Published in final edited form as:

J Neurosurg Pediatr. 2008 February ; 1(2): 118–123. doi:10.3171/PED/2008/1/2/118.

Time course of syringomyelia resolution following decompression of Chiari malformation Type I

Nicholas M. Wetjen, M.D.^{1,2}, John D. Heiss, M.D.², and Edward H. Oldfield, M.D.²

¹Department of Neurologic Surgery, Mayo Clinic, Rochester, Minnesota ²Surgical Neurology Branch, National Institute of Neurological Disorders and Stroke, National Institutes of Health, Bethesda, Maryland

Abstract

Object—To better understand syrinx pathophysiology, the authors performed a prospective study in which they used findings from serial clinical and magnetic resonance (MR) imaging examinations performed before and after craniocervical decompression to establish the time course of syrinx narrowing.

Methods—Serial clinical examinations and cervical MR imaging were performed in 29 consecutive patients with Chiari malformation Type I (CM-I) and syringomyelia before surgery, 1 week, and 3–6 months after surgery, and then annually. Time to narrowing of the syrinx (> 50% decrease in maximal anteroposterior diameter) following surgery was calculated using the Kaplan–Meier method.

Results—All syringes decreased in diameter and length (number of segments) on MR images at 3–6 months, 1 year, and 2 years or later. The syrinx diameter decreased from 6.9 ± 2.1 mm (mean $\pm$ standard deviation) preoperatively to < 1.5 mm at last evaluation ($p < 0.0001$). The median time to syrinx narrowing was 3.6 months following CM-I decompression (95% confidence interval 3.0–6.5 months). After surgery 94% of patients had improved symptoms, but symptoms resolved incompletely in 68% of patients; 52 and 59% of patients had residual dysesthesias and sensory loss, respectively. Clinical improvement occurred before partial or complete disappearance of the syrinx on MR images. Patient age, duration of symptoms, sex, preoperative syrinx diameter, and length of syrinx were unrelated to time to syrinx narrowing.

Conclusions—Most patients improve after decompression for CM-I, but many have residual symptoms. Syringes may continue to diminish for months to years after surgical decompression. A collapsed syrinx (absence of distention of the spinal cord) indicates that the pathophysiology has been reversed by treatment regardless of the completeness of elimination of the cavity on MR images.

Keywords

Chiari malformation Type I; outcome assessment; pediatric neurosurgery; syringomyelia

Chiari malformation Type I is a disorder in which the cerebellar tonsils protrude through the foramen magnum and into the spinal canal. This anomaly is the leading cause of syringomyelia and occurs with or without associated osseous abnormalities at the craniovertebral junction.^{3–5,7–10,12–14} In contrast to other types of Chiari malformations, which usually present in infancy, CM-I tends to present during or after the second decade of life. Overcrowding of the hindbrain by an underdeveloped posterior fossa commonly causes tonsillar ectopia in CM-I.^{1,5,8,9,11,15,16} Magnetic resonance imaging has revolutionized the diagnosis of CM-I and has led to the detection of cases that previously were not recognized or were erroneously identified as other conditions. The resulting increase in reported cases has invigorated investigators to better understand the pathogenesis, clinical manifestations, and response to treatment of CM-I and syringomyelia.

The object of surgical treatment of syringomyelia associated with the CM-I is to eliminate the pathophysiology underlying the syrinx and prevent progression of myelopathy. A reduction in syrinx size usually occurs after craniocervical decompression, but an MR imaging study to document this change may not be obtained routinely by the physician if symptoms of myelopathy resolve or remain stable after surgery. Although the resolution or stabilization of clinical symptoms has been reported to accompany neuroimaging-documented narrowing of syringomyelia, the time course of syrinx narrowing has not been examined by serial imaging.

The time course of narrowing of syringomyelia following craniocervical decompression for CM-I is examined in a prospective clinical research protocol (Heiss et al., Establishing the Physiology of Syringomyelia, NINDS 92-N-0226) in which neurological examinations and spinal MR imaging are repeated before and at defined time points after surgery. In addition to physiological measurements, this approach allows determination of the rate of clinical improvement and of reduction in syrinx size after surgery.

