The Laryngoscope

Swallowing Function After Laryngeal Cleft Repair: More Than Just Fixing the Cleft

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Objectives/Hypothesis: To evaluate and describe the swallowing function in children after laryngeal cleft repair.

Study Design: Ten-year (2002–2012) retrospective chart review. Setting: Academic tertiary care pediatric otolaryngology practice.

Methods: Records of 60 children who had surgical repair of laryngeal cleft (ages 2 weeks–14 years) and postoperative functional endoscopic evaluation of swallowing or videofluoroscopic swallow studies were examined retrospectively.

Results: Twenty-nine children had one postoperative swallow evaluation, 19 children had two, 4 children had three, 5 children had four, and 3 children had five. Median time to the first evaluation was 10.8 weeks (interquartile range [IQR]: 36.5, 231). On the final swallow evaluation, 34 (57%) children demonstrated normal swallowing parameters, 12 (20%) children showed penetration, and 14 (23%) children showed aspiration. Forty-three (72%) children were able to take everything by mouth normally or with minor behavioral modifications, 11 (18%) children required thickened fluids, and six (10%) children were kept nil per os (NPO). Mean improvement on the penetration-aspiration (pen-asp) scale was 2.13. On multivariable analysis, neurodevelopmental issues and gastronomy tube use were associated with the need for NPO status.

Conclusion: Despite a high rate of surgical success, a substantial minority of children have persistent swallowing dysfunction after laryngeal cleft repair. Swallowing dysfunction after repair is multifactorial and arises from concomitant neurologic, anatomic, or other comorbidities that contribute to oropharyngeal and pharyngeal dysphagia. Based on our results, we recommend a testing schedule for postoperative swallowing evaluations after cleft repair.

Key Words: Laryngeal cleft, swallowing, FEES, VSS, VFSS.

Level of Evidence: 4.

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INTRODUCTION

Laryngeal cleft is a rare congenital anomaly in which there is incomplete separation of the aerodigestive tract due to a midline defect in the common wall between the laryngotracheal and esophageal lumens. Clefts range from deep interarytenoid notches to those that extend below the vocal cords, through the cricoid, and into the trachea. Benjamin and Inglis[1] developed the most commonly used classification scheme for laryngeal clefts, and cleft grade correlates with symptom intensity.[2] Common presenting symptoms include hoarseness, stridor, chronic cough, aspiration with feeding, recurrent pneumonia, and respiratory distress.[3] Diagnosis of a laryngeal cleft requires a high index of suspicion—and typically direct laryngoscopy.

Small clefts (type I and II) can remain clinically silent, causing no symptoms at all. Even when symptoms are present, almost half of children with type I and type II clefts can be treated conservatively with medical and feeding modifications.[4] The remainder of small clefts and virtually all type III and IV clefts require surgery to close the cleft and to prevent aspiration and life-threatening pulmonary compromise. Studies of patients with laryngeal clefts have focused largely on indication for surgery, surgical methodology, and surgical success rates.[4–7] A detailed characterization of swallowing function in children who have had laryngeal cleft repair is missing from the literature.

Although the laryngeal cleft itself can lead to aspiration through incomplete separation of the respiratory and digestive tracts, dysfunctional swallowing in children with clefts is often multifactorial. Laryngeal clefts can be associated with other airway abnormalities or syndromes with craniofacial, aerodigestive, or neurological effects that contribute to oral motor and pharyngeal swallowing dysfunction. Furthermore, even otherwise normal children with laryngeal clefts may require prolonged periods of gastronomy tube (g-tube) feeding and nil per os (NPO) status, during which the complex oral

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and oropharyngeal motor patterns required for normal feeding and swallowing may regress or fail to develop. Additionally, decreased oral feeding over a prolonged time period is associated with the development of oral aversion and the lack of progression with oral feeding.[8] With these points in mind, we examined the postoperative swallowing function of children in our practice who underwent laryngeal cleft repair.

