

Lingual and Labial Movements of Pierre-Robin Syndrome Patients Treated by Beverly Douglas Procedure

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In 1923 Pierre Robin (6) described a condition in young children in which the tongue assumed a posterior position in the oral cavity because of hypoplasia of the mandible. Robin described associated difficulties of feeding and respiration. Since his writings, other information has been made available about the syndrome which bears his name. The Pierre-Robin Syndrome is usually thought to consist of a small jaw (micrognathia), a falling backward and downward on the tongue (glossoptosis); and often an isolated cleft palate.

In normals, according to Douglas (2) and Steigrad (9), the attachment of the genioglossus muscle and the frenulum of the tongue to the mandibular symphysis is the primary means by which the tongue is held forward. Goldberg (3) states that the genioglossi pull the tongue forward, thus overcoming the backward and downward pull of the hyoglossal muscle and the upward and backward pull of the styloglossal muscle. In micrognathia the tongue receives little, if any, support from the genioglossi attachments since these attachments are much more posteriorly placed. The tongue, consequently, acts as a ball valve and presses on the epiglottis, allowing expiration but preventing inspiration of air (2, 3, 7, 9, 12). According to Steigrad (9), and others (2, 3, 5, 6, 10), respiratory and feeding difficulties such as cyanosis, malnutrition, sternal retraction, pneumonia, or even death from suffocation may therefore occur.

A number of procedures have been utilized in treating glossoptosis of the tongue. Robin (6) utilized an intraoral supporting device called a monobloc. Goldberg (3) has described the Eley and Farber extraoral brace technique as well as the traction method of Callister. Davis and Dunn (1) have reviewed the bottle guard which is used as an attachment on the ordinary nursing bottle.

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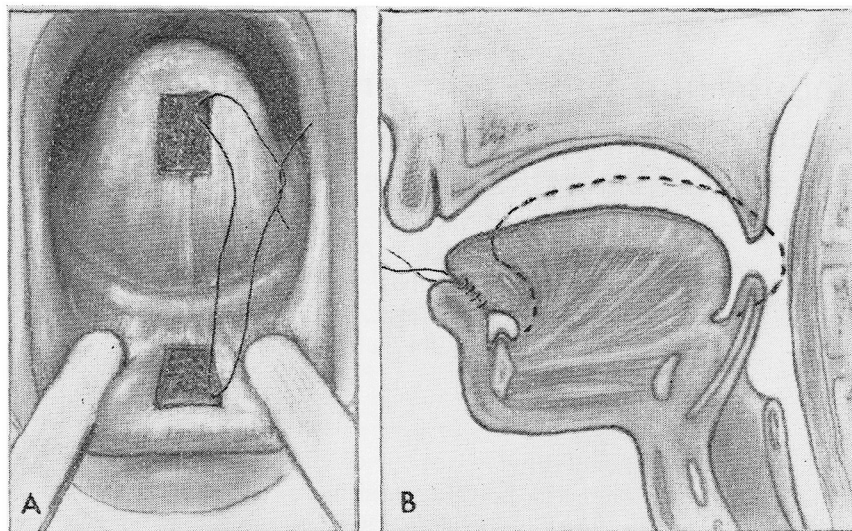


FIGURE 1. A; modification of the Beverly-Douglas operation. Under orotracheal anesthesia, a rectangular segment of mucosa is removed from the under surface of the anterior tongue. A similar segment of mucous membrane is removed from the lower central lip. Sutures of heavy white dacron or silk are inserted to anchor the musculature of the tongue forward to the musculature of the lip. B; sutures of 4-0 black silk or dacron are used in the mucosa. The operation will immediately bring the tongue forward and correct the obstruction, as shown by the dotted line.

In 1946, Beverly Douglas (2) developed an operation with the purpose of anatomically correcting the faulty position of the tongue. As shown in Figure 1, the operation is performed under orotracheal general anesthesia. A rectangular segment of mucous membrane is removed from the under surface of the tongue—approximately one centimeter transversely by 1.5 centimeters in the long axis of the tongue. Caution is exercised to avoid injury to Wharton's ducts. A similar segment of mucous membrane is removed from the buccal surface of the lower lip (Figure 1A). The musculature of the tongue is then anchored securely to the musculature of the lip using three or four sutures of 4-0 white silk or dacron. Sutures of 4-0 black silk are inserted to approximate the mucous membrane of the tongue with the mucous membrane of the lip (Figures 1B, 2B).

Our modification of the Douglas operation, as presented here, brings the tongue forward immediately and corrects the obstruction.

The surgical tongue-lip adhesion is allowed to remain until approximately six or seven months of age, when the lower central incisors erupt (Figure 2C). Under oro- or nasotracheal anesthesia, the tongue is freed from the lip. Interrupted sutures of catgut are used to close each defect. There should be no functional or cosmetic defect remaining (Figure 2D).

