Applications of Flexible Bronchoscopy in Infants with Congenital Vocal Cord Paralysis: A 12-Year Experience

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**Background:** Congenital vocal cord paralysis (VCP) is a common cause of congenital stridor. Before the widespread application of flexible bronchoscopy (FB) by pediatricians, congenital stridor in infants was usually attributed to laryngomalacia. Prompt recognition and careful follow-up is crucial for the management of congenital VCP.

**Methods:** We performed a retrospective chart review of newborn infants with congenital VCP diagnosed by FB over a 12-year period.

**Results:** During the 12-year period, FB was performed on a total of 356 infants. Fifteen (4%) infants were diagnosed with congenital VCP. There were eight males and seven females and the mean age at diagnosis was 76.6 days. Stridor with respiratory distress was the most prominent presenting symptom. The majority (93%, 14/15) demonstrated bilateral VCP, while one patient (7%) had unilateral VCP. Seven of the 15 (46%) patients had idiopathic VCP, while eight (54%) had VCP associated with neuromuscular disorders. Tracheotomy was necessary in four patients (26%). None of them underwent further surgical interventions. Spontaneous recovery occurred in 10 patients (71%), and of these, 90% (9/10) were treated without tracheotomy.

**Conclusion:** In order to allow prompt diagnosis of congenital VCP, FB should be performed in every newborn infant with stridor. Patients with congenital VCP should undergo additional imaging studies to detect any associated neurological abnormalities and intrathoracic comorbidities. The majority of patients can be managed conservatively and monitored carefully using serial FB. Corrective surgery should be reserved for those with a lack of resolution at prolonged follow-up, and those with significant comorbidities.

1. **Introduction**

Congenital vocal cord paralysis (VCP) is rare. Ungkanont et al and Murty et al estimated its annual incidence to be 0.75 cases per million births.¹,² Nonetheless, it is the second most common cause of neonatal stridor and accounts for 10% of congenital laryngeal anomalies.³,⁴ The etiologies of congenital VCP include anomalies of the central nervous system (CNS), birth trauma, and intrathoracic lesions.
but it is usually idiopathic. The initial presentations of congenital VCP vary from minor symptoms such as stridor, weak cry, feeding difficulties, and aspiration, to more severe problems including dyspnea, cyanosis, and respiratory failure.

Before the widespread application of flexible bronchoscopy (FB) by pediatricians, congenital stridor in infants was usually initially attributed to laryngomalacia, with a definite diagnosis of VCP being delayed and the optimal time for management therefore being missed. Today, with advanced instrumentation and anesthetic techniques, FB can be performed safely in small infants. Pediatric pulmonologists play an active role in the evaluation and follow-up of laryngeal pathologies. The currently available literature on VCP in infants is limited and demonstrates controversy regarding treatment strategies, surgical interventions and prognosis. We reviewed the literature, and the medical records of patients with congenital VCP treated at our hospital in order to determine the characteristics, comorbidities, optimal management strategies, and outcomes of such patients, with emphasis on the usefulness of pediatric FB.

2. Materials and Methods

During a 12-year period from 1995 to 2006, FB was performed on a total of 356 newborns and infants at the Children’s Hospital of Chang Gung Memorial Hospital (Kaohsiung, Taiwan), due to congenital stridor and respiratory distress. Prior to each examination, informed consent was obtained from the parents of each infant. All procedures were performed in the pediatric or neonatal intensive care units by pediatric pulmonologists. Patients with a diagnosis of VCP were included in this retrospective study.

An Olympus BF 3C20 bronchoscope (Olympus, Shinjuku-ku, Tokyo, Japan) with a distal outside diameter of 3.5 mm was used as the main exploratory tool for all procedures. The Olympus N20 with an outer diameter of 2.2 mm was also applied in selected cases. All the infants received a single dose of oral chloral hydrate (10%) 50 mg/kg for patient sedation. In infants older than 3 months, midazolam (5 mg/mL/amp) 0.1 mg/kg and fentanyl (0.5 mg/10 mL/amp) 2.5 μg/kg were added for sedation. Supplemental oxygen was given and the patient breathed spontaneously throughout the examination. Vital signs including blood pressure, heart rate, and oxygen saturation were monitored continuously. The bronchoscope was introduced into the larynx via the nasopharyngeal route, and motor function of the epiglottis, arytenoid cartilages, and vocal cords were carefully evaluated. The bronchoscope was then advanced to the trachea, carina, and segmental bronchial Airways. In patients with tracheotomies, the tube was occluded for a short period of time in order to observe laryngeal mobility. Bronchoalveolar lavage (BAL) fluid was obtained when possible, and was subjected to examination of cellular differentiation, cytolgy for lipid-laden macrophages, cultures for identification of bacteria and virus, and other laboratory investigations if necessary. Further additional investigations of the esophagus and stomach were performed simultaneously, using the same endoscope. All procedures were recorded and the videos were stored on compact discs. Case notes were reviewed and demographic data, clinical features, possible underlying causes of VCP, types of VCP, associated airway anomalies, courses of treatment, and outcomes were all recorded. Patients with VCP were divided into idiopathic and neurogenic groups, depending on etiology. Initial treatments were classified as tracheotomy or observation. The duration of the tracheotomy and resolution were evaluated. Recovery was confirmed by serial bronchoscopy.

