Cervical duraplasty with tenting sutures via laminoplasty for cervical flexion myelopathy in patients with Hirayama disease: successful decompression of a “tight dural canal in flexion” without spinal fusion

Clinical article

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Object. Hirayama disease, juvenile muscular atrophy of the distal upper extremity, is a rare type of cervical flexion segmental myelopathy and its etiology is still being debated. Two theories have been proposed: a “contact pressure” theory and “tight dural canal in flexion” theory. Previously reported treatments, including conservative neck collar therapy and surgical spinal fusion, used fixation of the cervical spine with the aim of avoiding contact pressure between the cord and anterior structures. On the other hand, treatment by duraplasty without spinal fusion has also been used, which aims at decompressing a tight dural canal in flexion by preventing abnormal forward displacement of the posterior dura mater without restricting cervical motion in young patients.

The authors developed a new surgical approach for treating a tight dural canal in flexion in patients with Hirayama disease: cervical duraplasty with tenting sutures via laminoplasty without spinal fusion. With this treatment they aimed to both decompress the spinal cord and preserve as much cervical motion as possible. The purpose of this study was to assess the clinical outcomes of patients who underwent this new surgical procedure and to investigate the etiology of Hirayama disease.

Methods. Six male patients (age range 17–23 years) with Hirayama disease underwent surgery between 2006 and 2012. The pre- and postoperative anteroposterior diameters of the dural canal in the flexed neck position, grip strength of the bilateral upper extremities, cervical alignment (C2–7), and cervical local flexion range of motion were compared. The presence or absence of surgical complications was assessed. To investigate the comparison group of Hirayama disease treated with spinal decompression, the PubMed database was searched for all relevant English-language case reports and series published between 1990 and 2013.

Results. The postoperative anteroposterior diameters of the dural canal were significantly expanded in the flexed neck position (7.2 ± 2.2 mm preoperatively vs 9.8 ± 1.7 mm postoperatively, p = 0.001). Grip strength of the upper extremities significantly improved bilaterally (20 ± 14 kg preoperatively vs 26 ± 15 kg postoperatively, p = 0.001). No significant difference was observed between pre- and postoperative cervical alignment in the neutral neck position (7.7° ± 8.1° preoperatively vs 9.0° ± 7.7° postoperatively, p = 0.74) or the cervical local flexion angle in the flexed neck position at the corresponding level of laminoplasty (16.6° ± 5.1° preoperatively vs 15.0° ± 9.4° postoperatively, p = 0.8). No surgical complications were noted, except for transient CSF leakage, which was resolved after lumbar drainage. The systematic review identified 37 cases from 7 reports: 26 with spinal fusion only, 5 with duraplasty without fusion, and 6 with combined duraplasty and fusion. In the largest series, in which 12 cases were treated with anterior fusion, cervical alignment was maintained, but local flexion motion was significantly decreased as a result of fixation. Although significant improvements in or stabilization of grip strength occurred in all 7 reported studies regardless of decompression procedures, one major delayed surgical complication was noted in a patient treated with anterior fusion. The patient developed severe kyphotic changes, which required reconstruction surgeries.

Conclusions. Cervical duraplasty with tenting sutures via laminoplasty prevented abnormal forward displacement of the posterior dura mater while preserving normal anterior structures and flexion motion of the cervical spine without major surgical complications. The clinical improvements achieved by the authors’ method support evidence that a tight dural canal in flexion largely contributes to segmental myelopathy in patients with Hirayama disease.

Key Words • cervical flexion myelopathy • systematic review • forward displacement of the dura mater • spinal fusion • complication • follow-up study

Abbreviation used in this paper: ROM = range of motion.

* Drs. Ito and Takai contributed equally to this work.
It is generally worsened by neck extension; however, rare
groups of cervical myelopathies are caused by neck flex-
ion.9

Hirayama disease, juvenile muscular atrophy of the
distal upper extremity, is a rare type of cervical flexion
segmental myelopathy. Major symptoms include a motor
deficit and muscular atrophy of the distal upper extrem-
ity with slight or no subjective or objective sensory dis-
turbances.4 This muscular atrophy is unilateral in most
patients and asymmetrically bilateral in others. The peak
age at onset is 15–17 years, with a marked male prepon-
derence, commonly slow onset and progression, and qui-
escence several years after its onset.18 Because Hirayama
disease differs from intrinsic motor neuron disease and
spinal muscular atrophy, early diagnosis and specific
treatments are important.

