IAPO-Interamerican Association of
Pediatric Otorhinolaryngology

Vocal Fold Immobility in the Pediatric Patient

Ellen M. Friedman and Tulio A. Valdez

Introduction

Vocal fold immobility (VFI) has been recognized as the second most common cause of stridor in children after laryngomalacia accounting for 10% of all congenital anomalies affecting the larynx [1]. Vocal fold immobility can be divided into unilateral or bilateral and congenital or acquired. The etiology, presentation and management of VFI in a child differ from those in adults and it often varies with the age of the patient. We will describe our approach to diagnosis and management of bilateral and unilateral vocal fold immobility in neonates and children.

Background

Technological limitations in the past made the diagnosis of vocal cord paralysis difficult. Flexible laryngoscopy has allowed otolaryngologists to make this diagnosis more accurately without the need of taking the patient to the operating room. The ability to digitally record and play back in slow motion has increased our ability to make this diagnosis and follow the course.

Murty et al. estimate the incidence of congenital bilateral vocal cord paralysis at 0.75 cases per million births per year [2]. There has been no association found with respect to gender [3-6] but there have been familial and genetic associations [7-10].

Etiology

The causes of vocal cord immobility in children are varied. These include neurologic, iatrogenic, inflammatory, and trauma among other etiologies similar to adults. However, when compared to the adult population, the presence of idiopathic or neurological conditions is higher among children [3-6].

In the neonate with VFI a complete examination is necessary with special emphasis to the central nervous system and the cardiovascular system. Arnold Chiari malformation (ACM) is the most common CNS anomaly associated with VFI. Although it is most commonly associated with bilateral VCP, unilateral cases have been reported. Any infant born with an ACM and a high-pitch, inspiratory stridor should be investigated for bilateral VFI. Arnold Chiari malformation Type II is implicated more frequently [11-15]. Children with ACM often have concomitant central respiratory problems and in some cases may continue to experience apneas and other respiratory difficulties even after a tracheotomy is performed [12-14].

Other less frequent CNS causes of VFI include leukodystrophy, amyotrophic lateral sclerosis, encephalocele, kernicterus, hydrocephalus, and cerebral or nuclear dysgenesis [16-22].

Peripheral nervous system anomalies may also affect vocal cord function in children. Conditions such as Myasthenia gravis, Myotonic dystrophy, Werdnig-Hoffman disease (i.e., infantile muscular atrophy), fascioscapulohumeral myopathy, benign congenital hypotonia, and Charcot-Marie-Tooth disease also have been implicated VFI [15-20].

Acquired iatrogenic nerve injury was not a common etiology in the pediatric population, however the significant improvement in the care of the premature infant has resulted on an increased number of surgical procedures involving patent ductus arteriosus as well as congenital heart and great vessels conditions putting the vagus or recurrent laryngeal nerve at risk. Zbar and Smith reported a frequency of 8.8% of left VFI after patent ductus arteriosus ligation [23]. The suspicion for this mechanism of injury should be high in patients that present with stridor or hoarseness following a thoracic or cervical surgical procedure. In these cases VFI is usually unilateral. New surgical techniques such as vagus nerve stimulators have also been associated with vocal cord immobility [24]. Trauma from endotracheal intubation and laryngeal masks have also been reported as a causes of vocal cord paralysis [25-28]. Trauma sustained during birth with forceps delivery or neck torsion may result in VFI. Vocal cord paralysis can also be the result of child abuse or head trauma [29-30]. Neoplastic conditions are a rare cause of VFI in children.

Evidence of hereditary VFI has been reported but is extremely uncommon. All cases reported have involved both cords. Cases of Familial VFI have been described in X-linked, autosomal recessive and autosomal dominant inheritance patterns [7-10].

Idiopathic paralysis ranks as the first or second most common cause of congenital VFI in children and accounts for 36% to 47% of the cases in a variety of series [3-6].

Clinical Presentation

Congenital bilateral VFI often manifests early in life .Cohen et al. [4] reported 100 children with VFI, 58% of whom presented within the first 12 hours of birth.

