Supraplacode spinal cord transection in paraplegic patients with myelodysplasia and repetitive symptomatic tethered spinal cord

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Object. The authors describe the technique of transecting the spinal cord in children born with myelomeningocele who have undergone multiple detherings and are functionally paraplegic.

Methods. The authors’ technique involves identifying the neural placode and sectioning the normal spinal cord just superior to this site. No postoperative complications have been identified in 14 patients undergoing this procedure over an 11-year period. No patient at last follow up was found to have symptoms referable to a tethered spinal cord. The advantage of this procedure is to excise the normally pia-coated cord, which is unlikely to retether compared with the neural placode, which is often covered with scar tissue and does not have a well-formed pial surface—hence, predisposing it to frequent dorsal adhesions.

Conclusions. The authors believe that this technique is of benefit in a small, carefully selected group of myelodysplastic patients with repetitive tethering of the spinal cord.

KEY WORDS • spina bifida • paraplegia • tethered cord syndrome • pediatric neurosurgery

RECURRENT tethering of the spinal cord is a troublesome problem for a select subset of patients with myelomeningocele. A central neurosurgical principle in the contemporary closure of an myelomeningocele at birth is that of sequentially reapproximating the normal anatomical layers that failed to separate during disordered embryogenesis. Despite this surgical restoration of normal anatomical planes, tethering of the distal spinal cord at the site of myelomeningocele closure is a common problem for many reasons. One of the most important is the absence of a normal pial plane along the neural placode. Another is the unavoidable contact between the placode and the reconstituted dural sleeve. Various authors pursue and have advocated a wide range of surgical techniques to reduce the incidence of symptomatic tethering of the spinal cord, yet the fact remains that no single technique has gained widespread favor as an effective means to prevent spinal cord tethering in the myelodysplastic population.

Tethering clinically manifests as progressive back pain and decline in neurological performance in the lower extremities and sphincters for patients who retain lower-extremity function and bowel and bladder continence. For paraplegic, incontinent patients, progressive severe pain in the midthoracic and lumbar region is most common. It can be difficult to distinguish between neurogenic pain from recurrent tethering and mechanical pain from degenerative disease; however, we have found that relentless and progressively severe back pain that is independent of positioning and activity is highly associated with spinal cord tethering in this subset of patients. Syringomyelia may also develop when the cord is tethered and may threaten either upper-extremity function or any preserved lower-extremity function. Approximately 40% of patients who have an myelomeningocele closed at birth will require surgical untethering for symptomatic tethered cord. A single surgical untethering is sufficient for long-term improvement and stabilization of symptoms in the majority of patients, but some patients with myelomeningocele suffer recurrent tethering. It is surprising that more patients do not have recurrences given the persistence of a placode that is not covered with pia and continued anatomical proximity of the dura and placode, despite adequate microsurgical untethering. Recurrent tethering is a very difficult problem to manage because each successive procedure finds increased scarring of neural tissue.

The conventional approach to recurrent tethering of the spinal cord is geared toward repeated efforts at microsurgical untethering, yet virtually all large clinical series of surgically treated children with myelodysplasia show a signifi-

Abbreviations used in this paper: CSF = cerebrospinal fluid; SCT = spinal cord transection.
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significantly higher incidence of neurological morbidity after repeated operations for untethering in children with retained neurological function. Success, as defined by long-term, pain-free rates, is also lower. The incidence of CSF leaks and other complications is also higher because the dysmorphic tissue holds sutures poorly and because of the abundance of scarring. For highly selected patients, we have found surgical transection of the distal spinal cord immediately above the placode to be a safe and highly effective approach for recurrent spinal cord tethering. This report summarizes a retrospective review of 14 patients thus treated over an 11-year period at the Children’s Hospital in Birmingham, Alabama.