Two theories have been proposed as the causes of
flexion myelopathy: a “contact pressure” theory and a
“tight dural canal in flexion” theory. The contact pressure
theory states that normal anterior structures, including
intact vertebral bodies and intervertebral discs, contrib-
ute to contact pressure over the spinal cord in flexion of
the cervical spine.20 On the other hand, the tight dural
canal in flexion theory states that abnormal forward dis-
placement of the posterior dura mater contributes to spi-
nal cord compression against normal anterior structures
from behind.7

Treatments based on the first theory, which aim to
relieve contact pressure by the fixation of normal cervi-
cal vertebras, include conservative neck collar therapy19
and anterior or posterior spinal fusion and have been
frequently reported.10,11,20 However, to the best of our
knowledge, treatments to decompress a tight dural canal
in flexion by preventing abnormal forward displacement
of the posterior dura mater without spinal fusion, such as
duraplasty without spinal fusion, have been reported in
only a few cases.1,3

We developed a new surgical treatment for a tight
dural canal in flexion in patients with Hirayama disease:
cervical duraplasty with tenting sutures via laminoplasty
without spinal fusion. With this treatment we aimed to
both decompress the spinal cord and preserve as much
cervical motion as possible. The purpose of this study
was to assess the clinical outcomes of patients who un-
derwent this new surgical procedure and to investigate
the etiology of Hirayama disease.

**Methods**

**Patient Population**

The study population included 6 consecutive male
patients (age range 17–23 years) in whom Hirayama dis-
ease was diagnosed at our institution between 2006 and
2012 (Table 1). The diagnosis of Hirayama disease was
made on the basis of the following clinical signs and ex-
amination results: onset between the age of 10 years and
the early 20s; unilateral or asymmetrical bilateral atrophy
of the intrinsic hand muscles and the flexor muscle group
of the forearm, except for the brachioradialis muscle (Fig.
1); no objective sensory disturbance; no objective find-
ings in the lower extremities; a neurogenic pattern on an
electromyographic examination restricted to the affected
upper extremity; and neuroradiological findings that in-
cluded the absence of disc herniations, spondylotic spur,
or hypertrophy of the yellow ligaments, as well as the
presence of forward displacement of the posterior wall of
the lower cervical dura mater in the flexed neck posi-
tion, leading to spinal cord compression and spinal cord
atrophy, as seen on CT myelography (Fig. 2) and/or MRI
(Figs. 3 and 4). Other causes of myelopathy, such as spinal
cord tumors and vascular malformations, syringomyelia,
motor neuron disease, and neuropathy, were excluded.

**Surgical Techniques**

Figure 3 shows illustrations of the surgical techniques
together with operative photographs. After a midline skin
incision was made, the vertebral arches were split in the
midline while preserving the spinous processes and nu-
clal ligament. Bilateral hinges were made by creating bur
holes through the outer cortical bone of the lamina-facet
borders (Fig. 3A and E). After double-door opening of the
vertebral arches, the engorged extradural venous plexus
was moved outward beyond the midline (Fig. 3F) and a
posterior dural incision was made under a microscope
while keeping the arachnoid intact (Fig. 3B and G). Dural
tenting sutures were placed between the edges of the du-
ral opening and yellow ligaments to lift the dura mater to
the vertebral arches and prevent forward displacement of
the lower cervical dura mater in the flexed neck position
(Fig. 3B and C). Duraplasty was made with the autolo-
gous nuchal fascia to cover the dural opening (Fig. 3B
and H). After duraplasty, small pieces of the autologous
bone graft were placed between the spinous processes.
The spinous processes were closed medially and secured
with sutures (Fig. 3C and D). The neck was fixed postop-
eratively with a cervical brace for the 1st and 2nd months,
and with a soft neck collar for the 3rd month.

