Prone Feeding of Infants with the Pierre Robin Syndrome

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Pierre Robin, a pediatric stomatologist in France, described a syndrome of glossoptosis and hypoplasia and retrusion of the mandible, with resulting difficulty in feeding and respiration (*7, 8, 9*). Though a cleft palate or insufficiency of the palate was frequently present, the difficulty presented by the infants with this syndrome was not due to the deformity of the palate, but due to the dorsal displacement of the tongue.

The development of a variety of therapeutic methods has resulted in a rapid and progressive improvement of oral function of these infants during the early postnatal period. These include the monobloc, an intraoral supporting device of Robin (*9*), the extraoral brace of Eley and Farber (*5*), the traction method of Callister (*2*), and the various surgical fixations of tongue to the anterior oral structures, notably those of Schukowsky (*12*) and of Douglas (*4*), which are designed to passively prevent the displacement of the mandible or the tongue. There have been, in addition, attempts to stimulate function of muscles related to the mandible and tongue by active exercise, such as the bottle guard of Davis and Dunn (*3*). The orthostatic feeding technique of Robin (*9*) can also be classified in this category.

For the past two years, infants with the Pierre Robin syndrome have successfully been treated by general maintenance of a prone position and by initiating prone feeding with a specially designed feeder described in some detail elsewhere (*13*).

Clinical Observation

DESCRIPTION OF SUBJECTS. The subjects in this study consisted of three girls and two boys, ranging in age from six days to nine months old at the time of admission (Table 1). In general, these infants exhibited various degrees of dehydration and malnutrition, depending upon their age and severity of dysphagia. The respiratory difficulty was manifested by the inspiratory retraction and intermittent episodes of cyanosis and stridor. In severe cases, the infants assumed an opisthotonic position as

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TABLE 1. Summary of additional information on cases.

Child	*M.D.*	*W.D.*	*T.E.*	*G.F.*	*P.G.*
Age at admission	1 month	6 days	5 months	9 months	7 months
Sex	female	male	male	female	female
Race	W	W	N	W	W
Family history	neg.	neg.	One sibling with Pierre Robin syndrome	neg.	neg.
Maternal age	41	19	34	25	29
Parity	5-3-0-2	1-0-0-1	17-9-1-5	3-0-0-3	4-0-0-4
Weight					
At birth	3.1 kg.	3.1 kg.	3.0 kg.	2.7 kg.	3.6 kg.
On admission	2.5 kg.	2.7 kg.	3.8 kg.	4.1 kg.	4.2 kg.
Cleft palate	yes	no	yes	no	yes
Previous surgery	none	none	Douglas, gastrostomy	none	none

a compensatory maneuver (Figure 1). The degree of respiratory difficulty varied also with the stage of alertness, and the ptosis of the tongue became particularly evident when they were at rest or during the gradation into the drowsy stage of sleep, predisposing these infants to continuous restlessness. Though the mandibles of these infants were hypoplastic and retruded, the degree of displacement of the tongue, in the dorsal direction, was much greater than to be expected from the micrognathia per se. The apposition of the tip of the tongue to the inner surface of the lower lip, as observed in normal infants at rest, was lost in the infants with the Pierre Robin syndrome. The tip of the tongue was drawn away from the lower lip and alveolar ridge, and the open-mouth was characteristic. Three of our five cases had clefts of the soft palate, extending in various degrees to the hard palate. These clinical findings were substantiated further by analysis of the laterally oriented roentgencephalometric films (Figure 2), from which quantitative measures of the degree of retrusion and displacement of the mandible and of the tongue-hyoid complex became possible (Figure 2).

It is of interest to note that one of our cases (P.G.) presented a trisomy of a minimum sized chromosome simulating those of the ⅋21–22 group, without the stigmata of Mongolism, though the other patients had normal chromosomal numbers and morphology.

Nursing Care and Treatment. The patients were admitted to the NIH Clinical Center Pediatric Unit, which provides nursing care for infants having problems of oral and pharyngeal impairment due to congenital malformations of the cranio-facial complex, for diagnostic evaluation and trial therapy. Since the greatest danger to these infants with the Pierre Robin syndrome was respiratory insufficiency, they were under extremely close observation. In the early stages of therapy, the nurses

