Recurrent Aspiration Pneumonitis in A Cleft Palate Child With Hamartoma of the Tongue

ROBERT J. DEMUTH, M.D.
DONNELL F. JOHNS, Ph.D.
Portland, Oregon, 97201

A patient with unilateral cleft lip and palate and a history of *recurrent aspiration pneumonitis* caused by a *benign hamartoma* at the base of the tongue is presented.

KEY WORDS: Cleft palate, aspiration pneumonitis, hamartoma, tongue

Introduction

Problems with deglutition are common in neonates with congenital abnormalities involving the craniofacial area, i.e., Pierre-Robin Anomaly, choanal atresia, or neurogenic abnormalities secondary to prematurity. These problems are often related to abnormalities in the relationships among the tongue, mandible, palate, and pharynx. The feeding problems associated with cleft lip and palate, although frequently encountered, are usually solved by revising the size, shape, or orifice of the feeding nipple and by positioning of the patient during feeding. Infants with clefts do not usually have swallowing problems per se. The cleft patient presented here, demonstrates feeding problems which were not solved by the usual simple techniques because the true cause of the difficulty, hamartoma at the base of the tongue, was not discovered until a palatoplasty was performed.

Case Report

This male Latin-American child was first seen at the age of two months by the Plastic Surgery team for evaluation of a unilateral cleft lip and palate. He was a full-term infant, spontaneously delivered without complications. There was no history of problems during the mother's pregnancy. Postnatal care was supervised by a pediatrician who was knowledgeable about the care of children with cleft lips and palates. Feeding and regurgitation problems were reported during the initial examination. The child was below the tenth percentile in weight and below the fifth percentile in length (NCHS) when first seen. The parents were given the usual advice and instructions regarding nipple change and positioning procedures during and after feeding.

At three months of age, a left cheiloplasty was performed under general endotracheal anesthesia without complication. At age six, nine, and eleven months, the patient was admitted to a hospital for recurrent pneumonitis. Each time, treatment consisted of respiratory care and systemic antibiotics. A barium swallow was performed at 11 months of age. Tracheal aspiration was confirmed, but no other abnormalities were noted (Figure 1). Consultations with other specialists at this time led to intensive dietary consultation, home visits by a visiting nurse, and focus on bilingualism, even though the mother communicated well and was thought to be reliable. Failure to thrive was evidenced by the fact that the child's height and weight at 11 months were both below the fifth percentile. This represented a more serious problem than the usual moderately impaired weight gain which occurs in cleft palate patients (Ranalli and Mazaheri, 1975).

At age 18 months, the patient's pulmonary

Dr. Demuth is affiliated with the Division of Plastic and Reconstructive Surgery, University of Oregon Health Sciences Center, Portland, Oregon 97201. Dr. Johns is affiliated with the Division of Plastic Surgery, University of Texas, Southwestern Medical School, Dallas, Texas 75235.

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status was acceptable, and it was decided to perform a palatoplasty under general endotracheal anesthesia. The endotracheal tube was placed without difficulty by an experienced pediatric anesthesiologist. Upon insertion of the Dingman-Dott mouth retractor, a pedunculated tumor was noted on the base of the tongue in the area of the foramen cecum (Figure 2). The tumor was cored out after frozen section histologic interpretation of the incisional biopsy suggested that it was benign. A push-back palatoplasty was completed without complication. The patient's hospitalization was extended to ten days because of postoperative fever and pneumonitis. Following discharge, the patient did well with no subsequent episodes of aspiration, pneumonia, or feeding problems. Permanent microscopic sections of the tumor demonstrated normal nerve elements, smooth muscle, skeletal muscle, and glands in abnormal proportions for the anatomic area inspected. The lesion was thought to be a benign hamartoma (Figure 3).

