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ORIGINAL ARTICLE

Factors contributing to improvement of syringomyelia after foramen magnum decompression for Chiari type I malformation

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Abstract
Background Although various surgical approaches have been proposed for treating syringomyelia associated with Chiari type I malformation, a standard method has yet to be established. we prospectively investigated the results of our surgical method: foramen magnum decompression combined with C1 laminectomy and excision of the outer layer of the dura mater.
Methods Twenty patients underwent surgery between 2000 and 2010 at our hospital. After surgery, the size of the syrinx decreased in 11 patients (decreased group) but remained unchanged in nine patients (unchanged group). The following parameters were compared: age at the time of surgery, duration of morbidity, improvement of preoperative symptoms, morphological type and length of the syrinx, presence or absence of scoliosis, cervical alignment, basal and clivo-axial angles, and postoperative subarachnoid space at the foramen magnum level.
Results Preoperative symptoms improved in all patients in the decreased group but in only one patient in the unchanged group. The average duration of morbidity was significantly shorter in the decreased group. Morphological examination revealed that the size of all central-type syrinxes decreased after surgery, whereas in all cases of deviated-type syrinx, size was unchanged. The average length of preoperative syrinx was significantly shorter in the decreased group. The postoperative subarachnoid space at the foramen magnum was enlarged in the entire decreased group, whereas residual narrowing of the space was observed in 44 % of patients in the unchanged group. No significant intergroup differences were observed in the other factors.
Conclusions In patients with syringomyelia, a longer and deviated type of syrinx, a longer duration of morbidity, and postoperative residual narrowing of the subarachnoid space are associated with a poor prognosis after the surgical procedure. The pathogenesis of syringomyelia is inconsistent, and the choice of surgical technique for each pathological condition is important.

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Introduction

Chiari type I malformation, characterized by downward herniation of the caudal cerebellum or medulla oblongata into the foramen magnum, is one of the major causes of syringomyelia [1, 2]. Although various surgical procedures to restore cerebrospinal fluid (CSF) circulation and decompress the neuraxis have been described, there is no consensus regarding the optimal surgical procedure. Some authors prefer foramen magnum decompression (FMD) with dural opening [3, 4], whereas others believe that duraplasty [5–8], sometimes with tonsillectomy [9–12], is required to expand the posterior fossa and restore normal CSF flow. Syringosubarachnoid shunt is also used either alone [13, 14] or combined with FMD [11, 15].

We prospectively investigated the results of our surgical method, FMD combined with C1 laminectomy and excision of the outer layer of the dura mater, which was advocated by Isu et al. [16] and is recognized as a minimally invasive surgical

technique for the spinal cord. The goals of this surgical procedure are to correct the circulatory disturbance of CSF at the foramen magnum and create a new cisterna magna, considering the dural inner part capability of expansion. So far, there are few reports applying this surgical procedure as a single method. Gambardella et al. [17] performed the surgery for eight patients with syringomyelia and focused on surgical techniques. The other published reports were from the same institution and focused on the scoliosis, cervical spinal motion, and neuropathic pain [18–20], respectively. The objectives of our study were to assess surgery-related results and identify factors influencing improvement by comparing symptoms, clinical results, and imaging findings.

Materials and methods

Twenty patients underwent surgery at our hospital between 2000 and 2010 for syringomyelia associated with Chiari type I malformation. There were two male and 18 female patients, ranging in age from 11 to 57 years (mean 31.0 years) at the time of surgery. Patients with syringomyelia associated with other diseases, such as spinal cord tumors or adhesive arachnoiditis, or those with spinal cord injuries, were excluded from this study. All patients underwent FMD combined with C1 laminectomy and excision of the outer layer of the dura mater. The postoperative period ranged from 1.3 to 10.8 years (mean 6.0 years). After surgery, patients in whom the size of the syrinx decreased on both sagittal and axial magnetic resonance imaging (MRI) were defined as the decreased group, and those in whom the size was relatively unchanged were defined as the unchanged group (Fig. 1). The following parameters were compared between groups: age at the time of surgery, duration of morbidity (from symptom onset to initial surgery), improvement of preoperative symptoms, type and length of the syrinx, subarachnoid space at the foramen magnum level in postoperative MRI (Fig. 2), basal and clivo-axial angles, cervical alignment from C2 to C7, and presence or absence of scoliosis in preoperative radiographs. Scoliosis was defined as a Cobb angle $\geq 10°$.