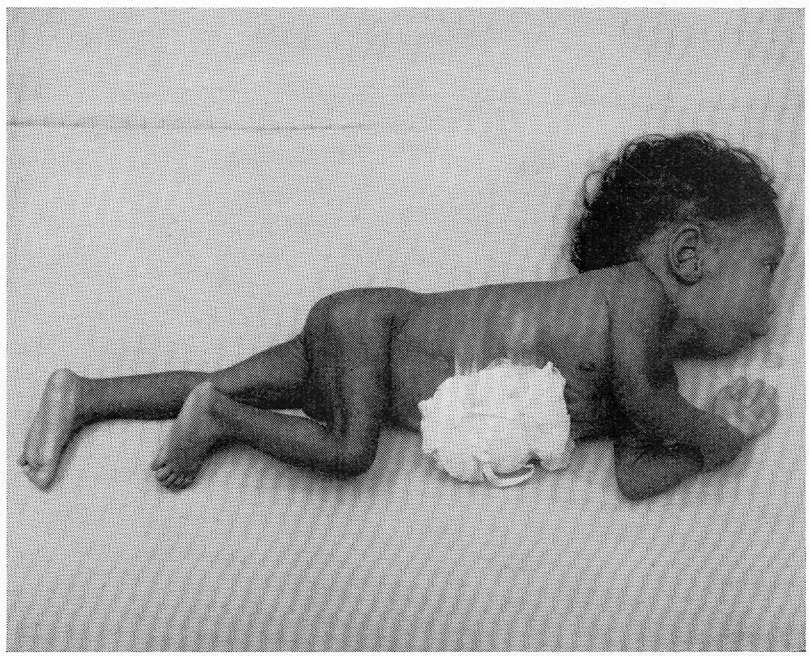

FIGURE 1. T. E. is a five-month-old Negro boy with Pierre Robin syndrome. He had been treated with Douglas procedures at age three, 10, and 14 days, respectively. When the tip of his tongue became necrotic, gastrostomy was performed at age six weeks. Note malnutrition and dehydration. The respiratory difficulty is manifested by the intercostal retraction and marked opisthotonus. The bandage was placed in the epigastric region following removal of the gastrostomy tube.

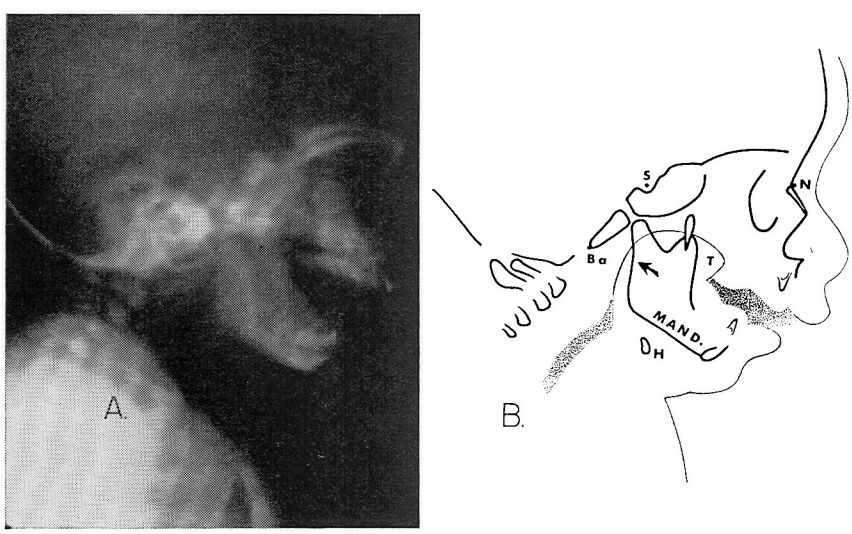

FIGURE 2. A (left), roentgenogram of T. E. taken in spine position. B (right), the tracing of the roentgenogram. Landmarks are: Nasion (N), Center of Sella turcica (S), Basion (Ba), Mandible (Mand), and Hyoid (H). Note hypoplastic mandible. The tip of the tongue is displaced dorsally and the tongue is obstructing the pharynx. The extent of glossoptosis is far greater than one would expect from this degree of micrognathia.

needed to be alert for signs of respiratory distress, even when the infant was in a prone position. Since the disability of oral functions was in part manifested by the failure of swallowing, suctioning equipment needed to be readily available to alleviate the accumulation of secretions and mucus in the pharynx.