Clinical Material and Methods

Operative Technique and Rationale

Patient candidates are identified in the course of routine care at a large multidisciplinary spina bifida clinic at the Children’s Hospital, Birmingham. Only patients with complete functional paraplegia are considered. When symptomatic retethering is recognized, an updated magnetic resonance image is obtained to visualize the distal placode. We agree with other authors who emphasize that spinal cord tethering is a clinical diagnosis recognized by progressive back pain and lower-extremity and sphincter dysfunction in the patient with myelodysplasia. Imaging is performed for surgical planning and to determine whether there is evidence of a syrinx. It is also essential to check meticulously for any evidence of a diastematomyelia or any other potentially tethering lesion at a higher level. Failure to recognize such a lesion could result in a failure to achieve untethering, despite a technically satisfactory surgical procedure. Sagittal images are particularly useful in planning the site of transection, which should be immediately superior to the placode. The distal cord is often tethered dorsally. The point of maximal adhesion of the spinal cord is identified and the level immediately rostral to it is targeted for transection. It is important that the transection be as low as possible so as to denervate as few of the intercostal muscles as possible. Children with myelodysplasia are frequently obese and as such have some degree of pulmonary compromise. Upward pressure on the diaphragm can compromise pulmonary capacity and compliance, so it is essential not to injure any functional intercostal muscles.

The procedure is performed after general anesthesia is induced in a latex-free environment. Patients are positioned prone, with bolsters under the iliac crests and superior chest. The previous myelomeningocele closure site is noted and an antiseptic skin preparation is performed. The incision is typically planned immediately rostral to the most rostral extent of the previous incision (or through the very top part of it if a generous incision was used for previous untethering). The tissue planes are normal at this level, so a conventional approach for laminectomy is used. After paraspinal muscle is retracted, a two-level laminectomy is performed with a high-speed drill or a rongeur. The normal dura is opened and the operating microscope is used. On several occasions we have observed the rostral extent of the suture line from previous untetherings at the very caudal extent of our exposure. Under the operating microscope, any arachnoidal adhesions are sharply dissected. The pia mater of the spinal cord is coagulated and sharply opened. Pial coagulation is gently performed with low bipolar settings to minimize contraction with resulting axial traction along an already tethered, compromised spinal cord. The cord is then directly divided with subpial aspiration and cut sharply with microscissors. A visible retraction of at least 1 cm is often evident between the two cut ends of the spinal cord. The dura is closed in a watertight fashion and the fascia, subcutaneous tissue, and the skin are closed in sequential watertight layers. The patient is kept flat for 2 to 5 days postoperatively and then permitted to ambulate ad libitum in a wheelchair.

Patient Population and Selection Criteria

We retrospectively identified 14 consecutive patients (seven male and seven female patients) with myelodysplasia and recurrent spinal cord tethering (13 having large lumbosacral decubiti and one having a CSF leakage) who underwent SCT between 1993 and 2004. Details of their clinical course and presentation are summarized in Table 1. Because the surgical division of the cord eliminates distal volitional neurological control, only patients with functionally complete paraplegia are considered candidates. All of the patients in our series met this stipulation. Three patients, however, had minimal movement in the lower extremities (for example, flicker movement of the great toe). These movements were so limited that they did not contribute significantly to the patient’s overall neurological capabilities and these patients were deemed functionally paraplegic. Twelve patients had experienced complete functional paraplegia since birth and two demonstrated later deterioration (from detethering surgery at an outside institution). The mean follow-up period was 5.5 years and ranged from 6 months to 11 years. Ages of the patients ranged from 8 to 22 years old (mean age 15.9 years). The most common clinical presentation for recurrent tethering was progressive and unremitting back pain, which occurred in 10 of the 14 patients. Patients were evaluated for other possible causes of back pain, such as spondylolisthesis, herniated disc, instability, and pyelonephritis. Although 12 patients demonstrated scoliosis, none was found to have any other spinal comorbidity to which their back pain could be reasonably attributed. Syringomyelia was also evident in three patients.

No patient had volitional control of bowel or bladder. Nine patients were on a catheterization and bowel program and five wore some form of diaper to maintain hygiene from spontaneous evacuation. Two patients had undergone a bowel or bladder diversion. Urodynamics were not routinely obtained in this group of patients. All patients continued to undergo close urological follow up as part of comprehensive care in the spina bifida clinic. No notable increase in the frequency of urological problems (reflux, urinary tract infections, and so forth) was observed following SCT.