It is the purpose of this investigation to compare the tongue and lip

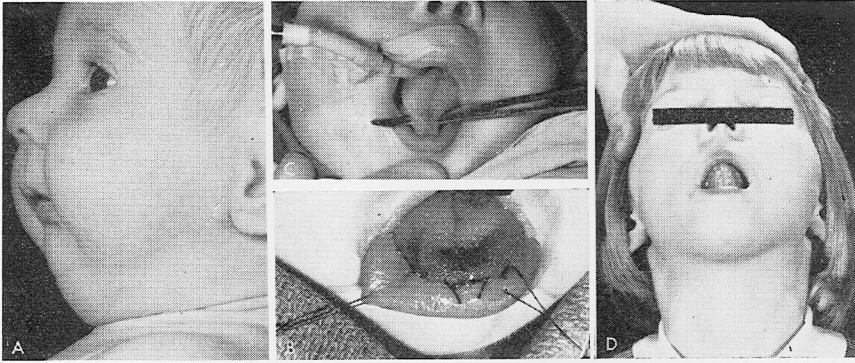


FIGURE 2. A, typical appearance of patient with micrognathia, or small lower jaw. Respiratory obstruction is due to backward displacement of the tongue, whether a cleft of the palate is present or not. The intermittent respiratory dyspnea, with sternal retraction, characteristic of the Pierre-Robin syndrome, may result in death unless corrected surgically. B, the tongue is brought forward to correct the glossoptosis and is anchored surgically to the lower lip. C, the tongue-lip adhesion is allowed to remain until the lower central incisor teeth erupt, usually at six to seven months. D, photograph of a 7-year-old girl showing perfect cosmetic and functional result of lips and tongue following tongue-lip adhesion operation performed in infancy.

mobility of a group of children who have had the Beverly-Douglas procedure with the tongue and lip mobility of a group of normal children.

Methods of Procedure

SUBJECTS. Five cleft palate Pierre-Robin Syndrome patients, aged $5\frac{1}{2}$ to 9 years, who had the Beverly-Douglas procedure in early infancy, were selected as the experimental group. The control group was composed of five children matched for age and with a negative history of oral-facial anomalies.

NONSPEECH AND SPEECH SAMPLE AND ANALYSIS. For assessing tongue and lip mobility, several tasks described by Westlake (11) were used. The following is a list of the activities, the time interval tested and the minimal number of repetitions suggested by Westlake.

Exercise I: The tongue was extended and retracted between the lip (five times in ten seconds).

Exercise II: The tongue was moved laterally from one corner of the lips to the opposite corner (ten times in ten seconds).

Exercise III: A rubber block was placed between the molars to stabilize the jaws and the tongue was elevated to the rugae (ten times in ten seconds).

Exercise IV: The upper and lower molars were in light contact and the lips were separated and brought together again (ten times in ten seconds).

Exercise V: The upper and lower molars were in light contact and the corners of the mouth were extended laterally and drawn medially so that the lips were puckered (five times in ten seconds).

The examiner also tested diadochokinetic rates for combined consonants and vowels, stressing sounds which required use of the two lips, the front of the tongue or the back of the tongue. The subject was required to repeat each combination for a period of ten seconds. The following consonant and vowel combinations were tested /PΛ/, /tΛ/, /kΛ/, /patakΛ/.

Each subject was also required to extend his tongue between his lips as far as possible. Then he was asked to move his tongue as high as possible towards his nose. Lastly, he had to move his tongue as far downward as possible towards his chin. Lateral cinefluorographic film of these movements was developed (4) and cine tracings and measurements were made of the frames showing the greatest exclusion.

Measurements were made from the base of the chin to the highest elevation of the tip of the tongue and from the point of tongue extension between the teeth to the lowest point towards the chin that the tongue moved.

Results

The randomization test for two independent samples was used to study the performance of the two groups. In this test, all possible 5-5 partitions of the 10 scores (5 treatment scores and 5 control scores) are considered. The probability of obtaining partitions more extreme than the observed is computed. This provides the significance level for the test (8). Of the ten measures used in the study, a significant difference was found on four tasks. The six measures in which there appeared no difference included movement of the tongue outward between the lips and back again; movement of the tongue from one corner of the mouth to the opposite corner; the lowest possible vertical movement of the tongue when it was extended between the lips. Also included in the group of tasks on which no significant differences were found was repetition of the consonants /tΛ/, /kΛ/, and /patakΛ/.

Three of the four exercises which were significantly different were at the 5 % level of confidence. These included elevation of the tongue to the rugae, the highest vertical movement of the tongue when it was extended between the lips; and lip protrusion-retraction. (In this exercise, the experimental group did significantly better than the control group.) One exercise, repetition of the consonant /pΛ/, was significant at the 1 % level of confidence.

It is interesting to note that although a significant difference between the two groups was evident on tongue elevation, there was not a significant difference between the two groups when speech was added as in the consonant /tΛ/.

Summary

The tongue and lip mobility of a group of 5 children with the Pierre-Robin Syndrome who had received the Beverly-Douglas plastic procedure was studied and compared with a group of normal children. Studies of tongue and lip activities, tests of diadochokinetic rates of the tongue and lips during speech, and cinefluorographic x-ray films and tracings were made of the tongue mobility of the two groups and results reported.

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