Published in final edited form as:

J Neurol. 2006 June ; 253(6): 707–713. doi:10.1007/s00415-006-0091-5.

The post-syrinx syndrome: stable central myelopathy and collapsed or absent syrinx

E. I. Bogdanov, MD,

Dept. of Neurology and Rehabilitation, Kazan State Medical University, Butlerov str. 49, Kazan, Russia 420012, Tel.: +7-8432/353308, Fax: +7-8432/360693

John D. Heiss, MD, and

Surgical Neurology Branch, National Institute of Neurological Disease and Stroke, National Institutes of Health, Bethesda, MD 20892-1414, USA, Tel.: +1-301/594-8112, Fax: +1-301/402-0380

E. G. Mendelevich, MD

Dept. of Neurology and Rehabilitation, Kazan State Medical University, Butlerov str. 49, Kazan, Russia 420012, Tel.: +7-8432/353308, Fax: +7-8432/360693

E. I. Bogdanov: enver_bogdanov@mail.ru; John D. Heiss: heissj@ninds.nih.gov

Abstract

Among 168 cases with neurologic findings of cervicothoracic syringomyelia and MRI findings of Chiari 1 malformation and/or underdevelopment of the posterior cranial fossa, 15 patients (9.1%) had collapsed, flat syrinxes and 14 patients (8.3%) did not have syrinxes. Both groups of patients had clinical findings of central myelopathy that had been stable for at least 3 years. Magnetic resonance imaging detected atrophy of the cervical spinal cord in both groups and spontaneous communications between the syrinx and the subarachnoid space in 3 patients of the group with collapsed syrinxes. Analysis of these results and review of the literature suggest that patients with clinical signs of syringomyelia and Chiari 1 malformation or underdeveloped posterior fossa, but with small or absent syringomyelitic cavities, have the “postsyrinx” state as a result of spontaneous collapse of distended syrinxes.

Keywords

syringomyelia; Arnold-Chiari malformation; skull base; spinal cord diseases

Introduction

Syringomyelia associated with CM1 presents clinically as a slowly progressive central cervical myelopathy distinct from other myelopathies and polyneuropathies. MRI of the spine confirms the diagnosis of syringomyelia. However, MRI does not confirm the presence of a syrinx or of other syringomyelia-like diseases in some patients with CM1, despite neurologic findings that suggest syringomyelia. For example, Elster et al.[11] did not

find syringes in 2 of 25 patients with CM1 and central myelopathy, Tanghe [53] in 4 of 19, and Milhorat et al. [37] in 83 of 126. The segmental signs and symptoms of central cord syndrome in these patients were unlike the signs and symptoms produced by long tract lesions at the cranio-vertebral junction (CVJ) level. In addition, the central cord syndrome was not a product of the “presyrinx” state, because swelling and edema of the spinal cord were not present on MRI studies [13, 28]. To elucidate the causes and mechanisms of central cord syndrome in patients without syringes, we compared clinical and MRI evaluations of patients with CM1 without syringomyelia to those of patients with CM1 with collapsed syringes.

Patients and methods

From 1997 to 2001, 168 patients were examined in our hospital with clinical signs and symptoms of syringomyelia associated with CM1 and/or underdevelopment of the PF; other diseases with similar manifestations were excluded. Specifically, at examination we excluded diseases that mimic syringomyelia: 1) adult form of occult spinal dysraphism and tethered cord syndrome, 2) cervical spinal canal stenosis, 3) spondylosis and cervical disc herniations, 4) post-traumatic central cord syndrome, 5) Tangier disease and 6) other myelo- and polyneuropathies. All examined patients were rural tenants of the Republic of Tatarstan in the Russian Federation where syringomyelia is very common, 130:100,000 inhabitants [8, 49]. Syringomyelia predominates in males and in those from isolated areas [8, 18, 47, 49].

Patients underwent one or more MRI scans of the brain and the cervical and thoracic spinal cord. Imaging was performed on the 1.0-T General Electric Signa Horizon or the 0.28-T Bruker with SGI workstation. Standard T1 and T2 weighted images were taken in sagittal, coronal, and axial planes. The ‘hydrography’ technique of long-echo RARE sequence was also used to show syrinx cavities in three planes. The widest anteroposterior diameter of the spinal cord at the C4 and C7 cervical levels was measured [48]. Measurements of CSF spaces, bony structures, and angles at CVJ and PF levels were performed in accordance with previous recommendations [16,37,40] and are listed in the Table.