**Data Collection and Statistical Analysis**

Medical records, radiological images, and operative
records were retrospectively reviewed to collect clinical
data. The postoperative follow-up period ranged between
12 and 75 months (mean 46 months). We compared the
pre- and postoperative anteroposterior diameters of the
dural canal at the superior region of the C-6 vertebral
body on T2-weighted MR images obtained with the pa-
tient in the flexed-neck position to determine whether the
cervical dural sac expanded. To confirm whether cervical
alignment was maintained, we compared pre- and post-
operative cervical alignment, which was measured as the
angle composed of the posterior lines of the C-2 and C-7
vertebral bodies in the neutral neck position on cervical
radiographs (Fig. 5). We then compared the pre- and post-
operative cervical local flexion angle, which was mea-
sured as the angle created by the posterior lines of the up-
per and lower vertebral bodies at the corresponding levels
of laminoplasty in the flexed neck position on cervical
radiographs to confirm whether the cervical flexion range
of motion (ROM) was maintained (Fig. 5). To assess clin-
cal outcomes, we compared the pre- and postoperative
Decompression of “tight dural canal in flexion”

TABLE 1: Summary of the preoperative clinical findings in 6 patients with Hirayama disease*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Duration of Symptoms (mos)</th>
<th>Presentation</th>
<th>Grip Strength (rt/lt, kg)</th>
<th>Location of Muscle Atrophy</th>
<th>Abnormal EMG Findings (neurogenic changes)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>20, M</td>
<td>81</td>
<td>lt hand weakness</td>
<td>34/7</td>
<td>lt forearm, hand</td>
<td>NA</td>
</tr>
<tr>
<td>2</td>
<td>19, M</td>
<td>36</td>
<td>bilat hand weakness</td>
<td>21/25</td>
<td>bilat hands</td>
<td>bilat Tri, FCR, FCU, FDI</td>
</tr>
<tr>
<td>3</td>
<td>17, M</td>
<td>15</td>
<td>bilat hand weakness</td>
<td>18/3</td>
<td>bilat forearms, hands</td>
<td>lt FDI, APB, FCR</td>
</tr>
<tr>
<td>4</td>
<td>15, M</td>
<td>6</td>
<td>rt hand weakness</td>
<td>6/30</td>
<td>rt hand</td>
<td>rt FCR, EDC, FDI</td>
</tr>
<tr>
<td>5</td>
<td>18, M</td>
<td>37</td>
<td>bilat hand weakness</td>
<td>6/19</td>
<td>rt forearm, hand</td>
<td>rt FCR, FCU, FDI, EDC, APB</td>
</tr>
<tr>
<td>6</td>
<td>23, M</td>
<td>48</td>
<td>lt hand weakness</td>
<td>50/24</td>
<td>lt forearm, hand</td>
<td>lt Del, Bi, Tri, FDI</td>
</tr>
</tbody>
</table>

* APB = abductor pollicis brevis; Bi = biceps; Del = deltoid; EDC = extensor digitorum communis; EMG = electromyography; FCR = flexor carpi radialis; FCU = flexor carpi ulnaris; FDI = first dorsal interosseous; NA = not available; Tri = triceps.

Grip strengths of the bilateral upper extremities. A hand grip dynamometer was used to measure grip strengths. We also assessed the presence or absence of perioperative surgical complications such as CSF leakage, infection, new radiculopathy, new myelopathy, subdural/epidural hematoma, bone graft displacement, vertebral arch fractures, worsened intractable neck pain, pulmonary embolism, and cardiopulmonary complications.

A paired t-test was used to statistically analyze the data. SPSS II for Windows (SPSS Japan Inc.) statistical software was used for statistical analysis. A p value of < 0.05 was considered significant.

To investigate the comparison group in the reported series of Hirayama disease patients treated with spinal decompression, we systematically reviewed the associ-

Fig. 1. Case 5. Photographs showing unilateral muscular atrophy of the right hand intrinsic (C) and flexor muscle group of the right forearm, except for the brachioradialis muscle (A and B).

