Management of unilateral true vocal cord paralysis in children

Jennifer Setlur and Christopher J. Hartnick

Purpose of review
Historically, information gained from the treatment of unilateral true vocal cord paralysis (UVCP) in adults was the same used to treat children. Today, there is a growing body of literature aimed specifically at the treatment of this condition in children. It is an area of growing interest as UVCP can significantly impact a child’s quality of life.

Recent findings
Children with UVCP may present with stridor, dysphonia, aspiration, feeding difficulties, or a combination of these symptoms. Diagnosis relies on laryngoscopy, but other adjuncts such as ultrasound and laryngeal electromyography may also be helpful in making the diagnosis and forming a treatment plan. In many instances, there is effective compensation by the contralateral vocal fold, making surgical intervention unnecessary. Children who cannot compensate for a unilateral defect may suffer from significant dysphonia that can affect their quality of life because their ability to be understood may be diminished. In these patients, treatment in the form of medialization or reinnervation of the affected recurrent laryngeal nerve may be warranted.

Summary
UVCP is a well recognized problem in pediatric patients with disordered voice and feeding problems. Some patients will spontaneously recover their laryngeal function. For those who do not, a variety of reliable techniques are available for rehabilitative treatment. Improved diagnostics and a growing understanding of prognosis can help guide therapy decisions along with the goals and desires of the patient and his or her family.

Keywords
dysphagia, hoarseness, laryngology, pediatric, vocal cord paralysis, voice

INTRODUCTION
The majority of otolaryngologists caring for children can expect to encounter patients with unilateral vocal cord paralysis (UVCP). The literature suggests that tertiary care pediatric otolaryngology practices see 4–10 patients a year [1–3]. Whereas bilateral immobility tends to present with signs and symptoms of airway obstruction, such as retractions, cyanosis, and even apnea, UVCP is more frequently associated with noisy breathing and symptoms associated with an incompetent larynx, such as difficulty with feeds [1,4]. The focus of this article is to review the differential diagnosis and workup of pediatric UVCP, followed by a focus on diagnostic techniques and the growing variety of surgical efforts available to treat children.

PRESENTATION
Stridor is present in up to 70% of young children with UVCP [5]. Additionally, poor feeding, weak cry, cyanosis, and aspiration are common in neonates and young children. Hoarseness and poor vocal projection are more often the chief complaints in older children and adolescents. History and physical examination should first determine the stability of the airway. Once a stable airway is either assessed or established, the onset of symptoms in relation to birth, intubation, surgery, recent illness, or medical therapy can be explored. In addition to a detailed head and neck examination, the chest should not be overlooked. History and
physical examination alone is successful in determining cause of unilateral vocal fold paralysis in two out of three adults with unilateral vocal fold immobility [6].

**CAUSES OF UNILATERAL VOCAL CORD PARALYSIS**

The causes leading to UVCP may be congenital or acquired. The age of the patient along with the history can help direct the physician to make an accurate diagnosis. Neonates who suffer cervical trauma in the setting of a complicated or prolonged vaginal delivery may present with UVCP [2]. In other children, central pathologies, such as Arnold Chiari malformations, may cause vocal fold immobility in the very young, and it is important to remember to track the entire course of the recurrent laryngeal nerve (RLN) from brain and brainstem down to the chest and back to the neck when evaluating affected children with no obvious cause. Congenital left VCP that occurs in association with thoracic vascular or cardiac anomalies is known as cardiovocal syndrome [2]. For as many as 30–50% of patients with UVCP at birth, a cause is never determined [2,7]. When a cause is determined, the most common of it is typically aberrant thoracic vasculature, but again, in many cases, a cause is not found.

For other children, iatrogenic injury may be the cause. This can occur in any age group, although it is seen most often after patent ductus arteriosus (PDA) ligation [8]. Rates of RLN injury after PDA ligation between institutions vary widely (8–62%) [9,10]. The rate of UVCP is on the rise due to the increased number of pediatric cardiothoracic procedures [5]. Extremely low birth weight (<1 kg) children undergoing PDA ligation appear to be at increased risk for UVCP [10]. UVCP is also associated with repair of other congenital anomalies, such as tracheoesophageal fistula or esophageal atresia [11]. Infection should not be overlooked as a cause of pediatric UVCP. Although this is more reported in the adult literature, children are also at risk. Reports of paralyzed vocal cords as a sequela of Epstein–Barr virus, polio, and Lyme disease have all been reported [12–14].

Neoplasms of the central nervous system, neck, or mediastinum can cause vocal fold paralysis by invasion. Alternatively, UVCP in the setting of cancer may be related to RLN neuropathy secondary to vinca alkaloid chemotherapy [15]. The majority of reported cases of VCP after chemotherapy administration have been bilateral, but there have been some reports of UVCP. It remains unclear exactly why the process is unilateral in some cases and bilateral in others. One assumption is that children with UVCP secondary to chemotherapy are experiencing a progressive neuropathy that has not yet advanced, given that neuropathies secondary to this treatment are sometimes not uniform in onset and severity. Childhood malignancies, such as lymphoma and sarcoma, are commonly treated with these types of agents, which should be considered as the cause of VCP.

