Between 15 and 25% of patients who undergo perinatal repair of myelomeningoceles will develop neurological symptoms related to spinal cord tethering, or STCS. Spinal cord tethering results in neuronal, metabolic, and vascular derangements, which can manifest clinically as pain, paresthesia, weakness, spasticity, or bladder and bowel dysfunction. Studies of the natural history would suggest that these symptoms are progressive without intervention. Surgical untethering is primarily aimed to prevent further neurological deterioration, and in some cases may allow for functional improvement. Untethering of the spinal cord after a previous myelomeningocele repair is, however, challenging because the structural and functional anatomy is distorted due to both developmental anomalies and scar from previous surgery. The placode and functional nerve roots can be difficult to identify and untether autonomously.

Electrophysiologically guided untethering of secondary tethered spinal cord syndrome

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Object. Many patients develop neurological symptoms related to spinal cord tethering after perinatal repair of myelomeningocele. This is referred to as secondary tethered cord syndrome (STCS). The authors describe their methodology and evaluate the intraoperative utility and postoperative outcomes of electrophysiologically guided untethering for STCS. In addition, the authors describe the use of electrophysiological guidance to identify an “autonomous placode” in the untethering of the cord in STCS.

Methods. The authors retrospectively identified 46 untethering procedures in 38 patients who had undergone perinatal myelomeningocele repair and in whom the index surgery was for tethered cord release at the site of the repair. In all cases, both passive (electromyography) and active (detection of compound muscle action potentials) electrophysiological monitoring was used. The proximity to neural elements was determined based on the current used; eliciting compound muscle action potentials with a ≤ 10-mA stimulation was assumed to represent direct neuronal stimulation. Clinical records were reviewed to evaluate the utility of electrophysiological guidance and patient outcomes.

Results. The median age at the time of untethering was 9.5 years (range 0.5–54 years). The median follow-up time was 42 months (range 3–172 months). Progressive bowel and bladder dysfunction, diagnosed either clinically or by cystometrogram, and low-back pain were the most common presenting symptoms. Intraoperative findings indicated that the most common causes of tethering were dense scar (76%) and a tethered placode (39%). Electrophysiological monitoring identified functional neural tissue near tethered elements and provided intraoperative guidance in all cases. In 41% of cases (19 cases), the untethering plan was noted to have been significantly influenced by intraoperative neurophysiological findings. Moreover, an autonomous placode was identified in 6 patients who were nonambulatory preoperatively and had presented with increasing pain and spasticity. In electrophysiologically silent areas, more aggressive dissection and untethering were possible. Symptoms of low-back pain, lower-extremity paresthesia, and lower-extremity spasticity were most likely to improve after untethering surgery (91, 88, and 82%, respectively). Sectioning above the electrophysiologically defined autonomous placode resulted in significant improvement in back pain and lower-extremity spasticity in 5 of 6 patients. There was 1 case of immediate postoperative neurological deterioration (fetal incontinence). All patients remained clinically stable or improved on long-term follow-up, except for 6 (16% of patients) who required a total of 7 additional procedures for recurrent symptoms (median time to repeat surgery 36 months). Complications were noted in 8 cases, including infections and CSF leaks.

Conclusions. Surgical untethering of STCS halts progression and often improves preoperative symptoms. Electrophysiological monitoring, using both a threshold-based interpretation system and continuous electromyography monitoring, provides an efficient, effective, and reliable method for intraoperative guidance, thereby limiting iatrogenic injury and providing a means to identify and untether autonomous placodes. Electrophysiological monitoring also allows for more aggressive dissection and untethering in functionally silent regions, possibly decreasing retethering rates. (DOI: 10.3171/2010.3.FOCUS09299)

Key Words • tethered spinal cord • secondary tethered cord syndrome • spinal cord untethering • intraoperative monitoring • myelomeningocele

Abbreviations used in this paper: CMAP = compound muscle action potential; CMG = cystometrogram; EMG = electromyography; STCS = secondary tethered cord syndrome.
to distinguish from scar, can be intimately involved with scar, or can be tethered themselves, placing the patient at significant risk of neurological deterioration from the untethering procedure. Measures are therefore necessary to minimize the risk of iatrogenic injury during untethering procedures.\textsuperscript{3,8,12,19,20}

