharged home three weeks after his operation, having gained 12 oz. in weight. On 21st April 1954 his weight was 12 lb. 8 oz. and in five weeks he had gained 2 lb. 8 oz. On 16th June 1955, seventeen months after operation, he weighed 20 lb. 2 oz., well within normal limits (Fig. 6, A and B, and Fig. 7, A and B).

Following the experience of the previous case, a palate repair was performed in the second case with micrognathia at 1 month of age.

Case 2.—B. S., female; date of birth, 3rd September 1954; weight, 6 lb. 11 oz. The micrognathia and cleft palate in this infant caused severe asphyxia which did not respond to postural treatment, and the baby was treated most of the time in an oxygen tent. In addition, all the feeds had to be gavaged. On 1st October 1954, when the child was a month old and 6 lb. 4 oz. in weight, an operation was performed for repair of the palate and the tongue was sutured forwards. The tongue suture was released in forty-eight hours and after three days the administration of oxygen by means of an oxygen tent was no longer necessary. The baby took feeds better and about half her feeds were by bottle; the remainder still had to be gavaged. Unfortunately there was paresis of the right phrenic nerve which interfered with the filling of the right side of the thorax. This predisposed to recurrent attacks of chest infection and the baby made only slow progress. However, she did gain 2 lb. in three months and breathing was very much easier than prior to the operation. On 3rd January 1955, when 4 months of age, she contracted a pulmonary infection and, with the inability of the right side of the diaphragm to move, she died from pneumonia.
FIG. 8
Technique of palate repair.
TECHNIQUE OF PALATE REPAIR

It is important that the repair of the palate should be carried out in one operation. The four-flap Wardill often leaves some defect in the hard palate, and the method advised in these cases of micrognathia with involvement of two-thirds of the hard palate is to raise two long palate flaps, leaving the posterior palatine artery intact. The flaps are mobilised from the medial pterygoid plate and the hamulus is fractured. The lining is obtained by reflecting the mucous membrane on each side of the vomer, which is midline in these cases. There is usually some difficulty in suturing the lining mucous membrane in the region of the junction of the hard and soft palate and sometimes a small gap will remain in the lining. However, if the palate flaps are mobilised adequately they should be able to give satisfactory mucous membrane covering on the palate side and the suture line should heal without any defect. In order that the palate flaps may remain in apposition to the lining, one or two silk sutures are passed through the palate flaps and also through the septum. This ensures that the suture does not cut out as may happen if the suture is passed through the nasal mucous membrane (Fig. 8, A, B, and C). Technically the repair of the palate in the small baby is a practical proposition. The greatest difficulty is from the anaesthetist's point of view, who may have much trouble in inserting a tube into the trachea in a baby with micrognathia with some distortion of the anatomy in the region of the epiglottis.

CONCLUSIONS

Early palate repair is advised in babies with a cleft palate and an associated micrognathia with glossoptosis, especially in those cases that do not respond to postural treatment. In selected cases the writer considers that the palate repair should be performed as an emergency operation during the first forty-eight hours of life.

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REFERENCES