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Chiari I malformation in children—the natural history

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Abstract

Purpose To review the natural history of asymptomatic and symptomatic pediatric Chiari I malformations with and without syringomyelia.

Materials and methods We reviewed the literature for case reports and case series describing the natural history of asymptomatic and symptomatic children with Chiari I malformations with and without syringomyelia. Our review included approximately 700 asymptomatic children without syringomyelia, 100 symptomatic children without syringomyelia, 22 asymptomatic children with syringomyelia, and 11 symptomatic children with syringomyelia. Symptomatic and imaging outcomes at the point of last reported follow-up were noted to describe the natural history of Chiari I malformations in children.

Results Our review of about 700 asymptomatic children with CM-I without syrinx revealed that most children do not exhibit new-onset symptoms (5–6%) or syrinx (2–3%). The nearly 100 published cases of symptomatic CM-I without syrinx suggest that about half of children report symptomatic improvement (48%) and few report symptomatic worsening (7%). New-onset syrinx is rarely observed (2%). Few cases have been published about asymptomatic and symptomatic CM-I with syrinx as syringomyelia are generally regarded to be an indication for surgical intervention. Nevertheless, all 22 children with asymptomatic CM-I with syringomyelia included in this study were asymptomatic at follow-up, with syrinx resolution observed in 18 children and tonsillar herniation improvement observed in 16 children. Overall, our review of asymptomatic pediatric CM-I with or without syringomyelia suggests that its natural history is much more favorable than previously acknowledged and that the literature generally favors conservative management of these cases.

Conclusion Our review of asymptomatic pediatric CM-I with or without syringomyelia suggests that its natural history is much more favorable than previously acknowledged and that the literature generally favors conservative management of these cases. Further study of symptomatic pediatric CM-I is necessary to better understand its natural history.

Keywords Chiari I malformation · Pediatric · Natural history · Syringomyelia · Conservative management · Review

Introduction

Chiari malformations (CM) consist of a variety of disorders characterized by displacement of the cerebellum with or without the inferior medulla into the spinal canal. Chiari malformations are frequently accompanied by anatomic abnormalities of the brainstem, cerebellum, and craniocervical junction [1]. CM were first described by John Cleland in 1883 and classified by Hans Chiari in 1891 [2, 3]. There are four types of CM. Chiari I malformations (CM-I) present with irregularly shaped cerebellar tonsils and downward displacement of the cerebellar tonsils below the foramen magnum.

CM-I is relatively common in pediatric patients, with an imaging incidence of 3.6 %. Nearly, a quarter of these patients present with a syrinx [4]. With that said, tonsillar herniation naturally decreases from childhood to adulthood, suggesting that some children may be able to outgrow a CM-I diagnosis [5]. On the other hand, CM-I may cause symptoms that significantly impair quality of life or can result in syrinx formation and expansion which can be resolved through neurosurgical intervention [6–9]. Currently, asymptomatic patients without syrinx are often followed clinically or with MRI surveillance for the interval development of syringomyelia. Highly symptomatic patients are generally managed surgically. The management of mildly symptomatic children without syringomyelia or children with stable syringes is debated. In this study, we review

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the natural history of asymptomatic and symptomatic children with and without syringomyelia.

Disease onset

Although the onset of CM-I was initially thought to occur in adolescence and adulthood, reviews of neuroimaging in asymptomatic children suggest that CM-I frequently presents in young children as well, though it is often asymptomatic [10, 11].

Epidemiology

CM-I is an imaging diagnosis, made when the cerebellar tonsils extend 5 mm or further below the foramen magnum. Determining the true population prevalence of CM-I is difficult, as neuroimaging is not typically obtained on asymptomatic patients. The imaging prevalence of CM-I is therefore likely an overestimate of the population prevalence. Strahle et al. reviewed the brain or cervical MRIs of 14,116 pediatric patients and found that 3.6 % of patients had CM-I [4]. The male to female ratio in this study was close to 1:1. Twenty-three percent of patients had a spinal cord syrinx with 86% of the syringes being found in the cervical spine. Syringes were almost 2.5× more common in females as males. Almost 70 % of patients were asymptomatic. Girls were more likely to be symptomatic than boys.

