Different surgical treatment techniques used by neurosurgeons and orthopedists for syringomyelia caused by Chiari I malformation in Japan

Clinical article

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Object. Syringomyelia is a rare disease commonly caused by Chiari I malformation. Surgery by neurosurgeons and orthopedists is a critical treatment for symptomatic patients, and surgical techniques are associated with improved symptoms for these patients. The aim of this study was to determine the different surgical techniques used by neurosurgeons and orthopedists in Japan to treat syringomyelia caused by Chiari I malformation.

Methods. Patients who had undergone a surgical treatment were identified from a 2-stage postal survey conducted in late 2009. The authors compared the type of surgery performed and its association with cavity size reduction, on the basis of whether patients were receiving care in a neurosurgery or orthopedics department.

Results. A total of 232 patients with syringomyelia caused by Chiari I malformation were included in this study. Two-thirds of patients were treated in a neurosurgery department and the other third in an orthopedics department. Neurosurgeons preferred foramen magnum decompression (FMD) with dural patch grafting, and orthopedists preferred FMD with dural dissection. Foramen magnum decompression with dural patch grafting was associated with better outcomes than was dural dissection with regard to the following: motor impairment (66% vs 39%, *p* < 0.05), sensory disturbance (60% vs 43%, *p* = 0.051), pain (67% vs 47%, *p* < 0.05), and cavity size (74% vs 58%, *p* < 0.05). Improved motor function was associated more with cavity size reduction than with sensory disturbance and pain.

Conclusions. Surgical procedures and outcomes differed, depending on whether the patient’s care was managed in a neurosurgery or orthopedics department. Outcomes were better after FMD with dural patch grafting.

(keywords: syringomyelia, nationwide survey, surgical procedure, neurosurgery, orthopedics, technique)

Syringomyelia is a rare disease characterized by abnormal fluid-filled cavities or cysts within the spinal cord. Syringomyelia is caused by various pathological conditions, the most common of which is a Chiari I malformation, which can disturb the flow of CSF between the posterior cranial fossa and the spinal canal. Other causes include trauma, arachnoiditis, Chiari II malformation, and idiopathic origin. The essential treatment for syringomyelia is surgery to repair the structural abnormality associated with impaired CSF flow and syrinx. Until the 1980s, the main procedure for treating syringomyelia with Chiari I malformation was FMD with plugging of the obex (Gardner operation). However, this procedure was accompanied by severe complications such as wound infection, CSF accumulation, and respiratory failure; thus, other surgical procedures were developed. Procedures that have recently become common are FMD with an additional procedure, such as dural patch grafting with or without intradural dissection and FMD with dural dissection (removal or microincision of the outer layer of the dura). With these improvements, the surgical treatment of syringomyelia is evolving. Various surgical procedures have been performed in neurosurgery and orthopedics departments, although variations in the treatments and their outcomes remain unclear. Therefore, to identify the different surgical techniques used by neurosurgeons and orthopedists to treat syringomyelia caused by Chiari I malformation in Japan, we examined data from a nationwide survey on the epidemiology of syringomyelia in Japan.

Methods

Data Source

This study was approved by the Institutional Review Board of Hokkaido University Hospital. We used a subset of syringomyelia patient data from a nationwide syringo-
myelomelia survey that was performed in late 2009. These data were collected via a 2-stage postal survey sent to departments associated with the treatment of syringomyelia, including neurosurgery, neurology, orthopedics, and pediatrics. The first stage of this postal survey was performed by using a stratified random process for selection of departments, based on a list of all hospitals in Japan with 20 or more beds, which was obtained from the Ministry of Health, Labour and Welfare. Sampling rates for general hospitals with 20–99, 100–199, 200–299, 300–399, 400–499, and 500 or more beds were approximately 5%, 10%, 20%, 40%, 80%, and 100%, respectively. In addition, all university hospitals were included in the survey because they were major institutions for the surgical treatment of patients with syringomyelia in Japan.

The first stage of the survey asked each department for the number of ambulatory patients with syringomyelia seen during the past year. The second stage of the survey collected detailed information on the patients reported in the first stage via a structured questionnaire, written by clinicians of each department and based on chart review. The questionnaire asked about patient demographics (sex, date of birth, date of disease onset, diagnosis, and family history), clinical signs and symptoms at diagnosis (motor deficits, sensory disturbances, autonomic failure, cranial nerve disturbances, and skeletal deformity), imaging findings (range of syrinx and morphologic asymmetry), surgical treatment (patient age and procedure performed, restricted to symptomatic patients), and clinical course of disease.

