Laryngeal Electromyography in the Management of Vocal Cord Mobility Problems in Children

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Objectives: To evaluate the efficacy and clinical usefulness of laryngeal electromyography (EMG) in the evaluation and management of vocal cord mobility problems in children; and to determine the ability of laryngeal EMG to differentiate vocal fold fixation versus paralysis. Study Design: Case-series review of 8 children with vocal cord immobility who underwent laryngeal EMG. Methods: Eight children with bilateral vocal fold immobility underwent microlaryngoscopy and electromyography. Bipolar concentric needle electrodes were used and implanted separately into both posterior cricoarytenoid (PCA) and both thyroarytenoid (TA) muscles. EMG activity was recorded during spontaneous ventilation under a light plane of anesthesia with propofol. EMG activity was correlated with the phases of the respiratory cycle. Results: Three patients had evidence of normal EMG activity with PCA activity peaking during early inspiration. Maximal TA activity occurred expiration. These patients were assumed to have vocal fold fixation. Two of these 3 patients underwent laryngotracheoplasty (LTP) with posterior glottic expansion with costal cartilage. One is being considered for LTP in the future. Five patients had abnormal EMG activity and remain tracheotomy-dependent. Four patients exhibited synkinetic activity with peak PCA activity during expiration and peak TA activity during expiration. Two patients had both fixation and unilateral vocal cord paralysis. One was successfully decannulated after posterior graft LTP and the other is planned for the future. Conclusions: Electromyography, which differentiates paralysis from fixation, is a useful diagnostic tool in the evaluation of children with vocal cord immobility.

Key Words: Electromyography (EMG), posterior cricoarytenoid (PCA), thyroarytenoid (TA), laryngotraceoplasty (LTP).

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INTRODUCTION

Electromyography (EMG) is a standard modality for diagnosing neuromuscular disorders. It can establish useful information about muscle function and innervation. Laryngeal EMG, which was introduced by Hirano and Ohala in 1969, is an accepted method for the diagnosis of movement disorders of the larynx. While laryngeal EMG has been easily performed while awake in adults, children present special challenges. Nevertheless, in recent years, EMG has been performed more frequently in children. However, the clinical indications and usefulness have not been defined and are still largely debated.

Vocal cord immobility in small children may be caused by vocal cord paralysis or glottic fixation from scarring. This later condition may be secondary to prolonged intubation, gastroesophageal reflux, autoimmune diseases, or cricoarytenoid arthritis. Awake flexible laryngoscopy will reveal diminished vocal fold motion, and often the diagnosis may be made on microlaryngoscopy with the aid of direct palpation. However, scarring may be subtle and it may at times be difficult to differentiate vocal cord paralysis versus fixation. Moreover, in some patients, vocal fold fixation may coexist with paralysis on one or both sides. Furthermore, the treatment is different for paralysis versus fixation. Children with bilateral vocal cord paralysis (BLVP) often require a tracheotomy, followed by vocal cord lateralization, or arytenoidectomy. In contrast, children with fixation respond best to laryngotracheal expansion with posterior grafting.

In our clinical practice we have used EMG to assist in the management of children with vocal cord immobility. In most cases the diagnosis of vocal cord fixation may be obvious by arytenoid palpation during microlaryngoscopy. However, the technique has helped most in surgical planning of patients in whom the differentiation of paralysis versus fixation may be difficult to make. In addition, la-
ryngeal EMG has helped elucidate the prognosis of patients with BLVP.

The technique of laryngeal EMG is more challenging in children. Children require general anesthesia because of discomfort with probe needle placement. Direct endoscopic electrode placement has been preferred over percutaneous placement.\(^4\)\(^–\)\(^7\) In addition, the electrode recordings may be affected by interference in the operating room, making interpretation more difficult. Therefore, one needs to be cautious in the analysis of the results. Also, the placement of electrodes into a small compromised airway may present some risk as edema develops. Nevertheless, this can be safely performed in infants and children of all ages if special precautions are taken. In this report, we outline our technique of laryngeal EMG in its application to children with vocal cord mobility problems. We also discuss the indications and limitations of laryngeal EMG.

MATERIALS AND METHODS

We reviewed all the patient records of those children who underwent laryngeal EMG on the Otolaryngology Service of The Children’s Hospital of Philadelphia between January 1998 and December 2001. The name and birth date of the patients were recorded. The surgical, medical, and neonatal history was recorded, including any intubations and periods of ventilation. The dates and results of all endoscopies and surgical procedures were recorded, including tracheotomy, laryngotracheoplasty, arytenoidectomy, and epiglottoplasty.