Previous reports, including a recent one by Al-Holou and colleagues,\textsuperscript{1} have described the benefits of surgical untethering in this patient population, clearly documenting the benefits of intervention.\textsuperscript{7} In this study, we again assess outcomes of untethering in this population. However, we focus on the role and contribution of electrophysiological guidance for surgical untethering. We have developed a systematic approach of using intraoperative neurophysiological mapping to guide repeat operations for spinal cord tethering in patients with myelomeningocele. We use neurophysiological monitoring intraoperatively to define the functionality of nerve roots and the placode and to guide surgical untethering. This allows determination of the proximity of functional neural elements for preservation, more aggressive untethering in electrophysiologically silent areas, and identification of and transection above autonomous placiodes, which could not otherwise be done. We describe our experience using this technique, including an assessment of intraoperative utility and patient outcomes.

Methods

Patient Selection

We retrospectively reviewed all surgeries for spinal cord untethering over a 13-year period (1992–2005), identifying 101 consecutive surgeries performed using neurophysiological guidance. Patients were included in the current analysis if they met the following criteria: 1) they had undergone perinatal myelomeningocele repair; and 2) the index surgery was for release of tethered cord at the site of myelomeningocele repair. We identified 46 untethering procedures in 38 patients meeting these criteria (6 patients underwent 2 surgeries and 1 patient underwent 3 surgeries).

Patients were selected for surgery if they presented to neurosurgical attention with signs and symptoms consistent with spinal cord tethering (as described in Table 1 and the Results section) and MR imaging demonstrated radiographic evidence of tethering. All patients underwent preoperative EMG and CMG evaluation.

Operative Technique (Including Electrophysiological Mapping)

In all cases, we used both passive and active monitoring. Free-run EMG was used throughout each surgery to detect spontaneous discharges from efferent nerves due to intraoperative manipulation or stretch of functional neural elements. Active monitoring consisted of stimulating suspected functional tissue by using a bipolar electrode (5-mm interelectrode distance) and detecting CMAPs peripherally. Recording sites were selected based on preoperative EMG examination, preoperative clinical examination, and either the nerve roots in question or those at risk during surgery. Typical muscles recorded included biceps femoris, anterior tibialis, gastrocnemius, and anal sphincter. As reported previously, we use a threshold approach to determine the proximity to viable neural elements. The CMAPs elicited with $\leq 10$ mA were interpreted as being a direct stimulation of the nerve.\textsuperscript{15} The CMAPs elicited with 11–25 mA were interpreted as being near a functional nerve, probably with intervening tissue. If currents $> 25$ mA are needed to elicit a CMAP, we consider functional tissue to be remote and to be activated via spread through adjacent tissue. All surgical untetherings were performed by the senior author (J.A.J. Sr.). All intraoperative neurophysiological monitoring was performed by a neurologist specializing in neuromonitoring (L.H.P).

Outcomes Assessment

Patient charts, including operative reports, were reviewed in detail to assess the utility of neurophysiological guidance for surgical untethering. All patients were clinically evaluated immediately postoperatively and 3 months after surgery. Long-term follow-up was also obtained to determine if patients had stabilization or progression of symptoms after surgery. In addition, all patients underwent follow-up CMG evaluation to identify subtle changes in bladder function.

Results

Patient Profile

Thirty-eight patients underwent 46 untethering procedures for STCS (6 patients underwent 2 surgeries and 1 patient underwent 3 surgeries). Eighteen of the patients were male. The median age at the time of surgery was 9.5 years (range 6 months–54 years), representing a similar age distribution to that reported by Herman and colleagues\textsuperscript{7} (Fig. 1). The median follow-up was 42 months (range 3–172 months). Eighteen patients had < 3 years of follow-up.

Most patients presented with multiple symptoms, as is common in tethered cord syndrome. Bowel and bladder dysfunction was the primary presenting symptom in nearly two-thirds of cases. Low-back pain was the second most frequent presenting symptom; it was found in 50% of cases. Presenting symptoms and corresponding frequencies are fully detailed in Table 1.