MATERIALS AND METHODS

Study approval was granted by the Cincinnati Children's Hospital Medical Center (CCHMC, Cincinnati, OH) Institutional Review Board (study number 2012–2035). Ten years (July 2002–June 2012) of records from the Otolaryngology–Head and Neck Surgery Clinic and the Aerodigestive and Esophageal Center (ADEC) were searched for children less than 18 years of age with a diagnosis of laryngeal cleft confirmed by direct laryngoscopy in the operating room. All children who underwent surgical repair of their cleft and who had postoperative evaluation of swallowing were included in our analysis.

We collected information on demographics, Benjamin and Inglis cleft grade (3 "deep interarytenoid groove" patients were included in the type I group), type of repair, other airway findings, neurologic comorbidities, syndromic associations, and swallowing outcomes (see Supplementary Information for a complete list).

Cleft repair was decided upon by the interdisciplinary aerodigestive and esophageal center group. Symptoms such as recurrent pneumonia and choking during feeds, as well as data from bronchioalveolar lavage, computed tomography, or preoperative swallow studies were used to guide this decision. Closure techniques were endoscopic or open, layered or simple, and with or without cartilage or periosteal graft—at the discretion of the operating surgeon. Revision was performed if breakdown was seen on surveillance endoscopy and if symptoms or laboratory data suggested continued aspiration.

Functional endoscopic evaluation of swallowing (FEES) or videofluoroscopic swallowing study (VSS/VFSS) was performed at the discretion of the ADEC physicians and speech pathologists. The airway protection ability of each child was rated using the penetration-aspiration scale (pen-asp scale) previously described.[9] Children are scored on a scale of 1 to 8: 1 = normal; 2 to 5 = penetration; 6 to 8 = aspiration. Occasionally, children with tracheostomies were evaluated with dye testing. These were graded in a binary fashion (aspiration or no aspiration) by the presence or absence of dye in the tracheal aspirate. The recommendations of the speech pathologist were grouped as follows: 1) safe for oral feeding with all consistencies; 2) safe for oral feeding with all consistencies with minor feeding modifications such as slow bolus presentation, limited volume boluses, or positional adaptations; 3) safe for oral feeding with altered fluid viscosity; and 4) unsafe for oral feeding. In revision cases, only swallowing evaluations performed after the last revision for a persistent cleft or fistula that was causing aspiration were considered. Standard clinical signs of aspiration (or the resolution thereof), such as choking or coughing with feeds, recurrent respiratory infections, and parental suspicion served as indications for repeated postoperative swallow evaluations.

Descriptive statistics including frequencies and proportions or medians with interquartile ranges (IQR) were calculated on all variables. Chi-square or Fisher's exact tests were used to examine relationships between categorical predictors and feeding recommendations. Logistic regression was used to examine multivariable relationships between predictors and swallowing outcomes and feeding modifications. An alpha level of 0.05 was considered significant. SAS (Version 9.3, Cary, NC) was used to conduct the analysis.

RESULTS

We found 115 children with laryngeal clefts seen in our practice over the study period. Of these, 89 children had surgery to repair the cleft and 60 children had postoperative swallowing evaluation (35 [58%] males and 25 [42%] females). Forty-four patients had one surgery to repair the cleft, 10 patients had one revision, and six patients had two revisions. The median ages at the first and last surgery were 27.5 and 37.1 months, respectively (ranges 1 week–14.1 years and 2 weeks–18.11 years, respectively). There were 21 (35%) grade I clefts, 21 (35%) grade II clefts, 17 (28%) grade III clefts, and one (2%) grade IV cleft. Twenty-nine patients who underwent surgery did not have a postoperative swallow evaluation, either due to an extremely encouraging clinical picture or because they returned to their referring center for ongoing care.

Twenty-nine patients had one postoperative swallowing evaluation; 19 patients had two; four patients had three; five patients had four; and three patients had five evaluations. Of these 114 studies, 28 (24.5%) studies were FEES; 77 (67.5%) studies were VFSS; and nine (7.9%) studies were clinical/dye tests in patients with tracheotomy. The median time between surgery and first postoperative swallowing evaluation was 9.9 weeks (IQR: 1.7, 6.1 years). In those children who had multiple swallow studies (n = 31), the median time between surgery and the last swallow evaluation was 8 months (IQR: 1.9, 28.4 months).