Fourteen patients (8.3%) with CM1 did not have syringes on their MRI scans and formed Group A. Group A consisted of 2 females and 12 males, age 19 to 61 (mean 44.9) years, who had symptomatic clinical syringomyelia associated with CM1 or underdeveloped PF for 8 to 35 years (mean 21.8) (Table, Fig. B, C, E, G). Fifteen patients (8.9%) with CM1 had flat, spontaneously-collapsed syringomyelitic cavities and formed Group B. Group B was comprised of 5 females and 10 males, age 42 to 75 (mean 52.2) years, with symptomatic syringomyelia for 4 to 47 (mean 28.7) years, who had clear MRI signs of flat, spontaneously-collapsed syringomyelitic cavities [5] (Fig. F, H–K). Syringes had cervical or cervico-thoracic localization, were 3–10 spinal segments in length, but did not communicate with the fourth ventricle. Three patients of Group B had spontaneous syringo-subarachnoidal communications visible on MRI images in the axial plane [4]. Patients in both groups had normal or slightly limited functional activity and had no history of disease progression for at least the previous three years. Spinal cord atrophy was defined as an AP diameter of the spinal cord of less than 8 mm (Table, Fig. G–K) [48]. Fisher exact test and

non-parametric Mann-Whitney *U* test were used for statistical analysis; *P* values < 0.05 were considered significant.

Results

Results are presented in the Table and Figure. Patients in groups A and B did not differ significantly in terms of mean age and duration of the symptomatic stage of disease. Males predominated in both groups. Patients in both groups had clinical findings typical for syringomyelia associated with CM1 and/or underdeveloped PF, with neurological deficits generally being more severe in patients of group B (Table). Segmental and long tract lesions at the cervical spinal cord level, trophic disturbances, and kyphoscoliosis were frequent clinical findings. MRI features were those of CM1 and/or under-development of the PF (Table). Dysesthetic pain, pyramidal signs, increased Boogard's angle, and narrow anterior and posterior CSF spaces at the FM level were more frequent in patients of group B (Table). The mean anteroposterior diameter of the spinal cord did not differ significantly between groups. 4 patients of group A [1,7,10,13] and 6 patients of group B [2-5,7,13] had cervical spinal cord atrophy (Table, Fig. F-K) [48]. A common factor in both groups was a narrow or obliterated cerebellomedullary cistern. MRI scans separated by an 8 year period in Patient A-13 indicated that in this interval a large diameter syrinx (Fig. D) resolved (Fig. C, E) and the cerebellar ectopia (Fig. A) lessened (Fig. B).

Discussion

Syringomyelia is a polyetiologic disorder characterized by intramedullary tubular cyst formation and progression. One current classification distinguishes four types of syringomyelia [38]. In most cases, syringomyelia is a secondary event affecting the spinal cord due to a distant cause [10]. The most frequent cause of syringomyelia development is the Chiari 1 malformation (CM1) [14]. Primary CM1 is created when an underdeveloped posterior cranial fossa (PF) with decreased volume cannot accommodate the normally developed hindbrain [3, 37, 40]. It is characterized by compression of cerebellar cisterns, herniation of the cerebellar tonsils, decreased PF volume, and varying degrees of dysplasia of the posterior aspect of the skull base [37]. Because patients with a small posterior fossa, but without herniation of the cerebellar tonsils, can present with syringomyelia identical to that seen in patients with CM1, it has been suggested that this state be denoted as the Chiari 0 malformation (CM0) [55]. Other skull base abnormalities (basilar invagination, platybasia, and short clivus) are also associated with a small posterior fossa [24, 50]. Familial aggregation of CM1 and CM0 cases suggests that there is a genetic predisposition for developing the malformation [37, 50]. It is hypothesized that a gene(s) mutation prevents normal growth of the bones of the skull base and reduces the volume of the posterior fossa enough to compress the cerebellum and displace the cerebellar tonsils through the foramen magnum [50]. The likelihood of syringomyelia is related to the severity of the morphological abnormalities expressed in patients with CM1 or CM0. External factors also contribute to the development of syringomyelia. For example, syringomyelia is more prevalent in those with occupations that require strenuous physical exertion [8,18,47,49]. In fact, as the number of workers engaged in occupations that required heavy labor decreased

in Western European countries between 1940 and 1980, the incidence of syringomyelia decreased proportionally [18, 47].

