Bilateral Vocal Cord Paralysis in Newborns With Neuraxial Malformations
—Two Case Reports—

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Abstract
Two neonates presented with inspiratory stridor due to bilateral vocal cord paralysis associated with occipital encephalocele, Chiari malformation, and hydrocephalus in one patient, and cervical meningomyelocele and Chiari malformation in the other patient. The clinical symptoms dramatically regressed after repair of the encephalocele or meningomyelocele with no requirement for craniovertebral decompressive procedures or shunts in the acute phase. Careful evaluation of neonatal stridor and recognition of vocal cord paralysis are important, as treatment of associated congenital central nervous system anomalies is likely to achieve satisfactory surgical results.

Key words: vocal cord paralysis, neonatal stridor, neuraxial malformation

Introduction
Unilateral or bilateral vocal cord paralysis is a significant cause of stridor in infants and children.2,4) Bilateral vocal cord paralysis causes more severe stridor. Bilateral vocal cord palsy is the second most common cause of neonatal stridor.4) Neonatal bilateral vocal cord paralysis may be caused by birth trauma or asphyxia, and central nervous system (CNS) anomalies. Chiari malformation is the most common congenital CNS anomaly associated with laryngeal palsy, especially bilateral vocal cord paralysis,1,4,7,8) and is characterized by herniation of the pons and vermis into the cervical canal. Other uncommon CNS causes of vocal cord palsy are encephalocele, hydrocephalus, and cerebral nuclear dysgenesis.3,5) The combination of meningomyelocele, hydrocephalus, and Chiari malformation is well known.4,7–9) Bilateral vocal cord paralysis in this condition is secondary to traction on the vagus nerve caused by caudal displacement of the cerebellum or brainstem.4,7,8)

We treated two newborns with bilateral vocal cord paralysis associated with Chiari malformation and hydrocephalus.

Case 1: A male infant was born at 38 weeks gestation by normal delivery after an uneventful pregnancy. He was the first child of nonconsanguineous healthy parents. One and five minute Apgar scores were 5 and 8, respectively. His birth weight was 3375 g, length was 48 cm, and head circumference was 35 cm. He was admitted to our neonatal intensive care unit at the age of 4 days because of respiratory distress, stridor, and occipital encephalocele. He was treated with nasal continuous positive airway pressure for about 1 week. Physical examination revealed laryngeal stridor, subcostal and intercostal retractions, and mild generalized hypotonia. There was a high cervical mass measuring 7 × 7 cm in the occipital region. No other abnormality was recognized. Routine laboratory examinations including hemogram, electrolytes, calcium, glucose, lactate, ammonia, blood gases, and urinalysis were within normal limits. Cranial computed tomography revealed occipital encephalocele and Chiari malformation with hydrocephalus (Fig. 1). Initial flexible fiberoptic laryngoscopy showed bilateral vocal cord immobility without adduction or abduction. A diagnosis of complete bilateral vocal cord palsy was made (Fig. 2). Neurosurgical repair was performed...
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at age 7 days. The occipital encephalocele was excised and watertight dural closure was performed. No craniovertebral decompressive or diversive procedure was instituted. Unilateral (left) recovery in vocal cord mobility was established on the 3rd postoperative day (Fig. 3). Full mobility of the folds had returned at age 3 months (Fig. 4).

Case 2: A female infant was born at 40 weeks gestation to a healthy 28-year-old mother after an uneventful pregnancy. Her parents were nonconsanguineous. Apgar score was 8 at 1 minute and 10 at 5 minutes. Her birth weight was 3450 g, length was 51 cm, and head circumference was 36 cm. She was admitted to our hospital with feeding problems, respiratory distress, and stridor at age 18 days. Nasal continuous positive airway pressure was instituted. Physical examination revealed suprasternal, subcostal, and intercostal retractions, and inspiratory stridor. There was a low cervical mass measuring $4 \times 3$ cm at the cervicothoracic junction. No other abnormalities were found. Laboratory findings including hemogram, electrolytes, calcium, glucose, lactate, ammonia, blood gases, and urinalysis were within normal limits. Flexible fiberoptic laryngoscopy showed that the bilateral vocal folds were paralyzed in the midway position. Cranial computed tomography revealed posterior fusion defect at the C-7 to T-1 levels associated with meningomyelocele and Chiari malformation (Fig. 5). The mass was excised and the dura repaired at age 28 days (Fig. 6). No craniovertebral decompressive procedure was performed. Unilateral (left) recovery

Fig. 1 Case 1. Cranial computed tomography scan showing occipital encephalocele.

Fig. 2 Case 1. Laryngoscopic image showing complete bilateral vocal cord paralysis.

Fig. 3 Case 1. Laryngoscopic image showing unilateral (left) recovery of vocal cord paralysis.

Fig. 4 Case 1. Laryngoscopic image showing complete recovery of bilateral vocal cord paralysis.
in vocal cord mobility was established on the 10th postoperative day. Bilateral vocal cord functions had recovered completely within 1 month after surgery. Six months follow-up examination was uneventful.

**Discussion**

Bilateral vocal cord paralysis was present in 30 of 389 patients with congenital anomalies.\(^6\) Twenty-four of these 30 patients had meningomyelocoele and hydrocephalus coexistent with Chiari malformation, and the other six had hydrocephalus without craniovertebral pathology. Sixteen of 102 patients with laryngeal paralysis had neurological anomalies, the most frequent of which was Chiari malformation (7 cases).\(^2\) Sixteen patients had cerebral malformations among 113 patients with vocal cord paralysis, half of which were Chiari malformation.\(^3\) Similar cases with neonatal bilateral vocal cord paralysis associated with CNS anomalies have been reported.\(^1,8\) The present cases illustrate that neonatal stridor due to bilateral vocal cord paralysis associated with occipital encephalocele or cervical meningomyelocoele are caused by traction on the vagus nerve resulting from caudal displacement of the cerebellum or brainstem by the mass effect.

Flexible laryngoscopy has become the main method for the diagnosis of vocal cord palsy. It is safe and well-tolerated by all age groups. Neonates and infants with suspected vocal cord palsy must be carefully evaluated for underlying diseases, especially CNS anomalies. Vocal cord paralysis may resolve with treatment of the underlying disease or congenital anomaly. Bilateral vocal cord paralysis may totally be reversible if the decompressive procedure is performed in time. The best known examples are early decompression of Chiari malformation and/or treatment of hydrocephalus.\(^6,10\) The present cases emphasize the significance and urgency of bilateral vocal cord paralysis in newborns with neuraxial malformations, and underline the fact that limited surgery such as excision of the encephalocele or meningocele/meningomyelocoele can achieve effective outcomes at the acute stage. By removing the mass and decreasing the traction force on the vagus nerve, coexistent with a narrow craniovertebral junction, vocal cord paralysis may be cured without the requirement for craniovertebral decompressive surgery which is a major procedure for a neonate. No hydrocephalic progression was observed in our Case 2 within 6 months. Unfortunately, Case 1 was lost to follow up.

Laryngeal stridor caused by bilateral vocal cord paralysis in newborns must be carefully evaluated as this devastating pathology may be due to concomitant CNS anomalies which can be immediately repaired with good results.

**References**

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