When neurosurgeons evaluate patients after craniocervical decompression, it may be helpful to them to compare the progress of their patients with a cohort of similarly treated patients. In this study we establish a reference for the expected time to neuroimaging-documented narrowing of the syrinx and improvement of clinical findings after craniocervical decompression. We also explore whether complete disappearance of the syrinx is more important than relief of spinal cord distension for clinical stabilization of syringomyelia. Finally, we report the frequency of signs and symptoms of residual myelopathy that occurs after surgery that corrects syrinx pathophysiology. Understanding the expected imaging and clinical outcomes after craniocervical decompression for CM-I and syringomyelia will be useful for advising patients and for avoiding unnecessary secondary surgery.

Clinical Material and Methods

Study Protocol

Twenty-nine patients, 8 men and 21 women, mean age 37 ± 12 years (range 16–61 years) with syringomyelia and CM-I were enrolled in the clinical research protocol, Establishing the Physiology of Syringomyelia (NINDS 92-N-0226). The diagnosis of CM-I was based on displacement of the cerebellar tonsils inferior to the foramen magnum (mean 12 ± 6 mm,

range 3–23 mm) which constricted the CSF pathways.⁸ The mean follow-up duration was 3.0 ± 2.9 years (range 0.25–12.1 years). The institutional review board of the NINDS approved the research protocol. Informed consent was obtained from each patient and each healthy volunteer. All patients were evaluated and treated at the Clinical Research Center of the National Institutes of Health, Bethesda, Maryland.⁵

Preoperative Evaluation

Clinical Evaluation—Signs and symptoms were graded as “absent,” “mild,” “moderate,” or “severe.” Grades for symptoms of headache, dysesthetic pain, extremity weakness, sensory loss, and impaired ambulation and for signs of weakness, atrophy, and ataxia were recorded for later comparison to evaluations after surgery. Ambulation and weakness were graded on standard scales, and grades were converted to our scale for purposes of analysis. The ambulation scale was as follows: unaffected (normal ambulation), mild (with cane), moderate (with walker), and severe (nonambulatory). The weakness scale was as follows: unaffected (5/5), mild (4/5), moderate (3/5, antigravity strength), and severe (1–2/5, less than antigravity).

Neuroimaging—Midsagittal T1-weighted MR images of the posterior fossa and the cervical and thoracic spine were evaluated. The maximum anteroposterior diameter of the syrinx and length of the syrinx were measured. Because a T2 signal abnormality may represent either edema or free fluid within the spinal cord and T1 signal hypointensity clearly defines syrinx fluid, the T1 signal was used for all measurements. The T1 signal hypointensity that distended the spinal cord was considered to be syringomyelia. Only syringes meeting these criteria without other imaging abnormalities of the spinal cord were included for analysis. The length of the syrinx was recorded in spinal segments with a spinal segment defined as a vertebral body and intervertebral disc.

Surgical Treatment

All patients underwent surgery in a prone, horizontal position with the head in a neutral or gently flexed posture. Surgical treatment included suboccipital craniectomy, cervical laminectomy of C-1, and C-2 if necessary, opening of the dura in a Y shape leaving the arachnoid intact, and duraplasty. The dural graft was fashioned from a triangular autograft of occipital pericranium measuring 5×5 cm that was sewn to the cut edges of the dura with a continuous 4-0 nylon suture.^{5,13}

Postoperative Evaluation

Clinical Evaluation—Clinical evaluations were performed 1 week, 3–6 months, 1 year, and at subsequent yearly intervals after surgery. The clinical results were evaluated at each postoperative visit.

Postoperative Imaging—During each clinical visit, patients underwent midsagittal T1-weighted anatomical MR imaging of the posterior fossa and of the cervical and thoracic spine.