Although the pathogenesis of CM1-associated syringomyelia is not completely understood, abnormal CSF flow at the foramen magnum (FM) and upper cervical spinal canal appears to influence the creation and progression of syrinxes [9, 16, 20, 43, 45]. Several pathophysiological factors are proposed to promote syringomyelia development: 1) reduction of subarachnoid compliance and accentuated pulse pressure in the cervical portion of the spinal subarachnoid space [16, 43], 2) increased intrasyrinx pressure [36], 3) pressure gradients within the intramedullary extracellular fluid [23], 4) intrasyrinx fluid pulsation [9], 5) pulsation of the intramedullary arteries [51], and 6) patency of the central canal of the spinal cord [33]. However, because communication between the syrinx and fourth ventricle is usually not found in radiographic or post-mortem studies, it cannot play a role in pathogenesis of most cases of syringomyelia [34, 43, 45].

According to Oldfield et al. [43], the movement of the herniated cerebellar tonsils acts as a piston to create enlarged systolic CSF pressure waves that act on the surface of the spinal cord and propel the passage of CSF into the spinal cord through the perivascular spaces. The same mechanism of syrinx formation and development is proposed for syringomyelia associated with CM0 [55]. Before the syrinx is visible on MR images, longitudinal intramedullary edema and swelling associated with clinical manifestations of syringomyelia denote the “presyrinx” state in CM1 patients [13, 28]. In general, neurologic deficits progress rapidly in the initial period of spinal cord “presyrinx” edema and syrinx formation. We previously compared the rate of disease progression in 21 patients with distended syrinxes (mean diameter 8.8 mm) with that in 50 patients with smaller syrinxes (mean diameter 4.7 mm). Disease progression was more rapid in patients with larger syrinxes [5]. However, neurologic function stabilizes later in about one-third to one-half of patients with syringomyelia for 10 or more years [1, 6, 7, 17, 29] and shrinkage of cavities may be encountered [5]. Despite these general rules, the prognosis for progression of myelopathy is uncertain in individual patients. The dimensions of the syrinx and remaining spinal cord are not strongly related to clinical findings [30] and some patients with large, distended cavities are asymptomatic for many years [42]. However, once myelopathy develops it is largely irreversible and it improves modestly, if at all, even if the syrinx is eliminated [25, 43, 54]. Factors other than syrinx diameter, such as intra- and extrasyrinx fluid pressures [16, 36] and gliosis around the cavity, may play a role in the development of myelopathy.

There are several reports of cases in which syringomyelia symptoms resolve spontaneously or fail to progress [1, 6, 7, 17, 29]. In a nationwide epidemiological survey in Japan [39], among 1,243 patients with syringomyelia, one-half of them with CM1, 39 patients (2.3%) had spontaneous resolution of symptoms and 202 patients (16.2%) had a stable course of disease. Our retrospective analysis of 103 unoperated cases of syringomyelia associated with CM1 and/or underdeveloped PF, with duration of disease from 1 to 46 years, revealed that syrinx diameter decreases at a late stage in the disease [5]. In the literature there are descriptions of 39 single cases [27] in which spontaneous radiological resolution of syringomyelia occurred in children and adults, with or without clinical restoration [2, 25, 26, 44, 46, 52, 54]. In some of these cases the collapse of the syrinx was associated with

spontaneous upward displacement of the cerebellar tonsils [2, 25, 26, 44, 46, 52]. With time, the syrinx may become smaller [2,54] or disappear completely [22, 26, 54], and may be accompanied by spinal cord atrophy, as in the case of an 84-year-old patient with CM1 who was left with spinal cord atrophy and a narrow syrinx cavity after spontaneous resolution of holocord syringomyelia [12].