In the pediatric population, VFI may be part of a multi-systemic anomaly and may be associated with central nervous system (CNS) malformations or other congenital anomalies involving cardiovascular and pulmonary malformations [4-6]. There is a high association of VFI with other laryngeal malformations such as clefts or stenosis [2-5].

The most common manifestation of bilateral VFI in children is biphasic stridor. Voice, respiration, or deglutition may also be abnormal in the pediatric patient with VFI. The signs and symptoms may vary from a clinically subtle presentation, as it is often the case in unilateral fold immobility, to severe with significant respiratory distress requiring immediate endotracheal intubation. Respiratory distress is more severe in cases of bilateral VFI. Loud biphasic stridor, cyanosis, and apnea are common findings in patients with bilateral VFI. In contrast, voice and cry may be virtually normal in children with bilateral VFI. Other symptoms such as aspiration, recurrent pneumonia, dysphonia, feeding difficulties, poor ability to cough and an abnormal cry are more often associated with unilateral VFI.

Diagnostic Evaluation

The evaluation of a child with suspected VFI requires a thorough history and physical examination. The focus of the examination should include an understanding of the timing and severity of the stridor, the quality of the voice or cry, and the presence of any associated feeding difficulties. The initial physical examination centers on the respiratory effort such as intercostal retractions, stridor, and respiratory rate. A quick survey for the stigmata of other congenital anomalies, such as myelomeningoceles present in Chiari II malformation, should be performed. Associated medical conditions or congenital anomalies, such as neurologic disorders or congenital heart disease, must be investigated.

With the introduction and wide availability of flexible endoscopic equipment, flexible laryngoscopy has become the best tool for diagnosis of pediatric VFI. The vast majority of children with VFI are diagnosed accurately using this technique. Nevertheless, flexible laryngoscopy can be challenging in the very young child with copious secretions, rapid respiration, frequent oxygen desaturations or associated laryngomalacia. In such cases, there is a role for direct laryngoscopy and ultrasound. The ability to record and play back the flexible laryngoscopy exam gives us the opportunity review and slow down the exam improving our ability to make a correct diagnosis. Conditions such as paradoxical vocal fold movement of the neonate, which can mimic a bilateral fold immobility had been unknown to most otolaryngologist until we had the ability to digitally record and play back in slow motion our examinations [31].

The diagnosis of VFI in a child should be followed by a search for the underlying cause. Clues in the history such as birth trauma, recent PDA ligation or hereditary causes may help with the diagnosis. The work up in an infant should always include an MRI of the brain and brain stem to rule out congenital anomalies such as ACM.

In patients in which, the history and physical exam give no clues of a possible etiology, a complete radiologic evaluation of the entire course of the vagus and recurrent laryngeal nerve should be pursued. MRI offers great definition of brain and brain stem anomalies without delivering the amount of radiation required for a CT scan.

Evaluation of the swallowing function is warranted once the diagnosis of vocal cord paralysis is made. This can be performed with FEES or a FEEST or with traditional techniques such as a barium swallow. A barium study may also document associated mediastinal anomalies, such as a vascular ring.

Rigid laryngoscopy and bronchoscopy continue to play a significant role in the identification of etiologies of VFI. Direct laryngoscopy is a critical part of evaluation in cases where endotracheal intubation preceded VFI. Unilateral or bilateral VFI must be differentiated from cricoarytenoid fixation, cricoarytenoid subluxation or posterior glottic stenosis, which can mimic VFI. Direct laryngoscopy allows close visual inspection and palpation of the arytenoid cartilage and the posterior glottis.

Laryngeal electromyography, which is commonly used in adults, has been difficult to implement in the pediatric population [32-34]. Currently research is being

performed using EMG to evaluate for recovery of fold motion and to distinguish from vocal fold fixation in pediatric patients.

Management

The management goal of VCP in a child is to restore and maintain an adequate balance of all laryngeal functions. Factors determining the treatment algorithm are directly related to the etiology, prognosis and coexisting comorbidities. We believe that the otolaryngologist should have three goals: the maintenance of a safe and stable airway, preservation of intelligible speech, and swallowing without aspiration.