Statistical Analysis

The difference between maximal syrinx diameter measured before and at established time points after surgery was the primary outcome variable. Secondary analyses included clinical outcome for each individual, as determined by comparing the grades for signs and symptoms before surgery with those after surgery and determining if they were “improved,” “without change,” or “worse” for that visit. Other secondary analyses investigated the effect of patient age, degree of tonsillar ectopia in millimeters, location of the syrinx (cervical or cervicothoracic), and duration of symptoms on the length of the interval from surgery to imaging-documented narrowing and clinical resolution of syringomyelia. Data are expressed as mean $\pm$ standard deviation. Measurements from patients before surgery were compared to those after surgery using the paired Student t-test. The association between narrowing of the syrinx and resolution of clinical symptoms at each follow-up interval was tested with the chi-square test.

Multivariate regression models were developed to investigate which factors were associated with narrowing of the syrinx or resolution of clinical symptoms. Clinical factors that were tested included age of the patient at the time of surgery, sex, and duration of symptoms. Imaging factors tested included maximal syrinx diameter on preoperative imaging and the number of spinal segments over which the syrinx extended.

Time to narrowing of syrinx following surgery was calculated using the Kaplan–Meier method.⁶ Narrowing of the syrinx after surgery was defined as a $> 50\%$ reduction in syrinx diameter at the location of maximal cord distension on preoperative MR images. Curves for the various subgroups were compared using the log rank test. The Cox proportional hazards model was used to identify the univariate and multivariate predictors of time to syrinx narrowing. Crude and adjusted rate ratios and their 95% CIs were calculated. A probability value < 0.05 was considered significant. All data were recorded and entered into a spreadsheet (Excel, Microsoft Corp.), and statistical analyses were performed using SAS (Enterprise Guide 3, SAS Corp.).

Results

Clinical Evaluation

Symptoms had been present for 3 months to 37 years (mean 4.5 ± 7.5 years) before surgical consultation (Table 1), except for 1 patient who had no symptoms referable to the syrinx. By 6 months after surgery, 96% of patients had improvement of their clinical symptoms, and 4% had no change (Table 2). By 1 year after surgery all but 1 patient had improved. Improvement in clinical symptoms persisted during later follow-up. Despite improvement in overall clinical symptoms, most patients continued to have residual symptoms postoperatively at each follow-up interval. The proportion of patients who continued to have at least some symptoms following surgery was 78% at 3 months, 71% at 1 year, and 68% at 2 years. The most common symptoms and signs to persist after surgical treatment included painful dysesthesias (52%) and loss of sensation reported by the patient (48%) and on examination (59%).

Neuroimaging Evaluation and Time Course of Narrowing of the Syrinx

Before surgery the spinal cord was distended in all patients by syringes that extended longitudinally from 1 to 19 spinal segments (mean 10.0 ± 5.3). No patient had fluid communication between the fourth ventricle and the syrinx on MR images. Ten syringes were confined to the cervical portion of the spinal cord and 19 extended from the cervical to thoracic region of the spinal cord. The maximal diameter of the syrinx did not correlate with the number of spinal segments over which the syrinx extended; some patients had large diameter syringes that extended over only 1–2 spinal segments (Fig. 1). The CM-I was observed to severely narrow the anteroposterior diameter of the CSF pathways at the foramen magnum ventral to the medulla oblongata and dorsal to the cerebellar tonsils. The cerebellar tonsils extended below the level of the foramen magnum and their tips consistently exhibited an abnormal peg shape (Figs. 2–4).⁵

