Vocal cord palsy in an infant with myelomeningocele

**Abstract**

In this article we describe the case of a four-month-old male infant with myelomeningocele, who presented with inspiratory stridor and vocal cord palsy (VCP). Hindbrain dysfunction is a leading cause of mortality and morbidity in children with neural tube defects. It is important to consider the above in the differential diagnosis of infants with breathing difficulties. A discussion of myelodysplasia, Arnold-Chiari malformations, bilateral VCP and anaesthesia management is presented.

**Case report**

A four-month-old infant presented to the emergency department of our hospital with an acute episode of difficulty in breathing for the last five days. The child was treated earlier by a local doctor for respiratory distress due to pneumonitis. On examination at admission, the child had inspiratory stridor, was afebrile and had no history of trauma or upper respiratory tract infection. The examination revealed a lumbar myelomeningocele with no apparent neurological deficit. A CT scan revealed severe hydrocephalus with associated Arnold-Chiari malformation (ACM) type II. A probable diagnosis of stridor due to ACM was made and an emergency ventriculoperitoneal shunt was planned to relieve the increased intracranial tension.

Anaesthesia was induced with thiopentone 3 mg/kg and fentanyl 2 µg/kg. An uncuffed endotracheal tube of size 4.0 was inserted after achieving neuromuscular blockade with 0.15 mg/kg of vecuronium. Positive-pressure ventilation was commenced with 66% nitrous oxide in oxygen, isoflurane and an intermittent dose of vecuronium. At the end of surgery, the neuromuscular block was reversed. The stridor persisted after extubation, requiring reintubation. The child was breathing spontaneously with the endotracheal tube in situ and oxygen through a T-piece with an oxygen flow of 2 l/minute. A second trial of extubation was attempted after 48 hours, but this failed. Tracheostomy was carried out and an uncuffed tracheostomy with tube size 3.5 mm was inserted. The lumbar myelomeningocele was repaired after three days of shunt surgery. Repeat direct laryngoscopy after five days showed improvement, with the return of right vocal cord movement. Bilateral mobile vocal cords were demonstrable after one week.

**Discussion**

Bilateral vocal cord palsy (VCP) accounts for 10–15% of cases of stridor in infants. VCP is often idiopathic and may be associated with neurological anomalies such as ACM, intraventricular haemorrhage, meningoencephalocele and hydrocephalus. Myelomeningocele with ACM and hydrocephalus is a common association, seen in infants with a congenital failure of neurulation.

Clinical manifestations of ACM type II result from downward displacement and herniation of the cerebellar tonsil and medulla through the foramen magnum and may include dysphagia (92%), stridor or bilateral VCP (69%), apnoea (54%), and, less commonly, aspiration, respiratory difficulty, weak cry and lower cranial nerve palsies. These symptoms are a result of dysfunction of the medullary respiratory centre and traction or elongation of cranial nerves IX and X.

Charney et al. classified patients with ACM on the basis of their symptoms and signs as follows:
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- Grade I: Stridor alone.
- Grade II: Stridor and apnoea.
- Grade III: Stridor, apnoea, cyanosis and dysphagia.

Grade I patients have a good chance of recovery with intervention, and of long-term survival. However, this deteriorates to 50% morbidity and a 75% chance of long-term survival in grade II, and minimal chances of recovery and a 40% chance of long-term survival in grade III.

The development of VCP in our case may have been due to the downward displacement of the brainstem secondary to increased intracranial pressure (ICP), resulting in compression of the fourth ventricle and stretching of the rootlets of the vagus nerve. The treatment of such symptoms requires decompression of increased ICP, either by shunting of the cerebrospinal fluid or decompression of the foramen magnum, thereby reducing the pressure difference between intracranial and intraspinal compartments. The relief of stridor should theoretically occur after decompression of the vagus nerve in the posterior fossa; however, a period of 48 hours after definitive neurosurgical management should be given before making the decision for tracheostomy. Rath et al. reported relief of symptoms as early as 16 hours after definite surgery of a ventriculoperitoneal shunt and foramen magnum decompression.

As emergency measures, intubation and immediate ventricular puncture are recommended to relieve increased ICP. A ventricular shunt should ideally be performed within 48 hours of onset of features of hindbrain dysfunction. A timely decrease in ICP may reverse the VCP. Delay in relieving the increased ICP may lead to degeneration of the nucleus ambiguous and irreversible paralysis of the vocal cords.

In the perioperative period, anaesthesiologists should be aware of complications occurring during induction and maintenance of anaesthesia. An increase in ICP at the time of induction and intubation may result in adverse cardiac events and even cardiac arrest. Measures should be taken to avoid an increase in ICP and further herniation. Inhalation induction with halothane should be avoided because of its propensity to increase ICP. Induction with thiopentone or propofol is preferred, owing to their cerebral vasoconstrictor properties and prevention of an increase of ICP. Intubation should be done after achieving adequate muscle relaxation with the use of nondepolarising muscle relaxants and mild hypocapnia. Intraoperatively, depth of anaesthesia should be maintained through the use of depolarising muscle relaxants, short-acting opioids such as fentanyl and inhalational agents such as sevoflurane or isoflurane. Mild hypocapnia in the range of 30 to 35 mmHg should be maintained.

In conclusion, we wish to highlight the problems associated with myelomeningocele during the perioperative period. A thorough understanding of the pathophysiology is essential when such patients present for surgery.

References