Spontaneous resolution of the syrinx has also been seen in the pediatric age group. Clinical and MRI examinations were performed serially over many years in 27 children and adolescents with scoliosis and syringomyelia; the syrinx resolved spontaneously and completely in 9 and was reduced in size in another 5 of them [54]. The authors attributed this phenomenon to growth of the PF bones and normalization of CSF dynamics at the FM level [54]. That report supports earlier work that showed that with increasing age the volume of the PF volume increases [3] and the cerebellar tonsils ascend [32], as occurred in Patient A-13 (Fig. A, B). Spontaneous syrinx collapse can also result from spontaneous rupture of an arachnoid membrane at the foramen magnum, which opens the CSF pathways, or from spontaneous syringo-subarachnoid drainage [4]. The neurologic improvement or stabilization that follows spontaneous collapse of the syrinx is similar to that seen after surgical procedures that collapse the syrinx [19, 21].

The reports in the literature of children and adults who have had complete resolution of their syringomyelic cavities support our contention that patients in group A had syringomyelia in the past. The time course of the disease (Table) and the radiographic findings (Fig. B, C, E, F, G) in group A also suggests that this is the case. In patients with scoliosis and kyphosis, syringomyelia probably arose in childhood (for example, patients A-7, A-9, A-13 in the Table). In patients in other series who undergo surgical treatment, the prognosis for improvement decreases as the duration of syringomyelia before surgery increases [41]. Irreversible injury to the spinal cord from chronically increased intra- and extramedullary pressure [16, 36] and from gliosis around the syrinx cavities [34] prevents clinical restoration when the syrinx collapses spontaneously or from surgery. It is unknown if gliosis can progress after resolution of the syrinx filling mechanism and collapse of the syrinx cavity.

Clinical findings in patients without syrinxes (Group A) were identical to those in patients with spontaneously collapsed cavities (Group B, Table) and were typical for syringomyelia associated with CM1 and/or underdeveloped PF. The persistence of dysesthetic pain (especially in group B) was characteristic for patients with collapse of their syrinxes [35]. Patients of both groups had similar bony and CSF space abnormalities at the CVJ and PF (Table) [16, 43, 45]. Syrinxes in some patients of group B were very narrow (Fig. F, H–K) and complete obliteration of these cavities might be observed in future MR scans. Some patients in both groups had prominent atrophy of the cervical spinal cord (Table, Fig. F–K), which resulted from loss of spinal cord substance before the syrinx cavity collapsed.

CSF flow disturbances at the CVJ and PF that promote the development of syringomyelia can spontaneously revert to a normal state that resolves the syrinx, such as in Case A-13, in which a syrinx that was seen 8 years before by MRI resolved spontaneously and completely on follow-up examination (Fig. A–E). Incomplete disappearance of the syrinx in patients of

group B may have resulted because the ventral and dorsal CSF spaces at the FM level were smaller in this group than in group A (Table).

On magnetic resonance imaging, an eccentric part of a syrinx may appear to approach or communicate with the spinal subarachnoid space [4] (Fig. L, M). Postmortem examination of the spinal cord of patients with syringomyelia associated with CM1 often demonstrates fissures in the spinal cord that connect the subarachnoid space with syrinx cavities that extend into the dorsolateral part of the spinal cord [34]. Syrinx fluid can drain into the subarachnoid space through these channels.

The symptoms of clinical syringomyelia in patients of group A are best explained by residual central myelopathy that persists after spontaneous resolution of syringomyelia. Other mechanisms were considered that did not require a syrinx as an intermediate condition for central cervical myelopathy. Specifically, cervical spinal stenosis was excluded. In addition, the “presyrinx” state, which may result in signs and symptoms that mimic syringomyelia, was not present because there were no MRI features of intramedullary edema and swelling, the disease was of long duration, and symptoms were stable for many years [13, 28]. Non-cystic myelopathy did not result from the chronic effect of pulsatile traction on the spinal cord [20] because this mechanism is applicable primarily to type II CM, tethered cord, and spina bifida [31]. In addition, focal traction on the spinal cord is unlikely to produce stable central spinal cord syndrome with widespread segmental signs. A final potential mechanism, that the myelopathy arose from basilar impression alone (Table), is not in keeping with previous observations that basilar impression 1) is not manifested by segmental lesions, 2) has distinctive clinical characteristics [15], and 3) does not cause syringomyelia unless it is associated with CM1. These mechanisms can certainly be excluded in patient A-13.