All syringes decreased in diameter and length on follow-up imaging studies at 3–6 months, 1 year, and 2 years or later after surgery (Fig. 5). The median time to syrinx narrowing (> 50% reduction in syrinx diameter) was 3.6 months after CM-I (95% CI 3–6.5 months). The mean time to narrowing of the syrinx (6.5 ± 1.8 months) was longer than the median time because some syringes took over 6 months to narrow. Mean syrinx diameter decreased from 6.9 ± 2.1 mm on the preoperative MR images to <1.5 mm at each postoperative interval beyond 1 week. The difference in mean diameter at preoperative evaluation to diameter at postoperative evaluation was statistically significant ($p < 0.0001$). The mean postoperative syrinx diameter at 3–6 months was 1.2 ± 1.5 mm, 1.2 ± 1.5 mm at 1 year, and 1.1 ± 1.4 mm at 2 years. The mean number of segments over which the syrinx extended after surgery was 3.9 ± 2.0 at 3–6 months, 2.3 ± 2.9 at 1 year, and 1.3 ± 2.6 at 2 years. At 6 months, 86% of syringes had decreased by > 50%; by 1 year, 91%; and by 2 years, 100%. The syringes initially decreased in size at a mean rate of 3.9 ± 8.0 mm/month. Many syringes (12 [41%] of 29) never disappeared completely but left a collapsed cavity in which fluid was still present. In those syringes that disappeared completely, the initial rate of reduction in syrinx diameter was 2.2 ± 2.4 mm/month. The difference in the initial rate of decrease between syringes that resolved completely after surgery and those that did not was not statistically significant ($p = 0.25$).

Univariate regression models and Cox proportional hazard models did not demonstrate a relationship between the time to narrowing of the syrinx and 1) patient age; 2) symptom duration; 3) patient sex; 4) preoperative syrinx diameter; 5) number of segments over which the syrinx extended; 6) location of the syrinx (cervical versus cervicothoracic); and 7) extent of tonsillar ectopia. Adjustment for any of these clinical and imaging factors did not result in a significant difference in time to narrowing of the syrinx.

Relationship of Narrowing of the Syrinx to Resolution of Symptoms

Clinical symptoms improved and the syrinx diameter was reduced following craniocervical decompression of the Chiari malformation. At 3–6 months, 96% of patients had improved clinically and 86% of the syringes had narrowed on MR images (Table 2). At 1 year 95% had improved clinically, and 91% of syringes had narrowed on MR images. Two or more years after surgery, 94% had improved clinically and 100% of syringes had narrowed. The

proportion of patients who were completely symptom free at each follow-up interval were 22% at 3 months, 29% at 1 year, and 32% at 2 years, with the remainder having residual signs and symptoms of myelopathy.

Discussion

Almost all of the syringes narrowed on imaging (diameter < one half of that before surgery) within the 1st year following decompression, and most of the syringes completely disappeared during the follow-up period. Although some patients had residual fluid within the spinal cord on serial MR images up to several years after surgery, their syringes were much smaller than before surgery and no longer distended the spinal cord. Because active syringomyelia resulted in loss of spinal cord substance, after surgery residual syrinx fluid filled this void as the spinal cord returned to its normal diameter. Patients had improvement in their clinical symptoms following decompression of the Chiari malformation, although complete resolution of symptoms was less common. Improvement in symptoms was similar in patients with complete and incomplete disappearance of the syrinx on MR images.

In patients with CM-I and syringomyelia, the piston-like movement of the cerebellar tonsils during the cardiac cycle causes accentuated cervical subarachnoid pressure waves that steadily drive CSF into the spinal cord, initiate syrinx formation, and, after the syrinx forms, create pulsatile motion of the syrinx fluid that extends the syrinx.^{5,13} Craniocervical decompression improves the flow of CSF across the foramen magnum, reduces the piston-like movement of the cerebellar tonsils on the cervical subarachnoid space, reduces syrinx fluid motion, and leads to syrinx narrowing.^{5,13} Although in our study craniocervical decompression reversed the pathophysiological process that led to the development of the syrinx, it could not reverse the anatomical destruction to the spinal cord caused by the distended syrinx before surgery, which is born out by the persistent signs and symptoms of myelopathy experienced by most patients after surgery despite the consistent collapse of the syrinx.

Only patients with distended syringes were treated in our study. Duration of symptoms did not correlate with time to neuroimaging-documented or symptomatic improvement in our patients. It is unknown how long each syrinx may have been present before producing symptoms. Therefore it is impossible to know the duration of each syrinx. It is reasonable to believe that patients with expanded syringes for longer intervals, even if asymptomatic, would have more loss of spinal cord tissue than those harboring syringes for shorter periods. One could speculate that the duration that the spinal cord had been under distension from an expanded syrinx might influence how quickly and completely the syrinx would resolve after surgery. In the current study relief of distension of the spinal cord rather than complete disappearance of syrinx fluid was the key to clinical improvement because patients with a small residual fluid-filled cavity within the spinal cord were not at greater risk of persistent symptoms than patients with complete disappearance of the syrinx.² The rate of syrinx narrowing after surgery could not be predicted by the size of the syrinx, age of the patient, or duration of symptoms before surgery.