The imaging prevalence of CM-I is higher in studies including only pediatric patients compared to studies that have included adults [4]. This is likely due in part to lifetime dynamics of cerebellar tonsil position. A review of 2,400 MRIs from patients of different age groups from Smith et al. suggests that downward displacement of the cerebellar tonsils relative to the foramen magnum decreases from childhood to early adulthood and then increases afterwards [5].

Clinical presentation

The most common symptom of CM-I is pain, particularly headache localized to the suboccipital or neck area. The pain is exacerbated by activities that increase intracranial pressure including laughing, coughing, running, and playing sports. Pain can also localize to the shoulder, back, and upper and lower extremities. Sensory changes, including numbness and paresthesia, can occur and can lead to unnoticed injuries or fractures. Less commonly, clumsiness, respiratory dysrhythmias, dysphagia, dysarthria, hoarseness, facial numbness, and urinary incontinence are observed. These presentations can lead to delayed or mistaken diagnoses such as cerebral palsy, especially in newborns [12]. Neonates may present with a rapid neurological decline within the first few days of life if brainstem function is impaired. Neurogenic dysphagia, manifesting as cyanosis during feeding, pooling oral secretions, and decreased gag reflex can be severe in neonates. Apneic spells, stridor, aspiration, arm weakness, opisthotonos, downbeat nystagmus, facial weakness, and weak or absent cry are other potential symptoms [13–15].

Findings on imaging

CM-I is diagnosed radiographically by identifying the abnormal extension of the cerebellar tonsils more than 5 mm below the foramen magnum. In infants, it is important to note that distances greater than 6 mm are required to be considered pathologic. Imaging should include the spinal cord given the high prevalence of patients with syringomyelia. While most commonly occurring in the cervical spinal cord, 20 % of cases of syrinx are holocord [16].

Natural history

Methodology

In this section, we review the natural history of asymptomatic and symptomatic children with CM-I with and without syrinx. We separated case series into these four separate groups wherever possible. However, if the authors did not report outcomes for these individual groups, then we discussed the case series with the group that was most appropriate. We explicitly emphasize this shortcoming in the discussion of the studies when relevant.

Asymptomatic CM-I without syringomyelia

We identified a total of seven studies which together followed about 700 asymptomatic or mildly symptomatic patients diagnosed with CM-I without syringomyelia (Table 1).

The study by Novegno et al. followed ten asymptomatic children with CM-I over an average of 5.9 years [6]. CM-I had been incidentally diagnosed in these children as a result of an MRI being ordered for other reasons, such as neck injury, macrocrania, epilepsy, development delay, paranasal sinusitis, trauma, visual defect, or precocious thelarche. Over the follow-up period, two children developed new-onset neck pain and one child developed a new-onset headache. One child that developed neck pain and one that developed new-onset headache also developed cervical syrinx. Syrinx formation was also observed in a child that was asymptomatic at the end of the study, for a total of 30% of children in this cohort developing syrinx. The two children with new symptoms and syrinx formation during this study were ultimately treated with third ventriculostomies. Following treatment, one child was subsequently asymptomatic and the other exhibited substantial symptomatic improvement. On imaging, tonsillar herniation worsened in two patients, improved in one patient, and was stable in the remaining seven patients. While one of the children with worsening tonsillar herniation had developed new-onset syrinx and new-onset neck pain, the other child

Table 1 The natural history of about 700 asymptomatic pediatric patients diagnosed with CM-I

Reference	Year	Group and sample size	Follow-up	New-onset symptoms	Tonsillar herniation at follow-up	New syrinx formation	Patients needing surgery
Castillo and Wilson	1995	Asymptomatic (1)	4 years	0	Resolved: 1	0	0
Novegno et al.	2008	Asymptomatic (10)	5.9 years (mean)	3	Worsened: 2; improved: 1	3	2
Aitken et al.	2009	Asymptomatic (17); asymptomatic with syrinx (2)	6.4 years (mean)	4	NA	0	NA
Strahle et al.	2011	Asymptomatic or mildly symptomatic with syrinx (134); asymptomatic or mildly symptomatic with syrinx (13)	4.6 years (mean)	9	Worsened: 6; improved: 45 (resolved: 7)	7	14
Benglis et al.	2011	Asymptomatic or symptoms not attributable to CM-I (110)	2.83 years (median)	0	NA	0 of 25	0
Pomeraniec et al.	2016	Asymptomatic (25)	5.5 years (mean)	0	NA	3	3
Leon et al.	2019	Asymptomatic (427)	2.13 years (mean)	NA	NA	5	15

was asymptomatic at follow-up and did not exhibit syrinx formation.

