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Chiari Type I Malformation in a Pediatric Population

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Abstract

Objective—The natural history of Chiari I malformation in children remains unclear.

Methods—In this population-based retrospective cohort study, we searched radiology reports from all head and spine MRI scans (n=5248) performed among 741, 815 children under age 20 within Kaiser Northern California, 1997–1998, for Chiari I. We reviewed medical records and imaging studies to determine clinical and radiographic predictors of significant neurologic symptoms defined as moderate to severe headache, neck pain, vertigo or ataxia.

Results—The 51 patients identified with Chiari I represented 1% of children who had head or spine MRI scans performed during the study period. Headache (55%) and neck pain (12%) were the most common symptoms. Syringomyelia was present in 6 patients (12%) at time of initial diagnosis; no new syrinxes developed during follow-up. Older age at time of diagnosis was associated with increased risk of headache (OR 1.3, 95% CI 1.1–1.5) and significant neurologic symptoms (OR 1.2, 95% CI 1.04–1.4).

Conclusions—Chiari I, an under-recognized cause of headaches in children, is also frequently discovered incidentally in children without symptoms. Larger and longer-term studies are needed to determine the prognosis and optimal treatment of pediatric Chiari I.

Introduction

The Chiari type I malformation is characterized by ectopia or herniation of the cerebellar tonsils through the foramen magnum. This malformation typically presents with neurologic symptoms during early adulthood. Although Chiari I is increasingly recognized in children [1–8], little is known about its natural history. Patients can be asymptomatic or can have a variety of neurologic symptoms including headache, neck pain, visual disturbances, vertigo

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and ataxia [1, 4, 5, 9–11]. Chiari I may lead to the development of syringomyelia or spinal cord cavitation, which can lead to additional neurologic deficits. Since surgical intervention can improve existing symptoms as well as prevent further neurologic deterioration from syringomyelia, there may be a benefit to early identification of these patients [1, 2, 9, 12–14].

The cerebellar tonsils rarely descend more than 3 mm below the foramen magnum in normal adults, whereas symptomatic patients usually demonstrate at least 5 mm of herniation [15, 16]. Thus, 5 mm has become widely adopted as a cutoff for defining Chiari I for both clinical and research purposes. To some degree this measurement is arbitrary and does not include other anatomic factors that may determine if a patient develops symptoms or not. Whether individuals with 2–4 mm of tonsillar ectopia may also exhibit Chiari symptoms that respond to surgical intervention remains a matter of debate [17].

With the increasing availability of diagnostic MRI, more asymptomatic patients are being identified [4, 5, 18]. Yet the neurologic prognosis for such patients remains unclear, as long-term natural history studies of Chiari I are lacking. Previous studies of Chiari I consist primarily of neurosurgical case series [1–3, 5, 9, 11, 19–23]. In order to gain a better understanding of Chiari I in children, we conducted to our knowledge the first population-based study of Chiari I.

Methods

This is a retrospective cohort study of all children under 20 years of age who were diagnosed with Chiari I within the Kaiser Permanente Medical Care Program (KPMCP) in northern California during 1997–98. We searched an electronic database to identify patients with cerebellar tonsillar ectopia noted on a head or spine MRI, and reviewed medical records and neuroimaging studies. This study was approved by institutional review boards at KPMCP and the University of California, San Francisco.

KPMCP is a large integrated health care organization that provides health care to more than 30% of people in Northern California. Our study population consists of all 741,815 KPMCP members who were less than 20 years of age during the period January 1, 1997 through December 31, 1998. We searched an electronic database which contained the reports for all 5248 MRI studies (4568 head and 680 spine) performed in our study population during the study period using the following text strings: “chiari,” “ectopi,” “tonsil,” “low-lying,” “Arnold,” “syrinx,” “syringomyelia,” “hydromyelia,” “spinal cord cavitation,” and “suboccipital craniectomy.” CHIARI I was defined as ≥ 5 mm of tonsillar ectopia [16]. Children with 2–4 mm of tonsillar ectopia were considered to have “borderline ectopia.” We excluded patients with Chiari type II malformations or mass lesions or hydrocephalus that could cause tonsillar herniation.

The available scans were independently reviewed by a neuroradiologist (CL) blinded to the clinical history. The degree of tonsillar ectopia was determined by measuring the distance between the tip of the cerebellar tonsils to the foramen magnum which was defined by the line extending from the inferior limit of the basion to the opisthion (Figure). The degree of retrocerebellar cerebral spinal fluid compression, or diminished size of the retrocerebellar cisterns, was subjectively graded as none, mild, or significant, and the morphology of the cerebellar tonsils were defined as normal, mildly pointed, or significantly pointed.