Our present and previous [4, 5] studies and review of the literature lead us to conclude that clinical signs and symptoms of syringomyelia in patients with visualized CM1 and/or underdeveloped PF, but without visible syringomyelitic cavities, result from syringes that have resolved spontaneously after spontaneous normalization of CSF dynamics at the CVJ level or drainage of the syrinx through syringosubarachnoid communications. This state can be denoted as the “post-syrinx” state and is one possible course in the natural history of syringomyelia associated with CM1 and/or underdeveloped PF. The post-syrinx state is characterized by 1) stable symptoms and signs of central myelopathy, 2) MRI evidence of Chiari I or Chiari 0 malformation and absent or collapsed syrinx, and 3) absence of other diseases that mimic syringomyelia. We hope that elaboration of this syndrome will be helpful in the clinical management of these patients.

Acknowledgments

We thank Dr. Pauline Monro for reviewing the manuscript carefully and medical student Shamil Bogdanov for technical assistance.

References

1. Anderson NE, Willoughby EW, Wrightson P. The natural history of syringomyelia. *Clin Exp Neurol.* 1986; 22:71–80. [PubMed: 3581513]

2. Avellino AM, Britz GW, McDowell JR, Shaw DW, Ellenbogen RG, Roberts TS. Spontaneous resolution of a cervicothoracic syrinx in a child. Case report and review of the literature. *Pediatr Neurosurg*. 1999; 30:43–46. [PubMed: 10202308]
3. 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]
4. Bogdanov EI, Ibatullin MM, Mendelevich EG. Spontaneous drainage in syringomyelia: magnetic resonance imaging findings. *Neuroradiology*. 2000; 42:676–678. [PubMed: 11071442]
5. Bogdanov EI, Mendelevich EG. Syrinx size and duration of symptoms predict the pace of progressive myelopathy: retrospective analysis of 103 unoperated cases with craniocervical junction malformations and syringomyelia. *Clin Neurol Neurosurg*. 2002; 104:90–97. [PubMed: 11932037]
6. Boiardi A, Munari L, Silvani A, Porta E, Scuratti A, Lodrini S. Natural history and postsurgical outcome of syringomyelia. *Ital J Neurol Sci*. 1991; 12:575–579. [PubMed: 1783536]
7. Boman K, Iivanainen M. Prognosis of syringomyelia. *Acta Neurol Scand*. 1967; 43:61–68. [PubMed: 6040325]
8. Borisova, NA.; Valikova, IV.; Kutchaeva, GA. Syringomyelia (in Russian). *Meditcina*; Moscow: 1989. p. 5-150.
9. Brugieres P, Idy-Peretti I, Iffenecker C, Parker F, Jolivet O, Hurth M, Gaston A, Bittoun J. CSF flow measurement in syringomyelia. *AJNR Am J Neuroradiol*. 2000; 21:1785–1792. [PubMed: 11110528]
10. Catala M. Neuroembryologic considerations on the so-called malformative syringomyelia. *Neurochirurgie*. 1999; 45(Suppl 1):9–22. [PubMed: 10420401]
11. Elster AD, Chen MY. Chiari I malformations: clinical and radiologic reappraisal. *Radiology*. 1992; 183:347–353. [PubMed: 1561334]
12. Finsterer J, Lubec D, Samec P. Generalised spinal cord atrophy, Chiari-I malformation, and syringomyelia. *Spinal Cord*. 2001; 39:184–188. [PubMed: 11326330]
13. Fischbein NJ, Dillon WP, Cobbs C, Weinstein PR. The “presyrinx” state: a reversible myelopathic condition that may precede syringomyelia. *AJNR Am J Neuroradiol*. 1999; 20:7–20. [PubMed: 9974051]
14. Gardner WJ, Angel J. The mechanism of syringomyelia and its surgical correction. *Clin Neurosurg*. 1959; 6:131–140. [PubMed: 13826542]
15. Goel A, Bhatjiwale M, Desai K. Basilar invagination: a study based on 190 surgically treated patients. *J Neurosurg*. 1998; 88:962–968. [PubMed: 9609289]
16. Heiss JD, Patronas N, DeVroom HL, Shawker T, Ennis R, Kammerer W, Eidsath A, Talbot T, Morris J, Eskioglu E, Oldfield EH. Elucidating the pathophysiology of syringomyelia. *J Neurosurg*. 1999; 91:553–562. [PubMed: 10507374]
17. Hertel G, Kramer S, Placzek E. Syringomyelia, Clinical follow-up studies in 323 patients. *Nervenarzt*. 1973; 44:1–13. [PubMed: 4687739]
18. Hertel, G.; Ricker, K. A geomedical study on the distribution of syringomyelia in Germany. In: den Hartog Jager, WA., editor. *Neurology*. Vol. 434. Excerpta Medica; Amsterdam: 1978. p. 353-365.
19. Hida K, Iwasaki Y. Syringosub-arachoid shunt for syringomyelia associated with Chiari I malformation. *Neurosurg Focus*. 2001; 11:Article 7.
20. Hofmann E, Warmuth-Metz M, Bendszus M, Solymosi L. Phase-contrast MR imaging of the cervical CSF and spinal cord: volumetric motion analysis in patients with Chiari I malformation. *AJNR Am J Neuroradiol*. 2000; 21:151–158. [PubMed: 10669242]
21. Iskandar BJ, Hedlund GL, Grabb PA, Oakes WJ. The resolution of syringohydromyelia without hindbrain herniation after posterior fossa decompression. *J Neurosurg*. 1998; 89:212–216. [PubMed: 9688115]
22. Jack CR Jr, Kokmen E, Onofrio BM. Spontaneous decompression of syringomyelia: magnetic resonance imaging findings. Case report. *J Neurosurg*. 1991; 74:283–286. [PubMed: 1988600]
23. Josephson A, Greitz D, Klason T, Olson L, Spenger C. A spinal thecal sac constriction model supports the theory that induced pressure gradients in the cord cause edema and cyst formation. *Neurosurgery*. 2001; 48:636–645. discussion 645–646. [PubMed: 11270555]