Fig. 2. Case 6. Preoperative sagittal (A and B) and axial (C and D) CT myelograms showing spinal cord compression more in the flexed neck position than in the extended neck position by forward displacement of the posterior wall of the lower cervical dura mater. Note the spinal cord indentation produced by the dura mater (arrowhead, D).
ated literature in the MRI era. Two reviewers (K.T., M.T.) searched the PubMed database for all relevant English-language case reports and case series, published between 1990 and 2013, using text word–based searches with the terms “((((Hirayama disease) OR cervical flexion myelopathy) AND surgery) AND (“1990”[Date - Publication]:”2013”[Date - Publication])) AND English[Language].” Case reports and case series treated without spinal decompression, including those treated with musculotendinous transfer, were excluded.

This study protocol was approved by the Institutional Review Board at the Tokyo Metropolitan Neurological Hospital. Because this was a retrospective and noninvasive study, written patient informed consent was not obtained. Instead, a public notice that provided information on this study was posted on the Tokyo Metropolitan Neurological Hospital website.

Results

The clinical and radiological findings of the 6 patients are summarized in Table 1, and clinical outcomes are shown in Table 2. After surgery, abnormal forward displacement of the posterior wall of the lower cervical dura mater was prevented in the flexed neck position (Fig. 3). Therefore, the anteroposterior diameters of the dural canal were significantly expanded in the flexed neck position (7.2 ± 2.2 mm preoperatively vs 9.8 ± 1.7 mm postoperatively, \( p = 0.001 \), paired t-test [Table 2, Fig. 4]). Grip strength of the upper extremities significantly improved (20 ± 14 kg preoperatively vs 26 ± 15 kg postoperatively, \( p = 0.001 \), paired t-test [Table 2]), not only in the hand affected with muscular atrophy, but also in the opposite hand without atrophy. However, no significant changes were observed in muscular atrophy in any case. No significant difference was observed between pre- and post-

Fig. 3. Case 6. Illustrations (A–C), intraoperative photographs (E–H), and axial T2-weighted MR image (D) demonstrating the surgical techniques. The vertebral arches were split in the midline, and bilateral gutters were made (A and E). Note the marked engorgement of the extradural venous plexus (white asterisks, A and F). A posterior dural incision was performed after double-door opening (B and G), and duraplasty was performed using the autologous nuchal fascia (black asterisks, B and H). Dural tenting sutures were placed between the dura and yellow ligaments to lift the dura mater to the vertebral arches (arrows, B–D). After duraplasty, small pieces of the autologous bone graft were placed between the spinous processes, which were then closed medially and secured with sutures (C and D). Copyright Keisuke Takai. Published with permission.
Decompression of “tight dural canal in flexion”

Fig. 4. Case 2. Preoperative axial (A) and sagittal (C) T2-weighted MR images revealing spinal cord compression in the flexed neck position by forward displacement of the posterior wall of the lower cervical dura mater. Axial images of the C-5, C5–6, C-6, C6–7, C-7, and C7–T1 levels are arranged from the top to the bottom (A). Note the marked passive engorgement of the extradural venous plexus (asterisks, A and C). Postoperative axial (B) and sagittal (D) T2-weighted MR images show significant expansion of the dural canal. Axial images of C-5, C5–6, C-6, C6–7, C-7, and C7–T1 levels are arranged in a similar manner (B). Note the dura mater lifted to the vertebral arch (arrows, B) and the absence of forward displacement in the flexed neck position (arrowheads, D).

operative cervical alignment in the neutral neck position (7.7° ± 8.1° preoperatively vs 9.0° ± 7.7° postoperatively, p = 0.74, paired t-test [Table 2, Fig. 6]), or between the pre- and postoperative cervical local flexion angle in the flexed neck position at the corresponding levels of laminoplasty (16.6° ± 5.1° preoperatively vs 15.0° ± 9.4° postoperatively, p = 0.8, paired t-test [Table 2, Fig. 6]). No perioperative surgical complications were noted, except for transient CSF leakage, which was resolved 1 week after lumbar drainage.