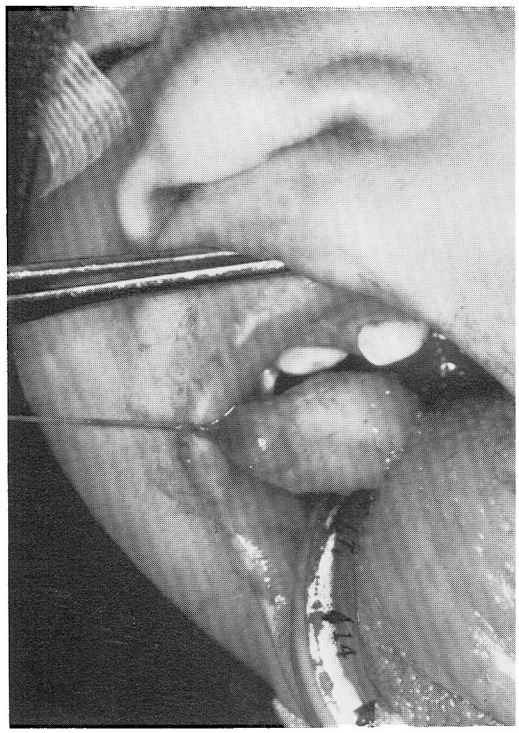

FIGURE 2. Base of the tongue pedunculated tumor exposed prior to removal at age 18 months during palatoplasty.

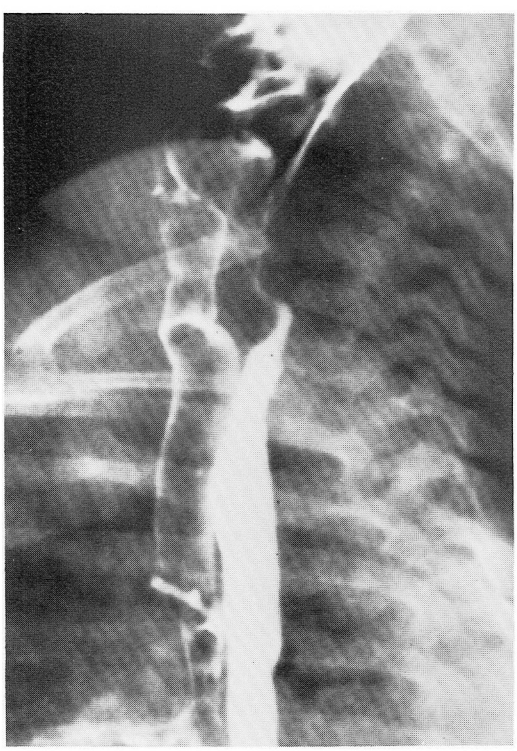

FIGURE 1. Barium swallow at age 11 months showing tracheal outline from aspiration of contrast material.

Six months following surgery, the patient weighed 27 pounds (greater than the 40th percentile) and was thriving and doing well. Postoperative barium swallow (Figure 4—right) confirmed the absence of aspiration. Physical examination demonstrated no evidence of tumor recurrence.

Discussion

Cleft lip and palate deformities are the most common malformations of the aerodigestive tract producing swallowing difficulties in childhood. These problems are usually solved without great difficulty. Other craniofacial abnormalities less frequent in incidence create similar aspiration problems because of anatomic or physiologic deformities of the palate or pharynx, i.e., Pierre-Robin Sequence, Stickler syndrome, Crouzon deformity, Apert deformity, or Treacher-Collins syndrome (Gilbert and Opitz, 1976).

Other causes of neonatal swallowing problems include rare space occupying lesions such as heterotopic brain tissue (Shapiro and Mix,

