Endoscopic Extended Ventriculotomy for Congenital Saccular Cysts of the Larynx in Infants

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Objective: To report a new procedure that has been successful in endoscopically treating congenital saccular cysts of the larynx without the need for a tracheostomy or an external incision.

Design: Retrospective chart review of a case series involving 4 patients who underwent a single endoscopic procedure for the treatment of their congenital saccular cysts.

Setting: Two pediatric tertiary care referral centers.

Patients: Four pediatric patients aged 1 to 7 weeks.

Interventions: Three of 4 patients underwent endoscopic extended ventriculotomy for treatment of a congenital laryngeal saccular cyst. The remaining patient had wide unroofing of the cyst through the floor of the vallecula.

Main Outcome Measures: The patients were followed up for breathing difficulties, dysphagia, and dysphonia. Routine flexible endoscopy was used to evaluate for recurrence of cysts.

Results: All 4 patients were successfully treated with a single endoscopic procedure. None has had a recurrence, and none required tracheostomy. One patient subsequently underwent conservative unilateral epiglottoplasty to remove redundant tissue caused by the cyst. Disease-free follow-up ranged from 2 to 6 years.

Conclusions: The endoscopic extended ventriculotomy procedure allowed successful endoscopic management of congenital saccular cysts of the larynx in 3 of 4 patients. Previously described management strategies for these difficult lesions have involved multiple failed endoscopic procedures or an external approach to the lesion and frequently required tracheostomy. With this procedure, we have avoided both a tracheostomy and an external approach to the lesion, which has minimized morbidity. Use of modern instrumentation and surgical adjuncts such as mitomycin C, as well as the support of the laryngeal framework for the continued patency of the ventriculotomy, has led to successful single-stage management of congenital saccular cysts of the larynx.


Saccular cysts of the larynx present an interesting diagnostic dilemma, as they represent a continuum of cystic lesions seen in the pediatric and adult larynx. The pathogenesis and classification of saccular cysts, laryngoceles, and other cystic lesions of the larynx have been described in detail in other publications.1,2 Making an accurate diagnosis when a saccular cyst is the causative lesion in pediatric stridor can be quite difficult because of the inherent difficulties in examining the pediatric larynx. Also, saccular cysts are rare and are therefore not commonly considered by the practitioner during the evaluation of an infant with stridor.

When the diagnosis of saccular cyst is made in a young patient, the next dilemma involves identifying a management strategy to correct the abnormality. Previously described methods include endoscopic measures, including needle aspiration and simple marsupialization.3-5 Unfortunately, most case series report a high rate of failure, with reaccumulation of the cyst necessitating numerous surgical procedures.6 Others advocate a variety of external approaches as a primary intervention or after endoscopic measures have failed.7,8 A high incidence of tracheotomy, either emergent or elective, has also been described.2,3,6,7

We report a case series involving infants with large saccular cysts of the larynx. We have devised a straightforward endoscopic procedure that has been successfully used as a single intervention to cure these abnormalities.

METHODS

After institutional review board approval, we reviewed the charts of 4 consecutive patients...
with a diagnosis of a saccular cyst of the larynx. We then developed a database to record the details of patient demographics, presenting symptoms, methods of diagnosis, surgical therapy, and clinical outcome.

All surgical procedures were performed under the supervision of the primary author (D.J.K.). Three of 4 patients underwent endoscopic extended ventriculotomy; a detailed description of the procedure follows: The patients underwent the induction of general anesthesia via a spontaneous ventilation technique. A rigid telescope was used to perform a detailed examination of the larynx to confirm the suspected diagnosis, which had previously been obtained during a flexible endoscopy with the patient in an awake state. The airway was secured using orotracheal intubation. The larynx was suspended using a laryngoscope designed to fit in the vallecula; this allows maximal visualization of the entire supraglottis. Mucosal vasoconstriction was attained with topical epinephrine applied to a cottonoid strip. A sickle knife was used to enter the cyst on the posterosuperior area of the false vocal fold (Figure 1). The cyst was decompressed, and the lumen of the cyst was definitively identified. Microlaryngeal scissors were used to perform a full-thickness cut along the superior surface of the false vocal fold, starting from the previously made puncture and extending anteriorly to the area of the anterior commissure. Scissors were then used to transect the false vocal fold as far anterior and posterior as possible. The final cut was made in a posterior to anterior direction through the laryngeal ventricle, along the superior surface of the true vocal fold. The resected tissue consists of much of the lateral wall of the supraglottis, including the false vocal fold and ventricle. Hemostasis was easily attained using topical epinephrine. Mitomycin C (0.4 mg/mL) (Bedford Laboratories, Bedford, Ohio) was applied to the surgical field for 2 minutes. The patient was kept intubated after surgery. Extubation was performed after the results of bedside direct laryngoscopy confirmed a safe airway. Perioperative steroids were administered at the usual doses. All patients were maintained on acid-suppressive medications for at least 4 months.