Management should be tailored to the individual patient based on the etiology of the paralysis, the prognosis and the associated comorbidities. Many authors suggest that children with a meningomyelocele, ACM, and bilateral VCP should not undergo an invasive airway procedure, such as tracheotomy, until a ventricle-peritoneal shunt or posterior fossa decompression procedure is performed to see if the vocal fold function improves [2-6].

Spontaneous resolution of unilateral or bilateral VCP is a more frequent occurrence in the pediatric population. The reported rate of recovery varies within the literature from 16 to 64 percent [2-5]. Recovery has been noted from 6 weeks to 5 years after the initial diagnosis [2-6].

Management of Unilateral Vocal Cord Paralysis

The management of unilateral VCP in children differs greatly from management in adults in several aspects. Unilateral VCP usually results in a weakened cry, an adequate airway, intelligible speech and minimal aspiration. Children adjust well to persistent, unilateral VCP with little sequelae. There is the possibility of spontaneous recovery from VCP, thus obviating the need for surgery [36-39]. Additional stresses such as trauma, vigorous activity, or an upper respiratory infection (URl) may not be well tolerated however. Nevertheless, surgical intervention rarely is required in cases of unilateral VCP in infants or children. In the review of 127 cases of pediatric unilateral VCP by Narcy et al, only 6 underwent surgical intervention [38]. Management should be based on the age of the patient and its symptoms. In the neonate with unilateral VCP a conservative approach is warranted. It is uncommon for a neonate with unilateral VCP to require a tracheotomy unless there are other significant associated upper airway anomalies or evidence of chronic aspiration [6,4,39,40].

In children above 1 year of age, the initial management should be conservative in the early part of childhood. Surgical management may be warranted in children with persistent dysphonia or with signs of aspiration. It is unclear what is the ideal duration of the observation and less clear at which age it is safe to intervene. There are very few descriptions of the surgical management of unilateral VFI in the pediatric population since most children do well without surgery. Sipp et al [40] reported in 2007 a series of 15 pediatric patients with unilateral vocal fold immobility requiring medialization techniques including injection laryngoplasty, thyroplasty and ansa cervicalis-recurrent laryngeal nerve reinnervation with good voice and swallowing outcomes [40].

Bilateral VCP

Children with bilateral VCP often present with respiratory distress accompanied with biphasic stridor and chest retractions. The initial management is to provide a secure airway and to elucidate the etiology of the paralysis. Most physicians agree that tracheotomy is necessary in over 50 percent of these cases [3-6]. Miyamoto et al recently reported a retrospective study with a mean follow up of 50 months in which 15 of 22 patients (68%) with bilateral vocal cord paralysis required tracheotomy [41]. De Gaudemar et al. followed fifty-two infants and children with bilateral VCP for an average of 4 years; only 19 % required tracheotomy [42].

Tracheotomy is usually the initial surgical procedure of choice in patients with bilateral vocal cord paralysis with a compromised airway. Tracheotomy is a potentially reversible procedure, which allows time for potential spontaneous laryngeal functional recovery. It maintains a stable airway and allows easy reevaluation of vocal cord function. After a tracheotomy is performed, the patient should be evaluated every 1-2 months for the possibility of return of laryngeal function. Approximately 50 percent of children requiring a tracheotomy for VFI will remain with their tracheotomies for more than 3 years [5,6,59]. Waiting at least 12 months before embarking on a more permanent procedure for VFI is recommended to allow for possible spontaneous laryngeal recovery. Once it is determined that enough time has been allowed for the recovery of vocal cord function, surgical techniques to achieve decannulation can be attempted.

Surgical Procedures to Achieve Decannulation

The goal of surgical management with these procedures is to achieve decannulation by widening of the glottis. The surgeon must balance voice, deglutition and airway patency issues. The family must be made fully aware of this trade-off prior to consenting for surgery. In adults various surgical procedures including external have been proposed such as, cordectomy or arytenoidectomy through various external approaches, including a lateral cervical approach, translaryngeal approach, or laryngofissure [43-47]. The use of external approaches is further complicated in children due to the small size of the airway and the potential for airway scarring.

