BILATERAL VOCAL CORD PARALYSIS ASSOCIATED WITH MENINGOMYELOCELE AND THE ARNOLD-CHIARI MALFORMATION.

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Stridor in the newborn or infant is generally viewed with considerable concern. Usually, causes other than vocal cord paralysis are apparent, *i.e.*, malacia of the laryngeal cartilages, congenital webs or congenital subglottic stenosis. Rarely, however, vocal cord paralysis does occur; but it is indeed rare, and the etiology is usually obscure.

Recently two infants with stridor have been investigated—both with bilateral vocal cord paralysis, myelomeningocele and the Arnold-Chiari malformation. This association, hitherto not described, would appear to be of interest insofar as it might account for some cases of bilateral vocal cord paralysis of unknown etiology occurring in infancy.

The Arnold-Chiari malformation consists essentially of a protrusion of medulla and cerebellum through the foramen magnum and conceivably can compress or exert traction upon the vagus nerves.

Upon reviewing the literature, only three previous reports[14,15,16] mention stridor in the infant with meningocele; however, this is probably a more frequent occurrence than the literature would indicate; perhaps the concern over the often accompanying hydrocephalus and meningocele sac overshadow at least the minor degrees of this condition.

CASE REPORTS.

Case 1. A full-term, 2600 gram, white female infant was born spontaneously of healthy parents after an uncomplicated delivery.

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At birth, a lumbar meningomyelocele and bilateral talipes calcaneovalgus were obvious.

The immediate postnatal period was uneventful. The head size and fontanelles were normal and remained so. The infant was discharged from the hospital at eight days of age.

At three weeks of age, the meningomyelocele sac collapsed, and clear fluid drained thereafter. Five days later, the infant developed stridor and was returned to the hospital. The stridor worsened in the succeeding few hours and the infant became semicomatose.

Direct laryngoscopy was performed. "There was no inflammation, edema, or tumor of the larynx. Both true cords were immobile, very near the midline—the glottis posteriorly being only about two millimeters wide. There was no abduction of the cords on deep inspiration, but they did approximate on phonation. A three millimeter bronchoscope was then passed with some difficulty through the larynx. The trachea and bronchi appeared normal in their entirety." A tracheostomy was performed following this examination.

Postoperatively the infant's airway remained adequate; however, feedings were taken progressively less actively. The infant remained afebrile but became increasingly lethargic. On the fourth postoperative day, following a convulsion, her respirations increased rapidly in rate, she became unresponsive and finally expired exactly one month following birth.

At post-mortem examination the body was normal except for the large lumbar spina fibida with meningomyelocele and bilateral talipes valgus. The larynx appeared normal. There was an Arnold-Chiari malformation manifested by elongation of the medulla and presentation of the tip of the midline cerebellum over the dorsal part of the medulla for approximately two centimeters below the foramen magnum. Both of these structures had herniated into and through the foramen magnum. Cervical nerve roots were directed cephalad.

Case 2. A ten-month-old child was stridorous from birth. He had a lumbar meningomyelocele and talipes calcaneovarus, probable congenitally dislocated hips and a dolichocephalic head.

When first seen, direct laryngoscopy was performed with no anesthetic. The vocal cords were immobile in the midline position with a very small glottic chink. A tracheostomy was performed. Seven months later the tracheostomy tube was temporarily removed; but stridor recurred, and the tracheostomy tube had to be replaced. Repeat direct laryngoscopy at that time again revealed bilateral vocal cord paralysis. Normal development continued and at 26 months of age direct laryngoscopy under general anesthesia with gradual lightening of the anesthetic plane revealed moderate motion of the left vocal cord, but paralysis of the right vocal cord. The tracheostomy tube was removed at this time without recurrence of the stridor. The child was seen again at 32 months of age with no stridor and with a strong voice. Direct laryngoscopy was not performed at this time. The patient was last seen at four years of age, at which time there were no additional abnormalities noted and no evidence of hydrocephalus. Four months later the child expired. No record of the terminal episode is available.

At post-mortem examination the body was normal except for moderate hydrocephalus, obvious spina bifida with myelomeningocele, and bilateral talipes calcaneovarus. The larynx was normal. Intracranial examination revealed evidence of prolonged increased intracranial pressure. The gyri