3. Results

The bronchoscopic findings in 356 infants with congenital stridor were laryngomalacia (214/356, 60%), subglottic stenosis (34/356, 9.5%), VCP (15/356, 4.2%), tracheomalacia (18/356, 5%), tracheal stenosis (8/356, 2.2%), laryngeal cyst (5/356, 1.4%), and other miscellaneous causes.

Fifteen infants were identified with congenital VCP. There were eight males and seven females and the mean age at diagnosis was 76.6 days. Stridor was the presenting symptom in all 15 infants. Other associated presentations included dyspnea (n=6, 40%), choking and recurrent aspiration (n=2, 13%), and weak cry (n=1, 7%). The majority (14/15, 93%) of patients had bilateral VCP; only one patient (7%) had left vocal cord abduction paralysis. Among those with bilateral VCP, 12 (86%) had abduction paralysis and two (14%) had adduction paralysis (Figure 1, Table).

Coexistent laryngotracheal anomalies included subglottic stenosis (n=6, 40%), laryngomalacia (n=3, 20%), and laryngeal web (n=2, 13.3%). Sixty percent of the patients had other associated abnormalities in either the upper or lower Airways. Seventy-three percent (11/15) had congenital heart disease diagnosed by echocardiography. The most common cardiac anomalies were atrial septal defect (67%), followed by pulmonary stenosis, and patent ductus arteriosus (Figure 2).
Eight patients (8/15, 57%) with associated neurogenic abnormalities were included in the neurogenic group. Three patients had choroid plexus cysts, two patients had bouts of neonatal asphyxia with hypoxic-ischemic encephalopathy (HIE), two patients had ventriculomegaly, and patient 8 had Down’s syndrome and a white matter disorder (diagnosed by brain echography). Patient 15 had Prader-Willi syndrome. The seven (47%) patients without obvious causes of VCP were included in the idiopathic group (Table).

Gross reflux esophagitis was found in 47% (7/15) of patients. BAL fluids were positive for lipid-laden macrophages in seven of nine patients (78%). Patients with either of these findings were treated with oral antireflux medications including lansoprazole...

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**Table**  Clinical characteristics of patients with congenital vocal cord paralysis

<table>
<thead>
<tr>
<th>Causes</th>
<th>Patient</th>
<th>Gender</th>
<th>Paralysis type</th>
<th>Age at diagnosis (days)</th>
<th>Tracheotomy (age, days)</th>
<th>Spontaneous recovery (final follow-up age, months)</th>
<th>Neuromuscular disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>Idiopathic</td>
<td>1</td>
<td>F</td>
<td>Bil ab</td>
<td>320</td>
<td>No</td>
<td>Lost to follow-up</td>
<td></td>
</tr>
<tr>
<td></td>
<td>2</td>
<td>F</td>
<td>Bil ab</td>
<td>46</td>
<td>No</td>
<td>Yes (4)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>F</td>
<td>Bil ab</td>
<td>55</td>
<td>No</td>
<td>Yes (5.5)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>4</td>
<td>F</td>
<td>Bil ab</td>
<td>65</td>
<td>No</td>
<td>Yes (3)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>5</td>
<td>M</td>
<td>Bil ab</td>
<td>1</td>
<td>Yes (13)</td>
<td>No (6)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>6</td>
<td>M</td>
<td>Bil ab</td>
<td>86</td>
<td>No</td>
<td>Yes (7)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>7</td>
<td>F</td>
<td>Bil ab</td>
<td>50</td>
<td>Yes (110)</td>
<td>No (63)</td>
<td></td>
</tr>
<tr>
<td>Neurogenic</td>
<td>8</td>
<td>F</td>
<td>Bil ab</td>
<td>306</td>
<td>No</td>
<td>Yes (22) Down’s syndrome, white matter disorder</td>
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</tr>
<tr>
<td></td>
<td>9</td>
<td>M</td>
<td>L ab</td>
<td>84</td>
<td>No</td>
<td>Yes (3) Ventriculomegaly</td>
<td></td>
</tr>
<tr>
<td></td>
<td>10</td>
<td>F</td>
<td>Bil ad</td>
<td>23</td>
<td>No</td>
<td>No (19) Neonatal asphyxia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>11</td>
<td>M</td>
<td>Bil ab</td>
<td>10</td>
<td>Yes (44)</td>
<td>No (3) Choroid plexus cyst</td>
<td></td>
</tr>
<tr>
<td></td>
<td>12</td>
<td>M</td>
<td>Bil ab</td>
<td>51</td>
<td>No</td>
<td>Yes (4) Choroid plexus cyst</td>
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<tr>
<td></td>
<td>13</td>
<td>M</td>
<td>Bil ab</td>
<td>18</td>
<td>Yes (20)</td>
<td>Yes (10) Ventriculomegaly, choroid plexus cyst</td>
<td></td>
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<tr>
<td></td>
<td>14</td>
<td>M</td>
<td>Bil ab</td>
<td>15</td>
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<tr>
<td></td>
<td>15</td>
<td>M</td>
<td>Bil ad</td>
<td>26</td>
<td>No</td>
<td>Yes (3) Prader-Willi syndrome</td>
<td></td>
</tr>
</tbody>
</table>