The introduction of the carbon dioxide laser in the early 1970's opened the possibility of endoscopic management of laryngeal conditions. $C0_2$ laser was first described for the management of bilateral VFI by Eskew and Bailey in 1983 [48]. Ossoff et al. were the first to report the clinical application of this technique achieving successful deccanulation in 10 of 11 adult patients with bilateral VFI [49]. Minimal dysphonia was noted after arytenoidectomy in these patients. Laser arytenoidectomy and cordectomy have proven to be excellent techniques in adults however they are not as equally effective in children [49-56]. Ossoff noted a higher rate of late failures in children compared to adults because of increased formation of scar tissue and the smaller glottic diameter in children [50]. It is difficult to achieve the proper balance between patency and voice issues in children where the overall size of the larynx is significantly smaller than in the adult. Other endoscopic CO_2 laser procedures such as vocal process resection, first described in children by Bigenzahn and Hoefler have shown promising results [53]. Friedman

et al. have used the CO_2 laser to resect a triangular portion of the posterior true and false vocal cord extending the endolaryngeal incision laterally to the level of the thyroid cartilage [54]. The incision is made immediately anterior to the vocal process with approximately 1/4 to 1/3 of the posterior vocal cord removed allowing improved respiratory function with minimal change in voice quality, and no reported problems with aspiration.

Bower et al reviewed the surgical treatment results of 30 children with bilateral VFI [55]. Decannulation was successful in 84 percent of the children after an external laryngeal approach compared with 56 percent of these undergoing laser arytenoidectomy. In a recent study by Hartnick et al [56] vocal cord lateralization procedures combined with a partial arytenoidectomy achieved the highest decannulation rate (17/24 or 71%) compared CO_2 laser cordotomy or arytenoidectomy procedures.

References

1. Dedo DD. Pediatric vocal cord paralysis. Laryngoscope. 1979;89:1378-1384.
2. Murty GE, Shinkwin C, Gibbin KP. Bilateral vocal fold paralysis in infants: tracheostomy or not? J Laryngol Otol. 1994;108:329-331.
3. Rosin DF, Handler SD, Potsic WP, Wetmore RF, Tom LW. Vocal cord paralysis in children. Laryngoscope. 1990;100:1174-1179.
4. Cohen SR, Geller KA, Birns JW, Thompson JW. Laryngeal paralysis in children a long-term retrospective study. Ann Otol Rhinol Laryngol. 1982;91:417-424.

4. Rothschild MA, Bratcher GO. Bilateral vocal cord paralysis and the pediatric airway. In: Myer CM, Cotton RT, eds. The Pediatric Airway: An Interdisciplinary Approach. Philadelphia, PA: Lippincott Williams & Wilkins; 1994:133-150.

5. Grundfast KM, Harley E. Vocal cord paralysis. Otolaryngol Clin North Am. 1989;22:569-597
6. Grundfast KM, Milmoe G. Congenital hereditary bilateral abductor vocal cord paralysis. Ann Otol Rhinol Laryngol. 1982;91:564-566.
7. Gacek RR. Hereditary abductor vocal cord paralysis. Ann Otol Rhinol Laryngol. 1976;85:90-93.
8. Isaacson G, Moya F. Hereditary congenital laryngeal abductor paralysis. Ann Otol Rhinol Laryngol. 1987;96:701-704.
9. Tucker HM. Congenital bilateral recurrent nerve paralysis and ptosis: a new syndrome? Laryngoscope. 1983;93:1405-1407.
10. Kirsch WM, Duncan BR, Black FO, Stears JC. Laryngeal palsy in association with myelomeningocele, hydrocephalus and the Arnold-Chiari malformation. J Neurosurg. 1968;28:207-214.
11. Oren J, Kelly DH, Todres ID, Shannon DC. Respiratory complications in patients with myelodysplasia and Arnold-Chiari malformation. Am J Dis Child. 1986;140:221-224.