Operative Findings and Utility of Mapping

Electrophysiological monitoring identified functional neural tissue near tethered elements and provided intraoperative guidance in all cases. In 41% of cases (19 cases),

<table>
<thead>
<tr>
<th>Presenting Symptoms</th>
<th>No. (% of 46 cases)</th>
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<tbody>
<tr>
<td>bladder/bowel dysfunction</td>
<td>30 (65)</td>
</tr>
<tr>
<td>low-back pain</td>
<td>23 (50)</td>
</tr>
<tr>
<td>lower-extremity weakness</td>
<td>20 (43)</td>
</tr>
<tr>
<td>lower-extremity spasticity</td>
<td>11 (24)</td>
</tr>
<tr>
<td>paresthesia</td>
<td>8 (17)</td>
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</table>
Electrophysiological untethering of secondary tethered cord

The untethering procedure was significantly influenced by intraoperative neurophysiological findings. For example, in 1 case, the surgeons noted that “further untethering may have caused injury” based on intraoperative neurophysiological findings. Conversely, more aggressive dissection and untethering were possible in electrophysiologically silent areas; the absence of identifiably functional tissue gave the primary surgeon the confidence to aggressively dissect and untether areas infiltrated with dense scar.

Multiple factors, even within an individual patient, can contribute to tethering of the spinal cord in STCS. In this series, the most common cause of tethering was dense scar (35 cases, 76%). A tethered placode was the next most common cause of cord tethering in this series (18 cases, 39%). The causes of tethering and relative frequencies are detailed in Table 2.

Autonomous Placode

We have previously described the concept of an autonomous placode. It is defined as a placode which, when stimulated at its caudal extent with low currents (≤10 mA), results in marked lower-extremity CMAPs, but it produces no lower-extremity CMAPs when stimulated in a slightly more rostral position, signifying an intact efferent pathway that is completely disconnected from the functioning (and more rostral) spinal cord. Because of the functional disconnection from the normal spinal cord and because the placode is often a source of tethering (Table 2), identification of an autonomous placode and the functionally silent area between this placode and the normal spinal cord can be critical for successful untethering. Because this is a functionally defined entity, the autonomous placode cannot be identified visually and requires intraoperative neurophysiological monitoring for localization and confirmation.

An autonomous placode was identified in 6 patients, all of whom were nonambulatory preoperatively and had presented with increasing back pain and spasticity. In all of these patients, the functionally silent area superior to the autonomous placode and inferior to the normal spinal cord was sectioned. Five of the 6 patients experienced significant improvement in back pain and lower-extremity spasticity as a result of sectioning.

Symptomatic Outcomes

The goal of surgical intervention is preventing clinical progression. With one exception (discussed below), all symptoms were either stable or improved in all patients at the 3-month follow-up. Symptoms of low-back pain, lower-extremity paresthesia, and lower-extremity spasticity were most likely to improve after untethering surgery (91, 88, and 82%, respectively). Symptomatic outcomes at 3 months are detailed in Table 3. There was a single case of immediate postoperative neurological deterioration (fetal incontinence), deemed a surgical complication, which had not improved at follow-up. Long-term follow-up revealed stable clinical status in all patients relative to 3-month postoperative assessments.

A secondary measure of the efficacy of untethering is an evaluation of rates of repeat operation for persistent symptomatology. With a median follow-up time of 42 months, all patients experienced lasting clinical stability or improvement, except for 6 patients (16%) who required a total of 7 additional procedures for recurrent symptoms (similar to the original presentation), with a median time to repeat surgery of 36 months (range 3–119 months). The patient who underwent 2 additional untethering procedures had these interventions 32 and 36 months apart. Interestingly, in the patient who underwent early repeat surgery (at 3 months) an autonomous placode was identified during the first surgery, but the placode was not sectioned due to a question of persistent sensory function in the lower extremities. After repeat surgery and sectioning above the autonomous placode, the patient experienced significant improvement in her spasticity symptoms.

Surgical Complications

As noted above, neurological worsening was noted

<table>
<thead>
<tr>
<th>TABLE 2: Tethering elements in 38 patients who underwent 46 procedures for spinal cord untethering</th>
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<tbody>
<tr>
<td>Tethering Element</td>
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<tr>
<td>---------------------------------------------------------------</td>
</tr>
<tr>
<td>scar</td>
</tr>
<tr>
<td>placode</td>
</tr>
<tr>
<td>filum</td>
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<tr>
<td>lipomatous tissue</td>
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<td>nerve roots</td>
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<tr>
<th>TABLE 3: Symptomatic outcomes at 3 months in 38 patients who underwent 46 procedures for spinal cord untethering</th>
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<tr>
<td>Symptom</td>
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<tr>
<td>bladder/bowel dysfunction*</td>
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<tr>
<td>low-back pain</td>
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<td>lower-extremity spasticity</td>
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<td>paresthesia</td>
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* Based on clinical or CMG evaluation.
in 1 patient, who had fecal incontinence after surgery. In addition, there were 8 cases of operative complications, including 2 infections (4%), 4 CSF leaks (9%), and 2 combined infection and CSF leaks (4%), all requiring operative intervention.