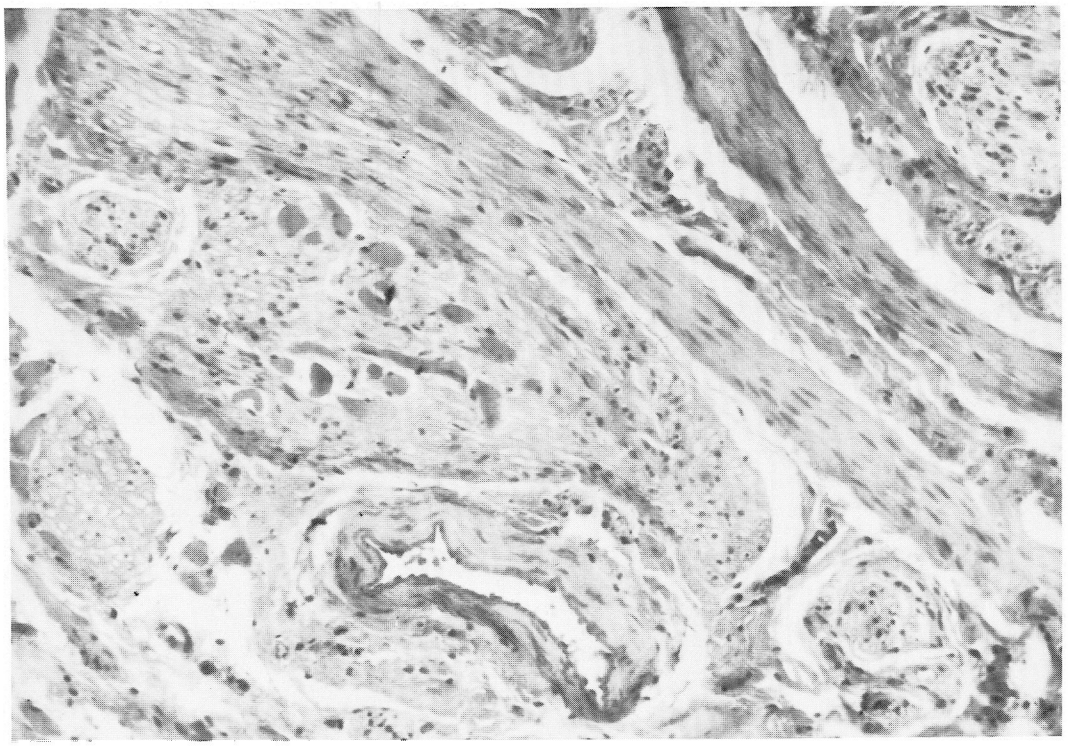

FIGURE 3. Photomicrograph of tongue tumor showing excessive amounts of variable type connective tissue including neural tissue, smooth, and striated muscle, blood vessels interpreted as benign hamartoma.

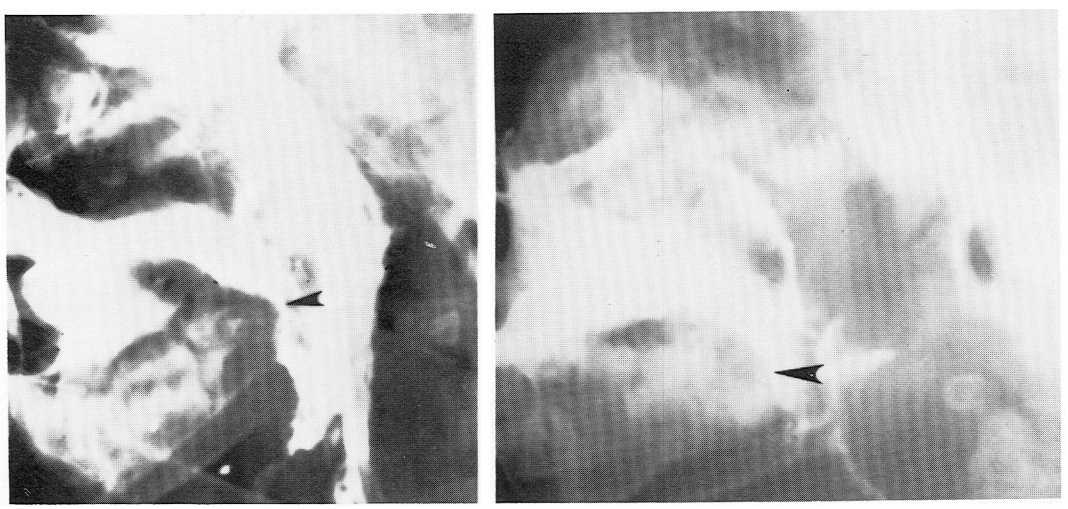

FIGURE 4. (Left) Barium swallow re-evaluation of study done at age 11 months with arrow suggesting tumor at base of the tongue. (Right) Follow-up barium swallow at age 24 months, 6 months following tumor removal showing smooth tongue surface without tumor.