Our systematic review identified a total of 37 cases from 7 reports (Table 3). Twenty-six cases (70%) were treated with multilevel anterior or posterior fusion only, 5 cases (14%) were treated with duraplasty via laminectomy or laminoplasty without spinal fusion, and 6 cases (16%) were treated with combined duraplasty and spinal fusion. After surgical decompression, significant improvements in or stabilization of grip strength was noted in all 7 reports, regardless of decompression procedures. Overall, there have been few objective data regarding postoperative biomechanical assessments of the cervical spine including cervical alignment and ROM. In the largest series,
in which 12 cases were treated with anterior fusion, cervical alignment was maintained, but local flexion ROM significantly decreased as a result of spinal fixation. In 2 reported cases treated with duraplasty via laminectomy without fusion, although postoperative imaging revealed that flexion ROM was maintained in the illustrative cases, objective measurements were not provided. One major delayed surgical complication was reported in a patient treated with anterior fusion (Table 3). The patient developed severe kyphotic changes at an adjacent level above the anterior fusion site, which were evident 3 months after surgery and had markedly progressed by 7 months. The patient required second and third reconstruction surgeries by means of anterior and posterior fusion procedures, which resulted in the correction of kyphosis with C3–7 immobilization.

Discussion

In the present study, we assessed 6 cases of Hirayama disease in which patients were treated with a newly developed nonfusion technique for a tight dural canal in flexion—cervical duraplasty with tenting sutures via double-door laminoplasty (Fig. 3, Table 1). In these patients, we arrested forward displacement of the posterior dura mater in the flexed neck position, which led to expansion of the cervical dural canal without spinal fusion. The spinal cord was decompressed without our having to remove the anterior structures of the normal cervical spine or restrict local flexion motion, which was also within the normal range (Figs. 4 and 6, Table 2). After this procedure, the grip strength of the upper extremities significantly improved (Table 2).

To the best of our knowledge, the present study is the first reported case series in which successful decompression of a tight dural canal in flexion was achieved without spinal fusion in young patients with Hirayama disease.

The etiology of Hirayama disease is still being debated. Patients with Hirayama disease have circulation
disturbances in the lower spinal cord, leading to an anterior horn–dominant impairment in which anterior horn cells are the most vulnerable to ischemia. Two theories have been proposed for the causes of these disturbances, a contact pressure theory and a tight dural canal in flexion theory. Critical Comparison of the Contact Pressure and Tight Dural Canal in Flexion Theories

The first theory, the contact pressure theory, is based on a classic cadaver study performed in the 1960s, in which specimens were divided into 3 groups and fixed in the flexed, neutral, or extended neck positions. Cervical spondylotic spurs were shown to contribute to spinal cord compression in the flexed neck position group, in which the spinal cord was stretched over abnormal anterior degenerative structures. In another cadaver study, by Reid17 contact pressure produced by cervical anterior structures over the anterior dura mater was most affected by the size of the spondylotic spurs, especially when they were 3 mm or larger.17

Previously reported treatments for Hirayama disease, including conservative cervical collar therapy and surgical spinal fusion, aimed at avoiding contact between the cord and anterior structures by restricting cervical flexion motion.10,11,15,19,20 Anterior spinal fusion decompresses the cord by removal of the intervertebral discs and a portion of the cervical vertebrae as well as fixation of the cervical spine.15,20 Posterior spinal fusion decompresses the cord by multilevel fixation of cervical vertebrae, leading to the arrest of normal flexion motion.3,10,11

In our case series, cervical vertebrae did not exhibit abnormal degenerative spondylotic changes, and the local cervical flexion angle was within the normal range (16.6° ± 5.1°), which was consistent with the findings of previous studies.20

Patients with Hirayama disease by definition do not have abnormal anterior degenerative spondylotic diseases of the spine, such as hard spondylotic spurs or soft-disc herniations5,18 or abnormal spinal subluxation;7 therefore, we concluded that there is no need to remove these normal structures, such as intact cervical vertebrae and discs, or restrict normal physiological cervical motion.9 Thus, the tight dural canal in flexion theory proved to be a more useful model in the treatment of these patients.