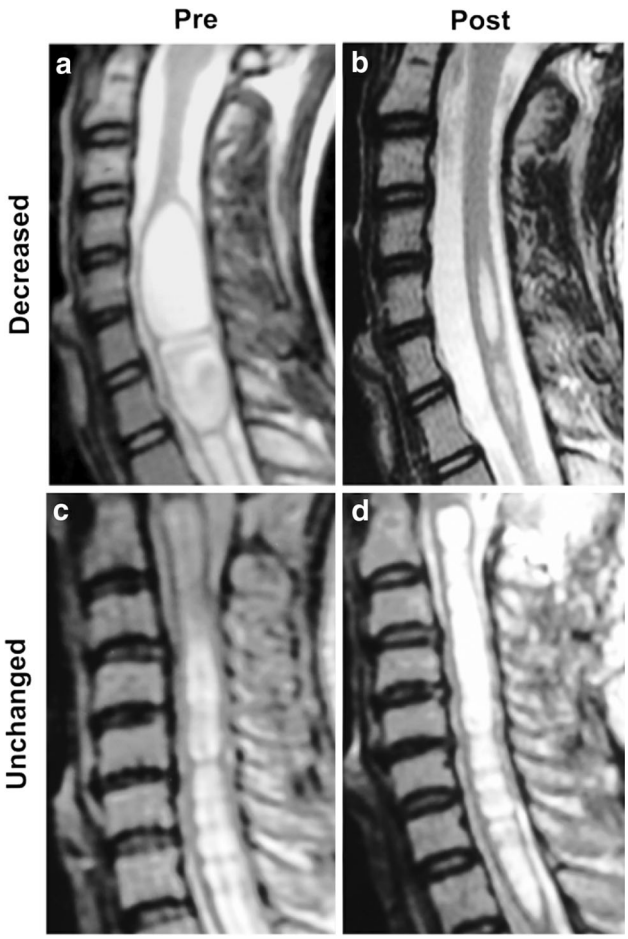

Fig. 1 Representative pre- and postoperative T2-weighted magnetic resonance (MR) images from patients in the decreased and unchanged group. Note the markedly decreased size of the syrinx in **a**, **b** in contrast to the unchanged size in **c**, **d**

Clinical results were evaluated on the basis of the outcome scale proposed by Bidzinski [21] (Table 1).

Syrinx morphology was classified into three types—central, enlarged, and deviated—based on axial MRI at the level corresponding to the dermatomal distribution of sensory disturbance and at the level where syrinx size was the greatest in asymptomatic scoliotic patients (Fig. 3) [22].

Statistical analysis

The Mann–Whitney *U* test was used to compare data between groups. Chi-square test for independence was used to evaluate clinical results, syrinx type, presence or absence of scoliosis, and enlarged subarachnoid space after surgery. Statistical significance was defined as a probability value <0.05. Values are presented as mean ± standard deviation (SD).

Statement of ethics

All applicable institutional and governmental regulations concerning the ethical evaluation of our patients were followed during the course of this research. This study was approved by the Institutional Review Board of School of Medicine, Keio University. We obtained signed consent forms from all patients.

Results

After surgery, syrinx size decreased in 11 patients (decreased group) and remained relatively unchanged in