Before the EMG evaluation all patients had undergone awake flexible fiberoptic laryngoscopy and were diagnosed with vocal cord immobility. Five children had a pre-existing tracheotomy and 3 children had severe stridor at the initial evaluation. All children underwent electromyographic analysis in the operating room of The Children’s Hospital of Philadelphia. This was performed under general anesthesia with spontaneous ventilation. Intravenous propofol was used as an anesthetic agent and was titrated to the desired level of analgesia. The patient underwent both flexible and rigid laryngoscopy and bronchoscopy evaluating vocal cord mobility, supraglottic dynamics, subglottic, and tracheal disease. The lack of vocal cord mobility was confirmed on endoscopy. Vocal fold fixation was diagnosed by direct palpation of the arytenoid with the laryngeal suction. In addition, neuro muscular blockade was administered at the end of the procedure to determine if the vocal cords lateraled or remained fixed.

The patient was then suspended with a Jako or Parson’s laryngoscope and the posterior portion of the larynx was exposed. Concentric bipolar needle electrodes (TECA NDMC 50 bipolar concentric needle electrode; Oxford Instruments, Pleasantville, NY; Fig. 1) were placed under direct visualization by one otolaryngologist (I.N.J.) into the belly of each posterior cricoarytenoid (PCA) and thyroarytenoid (TA) muscle (Fig. 2) using microlaryngeal forceps using standard EMG equipment (TECA “Synergy” model, software v 3.0; Fig. 3). EMG tracings were recorded and correlated with the respiratory phase by one neurologist (R.S.F.). A tracing was considered normal if there was: 1) no spontaneous activity with the muscle at rest (no fibrillation potentials, positive waves, or fasciculations); and 2) normal motor unit activity generated in a phasic, sustained fashion for the appropriate phase of breathing and full cessation of activity during the resting phase of breathing. The motor unit potentials in the PCA muscle normally recruit during the early phase of inspiration and increase in the TA muscle during the early phase of expiration. These muscles activate with rapid generation of a full interference pattern, precluding assessment of individual motor unit morphology. The right and left sides were compared. If there was any doubt about the electrode placement, the electrode was repositioned and recordings continued. At the end the electrodes were removed and the larynx was inspected for edema. The patient was given intravenous steroids if there was significant laryngeal edema (0.5 mg/kg dexamethasone).

RESULTS

There were no complications. All ambulatory patients were discharged the same day. Useful EMG record-
ings were made in all 8 patients from both PCA and TA muscles.

**Group 1**

Group 1 consisted of 3 patients (aged 24, 29, and 48 mo) with vocal cord immobility who had evidence of normal EMG activity. In all 3 patients the EMG recording of the PCA muscle demonstrated normal motor units with recruitment. Electrical activity peaked with inspiration (Fig. 4A). In contrast, the TA peaked with expiration (Fig. 4B). As the depth of anesthesia increased, the tracings diminished as well. Two of these 3 patients, who were confirmed to have normal laryngeal innervation, underwent laryngotracheal expansion surgery with posterior costal cartilage grafts. They were decannulated without difficulty. Postoperative endoscopy revealed improved vocal cord mobility. One patient will undergo laryngotracheoplasty in the future.

**Group 2**

Five patients in group 2 had abnormal EMG activity, confirming a neurogenic component to the vocal cord immobility. Four patients were noted to have synkinetic coactivation of the PCA and TA, i.e., the PCA activated in expiration and the TA in inspiration. One 18-month-old female patient, with idiopathic vocal cord paralysis, demonstrated clear-cut bilateral paradoxic activity with peak PCA activity during expiration and TA activity during inspiration in both vocal cords. As a result of the EMG and lack of significant vocal fold abduction, a tracheotomy was performed and lateralization will be considered in the future.

A 6-month-old male patient, who was a former premature infant with bronchopulmonary dysplasia (BPD) and who suffered a left vocal cord paralysis after a PDA ligation in early infancy, remains tracheotomy-dependent for both lower and upper airway disease. This child initially demonstrated reduced amplitude activity on the left and normal phasic activity on the right at 6 months of age. At that time a tracheotomy was placed. A repeat laryngeal EMG at 27 months of age revealed synkinetic activity on the left side and normal activity on the right. Because his right vocal cord motion is normal and his glottic airway have improved since the tracheotomy, he will be decannulated when he is weaned from pressure support for the lower airway.