All patients were closely followed up for symptoms of airway compromise, voice difficulties, and swallowing problems. Flexible fiberoptic examinations were performed on a frequent basis in the clinic. Rigid examination of the larynx was performed with the patient under general anesthesia only if clinical suspicion of a recurrent cyst was suspected. The patients were discharged from the clinic after they were symptom free for at least 2 years.

REPORT OF CASES

All patients presented to the otolaryngology service for evaluation of persistent, and sometimes progressive, inspiratory stridor. Symptoms of muffled cry and feeding difficulties were variable (Table). On initial consultation, all patients underwent flexible fiberoptic laryngoscopy, at which time a suspected diagnosis of saccular cyst was made. No preoperative radiographic imaging was performed in any patient. All patients were taken to the operating room within 24 hours for complete endoscopy and surgical management of the cyst. After surgery, all patients had immediate relief of all airway-related symptoms. All patients have had normal voices, and there have been no symptoms of swallowing difficulties.

<table>
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<th>Patient No.</th>
<th>Age at Presentation, d</th>
<th>Age at Operation, d</th>
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<th>Length of Follow-Up, y</th>
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*All patients underwent a single procedure, and no patient required a tracheostomy.*
CASE 1

A 7-week-old girl presented to the clinic for evaluation of progressively worsening stridor, which had been present since birth. She had a muffled cry and significant feeding difficulties, with poor weight gain. She underwent extended ventriculotomy for a huge right-sided cyst. She was extubated on postoperative day 2 after bedside direct laryngoscopy was performed. One month later, she was found to have a recurrence of mild inspiratory stridor, and a recurrent cyst could not be definitively ruled out with flexible laryngoscopy in the clinic. Therefore, she was returned to the operating room to undergo rigid examination of the larynx under general anesthesia. She was found to have a patent orifice into the area of the previous cyst, with no evidence of reaccumulation. However, she did have redundant, floppy tissue in the area of the right aryepiglottic fold, which mimicked laryngomalacia. A conservative right-sided epiglottoplasty was performed, with trimming of the redundant tissue. The patient has been followed up for more than 3 years and remains symptom free.

CASE 2

A 51-day-old girl presented to the clinic for evaluation of progressive stridor. She also had a muffled cry, but feeding disturbance was minimal. She underwent extended ventriculotomy for a left-sided cyst, as previously described. She self-extubated later the same day while in the intensive care unit. She had no signs of airway distress, so she was not reintubated. She was discharged to home on postoperative day 2. She has remained symptom free for more than 5 years.

CASE 3

A 1-month-old girl presented to the clinic for evaluation of progressively worsening stridor. She had a very muffled cry and significant feeding difficulties, with poor weight gain. She underwent extended ventriculotomy for a huge right-sided cyst. She was extubated on postoperative day 2 after bedside direct laryngoscopy was performed. One month later, she was found to have a recurrence of mild inspiratory stridor, and a recurrent cyst could not be definitively ruled out with flexible laryngoscopy in the clinic. Therefore, she was returned to the operating room to undergo rigid examination of the larynx under general anesthesia. However, she did have redundant, floppy tissue in the area of the right aryepiglottic fold, which mimicked laryngomalacia. A conservative right-sided epiglottoplasty was performed, with trimming of the redundant tissue. The patient has been followed up for more than 3 years and remains symptom free.

CASE 4

A 1-week-old boy was transferred to the neonatal intensive care unit from a community hospital for evaluation of stridor. He had a muffled cry and a fragmented feeding pattern. The pulmonary service was consulted, and the patient underwent flexible bronchoscopy, during which a lesion was found in the larynx. Our service was then consulted, and on review of the recorded bronchoscopic examination findings, a diagnosis of saccular cyst was made (Figure 2). The patient was taken to the operating room and underwent extended ventriculotomy for a large left-sided cyst (Figure 3). He was extubated on postoperative day 2 after undergoing bedside direct laryngoscopy. He has remained symptom free for more than 2 years.