24. Karagoz F, Izgi N, Kapijicijoglu Sencer S. Morphometric measurements of the cranium in patients with Chiari type I malformation and comparison with the normal population. *Acta Neurochir (Wien)*. 2002; 144:165–171. discussion 171. [PubMed: 11862517]
25. Kastrup A, Nagele T, Topka H. Spontaneous resolution of idiopathic syringomyelia. *Neurology*. 2001; 57:1519–1520. [PubMed: 11673607]
26. Klekamp J, Iaconetta G, Samii M. Spontaneous resolution of Chiari I malformation and syringomyelia: case report and review of the literature. *Neurosurgery*. 2001; 48:664–667. [PubMed: 11270558]
27. Kyoshima K, Bogdanov EI. Spontaneous resolution of syringomyelia: report of two cases and review of the literature. *Neurosurgery*. 2003; 53:762–768. discussion 768–769. [PubMed: 12943593]
28. Levy EI, Heiss JD, Kent MS, Riedel CJ, Oldfield EH. Spinal cord swelling preceding syrinx development. Case report. *J Neurosurg Spine*. 2000; 92:93–97.
29. Mariani C, Cislighi MG, Barbieri S, Filizzolo F, Di Palma F, Farina E, D'Aliberti G, Scarlato G. The natural history and results of surgery in 50 cases of syringomyelia. *J Neurol*. 1991; 238:433–438. [PubMed: 1779249]
30. Masur H, Oberwittler C, Reuther G, Heyen P. Cerebellar herniation in syringomyelia: relation between tonsillar herniation and the dimensions of the syrinx and the remaining spinal cord. A quantitative MRI study. *Eur Neurol*. 1995; 35:162–167. [PubMed: 7628496]
31. Meadows J, Guarnieri M, Haroun R, Kraut M, Carson B. Type I Chiari malformation: a review of the literature. *Neurosurg Quart*. 2001; 11:220–229.
32. Mikulis DJ, Diaz O, Egglin TK, Sanchez R. Variance of the position of the cerebellar tonsils with age: preliminary report. *Radiology*. 1992; 183:725–728. [PubMed: 1584927]
33. Milhorat TH, Nobandegani F, Miller JJ, Rao C. Noncommunicating syringomyelia following occlusion of central canal in rats. Experimental model and histological findings. *J Neurosurg*. 1993; 78:274–279. [PubMed: 8421210]
34. 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]
35. Milhorat TH, Kotzen RM, Mu HT, Capocelli AL Jr, Milhorat RH. Dysesthetic pain in patients with syringomyelia. *Neurosurgery*. 1996; 38:940–946. discussion 946–947. [PubMed: 8727819]
36. Milhorat TH, Capocelli AL Jr, Kotzen RM, Bolognese P, Heger IM, Cottrell JE. Intramedullary pressure in syringomyelia: clinical and pathophysiological correlates of syrinx distension. *Neurosurgery*. 1997; 41:1102–1110. [PubMed: 9361064]
37. Milhorat TH, Chou MW, Trinidad EM, Kula RW, Mandell M, Wolpert C, Speer MC. Chiari I malformation redefined: clinical and radiographic findings for 364 symptomatic patients. *Neurosurgery*. 1999; 44:1005–1017. [PubMed: 10232534]
38. Milhorat TH. Classification of syringomyelia. *Neurosurg Focus*. 2000; 8:Article 1.
39. Moriwaka F, Tashiro K, Tachibana S, Yada K. Epidemiology of syringomyelia in Japan—the nationwide survey. *Rinsho Shinkeigaku*. 1995; 35:1395–1397. [PubMed: 8752408]
40. 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]
41. Nishikawa M, Sakamoto H, Kitano S, Hakuba A, Nakanishi N. A clinical study on long-term outcome in patients with syringomyelia associated with Chiari malformation. *No To Shinkei*. 1999; 51:318–323. [PubMed: 10363266]
42. 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; 49:637–640. discussion 640–641. [PubMed: 11523674]
43. 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]

