With the widespread use of MR imaging in recent years, syringomyelia is being diagnosed in an increasing number of patients. The authors of recent reports have suggested that early diagnosis and the establishment of surgical techniques such as foramen magnum decompression and creation of a syringosubarachnoid shunt have made it possible to reduce the size of the syrinx. Improvements in syringomyelia-related symptoms, however, do not always correspond to reductions in the size of the syrinx, and this may pose treatment difficulties. Pain is one of the major disturbing symptoms in patients with syringomyelia, but the mechanisms of its development are not fully understood, and postoperative improvement in such pain is difficult to predict.

The objectives of this study were to assess the surgery-related results obtained in patients who underwent treatment for syringomyelia associated with Chiari I malformation, particularly related to pain status, and to identify factors that may influence improvement in postoperative pain by comparing pre- and postoperative magnetic resonance (MR) imaging findings.

Clinical Material and Methods

Twenty-five patients underwent surgery for syringomyelia associated with Chiari I malformation at our hospital between 1984 and 2001. There were four male and 21 female patients who ranged in age at the time of surgery from 13 to 57 years (mean 37 years). Patients with syringomyelia associated with other diseases, such as spinal cord tumors, adhesive arachnoiditis, and spinal cord injuries, were excluded from this study. Twelve patients underwent placement of a syringosubarachnoid shunt, 11 underwent foramen magnum decompression, and two patients received both treatments. The postoperative period ranged from 1.5 to 15.5 years (mean 5.3 years). The patients were divided into two groups according to the presence or absence of preoperative pain. The patients in the pain group were further divided into those in whom pain improved and those in whom it did not postoperatively. Overall parameter

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

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Object. Pain is one of the major symptoms in patients with syringomyelia; however, its origin is not fully understood, and postoperative improvement of pain is difficult to predict. The objectives of this study were to assess the surgery-related results obtained in patients who underwent treatment for syringomyelia associated with Chiari I malformation, particularly related to pain status, and to identify factors that may influence improvement in postoperative pain by comparing pre- and postoperative magnetic resonance (MR) imaging findings.

Methods. The correlation between pre- and postoperative changes in the size and the location of the syrinx and pain improvement was investigated in 25 patients. The shapes of the syrinxes were classified into three types: central, enlarged, and deviated. In most cases in which the syrinx deviated toward the posterolateral aspect of the spinal cord at the level corresponding to dermatomal distribution of preoperative pain, the lesion remained at the same position postoperatively, and improvement in pain was poor. On the other hand, enlarged-type syrinxes were the most frequently observed prior to surgery, exhibited diverse changes postoperatively, and improvement in pain status was difficult to predict. When postoperative MR imaging revealed a transformation to the deviated type, poor pain improvement was noted.

Conclusions. Neurons in the dorsal horn were thought to be involved in the development of pain as a result of the deafferentiation mechanism in cases of syringomyelia.

KEY WORDS • Chiari I malformation • syringomyelia • pain • magnetic resonance imaging

Abbreviation used in this paper: MR = magnetic resonance.
ters examined were age at the time of surgery, duration of disease, and changes in the location and size of the syrinx based on pre- and postoperative MR images. The morphology of the syrinx was classified into three types—central, enlarged, and deviated—based on the axial MR imaging findings at the level corresponding to the dermatomal distribution of pain in the pain group and at the level where the size of the syrinx was maximum in the nonpain group.

Results

Clinical Symptoms

The initial symptoms were segmental dysesthesia in 14 patients (56%), radicular pain in 10 (40%), and upper-extremity weakness in three patients (12%). The interval between onset of the initial symptoms to surgery ranged from 0.5 to 12 years (mean 2.9 years). The most common preoperative clinical symptoms were segmental pain (17 patients [68%]), sensory loss (15 [60%]), and motor weakness (eight patients [32%]). In the 17 patients with pain (the pain group), postoperative improvement was noted in six (pain-improvement group), whereas in 11 little improvement was demonstrated (no pain-improvement group). None of the eight patients without preoperative pain (non-pain group) developed pain postoperatively.