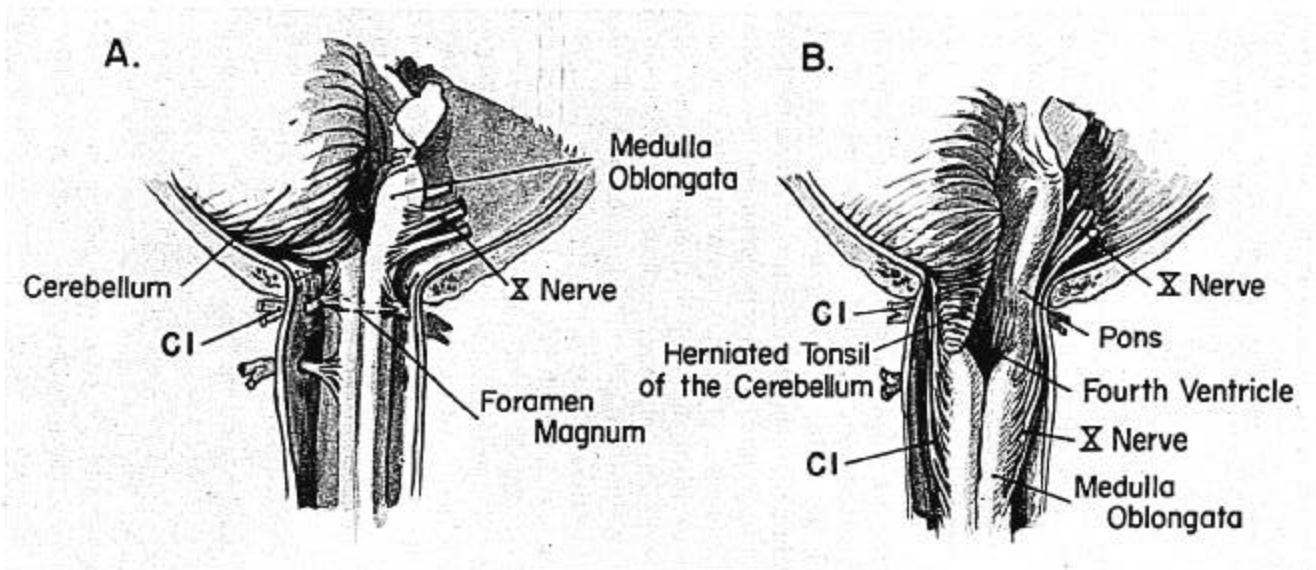

Fig. 1. Dorsal view of the posterior fossa, foramen magnum and upper cervical spine. a. Normal. b. Arnold-Chiari malformation. Left—cerebellar hemisphere in place. Right—cerebellar hemisphere removed.

Note: 1. Level of the foramen magnum relative to the medulla oblongata and the cerebellar hemisphere. 2. Upward course of the lower cranial nerves and the upper cervical nerves. 3. The pinching effect of the foramen magnum upon the upward coursing nerves and the caudally displaced medulla and cerebellar tonsils.

were flattened, and the sulci were narrowed. Chronic herniation of the cerebellum and medulla oblongata through the foramen magnum was apparent. The cause of death was felt to be secondary hydrocephalus due to herniation of the pons, medulla, and cerebellum through the foramen magnum in the presence of a lumbar meningomyelocele.

DISCUSSION.

The Arnold-Chiari malformation (Fig. 1-b) first noticed by Arnold[1] and soon after elaborated upon by Chiari[2] was first well described by Guedig and Schwalbe.[3] Russell and Donald's[4] gross description follows:

"A tongue consisting of cerebellar tissue and greatly elongated medulla oblongata protrudes through the foramen magnum downward into the spinal canal. It overlaps and compresses the upper segments of the cervical cord, distending the dural theca in this neighborhood and filling the foramen magnum. The cavity of the fourth ventricle extends caudally into this tongue, lying between the cerebellar and medullary components. The maximum diameter of the ventricle is usually within the vertebral canal at and below the level of the foramen magnum, and here, too, or at a still lower level are found the choroid plexuses and the foramina with which they are associated. The plexuses are sometimes united into one mass which lies in the midline in the ventricular cavity. The floor of the ventricle, formed by the elongated medulla oblongata, presents the characteristic markings of the rhomboid fossa, and in the midline toward its caudal extremity is a minute pore which provides a horizontal communication between the ventricle and the central canal of the ventrally placed spinal cord. The cavity of the ventricle usually extends beyond this pore as a diverticulum into the tip of the tongue-like mass."

Other structural abnormalities frequently seen in association with this anomaly are small spinal cord, cephalic covering of the upper cervical roots, hydromyelia, and hypoplasia of the cerebellum. The pons and medulla are greatly elongated while the tonsils and flocculi are seldom identifiable. The cranial nerve roots arising from the pons and medulla can be greatly elongated.