All patients had undergone at least one prior surgical intervention for retethering. As mentioned earlier, two patients had become functionally paraplegic as a result of surgical untethering. Nine patients had previously undergone more than one untethering operation.

One patient underwent SCT when she presented with CSF emanating from massive, deep lumbar pressure sores that were infected. She was an obese, paraplegic teenager...
with myelodysplasia whose care needs simply exceeded the capability of her elderly parents. As a result, she was in-
frequently repositioned and large (15 × 20 × 4 cm) lumb-
osacral decubiti developed that became secondarily in-
fected. A general surgeon in the local community was
consulted and local debridement was undertaken in the of-
office. This procedure resulted in liberal drainage of CSF
from the depths of one of the largest decubiti. The girl un-
derwent SCT with oversewing of the distal thecal sac and
concomitant myofascial flaps by the plastic surgery service
as a successful means to control this problem.

An additional patient, one who is not included in this co-
hort of myelodysplastic patients, underwent SCT for pro-
gressive syringomyelia/syringobulbia from tethering relat-
ed to penetrating cervicothoracic spinal cord injury from a
gunshot wound.

Results

All patients underwent successful transection of the spi-
nal cord in a single operation. No significant intraopera-
tive complications occurred, and no patient required a
blood transfusion. Transient decreases in blood pressure
were occasionally observed on transection of the cord, but
all were minor and of short duration (1–3 minutes). De-
clines typically ranged from 10 to 20 mm Hg. No patient
required pressors or prolonged fluid resuscitation.

Patients were characteristically observed overnight in
the intensive care unit during the early part of our experi-
ence, but we now routinely send patients who have under-
gone SCT to the ward. No postoperative hypotension was
observed. No postoperative CSF leaks or positional
headaches were observed.

Neurologically, all patients retained the same functional
level that they demonstrated preoperatively. As mentioned
earlier, three patients had minimal although not functional-
ly significant movement in the lower extremities that was
lost with SCT. Similarly, two patients demonstrated a tem-
porary change in bladder behavior but did not change over-
all continence/hygiene capability. Three patients reported
transient periumbilical loss of sensation that was not pres-
ent preoperatively. No patient demonstrated progressive
postural problems associated with unopposed hip flexors.
None of these neurological changes adversely affected
these patients’ overall well being.

Every patient who had significant back pain prior to the
procedure experienced significant relief. Complete pain re-
relief was achieved in nine of 10 patients, and the remainder
had only intermittent discomfort, an improvement. No pa-
in this series has demonstrated any clinical evidence
for recurrence of tethering. Routine postoperative imaging
was not undertaken.

Nine patients had preoperative scoliosis that required in-
strumented fusion. No patient who underwent SCT dem-
strated any loss of neurological function associated with
instrumented fusion.

Discussion

Spinal cord transection in selected patients as a means of
achieving immediate and lasting therapeutic benefits has
been performed in preparation for vertebral osteotomy in
myelodysplastic patients,14 resection of tumors of the conus
medullaris,4 drainage of syringomyelia,2,12,13 treatment of in-
tractable pain in posttraumatic paraplegic patients,11 dysau-
tonomia in quadriplegic patients,10 and severe spasticity.1
McLaughlin, et al.,7 sectioned the conus medullaris in sev-
en patients with thoracolumbar myelomeningoceles who
presented with recurrent deformities of the lower extremi-
ties caused by persistent spasticity that was severe enough
to preclude sitting in a wheelchair and that interfered with
activities of daily living. All patients were free of spastic-
ty and had manageable lower extremities at a mean follow
up of 5.3 years. Two patients had improvement of bladder
continence. The precedent for cordectomy in the manage-
ment of spastic paraplegia was set by MacCarty and Kiefer6
in 1949. Indeed, in 1954 MacCarty7 stated that “spasticity
frequently induces such overwhelming situations [for ex-