M = male; F = female; L = left; Bil = bilateral; ad = adduction paralysis; ab = abduction paralysis.
(proton pump inhibitor) 1 mg/kg/day and metoclopramide (0.3 mg/kg/day, in three divided doses) for at least 2 weeks.

One patient was lost to follow-up after 5 years of conservative treatment. Spontaneous recovery was identified by serial FB in nine of 10 (90%) patients without tracheotomy (Table). In these patients, the mean age at recovery was 5.9 months (range 20 days to 1 year and 10 months). The overall spontaneous recovery rate was 71% (10/14). Tracheotomy was required for treatment in four patients (26%), two in the idiopathic group, and two in the neurogenic group. One patient (patient 13) in the neurogenic group was decannulated successfully at 10 months of age, while the other three patients could not be decannulated during follow-up for 3 months, 6 months, and 5 years, respectively.

4. Discussion

Among the 356 infants with congenital stridor included in our study, laryngomalacia was the commonest congenital laryngeal anomaly (60%). Subglottic stenosis was the second most common cause (9.5%). Congenital VCP was responsible for only 4.2% of stridor cases in our patients, but was still the third most common cause. Holinger and Brown found that VCP accounted for 10% of congenital laryngeal anomalies,4 while Wood’s study showed that VCP was responsible for stridor in 9.3% of 225 patients.5 One possible reason for the relatively low prevalence of stridor in our study was that some patients with unilateral VCP were not referred to our department for further evaluation, due to lack of significant stridor. This possibility was supported by the fact that there was only one case of unilateral VCP in our patient series. In Gentile’s review, stridor was present in all 12 patients with bilateral VCP, but only in seven of 10 patients with unilateral VCP.6 General practitioners should be acutely alert to the symptoms and signs of unilateral VCP, such as weak cry, swallowing disturbances, aspiration pneumonia, and hoarseness with a breathy, raspy quality.7

Before common application of FB by pediatric pulmonologists, congenital stridor in newborn infants was frequently attributed to laryngomalacia. In intractable cases, a definitive diagnosis depended on laryngoscopy performed by an otolaryngologist. This process had many flaws, and diagnosis and management were therefore sometimes delayed. FB is now a well-established technique used in pediatric respiratory practice allowing for the early diagnosis of many congenital upper and lower airway anomalies. Since multiple laryngeal anomalies can coexist, and because stridor can also result from tracheal abnormalities, we recommend FB in preference to laryngoscopy, as the gold standard for the evaluation of infants with congenital stridor. In view of possible comorbidities and subsequent treatment planning, FB is most appropriately performed by pediatricians in the intensive care unit or in offices with cardiovascular monitoring and oximeters, so long as a competent pediatric bronchoscopist and a well-trained team are present.

Information on the etiology of congenital VCP is still evolving. The causes include birth trauma, asphyxia, surgical trauma of the recurrent laryngeal nerve, infections, and CNS anomalies. Chiari malformation is the commonest CNS-related cause, followed by encephalocele, hydrocephalus, and cerebrospinal fluid dysgenesis.6,8 Several studies have revealed that CVP in patients with CNS anomalies

![Figure 2](image-url)  Comorbidity of congenital vocal cord paralysis. ASD = atrial septal defect; PS = pulmonary stenosis; PDA = patent ductus arteriosus.
was secondary to traction on the vagus nerve caused by caudal displacement of the cerebellum or brainstem, which may be reversible if decompression is performed in time.\textsuperscript{3,9,10} About half (8/15) of the patients in our study had associated neuromuscular diseases. The commonest CNS lesions were choroid plexus cysts, followed by ventriculomegaly and neonatal asphyxia. However, we failed to find a convincing cause and effect relationship between congenital VCP and choroid plexus cysts, based on our limited experience. We recommend that magnetic resonance imaging of the brain, rather than ultrasonography, should be performed in all patients with congenital VCP in order to identify possible CNS etiologies and associated intracranial lesions.