Conclusions

This longitudinal prospective study defines the time course of neuroimaging-documented syrinx narrowing and symptomatic improvement after craniocervical decompression and duraplasty for CM-I malformation and syringomyelia. It confirms previous observations that surgical decompression of the CM-I reliably results in syrinx reduction. A favorable clinical outcome resulted from relief of spinal cord distension as the syrinx became smaller and did not require complete disappearance of syrinx fluid on MR images. Symptoms usually improved incompletely after surgery; residual signs and symptoms did not signal a failure of therapy, but rather confirmed that the syrinx permanently injured the spinal cord before surgery.

Acknowledgments

This work was supported by the Intramural Research Program of the National Institute of Neurological Disorders and Stroke, National Institutes of Health.

Abbreviations used in this paper

CI	confidence interval
CM-I	Chiari malformation Type I
CSF	cerebrospinal fluid
MR	magnetic resonance
NINDS	National Institute of Neurological Disorders and Stroke

References

1. Badie B, Mendoza D, Batzdorf U. Posterior fossa volume and response to suboccipital decompression in patients with Chiari I malformation. *Neurosurgery*. 1995; 37:214–218. [PubMed: 7477771]
2. Bogdanov EI, Heiss JD, Mendelevich EG. The post-syrinx syndrome: stable central myelopathy and collapsed or absent syrinx. *J Neurol*. 2006; 253:707–713. [PubMed: 16511636]
3. Dyste GN, Menezes AH. Presentation and management of pediatric Chiari malformations without myelodysplasia. *Neurosurgery*. 1988; 23:589–597. [PubMed: 3200390]
4. Dyste GN, Menezes AH, VanGilder JC. Symptomatic Chiari malformations. An analysis of presentation, management, and long-term outcome. *J Neurosurg*. 1989; 71:159–168. [PubMed: 2746341]
5. Heiss JD, Patronas N, DeVroom HL, Shawker T, Ennis R, Kammerer W, et al. Elucidating the pathophysiology of syringomyelia. *J Neurosurg*. 1999; 91:553–562. [PubMed: 10507374]
6. Kaplan EL, Meier P. Nonparametric estimation for incomplete observations. *J Am Stat Assoc*. 1958; 53:457–481.
7. Milhorat TH, Capocelli AL Jr, Anzil AP, Kotzen RM, Milhorat RH. Pathological basis of spinal cord cavitation in syringomyelia: analysis of 105 autopsy cases. *J Neurosurg*. 1995; 82:802–812. [PubMed: 7714606]
8. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, et al. Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. *Neurosurgery*. 1999; 44:1005–1017. [PubMed: 10232534]

9. Nishikawa M, Sakamoto H, Hakuba A, Nakanishi N, Inoue Y. Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. *J Neurosurg.* 1997; 86:40–47. [PubMed: 8988080]
10. Nishizawa S, Yokoyama T, Yokota N, Tokuyama T, Ohta S. Incidentally identified syringomyelia associated with Chiari I malformations: is early interventional surgery necessary? *Neurosurgery.* 2001; 49:637–641. [PubMed: 11523674]
11. Nyland H, Krogness KG. Size of posterior fossa in Chiari type 1 malformation in adults. *Acta Neurochir (Wien).* 1978; 40:233–242. [PubMed: 676804]
12. Oldfield EH. Cerebellar tonsils and syringomyelia. *J Neurosurg.* 2002; 97:1009–1010. [PubMed: 12450018]
13. Oldfield EH, Muraszko K, Shawker TH, Patronas NJ. Pathophysiology of syringomyelia associated with Chiari I malformation of the cerebellar tonsils. Implications for diagnosis and treatment. *J Neurosurg.* 1994; 80:3–15. [PubMed: 8271018]
14. Pillay PK, Awad IA, Little JR, Hahn JF. Symptomatic Chiari malformation in adults: a new classification based on magnetic resonance imaging with clinical and prognostic significance. *Neurosurgery.* 1991; 28:639–645. [PubMed: 1876240]
15. Schady W, Metcalfe RA, Butler P. The incidence of craniocervical bony anomalies in the adult Chiari malformation. *J Neurol Sci.* 1987; 82:193–203. [PubMed: 3440866]
16. Stovner LJ, Bergan U, Nilsen G, Sjaastad O. Posterior cranial fossa dimensions in the Chiari I malformation: relation to pathogenesis and clinical presentation. *Neuroradiology.* 1993; 35:113–118. [PubMed: 8433785]

