Pediatric Vocal Fold Paralysis

A Long-term Retrospective Study

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Objective: To review our experience of pediatric vocal fold paralysis (VFP), with particular emphasis on etiological factors, associated airway pathologic conditions, and treatment and prognostic outcomes.

Design: Retrospective case review of a cohort of patients presenting with VFP.

Setting: Tertiary referral center.


Results: There was an almost equal distribution of unilateral (52% [n = 53]) and bilateral (48% [n = 49]) VFP. Iatrogenic causes (43% [n = 44]) formed the largest group, followed by idiopathic VFP (35% [n = 36]), neurological causes (16% [n = 16]), and finally birth trauma (5% [n = 5]). Associated upper airway pathologic conditions were noted in 66% (n = 23) of patients who underwent tracheotomy. Tracheotomy was necessary in only 57% (n = 28) of children with bilateral VFP. Prognosis was variable depending upon the cause, with neurological VFP having the highest rate of recovery (71% [5/7]) and iatrogenic VFP the lowest rate (46% [12/26]).

Conclusion: Recovery after an interval of up to 11 years was seen in idiopathic bilateral VFP; this has significant implications when considering lateralization procedures in these patients.


Vocal fold paralysis (VFP) is the absence of movement of the vocal folds following dysfunction of the motor nerve supply to the larynx. Presenting symptoms in children include stridor, a weak cry or voice, feeding difficulties, and aspiration. It is the second most common cause of neonatal stridor and has been reported to account for 10% of all congenital anomalies affecting the larynx.1

Vocal cord paralysis may be unilateral or bilateral. Stridor is the most common presenting symptom and is often the reason for the failure of neonates to wean from nasal continuous positive airway pressure therapy. In bilateral cases, the stridor is usually more severe; in the past, tracheotomy has traditionally been recommended for these patients. Dysphonia is not a typical feature of bilateral VFP, since most cases are abductor palsies, with the folds in close apposition to each other. In contrast, dysphonia is usually found in unilateral VFP.

There are a number of causes of VFP, including iatrogenic, neurological disorders, birth trauma, and idiopathic. Diagnosis depends on careful evaluation of the larynx by an experienced pediatric otolaryngologist. This may be performed by direct laryngoscopy with the patient under general anesthesia, by dynamic assessment of the larynx during the lightening of anesthesia, or by flexible fiberoptic endoscopy; however, the last technique may not identify other associated airway pathologic conditions. Ultrasonography of the larynx is a useful adjunctive examination that we now use routinely.2

The treatment of VFP depends primarily on a patient’s symptoms, particularly the extent of airway compromise. The presence of associated laryngeal pathologic conditions is an important consideration. Patterns do emerge, however, depending on the type of VFP and the cause. Greater morbidity is generally seen in bilateral VFP, but, interestingly, in idiopathic bilateral VFP, recovery is more likely.

In this review, we present a comprehensive summary of 102 consecutive cases of VFP seen at Great Ormond Street Hos-
RESULTS

One hundred two cases of VFP were identified over a 14-year period. There was an almost equal distribution of bilateral (n = 49) and unilateral (n = 53) cases; the majority of unilateral cases (77% [n = 41]) were left-sided. Most patients (68% [n = 69]) were diagnosed before age 12 months; of these, 65% (n = 45) presented with symptoms at birth.

PRESENTING SYMPTOMS

The most common presenting symptom was stridor (86% [n = 88]), present in 96% (47/49) of patients with bilateral VFP and 77% (41/53) of patients with unilateral VFP. An abnormal cry or dysphonia was noted more often in patients with unilateral VFP (51% [27/53]) than bilateral VFP (2% [1/49]). Feeding difficulty was also more common in unilateral (23% [12/53]) compared with bilateral (4% [2/49]) VFP (Table 1). Cyanosis, severe chest retraction, and apneas were also observed, but these occurred predominantly in patients with coexisting cardiac or neurological anomalies.

CAUSES OF VFP

Birth Trauma

Five patients had symptoms of VFP presenting at birth and a documented history of birth trauma. All of these patients had bilateral VFP and most (80% [4/5]) resulted from forceps delivery. Tracheotomy was necessary in 1 of these patients.

Iatrogenic Causes

Of the 44 patients in this group, 39 had unilateral VFP and 5 had bilateral VFP. Cardiac surgery was the most common cause of VFP, resulting in 33 cases. As expected, the majority of these (88% [n = 29]) were patients with left-sided VFP; in 11 patients VFP occurred following patent ductus arteriosus ligation.