### Table 1

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Follow Up</th>
<th>MM Level</th>
<th>Detetherings†</th>
<th>Symptoms/Signs</th>
<th>Resolved/Improved</th>
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<tbody>
<tr>
<td>1</td>
<td>10, F</td>
<td>6 mos</td>
<td>TL</td>
<td>2</td>
<td>back pain/scoliosis</td>
<td>yes</td>
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<td>2</td>
<td>15, M</td>
<td>10 yrs</td>
<td>TL</td>
<td>3</td>
<td>progressive scoliosis/preorthopedic fusion</td>
<td>yes</td>
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<tr>
<td>3</td>
<td>18, M</td>
<td>5 yrs</td>
<td>L</td>
<td>3</td>
<td>UE dysfunction/scoliosis/back pain</td>
<td>yes</td>
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<td>22, M</td>
<td>6 yrs</td>
<td>LS</td>
<td>1</td>
<td>UE dysfunction/back pain</td>
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</tr>
<tr>
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<td>22, M</td>
<td>6 yrs</td>
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<td>yes</td>
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<tr>
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<tr>
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<tr>
<td>8</td>
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<td>3</td>
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<tr>
<td>9</td>
<td>20, F</td>
<td>1 yr</td>
<td>LS</td>
<td>1</td>
<td>huge decubiti w/CSF leak</td>
<td>yes</td>
</tr>
<tr>
<td>10</td>
<td>15, M</td>
<td>9 yrs</td>
<td>LS</td>
<td>2</td>
<td>huge holocord syringomyelia w/progressive scoliosis</td>
<td>yes</td>
</tr>
<tr>
<td>11</td>
<td>16, F</td>
<td>6 mos</td>
<td>TL</td>
<td>1</td>
<td>progressive UE dysfunction</td>
<td>yes</td>
</tr>
<tr>
<td>12</td>
<td>15, F</td>
<td>4 yrs</td>
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<td>2</td>
<td>back pain</td>
<td>yes</td>
</tr>
<tr>
<td>13</td>
<td>8, F</td>
<td>3 yrs</td>
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<td>1</td>
<td>holocord syringomyelia w/progressive scoliosis</td>
<td>yes</td>
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<tr>
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<td>2</td>
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</tr>
</tbody>
</table>

† Number of past surgical detetherings (not including current SCT).

* L = lumbar; LS = lumbosacral; TL = thoracolumbar; TLS = thoracolumbosacral; UE = upper extremity.
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ample, urological complications or bedsores] that death may result [so that] it becomes necessary to produce flaccidity in the lower extremities and bladder so that these structures can be adequately cared for.” Ragnarsson and colleagues sectioned the cord in a patient with complete T-5 paraplegia who presented with a tethered cord manifesting as chronic progressive cervical myelopathy. The procedure resulted in immediate improvement in intraoperative, somatosensory evoked potentials and marked postoperative clinical improvement. Durward, et al., have reported that neurogenic leg pain was not relieved in their patients undergoing selective spinal cordectomy.

A few of our patients exhibited intraoperative hypotension following transection of their cord. This occurrence was followed without intervention and was noted to reverse itself prior to extubation. Winston, et al., have reported acute elevation of intracranial pressure in patients with myelomeningocele following SCT performed at T-12 in preparation for vertebral osteotomy. Cardiac arrest occurred at 1 and at 18 hours following SCT in these patients. These authors speculate that an unrecognized CSF shunt that malfunctioned was unable to accommodate CSF production once the distal subarachnoid space was ligated.

Egress of CSF from infected decubiti as seen in one of our patients is a difficult and potentially dangerous problem. Paraplegic patients with myelodysplasia are at high risk for decubiti due to sensory loss and paraplegia. Obesity is a separate, although frequently concomitant, risk factor. Any (ill-advised) attempts at local debridement by inexperienced providers may result in CSF leaks from the depths of infected decubiti. Because the tissue planes are distorted, the tissue is dysmorphic and closure is exceptionally difficult or impossible. We found in one patient that SCT through the most rostral normal planes allowed good control of the dural sleeve, with resultant cessation of CSF egress from the wound.

Despite the favorable results of SCT, controversy surrounds the procedure for several reasons. Dividing the spinal cord is an aesthetically difficult and counterintuitive endeavor for neurosurgeons, who customarily go to great lengths to protect this delicate structure. Some may harbor speculative concerns may have some legitimacy, our collected experience demonstrates without question that pain is uniformly improved, complications are minimal, and recurrent tethering does not appear to occur at long-term follow-up after SCT.

References