Factors Influencing Preoperative Pain

The age at the time of surgery and the duration of the disease were compared between those with and those without pain as factors that might be related to the development of preoperative pain. Although no significant intergroup difference was observed in the age at the time of surgery, the duration of the disease in the pain group was significantly longer than that in those without pain prior to surgery (p < 0.05) (Fig. 2).

Analysis of the morphological features of the syringes in patients with and without preoperative pain revealed that nine patients (53%) harbored an enlarged-type syrinx and eight (47%) a deviated-type lesion in the pain group; the central-type syrinx was not observed in this group. In the nonpain group, an enlarged-type syrinx was found in six patients (75%) and a central-type lesion in two patients (25%); no deviated-type syrinx was demonstrated (Table 1).

![Fig. 1. Classification of syrinx type. The morphology of the syrinx was classified into three types: central, enlarged, and deviated type, based on axial MR images obtained at the level corresponding to the dermatomal distribution of pain in the pain group and at the level where the size of the syrinx was maximum in the nonpain group.](image1)

![Fig. 2. Graphs showing comparisons of age at surgery and the disease duration between patients with pain and those without pain. Although no significant intergroup difference was observed in age at the time of surgery, the duration of the disease in the pain group was significantly longer than that in the other group (p < 0.05).](image2)

![Fig. 3. Graphs demonstrating comparisons of age at surgery and disease duration between patients in whom pain improved and those in whom it did not. Although no significant intergroup difference in age at surgery was observed, disease duration in the nonpain-improvement group was significantly longer than that in the other group (p < 0.05).](image3)
Factors Influencing Postoperative Pain Improvement

Age at the time of surgery and the duration of disease were compared between the group in which pain improved and that in which pain failed to improve as factors that might be related to postoperative improvement. Although no significant intergroup difference in the age at the time of surgery was observed, the duration of the disease was significantly longer in those in whom pain did not improve than in those in whom it did ($p < 0.05$) (Fig. 3).

Analysis of the morphological features of the syringes in patients with improved postoperative pain status revealed that in four of six patients, the syrinx transformed from an enlarged-type to a central-type lesion. In the remaining two patients, the enlarged-type syrinx persisted in one and disappeared in another in whom the deviated type had been present preoperatively. On the other hand, of the 11 patients in whom pain did not improve, the enlarged-type syrinx transformed to the deviated type in three, and the deviated type persisted in six. Thus, the number of patients in whom the deviated-type syrinx eventually appeared (nine patients [82%]) in the no pain-improvement group was significantly higher ($p < 0.05$) than that in the pain-improvement group (Table 2).

### Discussion

The acceleration of pain sensitivity resulting from glial scar formation and disinhibition of the medial lemniscus is thought to be the underlying mechanism of central pain in spinal diseases such as syringomyelia and spinal cord injury. In recent years, the mechanism of central pain, especially that due to deafferentation, has been more clearly elucidated. When any abnormality appears in the afferent tract of pain, especially in the spinothalamic tract, abnormal nervous discharges mediated by $N$-methyl-$D$-aspartate receptors occur at the thalamic dorsoventral nucleus, leading to the development of spontaneous pain, hyperalgesia, and so-called allodynia. On the other hand, central pain can also be induced by injury to the pathway of pain at any site from the dorsal horn of the spinal cord to the cerebral cortex. In the case of syringomyelia, dysfunction of the dorsal horn is thought to cause central pain. To the best of our knowledge, however, no previous studies have been conducted to compare changes in the syrinx morphology before and after surgery and the relation between these changes and pain relief in patients with Chiari I malformation–associated syringomyelia.

