Adult tethered cord syndrome in patients with postrepair myelomeningocele: an evidence-based outcome study

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Object. As patients with myelomeningocele age, tethered cord syndrome (TCS) in adults with postrepair myelomeningocele has become more common. The authors have structured an evidence-based review of the literature for the purpose of addressing the following questions. 1) How is the diagnosis made? 2) What is the natural history that occurs in adults with postrepair myelomeningocele and TCS? 3) What are the criteria for operative intervention? 4) What is the functional outcome for patients with postrepair myelomeningocele?

Methods. A computerized search of the National Library of Medicine of the English-language literature published from 1966 to 2003 was performed. Articles pertaining to the clinical aspects and management of postrepair myelomeningocele in adults, TCS in adults resulting from closed defects, and pediatric patients with postrepair myelomeningocele were reviewed.

No Level 1 or 2 data were located; however, the search yielded Level 3 and 4 evidence in the literature in which the clinical syndrome, underlying pathology, and the imaging and electrophysiological evaluation for TCS in adults are discussed.

Conclusions. Analysis of the available data indicates the following. 1) A lower lesion level predisposes patients to symptomatic tethering; moreover, orthopedic and urological deterioration will occur in the majority of these patients. 2) Tethered cord release should be considered for adult patients with postrepair myelomeningocele when clinical symptoms, imaging studies, urodynamics, and somatosensory evoked potentials are consistent with TCS. 3) Prompt, aggressive untethering surgery within 5 years of symptom onset, along with long-term follow up to check for delayed retethering, is recommended. The overall outcome for patients with postrepair myelomeningocele may not be as good as the outcome for adults with closed dysraphism.

KEY WORDS • adult • tethered cord • myelomeningocele • evidence-based analysis • pediatric neurosurgery

The tethered spinal cord in adult patients was initially recognized more than 100 years ago. Despite this early observation, investigators have focused on the effects of tethering in the pediatric populations with spinal dysraphism. As many as two thirds of all procedures for effects of tethering in the pediatric populations with spinal dysraphism. As many as two thirds of all procedures for effects of tethering in the pediatric populations with spinal dysraphism have been the subject of several reviews, split cord malformations, thickened terminal filum, spinal lipomas, meningoceles. The clinical presentation and functional patients with postrepair myelomeningocele. Adult tethering in adults, TCS in adults resulting from closed defects and pediatric patients with postrepair myelomeningocele in adults, TCS in adults resulting from closed defects and pediatric patients with postrepair myelomeningocele have, been poorly described. With many more healthy and functional patients with postrepair myelomeningocele now entering adulthood, treatment and research guidelines need to be firmly established.

Evidence-based outcome in adult TCS

Adult Tethered Cord Syndrome. As patients with myelomeningocele age, tethered cord syndrome (TCS) in adults with postrepair myelomeningocele has become more common. The authors have structured an evidence-based review of the literature for the purpose of addressing the following questions. 1) How is the diagnosis made? 2) What is the natural history that occurs in adults with postrepair myelomeningocele and TCS? 3) What are the criteria for operative intervention? 4) What is the functional outcome for patients with postrepair myelomeningocele?

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The symptoms of adult TCS include progressive difficulty with ambulation, gait disturbance, back pain, nondermatomal sensory changes, lower-extremity weakness, sensory deficits, and bowel and bladder dysfunction. The symptoms of adult TCS worsen the condition of patients with this lesion.

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Clinical Material and Methods

A computerized search of the National Library of Medicine was performed to locate literature published between 1966 and 2003. The search was limited to reports available in the English language. The medical subject headings “tethered spinal cord,” “tethered cord,” and “myelomeningocele” yielded 3157 citations. These citations were retrieved when used in combination with “adult” yielded 41 citations. The abstracts were reviewed for those articles in which functional or surgical outcome for tethered cord in adult patients with myelomeningocele was described. Articles in which the clinical aspects and management of adult patients with postrepair myelomeningocele or tethered cord resulting from closed defects and pediatric patients with tethering and postrepair myelomeningocele were used to generate guidelines. Because of the limited number of articles, case reports were included as long as they met the basic inclusion criteria. Moreover, the bibliographies of the selected articles were reviewed to identify additional articles.

Evidence-based outcome in adult TCS

Conclusions. Analysis of the available data indicates the following. 1) A lower lesion level predisposes patients to symptomatic tethering; moreover, orthopedic and urological deterioration will occur in the majority of these patients. 2) Tethered cord release should be considered for adult patients with postrepair myelomeningocele when clinical symptoms, imaging studies, urodynamics, and somatosensory evoked potentials are consistent with TCS. 3) Prompt, aggressive untethering surgery within 5 years of symptom onset, along with long-term follow up to check for delayed retethering, is recommended. The overall outcome for patients with postrepair myelomeningocele may not be as good as the outcome for adults with closed dysraphism.