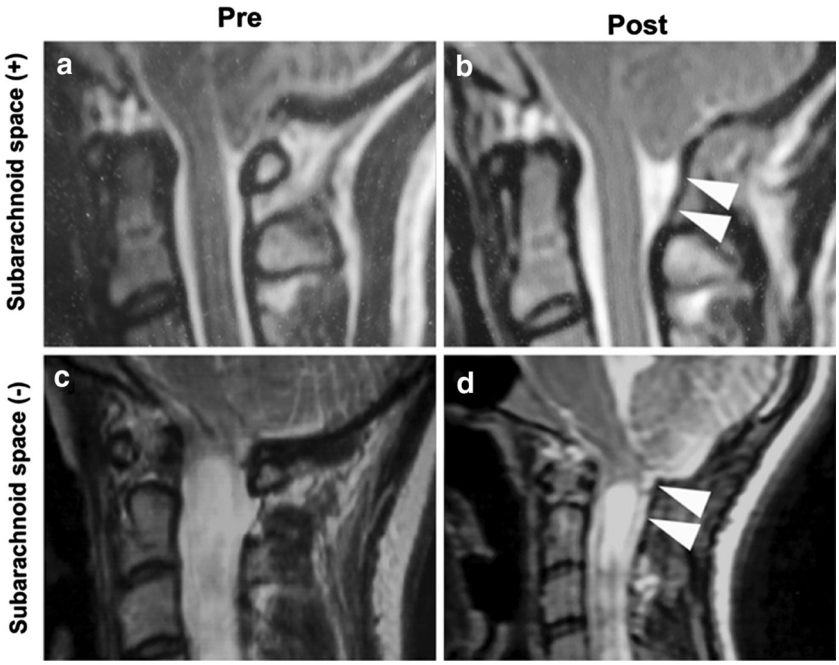

Fig. 2 Representative pre- and postoperative T2-weighted magnetic resonance (MR) images. **a**, **b** Note that the subarachnoid space at the level of the foramen magnum expanded after surgery in the decreased-group patient (*arrowheads*). **c**, **d** This is in contrast to residual narrowing of the subarachnoid space in the patient in the unchanged group

Table 1 Outcome scale for syringomyelia proposed by Bidzinski [21]

Outcome	Results
Very good	Marked postoperative improvement with further stabilization
Good	Slight postoperative improvement with further stabilization
Poor	Temporary or no postoperative improvement with further deterioration

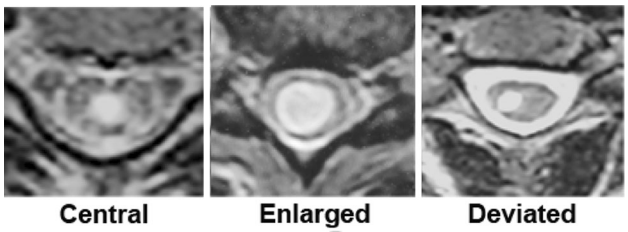

Fig. 3 Classification of syrinx type. Syrinx morphology was classified into three types—central, enlarged, and deviated—based on axial magnetic resonance (MR) images obtained at the level at which syrinx size was maximum

nine (unchanged group). Initial symptoms were segmental dysesthesia in 11 patients, numbness in 11, radicular pain in seven, and upper-extremity weakness in one. Two patients with scoliosis, whose syringomyelia was discovered and diagnosed by MRI, had no symptoms. Clinical results were analyzed in the 18 patients presenting with obvious symptoms. Following Bidzinski's [21] outcome

Table 2 Clinical results for the two groups

	Clinical result (no. of patients)		
	Very good	Good	Poor
Decreased	4	6	0
Unchanged	0	1	7

Of 20 patients, clinical results were analyzed in 18 symptomatic patients, and two asymptomatic patients with scoliosis were excluded. The difference in clinical outcomes between groups was significant ($p = 0.0007$)

scale, of the ten patients in the decreased group, four were classified as very good and six as good. In contrast, in the unchanged group, only one patient was classified as good and seven were classified as poor (Table 2). The difference in clinical outcomes between groups was significant ($p < 0.01$), suggesting that a reduction in syrinx size is correlated with clinical improvement. There were no complications, such as CSF collection in the operative wound, pseudomeningocele, or meningitis, in either group.

There was no significant difference in age at the time of surgery between groups (Table 3). Average morbidity duration was analyzed for the 18 symptomatic patients, excluding two scoliotic patients whose symptoms from syringomyelia were not obvious. The decreased group had suffered from syringomyelia for 14.1 months and the unchanged group for an average of 43.5 months. This difference in mean morbidity duration between groups was significant (Table 3), suggesting there could be a correlation between longer morbidity and unchanged syrinx size following surgery.