Although 14 neuroimaging studies could not be reviewed because the films had been destroyed, the clinical reports provided measurements of the degree of tonsillar ectopia. We reviewed imaging studies from 25 cases and found that the degree of tonsillar ectopia noted in the radiology reports correlated well with our study neuroradiologist’s interpretation ($r =$

0.97, mean difference = 1.4 mm, S.D. 1.1 mm). Thus, we included the 14 missing imaging studies in our study, and relied on the clinical reports to determine the degree of tonsillar ectopia. We excluded an additional 19 neuroimaging studies from further analysis since these films had been destroyed, and the degree of tonsillar ectopia was not included in the report.

The following data were obtained from medical records: demographics, age at diagnosis, clinical presentation, neurologic exam abnormalities, surgical interventions, and clinical course during follow-up within KPMCP. We recorded headache location, as well as any factors that were reported to trigger head pain. Severity of headache syndrome was classified as follows: *severe* = daily or weekly headaches reported in multiple outpatient visits spanning more than one year, that are impacting the child's function (e.g., frequently missing school); *moderate* = headaches causing multiple outpatient visits spanning more than one year, but with no or minimal impact on function; *mild* = infrequent outpatient visits for headache without evidence of chronic pain or functional limitation.

We studied clinical and radiologic features that might predict the presence of headache or "significant neurologic symptoms" defined as any of the following: moderate to severe headache syndrome, neck pain, vertigo or ataxia. Odds ratios (OR) and 95% confidence intervals (CI) were calculated using the Cornfield method. Analyses were performed with Stata 9.0 statistical software (StataCorp, College Station, TX, USA).

Results

During the interval from January 1997 to December 1998, 51 children had a Chiari I identified on a head or spine MRI study. This diagnosis was present in 1.0% of all 5248 head and spine MRI scans performed in the study population. The frequency of Chiari I diagnoses in the KPMCP pediatric population was 0.7 per 10 000. Mean age at diagnosis was 11 years (S.D. 4.8).

Neuroimaging

The degree of tonsillar ectopia in children with Chiari I ranged from 5 to 32 mm (median 7, interquartile range 6–9). Of the 51 patients, 11 (22%) had tonsils that extended more than 10 mm below the foramen magnum. The cerebellar tonsils were abnormally pointed in half (55%) of all cases, with 18% of children demonstrating *severely* pointed tonsils. A similar proportion (57%) of children with Chiari I demonstrated compression of the retrocerebellar CSF space, with severe compression noted in 22%. Abnormally pointed tonsils and retrocerebellar CSF compression were typically seen together; if one was present, the likelihood that the other abnormality was also present was 80–82%.

Syringomyelia was present in 6 children (12%) at the time of initial Chiari I diagnosis. All syrinxes involved the cervical spinal cord, while 4 of the 6 extended into the thoracic cord. The width of the syrinxes ranged from 5 to 10 mm.

Clinical presentation

Of the 51 patients, 32 (63%) were symptomatic. The most common presenting symptoms were headache (55%), neck pain (12%), vertigo (8%), sensory changes (6%) and ataxia or poor coordination (6%). Other symptoms present at the time of diagnosis included leg weakness, tinnitus, hearing loss, dysarthria, loss of consciousness and scoliosis (Table 1). Of the symptomatic patients with Chiari I, 50% were diagnosed within 14 months of onset of symptoms (interquartile range 2–106 months). In children with syringomyelia, 3 presented with extremity numbness or weakness, 1 with progressive scoliosis, and 2 without symptoms attributed to the syrinx.

Nineteen (37%) children with Chiari I were diagnosed incidentally when they received a head or spine MRI for symptoms unlikely to be related to the tonsillar ectopia, such as seizure (6), developmental delay (4), psychosis (2), chorea (1), neurofibromatosis 1 (1), follow-up of arteriovenous malformation (1), Bell's Palsy (1), retinal dystrophy (1), microphallus (1), and scoliosis without evidence of syrinx (1).

Clinical course

Children with Chiari I were followed for a mean of 6.4 years (S.D. 4.1). No new syringes were observed during follow-up. Headaches were the most common neurologic complaint, occurring in 63% of all children diagnosed with Chiari I during the entire period of study. All but 1 of the 33 children who developed symptoms complained of headaches. Although headaches were usually present at the time of diagnosis, an additional 3 children (6%) developed headaches during follow-up. Over half (58%) of the 31 children with headaches experienced either moderate (11) or severe (5) headache syndromes, leading to repeated physician visits spanning more than a year. Although adults with Chiari I frequently report headaches in the occipital region, only 6% of our pediatric Chiari I headache patients described occipital pain. Head pain triggered by Valsalva maneuver or cough was similarly infrequent (4%) in our cohort.