44. Pierallini A, Ferone E, Colonnese C. Magnetic resonance imaging of a case of spontaneous resolution of syringomyelia associated with type I Chiari malformation. *Radiol Med (Torino)*. 1997; 93:621–622. [PubMed: 9280949]
45. Pinna G, Alessandrini F, Alfieri A, Rossi M, Bricolo A. Cerebrospinal fluid flow dynamics study in Chiari I malformation: implication for syrinx formation. *Neurosurg Focus*. 2000; 8:Article 3.
46. Santoro A, Delfini R, Innocenzi G, Di Biasi C, Transimeni G, Gualdi G. Spontaneous drainage of syringomyelia. Report of two cases. *J Neurosurg*. 1993; 79:132–134. [PubMed: 8315453]
47. Schergna E, Armani M. Catamnestic findings in syringomyelia in the province of Padua. *Minerva Med*. 1985; 76:1699–1704. [PubMed: 4047455]
48. Sherman JL, Nassaux PY, Citrin CM. Measurements of the normal cervical spinal cord on MR imaging. *AJNR Am J Neuroradiol*. 1990; 11:369–372. [PubMed: 2107721]
49. Sirotkin VM. Regional peculiarities of syringomyelia (in Russian). *Genetica*. 1970; 6:166–172.
50. Speer MC, George TM, Enterline DS, Franklin A, Wolpert CM, Milhorat TH. A genetic hypothesis for Chiari I malformation with or without syringomyelia. *Neurosurg Focus*. 2000; 8:Article 12.
51. Stoodley MA, Jones NR, Yang L, Brown CJ. Mechanisms underlying the formation and enlargement of non-communicating syringomyelia: experimental studies. *Neurosurg Focus*. 2000; 8:Article 2.
52. Sudo K, Tashiro K, Miyasaka K. Features of spontaneous improvement in syringomyelia with low-situated cerebellar tonsils. *Acta Neurol Belg*. 1998; 98:342–346. [PubMed: 9922822]
53. Tanghe HL. Magnetic resonance imaging (MRI) in syringomyelia. *Acta Neurochir (Wien)*. 1995; 134:93–99. [PubMed: 7668137]
54. Tokunaga M, Minami S, Isobe K, Moriya H, Kitahara H, Nakata Y. Natural history of scoliosis in children with syringomyelia. *J Bone Joint Surg Br*. 2001; 83:371–376. [PubMed: 11341422]
55. Tubbs RS, Elton S, Grabb P, Dockery SE, Bartolucci AA, Oakes WJ. Analysis of the posterior fossa in children with the Chiari 0 malformation. *Neurosurgery*. 2001; 48:1050–1054. discussion 1054–1055. [PubMed: 11334271]

Fig.