We found a high percentage of patients with coexisting congenital heart disease (11/15, 73%). The commonest cardiac anomalies were atrial septal defect, followed by pulmonary stenosis and patent ductus arteriosus (Figure 2). De Gaudemar et al reviewed 113 patients with congenital VCP, and only six had heart malformations.\textsuperscript{8} We recommend that infants with congenital VCP undergo three-dimensional cardiac ultrasonography and chest computed tomography in order to detect associated cardiovascular anomalies or other underlying intrathoracic comorbidities.

Other laryngeal anomalies coexistent with congenital VCP included subglottic stenosis and laryngomalacia. Interactions between these upper airway anomalies should be carefully investigated; follow-up studies can be complicated and confusing and previous video recordings should be carefully reviewed before each procedure to avoid misinterpretation.

Previously reported tracheotomy rates for the treatment of congenital VCP vary from 0–73% for bilateral VCP.\textsuperscript{2,11–13} Four patients in our study required tracheotomies (27%), all of who had bilateral abduction VCP. Two patients were in the idiopathic group and two were in the neurogenic group. In the neurogenic group, one had a choroid plexus cyst and one had ventriculomegaly plus a choroid plexus cyst. All the infants who needed tracheotomies had experienced at least two extubation failures. We suggest that, if a patient with congenital VCP has experienced respiratory failure and become endotracheal tube-dependent, they should undergo a tracheotomy first, before other corrective surgical interventions are considered.

The association between gastroesophageal reflux (GER) and laryngeal disorders has been recognized for decades. In infants, GER and laryngomalacia can both be manifestations of a common immaturity of the autonomic nerve system, or GER can sometimes be secondary to physiological disturbances caused by abnormal respiratory mechanisms. GER has long been thought to be a significant aggravating factor for laryngomalacia, but the link between GER and VCP is unclear. In this study, gross reflux esophagitis and posterior or reflux laryngitis consisting of edema, epithelial hypertrophy, and bluish color of the interarytenoid space, occurred in the majority of our patients. Significantly, lipid-laden macrophages were found in the BAL fluid of seven out of nine (77.7%) patients during the first procedure. We suggest that patients with VCP be treated with antireflux medications to avoid reflux aspiration and further irritations to the upper airways. Twenty-four-hour dual-probe pH monitoring is indicated in patients whose BAL fluids are positive for lipid-laden macrophages. The time to resolution can be shortened in the same manner as in laryngomalacia. Further prospective clinical studies are required to provide interpretation of bronchoscopic findings and clarification of the causal relationships between GER and VCP.

According to previous studies, the spontaneous recovery rate for congenital VCP is greater than 50%, often occurring before 6 months, and rarely after 36 months.\textsuperscript{6,8,13,14} Accordingly, VCP resolved in the vast majority of our patients in relatively short periods of time without the need for tracheotomies. Our patients’ spontaneous recovery rate was 71%, which is comparable with those in previous reports. There was no significant difference in recovery rates between idiopathic and neurogenic VCP, though the elapsed time for resolution was longer in the neurogenic group. Most authors have favored conservative management of VCP in order to avoid unnecessary surgical procedures. When indicated, tracheotomy remains the gold standard for treatment of patients with congenital bilateral VCP, because it is the least aggressive surgical technique for infants and it is a reversible, allowing time for potential spontaneous recovery. Some authors recommended waiting at least 1 year before undertaking other surgical interventions. Some patients recovered spontaneously at 9–11 years of age,\textsuperscript{13,14} and the parents of patient 7, who was managed with a tracheotomy for 5 years, decided to wait until adolescence due to concern over voice quality. Based on our experience, the decision of whether to manage VCP conservatively with observation and tracheotomy, or to undertake other corrective surgical procedures, should take into account the patient’s other concomitant medical conditions, including their respiration, swallowing, and growth.

In summary, FB is the gold standard for the evaluation of newborn infants with congenital stridor. Pediatric pulmonologists should play a more active role in the diagnosis and management of congenital VCP. The majority of patients can be managed conservatively and should be monitored carefully...
using serial FB. When indicated, tracheotomy is the preferred surgical procedure.

References