Mediastinal surgery caused 3 cases of VFP, all of which were unilateral. Three more cases of unilateral VFP occurred after neck surgery, and 3 cases of VFP occurred after tracheoesophageal fistula repair, 2 of which were unilateral and 1 bilateral. In 1 patient, VFP occurred after the surgical removal of an epidermoid; in another, unilateral VFP developed after a Girdlestone procedure.

Neurological Group

Among the 16 patients in this group, Arnold Chiari malformation was the most common condition and occurred in 7 patients (44%). In all 7 of these cases, it caused bilateral VFP.

Other central neurological causes (n = 7) included severe hypoxia (n = 2), corpus callosum agenesis (n = 2), congenital hydrocephalus (n = 2), and neurofibromatosis (n = 1). Except for 1 case of corpus callosum agenesis that resulted in unilateral left-sided VFP, the remaining central neurological causes all resulted in bilateral VFP.

There were 2 patients with presumed peripheral neurological disease, one with hereditary distal spinal muscular degeneration and the other with Horner syndrome affecting the same side as the VFP. Both of these patients had unilateral VFP.

Idiopathic

For 36 patients, no specific etiological factors could be identified. Among these, 26 patients had bilateral VFP and 10 had unilateral VFP. Twenty-eight patients (78%) presented with symptoms from birth; in 7 (19%), symptoms developed within the first 3 months. One child developed bilateral VFP at age 5 years.

Miscellaneous

One child had an aberrant innominate artery causing right-sided tracheal compression and a right-sided VFP.
Associated Upper Airway Disease

Associated upper airway disease was found in 46 patients. Ten patients had more than one associated upper airway pathologic condition.

<table>
<thead>
<tr>
<th>Associated Upper Airway Disease</th>
<th>No. of Patients</th>
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<tbody>
<tr>
<td>Laryngomalacia</td>
<td>17</td>
</tr>
<tr>
<td>Tracheobronchomalacia</td>
<td>16</td>
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<tr>
<td>Subglottic stenosis</td>
<td>13</td>
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<tr>
<td>Intubation granulomas</td>
<td>5</td>
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<tr>
<td>Cricothyroid joint fixation</td>
<td>3</td>
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<tr>
<td>Laryngeal web</td>
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*Ten patients had more than one associated upper airway pathologic condition.*

The common associated findings were laryngomalacia, tracheobronchomalacia, and subglottic stenosis. In 10 of these patients, more than 1 associated pathologic condition was present.

SURGICAL TREATMENT

Forty-four patients underwent surgery to treat their symptoms. Thirty-five patients underwent tracheotomy; 28 (80%) of these patients had bilateral VFP and 7 (20%) had unilateral VFP.

In 2 patients with unilateral VFP, however, tracheotomy was performed for associated conditions, including chronic lung disease and severe obstructive sleep apnea secondary to micrognathia. Thirteen patients (36%) from the idiopathic group required tracheotomy. All of these patients had bilateral VFP, and 9 had associated upper airway disease. One patient with a tracheotomy died after his tube became displaced.

Of the 43 patients who developed VFP following surgery, 10 (23%) required tracheotomy; 6 had unilateral VFP and 4 had bilateral VFP. Seven of these patients had previously undergone cardiac surgery; 3 had bilateral VFP and 4 had unilateral VFP. The unilateral cases all had associated subglottic stenosis or tracheomalacia.

Two patients in the noncardiac iatrogenic group required tracheotomy after developing VFP following tracheoesophageal repair. One of these patients had unilateral VFP, associated tracheomalacia, and intubation granulomas. The other child had bilateral VFP and associated intubation granulomas. Another patient who had not undergone cardiac surgery developed left VFP following a Girdlestone procedure. Her tracheotomy, however, was performed because of severe obstructive sleep apnea.

Eleven (69%) of the children in the neurological diseases group required tracheotomy. However, 10 (62%) of these patients had associated upper airway abnormalities, symptoms of reflux, or sleep apnea.

Five (71%) of the patients with Arnold Chiari malformation required tracheotomy. All of these patients had bilateral VFP, and 3 of them had associated upper airway disease. Six patients with different neurological diseases also required tracheotomy. Five had bilateral VFP and 4 had associated upper airway disease. The other patient with bilateral VFP had a tracheotomy performed for severe aspiration. The patient with unilateral VFP had a tracheotomy performed for chronic lung disease.