Fig. 1.

On T1-weighted sagittal MR images, a syrinx that is present before surgery (A) is little changed by 1 week after surgery (B), but then progressively becomes smaller by 3 months (C) and 1 year (D) after surgery.

Fig. 2. Sagittal T1-weighted MR images obtained before surgery (A) and 3 months (B), 1 year (C), and 3 years (D) after surgery, demonstrating a typical course of syringomyelia narrowing following decompression for CM-I.

Fig. 3.

On T1-weighted sagittal MR images, a syrinx that is present before surgery (A) becomes smaller by 1 week (B), and has nearly disappeared by 3 months after surgery that opens CSF pathways at the foramen magnum.

Fig. 4.

On T1-weighted sagittal MR images, a syrinx associated with CM-I (A) is present before surgery; after surgery to open the CSF pathways at the foramen magnum, the syrinx becomes smaller by 1 week (B), is undetectable by 3 months (C), and does not recur at the 1- (D), 3- (E), and 5- (F) year follow-up visits.

Fig. 5. Graph depicting the proportion (*black line*) of patients with neuroimaging-documented narrowing of syringomyelia following surgical decompression. Dotted lines indicate the 95% CI.

TABLE 1

Clinical presentation in 29 patients undergoing surgical decompression for CM-I and syringomyelia

Symptoms & Signs	No. of Patients (%)			
	Preop Severity			
	Absent	Mild	Moderate	Severe
symptom				
headache at rest	14 (48)	4 (14)	10 (34)	1 (3)
headache w/cough	21 (72)	1 (3)	7 (24)	0 (0)
dysesthetic pain	5 (17)	6 (21)	16 (55)	2 (7)
subjective weakness	19 (66)	9 (31)	0 (0)	1 (3)
sensory loss noted by patient	6 (21)	7 (24)	13 (45)	3 (10)
impaired ambulation *	20 (69)	8 (28)	0 (0)	1 (3)
increased pain w/cough	17 (59)	2 (7)	7 (24)	3 (10)
sign				
weakness by examination †	24 (83)	4 (14)	1 (3)	0 (0)
atrophy	26 (90)	2 (7)	1 (3)	0 (0)
spasticity	16 (55)	10 (34)	2 (7)	1 (3)
ataxia	23 (79)	4 (14)	2 (7)	0 (0)
sensory loss by examination	8 (28)	4 (14)	16 (55)	1 (3)

* Ambulation scale: unaffected (independent); mild (with cane); moderate (with walker); severe (nonambulatory).

† Weakness scale: unaffected (5/5); mild (4/5); moderate (3/5, antigravity strength); severe (1–2/5, < antigravity).

TABLE 2

Surgical outcome in patients undergoing surgical decompression for CM-I and syringomyelia

Surgical Outcome	Follow-Up Visit		
	3–6 Mos	1 Yr	2 Yrs
clinical result			
improved	27/28 (96) *	21/22 (95) *	17/18 (94) *
no change	1 (4)	1 (5)	1 (6)
worsened	0	0	0
neuroimaging result			
syrinx resolved [†]	25/29 (86)	21/23 (91)	19/19 (100)

* Because 1 patient was asymptomatic before and after surgery, the group with clinical results included 1 patient fewer than the group with radiological findings.

[†] Greater than 50% decrease in maximal anteroposterior diameter.