Key Words • adult • tethered cord • myelomeningocele • evidence-based analysis • pediatric neurosurgery

Abbreviations used in this paper: CSF = cerebrospinal fluid; CT = computed tomography; MR = magnetic resonance; mRSS = modified Rankin Scale; SSEP = somatosensory evoked potential; TCS = tethered cord syndrome.

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to the topic but did not meet the inclusion criteria, which were based on guidelines provided by the Oxford Centre for Evidence-Based Medicine (Tables 1 and 2). These reports, which include case series of adult tethered cord syndrome and review articles, are discussed in the Scientific Foundation section and are cited in the References. Summaries of the seven articles that meet inclusion criteria are provided in Table 3.

Results

Scientific Foundation

Adult Tethered Cord Syndrome. Articles concerning adults with tethered cord have been focused primarily on the clinical presentation and surgical outcome in patients with closed dysraphic lesions and have helped define the adult TCS.

The types of dysraphic defects include fatty and thickened terminal filament, spinal lipomas, meningoceles, split cord malformations, and postrepair myelomeningocele. Dysraphic lesions cause symptoms by attaching the spinal cord to a fixed vertebral structure. The spinal cord is stretched and cord motion is decreased, leading to impaired cellular metabolism and neuronal function. Particularly in adults, associated conditions may lead to spinal cord dysfunction when tethering is present. Degenerative spondylosis and spondylolisthesis, hydrocephalus, and thickened terminal filum, spinal lipomas, meningoceles, split cord malformations, and postrepair myelomeningocele can mimic TCS and/or worsen the condition of patients with this lesion.

The symptoms of adult TCS include progressive diffuse back pain, nondermatomal leg pain, pain in the anorectal region, lower-extremity weakness, sensory deficits, and bladder and bowel dysfunction in the setting of a low-lying spinal cord lesion (that is, below L-2). The precipitous onset of symptoms following lithotomy or flexion positioning, exercise, heavy lifting, childbirth, intercourse, and spine trauma has been reported in 60 to 90% of adult patients with tethered cords. Clinical symptoms are generally similar in adult patients with postrepair myelomeningocele or closed dysraphic defects. In two articles, however, divergent symptoms are described. Filler, et al., reported on two patients with postrepair myelomeningocele who presented with motor dysfunction as the main symptom and no pain. Oi, et al., described three young adults with postrepair myelomeningocele in whom pain was the common presenting symptom. Preexisting stable childhood musculoskeletal, urological, or neurological deficits were found in 44% of patients.

Diagnosis. Magnetic resonance imaging has become the gold standard for diagnosing dysraphic anomalies, determining the vertebral level at which they occur, and ruling out tandem or unsuspected lesions. In the evaluation of impaired cord motion, dynamic MR imaging may have an impact on determining tethering outcomes. Johnson and Levy evaluated two groups of children with tethered cords in which MR imaging was used to assess spinal cord motion. The first group comprised 17 asymptomatic children with normal or slightly decreased motion in whom surgery had not been performed. The second group consisted of 15 symptomatic patients with decreased cord motion in whom the cord was un tethered. None of the patients who did not undergo surgery became symptomatic. Children with poorer outcomes included those who were 10 years of age or older, those patients with postrepair myelomeningocele, and those in whom markedly decreased cord motion was demonstrated on MR imaging. No dynamic MR imaging studies that were performed specifically in adults have been reported.

Plain radiographs and CT scans are both useful in evaluating the degree of abnormal spinal curvature as well as vertebral segment anomalies. Myelography combined with CT scanning can provide neuroimaging in cases in which spinal instrumentation interferes with MR imaging or unusual cystic structures require better definition. The role of ultrasonography has been established in children, but remains unclear in adults.

In patients with TCS, electrophysiological evaluation of lower-extremity function with the aid of SSEP monitoring may be helpful in establishing the diagnosis and predicting outcome. There are currently no studies available in which adult patients only are prospectively evaluated. Two studies prospectively evaluated SSEPs in both children and adults with TCS of various origins. Roy, et al., evaluated 22 consecutive patients with asymptomatic TCS; the patients were divided into two groups based on myelographic presence or absence of a dysraphic lesion. In the group of patients in whom no lesion was detected by radiography, posterior tibial nerve SSEPs were within normal limits, thus indicating normal physiological function. In the second group, posterior tibial nerve SSEPs demonstrated a significant correlation between the severity of clinical findings and abnormal evoked potentials. Postoperatively, in eight patients SSEPs reflected improved function in relation to the level and type of dysraphic lesion present.

In a series of 29 children and nine adults, Kale and Mahapatra reported that abnormal SSEPs were evident in 72% of the children and 44% of the adults. Postoperatively, improved SSEPs were demonstrated in 22 children and four adults, and clinical improvement was observed in all of the children and two of the four adults at the 3-month follow-up examination. Improvement in SSEPs preceded clinical improvement in 60% of patients.

