

Congenital Bilateral Vocal Cord Paralysis

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Summary

Congenital upper airway obstruction is a relatively rare but important cause of major respiratory problems in the neonatal period. Vocal cord paralysis is the second most common cause of congenital airway obstruction presenting with neonatal stridor. It is often the reason for the failure of neonates to wean from the respiratory support. A retrospective analysis of medical record review was conducted. There were seven paediatric patients diagnosed with bilateral vocal fold paralysis in the past three years, of which five were recently diagnosed. All patients underwent flexible with/without rigid bronchoscopes to confirm the diagnosis. This case series highlight our experience in managing the problem of bilateral vocal cord paralysis in the paediatric population, with particular emphasis on their clinical presentations, associated complications and both upper and lower airway abnormalities. The management options and outcome of these patients will also be discussed.

Key Words: Congenital, Bilateral vocal cord palsy

Introduction

Congenital vocal cord paralysis (VCP) is the second most common cause of stridor in newborn, after laryngomalacia. It accounts for 10% of all congenital laryngeal anomalies¹. VCP in children can be unilateral or bilateral. Bilateral VCP is often symptomatic early in life, whereas unilateral paralysis may be missed. The fixation of both cords in adduction leads to significant airway compromise and results in stridor and dyspnoea. The airway intervention such as tracheotomy is required in more than 50% of affected patient². We present seven cases of congenital bilateral vocal cord paralysis, of which five needed a tracheotomy to overcome significant upper airway obstruction.

Materials and Methods

A retrospective analysis of medical record review was conducted. All patients underwent flexible

bronchoscopy with or without rigid bronchoscopy to confirm the diagnosis. Flexible bronchoscopy was performed by a paediatric chest physician and rigid bronchoscopy done by Paediatric ENT surgeon at the same setting to confirm the diagnosis and to decide on the intervention. The procedure was performed under general anaesthesia. The patient was lightened from the anaesthesia to observe the movement of the vocal cord.

Results

There were seven paediatric patients diagnosed with bilateral VC paralysis in the past three years (2003 – 2005). Five of them were male and two were female. All the patients were Malays except for one Bidayuh baby. The cause of bilateral VC palsy is not known but five patients were syndromic babies (Table I). The type of vocal cord palsy and associated upper and lower airway anomalies are shown in Table I.

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Clinical symptoms and diagnosis

Their presentations were almost similar. All infants presented early in life with stridor and/or weak cry associated with variable degrees of upper airway obstruction and difficulty in controlling secretions associated with recurrent aspiration and feeding problems (Table I). The median age of diagnosis was three months. However, the diagnosis was delayed up to nine months of age (in patient 7) due to late referral.

Four patients (2, 3, 6 & 7) had associated gastroesophageal reflux disease (GERD) that further complicates feeding difficulties and increased risk of aspiration pneumonia. Fundoplication and gastrostomy was performed in these patients.

Progress and Outcome

a) Children with tracheotomy

Five patients (patient 1, 3, 4, 5 and 7) required tracheotomy due to severe upper airway obstruction. Four of them (patient 1, 3, 4, 7) were ventilator-dependent up to one month and one patient (patient 5) was oxygen-dependent up to four months.

Patient 1, currently is still on home Continuous Positive Airway Pressure (CPAP). The diagnosis of vocal cord palsy was made at two weeks of life. After tracheotomy, he was managed to be weaned off ventilator within 48 hours and discharged home without oxygen two weeks later. Unfortunately, at six months of age, he had Mycoplasma pneumoniae infection and Respiratory Syncytial Virus (RSV) pneumonia. Following that, he required recurrent and prolonged ventilation and later developed bronchiolitis obliterans and tracheobronchomalacia. He was on home Bi-level Positive Airway Pressure (BiPAP) with oxygen for three years. Up to now, he has no voice but able to communicate using a sign language. A repeat flexible bronchoscopy at three year's of age and showed a slight improvement of vocal cord movements and tracheobronchomalacia.

Patient 3, 4 and 7 were weaned off ventilation within 48 hours after tracheotomy and patient 5 was weaned off oxygen two weeks after tracheotomy. All of them were discharged without oxygen.

In patient 3, a repeat bronchoscopy was recently done which showed normal movements of vocal cord. Decanulation of tracheotomy was planned in 1 - 2 months time. The rest of the patients had no repeat bronchoscopy done as yet.

Patient 7 was referred at the age of nine months old. The symptoms of stridor and weak cry were present at birth. He had multiple episodes of aspiration and nosocomial pneumonia required recurrent and prolonged mechanical ventilation. The tracheotomy was done at the age of two months.

b) Children without tracheotomy

Patient 2 had a milder form of upper airway obstruction and did not require ventilation or oxygen. However, she had severe GERD with hyperactive airway disease. As the risk of aspiration was higher in the presence of vocal cord palsy, fundoplication and gastrostomy insertion were done at two months of life. Currently, she is still on gastrostomy feeding and has had no further episodes of aspiration pneumonia.

Patient 6 (Figure 1) had severe upper airway obstruction and required non-invasive ventilation (BiPAP) for two months. He did not require tracheotomy as his symptoms improved and subsequently was discharged well without oxygen. This child also had severe GERD and the fundoplication and gastrostomy were done for the same reason. He was well for a month, but unfortunately was readmitted with severe sepsis and died two weeks later.