On final swallowing evaluation, 34 children had normal swallowing parameters, 12 demonstrated some degree of penetration, and 14 demonstrated some degree of aspiration (Fig. 1). Forty-three children were ultimately able to take all consistencies by mouth with minor or no feeding modifications, 11 children required modified consistencies, and six children remained unsafe for oral intake (Fig. 2).

Of the six children who were unsafe for oral intake, two children had a persistent cleft or fistula through which they aspirated. One child had a recurrent type II cleft, which is scheduled for revision. The second child had a type III cleft that was repaired but had a small tracheoesophageal fistula near the apex of the repair. This patient visited our center, did not remain under our care to have this fistula addressed, and was thus lost to follow-up. Interestingly, three children with normal swallowing parameters also had some degree of persistent clefting on follow-up direct laryngoscopy. These children were not revised because of their normal swallowing parameters.

Criteria for proceeding to cleft repair without a preoperative swallow evaluation at our institution included type III or IV cleft, strongly suggestive clinical symptoms of aspiration, or a swallow evaluation at the patient's home institution (which may not have been scored by our speech pathologists). Given those limitations, 41 children in the current study had preoperative

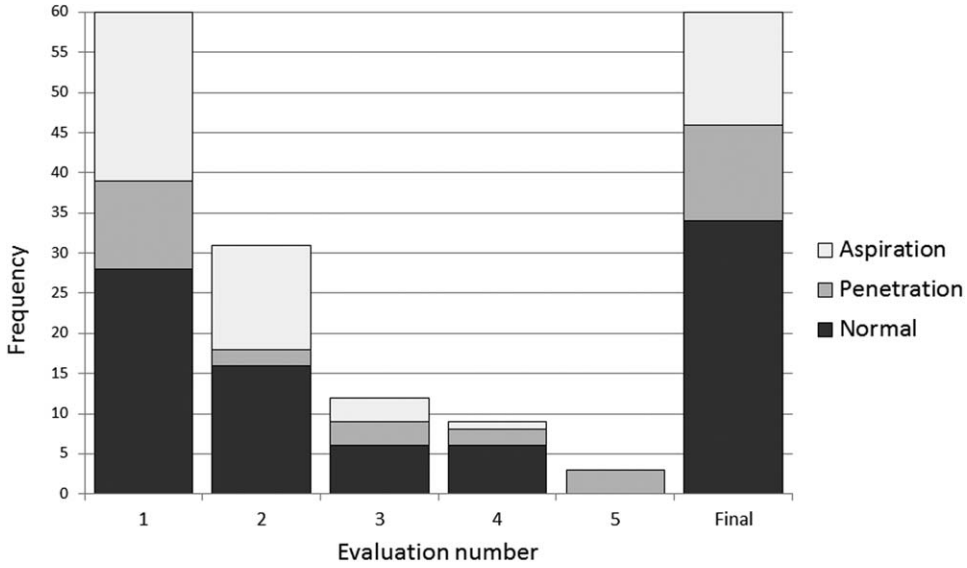

Fig. 1. Results of swallowing evaluations of children after laryngeal cleft repair. The results describing the degree of airway protection seen during swallowing evaluations after laryngeal cleft repair are shown. Proportions of children falling into each category are shown.

swallow evaluations that we could score. Preoperative and postoperative evaluations are compared in Table I. Children with normal swallow studies demonstrated clinical symptoms that warranted repair of the cleft in the opinion of the treating physician. The mean score on the pen-asp scale decreased from 5.33 to 3.2 ($P < 0.05$, paired t test).