COMMENT

The classification of laryngeal cysts has been addressed in several publications. After an exhaustive review of the historical literature, DeSanto et al1 reviewed their 20-year experience with clinical and pathologic specimens and proposed a classification of laryngeal cysts. In their article, a saccular cyst was described as a submucosal cystic dilation in the plane of the laryngeal ventricle. Saccular cysts were described as being anterior, usually small, and located near the saccular orifice or lateral, in which case they can become large enough to distort the entire supraglottis and cause airway obstruction. It is thought that most congenital saccular cysts are of the lateral variety. DeSanto and colleagues also described ductal cysts, which are intramucosal and can arise from any mucous-producing gland within the laryngopharynx.

The pathogenesis of saccular cysts is varied. A congenital saccular cyst is formed when the laryngeal saccule fails to canalize into the laryngeal lumen. Acquired saccular cysts can be seen in older children and adults. They occur when the opening of the saccule is obstructed by mucosal trauma.
or masses such as papillomas or other tumors. Without regard to the pathogenesis of saccular obstruction, mucous glands within the saccule continue to produce mucus, which can cause distention of the resulting saccular cyst. As the cyst enlarges, it expands along the path of least resistance. In the case of a lateral saccular cyst, the enlarging cyst can involve the area of the false vocal fold, aryepiglottic fold, and pharyngoepiglottic fold. Herniation through the thyrohyoid membrane to form an extralaryngeal component is possible but uncommon in congenital saccular cysts.

Treatment of an infant with a saccular cyst of the larynx can be a complex process. Two factors that make management of this particular lesion problematic are the rarity of the lesion itself and the variety of surgical interventions that have been described. Inspiratory stridor, when seen in an infant, is most likely to be caused by laryngomalacia. In fact, many clinicians quickly jump to this diagnosis before considering endoscopic evaluation of the airway or further workup to rule out other possible causative lesions. Saccular cysts, when small, can cause a similar type of inspiratory stridor to that seen in laryngomalacia and can easily be mistaken for the latter. Compounding this is the fact that diagnosing a small saccular cyst in an awake, crying infant can be challenging during the course of a flexible fiberoptic examination of the larynx. However, infants with saccular cysts frequently present when the cyst is large, and in this situation, it is possible to confuse the presenting symptoms with other causes of stridor. Larger saccular cysts will significantly affect glottic function, causing voice changes, which can be perceived as a hoarse or muffled cry. Laryngomalacia, however, never causes a change in the quality of the voice, and this distinction is important for the clinician to identify. Larger saccular cysts, like many other causes of significant stridor, can cause a frankly disrupted breathing pattern, which can disorganize the complex swallowing mechanism of an infant, resulting in significant feeding difficulties. Ultimately, enlarging saccular cysts may cause complete airway obstruction, requiring emergent intervention. It is sobering to learn that an early review of the topic showed that more than 50% of reported cases of congenital saccular cysts were discovered at autopsy.9

Because saccular cysts in infants are so rare, determining the optimal surgical management strategy has been difficult. Most articles published on this topic in the past 30 years involve isolated case reports or small case series. However, a small number of larger case series are available for review. In 1978, Holinger et al1 described their experience with 10 infants with saccular cysts, and then, in 1992, Civantos and Holinger described 7 more infants who also had saccular cysts. In their 28-year review of this entity, Civantos and Holinger stress the diagnostic and therapeutic difficulties in treating children with this particular type of airway lesion. These articles describe using multiple needle aspirations of the cyst as the primary modality of management, reserving an open surgical approach for cases in which multiple endoscopic interventions have failed. The average number of procedures was 6, with 7 patients requiring at least 10 procedures and 11 of 17 patients requiring at least 1 tracheostomy during the course of their therapy.6