Table 3 Comparison of patient characteristics

	Decreased ($n = 11$)	Unchanged ($n = 9$)	P value
Age at the time of surgery	30.2 ± 18.7	31.9 ± 13.3	0.6208
Mean duration of morbidity (months)	14.1 ± 17.6	43.5 ± 46.7	0.0406*
Mean length of syrinx[a]	9.5 ± 4.1	14.3 ± 2.8	0.0113*
Enlarged subarachnoid space after surgery (%)	100	55.6	0.0260*
Patients with scoliosis (%)	63.6	55.6	0.5350
Cervical alignment from C2 to C7 (°)	−11.7 ± 16.1	-5.6 ± 11.8	0.4620
Basal angles (°)	136.3 ± 13.8	132.7 ± 8.7	0.3689
Clivo-axial angles (°)	141.8 ± 10.6	148.8 ± 8.5	0.1527

Mean ± standard deviation

[a] Values reflect the number of vertebral bodies spanned by the lesion

* Significant difference ($p < 0.05$) between groups

Table 4 Preoperative syrinx morphology

	Type of syrinx (no. of patients)		
	Central	Enlarged	Deviated
Decreased	5	6	0
Unchanged	0	3	6

$p = 0.0258$

Analysis of morphological features of preoperative syrinxes revealed that five patients in the decreased group had the central type of syrinx, whereas six had the enlarged type (Table 4). Of the nine patients in the unchanged group, six had a deviated syrinx and three an enlarged syrinx. The differences in morphology of preoperative syrinx between groups were significant, with patients in the decreased and unchanged groups having central and deviated syrinxes, respectively.

The average length of the preoperative syrinx was significantly shorter in the decreased group (9.5 vs 14.3 vertebral bodies) (Table 3). Notably, syrinx length in all patients in the unchanged group was more than ten vertebral bodies (Fig. 4). These results suggest that FMD might be a good indication for syringomyelia shorter than ten vertebral bodies.

Postoperative subarachnoid space at the foramen magnum was enlarged in every member of the decreased group, whereas residual narrowing was observed in four of the nine patients in the unchanged group, with a statistically significant intergroup difference (Table 3). In contrast, there were no significant differences in basal or clivoaxial angles, cervical alignment from C2 to C7, or presence or absence of scoliosis in preoperative radiographs between groups (Table 3). In the unchanged group, a

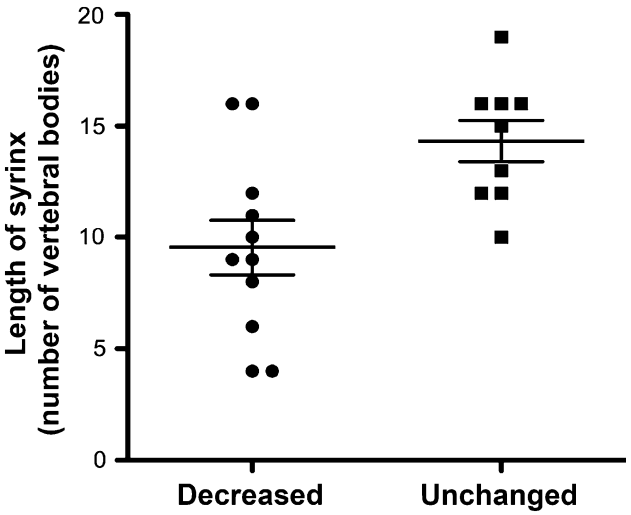

Fig. 4 Comparison of syrinx length between groups

syringosubarachnoid shunt was later performed in seven of the nine patients. Average duration between FMD and shunt was 16.3 months (range 6–24 months). Because there are some cases in which syrinxes gradually decrease after FMD, we wait for approximately 12 months before performing the second operation. After shunt treatment, the size of all syrinxes decreased, and patients' symptoms were relieved. The other two patients underwent conservative treatment consisting solely of medication. Although these two patients showed slight improvement, their syrinx size remained unchanged, and they continued to experience symptoms such as numbness and dysesthesia.