Only 4 of 19 patients (21%) who had been symptom-free at the time of initial diagnosis developed new neurologic problems. Of these 4 children, 3 developed headaches, including 1 with a severe headache syndrome, and 2 developed tremor and poor coordination during follow-up. Two children with symptomatic Chiari I received a physician diagnosis of depression during follow-up.

Of the 51 children with Chiari I, 8 (15%) underwent sub-occipital decompression surgery. The 5 children who received surgery due to syringomyelia all experienced a significant decrease in syrinx size following surgical decompression. Three children underwent surgical treatment for daily intractable headaches; two experienced complete resolution of headaches for up to 7 years of follow-up. The third child's intractable headaches resolved soon after surgery, but resumed with equal intensity 12 months later.

Predictors of neurologic symptoms

Half (49%) of all children with Chiari I had significant neurologic symptoms defined as moderate to severe headache syndrome, neck pain, vertigo or ataxia. Age at diagnosis was a predictor of significant neurologic symptoms, with older children showing an increased risk (OR 1.2, 95% CI 1.04–1.4). Older age at diagnosis was also predictive of headache occurrence (OR 1.3, 95% CI 1.1–1.5). In contrast, none of the radiologic characteristics we measured, including degree of tonsillar ectopia, tonsillar morphology or retrocerebellar cerebrospinal fluid compression, were predictive of headaches or significant neurologic symptoms.

Borderline ectopia

Borderline tonsillar ectopia (2–4 mm) was noted in 19 patients, or 0.4% of all head and spine MRI's performed in our study population. Interestingly, although this mild degree of tonsillar ectopia is typically considered to be a normal variant, headaches were present in 74% of these children, and a severe headache syndrome was described in 16%. Occipital headache was present in 2 children (11%), and headache upon Valsalva in 1 child (5%). Whereas pointed tonsils or retrocerebellar CSF compression was present in the majority of patients with Chiari I, none of the children with borderline tonsillar ectopia exhibited these radiologic abnormalities ($P<0.0001$).

Discussion

Despite having first been recognized over a century ago [24], Chiari I was difficult to diagnose before the advent of MRI, and little is known about the epidemiology and natural history of Chiari I, particularly in children. The prevalence of Chiari I remains unknown. In this first population-based study of Chiari I in children, we found that one-third of children diagnosed with Chiari I were asymptomatic at the time of diagnosis, and the majority of symptomatic patients complained of headaches. Older age at diagnosis predicted worse neurologic symptoms.

Chiari I is a multifactorial condition that is thought to result from a congenitally small posterior fossa in many cases [10, 25–27]. Although familial cases suggest a genetic component [10, 28], acquired conditions such as spontaneous intracranial hypotension can also mimic Chiari I [29]. The classic clinical syndrome seen in young adults with Chiari I consists of occipital pain precipitated by cough or Valsalva [10]. Although children also present with headache, occipital and cough-induced pain are less common [4]. Other neurologic symptoms that have been described in children with Chiari I include ataxia, sensory and motor deficits, and lower cranial nerve abnormalities such as apnea and hoarseness [5, 10, 22, 30–35].

We found that older age at diagnosis predicted more severe symptoms. This is consistent with the observation that symptoms of Chiari I take time to develop, often becoming clinically apparent during the third and fourth decades [20]. Compared to adult series in which only 14–30% of patients are asymptomatic [18, 36], as many as 37–57% of children are symptom-free at the time of initial Chiari I diagnosis [4].

Adults are more likely to exhibit syringomyelia (59–76%) [10, 20, 37] than are children (14–58%) [4, 11]. Although the pathogenesis of syringomyelia in the setting of Chiari I is debated [16, 38–41], it has been suggested that increased pulse pressure in the subarachnoid space causes cerebrospinal fluid to move through the interstitial spaces of the spinal cord toward the central canal resulting in edema of the spinal cord, which may progress over time to expansion of the central canal [38].

Much remains unknown about the natural history of Chiari I. In particular, few data exist regarding the progression of symptoms in individuals with asymptomatic Chiari I. In one study, 89% of asymptomatic patients with more than 10 mm tonsillar ectopia remained symptom-free after 10 years [42]. We found that 21% of asymptomatic children with Chiari I subsequently developed headaches or tremor during available follow-up. A particular problem is whether a new headache is related to Chiari I, or if a common alternate etiology, such as migraine or tension headaches, is responsible for the patient's symptoms. Of note, spontaneous resolution of childhood Chiari I has been described in several cases [43, 44].

