Laryngeal Palsy in Association with Myelomeningocele, Hydrocephalus, and the Arnold-Chiari Malformation*

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Although disturbances of laryngeal motor function in infants and children with congenital hindbrain malformations have been described and recognized in both the otolaryngological and pediatric literature,1,3,8,10,16 these problems have received comparatively scant neurosurgical attention. In this report, we present five cases in which mild to severe laryngeal motor deficits occurred in association with the Arnold-Chiari malformation, myelomeningocele, and aqueductal stenosis in varying combinations. We have reviewed the cases from the standpoint of the pathological physiology of abductor vocal cord paralysis, as well as the clinical aspects pertinent to the management of this perplexing, and at times, life-threatening disarrangement.

Case Reports

Case 1. A 2-day-old baby, the product of a full-term uncomplicated pregnancy and delivery, was transferred to the University of Colorado Medical Center because of a leaking myelomeningocele in the lumbosacral area.

Examination. On admission the child was alert, had spontaneous movements at the hips, but showed a sensory and motor loss below the L-3 spinal segmental level, as well as urinary and fecal incontinence. The myelomeningocele sac intermittently drained clear cerebrospinal fluid.

During the initial 12 days of hospitalization the head circumference increased 3 cm, the anterior fontanelle became tense, and marked stridor with suprasternal retraction supervened. Radiological examination demonstrated a lacunar skull, and a caudally coned foramen magnum which may be a characteristic of the Arnold-Chiari malformation (Fig. 1). When the child was 2 weeks old, a ventriculogram (Fig. 1) showed significant supratentorial hydrocephalus with the medulla, cerebellar tonsils, and caudal fourth ventricle herniated caudally to the C-2 arch. Functional assessment of the aqueduct and of the cisterns was not made.

Operation. A ventriculoatrial shunt (Pudenz) was inserted. Immediately after this operation stridor disappeared, but 8 days later the shunt was not functioning and the fontanelle again became tense. Severe stridor returned and laryngoscopy demonstrated bilateral abductor paralysis of the vocal cords. The shunt was revised and a clot found in the cranial limb of the Pudenz valve. After the shunt revision, stridor subsided, and the myelomeningocele was closed when the child was 6 weeks old.

The child's subsequent course has been free of laryngeal symptomatology, and the vocal cords have been observed to move normally. At age 20 months, the shunt was again revised because of an abnormal progression of head circumference, with a subsequent regression to a normal rate of head enlargement. At age 30 months, the valve is functioning well, but intellectual performance indicates mild retardation in both language development and motor skills.

Case 2. A 2-day-old boy, born after a full-term pregnancy complicated by persistent vomiting and severe peripheral edema, was transferred to the University of Colorado Medical Center because of a large lumbosacral myelomeningocele measuring 8×10 cm, as well as bilateral club feet.

Examination. The infant was noted to have a superficially infected myelomeningocele sac (nonhemolytic staphylococcus), flaccid paralysis of both legs, absent sensation below the T-12 dermatomal level, urinary and fecal incontinence, bilateral club feet, and dislocated

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hips. The myelomeningocele sac was treated with local application of 1% silver nitrate dressings, and cultures from the surface of the sac were sterile within 24 hours.

The head circumference increased abnormally during the first 3 weeks of life. A ventriculogram at 3 weeks demonstrated marked supratentorial hydrocephalus, total occlusion of an undeformed and midline rostral aqueduct, and foramen magnum caudal coning suggesting the presence of an Arnold-Chiari malformation. No soft tissue evaluation of the posterior fossa or cisterns could be made.

Operation. A ventriculoatrial shunt (Holter, "normal pressure") was inserted. On the 20th postoperative day, marked inspiratory stridor developed in association with an abnormal increase in head circumference (2.5 cm in 10 days). Direct laryngoscopy demonstrated adductor paralysis of the vocal cords. The respiratory distress became so severe that tracheostomy was necessary. The ventriculoatrial shunt was revised and the Holter valve was found to operate at a pressure of 120 mm of water instead of an expected pressure of approximately 70 mm of water. After the revision the head circumference decreased and at the age of 4 months the myelomeningocele was closed. At the age of 5 months, laryngoscopy revealed normal movement of the vocal cords, and the tracheostomy was gradually closed over a 9-day period. At the age of 12 months, the patient is free of stridor.

Case 3. At the age of 13, an otherwise normally developing girl began to complain of suboccipital headache and paresthesias in her finger tips. These symptoms became progressively more severe over a 2-year period, and at the age of 15 she developed truncal ataxia, dysphagia, and hoarseness.

Examination. The patient was first seen at the University of Colorado Medical Center at the age of 15 years. On physical examination she was alert and cooperative, but had a nasal, hoarse voice, difficulty in clearing pharyngeal secretions, decreased palatal movements, and a markedly reduced gag reflex. Unfortunately, direct laryngoscopy was not performed in this case. The fundi were normal except for healed chorioretinitis in the left eye, There was generalized weakness in the arms as well as bilateral extensor plantar responses and marked disequilibrium on attempted tandem gait. A cine-esophagram demonstrated a delayed initiated swallowing, feeble disorganized pharyngo-esophageal peristalsis, and laryngotracheal aspiration with no cough reflex. A Tensilon test was normal. X-rays demonstrated skeletal and soft tissue craniovertebral anomalies with mild dorso-rostral basilar invagination, and minimal symmetrical supratentorial hydrocephalus; the left cerebellar tonsil extended down to the level of C-2. The cisterns were patent, and the functional level of the obstruction in the CSF pathway was not proven.