Each domain of clinical signs and symptoms was defined on the questionnaire as follows. Motor deficits included weakness, muscle atrophy, spasticity, hypotonia, and abnormal plantar reflex. Sensory disturbances included those of the neck, trunk, and limbs. Autonomic failure included Horner syndrome, anisocoria, dyshidrosis, abnormal nail development, limb hypertrophy, bladder and rectal disturbance, orthostatic hypotension, impotence, and neurogenic arthropathy. The clinical course of disease comprised progression after initial diagnosis and outcomes after the first surgical treatment in 4 domains (cavity size, motor impairment, sensory disturbance, and pain) and 3 categories (improved, not changed, and worse).

The study included only patients with syringomyelia caused by Chiari I malformation. Patients treated with syringosubarachnoid shunt only were excluded from the study because it was unclear whether the cause of syringomyelia in these patients was Chiari I malformation.

**Statistical Analysis**

The statistical analyses were performed by using JMP statistical software version 8 (SAS Institute). Results are given as the mean with SD or proportion for each item. The chi-square test was used to evaluate frequencies. Statistical significance was defined as p < 0.05. The association between cavity size and symptoms was evaluated by using the Cohen kappa statistic.

**Results**

Survey response rates were 73% for the first survey.
Among the 376 patients who had undergone surgical treatment for syringomyelia, this study included 232 patients with syringomyelia caused by Chiari I malformation (Table 1). Most patients had undergone surgical treatment within 2 years after the initial diagnosis of syringomyelia. Twice as many patients received care in a neurosurgery department as in an orthopedics department. The most frequent range of syrinx was the cervical-thoracic cavity. The most frequent symptom was sensory disturbance. To treat syringomyelia with Chiari I malformation, neurosurgeons preferred to perform FMD with dural patch grafting, whereas orthopedists preferred to perform FMD with dural dissection (Fig. 1A). There was no obvious association between syrinx range and type of surgery (Fig. 1B). Outcomes were better after FMD with dural patch grafting than after FMD with dural dissection (Fig. 1C) regarding motor symptoms (66% vs 39%, p < 0.05), sensory disturbance (60% vs 43%, p = 0.051), pain (67% vs 47%, p < 0.05), and cavity size (74% vs 58%, p < 0.05). Rates of reoperation after FMD with dural patch grafting and after FMD with dural dissection were 8.7% and 16.2%, respectively. Motor impairment was the symptom most associated with cavity size, but it was not strongly associated (Table 2).

![Fig. 1. A: Distribution of surgical procedures performed for syringomyelia patients with Chiari I malformation in each department. B: Distribution of surgical procedures performed for syringomyelia patients with Chiari I malformation by syrinx range. C: Improvement of each symptom after surgical treatment, according to the procedure performed. Statistical analysis was performed by using the chi-square test.](image-url)
Different surgical techniques for syringomyelia

Discussion

This study revealed 3 main findings regarding the surgical treatment of patients with syringomyelia. First, there was a significant difference between neurosurgeons and orthopedists regarding their preference of surgical procedure for syringomyelia with Chiari I malformation. Second, FMD with dural patch grafting yielded better outcomes than did dural dissection. Third, the degree of motor impairment was more strongly associated with cavity size than with sensory disturbance and pain after surgical treatment.

The surgical treatment of syringomyelia varied from direct intervention of the syrinx to modification of the associated structural abnormalities, particularly the Chiari malformation. Neurosurgeons have contributed to the elucidation of the pathogenesis and treatment of syringomyelia, whereas orthopedists have operated on patients with syringomyelia coexisting with scoliosis. Hence, the surgical treatments for patients with syringomyelia should involve neurosurgeons and orthopedists. A previous study showed that the number of ambulatory patients with syringomyelia in neurosurgery and orthopedics departments is approximately the same. In the present study, the number and percentage of patients with syringomyelia who had undergone surgery were greater in neurosurgery than in orthopedics departments. These results might be explained by differences in patient characteristics or reporting biases.