For these and other reasons, there is controversy regarding the indications for surgery [45–47]. In patients who are clearly symptomatic, studies indicate that surgical treatment of Chiari I often results in either improvement or stabilization of symptoms [11, 14, 20, 37, 48, 49], and that outcomes are particularly favorable in children [5, 12]. It is also well-established that syringomyelia either completely resolves or decreases in size following Chiari I decompression [6, 20, 23, 49–51], as was observed in our study. Although the long-term outcome of untreated syringomyelia is unclear, it seems reasonable that the majority of neurosurgeons would surgically treat a Chiari I malformation in the setting of syringomyelia [44, 45]. In contrast, based on an international survey of neurosurgeons, only 46% would operate on a patient with occipital headaches as the only symptom in the absence of syringomyelia [45]. This is due, in part, to the variability of headache severity and the subjective quality of pain perception that may determine if an individual patient is

appropriate for surgical treatment. Surgeons are also aware that there are no clinical trials of the benefits and harms of surgery, nor are there adequate case-series of the decades-long consequences of the competing approaches to surgical decompression. Thus, mild headaches or headaches that are clearly migrainous in origin should be primarily managed medically.

Another group that generates controversy involves patients with asymptomatic Chiari I, whose diagnoses are made from imaging studies obtained for unrelated reasons. Our results indicate that the majority of asymptomatic Chiari I patients do not progress to new headaches or neurologic signs. Our study was too small, however, to reveal features in the asymptomatic group that would determine which patients are more likely to become symptomatic.

The frequency of Chiari I reported in other consecutive head MRI series carried out for clinical indications in adults (0.6–0.9%) [4, 18, 36] is similar to the 1% frequency we observed in our population. One recent study, which examined a consecutive series of healthy adults unselected for symptoms and signs that could represent Chiari I, found the prevalence of Chiari I to be 0.9% [52], suggesting that it may be surprisingly common for people with Chiari I to live their whole lives with few or no symptoms.

Unlike previous series in tertiary care settings that are limited by selection bias, this study provides valuable information regarding the symptoms and natural history of Chiari I in a general pediatric population. However, the cohort of patients with Chiari I were selected from children who underwent MR imaging for symptoms related either to the Chiari I or to other diagnoses. Although this reflects the frequency of Chiari I in children who undergo MR imaging, the true prevalence of asymptomatic Chiari I in the general pediatric population remains unknown. As in any retrospective study, errors may result from inaccurate or incomplete coding of medical records. We may also have missed cases of Chiari I in which the treating radiologists failed to recognize or record the presence of tonsillar ectopia in the clinical report.

Although 5 mm of tonsillar ectopia has been widely adopted as a “cut-off” for diagnosing Chiari I [18, 53, 54], there is increasing recognition that patients with lesser degrees of tonsillar ectopia may also develop classic neurologic symptoms and syringomyelia that are amenable to neurosurgical intervention [10, 55]. Thus, the definition of Chiari I malformation may need to be revised [30]. Furthermore, the cerebellar tonsils typically ascend with age and are situated lower during early childhood [56], raising the concern that a 5 mm cut-off may not be appropriate in the pediatric population. Additional work is needed to refine our definition of Chiari I, to improve our understanding of the neurologic symptoms and natural history of Chiari I, and to develop evidence-based strategies for deciding which patients should undergo neurosurgical intervention.

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Abbreviations

KPMCP Kaiser Permanente Medical Care Program

OR	odds ratio
CI	confidence interval

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Figure.

17 year old girl with leg weakness and headaches. On this MRI sagittal T1 Spin Echo Image (TR 450, TE 13), the line demarcates the foramen magnum. Cerebellar tonsils (bold arrow) are slightly pointed, 7 mm below the foramen magnum with significant compression of the retrocerebellar cerebrospinal fluid space. A small cervical syrinx (arrow) is present.

Table 1

Neurologic Symptoms Reported in 51 children with Chiari Type I Malformation within a General Pediatric Population.

Neurologic Symptoms	Present at Diagnosis		Developed in Follow-up*		Present Ever	
	N	%	N	%	N	%
Headache	28	55	3	6	61	61
Neck pain	6	12	5	10	22	22
Vertigo	4	8	3	6	14	14
Any upper extremity numbness	3	6	4	8	14	14
Ataxia	3	6	2	4	10	10
Upper or lower extremity weakness	1	2	2	4	6	6
Tinnitus or hearing loss	1	2	2	4	6	6
Dysarthria	1	2	1	2	4	4
Loss of consciousness	1	2	1	2	4	4
Scoliosis with syrinx	1	2	0	0	2	2
Depression	0	0	2	4	4	4
None of above [†]	19	37	N/A	N/A	31	31

* Mean follow-up was 6.4 years (SD 4.1).

[†] 19 children, with none of the symptoms listed above, received neuroimaging for the following indications: seizure (6), developmental delay (4), psychosis (2), chorea (1), neurofibromatosis type I (1), follow-up of arteriovenous malformation (1), Bell's Palsy (1), retinal dystrophy (1), microphallus (1), and scoliosis without evidence of syrinx (1).