Aitken et al. reported on the natural history of 19 children incidentally diagnosed with CM-I due to seizure, developmental delay, psychosis, chorea, neurofibromatosis I, arteriovenous malformation, Bell's palsy, retinal dystrophy, microphallus, and scoliosis [7]. Four of the 19 children developed symptoms over this period. Three developed headaches, two developed tremor and poor coordination, and two were diagnosed with depression.

Strahle et al. reported the natural history of 147 children with CM-I that were asymptomatic or minimally symptomatic [17]. While we discuss all patients included in Strahle et al.'s study in this section, it should be noted that 11 of the patients in this study were symptomatic at the beginning of the study and 13 patients had syrinx at the beginning of the study. While we are unable to differentiate the outcomes of these different groups from each other, we discuss the cohort as a whole in this section because the majority of patients were asymptomatic without syrinx. Five patients were symptomatic throughout the entirety of the study, six patients were symptomatic at the beginning of the study but not at the end, and nine patients exhibited new-onset of symptoms. Syrinx was present in 13 patients at the beginning of the study and seven patients exhibited new-onset syrinx. The mean time to syrinx development was 28 months. Tonsillar herniation increased in six patients and decreased in 45 patients, with a resolution of CM-I in seven patients. The authors noted that patients younger than 6 years old exhibited an increase in tonsillar herniation over the course of the study, whereas children between the ages of six and 18 exhibited decreased tonsillar herniation. Fourteen patients in this cohort ultimately underwent surgery due to headaches, sleep apnea, and syrinx progression. The mean time to surgery in this cohort was 2.1 years.

Benglis et al. followed 110 children with CM-I who were asymptomatic or whose symptoms were not attributed to CM-I according to the neurosurgeons responsible for their care [8]. None of the patients developed new symptoms over the follow-up period (average of 2.83 years) that the treating neurosurgeon attributed to CM-I. Twenty-five of these patients had multiple brain and spine MRIs, and syrinx formation was not seen in any of these patients over the follow-up period. All 110 children were managed conservatively throughout the length of this study.

A previous study from our institution described the natural history of 25 asymptomatic children with CM-I without syrinx followed over an average of 5.5 years [9]. Although none of these children developed new-onset symptoms at the time of the last follow-up, three children exhibited new syrinx formation. These syringes were deemed to be unstable and so these three children were ultimately treated surgically.

Leon et al. reported the natural history of 427 asymptomatic pediatric patients diagnosed with CM-I (median length of follow-up, 2.1 years) [18]. Fifteen patients ultimately were managed surgically over the course of the study. The median time to surgery was 21.0 months. Surgical intervention was recommended due to new-onset syrinx and headaches. Five patients developed new-onset syrinx over the course of the study. Children with headaches, syringomyelia, scoliosis, extremity weakness and numbness, and sleep apnea exhibited improvement following surgery. Age, sex, and insurance type were not predictors of need for surgical intervention in this cohort.

Symptomatic CM-I without syringomyelia

We identified a total of four studies describing the natural history of almost 100 symptomatic children with CM-I without syringomyelia (Table 2).

Table 2 The natural history of about 100 symptomatic pediatric patients with CM-I without syringomyelia

Reference	Year	Group and sample size	Follow-up	Symptoms at follow-up	Tonsillar herniation at follow-up	New-onset syrinx	Patients requiring surgery
Novegno et al.	2008	Symptomatic without syrinx (11)	5.9 years (mean)	Improved: 9 (asymptomatic: 7); worsened: 2	Improved: 3 (resolved: 1); stable: 8	0	1
Aitken et al.	2009	Symptomatic without syrinx (28); symptomatic with syrinx (4)	6.4 years (mean)	NA	NA	0	NA
Benglis et al.	2011	Symptomatic without syrinx (12)	2.83 years (median)	Improved: 5; worsened: 3	NA	0	Recommended: 3 (parents refused)
Pomeraniec et al.	2016	Symptomatic without syrinx (46); symptomatic with syrinx (2)	5.5 years (mean)	Improved: 20	NA	2	0