The “tight dural canal in flexion” theory was originally described in a flexion-extension myelographic study.
TABLE 3: A systematic review of the reported series of Hirayama disease treated with spinal decompression*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No of Cases</th>
<th>Operative Procedure</th>
<th>Spinal Canal or Cord Diameter</th>
<th>Mean C2–7 Alignment</th>
<th>Mean Flexion ROM</th>
<th>Mean Grip Strength (kg)</th>
<th>Muscle Atrophy</th>
<th>Complications</th>
<th>FU Period (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lin et al., 2010</td>
<td>4</td>
<td>AF</td>
<td>NA</td>
<td>NA</td>
<td>decreased†</td>
<td>improved in 2 cases</td>
<td>slightly improved in 2 cases</td>
<td>none</td>
<td>40</td>
</tr>
<tr>
<td>Lin et al., 2010</td>
<td>1</td>
<td>D-plasty via laminectomy w/o fusion</td>
<td>NA</td>
<td>NA</td>
<td>maintained‡</td>
<td>unchanged</td>
<td>improved</td>
<td>none</td>
<td>39</td>
</tr>
<tr>
<td>Arrese et al., 2009</td>
<td>1</td>
<td>D-plasty via laminectomy w/o fusion</td>
<td>increased in flexion§</td>
<td>NA</td>
<td>maintained‡</td>
<td>unchanged</td>
<td>unchanged</td>
<td>none</td>
<td>24</td>
</tr>
<tr>
<td>Patel et al., 2009</td>
<td>1</td>
<td>D-plasty via laminectomy w/ PF</td>
<td>NA</td>
<td>slight kyphosis</td>
<td>NA</td>
<td>improved</td>
<td>NA</td>
<td>none</td>
<td>3</td>
</tr>
<tr>
<td>Watanabe et al., 2005</td>
<td>12</td>
<td>AF</td>
<td>increased in neutral</td>
<td>maintained, ~4.7° → ~1.5°</td>
<td>decreased, 11.8° → 2.2°</td>
<td>improved, 16.3 → 18.6</td>
<td>unchanged</td>
<td>none</td>
<td>63</td>
</tr>
<tr>
<td>Fujimoto et al., 2002</td>
<td>3</td>
<td>D-plasty via laminectomy w/o fusion</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>improved in 3 cases</td>
<td>unchanged, later worsened in 2 cases</td>
<td>none</td>
<td>41</td>
</tr>
<tr>
<td>Kohno et al., 1999</td>
<td>10</td>
<td>AF in 7 cases, PF in 3 cases</td>
<td>NA</td>
<td>severe kyphosis in 1 case</td>
<td>decreased†</td>
<td>improved, 20.0 → 26.4</td>
<td>improved in 1 case, slightly improved in 1 case</td>
<td>reconstructive ops in 1 case¶</td>
<td>45</td>
</tr>
<tr>
<td>Konno et al., 1997</td>
<td>5</td>
<td>D-plasty via laminectomy w/ PF</td>
<td>increased in neutral</td>
<td>NA</td>
<td>NA</td>
<td>improved, 9.3 → 18.3</td>
<td>slightly improved in 5 cases</td>
<td>none</td>
<td>24</td>
</tr>
</tbody>
</table>

* AF = anterior fusion; laminectomy = laminectomy; PF = posterior fusion.
† Postoperative imaging showed a decreased flexion ROM in the illustrative case (objective measurements were not provided).
‡ Postoperative imaging showed maintained flexion ROM in the illustrative case (objective measurements were not provided).
§ Postoperative imaging showed an increased spinal canal diameter in the flexed neck position in the illustrative case (objective measurements were not provided).
¶ Severe kyphosis progressed several months after anterior fusion and required second and third reconstructive surgeries in 1 case.