Six patients underwent other surgical procedures to treat their symptoms. Two patients with bilateral abductor palsy underwent arytenoidectomy, 1 at age 13 years and the other, who was dependent on tracheotomy, at age 8 years. This patient underwent successful decannulation following arytenoidectomy. No follow-up records were available for the first patient.

Three patients had medialization procedures. Two underwent Teflon injection; both had unilateral left VFP following cardiac surgery. One of these patients underwent the procedure 18 months after surgery and the other 5 years after surgery. A successful outcome with an improvement in voice was seen in one patient. The other developed a granuloma following Teflon injection that required surgical removal. There was no documentation regarding voice outcome in this patient.

Thyroplasty with a Silastic implant was performed in a child with unilateral left-sided VFP following tracheoesophageal fistula repair. This was performed at age 3 years, with minimal improvement in voice quality.

One child with bilateral VFP underwent a Tucker reinnervation procedure at age 9 years, with subsequent recovery of left vocal fold function. The right vocal fold had recovered spontaneously at age 6 years. Decannulation was possible 4 months after surgery, and a significant improvement in his voice was noted.

There were 3 patients with both VFP and laryngomalacia who underwent aryepiglottoplasty.

FOLLOW-UP

At our hospital, we recommend that patients with newly diagnosed VFP return for follow-up every 3 months for the first year, every 6 months for the second year, and annually thereafter. Those patients with tracheotomies usually undergo endoscopic examination with dynamic assessment under general anesthesia at the same intervals. Follow-up records with specific reference to outcome of VFP, either by clinical impression or endoscopic examination, were available for 65 (64%) of the 102 patients included in our study (Table 2). If examination was not possible because of difficulties in assessing the infant larynx, then a clinical assessment of symptom improvement was undertaken. The range of follow-up extended from 3 months to 13 years. Recovery was noted in 36 (56%) of the 64 patients with follow-up records reporting VFP outcome; of these 36 patients, 25 (69%) recovered within 2 years.

Patients with VFP secondary to neurological disease (n = 7) had the highest recovery rate (71% [5/7]); all 5 of these patients recovered within 2 years, and they all had bilateral VFP.

In the idiopathic group (n = 28), 18 patients (64%) recovered within a range of 6 months to 11 years. Within
4 years, 13 patients (72%) had recovered. Five patients with bilateral VFP (18%) took between 5 and 11 years to recover.

Of the patients with VFP secondary to surgery (n = 26), 12 (46%) recovered within 5 years.

There were 10 deaths, 1 of which was tracheotomy related. The remaining 9 deaths were non–airway related.

### CAUSES OF VFP

The most frequent cause of VFP in this study was surgery (43% [44/102]). Although similar figures were reported by Zbar and Smith, other series reported lower rates. This variation is probably institution-specific and related to the presence and activity of a pediatric cardiothoracic surgery service. The closure of a patent ductus arteriosus was the most common cause of VFP in this group, occurring in 11 (25%) of the 44 patients who underwent surgery. Because the left recurrent laryngeal nerve is highly susceptible to damage during surgery, this would account for the almost equal distribution of patients with bilateral and unilateral VFP in our study in contrast to the predominance of patients with bilateral VFP in other series.

Patients with VFP of idiopathic origin constituted the next most common group, with rates similar to those reported by Cohen et al and de Gaudemar et al. Interestingly, patients with bilateral VFP dominated this group (26/36 [72%]), a finding that has also been observed in other studies.

Neurological disease is an important factor in bilateral VFP, and all patients with VFP should have a thorough neurological assessment, including a magnetic resonance imaging scan. Arnold Chiari malformation is the most frequent neurological condition causing VFP, and it invariably results in bilateral VFP. Recovery from VFP following decompression has been reported, but we did not observe this in our group of patients.

Birth trauma resulted in fewer cases (5/102 [5%]) in our study compared with other series (approximately 20%). Our lower rate may be explained by the absence of an on-site maternity department. Within this group, it was surprising that all patients had bilateral VFP, since one would have expected unilateral VFP to be more likely. Although Emery and Fearon reported a predominance of left-sided VFP, other series have also noted a higher incidence of bilateral VFP. It is possible that these were cases of congenital idiopathic VFP and the history of birth trauma was simply coincidental.