The study by Novegno et al. included 11 children whose symptoms could be attributed to CM-I [6]. Symptoms included headache ($n = 6$), vertigo ($n = 2$), neck pain ($n = 2$), gait instability ($n = 1$), nausea and vomiting ($n = 1$), and papilledema ($n = 1$). At the end of the study, seven of the 11 children were asymptomatic. One child with neck pain and one with headache reported symptomatic improvement and one child with headache and vertigo and another with headache reported worsening symptoms. The child with worsening headache and vertigo ultimately underwent suboccipital craniectomy. By the end of the study, her vertigo had resolved but her headaches continued to persist. On follow-up imaging, tonsillar herniation improved in three patients and was stable in the remaining eight children. CM-I entirely resolved in one of the children showing improvement on imaging.

Aitken et al. described the natural history of 32 children with symptomatic CM-I [7]. Four of these children were also diagnosed with syringomyelia. The most common symptoms exhibited by these children included headache (55%), neck pain (12%), vertigo (8%), sensory changes (6%), and poor coordination (6%). Three of the children with syringomyelia presented with extremity weakness or numbness. While the authors did not report whether or not the children improved symptomatically at the end of the study, they did report that children in this group exhibited new-onset neck pain ($n = 5$), vertigo ($n = 3$), tinnitus or hearing loss ($n = 2$), dysarthria ($n = 1$), or loss of consciousness ($n = 1$) during the duration of the study. None of the 28 children with symptomatic CM-I without syringomyelia developed syringes over the course of this study.

The study by Benglis et al. included 12 children with symptomatic CM-I without syringomyelia [8]. Children presented with posterior headache ($n = 11$), neck pain ($n = 2$), and vomiting ($n = 1$). The median length of follow-up of this study was 2.83 years. No new objective neurological deficits were demonstrated in any children over the course of this study. Subjectively, five children reported improvements in their symptoms and three reported worsening of their symptoms by the end of the study. Surgery had been recommended to the parents of three children included in this cohort based on symptom severity, two with posterior headaches and one with vomiting, though the parents refused surgery. The children with posterior headaches reported worsening of their headaches over time whereas the child with vomiting reported symptomatic improvement.

A previous study from our institution included 48 symptomatic children with CM-I, two of which also presented with syringomyelia [9]. The mean follow-up time for our study was 5.5 years. Children presented with headache ($n = 40$), swallowing difficulty ($n = 7$), nausea and vomiting ($n = 7$), motor deficits ($n = 3$), sensory complaints ($n = 2$), and sleep apnea ($n = 1$). New syrinx was observed in two children. None of the patients in this group were ultimately surgically managed. Twenty of the 48 patients in our cohort reported symptomatic improvement at the time of the last follow-up.

Asymptomatic CM-I with Syringomyelia

We identified a total of seven studies describing the natural history of 22 asymptomatic children with CM-I with syringomyelia (Table 3). Given the small sample size, it is possible that a review of asymptomatic CM-I with syringomyelia based only on these studies may be affected by publication bias. Nevertheless, we present what is known about the natural history below.

Avellino et al. described a case of a 5-year-old boy incidentally diagnosed with CM-I and syringomyelia due to complex partial seizure disorder [19]. The syringes extended from C3 to C7 and from T5 to T8. The syringes had decreased in size by the time he was 7.5 years old and had entirely resolved by

Table 3 The natural history of 22 asymptomatic children with CM-I and syringomyelia