The Natural History of TCS. Knowledge of the natural history of TCS in patients with postrepair myelomeningocele, which might help predict the chance for delayed de-

TABLE 1

Levels of evidence

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<thead>
<tr>
<th>Level</th>
<th>Recommended Evidence</th>
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<tr>
<td>1a</td>
<td>obtained from metaanalysis of randomized controlled trials</td>
</tr>
<tr>
<td>1b</td>
<td>obtained from ≥1 randomized controlled trial</td>
</tr>
<tr>
<td>2a</td>
<td>obtained from ≥1 well-designed controlled study without randomization</td>
</tr>
<tr>
<td>2b</td>
<td>obtained from ≥1 type of well-designed quasi-experimental study</td>
</tr>
<tr>
<td>3</td>
<td>obtained from well-designed nonexperimental descriptive studies, such as comparative studies, correlation studies, &amp; case studies</td>
</tr>
<tr>
<td>4</td>
<td>obtained from expert committee reports or opinions &amp;/or clinical experiences of respected authorities</td>
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</table>

* Adapted from the Oxford Centre for Evidence-Based Medicine Levels of Evidence.
terioration or identify mitigating factors. Phuong, et al.,33 outlined a timeframe for the progression of the symptoms and signs of tethering. This series represents Level 2b evidence (see Table 2). These authors retrospectively reviewed data regarding 45 patients with postrepair myelomeningocele and defined a cohort of patients with symptoms of bladder dysfunction or orthopedic foot deformity who were followed for a mean of 12.2 years. Overall, 40 (88.9%) of 45 patients required orthopedic or urological procedures due to further symptoms. The incidence of progression was 27.5% at 1 year, 40% at 2 years, and 60% at 5 years. Patients with the more common symptoms of tethering, such as pain and/or lower-extremity weakness, were excluded; therefore, only the natural progression of urological and distal orthopedic symptoms can be inferred.

Oi, et al.,30 correlated the MRI imaging–documented vertebral level of the conus medullaris to the onset of late neurological deterioration in pediatric and adult patients with postrepair myelomeningocele. Of the 26 total patients with the conus medullaris level at L2–S3, only those patients in whom the conus level was S-1 or below exhibited the delayed neurological symptoms of tethering at follow-up. Specifically, eight (50%) of the 16 patients in whom the conus was observed to be at S-1 or below developed symptoms. This Level 2b evidence indicates that only a very low level of the conus medullaris predisposes the patient with postrepair myelomeningocele to symptomatic tethering.

Clinical Outcome in Adults With Closed Defects

In retrospective case series adults with closed dysraphic defects were evaluated (constituting Level 3 data). Surgical untethering was generally advocated. In several articles it is recommended that surgery be performed promptly—within 5 years of symptom onset.15 In another study, however, Gupta, et al.,11 failed to demonstrate differences in postsurgical outcomes when comparing adult patients who were asymptomatic during childhood with those with those who had suffered long-term preexisting static skeletal or neurological deficits since childhood.

An aggressive surgical strategy of untethering and repairing the lesion has been advocated. Moreover, the laser can provide a useful adjunct in lipoma surgery. According to Huttman, et al.,17 incomplete surgery or untethering can result in symptoms developed 5 years after surgery; symptoms recurred in only 16% of patients in whom complete untethering was achieved. Furthermore, decreased operation stabilized symptoms in 70% of patients.27

Postoperative improvement varied based on the type of presenting symptomatology. Among the 275 patients cumulatively described in multiple case studies, the greatest improvement in symptoms was pain reduction, which occurred in up to 86% of patients. Sensorimotor deficits improved in 35 to 71%, whereas bladder and bowel dysfunction, a category in which poor outcome was consistently demonstrated, improved in only 16 to 60% of patients. In one series musculoskeletal deformities, which are typically not associated with adult patients, were reported to have improved after untethering.44

Urological improvement was consistently minimal in all series, although the worsening of symptoms was uncommon.1,2,6,13,17,25,35,44,54 The types of urological dysfunction were urgency and urge incontinence, urinary tract infection, and voiding difficulties. Urodynamical testing in patients with TCS revealed hyperreflexia, external detrusor–sphincter dysynergia, decreased sensation, decreased compliance, and hypercontractile detrusor. The results of the two small case series by Satar, et al.,46 and Adamson, et al.,1 indicated that improvement in bladder function is uncommon following cord release and that the urological outcome was due to irreversible dysfunction. Giddens, et al.,12 compared pre- and postoperative urodynamic testing in 14 patients and demonstrated that in four (29%) of the patients the symptoms improved and in 10 (71%) symptoms were unchanged. The best outcome was demonstrated in a subset of patients with preoperative external detrusor–sphincter dysynergia which resolved in three of four patients and remained unchanged in one postoperatively. Furthermore, Basar, et al.,9 reported improvement of urological symptoms in 67% of patients and improvement of urodynamic function in 49%; the best response, they noted, occurred in those patients treated early after diagnosis.