A 3-year-old male patient, with a history of tracheoesophageal fistula repair, underwent tracheotomy in early life for bilateral vocal cord immobility. This child has been unable to be decannulated, having failed a tracheotomy tube capping trial. We had abnormal EMG activity on the left side. He inappropriately initiated PCA activity during the end of expiration. This continued into mid-inspiration, but with an abnormal, biphasic EMG pattern (Fig. 5A). TA

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**Fig. 4.** (A) Normal EMG tracing of the PCA muscle showing peak activity during inspiration. The arrows indicate the start of inspiration. (B) Normal EMG tracing of the TA muscle showing peak activity during expiration. The arrows indicate the start of expiration.

**Fig. 5.** (A) Abnormal EMG tracing of the left PCA. There was inappropriate initiation of EMG during late expiration. This continued into mid-inspiration with an abnormal biphasic EMG pattern. The arrows indicate the start of inspiration. (B) TA activity initiated during late expiration and continued into inspiration. The arrows indicate the start of expiration.
activity was initiated inappropriately late into expiration and was maintained into early inspiration (Fig. 5B). The right side demonstrated normal EMG activity in the appropriate phases of respiration. His endoscopic examination under spontaneous ventilation revealed a quivering motion on the left vocal fold and reduced abduction on the right. A left-sided vocal cord lateralization is planned.

The fourth patient, a 5-year-old boy from Central America, sustained a laryngotracheal separation at 2.5 years of age after being thrown through a glass windshield in a car accident. An emergent repair of the separation saved his life but left him with severe stridor. Initial microlaryngoscopy demonstrated limited abduction of the left vocal cord and no motion of the right vocal cord. Microlaryngoscopy demonstrated vocal cord fixation with asymmetric motion of the left side and a 2- to 3-mm glottic airway. A laryngeal EMG was performed at age 5 and demonstrated a small degree of low-amplitude motor unit activity on the left in phase with respiration. The right revealed electrical silence probably from nerve transection. A tracheotomy was performed immediately after the EMG and a laryngotraheoplasty with a posterior costal cartilage graft was performed with a short stent at 6 years of age. The graft was placed below the arytenoids because of concerns about aspiration. He was decannulated 5 months after surgery. He has a persistent right vocal cord paralysis and normal left vocal motion with an excellent airway at this time. There have been no problems with aspiration but he still has a hoarse voice.

A fifth patient, a 5-year-old girl with vocal cord fixation from prolonged intubation, had asymmetric vocal cord motion with no motion of the right vocal cord and decreased motion of the left. The EMG demonstrated synergistic coactivation of the right vocal cord and normal motor unit activity with recruitment in the left vocal cord in phase with respiration. Her vocal cords did not lateralize after neuromuscular blockade, indicating vocal cord fixation as well. Palpation also demonstrated this. Laryngotracheal expansion surgery is planned.

**DISCUSSION**

Laryngeal EMG helped us differentiate vocal cord paralysis from fixation in complicated patients. Although most cases of laryngeal stenosis and paralysis do not require laryngeal EMG, the modality helped us with the more difficult or ambiguous cases. The laryngeal EMG is helpful in three specific clinical situations in children. The first is when there is a subtle degree of laryngeal scarring and fixation, which is not obvious, such as a discreet interarytenoid scar band. In such cases there may be periarticular scarring around the periphery of the arytenoid cartilage, which may restrict motion. These cases may be mistakenly diagnosed as pseudo-paralysis. The second situation for EMG is in cases in which there are combined pathologies, and it may be easy to overlook a unilateral or bilateral paralysis in cases of laryngeal stenosis. An asymmetric vocal cord immobility may result. This occurred in 2 patients. The EMG is helpful for sorting out issues related to pathogenesis of vocal cord immobility and may influence surgical decision-making. One may anticipate problems with posterior laryngeal expansion when there is vocal fold paralysis and potential for glottic incompetence and aspiration. The third clinical indication for EMG is to better define the extent of denervation and reinnervation with vocal cord paralysis and determine the prognosis. Knowledge of the prognosis would certainly influence treatment planning and help in airway management. In paralysis cases, the EMG may also help localize the side for lateralization or arytenoidectomy. The awareness at a very early age that recovery is unlikely would indicate early surgical intervention rather than watchful waiting for many years with a tracheotomy tube in place.

