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,4,7,22,24,26 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 ventriculostomy 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 abductor paralysis of the vocal cords. The respiratory distress became so severe that tracheostomy was necessary. The ventriculostomy 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.
Laryngeal Palsy

Operation. A suboccipital craniectomy was performed immediately after the ventriculogram. Symmetrical prolongations of the cerebellar tonsils extended caudally to the second cervical cord segment, with spinal and bulbar nerves coursing cephalad. A wide suboccipital decompression was performed with resection of the laminar arches of first and second cervical vertebrae.

The patient improved rapidly following surgery, and within 2 weeks her voice and swallowing were normal. When seen 1 year after surgery she was entirely asymptomatic and doing well in school.

Case 4. A baby girl, the product of a full-term pregnancy and uncomplicated delivery, was born with a myelomingingocele extending from the mid-thoracic to the upper lumbar spine and an obviously enlarged head. At the age of 15 days, ventriculography demonstrated severe communicating hydrocephalus. A ventriculoatrial shunt (Pudenz) was inserted and at the age of 23 days the thoracic myelomingingocele was repaired.

First examination. The patient was first seen at the University of Colorado Medical Center at the age of 13 months, and on examination exhibited minimal movement of the right hip flexors, but otherwise completely paralyzed legs. Urographic studies demonstrated a flaccid bladder with reflux into duplicated left ureters, and severe left hydronephrosis. The Pudenz shunt mechanism was functioning.

Second examination. At the age of 16 months the patient was readmitted to the University of Colorado Medical Center because of stridor, vomiting, and lethargy. The Pudenz shunt was not functioning. She was cyanotic and desperately ill, with profound inspiratory stridor and suprasternal retraction.

Operation. Laryngoscopy revealed bilateral abductor paralysis of the vocal cords, and a tracheostomy was performed. Subsequent to this, ventilation improved and she became more alert. The Pudenz valve was revised and found to be obstructed at the cardiac limb. Nine days after the shunt revision, laryngoscopy revealed normal movements of both vocal cords during both inspiration and crying; the tracheostomy tube was removed, and the patient was discharged.

Third examination. Only 2 days later she was readmitted in coma with severe stridor and a malfunctioning shunt. A tracheostomy was re-established, and the shunt again revised. She improved over the subsequent few days and tolerated prolonged plugging and eventual removal of the tracheostomy tube within 1 week. During the past 2 years she has been seen at 3- to 4-month intervals and her neurological and respiratory status found to be stable. At 36 months of age her mental development has been assessed as functioning at a level of 18 to 24 months.

Case 5. A 3-day old girl was transferred to the University of Colorado Medical Center with a superficially infected lumbar myelomingingocele (nonhemolytic staphylococcus). She had been the product of a pregnancy complicated by exposure to infectious hepatitis. Her family history was pertinent in that a maternal cousin died at age 6 months with a myelomingingocele and hydrocephalus, a paternal cousin had club feet, and a paternal uncle was congenitally blind.

First examination. The infant had flaccid paralysis of both legs except for slight quadriceps movement, as well as urinary and fecal incontinence. Her head was not significantly enlarged nor was respiratory distress evident. Local silver nitrate applications (4%) rendered the sac covering sterile and promoted complete epithelization except in deeply furrowed areas.

By the age of 5 weeks her head circumference had increased 7 cm (from 29 to 36 cm) and ventriculography revealed moderate hydrocephalus, total aqueductal occlusion, lacunar skull, and a coned foramen magnum. A ventriculoatrial shunt (Pudenz) was established. The myelomingingocele flattened after the shunt and the patient was discharged.

Second examination. The patient was readmitted at the age of 4 months because of lethargy, vomiting, and inspiratory stridor. The shunt was malfunctioning. Her respiratory difficulty was initially thought to be secondary to "subglottic edema" and a tracheostomy was performed. After revision of the ventriculoatrial shunt, she became more alert, stridor subsided, and the tracheostomy was removed.

While at home, she would have intermittent stridor in association with upper respiratory infections, and laryngoscopy at the age of 14 months revealed bilateral abductor paralysis of the vocal cords with a 2 mm glottic "chink."

Third examination. In spite of a "perma-
tracheosotomy, she was readmitted at the age of 15 months because of progressive lethargy. When examined she was drowsy, and had a right peripheral facial weakness, absent gag reflex, spasticity of the arms, andneck stiffness.

The valve was functioning well. An esophagogagram demonstrated nasopharyngeal reflux, abnormal peristalsis, and marked swallowing dysfunction. The spinal fluid was sterile.