Reference	Year	Group and sample size	Follow-up	Symptoms at follow-up	Tonsillar herniation at follow-up	Syrinx at follow-up	Patients requiring surgery
Avellino et al.	1996	Asymptomatic with syrinx (1)	7 years	Asymptomatic: 1	Improved: 1	Resolved: 1	0
Sun et al.	2000	Asymptomatic with syrinx (1)	2 years	Asymptomatic: 1	Resolved and subsequently recurred: 1	Resolved and subsequently recurred: 1	0
Sun et al.	2001	Asymptomatic with syrinx (1)	7 years	Asymptomatic: 1	Resolved: 1	Resolved: 1	0
Tokunaga et al.	2001	Asymptomatic with syrinx (15)	4.9 years (mean)	Asymptomatic: 15	Improved: 13; unchanged: 2	Resolved: 15	0
Jatavallabhula et al.	2006	Asymptomatic with syrinx (1)	5 years	Asymptomatic: 1	Resolved: 1	Resolved: 1	0
Novegno et al.	2008	Asymptomatic with syrinx (1)	19 years	Asymptomatic: 1	Stable: 1	Stable: 1	0
Massimi et al.	2011	Asymptomatic with syrinx (2)	5 years (mean)	Asymptomatic: 2	Stable: 2	Stable: 2	0

the time he was 10.5 years old. He was followed until age 12 and had remained asymptomatic throughout all 7 years. Three other case reports reported the natural history of children with CM-I and syringomyelia who were asymptomatic at presentation [20–22]. All three children were asymptomatic at follow-up. Syrinx resolution was observed in two out of the three children. In the remaining case, the syrinx was found to have resolved and then had subsequently recurred [21].

Tokunaga et al. described the natural history of children with scoliosis and syringomyelia [23]. Fifteen of these children were asymptomatic and had also been diagnosed with CM-I. After an average follow-up of 4.9 years, the authors found that all of the children were asymptomatic at follow-up. Tonsillar herniation had decreased in 13 of the children and remained stable in two. No children had syrinx at follow-up.

The study by Novegno et al. included one patient with CM-I and syringomyelia incidentally diagnosed due to developmental delay [6]. His syrinx extended from C-4 to C-7 and had a width less than 4 mm. He also exhibited platybasia and mild ventriculomegaly. Over the course of 19 years, he remained asymptomatic and his tonsillar herniation and the size of his syrinx remained stable.

The study by Massimi et al. included two asymptomatic children with CM-I and syringomyelia [24]. They were incidentally diagnosed due to seizures and behavioral disturbance. One patient was followed for 3 years and the other was followed for 7 years. Both were asymptomatic at the end of the study and had stable syringes.

Symptomatic CM-I with syringomyelia

As with asymptomatic CM-I with syringomyelia, the field's understanding of the natural history of symptomatic CM-I with syringomyelia is limited as most symptomatic patients with CM-I and syringomyelia are surgically managed [9]. Nevertheless, we identified 17 cases of conservatively managed CM-I with syringomyelia that provide insight into the natural history of children with this diagnosis (Table 4).

Sudo et al. reported a case of a child with pain and weakness with CM-I and syringomyelia who exhibited symptomatic and radiologic resolution of CM-I and syringomyelia after 2.25 years [25]. Avellino et al. reported a case of a 5-year-old girl presenting with unsteady gait who was subsequently diagnosed with CM-I and syringomyelia [19]. By age 16, she reported substantial improvement of her unsteady gait. Tonsillar herniation and the syrinx were also substantially improved.

The study by Tokunaga et al. included six symptomatic children with scoliosis, syringomyelia, and CM-I. Headache ($n = 6$) and muscle weakness ($n = 2$) were the most commonly reported symptoms in these patients. After an average of 4.9 years, two of the patients exhibited decreased tonsillar herniation and four exhibited no change in tonsillar herniation. The syringes had resolved in all six patients at the end of the study. Objective neurological deficits were found to have improved in two patients and remained unchanged in the other four.

The series by Benglis et al. included seven children with symptomatic CM-I and syringomyelia [8]. Children presented with headache ($n = 6$), low back pain ($n = 1$), and neck pain ($n = 1$). Six of the patients reported improvement in their symptoms but one of the children with headaches reported worsening of her headaches. Although surgery was recommended to her, her parents refused surgery.