While the gross description is clear, there is not unanimity of opinion as to the etiology. Because it is so frequently associated with myelomeningomyocele, a causal relationship has been postulated by many.[5,6] Others,[9] from their studies of human embryos showing various types of myeloschisis, conclude that the abnormality is an overgrowth of the central nervous system commencing before the fourth week of development. The most striking overgrowth is in the lumbosacral region of the spinal cord. The medullary folds here become so large that normal closure of the neural tube is prevented and myeloschisis is produced; however, overgrowth of the cerebrum displaces the tentorium cerebelli more caudally, leaving much less room for the cerebellum and medulla to develop; thus they inevitably protrude downward through the foramen magnum into the cervical canal. There may be localized overgrowth only, giving either the Arnold-Chiari malformation or myeloschisis alone; the rule, with few exceptions,[7] being an association of the two anomalies.[4,8]

It is well recognized that there is a frequent association of internal hydrocephalus (usually of the communicating type) with this anomaly. This is readily explained mechanically. The foramina of Luschka and Magendie are patent and below the level of the foramen magnum; thus cerebrospinal fluid can exit freely, but re-entry is prevented by compression of the subarachnoid space at the foramen magnum. There is thus a damming back of cerebrospinal fluid and a resultant internal hydrocephalus. Escape, therefore, of the fluid through a defect in the meningomyocele sac would prevent back pressure and protect against hydrocephalus (Case 1). The association of hydrocephalus and the Arnold-Chiari malformation is considered by most workers the result rather than the cause of the malformation.[4,6,9] This appears substantiated from the fact that decompression of the foramen magnum area produces relief of the symptoms and signs of the accompanying hydrocephalus.[5,7,10]

The herniation of the posterior cranial fossa contents through the foramen magnum eventually blocks the subarachnoid space at this very point. In all of Ingraham and Scott's

20 cases, hydrocephalus was present. Of ten cases reported by Russell, all but two had hydrocephalus. In the two that did not, cerebrospinal fluid escaped freely from the body through a fistula in the membrane covering the spinal defect; thus, there was no back pressure which could produce hydrocephalus.

It is well substantiated[4,5] that the nerve roots of the lower cranial and upper cervical nerves are displaced caudally to a significant degree. Specific mention of cranial nerve involvement is scanty.[4,6,12] Clinical manifestations of these cranial nerve involvements are discussed more specifically by Ogryzlo,[7] Penfield,[5] Stookey,[13] and Smith.[14] Smith discusses two cases in which myelomeningomyocele was associated with acute laryngeal stridor. In both cases, the stridor was transitory. An autopsy, obtained on only one of these two cases, revealed hydrocephalus without a definite Arnold-Chiari malformation. Work[15] mentions briefly, bilateral vocal cord paralysis in a newborn with sacral meningocele, while Bigler *et al.*[16] mention that two of four infants with bilateral vocal cord paralysis had cervical myelomeningocele.

The vagus nerve contains both motor and sensory fibers. The motor fibers are derived from two nuclei in the medulla. The dorsal nucleus of the vagus is situated near the midline, a little beneath the floor of the fourth ventricle. The nucleus ambiguus is a column of grey matter situated deep in the medulla between nuclei of the trigeminal nerve. The vagus nerve leaves the medulla by a series of radicles with roots of the glossopharyngeal nerve above and accessory below. These roots form a single trunk running horizontally to leave the skull through the jugular foramen (Fig. 1-a).

The nucleus ambiguus receives cortical fibers directly or indirectly from the opposite aberrant pyramidal tract; these control voluntary movements of the pharynx and larynx. The reflex pathways conveying impulses from the terminal sensory nuclei are unknown, but probably form part of the intricate mass of fibers constituting the reticular formation. The recurrent laryngeal nerves spring from the vagus at the root of the neck. The left arises as the vagus crosses the

aortic arch. After hooking below the arch it passes upward in the superior mediastinum to the neck where it ascends in the groove between esophagus and trachea. It disappears under cover of the inferior constrictor muscle, to end in laryngeal branches which supply all the intrinsic muscles of the larynx except the cricothyroideus.

The right recurrent laryngeal nerve arises as the vagus crosses the right subclavian artery, hooks beneath and ascends into the neck in the same fashion as does the left recurrent nerve.

Only three mechanisms for bilateral vocal cord paralysis can be postulated, all implying the involvement of the vagus nerve at a more cephalad level than the recurrent nerve origin: *a.* brain stem disease; *b.* compression of the vagus nerves in their abnormal course upward through the foramen magnum; and *c.* traction on the Xth cranial nerve roots by caudal displacement of the brain stem (Fig. 1-b). One or more of these factors may play a role in any given case.

In an infant the only obvious manifestation of vagus nerve dysfunction would be vocal cord paralysis. This is most dramatic when bilateral involvement produces stridor. One can only theorize as to the uncommon occurrence of this symptomatology. Possibly it is because many infants die of meningitis before these symptoms can develop.

CONCLUSION.

Two cases of bilateral vocal cord paralysis associated with meningomyelocele and the Arnold-Chiari malformation have been presented with the results of post-mortem examination.

Attention has been drawn to this entity as a possible cause for stridor in infancy and cranial nerve signs and symptoms in adulthood.

The gross description of this malformation and its probable etiology have been considered.

A consideration of the possible causes of vagus nerve involvement in this abnormality has been given.

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