Operation. A suboccipital craniotomy was performed, with resection of the posterior elements of the first and second cervical vertebrae. Numerous aberrant dural sinuses were noted in the dura. The posterior fossa appeared small, and upon opening the dura an Arnold-Chiari malformation involving cerebellar tonsils, medulla, and emerging nerve roots was encountered. A wide suboccipital decompression was performed.

Postoperative course. The patient improved; 2 weeks postoperatively her vocal cords began to abduct feebly, and she was able to breath around the tracheostomy tube without stridor or hoarseness. Progressive improvement in swallowing has been evident. The child, now 28 months of age, is free of the tracheostomy; swallowing is normal and repeated laryngoscopy has demonstrated normally functioning vocal cords.

Discussion

The Physiological Basis of Laryngeal Palsy with Special Reference to Abductor Dysfunction. Original observations on the neural mechanisms governing laryngeal regulation made by a number of pioneers in the neurological sciences have been confirmed and amplified by recent studies. Krause in 1884 reported aphony in dogs after bilateral extirpation of the gyrus precrucialis, and Cushing. Foerster and Penfield described coordinated laryngeal movements and phonation incident to stimulation of the human motor cortex at its caudal extremity adjacent to Broca’s area. Foerster postulated the necessity of bilateral lesions as a requisite for supranuclear laryngeal disturbances. Semon and Horsley defined the topography of medullary laryngeal centers in the dog by correlating regional stimulation along the fourth ventricular floor with direct laryngoscopy. Stimulation in the region of the ala cinera was found to result in vocal cord abduction, whereas more caudally direct stimulation near the calamusscriptorius induced adduction of the cords.

Furstenberg and Magielski, applying stereotaxic techniques to this problem, have confirmed and extended these observations, explicitly defining the functional pattern of cellular organization within the primate nucleus ambiguus. This nucleus group is considered to be the substrate for peripheral motor innervation for the ipsilateral branchiomerically derived skeletal muscle of the larynx, esophagus, and pharynx in both the primate and human. The human nucleus ambiguus is an elongated structure approximately 2.0 cm in length and 0.3 cm in width, extending from the upper border of the medulla and striate medullaris caudalward to the level of the motor decussation. The small rostral segment of the nucleus is the somatomotor center for both palate and pharynx with a motor outflow via the glossopharyngeal and cephalad vagal rootlets. The larger caudal extent of the nucleus subserves laryngeal function exclusively and mediates its effect via the vagus nerve. Vocal cord movements are provided by four paired (cricothyroids, posterior cricoarytenoids, lateral cricoarytenoids and thyroarytenoids) and one unpaired (arytenoid) muscle groups, which, according to current opinion, have discrete regional representation within the nucleus ambiguus.

Though volitional human vocal cord movements are an exceptionally complex interplay of intrinsic muscular activities, and the proprioceptive role of the superior laryngeal nerve within the larynx remains unsettled, a general outline of laryngeal innervation and function has been formulated which serves as a valuable guide. The intrinsic laryngeal musculature is innervated unilaterally by two branches of the vagus nerve, the recurrent laryngeal and external branch of the superior laryngeal nerves. The latter nerve, which supplies the cricothyroid muscle, a cord tensor and adductor, originates from cell bodies situated in the rostral position of the laryngeal segment of the nucleus ambiguus. The posterior cricoarytenoids, which function as cord abductors, are innervated by recurrent laryngeal fibers derived from cells located immediately subjacent to the cricothyroid cell group. The remaining arytenoid muscles (vocal cord adductors) receive their nerve supply from the most
caudally disposed cells within the nucleus ambiguus through the recurrent laryngeal nerve.

In addition to the rostral-caudal localization of laryngeal function, nerve fibers subserving cord adduction and abduction can be further differentiated on the basis of their caliber. Fibers supplying the adductor muscle are larger with heavier myelin sheaths than axons destined for abductors. Consistent with these histological differences are the physiological observations of Murtagh and Campbell that adductor fibers continue to propagate action potentials despite chemical or thermal blockade sufficient to paralyze abductor function. In addition, the chronaxie of adductor fibers, one of the most rapid thus far identified (0.1 msec), is tenfold smaller than that found for fibers subserving abduction.

The differential susceptibility to progressive injury of adductor and abductor fibers within the recurrent laryngeal nerve was first recognized by Semon in 1881, and was based on the study of patients with partial or slowly progressive lesions of the recurrent laryngeal nerves. Semon stated that partial or progressive lesions of the recurrent laryngeal nerve resulted initially in adducted cords with a narrow rima, and as the nerve injury progressed to completeness the cords would subsequently be fixed in the "cadaveric" or semi-adducted position. From these clinical observations, Semon concluded that fibers subserving vocal cord abduction were perhaps more vulnerable to injury than those responsible for adduction, and speculated that the former fibers were located about the periphery of the recurrent laryngeal nerve and thus more exposed to injury.