**INITIAL DIAGNOSIS**

When a child is suspected to have UVCP, one should proceed with a complete evaluation of the airway, even if the diagnosis is evident. Synchronous airway lesions including laryngomalacia, tracheomalacia, subglottic stenosis, intubation granuloma, cricoarytenoid joint fixation, or laryngeal webs can be present in up to 45% of patients [1]. Patients who present with a primary aerodigestive complaint should undergo flexible fiberoptic laryngoscopy in the office. Scopes as small as 2.9 mm in diameter allow for improved physical examination of these patients [16]. Recording these examinations is particularly helpful, as a real-time view of the larynx in the infant or small child can be challenging, and viewing the examination in slow motion is necessary. In addition, videos can also be archived for pretreatment and posttreatment comparisons. There are still rare occasions when a child cannot be fully evaluated in the office. This is especially true for the small infant who presents in frank distress or who has a significant cardiac history. In these settings, office-based laryngoscopy may be unsafe. Examination in the operating room with directly laryngoscopy and bronchoscopy can be done to assess for other airway lesions. Vocal cord motion can be assessed via a laryngeal mask airway as the child is recovering from anesthesia. Even if a good view of the vocal cords is obtained in this manner, one should be extremely cautious in proceeding with treatment planning based on this type of examination. The senior author has had one experience in which office-based laryngoscopy revealed left-sided UVCP and the patient was brought to the operating room for a left injection laryngoplasty. At the time of the procedure, it appeared that the child suffered from right-sided UVCP. The child was woken up from anesthesia because of this conflicting examination, and repeat office laryngoscopy revealed left-sided UVCP. The surgeon felt that perhaps there was an issue of synkinesis not fully appreciated by either examination (office or operative) alone.

Operative endoscopy to diagnose UVCP is done at some centers on a more regular basis than at others. Because this requires general anesthesia,
the help of well trained pediatric anesthesiologists is a must in order to develop the correct plane of anesthesia. This may be met by some resistance from parents. In patients who cannot tolerate office laryngoscopy and who cannot undergo operative examination, some may choose to use ultrasound to aid in the diagnostic process. A recent study of 13 children with VCP showed that ultrasound can yield quantitative results that can be used to diagnose immobility [17**]. Obtaining useful results from this examination is technician-dependent and entails a steep learning curve. Nevertheless, it may be helpful in certain situations.

Assessment of patients with UVCP is not uniform among providers. The specifics of the assessment after full evaluation of the airway are tailored on a case-by-case basis. For instance, an extensive assessment may not be warranted for the child who suffers from UVCP after a PDA ligation. In other instances, in which the cause may be less clear, examination of the child should include a thorough physical examination of the head and neck and magnetic resonance imaging from the brain to the thoracic inlet to image the entire course of the RLN. In our practice, we include Lyme titers as part of this work-up as we live and work in an endemic area. For the most challenging cases in which a cause still cannot be found, we proceed with adjunct diagnostics (see below).

**VOICE-RELATED QUALITY OF LIFE**

For patients who present with a primary voice complaint, and in general for older children and adolescents, specific subjective and objective voice measures should be obtained. A complete evaluation by a voice therapy specialist is important. The aerodynamic profile allows for subjective measurement of voice changes following therapy. A second method of obtaining subjective measurements involves administering quality-of-life surveys. The pediatric voice-related quality of life questionnaire (PVRQoL) is a validated instrument that is simple to administer and complete and is responsive to changes in voice-related quality of life [18]. It can be used in the general pediatric patient population and is a 10-question survey that is scored on a scale of 0–100. Lower scores reflect less impact on quality of life than higher scores. The Pediatric Voice Handicap Index (pVHI) is the other commonly cited vocal quality-of-life instrument and has been validated in children [19]. Scaled scores range from 0 to 120 points with higher scores representing worse perceived quality of life. It is used to differentiate between the different areas in a child’s life that a voice disorder can affect, but can take more time than the PVRQoL to complete.

**ADJUNCT DIAGNOSTICS**

Management of UVCP in children can depend on the underlying cause, as there may be a possibility that a unilateral deficit may spontaneously recover. Recovery of function after an 11-year interval has been previously reported [1]. Adjunct testing may be helpful in predicting recovery of vocal cord motion. Intraoperative laryngeal electromyography (LEMG) has been shown to provide this information and is safe in children and easy to use. First, it can help differentiate paralysis from fixation [20]. Second, it can give general prognostic information as demonstrated by Maturo et al. [21]. In this longitudinal, prospective study, 25 children with vocal fold immobility (18 unilateral and seven bilateral) and a mean age of 21.4 months were followed over a 1-year period of time. Twelve children had UVCP after a PDA ligation. Results showed that in these children, if there are no normal-appearing motor unit action potentials (MUAPs) by 6 months, then it is highly unlikely that vocal fold function will recover. Overall, for children with UVCP irrespective of cause, LEMG accurately predicted the outcome of (return or persistent absence) vocal cord function in 17 of 18 patients.