Upon feeding, the infants were placed in a prone position with cervical extension. With less vigorous infants, or with those in the younger age group in which the neurological maturity of head-neck stabilization was less established, the extended head was supported at the forehead with the nurse's hand, or the face was turned to the side maintaining the extension at the neck (Figure 3). The initial feeding of the newborn infant

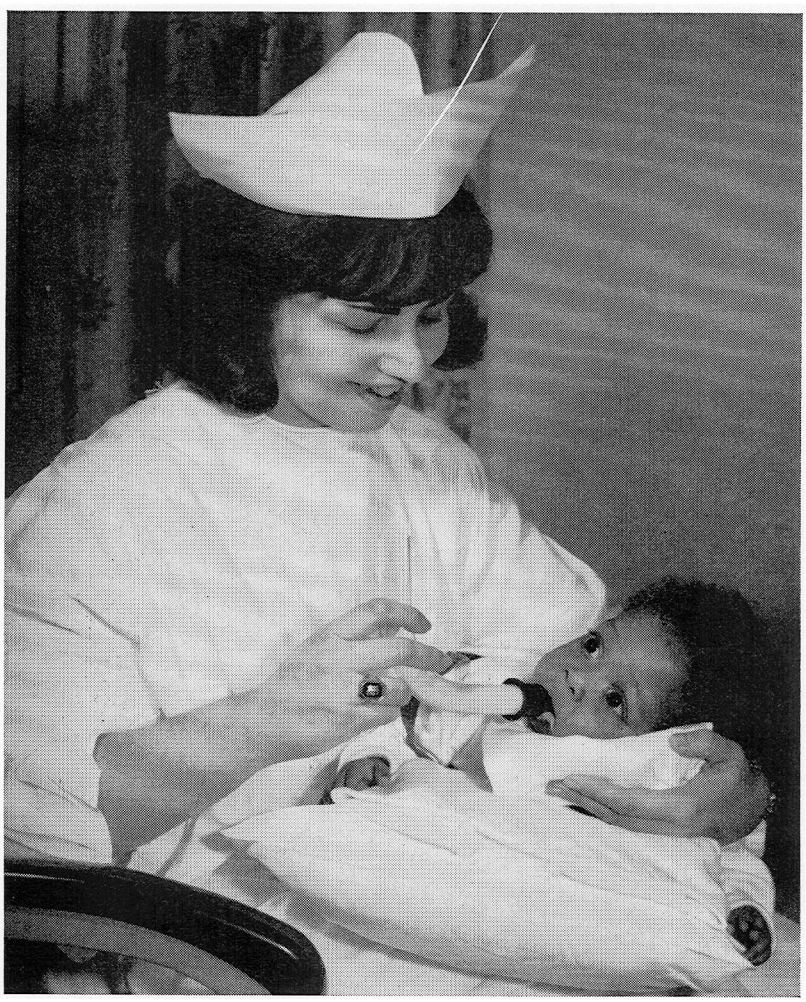

FIGURE 3. Feeding of T. E. in prone position using curved bottle with lamb nipple attached. The patient is placed in a prone position. In this instance, the head is turned to the side and held in extension.

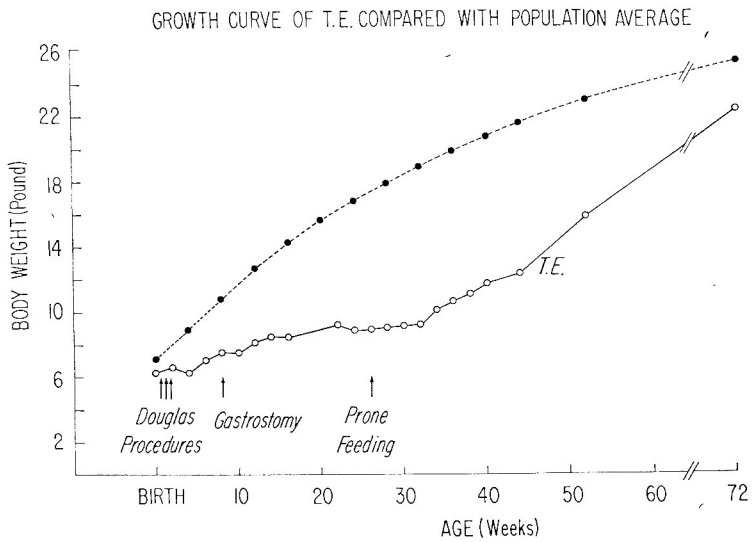

FIGURE 4. Growth chart of T. E. (solid line) compared with the normal population average (dotted line) (*10*). The approximate dates of repeated Douglas procedures, gastrostomy, and initiation of prone feeding are indicated with arrows. Note a time lag following prone feeding previous to the steady gain in weight.