Considerable controversy has existed, however, regarding the validity of Semon's conclusions. Objections are based upon numerous examples of laryngeal palsy secondary to complete, acute recurrent nerve injury in which the vocal cords have remained in the adducted position, rather than the cadaveric position. The retention of cord adduction in the presence of a complete recurrent laryngeal nerve injury has been demonstrated to be due to contraction of the cricothyroid muscle innervated by the external branch of the superior laryngeal nerve. This alternative route for vocal cord adduction has led to the questioning of Semon's provisional statement regarding the differential susceptibility of adductor and abductor fibers to injury. As Pressman and Keleman and Clerf and Baltzell have indicated, however, Semon's pronouncements were based on a study of slowly progressive disturbances involving the vagus nerve, and not on acute disruptions of the recurrent laryngeal nerve.

The examples of abductor cord paralysis in this series of cases appear to fall within the scope of Semon's hypothesis, in that abductor function failed in the presence of intact adduction. Although the Arnold-Chiari malformation was directly demonstrated in only two of the five cases, it is highly likely that the other three cases with myelomeningoceles and hydrocephalus have a similar hindbrain deformation. In ascribing the responsible factor for the observed laryngeal dysfunction, it is impossible to fractionate the role of vagal angulation and tension from that of medullary compression at the cranio-vertebral junction. The significant facts are that, in these cases, the vagal disturbances were slowly progressive and fluctuating, with the cardinal defect being in the realm of cord abduction, and the cord disturbances were all reversible. Although none of the cases developed vocal cord positions of the "cadaveric type," the striking loss of abductor function with persistent adductor positioning is highly suggestive of increased susceptibility of the former fibers to injury. The reversible nature of these disturbances would be evidence against a selective structural lesion within the nucleus ambiguus, such as a syringomyelic cleft or congenital aplasia.

Clinical Features Related to the Management of Disturbed Laryngeal Function. The incidence of abductor cord paralysis in association with congenital hindbrain malformations is unknown, but probably occurs intermittently, or in a subclinical form in a higher frequency of cases than has been generally appreciated. Symptoms secondary to laryngeal dysfunction fall into a continuum whose spectrum is comparable to the variance encountered when attempting to classify the underlying anatomical abnormalities. Our cases presented symptoms which ranged from life-endangering stridor to hoarseness coupled with varying combinations of hydrocephalus, myelomeningocele, and the Arnold-Chiari
malformation. A summary of the pertinent clinical and radiographic features of each of these cases is given in Table 1.

The clinical course of the infants in this series corresponds to other descriptions in isolated related case reports. The child is born with an obvious myelomeningocele, but significant airway difficulties usually do not appear until a few weeks or months postpartum, at which time hydrocephalus has become manifest. Changes in quality of the voice or cry are not reliable indicators of the severity or progression of abductor cord paralysis. The degree of laryngeal palsy fluctuates in individual cases, thus necessitating repeated direct laryngoscopic examination for an adequate appraisal of vocal cord function. Stridor usually appears precipitously, aggravated by crying, and as abductor palsy progresses, stridor is present during both inspiratory and expiratory phases of respiration. As in previously reported cases, three of the four infants in this group showed close correlation between the degree of increased intracranial pressure and the severity of vocal cord paralysis. All of the infants in this series had significant airway compromise when the fontanelle became tense with the infant sitting, or when a significant acceleration in the rate of growth of the head was detected. With the exception of Case 3, stridor was relieved by the reduction of intracranial pressure either by ventricular puncture or ventriculostriatal shunting.

It should be emphasized that, in infants, the diagnosis of laryngeal palsy can be established with certainty only by direct laryngoscopic examination. The tip of the laryngoscope should be inserted in the vallecula just anterior to the epiglottis (without anesthesia) in order to view the larynx without placing tension on the aryepiglottic fold (Fig. 2A). Traction on a fold may partially or completely immobilize the ipsilateral vocal cord by limiting arytenoid motion, thus leading to an erroneous diagnosis.