Two patients (1 and 3) developed complications related to tracheotomy i.e. granulation tissue formation required revision and refashioning/dilatation (done by ENT surgeon under general anaesthesia). Prior to discharge, all the parents were trained to give gastrostomy feeding and to change tracheotomy tube. They were also taught cardiopulmonary resuscitation. Once confident to do those tasks, they were allowed home.

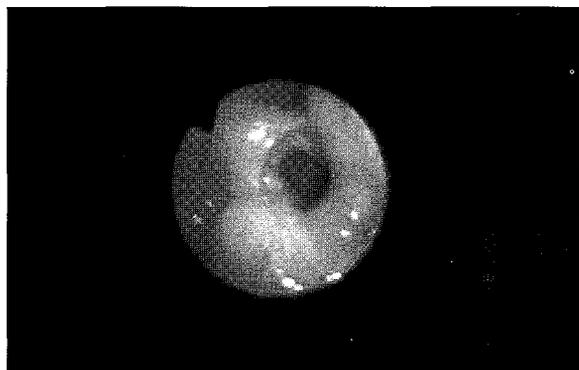


Fig 1: Abnormal vocal cord with bilateral adductor paralysis with glottic & supraglottic web (patient 6)

Table 1: Summary of characteristics of seven patients with bilateral vocal cord paralysis

| Patient | Age at diagnosis | Current age | Type of vocal cord palsy | Presentation | Other airway abnormalities | Associated syndrome |
|---------|------------------|-------------|------------------------------|--|--|--|
| 1 | 2 weeks | 3.5 years | Bilateral abductor paralysis | Stridor @ at birth, ventilator dependent | Tracheobronchomalacia 2' prolonged ventilation | Nil |
| 2 | 2 weeks | 2 years | Bilateral abductor paralysis | Stridor @ at birth, no voice, cyanosis on feeding | Laryngomalacia | Chromosomal abnormalities 46XX, t(q: 15) (q22.1;q26.3) |
| 3 | 3 months | 8 months | Bilateral adductor paralysis | Stridor @ 2 wks old Weak cry, feeding difficulties, cyanosis Recurrent pneumonia | Nil | Pierre Robin Sequence |
| 4 | 1 month | 6 months | Bilateral abductor paralysis | Stridor @ day 3 Ventilator dependent | Nil | Golden Har Syndrome |
| 5 | 3 months | 7 months | Bilateral abductor paralysis | Stridor @ day 3, weak cry, feeding difficulty, cyanosis, O2 dependent | Nil | Syndromic, congenital microcephaly |
| 6 | 5 months | 10 months | Bilateral adductor paralysis | Stridor @ at birth, abnormal cry, feeding difficulties, recurrent pneumonia | Glottic & Supraglottic web | Nil |
| 7 | 9 months | 1 year | Bilateral abductor paralysis | Neonatal Stridor, feeding difficulties, cyanosis, recurrent pneumonia | Tracheobronchomalacia | Larsen syndrome |

Discussion

Congenital bilateral vocal cord paralysis is often symptomatic in early life. The diagnosis should be considered in any infant with a weak or absent cry, significant stridor (typically biphasic), feeding and swallowing difficulties and evidence of recurrent aspiration.

In this series, five patients had bilateral VC abductor paralysis and two patients (3 & 6) had bilateral VC

adductor paralysis. VC abductor paralysis is more common, whereas VC adductor paralysis is rare. The features of laryngeal incompetence are more pronounced in VC adductor paralysis.

During bronchoscopy, apart from observing the vocal cord movements to confirm the diagnosis, it is also important to do a complete assessment of upper and lower airway to detect other airway disease to prognosticate the outcome.

The causes of VC palsy include birth trauma, iatrogenic either secondary to cardiac or mediastinal surgery, neurological (e.g. Arnold-Chiari Malformation and congenital hydrocephalus). Some cases are idiopathic. In this series, the cause of vocal cord palsy was most likely associated with dysmorphism or chromosomal abnormalities in five of seven patients. In other two patients the cause is not known. MRI Brain is recommended in patients with bilateral VC palsy. Patient 2 and 3 had normal MRI of the Brain. The rest of the patients had no MRI Brain as yet because of the risk of respiratory depression and worsening of upper airway obstruction if given sedation.

Early referral and diagnosis is very important as it does make a difference to the management and outcome of the patients. As clearly illustrated in patients 1, 3, 4 and 7, once the diagnosis was made and tracheotomy were performed, they were weaned off ventilator and were discharged without oxygen within two weeks. Patient 3 and 7 were referred from the district hospital and

were diagnosed late compared to patients 1 and 3 who were in-patients. The risk of recurrent aspiration pneumonia and lung damage could have been avoided in both patients and the duration of hospitalization would be much shorter in patient 7 (who was managed in the district hospital for nine months) if the diagnosis was made earlier.

Based on this case series, we felt that the treatment options of bilateral vocal cord palsy is determined by patient's symptoms, particularly by the severity of airway obstruction, which may exacerbated by coexisting upper or lower airway disease. The long-term outcome is variable depending on:-

- the degree of paralysis
- the aetiology and presence or absence of other congenital anomalies
- the complications related to the problem e.g. degree of lung damage secondary to recurrent aspiration pneumonia.

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