When we examined potential predictors of feeding modifications, there was no association detected between cleft grade and final feeding recommendations (Fig. 3). We considered other factors that might influence the ability to gain functional swallowing, such as g-tube use prior to surgery, neurologic comorbidities, syndromic associations, age at repair, method of repair (endoscopic vs. open), and additional airway findings. Upon multivariable analysis, the presence of neurologic comorbidities (Coloboma Heart abnormalities, choanal Atresia, growth Retardation, Genitourinary abnormalities, and Ear abnormalities (CHARGE) syndrome, Opitz syndrome, trisomy 21, cerebral palsy, and global developmental delay) and g-tube use predicted the need to modify diet (minor feeding modifications, thickeners, or NPO status). Children with neurodevelopmental issues had 6 times greater odds of having modified feeding recommendations compared to those without neurodevelopmental issues (95% CI 1.4–26.6). Those with g-tubes had 3.6 times greater odds of diet modification (95% CI: 1.02–13.0). Although feeding modifications are a restriction, they do not represent the same lifestyle impact and burden of care that the use of thickeners and NPO status represent. Accordingly, we separated children into two groups: those children who could take a normal diet without modifications or with slight modifications and those children who required the use of thickeners or NPO status. When these alternative groups were considered, only neurodevelopmental issues remained as a predictor of the need for thickeners or NPO status (OR: 5.8, 95% CI: 1.5–22.7).

Taking those 43 children who were ultimately cleared for per os (PO) intake of all consistencies with no or only minor behavioral modifications, 20 (45%) of the children

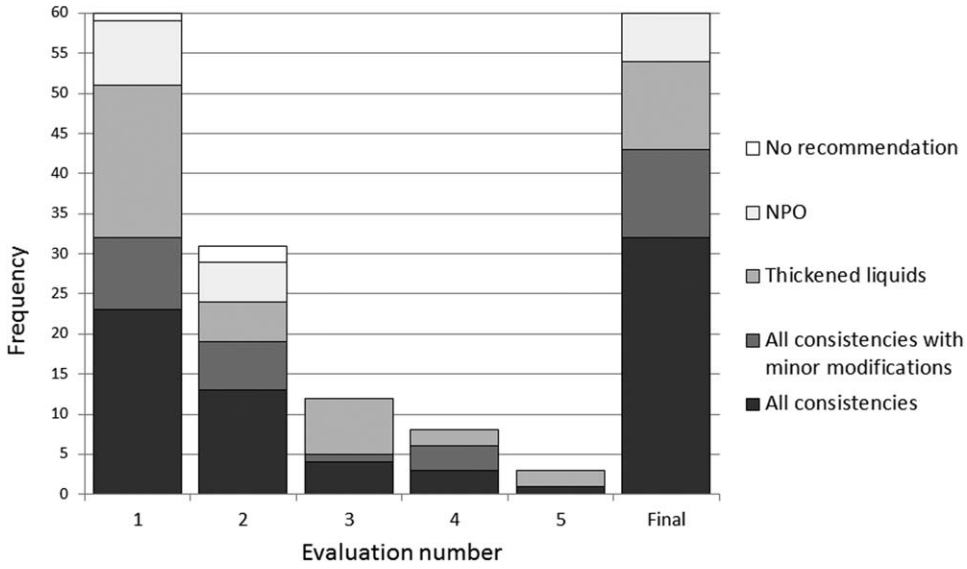

Fig. 2. Speech pathologist's recommendations following swallowing evaluations after laryngeal cleft repair. The recommendations regarding per os intake based on the swallowing evaluations after laryngeal cleft surgery are described, and proportions of children falling into each category are shown. In rare instances, the child was evaluated using the penetration-aspiration scale, but no formal recommendation by the speech pathologist was recorded in the chart on how to proceed with feeding. These studies are represented as "no recommendation."

TABLE I.
Comparison of Preoperative and Postoperative Swallow Studies.

	Normal	Penetration	Aspiration
Preoperative	13	2	26
Postoperative	25	7	9

had evaluations within the first 3 months after their final surgery that demonstrated safety for intake of all consistencies (Fig. 4). Cumulatively, 32 (74%) children were cleared for PO intake of all consistencies within the first year, and 11 children took more than 1 year. Of those individuals who took more than 2 years to be cleared for all consistencies (n = 7), two patients did not have their first evaluation until more than 5 years after surgery; however, the remaining patients had regular swallow studies at roughly 1-year intervals until they were cleared for all consistencies. Thus, a small number of individuals (in this case 5 out of 43 [11%]) can truly take many months to achieve normal swallowing after cleft repair.