Discussion

We performed FMD with excision of the outer layer of the dura mater in patients with syringomyelia due to Chiari type I malformation and prospectively examined the factors contributing to the surgical outcome. After the surgery, 55 % of patients showed an improvement in clinical symptoms, along with a decrease in syrinx size. Clinical symptoms of the other patients did not improve, and there was no change in syrinx size. Patients with deviated syrinx, a long syrinx, and a long duration of morbidity showed a poor prognosis after FMD. Furthermore, patients had a poor prognosis when postoperative MRI showed residual narrowing of the subarachnoid space. Thus, the ability of this surgical procedure to improve clinical symptoms is not consistent, being dependent upon the underlying pathogenesis of the syringomyelia. Considering that the condition of most patients with an unchanged syrinx size was improved by additional procedures, such as syringosubarachnoid shunt, we conclude that the choice of surgical procedure for each pathological condition is critical for the

effective treatment of the syringomyelia in Chiari type I malformation.

These results suggest that syrinx size can be reduced by FMD in patients with a shorter morbidity duration and a short syrinx (<10 vertebral bodies) and of the central type. In agreement with our results, previous studies demonstrated that these factors contribute to clinical improvement after this surgical procedure [9, 18, 23, 24]. In contrast to the occasional report of postoperative complications, such as CSF collection in the operative wound, pseudomeningocele, or meningitis, when FMD was used with dural opening or duraplasty [3–6, 9, 10, 12, 15], there is no risk of such complications when FMD is used with excision of the dural outer layer [16–18]. Furthermore, previous reports demonstrated that FMD without duraplasty has a lower risk of complications than that with duraplasty [25, 26]. Therefore, treatment consisting of FMD with excision of the outer dural layer may provide optimal benefit with a low risk of postoperative complications if the patient has the prognostic factors for improvement.

Few studies have demonstrated a relationship between syrinx morphology and postoperative results in patients with Chiari type I malformation. In this study, treatment with FMD produced both clinical improvement and a smaller syrinx in all patients with a central syrinx. Similar to these results, previous studies demonstrated that patients with the central type of syrinx rather than the deviated type tend to recover well from preoperative neuropathic pain after surgery [20, 23]. Milhorat et al. [22, 27] demonstrated that the central syrinx is anatomically continuous with the fourth ventricle. Therefore, decompression at the level of the foramen magnum for this type of syrinx is a reasonable approach for releasing an obstruction in CSF circulation and allowing communication between syrinx and the fourth ventricle. In contrast, we found that FMD did not reduce the syrinx size for all patients with the deviated type. This form of syrinx originates in the gray or white matter of the parenchyma and does not communicate with the central canal [27]. The deviated syrinx is thought to be produced by cleft formation and extracellular fluid accumulation in the parenchyma [28]. Thus, even if the CSF circulation at the level of the foramen magnum recovers following FMD, the deviated syrinx, which separates from the central canal, does not decompress and remains unchanged. Because patients in this study with a deviated syrinx experienced improvement after undergoing an additional operation (syringosubarachnoid shunt), direct decompression using a shunt may be adequate treatment for this syrinx type. Intriguingly, Hida et al. [14] placed a syringosubarachnoid shunt in patients as the initial treatment and reported no case of shunt malformation after shunt tube insertion into the ventral subarachnoid space. Although there are no other reports in which this procedure was followed, the syringosubarachnoid shunt alone may be an appropriate surgical choice for improving syringomyelia. In regard to cases with an enlarged syrinx reviewed in this study, surgical outcomes of FMD were not consistent. Therefore, for such cases, other prognostic factors, including symptom duration and syrinx length should be taken into account when determining the surgical indication of FMD or syringosubarachnoid shunt.

In this study, 44 % of patients with an unchanged syrinx size had residual narrowing of the subarachnoid space in the cerebellomedullary cistern after surgery, necessitating additional surgery. These results indicated that an adhesive arachnoiditis concomitantly developed at the level of the foramen magnum. In fact, Alfieri and Pinna [9] pointed out the existence of adhesive arachnoiditis at the level of the foramen magnum in visual inspection during duraplasty. Aghakhani et al. [5] reported surgical outcomes of Chiari-related syringomyelia and demonstrated that one factor associated with poor outcome was arachnoiditis. They also showed that the presence of arachnoiditis was associated with poor clinical presentation [5]. Previous reports demonstrated that FMD without duraplasty is associated with a higher risk of reoperation compared with that with duraplasty [26]. Although the reported risk of complications, including CSF leakage or meningitis, ranges from 2.7 % to 36 % [5, 6, 9, 10, 12, 15], FMD with duraplasty could be an appropriate choice for releasing subarachnoid space adhesion. Alternatively, syringosubarachnoid shunt could be considered as a means by which to directly decompress the syrinx, regardless of adhesive arachnoiditis at the level of the foramen magnum [13, 14]. Further studies are needed to determine the most suitable surgical procedures for treating adhesive cisterna associated with syringomyelia.