Discussion

Regardless of the cause of the condition, the goal of tethered cord surgery is to alleviate the neuronal, metabolic, and vascular derangements induced by spinal cord stretch.24 In doing so, we hope to thwart further neurological deterioration, to reverse current neurological deficits, to avoid iatrogenic injury, and to prevent retethering. Achieving these goals requires aggressive and maximal untethering, which is complicated in STCS by prior repair of spinal dysraphism and disrupted anatomy of the caudal spine. Although others have reported the use of neurophysiological monitoring for tethered cord release, this is the largest series specifically analyzing the utility and outcomes of this technique in STCS.1,13,14,17,22

Secondary Tethered Cord Syndrome

Although radiographically demonstrated spinal cord tethering occurs in nearly all patients after repair of spinal dysraphisms, not all patients are symptomatic. The use of CMG diagnostics can be particularly helpful in identifying patients who are symptomatic from secondary tethered cord, and can be highly useful for operative decision making.4 Although patients may present at any age, the most common age at presentation is between 5 and 9 years, concurrent with the period of rapid growth.1,7 Because children may not always recognize bladder dysfunction, a CMG evaluation should be included in the standard assessment of STCS. It is imperative that symptomatic secondary tethered cords are recognized early because delayed recognition is associated with irreversible injury.16

Neurophysiological Monitoring for Cord Untethering

Multimodality neurophysiological monitoring for tethered cord release has been reported previously. For example, von Koch and colleagues22 reported using neurophysiological monitoring for tethered cord release in 25 pediatric patients with a thickened filum and low-lying conus. Consistent with this report and as might be expected, they state that functional tissue can easily be differentiated from thickened filum based on stimulation thresholds, the latter requiring thresholds of nearly 100 times that of functional tissue. The same group reported similar results and utility of neurophysiological monitoring for tethered cord release in 15 adults with a diverse number of causes of tethered cord, including thick filum, spinal cord malformations, and low-lying conus.17 Paradiso and colleagues41 have also reported success with identifying viable tissue intraoperatively based on stimulation thresholds. Although the pathophysiological mechanisms of neurological deterioration are probably similar across causes for tethered cord syndromes, the techniques used for untethering cannot necessarily be extrapolated to patients with STCSs, in which the filum may not be readily visually identifiable, and in which little normal comparative anatomy is available.

In this series, a threshold approach to interpreting intraoperative stimulation provided a rapid means of assessing the in situ functional architecture of the secondary tethered cord. Neurophysiological monitoring in our series was effective in minimizing iatrogenic injury. In all cases, functional tissue was identified near the site of tethering. Moreover, 41% of cases were associated with an altered plan for untethering based on intraoperative findings. Whereas neurological deterioration has been reported in up to 9% of cases in other series of secondary tethered cord surgery regardless of the use of neurophysiological monitoring, neurological worsening in our series was limited to 1 surgery (2.2%), a rate similar to that reported in other series in which neurophysiological monitoring was used.9,17 Having this rapid means of assessment is essential, considering that the two most common tethering elements are scar and placode, entities that are better defined functionally than visually.

Posttreatment Outcomes

Although the goal of spinal cord untethering is primarily to halt clinical progression, the majority of patients experience significant improvement, especially with respect to pain, paresthesia, and spasticity (see Table 3). Overall, approximately 75–90% of patients experience improvement or stabilization of symptoms.1,7,9,12,18 Pain is most amenable to improvement; Al-Holou and colleagues1 recently reported improvement in pain in 75% of patients.9,10,12,20 Bowel and bladder function, on the other hand, are less likely to improve, with only 13–46% of patients reporting improved function (20% in this series).1,7,17,22 In addition, we suspect that untethering contributed to the stabilization of bowel and bladder function in the other patients who presented with these symptoms.