11. Papasozomenos S, Roessmann U. Respiratory distress and Arnold-Chiari malformation. Neurology. 1981;31:97-100.
12. Sieben RL, Hamida MB, Shulman K. Multiple cranial nerve deficits associated with the Amold-Chiari malformation. Neurology. 1971;21:673-681.
13. Bluestone CD, Delerme AN, Samuelson GH. Airway obstruction due to vocal cord paralysis in infants with hydrocephalus and meningomyeloce. Ann Otol Rhinol Laryngol. 1972;81:778-783.
14. Gentile RD, Miller RH, Woodson GE. Vocal cord paralysis in children 1 year of age and younger. Ann Otol Rhinol Laryngol. 1986;95:622-625.
15. Fitzsimmons JS. Laryngeal stridor and respiratory obstruction associated with meningomyelocele. Arch Dis Child. 1965;40:687-688.
16. Zager EL, Ojemann RG, Poletti CE. Acute presentations of syringomyelia. Report of three cases. J Neurosurg. 1990;72:133-138.
17. Willis WH, Weaver DF. Syringomyelia with bilateral vocal cord paralysis. Report of a case. Arch Otolaryngol. 1968;87:468-470.
18. Holinger PH. Clinical aspects of congenital anomalies of the larynx, trachea, bronchi and oesophagus. J Laryngol Otol. 1961;75:1-44.
19. Stuart WD. The otolaryngologic aspects of myasthenia gravis. Laryngoscope. 1965;75:112-121.
20. Fairley JW, Hughes M. Acute stridor due to bilateral vocal fold paralysis as a presenting sign of myasthenia gravis. J Laryngol Otol. 1992;106:737-738.
21. Zbar RIS, Smith RJH, Vocal fold paralysis in infants twelve months of age and younger. Otolaryngol Head Neck Surg. 1996;114:18-21.
22. Shaffer MJ, Jackson CE, Szabo CA, Simpson CB. Vagal nerve stimulation: clinical and electrophysiological effects on vocal fold function. Ann Otol Rhinol Laryngol. 2005;114:7-14.
23. Nuutinen J, Karja J. Bilateral vocal cord paralysis following general anesthesia. Laryngoscope. 1981;91:83-86.
24. Brandwein M, Abramson AL, Shikowitz MJ. Bilateral vocal cord paralysis following endotracheal intubation. Arch Otolaryngol Head Neck Surg. 1986;112:877-882.
25. Cavo JW Jr. True vocal cord paralysis following intubation. Laryngoscope. 1985;95:1352-1359.
26. Inomata S, Nishikawa T, Suga A, Yamashita S. Transient bilateral vocal cord paralysis after insertion of a laryngeal mask airway. Anesthesiology. 1995;82:787-788.
27. Myer CM 3rd, Fitton CM. Vocal cord paralysis following child abuse. Int J Pediatr Otorhinolaryngol. 1988;15:217-220.
28. Pfenninger J. Bilateral vocal cord paralysis after severe blunt head injury--a cause of failed extubation. Crit Care Med. 1987;15:701-702.
29. Omland T, Brøndbo K. Paradoxical vocal cord movement in newborn and congenital idiopathic vocal cord paralysis: two of a kind?Eur Arch Otorhinolaryngol. 2008 Jul;265(7):803-807