Previous studies demonstrated that Chiari patients with syringomyelia have a significantly smaller foramen magnum volume than control individuals [29]. These results indicate that the Chiari type I malformation is a disorder of the paraxial mesoderm that induces underdevelopment of the occipital bone and overcrowding in the posterior fossa [28]. In this study, our comparison of bone morphological factors, such as scoliosis, cervical alignment, and angles around the foramen magnum, revealed no significant differences between patients with a decreased versus an unchanged syrinx size. In agreement with our results, Ono et al. [18] compared the decrease in syrinx size after FMD in scoliotic versus nonscoliotic patients and found no statistically significant intergroup difference. Taken together, results of our study suggest that decreased or unchanged syrinx size after surgery is not affected by a mesodermal abnormality but by syrinx morphology or choice of surgical technique.

Conclusion

In patients with syringomyelia associated with Chiari type I malformation, this study showed that: (1) a longer length and (2) laterally deviated syrinx, (3) a longer morbidity

duration, and (4) postoperative residual narrowing of the subarachnoid space in the cerebellomedullary cistern are prognostic factors for an unchanged syrinx size following FMD. The pathogenesis of syringomyelia in Chiari type I malformation is not consistent, and the choice of surgical technique for each pathological condition is important.

Conflict of interest None.

References

1. 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.
2. Rhoton AL Jr. Microsurgery of Arnold-Chiari malformation in adults with and without hydromyelia. J Neurosurg. 1976;45:473–83.
3. Kotil K, Ton T, Tari R, Savas Y. Delamination technique together with longitudinal incisions for treatment of Chiari I/syringomyelia complex: a prospective clinical study. Cerebrospinal Fluid Res. 2009;6:7.
4. Di Lorenzo N, Palma L, Palatinsky E, Fortuna A. "Conservative" cranio-cervical decompression in the treatment of syringomyelia-Chiari I complex. A prospective study of 20 adult cases. Spine (Phila Pa 1976). 1995;20:2479–83.
5. Aghakhani N, Parker F, David P, Morar S, Lacroix C, Benoudiba F, Tadie M. Long-term follow-up of Chiari-related syringomyelia in adults: analysis of 157 surgically treated cases. Neurosurgery. 2009;64:308–15.
6. Munshi I, Frim D, Stine-Reyes R, Weir BK, Hekmatpanah J, Brown F. Effects of posterior fossa decompression with and without duraplasty on Chiari malformation-associated hydromyelia. Neurosurgery. 2000;46:1384–9.
7. Vanaclocha V, Saiz-Sapena N, Garcia-Casasola MC. Surgical technique for cranio-cervical decompression in syringomyelia associated with Chiari type I malformation. Acta Neurochir (Wien). 1997;139:529–39.
8. Tubbs RS, McGirt MJ, Oakes WJ. Surgical experience in 130 pediatric patients with Chiari I malformations. J Neurosurg. 2003;99:291–6.
9. Alfieri A, Pinna G. Long-term results after posterior fossa decompression in syringomyelia with adult Chiari Type I malformation. J Neurosurg Spine. 2012;17:381–7.
10. Depreitere B, Van Calenbergh F, van Loon J, Goffin J, Plets C. Posterior fossa decompression in syringomyelia associated with a Chiari malformation: a retrospective analysis of 22 patients. Clin Neurol Neurosurg. 2000;102:91–6.
11. Guyotat J, Bret P, Jouanneau E, Ricci AC, Lapras C. Syringomyelia associated with type I Chiari malformation. A 21-year retrospective study on 75 cases treated by foramen magnum decompression with a special emphasis on the value of tonsils resection. Acta Neurochir (Wien). 1998;140:745–54.