In patients with clinical signs of syringomyelia, midsagittal and axial MR images of the cervical spinal cord demonstrated some patients without radiographic evidence of syringomyelia (**B, C, E, F, G**), (Group A) and some with small, collapsed syrinxes (**H–N**), (Group B). Patient A-7 (**F, G**) has spinal cord atrophy and a small central canal. Patient A-13 had a large syrinx (**A, D**) and tonsillar ectopia (**D**), which extended below the C1 level on an initial scan; 8 years later the syrinx had resolved (**B, C, E**) and the cerebellar tonsils (**B, E**) had ascended. Patients B-2 (**H–K**) and B-5 (**N**) have spinal cord atrophy and collapsed syrinxes

Clinical and MRI findings in patients with CM1 and clinical signs and symptoms of cervico-thoracic syringomyelia. On recent MRI studies, Group A patients do not have syrinxes and Group B patients have narrow syrinx cavities

Group Patient number	A															B															
	1	2	3	4	5	6	7	8	9	10	11	12	13	14	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15		
Age	55	48	27	61	37	43	42	52	45	51	45	57	19	47	50	64	64	75	42	47	41	47	45	45	48	63	46	46	60		
Age at symptom onset	47	28	20	48	30	22	12	32	12	29	10	20	11	17	21	24	44	30	12	31	37	17	11	17	21	16	14	18	39		
Segmental signs:																															
dysesthetic pain	+							+	+							+				+	+	+		+	+	+	+	+	+	p < 0.017	
sensory loss	+	+		+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	
trophic disturbance	+		+	+						+						+		+					+		+	+	+	+	+	+	
muscular atrophy			+						+	+		+			+	+		+	+		+		+							+	
weakness	+	+		+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	
Long tracts signs																															
pyramidal signs					+	+	+				+	+	+	+	+	+	+		+	+	+	+	+	+	+	+	+	+	+	p < 0.036	
impaired position sense				+								+			+	+	+	+	+				+							+	
FM level signs																															
suboccipital headache																+									+						
nystagmus						+	+								+						+	+			+	+		+	+		
ataxia							+	+		+								+	+		+	+			+	+					
dizziness/vertigo						+	+										+					+	+		+	+					
impaired CN IX, X	+			+			+										+			+	+			+							
Scoliosis/kyphosis							+		+		+		+	+	+	+		+	+	+	+	+	+	+	+	+	+	+	+	+	
MRI features																															
cerebellar cisterns																															
obliterated	+		+		+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	
small		+		+	+										+													+			
tonsillar herniation (mm)																												5			
basilar invagination		+	+	+	+		+	+		+		+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	+	
Cervicomedullary kinking		+																	+		+	+		+	+	+	+	+	+		
short supraocciput	+		+				+			+	+					+			+	+	+	+	+	+	+	+	+	+	+	+	

Group Patient number	A														B														
	1	2	3	4	5	6	7	8	9	10	11	12	13	14	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15
short clivus			+				+								+				+	+	+	+		+				+	
Boogard's angle increased				+								+	+		+	+	+	+	+		+	+	+	+	+	+	+	+	
platybasia	+			+	+						+	+	+						+	+						+	+	+	+
Obliterated CSF space																													
Ventral Foramen magnum									+			+			+	+	+	+	+	+	+	+	+	+	+	+	+	+	
Dorsal Foramen magnum	+		+			+	+	+					+		+	+	+	+	+	+	+	+	+	+	+	+	+	+	
Spinal cord AP diameter (mm)	6.5	11	12	10	11	9	6	8.5	10	6	11	8.5	7	10	8.5	6	6	7.5	7	9.5	6.5	8	10	9.5	9	8	7.5	9.5	15
Syrinx AP diameter (mm)															1	1	3	3	3	3	1	2	4	1	3	2	4	1	3
Spontaneous syrx drainage															+					+	+	+							