1968), nasal gliomas (Lofgren, 1963), neurogenic hamartoma (Gallo and Smith, 1975), lingual cysts (Lofgren, 1963), thyroglossal duct cysts, dermoids, lingual thyroid or angiomas.

Lack of palatopharyngeal coordination in both cleft and non-cleft patients without anatomic or space occupying defects produce difficulties in swallowing, regurgitation into the nasopharynx, and tracheal aspiration. A

transient form of palatopharyngeal dysfunction is often seen in the newborn infant and presumably represents delayed maturation and coordination in the swallowing mechanism (Frank and Gatewood, 1966). This is common in the premature infant because palatopharyngeal coordination is not fully developed. These disturbances of swallowing in children with dystonic syndromes are often undiagnosed until later in life when failure of growth and development become more obvious.

Studies of normal growth, development, and function of the aerodigestive tract have not been extensively done (Maue-Dickson, 1979). Studies by Crelin and associates seem to best delineate the transition which occurs in infancy from obligatory nasal breathing to oral breathing (Sasaki, et al., 1977, and Crelin, 1976). In adult mammals and the human infant, the larynx normally can be elevated so that the epiglottis lines up behind the soft palate. This in combination with the medial movement of the posterolateral walls of the nasopharynx creates a tight seal between the naso- and oropharynx separating the olfactory respiratory pathway from the oral cavity. With the soft palate locked into place by the epiglottis and the larynx locked into place by the posterolateral walls of the pharynx, the suckling human newborn infant can breathe while swallowing liquid. Liquid passes through the isthmus faucium to the pharynx along both sides of the elevated larynx (Figure 5). The ability to swallow liquids and breathe simultaneously is lost in humans by the end of the first six months.

We postulate that, when the child presented here was intubated at age three months for the cleft lip repair, the tumor was either not seen by the anesthesiologist or was not large enough to be of significance. Recurrent aspiration pneumonia may have been caused by tumor growth or could have been because the child could not use the epiglottis adequately and could not elevate the larynx to control the flow of liquid passing around it so that simultaneous feeding and nose breathing