The prognosis for children with bilateral vocal cord paralysis (BLVP) depends on the underlying etiology. The common causes include trauma (birth-related and from prolonged intubation), asphyxia, neoplasms (rare in children), neuromuscular disorders, central nervous system disorders (mostly the Arnold-Chiari malformation), and idiopathic. The prognosis is worse for cases associated with severe neuromuscular and central nervous system disorders than with reversible neurologic conditions or birth trauma-related cases. With global neuromuscular disorders the vocal cord dysfunction may be progressive. Treatable central nervous system lesions such as hydrocephalus or Arnold-Chiari malformation may respond to neurosurgical intervention with ventricular shunt placement or posterior fossa decompression. Recovery of vocal cord function generally depends on the degree of preexisting brain stem damage.

Nevertheless, most children with BLVP paralysis require surgical intervention for the airway. Tracheotomy is required in over 50% of patients with BLVP resulting from severe airway obstruction. Spontaneous recovery may occur in many cases and watchful waiting for at least 12 months is advocated. If the airway does not improve, then irreversible glottic expansion surgery may be performed. Confirmation of poor neurologic recovery with laryngeal EMG (chronic denervation features with poor reinnervation) is generated from the coordinated interference pattern. The standard options for glottic enlargement include endoscopic partial or complete arytenoidectomy, laser cordectomy, or external arytenoidectomy or lateralization. The results in children may not be as good as in adults.

The normal EMG tracing from skeletal muscle includes identifying features of acute denervation and those of reinnervation. Acute denervation typically occurs within 21 days of an acute neurogenic lesion affecting the axon of the nerve and is characterized by abnormal “spontaneous activity” at rest (fibrillation potentials and positive waves). With volitional or involuntary contraction of skeletal muscle, a single muscle motor unit potential begins to fire initially and then increases in frequency, followed by recruitment of additional motor units with increased effort. Eventually, with full effort, a full “interference pattern” is generated from the coordinated activation of maximal motor units. Signs of reinnervation include motor units of large amplitude firing at increased frequencies at low-level contraction. With complete rei-
nervation, a full interference pattern is generated, but with limited reinnervation an incomplete interference pattern is achieved.25,26

Additionally, a proximal axon may reconnect to a different distal myelin sheath than originally intended, resulting in anomalous reinnervation with synkinesis, e.g., as can be seen after Bell’s palsy, with eye winking when activating the lower facial muscles during periods of increased physiological activity.

This standard EMG approach to the study of denervation and reinnervation of muscle must be adapted to the study of laryngeal muscles, because they generate contraction in rapid phasic bursts, not gradually. Thus, individual motor units and early recruitment patterns cannot be easily deciphered. Additionally, there is no normative data for the normal range of amplitude for the PCA and TA muscle in children. Spontaneous activity in the resting state is often difficult to assess given a degree of background noise in the baseline of the EMG that is nearly unavoidable in the operating room environment. Nonetheless, useful EMG data can be obtained from the laryngeal muscles by assessing the qualitative nature of the response in relation to the phase of breathing. First, one can determine whether any EMG activity is elicited when the PCA or TA is expected to contract. A total absence of EMG activity may reflect a total lack of reinnervation in that muscle, but must be interpreted with caution, however, because one cannot in such instance entirely exclude that the electrode is not correctly placed in muscle. When at least some motor unit activity is identified, that potential problem is avoided. Next, the EMG activity can be analyzed as to whether a normally sustained phasic contraction is generated and whether it does so during the correct phase of the breathing cycle. Finally, by comparing the PCA and TA responses, a conclusion can be drawn whether there is anomalous reinnervation with inappropriate coactivation of both muscles.

In small children it is difficult to test phonation awake. Therefore, it is performed with spontaneous ventilation under general anesthesia. One should see peak activity bursts of recruitment in the PCA during inspiration and in the TA muscle during expiration. There may be additional recruitment as the patient awakens from anesthesia or if the tracheotomy tube is occluded. EMG of the laryngeal muscle has its limitation and caution must be taken not to overly rely on the findings out of the clinical context. Berkowitz, for example, found normal innervation in children with idiopathic bilateral vocal cord paralysis, suggesting an unclear site of lesion. In such cases, when there is clearly no sign of vocal cord fixation, the EMG may not be prognostic because recovery is still unlikely.27

In a number of patients we found evidence of synkinetic activity on one or both sides. In such cases, in which there is partial nerve paralysis, there may be misdirected reinnervation of the abductors with adductors and vice versa. Physiologically one would see peaked activity of the laryngeal abductors (PCA) during the expiratory phase and the adductors during the expiratory phase resulting in an ineffective opening of the glottis. There may even be paradoxical closure during inspiration. Because reinnervation has already occurred, one would also conclude that future clinical improvement would be unlikely and glottic expansion surgery could be considered.