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in 1987. The posterior surface of the lower cervical dura mater was shown to migrate forward more in the flexed neck position than in the neutral and extended neck positions. Abnormal forward displacement of the dura mater was shown to compress the cervical spinal cord against normal anterior vertebral bodies, even though there was no cervical spondylosis and the cervical flexion ROM was within normal limits. A tight dural canal was attributed to uneven growth of the spine and dura.5

After the introduction of MRI, abnormal forward displacement of the dura mater and flattening of the spinal cord could easily be identified using flexion MRI.12,13 Considerable attention has recently been paid to these neuroradiological findings because they have high sensitivity and specificity in the diagnosis of Hirayama disease. Although forward displacement of the dura mater also occurs in normal control subjects, flattening, widening, and distortion in the spinal cord do not occur, because forward displacement is generally 1 mm on average. On the other hand, forward displacement of the dura mater has been shown to be several times greater in patients with Hirayama disease than in normal subjects.12 As a result, the spinal cord is compressed against normal anterior structures by the abnormally shifted dura mater, even though there is no spondylosis or subluxation.9 Forward displacement of the dura mater was shown to migrate forward more in the flexed neck position,6 anterior dural shifting has also been shown to contribute to enlargements of the posterior epidural space, leading to passive posterior epidural vertebral venous engorgement in the flexed neck position without epidural pressure changes.16

In the largest reported case series study on surgical spinal fusion in patients with Hirayama disease, the local cervical flexion angle at the corresponding levels of the lower cervical spine was “corrected” from 11.8° ± 7.0° to 2.2° ± 2.9° by anterior fusion.20 Although grip strength significantly increased after surgery in this series, it is reasonable to conclude that in the patients in this case series, the removal of normal anterior structures and fixation of normal cervical motion contributed to the prevention of cervical flexion motion, which had an indirect influence on forward displacement of the posterior dura mater because forward displacement of the dura occurs only in the flexed neck position. Although the cervical flexion angle was preserved in an equal measure with the preoperative value in our series and the spinal cord shifted anteriorly in the flexed neck position, grip strength increased by 30% of preoperative values because the dura did not shift anteriorly and the spinal cord was free from compression by the dura mater. In a previous study, a patient required additional reconstructive surgeries after anterior fusion because delayed severe kyphotic changes occurred at an adjacent level above the fusion site.19 Our series was not complicated by adjacent-level kyphosis or restenosis during the follow-up period. The immobilization of normal physiological movement may lead to adjacent-level problems in the long term.

We believe that significant expansion of the dural canal in the flexed neck position by duraplasty is theoretically reasonable if the anteriorly shifted posterior dural mater contributes to cervical flexion myelopathy. Not only duraplasty, but also dural tenting sutures to lift the dura to the vertebral arches, prevented forward displacement of the dura mater in the flexed neck position, resulting in significant expansion of the dural canal by 2.6 mm on average (36% of preoperative diameter), which was 2 times greater than previous findings.11 Passive engorgement of the posterior epidural venous plexus also disappeared following surgery (Fig. 4). Although Case 2 developed a CSF fistula, it was resolved after lumbar drainage. Therefore, it was important to keep the arachnoid intact when the dural incision was made to prevent a CSF fistula. The clinical improvements observed in grip strength by our method support evidence that a tight dural canal in flexion induced by forward displacement of the lower cervical dura mater contributes largely to segmental myelopathy in patients with Hirayama disease.

Limitations

This study has some limitations. Because the incidence of Hirayama disease is very low, this study was limited to a small number of patients, and assessments of the outcomes of patients treated by our technique were not compared prospectively with those of patients treated with other medical or surgical techniques. Therefore, further well-designed studies are needed using larger numbers of patients.

Conclusions

Cervical duraplasty with tenting sutures via laminoplasty led to spinal cord decompression with the preservation of cervical alignment and local physiological motion in young patients with Hirayama disease without major complications. The clinical improvements achieved by our method support the theory that a tight dural canal in flexion largely contributes to segmental myelopathy in patients with Hirayama disease.

Disclosure

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

Conception and design: Takai, Taniguchi. Acquisition of data: all authors. Analysis and interpretation of data: Takai. Drafting the article: Takai, Ito. Critically revising the article: Takai. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Takai. Statistical analysis: Takai. Study supervision: Takai, Taniguchi. Medical illustrations: Takai.

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