30. Jacobs IN, Finkel RS. Laryngeal electromyography in the management of vocal cord mobility problems in children. Laryngoscope. 2002 Jul;112(7 Pt 1):1243-1248
31. Berkowitz RG. Laryngeal electromyography findings in idiopathic congenital bilateral vocal cord paralysis. Ann Otol Rhinol Laryngol. 1996;105:207-212.
32. Wohl DL, Kilpatrick JK, Leshner RT, Shaia WT. Intraoperative pediatric laryngeal electromyography: experience and caveats with monopolar electrodes. Ann Otol Rhinol Laryngol. 2001;110:524-531.
33. Parnes SM, Satya-Murti S. Predictive value of laryngeal electromyography in patients with vocal cord paralysis of neurogenic origin. Laryngoscope. 1985; 95:1323-1326.
34. Swift AC, Rogers J. Vocal cord paralysis in children. J Laryngol Oto1 1987;101:169-171.
35. Tucker HM. Vocal cord paralysis in small children: principles in management. Ann Otol Rhinol Laryngol. 1986;95:618-621.
36. Narcy P, Contencin P, Viala P. Surgical treatment for laryngeal paralysis in infants and children. Ann Otol Rhinol Laryngol. 1990;99:124-128.
37. Daya H, Hosni A, Bejar-Solar I, Evans JN, Bailey CM. Pediatric vocal fold paralysis: a long-term retrospective study. Arch Otolaryngol Head Neck Surg. 2000;126:21-25.
38. Sipp JA, Kerschner JE, Braune N, Hartnick CJ Vocal fold medialization in children: injection laryngoplasty, thyroplasty, or nerve reinnervation? Arch Otolaryngol Head Neck Surg. 2007 Aug;133(8):767-71
39. Miyamoto RC, Parikh SR, Gellad W, Licameli GR.Bilateral congenital vocal cord paralysis: a 16-year institutional review. Otolaryngol Head Neck Surg. 2005 Aug;133(2):241-5 de Gaudemar I, Roudaire M, Francois M, Narcy P. Outcome of laryngeal paralysis in neonates: a long-term retrospective study of 113 cases. Int J Pediatr Otorhinolaryngol. 1996;34:101-110.
40. Gardner GM, Altman JS, Balakrishnan G. Pediatric vocal fold medialization with silastic implant: intraoperative airway management. Int J Pediatr Otorhinolaryngol. 2000;52:37-44.
41. Woodman DG. A modification of the extralaryngeal approach to arytenoidectomy for bilateral abductor paralysis. Arch Otolaryngol. 1946;43:63-65.
42. Amedee RG, Mann WJ. A functional approach to lateral fixation in bilateral abductor cord paralysis. Otolaryngol Head Neck Surg 1989;100:542-545.
43. Thornell WC. Intralaryngeal approach in arytenoidectomy in bilateral abductor vocal cord paralysis. Trans Am Acad Ophthalmol Otolaryngol. 1949;53:631-635.
44. Downey WL, Kennon WG Jr. Laryngofissure approach for bilateral abductor paralysis. Arch Otolaryngol. 1968;88:513-517.
45. Eskew JR, Bailey BJ. Laser arytenoidectomy for bilateral vocal paralysis. Otolaryngol Head Neck Surg. 1983;91:294-298.
46. Ossoff RH, Sisson GA, Duncavage JA, Moselle HI, Andrews PE, McMillan WG. Endoscopic laser arytenoidectomy for the treatment of bilateral vocal cord paralysis. Laryngoscope. 1984;94:1293-1297.

47. Ossoff RH, Duncavage JA, Shapshay SM, Krespi YP, Sisson GA Sr. Endoscopic laser arytenoidectomy revisited. Ann Otol Rhinol Laryngol. 1990;99:764-771.
48. Lim RY. Laser arytenoidectomy. Arch Otolaryngol. 1985;111:262-263.
49. Crumley RL. Endoscopic laser medial arytenoidectomy for airway management in bilateral laryngeal paralysis. Ann Otol Rhinol Laryngol. 1993;102:81-84.
50. Holm AF, Wonters B, Van Averbeek JJM. CO2 laser cordectomy for bilateral vocal cord paralysis. Lasers Med Science. 1989;4:93-96.
51. Eckel HE, Thumfart M, Wassermann K, Vossing M, Thumfart WF. Cordectomy versus arytenoidectomy in the management of bilateral vocal cord paralysis. Ann Otol Rhinol Laryngol. 1994;103:852-857.
52. Dennis DP, Kashima H. Carbon dioxide laser posterior cordectomy for treatment of bilateral vocal cord paralysis. Ann Otol Rhinol Laryngol. 1989;98:930-934.
53. Bigenzahn W, Hoefler H. Minimally invasive laser surgery for the treatment of bilateral vocal cord paralysis. Laryngoscope. 1996;106:791-793.
54. Friedman EM, de Jong AL, Sulek M. Pediatric bilateral vocal fold immobility: the role of carbon dioxide laser posterior transverse partial cordectomy. Ann Otol Rhinol Laryngol. 2001;110:723-728.
55. Bower CM, Choi SS, Cotton RT. Arytenoidectomy in children. Ann Otol Rhinol Laryngol. 1994;103:271-278.
56. Hartnick CJ, Brigger MT, Willging JP, Cotton RT, Myer CM 3rd.Surgery for pediatric vocal cord paralysis: a retrospective review. Ann Otol Rhinol Laryngol. 2003;112:1-6.