12. Ma J, You C, Chen H, Huang S, Ieong C. Cerebellar tonsillectomy with suboccipital decompression and duraplasty by small incision for Chiari I malformation (with syringomyelia): long term follow-up of 76 surgically treated cases. Turk Neurosurg. 2012;22:274–9.
13. Tator CH, Meguro K, Rowed DW. Favorable results with syringosubarachnoid shunts for treatment of syringomyelia. J Neurosurg. 1982;56:517–23.
14. Hida K, Iwasaki Y. Syringosubarachnoid shunt for syringomyelia associated with Chiari I malformation. Neurosurg Focus. 2001;11:E7.
15. Ergun R, Akdemir G, Gezici AR, Tezel K, Beskonakli E, Ergungor F, Taskin Y. Surgical management of syringomyelia-Chiari complex. Eur Spine J. 2000;9:553–7.
16. Isu T, Sasaki H, Takamura H, Kobayashi N. Foramen magnum decompression with removal of the outer layer of the dura as treatment for syringomyelia occurring with Chiari I malformation. Neurosurgery. 1993;33:844–9.
17. Gambardella G, Caruso G, Caffo M, Germano A, La Rosa G, Tomasello F. Transverse microincisions of the outer layer of the dura mater combined with foramen magnum decompression as treatment for syringomyelia with Chiari I malformation. Acta Neurochir (Wien). 1998;140:134–9.
18. Ono A, Suetsuna F, Ueyama K, Yokoyama T, Aburakawa S, Numasawa T, Wada K, Toh S. Surgical outcomes in adult patients with syringomyelia associated with Chiari malformation type I: the relationship between scoliosis and neurological findings. J Neurosurg Spine. 2007;6:216–21.
19. Ono A, Suetsuna F, Ueyama K, Yokoyama T, Aburakawa S, Takeuchi K, Numasawa T, Wada K, Toh S. Cervical spinal motion before and after surgery in patients with Chiari malformation Type I associated with syringomyelia. J Neurosurg Spine. 2007;7:473–7.
20. Ono A, Numasawa T, Wada K, Yokoyama T, Takeuchi K, Suetsuna F, Ueyama K, Toh S. Surgical outcomes of foramen magnum decompression for syringomyelia associated with Chiari I malformation: relation between the location of the syrinx and body pain. J Orthop Sci: Off J Japan Orthop Assoc. 2010;15:299–304.
21. Bidzinski J. Pathological findings in suboccipital decompression in 63 patients with syringomyelia. Acta Neurochir Suppl (Wien). 1988;43:26–8.
22. Milhorat TH, Johnson RW, Milhorat RH, Capocelli AL Jr, Pevsner PH. Clinicopathological correlations in syringomyelia using axial magnetic resonance imaging. Neurosurgery. 1995;37:206–13.
23. Nakamura M, Chiba K, Nishizawa T, Maruiwa H, Matsumoto M, Toyama Y. Retrospective study of surgery-related outcomes in patients with syringomyelia associated with Chiari I malformation: clinical significance of changes in the size and localization of syrinx on pain relief. J Neurosurg. 2004;100(3 Suppl Spine):241–4.
24. Batzdorf U, McArthur DL, Bentson JR. Surgical treatment of Chiari malformation with and without syringomyelia: experience with 177 adult patients. J Neurosurg. 2013;118:232–42.
25. Sakushima K, Hida K, Yabe I, Tsuboi S, Uehara R, Sasaki H. Different surgical treatment techniques used by neurosurgeons and orthopedists for syringomyelia caused by Chiari I malformation in Japan. J Neurosurg Spine. 2013;18:588–92.
26. Durham SR, Fjeld-Olenec K. Comparison of posterior fossa decompression with and without duraplasty for the surgical treatment of Chiari malformation Type I in pediatric patients: a meta-analysis. J Neurosurg Pediatr. 2008;2:42–9.
27. 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;82:802–12.
28. Koyanagi I, Houkin K. Pathogenesis of syringomyelia associated with Chiari type 1 malformation: review of evidences and proposal of a new hypothesis. Neurosurg Rev. 2010;33:271–84.
29. Badie B, Mendoza D, Batzdorf U. Posterior fossa volume and response to suboccipital decompression in patients with Chiari I malformation. Neurosurgery. 1995;37:214–8.