![Fig. 2 Schematic endoscopic views of the larynx with abductor paralysis. Note position of laryngoscope blade. A. In the adult, the vocal cords are flaccid, slightly bowed and lie in a median position. During respiration, stridor is present during expiration but is most marked during inspiration. B. In some cases with the Arnold-Chiari malformation, infants and children have only a posterior glottic chink upon inspiration. The vocal cords are less flaccid than in the adult and coapt for an almost normal cry. Inspiratory stridor is present, but severity varies with the size of the glottic opening.](image)
of vocal cord paralysis. Examination should include observations during both inspiration and expiration, as well as coughing or crying if possible. Since the child is usually dyspneic, multiple insertions of the laryngoscope are usually required if a tracheotomy has not been performed. Vocal cord movements will occur with respiration within the limits of the degree of abductor paresis, even with a tracheotomy tube in place.

In cases of bilateral abductor cord paralysis, the cords will be either motionless (except passively) in the median or paramedian position (Fig. 2 B), or closed anteriorly with only a 2–3 mm “chink” posteriorly upon inspiration. Our cases demonstrated many variations in the degree of abductor paresis, which appeared to be a function of the severity of increased intracranial pressure. At times, one cord appeared totally paralyzed, whereas the other was mildly paretic. The voice or cry as a result of unilateral abductor cord paralysis will be weak or “breathy,” in contrast to the strong cry and inspiratory stridor of bilateral abductor paralysis. Since disturbances of deglutition may exist pari passu with laryngeal dysfunction, as in Cases 3 and 5, aspiration with mechanical obstruction as a cause for stridor must be considered. Bronchoscopy may be indicated in some instances (Case 5) as both a therapeutic measure to accomplish an adequate tracheal toilet and respiratory exchange, as well as determining possible tracheal abnormalities.

Although a plastic nasotracheal tube may be utilized as an initial therapeutic maneuver for short periods of time, the definitive procedure of choice in the face of the usual clinical course posed by severe stridor is tracheostomy. Three of the four infants in this series required tracheotomies during the course of their hospitalizations. Airway obstruction is the immediate cause of death in a high percentage of patients with the Arnold-Chiari malformation, and it is our opinion that once a tracheostomy has been performed, it should be maintained until there is evidence of abductor function recovery by direct laryngoscopy. The three cases in this series required a tracheostomy for periods of time ranging from 2 weeks (Case 4) to 8 months (Case 5). Restoration of adequate laryngeal function in Cases 1, 2, and 4, as well as in previously reported cases, appeared to be dependent on reducing intracranial pressure by ventriculoatrial shunting procedures.

It is noteworthy, however, that in Cases 3 and 5, laryngeal dysfunction continued despite normal intracranial pressure in the presence of the Arnold-Chiari malformation. The relationship of the malformation per se to motility disturbances of the larynx and pharynx is uncertain. Although Ogryzlo, Rullan, and Penfield and Coburn have described adults with multiple lower cranial nerve deficits (dysphagia, dysarthria, hoarseness) in association with the Arnold-Chiari malformation, these manifestations are considered to be distinctly unusual in childhood cases. Both laryngeal and pharyngeal motility returned to normal in Cases 3 and 5 following suboccipital and rostral posterior cervical decompression. In contrast to these observations, however, Snow and Rogers have reported a case of bilateral abductor cord paralysis in an infant with the Arnold-Chiari malformation in whom laryngeal function failed to improve after suboccipital and posterior cervical decompression as the initial surgical procedure. Improvement subsequently followed the relief of hydrocephalus by the placement of a ventriculoatrial shunt.

Rullan has reported an adult with the Arnold-Chiari malformation and bilateral abductor cord paralysis which persisted after suboccipital and cervical decompression. Furthermore, attempts to evaluate the effects of surgery upon abductor paralysis of the cords are rendered even more complex because of reports of spontaneous improvement in the absence of any form of surgical intervention. Finally, it should be emphasized that suboccipital decompressive surgery for the treatment of the Arnold-Chiari malformation may be a hazardous venture, with a significant mortality rate.

Blanket statements regarding the management of these difficult and distressing cases are inappropriate in view of the subtle, as well as extreme, variations in pathological anatomy that form the basis of the underlying physiological disturbance.

Summary

We have described five cases of disturbed laryngeal function (abductor paralysis) that occurred in children with a variety of congenital malformations of the neuraxis which
included the Arnold-Chiari malformation, hydrocephalus, and myelomeningocele. We have also discussed the pathological physiology of abductor cord paralysis as well as certain clinical and radiographic features pertinent to the management of these cases. Tracheostomy appears to be a necessary procedure as an emergency measure until the hydrocephalus can be controlled; ventricular taps also have a temporizing emergency value. Shunting procedures for the relief of hydrocephalus are of definite benefit. Although the role of suboccipital and high cervical decompression remains uncertain, we have given two instances of improved laryngeal function following use of this procedure.

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