In addition to neurological improvement, one of the goals of cord untethering is to prevent retethering or a repeat operation. Retethering requiring a repeat operation is estimated to occur in 20–30% of all patients, and in up to 50% of pediatric patients.2,6,18 Techniques have been described to prevent retethering, including the use of intradural retention sutures and placement of implants (or dural substitutes) to reconstruct and maintain the intradural space.21,25 We had to perform 7 repeat operations, one of which was needed due to incomplete untethering. Excluding this case, the rate of retethering requiring repeat operation was 15.4%, which is favorable relative to previous reports. Considering that the median time to the repeat untethering in this series was 36 months, and that untethering procedures have been reported in the literature several years after the index surgery, the reported retethering rate may be artificially low due to the number of patients in this series in whom the follow-up was less than 3 years.18 Alternatively, more aggressive untethering with neurophysiological monitoring may have contributed to a decreased retethering rate.

Placode Transection

Tethering at the level of the placode is a common
Electrophysiological untethering of secondary tethered cord

finding and has been postulated to be due in part to the absence of a normal pial plane, and to the unavoidable contact of the placode with the reconstructed canal.\(^5\)\(^,\)\(^6\)\(^,\)\(^11\)\(^,\)\(^23\) As we have previously noted, and report in additional cases in this paper, transection above the level of an autonomous placode is an efficacious treatment for paraplegic patients (for whom there is no risk of neurological deterioration) presenting with progressive spasticity. Although with some variation (for example, transection immediately rostral to the point of maximal placode adhesion), similar placode transection approaches have been previously described, with excellent results: nearly 100% pain and spasticity relief.\(^2\)\(^,\)\(^6\)\(^,\)\(^11\)\(^,\)\(^23\) Although it has been suggested that transection might be reserved for those with recurrent retethering,\(^2\) because of its efficacy (83% improvement), we advocate consideration of transection above the level of the autonomous placode at the time of initial untethering. Importantly, because the “autonomous placode” is functionally defined, it necessitates neurophysiological monitoring for identification.

**Limitations of Monitoring**

Although we believe that neurophysiological monitoring is critical to preserve function and to maximize untethering in STCS, we recognize the limitations. First, the success of motor root mapping is limited by the selection and placement of electrodes, which relies on a thorough preoperative clinical and electromyographic examination as well as a clear operative plan that recognizes which spinal levels may be at risk during surgery. Second, passive EMG monitoring for stretch-induced discharges has limited sensitivity; nerves may be stretched or injured without spontaneous discharges. It is therefore imperative not only to rely on passive monitoring, but also to use active mapping with a threshold-based interpretation system, providing a reliable and rapid assessment of functional architecture. Third, electrodiagnostic testing during surgery probably increases operative and anesthetic times. We believe that the time increase is modest when the test is routinely performed by experienced neurophysiologists and technicians, and that the benefits (for example, decreased iatrogenic injury) justify the potentially increased time of surgery and anesthesia.

This study is also limited by its design, being based on an institutional, retrospective, uncontrolled series. To determine whether this technique improves outcomes would technically require a prospective randomized trial. It is unlikely that this could be accomplished. We believe that intraoperative electrophysiological monitoring provides the surgeon with information that is otherwise unattainable, thereby making the surgery safer (preserving nerves) and more efficacious (identifying autonomous placodes).

**Conclusions**

Surgical untethering of secondary tethered cord syndrome is effective at halting symptomatic progression of bowel and bladder incontinence and weakness, and it improves preoperative symptoms of pain, spasticity, and paresthesia. Intraoperative neurophysiological monitoring, using both passive and active mapping and a threshold-based system for interpreting maps, provides an efficient, effective, and reliable method for intraoperative guidance, thereby limiting iatrogenic injury during untethering of STCS, and providing a means to identify autonomous placodes that can be detached from the tethered spinal cord. Moreover, using electrophysiological monitoring allows for more aggressive dissection and untethering in functionally silent regions, hopefully resulting in a decreased rate of retethering and repeat operation.

**Disclosure**

Dr. Elias receives an honorarium from the Focused Ultrasound Surgery Foundation.

Author contributions to the study and manuscript preparation include the following. Conception and design: Pouratian. Acquisition of data: Elias. Analysis and interpretation of data: Pouratian, Elias. Drafting the article: Pouratian. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: all authors. Statistical analysis: Pouratian. Study supervision: Elias, JA Jane Jr, JA Jane Sr.

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