DISCUSSION

We present the first detailed analysis of swallowing function after laryngeal cleft repair. Thirty-four (57%) children ultimately achieved normal swallowing as confirmed by FEES, VFSS, or dye testing; and 43 (72%) children were cleared for a normal diet with no or only minor feeding modifications. Some children who demonstrated penetration or aspiration did so only under certain circumstances such as rapid chain swallows or with large volumes. These children can often take thin liquids safely with adequate pacing of intake or with changes in positioning. We feel that there is a natural distinction between children who are given a final recommendation for normal PO diet or normal diet with minor feeding modifications and those children who require the use of thickened liquids or are kept NPO. Both NPO status and the need for thickened fluids present a large impact on quality of life for children and their caretakers, while minor feeding modifications are easily adopted, develop naturally, or are sometimes ignored—essentially placing the child on a normal PO diet without modifications.

We anticipated that more severe cleft grade, later age at surgical repair, use of a g-tube, method of repair, and the presence of other medical comorbidities or aerodigestive findings would influence the chance of acquiring normal swallowing. Only g-tube use and neurodevelopmental comorbidities predicted the need for feeding modifications; and neurodevelopmental compromise was the strongest predictor. That neurodevelopmental abnormalities predict the need for NPO status or the use of thickeners is expected. The relationship between neurodevelopmental disorders and dysphagia has been extensively studied.[10–12] We included children with Trisomy 21, CHARGE syndrome, and Opitz syndrome in our group of children with neurodevelopmental disorders. Despite the fact that these syndromes may have comparatively mild neurodevelopmental defects compared to cerebral palsy or severe global developmental delay, a significant portion of these children had difficulty gaining normal swallowing after cleft repair. Thus, the complex oral and oropharyngeal motor patterns of safe swallowing in these individuals may be sensitive to moderate perturbations brought about by laryngeal surgery and developmental delay. Additionally, it is difficult to separate the effects of neurodevelopmental delay from the concomitant craniofacial abnormalities that are present in some of these children. The true picture of dysphagia in these cases is likely a combination of neurologic, anatomic, and medical factors.[13]

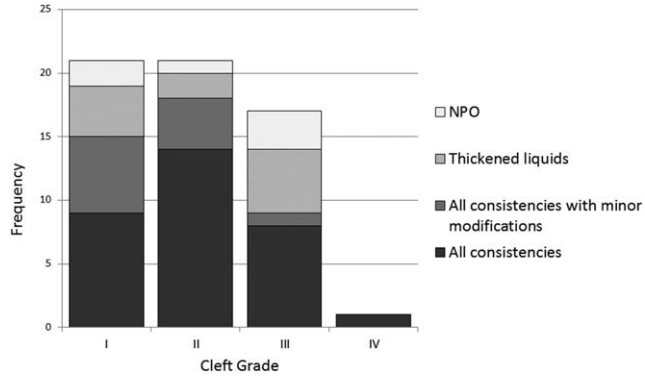

Fig. 3. Final speech pathologist recommendation shown with respect to initial cleft grade.

It is not surprising that g-tube use might predict worse swallowing function postoperatively. Many children with type I or II clefts can partially or entirely compensate for the cleft to prevent aspiration. If a g-tube is needed, it might indicate that the child had worse compensatory mechanisms to begin with. Additionally, evidence suggests a critical window of neuromotor development for the coordination of swallowing and breathing, which can be disrupted if the infant engages in nonnutrative sucking alone.[14] Thus, reliance on a g-tube early in life might impair development and hinder postrepair swallowing. In our study, even children who were ultimately cleared for a normal diet with no or minor modifications demonstrated a high rate of oral and oropharyngeal dyscoordination, highlighting the sensitivity of these motor patterns to disruption.