Our prior study included two children with symptomatic CM-I and syringomyelia managed conservatively due to stable syrinx and abnormal skull base development [9]. One presented with motor symptoms and the other with a

Table 4 The natural history of 11 pediatric patients with CM-I and syringomyelia

Reference	Year	Group and sample size	Follow-up	Symptoms at follow-up	Tonsillar herniation	Syrinx at follow-up	Patients requiring surgery
Sudo et al.	1990	Symptomatic with syrinx (1)	2.25 years	Resolved: 1	Improved: 1	0	0
Avellino et al.	1999	Symptomatic with syrinx (1)	11 years	Improved: 1	Improved: 1	0	0
Tokunaga et al.	2001	Symptomatic with syrinx (6)	4.9 years (mean)	NA	Improved: 2; unchanged: 4	0	0
Benglis et al.	2011	Symptomatic with syrinx (7)	2.83 years (median)	Improved: 6 (resolved: 1); worsened: 1	NA	NA	Recommended: 1 (parents refused)
Pomeraniec et al.	2016	Symptomatic with syrinx (2)	5.5 years (mean)	Stable: 1; worsened: 1	NA	Stable: 1; worsened: 1	0

headache. The child with motor symptoms had an unchanged syrinx and reported no change in her symptoms at the end of the study, whereas the child who reported headaches exhibited enlargement of the syrinx and worsening headaches.

Summary statistics and conclusion

The increased use of MRI has resulted in the increased incidence of CM-I in pediatric populations with a previous study reporting the imaging incidence as 3.6% [4].

Because herniation of the cerebellar tonsils naturally decreases from childhood to adulthood, deciding whether or not a child with CM-I should be treated surgically is challenging [5].

As part of our review of the natural history of children with CM-I without syrinx, we found that a small percentage of children (5–6%) develop new-onset symptoms. Tonsillar herniation frequently improves (29%) or at least remains stable (65%). Worsening tonsillar herniation is rare (5%), as is new-onset syrinx (2–3%). Our findings validate the field's general approach to managing these children through regular follow-up, reserving surgical consideration for children with new-onset symptoms or syrinx.

Less is known about children presenting with symptomatic CM-I without syrinx, as we were only able to review about 100 previously published cases. Many patients reported symptomatic improvement (48%) with a small percentage reporting worsening of their symptoms (7%). New-onset syrinx formation is quite rare (2%). There are currently no validated methodologies for predicting which children will exhibit worsening symptoms and which will exhibit symptomatic improvement, including the initial symptom that the child presented with. In our previous study, we had found that children with CM-I and sleep apnea or dysphagia managed surgically exhibited high rates of symptomatic improvement [9]. Therefore, at this point, deciding whether or not surgery should be recommended should be based on how significantly the child's symptoms are impairing daily life and the family's preferences for surgical management.

Our understanding of the natural history of CM-I with syrinx may be affected by publication bias. We described the natural history of 22 children with asymptomatic CM-I and syrinx in this review, all of which were asymptomatic at follow-up. Tonsillar herniation improved in 16 children (72%) and was unchanged in five (23%). At follow-up, the syringes were found to be resolved in 18 children (82%) and stable in three (14%). None of the patients required surgery. We identified 17 cases of symptomatic CM-I with syrinx. We found that most children reported symptomatic improvement (73%) and only one of the total of 11 children included in studies with symptom outcome information reported worsening symptoms (9%). Of the eight cases for which tonsillar herniation outcomes were described at follow-up, tonsillar herniation was found to be improved in four patients (50%) and unchanged in the other four patients (50%). Of five cases reporting syrinx imaging results at follow-up, the syrinx was resolved in three cases (60%), unchanged in one case (20%), and worsened in one case (20%). The decision of whether or not to operate on children with syrinx varies from one institution to another, though many neurosurgeons view syringomyelia as an indication for surgical intervention. Future case series reporting the natural history of children managed conservatively due to their parents refusing surgery may provide the field with a more unbiased understanding of the natural history of CM-I with syrinx, though patient selection bias will likely remain a limitation of these studies.

Overall, this study suggests that the natural history of asymptomatic Chiari I malformation in children with or without syringomyelia is much more favorable than previously acknowledged. Pediatric neurosurgeons should be aware that, in general, the literature favors conservative management of asymptomatic Chiari I malformation in children. Further study of symptomatic Chiari I malformation is necessary to better understand its natural history.

Compliance with ethical standards

Conflict of interest The authors have no conflicts of interest to report.

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