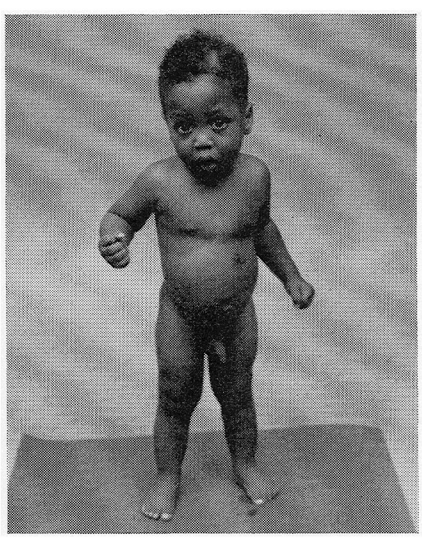

FIGURE 5. T. E. at age 72 weeks. His growth is essentially normal.

in the prone position caused less difficulty than that of older infants since a feeding pattern had not been established. Considerable patience was required of the staff during the first few days of the new feeding procedure. The staff assigned to feed infants in this position needed to be aware of the fact that older infants are quite active during the procedure.

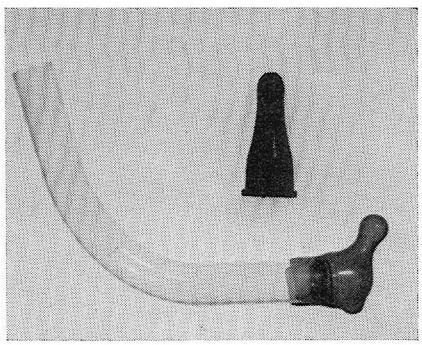

FIGURE 6. Curved glass tube feeder, to which 'ducky' nipple is attached. 'Lamb' nipple is shown separately.

Therefore, all equipment must be assembled and readily available before positioning the infant. The prescribed formula, the curved glass feeder with the appropriate nipple attached, and any solid food ordered for an older infant should be placed on a table within easy reach of the nurse. A 'ducky' latex nipple was used to feed infants without a cleft palate, whereas a 'lamb' nipple was used for those with a cleft (Figure 6). The abruptness of change in suckle and swallow performances during prone feeding was clinically evident. The improvement in oral intake and feeding schedule was associated with a change in turgor of the skin and subcutaneous tissues and with signs of feeding satisfaction prior to the onset of steady gain in weight. The sign of feeding satisfaction and acquisition of sleep uninterrupted by glossoptosis were particularly significant, for the infants with the Pierre Robin syndrome suffer from continued restlessness due to pharyngeal obstruction as much as they do from malnutrition. The frequent incidences of aspiration encountered in supine feeding of these infants were successfully avoided by the prone feeding method. There was much individual variation in the adaptation to the prone feeding and to the acquisition of progressive gain in weight. The degree of chronicity of nutritional and oral deprivation appeared to be the dominant factor in this variation. After the definite signs of improvement and steady gain in weight were noted, the parents were instructed in the general care of these handicapped infants and in the technique of feeding in a prone position prior to the discharge from the Clinical Center.

Information regarding one subject, T. E., is presented in figures 1 through 5.

Discussion

Previously proposed methods of treatment of infants with the Pierre Robin syndrome, either by passive stabilization of the mandible and tongue or by active exercise of the oral structures, have met with fair success. The method of feeding described in this article is simple to prac-

tice, and eliminates the difficulty and trauma encountered in the surgical procedures and orthopedic practice. Once the feeding pattern is established, these infants show rapid and progressive improvement of the oral and pharyngeal function. Thus, the necessary period of maintaining the prone feeding procedure is relatively short. The experience of this medical and nursing group in the care of infants with the Pierre Robin syndrome also confirmed the observation of Pierre Robin that these infants develop compensatory posture of tongue and related structures so as to maintain adequate pharyngeal airway in the variety of head-neck postures.

The basic mechanisms contributing to the change in oral and pharyngeal performances in prone feeding are not well understood. Feeding an infant in prone position requires extension of the infant's neck. This cervical extension is the familiar maneuver used to facilitate swallow in paralytic dysphagia (*11*), to compensate for dyspnea, or to facilitate loud speech or shouting. The radiographic observation of the effect of head-neck posture upon the pharyngeal airway revealed that the extension at the neck was consistently associated with the opening of the airway by the ventral displacement of the anterior cervical structures (*12*). In general, the feeding of infants in a prone position is the universal method of feeding among the mammals. This physiological method may also be applied to the infants with a variety of neurological impairments and to the premature infants in which the cervical musculature and head-neck stabilization are less developed.

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