Neurosurgeons and orthopedists differed in their preference of surgical procedure for the treatment of syringomyelia with Chiari I malformation. Neurosurgeons preferred to perform FMD with dural patch grafting (in Japan, almost all seem to use Gore-Tex, W. L. Gore & Associates), whereas orthopedists preferred to perform FMD with dural dissection (although neurosurgeons in Japan have proposed a procedure involving removal of the outer layer of the dura). Surgical complications, such as CSF leakage, seemed to be more frequent after FMD with dural patch grafting than after FMD with dural dissection. The results of the present study show that with regard to the symptoms caused by syringomyelia, outcomes were better after FMD with dural patch grafting than after FMD with dural dissection. Munshi et al. reported the superiority of FMD with dural patch grafting (duralplasty) over FMD without duraplasty in a patient with Chiari I malformation. Other authors, however, have reported that suboccipital craniectomy with C-1 laminectomy or FMD without duraplasty for treatment of Chiari I malformation had adequate outcomes and electrophysiologic results. The optimal procedure for the treatment of syringomyelia with Chiari I malformation remains undetermined; however, the results of this study indicate that FMD with dural patch grafting might be preferable for symptom improvement. The different procedures chosen by neurosurgeons and orthopedists might have been affected by a political issue; however, our results should be taken into consideration for the improvement of surgery outcomes by adopting the best procedure.

The evaluation of the association between cavity size and symptoms after surgery indicated that motor impairment exhibited a stronger correlation with cavity size than it did with sensory disturbance and pain. Generally, the reduction of the cavity after surgery is accompanied by improvement of symptoms caused by this structure. A difference was found in symptom improvement according to cavity size, although the outcomes of surgery in our study were evaluated by using 3 broad categories.

This study had some limitations. The first limitation stemmed from using a survey method to interpret the results. This study was not an inventory survey and was performed by using a limited-source population. However, the representativeness of the patients recruited might have been maintained because we adopted a stratified random sampling method for selecting departments and achieved a relatively good response rate. In addition, this study enrolled all university hospitals and large general hospitals, where most patients would have undergone surgery. The second limitation is that technical proficiency was not considered in this study.

<table>
<thead>
<tr>
<th>Impairment</th>
<th>Cavity Size</th>
<th>Cohen κ (95% CI)*</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Improved</td>
<td>Not Changed</td>
</tr>
<tr>
<td>motor impairment</td>
<td></td>
<td></td>
</tr>
<tr>
<td>improved</td>
<td>37</td>
<td>6</td>
</tr>
<tr>
<td>not changed</td>
<td>11</td>
<td>23</td>
</tr>
<tr>
<td>worse</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>sensory disturbance</td>
<td></td>
<td></td>
</tr>
<tr>
<td>improved</td>
<td>55</td>
<td>8</td>
</tr>
<tr>
<td>not changed</td>
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<td>28</td>
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<tr>
<td>worse</td>
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<td>0</td>
</tr>
<tr>
<td>pain</td>
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</tr>
<tr>
<td>improved</td>
<td>53</td>
<td>8</td>
</tr>
<tr>
<td>not changed</td>
<td>17</td>
<td>23</td>
</tr>
<tr>
<td>worse</td>
<td>3</td>
<td>3</td>
</tr>
</tbody>
</table>

* Interpretation of Cohen kappa: < 0.20, poor; 0.21–0.40, fair; 0.41–0.60, moderate; 0.61–0.80, strong; 0.81–1.00, almost perfect.
patients with syringomyelia are affected not only by the choice of procedure but also by the surgeon’s proficiency. Therefore, the difference in outcomes between FMD with dural patch grafting and FMD with dural dissection might have been affected by surgeon proficiency.

Conclusions

Neurosurgeons and orthopedists preferred different surgical procedures for the treatment of syringomyelia. Better outcomes were achieved by FMD with dural patch grafting.

Disclosure

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The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Sakushima. Acquisition of data: Sakushima. Analysis and interpretation of data: Sakushima. Drafting the article: Sakushima. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Sakushima. Administrative/technical/material support: Hida. Study supervision: Hida, Yabe, authors. Statistical analysis: Sakushima. Administrative/technical/material support: Hida. Study supervision: Hida, Yabe, Tsuoi, Uehara, Sasaki.

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References


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