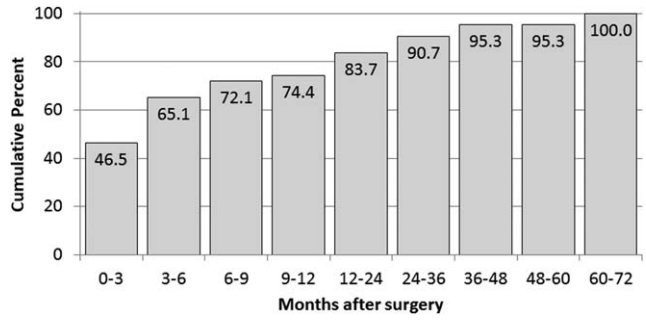

Fig. 4. Time to clearance for a normal per os diet with no or minor feeding modifications after repair of laryngeal cleft. For those children who were ultimately cleared for a full diet with no or only minor behavioral modifications (n = 43), the cumulative frequency of those cleared is displayed as a function of time after cleft repair.

Almost half of the children who were ultimately cleared for a normal diet with no or minor modifications were so cleared within the first 3 months after surgery. In the second 3 months after surgery, another 18% of patients were cleared for a normal diet. Approximately 10% of patients were cleared in the next 6 months, after which the rate of clearance fell dramatically. Given these rates, we recommend swallow evaluation at 3 months after surgery. Those with persistent swallowing problems should have evaluations at 6 and 12 months and then annually, while problems persist. Although most children do recover normal swallowing within 24 months of surgery, a small minority of children recover normal swallowing after this time. This raises the question of when to stop the evaluation of swallowing in the child who persistently aspirates after cleft repair. It is here that the clinicians must exercise their judgment. The degree of dysfunction, neurologic status, and other factors such as progress with the speech therapist and parental reports must be considered. If children undergo multiple swallowing evaluations, nonirradiating studies should be used when appropriate.

Interpretation of the above results is hindered by the most obvious limitation of our study, namely that there was no set protocol for the timing or indications for postoperative swallowing evaluations. Some children in our study had their first swallowing evaluation many months after surgery. This artificially inflated the postsurgical time to normal swallowing, and many children likely recovered normal swallowing earlier than indicated in Figure 4. This strengthens the argument for less frequent swallow evaluations after the first 6 months; even fewer children would be expected to recover normal swallowing after this time if evaluated regularly. Despite the lack of a strict protocol, the current study does allow broad guidelines to be established for the timing of postoperative swallowing evaluation of patients after laryngeal cleft repair.

A set protocol would ideally clearly delineate clinical indications for repeat studies. In the current series, the timing of and indications for a repeat swallowing evaluation was decided by the managing physician and speech therapist, with standard clinical signs of aspiration such as choking or coughing with feeds, recurrent respiratory infections, and parental suspicion serving as guiding factors. Additionally, the choice of which test was performed was made partially subjectively. Although VFSS was our preferred means of evaluation, if patients were unable to take significant amounts of contrast or if they had already had a number of irradiating VFSS evaluations, then FEES was performed. Although we have pooled the data from VFSS and FEES studies, little correlation exists between VFSS and FEES scores.[15] This underscores the importance of taking into account clinical, laboratory, and temporal data when assembling a picture of aspiration.

CONCLUSION

We have performed a retrospective analysis of swallowing function after laryngeal cleft repair. A substantial minority of children (28%) remained NPO or required the use of thickeners to achieve airway protection during swallowing after surgery, and neurodevelopmental delay was the best predictor of falling into this category. Based on our analysis of children who ultimately regained normal swallowing, we recommend swallow evaluations at 3, 6, 12, and 24 months after surgery, until normal swallowing is observed. The chance of recovering normal swallowing more than 24 months after surgery is small, so the physician must balance patient factors, the availability and quality of swallowing therapy, and parental wishes when deciding how long to follow swallowing function after surgery.

Acknowledgments

Study data were collected and managed using research electronic data capture (REDCap (developed by Vanderbilt University, Nashville TN), CCHMC) electronic data capture tools hosted at CCHMC.[16] REDCap is a secure, Web-based application designed to support data capture for research studies, providing: 1) an intuitive interface for validated data entry; 2) audit trails for tracking data manipulation and export procedures; 3) automated export procedures for seamless data downloads to common statistical packages; and 4) procedures for importing data from external sources. Institutional guidelines as well as our license agreement for REDCap usage mandate this precise text be used in all papers published in which REDCap was used. REDCap is made possible at CCHMC by the Center for Clinical and Translational Science and Training grant support (UL1-RR026314).

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