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Management and outcomes of pediatric vocal cord paresis in Chiari malformation

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ABSTRACT

Introduction: Pediatric vocal cord paresis (VCP) has a variety of etiologies, including congenital neurologic disease. Arnold-Chiari Malformation (ACM) is one such disease with known VCP association. However, the natural history, need for tracheostomy, and rate of decannulation in this patient population is not well characterized.

Objective: To provide prognostic information on infants with ACM and VCP.

Methods: A retrospective chart review was conducted of patients with both ACM and VCP at a single institution. Clinical outcomes and disease progression were determined using flexible laryngoscopy, serial clinical exams, and operative reports from otolaryngology and neurosurgery services.

Results: Eighteen patients were included in this study, four with ACM Type I and 14 with ACM Type II. These groups were analyzed separately. For ACM I, the average age at diagnosis was 25 months and two (50%) required tracheostomy. Three subjects (75%) achieved VCP resolution, with two doing so after neurosurgical decompression. For ACM II, the average age at diagnosis was eight months and 12 patients (86%) underwent tracheostomy. Four subjects with tracheostomy (33%) achieved decannulation, with three of these demonstrating VCP resolution. In total, six ACM II patients had complete and one had partial VCP resolution, all of whom underwent decompression. Two patients initially had normal endoscopic exams despite stridor and VCP was only noted on serial exams.

Discussion: This study represents the largest series of pediatric patients with VCP and ACM. The majority needed decompression (80%) and tracheotomy (78%). Tracheostomy decannulation typically occurred only after decompression and resolution of VCP. No children diagnosed at age < 1 month were decannulated. Early decompression was associated with successful avoidance of tracheostomy in majority of Chiari I but not Chiari II patients. Serial endoscopies were required to confirm VCP in some patients. This information could potentially aid in management and counseling parents of children with VCP and CM.

1. Introduction

Vocal cord paresis in children may be due to a variety of etiologies, such as acquired after surgery, intracranial injury, or birth trauma; congenital due to underlying neurologic co-morbidities; or idiopathic. Prior review of over 400 cases of pediatric vocal cord paresis at our own institution showed that 7% may be attributed to underlying neurologic disease [1]. Within this group, Arnold-Chiari malformation with associated hydrocephalus or myelomeningocele are the most frequently named congenital abnormalities. Gentile et al. examined 22 children younger than 1 year of age with vocal cord paralysis and noted > 20% of this group had Arnold Chiari malformation [2]. Similarly, Holinger et al. noted that out of 149 infants and children with bilateral vocal cord paralysis, 16% had concomitant Arnold-Chiari malformation [3].

Arnold Chiari malformations are a series of anatomic deformities that result in hindbrain herniation, and are classified into four different subtypes which are associated with distinct natural histories. Type I is associated with a small posterior fossa, which subsequently causes inferior descent of the cerebellar tonsils, and is often an isolated finding. Type II involves herniation of the cerebellar vermis, medulla, and fourth ventricle, and is frequently associated with hydrocephalus and myelomeningocele. It has also been associated with histopathologic changes of the brainstem, such as absence, hypoplasia, and/or necrosis of cranial nerve nuclei. Type III involves some of the findings seen in

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Type II, and with the serious addition of occipital encephalocele. Type IV is associated with cerebellar hypoplasia. Types I and II are the most common, while Types III and IV are rare. The natural histories of the various subtypes are also distinct, with patients with Type I often not diagnosed until symptoms present during young adulthood or later, while those with Type II are typically diagnosed at birth due to myelomeningocele and associated hydrocephalus in 80–90% [4–6].

Although vocal cord paresis (VCP) secondary to Arnold-Chiari malformation (ACM) is a known cause of neonatal stridor, the natural history, need for tracheotomy, and rate of tracheostomy decannulation is unknown. Here we provide the largest case series to date examining these questions.

2. Methods

A retrospective search of patients previously seen at the Children's Hospital of Wisconsin from 1999 to 2016 with a diagnosis of both ACM and VCP was performed. The study protocol was approved by the Children's Hospital of Wisconsin Institutional Review Board. Twenty-one patients were identified as candidates for the study with dates of birth ranging from August 1984 to May 2015.

Resulting patients' charts were searched for documentation of treatment for Chiari malformation and vocal cord paresis. Parameters examined included findings on endoscopic evaluation with flexible fiberoptic laryngoscopy (FFL) or bronchoscopy, direct laryngoscopy with telescopic examination, as well as otolaryngologic and neurosurgical interventions. To determine persistence or resolution of vocal cord paresis in each subject, subjects' records were searched for airway endoscopy reports. Resolution of VCP was defined as normal mobility of both true vocal cords. If there was some improvement in vocal cord mobility after the initial diagnostic exam, but one or both vocal cords had persistently decreased mobility, this was defined as partial resolution of VCP. If there was no change in vocal cord mobility after the initial diagnosis, this was defined as no resolution of VCP. Patients who did not have resolution of VCP had serial examinations (endoscopic and/or clinical) for at least 2 years after decompression.

Records were also examined for operative reports to determine which surgical interventions were undertaken by the neurosurgery and otolaryngology services. If operative reports were not found, histories from office notes were utilized to determine surgical interventions. The following neurosurgical procedures were considered indicative of decompression: cervical laminectomy inclusive of C1 laminectomy, suboccipital craniectomy, cerebellum tonsillar reduction, and/or posterior fossa decompression.

3. Results

Twenty-one patients with diagnoses of both ACM and VCP were identified. Three patients were excluded, due to incomplete records and/or uncertain history and diagnosis of VCP. Of the remaining 18 subjects, 4 were diagnosed with ACM Type I while the remaining 14 had ACM II. No patients with ACM Types III or IV were identified. Given the distinct natural histories of ACM Type I vs. Type II, the outcomes of the two groups were analyzed separately.

In patients with ACM Type I and VCP (n = 4, Table 1), the average age at VCP diagnosis was 25 months (range 0–72, mean 14.5, standard deviation 32 months, n = 4). Two required tracheostomy; these subjects had VCP diagnosed at a younger age (0 and 8 months) than the patients that did not require tracheostomy (21 and 72 months). Three subjects underwent decompression. Three subjects had resolution of VCP. Two subjects had resolution after decompression. Serial endoscopic examination after decompression was only available for subject #1, and VCP resolved within 2 months after neurosurgical intervention for this child. Subject #3 was lost to follow up after decompression at an outside institution, and therefore time to VCP resolution could not be accurately determined. Of the subjects who had undergone

Table 1
Natural history of VCP (vocal cord paresis) and need for tracheostomy in patients with Chiari I malformation.

Subject No.	Type of VCP	Age at VCP Diagnosis (months)	Age at decompression (months)	Resolution of VCP	Months to normal laryngoscopy after decompression	Age at trach (months)	Months to decannulation after trach placed	Comments
1	Bilateral	21	22	Yes	2	N/A	N/A	
2	Bilateral	0	0.2	No	N/A	0.85	N/A	Also had dysgenesis of corpus callosum. Remained trach/vent dependent with severe mixed sleep apnea
3	Bilateral	72	72–75	Yes	Unknown	N/A	N/A	Exact date of decompression unknown (performed at outside hospital), then lost to ENT follow up
4	Bilateral	8	None	Yes (at age 132 months)	N/A	8	Unknown	Was working towards decannulation but subsequently lost to follow up