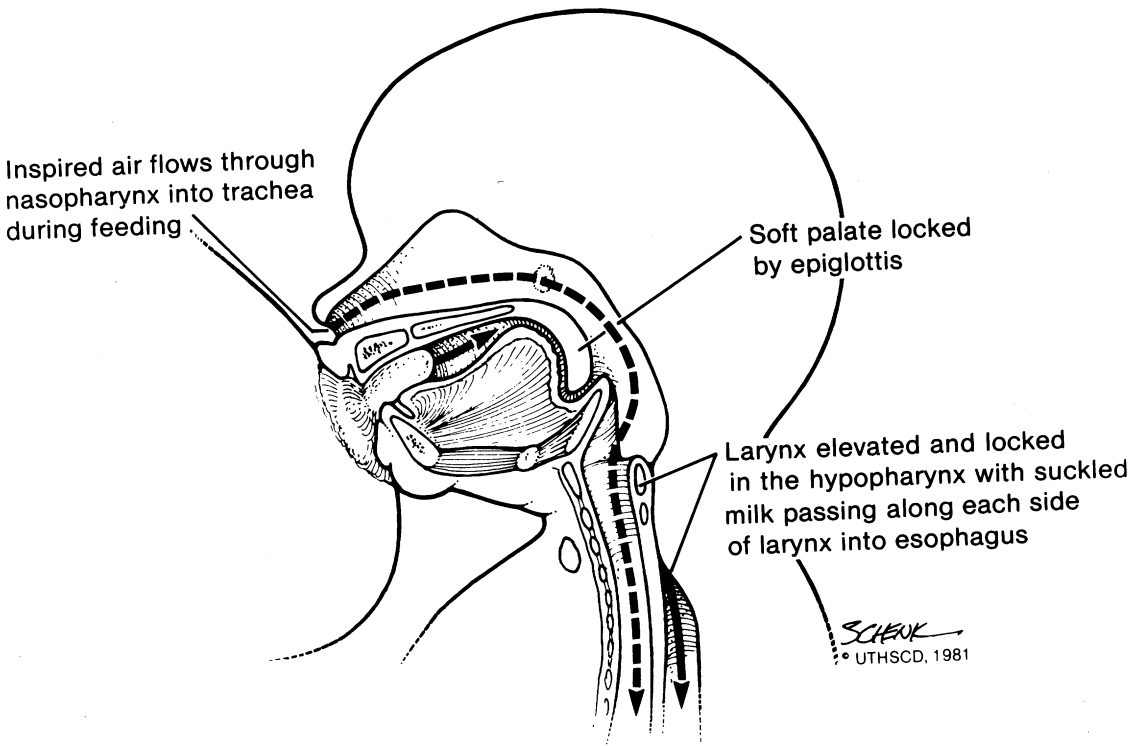

FIGURE 5. Diagram of normal infant feeding showing position of epiglottis with relation to palate allowing simultaneous nasal breathing and swallowing.

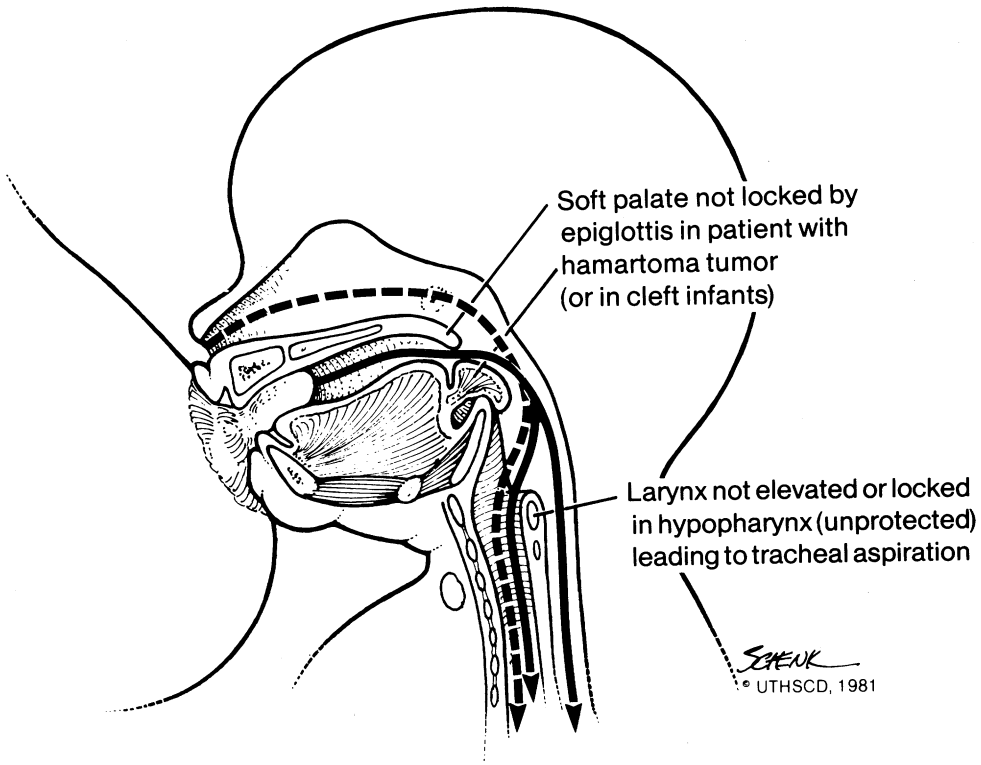

FIGURE 6. Diagram showing tumor at the base of the tongue influencing tracheal aspiration and interfering with normal neonatal swallowing.

could be carried out (Figure 6). After a review of the barium swallow performed at 11 months of age, a small filling defect on the surface of the base of the tongue at the site of the tumor suggested that it was present at that time (Figure 4—left).

All of the efforts of the multi-disciplinary team, including the plastic surgeon, pediatrician, pediatric dietician, bilingual social worker, and visiting nurses, and two direct laryngoscopic endotracheal tube placements did not lead to the correct diagnosis in this patient until the tumor was discovered at the time of palatoplasty. Because feeding difficulties in the cleft palate patient do not usually produce major problems in growth and development, this case report is submitted as a reminder to all of us to increase our index of suspicion when things do not go well with feeding in the patient born with cleft palate.

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