### TREATMENT AND PROGNOSIS

Because of the nature of the referral sources at Great Ormond Street Hospital for Children, where many children come from far afield and some from overseas, local follow-up is encouraged. This is reflected in our lower follow-up rate (64% [63/102]) compared with other series (Emery and Fearon, 92%; de Gaudemar et al, 79%).

The treatment of VFP is determined by a patient's symptoms, particularly by the severity of airway obstruction. Before surgical treatment is considered, parents are advised to position the child so that he or she is sitting up and thicken the food. If gastroesophageal reflux is suspected, then this is also treated. In addition, all children with VFP are seen by a speech pathologist. An assessment of the child's ability to cope with VFP is made initially by observation and oxygen saturation monitoring and thereafter by observation alone if the child's condition is stable. No specific recommendations about restriction of activity are given, since we believe that the child will do this automatically.
When airway obstruction is caused by VFP, the severity may be exacerbated by coexisting upper airway disease, which in this series was present in 46 (45%) of 102 patients. Of the 35 children who required tracheotomy, 22 (63%) had associated upper airway disease. As expected, more tracheotomies were necessary in patients with bilateral VFP than in those with unilateral VFP. All the patients with unilateral VFP who required tracheotomy had associated upper airway disease or had tracheotomy performed for chronic lung disease or sleep apnea. Twenty-two (79%) of the 28 patients with bilateral VFP who required tracheotomy had associated upper airway disease. Although in adults tracheotomy is almost always indicated in bilateral abductor VFP, this does not seem to be the case in children. We found that tracheotomy was necessary in only 57% (28/49) of patients with bilateral VFP; this finding is similar to the experience reported in the series of Murty et al. When examining patients with VFP grouped according to cause, 4 (80%) of 5 patients with iatrogenic VFP and 10 (77%) of 13 patients with bilateral neurological VFP required tracheotomy compared with 13 (50%) of 26 patients with bilateral idiopathic VFP.

Prognosis was also variable depending on the cause of VFP. Patients with VFP secondary to neurological disease had the highest rate of recovery (71% [5/7]) and those with iatrogenic VFP the lowest (46% [12/26]) (Table 2). A prolonged recovery time was seen in patients with idiopathic VFP, with 5 (28%) of 18 patients who recovered showing evidence of recovery after 5 to 11 years. Although a high early recovery rate (within 6 months to 1 year) has been reported in other series, this was not our experience. We found that 36 (55%) of 65 patients for whom recovery follow-up information was reported recovered, with 16 (44%) of these 36 patients recovering within 1 year. Assessment of recovery was based on examination of the larynx in the outpatient setting, using either a flexible endoscope or indirect laryngoscopy. In the event that this was not possible, a clinical assessment of the child’s symptoms was used to determine whether recovery had occurred. Patients who underwent tracheotomy also underwent endoscopic examination under anesthesia. Laryngeal electromyography was not used as a diagnostic tool because of concerns regarding the morbidity associated with the procedure and because its role in pediatric VFP has yet to be established.

The decision to perform other surgical procedures to treat symptoms of VFP is difficult, since recovery from VFP in children may occur only after several years. This is different in adults, for whom mediationization procedures or arytenoidectomy may reasonably be recommended after 1 year. In children, particularly those with bilateral idiopathic VFP, we would recommend a conservative approach, avoiding procedures such as arytenoidectomy until adolescence.

This variation in prognosis and tracheotomy rate suggests that there are different pathophysiological processes occurring within the different etiological groups. Previous authors have tended to relate prognostic and treatment indications to the type of paralysis rather than the underlying cause. The decision to perform tracheotomy in patients with VFP is dependent upon the severity of airway obstruction. We found that bilateral VFP with an iatrogenic or neurological cause was more likely to require tracheotomy than bilateral idiopathic VFP. We also found that that prognosis was dependent upon the cause of the paralysis rather than whether it was unilateral or bilateral.

Interestingly, all patients with bilateral idiopathic VFP had vocal folds in the adducted position. Despite this, only 50% (13/26) of bilateral idiopathic VFP cases required tracheotomy. In those patients who did not require tracheotomy, there must have been sufficient adductor muscle function to maintain an airway. It has been postulated that the pathophysiologic characteristic of this condition is an imbalance of the muscle groups of the larynx, with increased activity of the adductor muscles. Perhaps this represents an immature state that resolves with maturity, explaining the high recovery rate within this group. Laryngeal electromyography, although difficult to perform on children, may shed more light on this hypothesis.

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REFERENCES