The external approach is generally reserved for endoscopic failures or complex lesions. The external approach has been modified to address the technical difficulties involved in removing saccular cysts from the infant larynx. The usual approach that is described in the adult involves a dissection through the thyrohyoid membrane. However, in the infant larynx, the surgical field may need to be enlarged by opening the thyroid cartilage. This has traditionally been done through a midline or paramedian vertical thyrotomy. Malis and Seid described the fold-down thyroplasty, during which an inferiorly based, pedicled window is developed in the thyroid cartilage. This minimizes risk to the anterior commissure and provides wide access to the paraglottic space for removal of the lesion. Any external approach through the thyrohyoid membrane puts the internal branch of the superior laryngeal nerve at significant risk. Also, external approaches are usually carried out with a tracheotomy in place. However, it has been shown in a small number of infants that the tracheostomy can be avoided.7,10

The literature is replete with descriptions of successful endoscopic surgical procedures for laryngoceles and saccular cysts. The articles describe a variety of techniques, and the vast majority of patients have been adults. Definitive endoscopic interventions range from simple marsupialization to marsupialization with laser vaporization of the lining of the cyst to complete removal of the cyst lining.2-5,11 The small confines of the infant larynx and lack of appropriate instrumentation can make extensive endoscopic dissection of cyst lining impossible. This dissection would certainly risk distorting or destroying the delicate tissues of the supraglottis during the attempt to gain access to the lateral extent of a large saccular cyst. Laser vaporization of the cyst lining carries the risk of thermal injury to the supraglottis, causing permanent injury. Incomplete vaporization of the cyst lining is likely given the difficulties of effectively delivering adequate laser energy to the entire cyst cavity. This could lead to rests of buried mucous-producing glands being left behind, which could cause more difficult problems in the future.

Endoscopic marsupialization has been described as a potentially successful modality for treating saccular cysts in infants. However, surgical failures have been common, and Civantos and Holinger described the technique as “rarely adequate.” None of the articles that describe simple marsupialization elaborate on the finer details of the surgical technique to document exactly how or where the saccular cyst is opened. For simple marsupialization to be successful, the opening that is made into the cyst must remain open to allow continued decompression. Depending on where the cyst is opened, the laxity of the tissues at the orifice may not facilitate continued patency. Intuitively, the cyst should be opened at or near the natural drainage point of the normal laryngeal saccule. The procedure that is described herein addresses both of these issues. By working within the laryngeal framework, the margins of the opening into the cyst are held open by relatively sessile tissues of arytenoid posteriorly, and the anterior commissure and petiole anteriorly. The aryepiglottic fold is not disrupted, which helps maintain the architecture of the supraglottic larynx and possibly decrease the risk of aspiration. To our knowledge, this
is the first report of the use of mitomycin C to augment the surgical success of endoscopic marsupialization. This technique has been adapted from our extensive experience in using this chemical to decrease scarring in cases of subglottic stenosis and choanal atresia.

We have elected to keep our patients intubated for a short period of time after the procedure for airway protection. Experience has shown that very little laryngeal edema is encountered in the perioperative period. Thus, intubation after the procedure is likely not necessary in the uncomplicated patient. We have also chosen to empirically treat our patients with medicines to decrease gastric acid production for several months after surgery.

We have not seen any complications from using the endoscopic extended ventriculotomy in our series. The true vocal fold is not manipulated, so subjective voice outcomes have been excellent. We have not encountered any swallowing difficulties. No patients have shown signs of aspiration, although no diagnostic tests have been performed to definitively rule this out. No patients have required tracheostomy. During the operation, the larynx is intubated with a small endotracheal tube that does not obstruct the surgical exposure to the area. We have not felt it necessary to use the laser to perform this dissection. The carbon dioxide laser would certainly be useful as a cutting tool for this procedure, and the hemostatic properties could potentially be valuable. However, bleeding is minimal when epinephrine is used topically for mucosal vasoconstriction. Cold dissection also eliminates the risk of airway fire and potential thermal injury to the larynx.

In conclusion, we have introduced a new method to marsupialize congenital lateral sacculary cysts of the larynx as a definitive management strategy in pediatric patients. The procedure could be modified for use in similar adult laryngeal cases. The procedure described herein is similar to the surgical approach described for complete endoscopic removal of a laryngocele in an adult, except that the cyst lining is left intact. In our small case series, we have not encountered any recurrences or complications. Given the documented high failure rate of other endoscopic interventions, we would advocate extended ventriculotomy as the endoscopic procedure of choice for treating infants with a lateral sacculary cyst of the larynx.

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REFERENCES