There have been significant developments in terms of needle electrodes. The type of needle will affect the results.28 One can use either surface electrodes or needle electrodes. Surface electrodes, which are non-invasive and eliminate trauma to muscle, are susceptible to contamination by nearby muscle. The small PCA and TA responses would be hard to distinguish from neighboring laryngeal and chest wall muscles and could not be identified individually. Intramuscular needle electrodes, therefore, are necessary in pediatric laryngology. In terms of needle electrodes, one can use either monopolar or bipolar needle electrodes. Monopolar electrodes include one electrode placed into muscle and a second distant reference electrode.5 Placing a reference electrode within the larynx generates excessive motion-related artifact from breathing. In contrast, a bipolar electrode has the reference electrode on the outside barrel of the needle, in close proximity to the active electrode at the needle tip. The bipolar needle electrode reduces the risk of contamination from breathing or from neighboring muscle but records from a smaller volume of muscle. This in some ways is ideal for studying small linear muscles like the PCA and TA. Caution with exact electrode placement, however, is critical and it may need to be repositioned several times to obtain accurate readings. One can also choose a hooked wire electrode, which remains in place during vigorous activity but is more difficult to reposition and may cause more local trauma to muscle.29 In our experience, concentric bipolar needle electrodes did not dislodge during quiet breathing with a light plane of general anesthesia. They appear to be the optimal choice for EMG study of laryngeal muscles in children under anesthesia, which is in agreement with the literature.9,27

The laryngeal EMG may be difficult to interpret because of significant background activity and electrical interference in the operating room. The other electrical equipment in the room may cause generate electrical noise.30 Nevertheless, the newer EMG equipment reduces the extent of background noise to an acceptable degree. In approximately half of our cases it was difficult to address whether spontaneous activity might be present with the muscle at rest as a result of this continuous low-level background noise. We did not experience difficulty in any of our cases in discerning motor unit activity from baseline noise, as the muscle EMG amplitude was roughly 5- to 10-fold higher in amplitude. There may be a false-positive response if the electrode is incorrectly placed into an adjacent muscle. Alternately, a normally innervated muscle may not produce a normal EMG recording when one is not in the right area of the muscle, i.e., at a musculotendinuous junction. One needs to repeat the electrode placement several times before concluding a negative result. It is always more reassuring to identify distinctly abnormal findings such as aberrant reinnervation patterns than no activity at all. Abnormal “spontaneous activity” does not necessarily indicate acute denervation because it can be seen in some congenital myopathies. One should also identify signs of chronic reinnervation before concluding that
there is a neurogenic process present. Thus, EMG of laryngeal muscle has its complexities and is dependent on the competence and experience of the otolaryngologist placing the electrode into the PCA and TA and the electromyographer to interpret the EMG data within the clinical context.

In addition, there are also a number of risks of the endoscopic electrode placement, including the risk of general anesthesia and laryngeal edema or bleeding from the electrode placement. These problems may be alleviated by use of corticosteroids, topical vasoconstrictors, or humidified air and oxygen. Occasionally intensive-care monitoring may be needed. None of the patients in our series developed severe problems.

Moreover, the laryngeal EMG supplements but never replaces the routine endoscopic evaluation of vocal fold motion problems in children. It is never a substitute for awake flexible fiberoptic laryngoscopy and microlaryngoscopy under general anesthesia with spontaneous ventilation with direct palpation of the arytenoids. We also routinely evaluate vocal fold mobility issues with neuromuscular blockade. Paralyzed vocal folds will laterize with neuromuscular blockade whereas fixed vocal folds will remain in the midline.31 The EMG may help with the more ambiguous cases and may supplement the endoscopic data to help in the surgical decision-making, but is never a substitute for a complete endoscopic examination. One must always be cognizant of the risks and special issues of laryngeal EMG in children.

CONCLUSION
Laryngeal EMG is a safe, reliable, and reproducible diagnostic procedure that may help in the management of children with vocal cord immobility problems. One must interpret results with caution within the clinical context. It is helpful for select cases, such as when the diagnosis is in question or when there are combined pathologies. However, one must be aware of the risks involved. Nevertheless, laryngeal EMG with bipolar concentric needle electrodes may facilitate the surgical decision-making of complex children with vocal fold immobility and help with surgical decision-making.

BIBLIOGRAPHY