### Table 2

**Summary of postoperative changes of the morphology of the syrinx in relation to pain improvement**

<table>
<thead>
<tr>
<th>Status (no. of cases)</th>
<th>Type of Syrinx (no. of cases)</th>
<th>Preop</th>
<th>Postop</th>
</tr>
</thead>
<tbody>
<tr>
<td>no pain (8)</td>
<td>enlarged (6) → central (3)</td>
<td></td>
<td>disappeared (3)</td>
</tr>
<tr>
<td></td>
<td>central (2) → disappeared (2)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>pain (17)</td>
<td>enlarged (5) → deviated (1)</td>
<td>enlarged (1) → disappeared (1)</td>
<td></td>
</tr>
<tr>
<td>improved (6)</td>
<td>central (4) → deviated (3)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>not improved (11)</td>
<td>enlarged (4) → deviated (6)</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>deviated (7) → deviated (1)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* Arrows indicate the transformation (or lack of transformation) of one syrinx type into another.

### Fig. 4

Diagrams illustrating mechanism of pain. **A:** An enlarged syrinx that disappears or changes to the central type after the operation is more likely to be one formed as a result of central canal enlargement. The afferent nerve fibers passing through the anterior gray matter commissure (lateral spinothalamic tract) are likely to regain their function postoperatively as a result of the decompressive effect produced by the reduction in syrinx size, and pain resulting from deafferentation therefore should improve. **B:** In a residual deviated-type syrinx, the nerve cells in the dorsal horn around the Rex III to V layers may already have sustained irreversible damage, and no satisfactory improvement in pain due to deafferentation may be obtained, even if the syrinx diminishes in size postoperatively.
In the present study, patients in whom preoperative MR imaging revealed a syrinx deviated toward the posterolateral aspect of the spinal cord at the level corresponding to the dermatomal distribution of the pain, the syrinx often remained at the same site postoperatively, even if it had reduced in size; improvement in pain status was poor. On the other hand, patients harboring an enlarged-type syrinx (the most frequently observed type of syrinx prior to operation) exhibited diverse postoperative changes, making prediction of postoperative pain improvement difficult. Although a postoperative improvement in pain might be expected in patients in whom the syrinx reduced in size and shifted toward the center, such improvement is unlikely in those in whom the syrinx remains at the posterolateral aspect of the spinal cord. In cases of enlarged-type syringes, subsequent pain relief may vary depending on whether the syrinx changes to a central-type or to a deviated-type lesion after surgery. As suggested by Milhorat, et al.,7 an enlarged syrinx that disappears or changes to the central type after surgery is more likely to be one that formed as a result of central canal enlargement. The afferent nerve fibers passing through the anterior gray matter commissure (lateral spinothalamic tract) are likely to regain their function postoperatively as a result of the decerebration effect due to the syrinx’ reduced size, and pain resulting from deafferentiation should improve thereafter (Fig. 4A). On the other hand, a residual deviated-type syrinx may be one of the following two types: 1) one that is the consequence of the progressive enlargement of the central canal up to the parenchyma of the dorsal horn; or 2) one present in the dorsal horn preoperatively. In both cases, the nerve cells in the dorsal horn around the Rex III to V layers may have already sustained irreversible damage, and no satisfactory improvement in pain as a result of deafferentation can be obtained, even if the syrinx diminishes after the operation (Fig. 4B). This notion was supported by our finding that the duration of the disease in patients without pain improvement was significantly longer than that in those in whom pain did improve. As has been previously reported,10 irreversible changes in the spinal cord, such as neuronal loss, glial scar formation, and myelomalacia, may occur in cases in which the duration of disease has been lengthy. Taken together, our results suggest that postoperative postrolateral deviation of the syrinx and a long disease duration may indicate a poor prognosis for improved pain status.

Conclusions

In patients with syringomyelia associated with Chiari I malformation, a long disease duration and the presence of an MR imaging–documented deviated syrinx are the prognostic indicators of poor pain improvement. Nerve cells in the dorsal horn were thought to be involved in the development of the pain as a result of deafferentation in cases of syringomyelia.

References


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