**TREATMENT**

Much of the decision-making depends not only upon the age and symptoms of the child but also upon the skills and experience of the surgeon. The desires of the parents also play an important role as do those of the child if he or she is old enough to participate in the discussion.

The treatment of neonates, or children younger than school age, should focus on airway protection and alimentation. Tracheotomy can have a role, but is usually necessary only if synchronous airway lesions are present. Speech therapy evaluation and modified barium swallow can clarify dietary recommendations. Enteral nutrition via gastric or nasogastric tubes may be needed, but simple thickening of feeds is often all that is necessary.

In many instances (80% of cases of UVCP), there is effective compensation by the contralateral vocal fold, making intervention unnecessary. For those who have a small but persistent glottal aperture defect (20% of cases), speech therapy can be used to strengthen compensatory methods of glottic closure while minimizing risk of hyperfunctional dysphonia [22]. This can improve feeding in addition to dysphonia. For children who improve
with conservative measures, meaning that the airway is adequate and there are no feeding difficulties, no further treatment is needed. If there are persistent symptoms despite conservative management, surgical treatment should be considered. These patients should be divided into three age groups: the young infant, the mature adolescent, and the age in between.

For the very young infant who presents with aspiration, treatment is focused upon protecting against aspiration while supporting the airway. In this group, an injection laryngoplasty may be the first-line treatment. The availability of a variety of injectable materials makes this intervention applicable to a broader group of patients. The senior author prefers to use Radiesse Voice (Merz Aesthetics, San Mateo, California, USA) for this procedure. This allows for immediate results, but they are short-term and the child may need repeated injections to remain symptom-free. Despite the need for repeat procedures to maintain effect, it is a safe procedure in the pediatric population \[23^*\]. Long-term effects of serial vocal cord injection, such as scarring, have not been studied. Because injection laryngoplasty is not a permanent solution, it can be used in patients who may one day regain vocal cord function. More invasive and more definitive treatments can then be reserved for patients in whom function is not expected to return.

For the mature adolescent who is otherwise healthy and who can tolerate an awake medialization thyroplasty, this may be a viable option. The patient should be one who can tolerate local anesthesia; this allows the medialization to be fine-tuned for improved dysphonia, as the patient can phonate during the procedure while the surgeon manipulates the implant \[8,24**\]. The benefit is a definitive procedure that limits the need for future intervention. The disadvantage is that a thyroplasty is static, whereas the size, thickness, and composition of the larynx changes as a person progresses from childhood to adulthood. Good results obtained initially may decline as the child ages and grows. Because this procedure is done to treat dysphonia, which is not life-threatening, it may be prudent in some cases to wait for spontaneous recovery or to abstain from surgical intervention until a child has gone through puberty in order to get the best result.

For the child who is in between these ages with a primary voice disturbance, reinnervation procedure should be considered \[8\]. The goal of this procedure is not to produce a mobile vocal fold, but rather to restore its tone, thereby medializing it to a position wherein the contralateral mobile vocal fold can make adequate contact and allow for appropriate glottal closure. The theoretic advantage of this procedure over traditional thyroplasty that makes it attractive for pediatric application stems from it being performed while the child is intubated under general anesthesia without the need for intraoperative patient compliance for vocal tuning; the results of reinnervation are maintained long-term \[25\]. The results of ansa-RLN anastomosis may take 3–6 months to show evidence of effect. The time delay for reinnervation can be palliated by performing injection laryngoplasty at the same time as the reinnervation procedure. Reinnervation negates any chance of spontaneous recovery; therefore, it should be considered only after an observational period. LEMG should be performed in the preoperative assessment period, as any detection of activity should steer the surgeon away from reinnervation and toward a more temporary treatment (injection laryngoplasty). Because the laryngeal skeleton is not altered, additional phonosurgery is not compromised if needed later in life. Choosing between a medialization thyroplasty and a reinnervation procedure, as both are long-term solutions, depends on the individual case and the experience of the surgeon.

CONCLUSION

Evaluation and treatment of the child with UVCP requires an understanding of the diverse causes, aerodigestive sequelae, various diagnostic choices, and surgical options. As diagnostic techniques continue to improve, physicians and their patients will have more choices to evaluate the problem without causing significant morbidity to the patient. Surgical variety is likewise improving, allowing a broader group of patients to be treated.

Acknowledgements

None.

Conflicts of interest

There are no conflicts of interest.

REFERENCES AND RECOMMENDED READING

Papers of particular interest, published within the annual period of review, have been highlighted as:

- of special interest
- of outstanding interest

Additional references related to this topic can also be found in the Current World Literature section in this issue (p. 543).