Long-term outcome after surgery for TCS appeared promising. Iskander, et al.,19 conducted telephone interviews in 22 of 28 patients during a mean follow-up period of 8.6 years after surgery. Twenty-nine (95%) of the 28 patients said that the operation was a long-term success; these patients experienced sustained significant postoperative improvement (and not just stabilization) in pain and/or neurological function. Furthermore, of 22 employed patients 19 (86%) returned to work after surgery. Two (33%) of six patients who were not employed before surgery gained full-time employment postoperatively. Two patients who had been receiving Workers’ Compensation returned to work. No formal quality of life or outcome scale measures were used in this study.

T. M. George and L. H. Fagan

<p>| TABLE 2 |
| Levels of evidence and grades of recommendation* |</p>
<table>
<thead>
<tr>
<th>Grade</th>
<th>Evidence Level</th>
<th>Requirement</th>
</tr>
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<tbody>
<tr>
<td>A</td>
<td>1a, 1b</td>
<td>≥1 randomized controlled trial as part of the body of literature of overall good quality &amp; consistency addressing the specific recommendation</td>
</tr>
<tr>
<td>B</td>
<td>2a, 2b, 3</td>
<td>availability of well-conducted clinical studies but no randomized clinical trials on the topic of recommendation</td>
</tr>
<tr>
<td>C</td>
<td>4</td>
<td>evidence obtained from expert committee reports or opinions &amp;/or clinical experiences of respected authorities; indicates absence of directly applicable clinical studies of good quality</td>
</tr>
<tr>
<td>GPP</td>
<td></td>
<td>evidence obtained from expert committee reports or opinions &amp;/or clinical experiences of respected authorities; indicates absence of directly applicable clinical studies of good quality</td>
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</table>

* Adapted from the Oxford Centre for Evidence-Based Medicine Levels of Evidence. Abbreviation: GPP = good practice points.

Evidence-based outcomes in Adult Myelomeningocele

Discrepancies in the terminology used in the literature have made it difficult to compare results in adult patients with myelomeningocele. The inconsistencies are important because patients with meningoceles typically exhibit normal neurological function at birth compared with those with meningoceles. The inconsistency of the descriptions used in studies is important because patients with meningoceles typically exhibit normal neurological function at birth compared with those who have suffered long-term preexisting static skeletal or neurological deficits since childhood.

Another possible discrepancy in terminology is that patients who were thought to harbor meningoceles may have been patients with postrepair meningoceles and defined a cohort of patients with symptoms and signs of tethering. This series represents Level 2b evidence. These authors retrospectively obtained evidence (see Table 2). These authors retrospectively obtained evidence from expert committee concerning patients with "occult myelomeningocele" that were probably meningoceles. The outcome of these two case reports may pertain to patients with meningoceles and, therefore, not to myelomeningoceles.

As expected, the addition of the medical condition that is the aforementioned condition of the patient with the median stalk of the spinal cord was not reported in the literature. The outcome in four adults who have suffered long-term postrepair myelomeningocele (mean 19 months) was evaluated by the afore mentioned cases. The patients appeared to retain much more distal sacral bladder and bowel incontinence and leg weakness. Each of these patients apprised normal neurological function at birth compared with those who have suffered long-term preexisting static skeletal or neurological deficits since childhood.

Clearer descriptions of tethering and repair myelomeningocele are listed in Table 2. Discrepancies in the terminology used in the literature were insufficient to draw conclusions.

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Outcomes in Adult Patients With Postrepair Myelomeningocele

Discrepancies in the terminology used in the literature have made it difficult to extrapolate outcome data for the adult patient with postrepair myelomeningocele. Meningocele is sometimes termed myelomeningocele and vice versa. The inconsistency of the descriptions used in studies is important because patients with meningoceles typically exhibit normal neurological function at birth compared with that of patients with myelomeningoceles; therefore, patients with meningoceles are not likely to harbor the same risk for the deterioration in their conditions. Logan, et al., described a patient with postrepair myelomeningocele who, after having experienced a normal childhood and young adulthood, was an active-duty Marine for 20 years before presenting with late symptoms of tethering. During clinical examination, a large well-healed lumbar incision was found and radiographs revealed an L-5 bifid lumbal lamina. In another case, Wilden and Hadley described a 72-year-old woman who was by her own account a patient with postrepair myelomeningocele; she presented with abrupt onset of bladder and bowel incontinence and leg weakness. Each of these patients appeared to retain much more distal sacral function than that which would be anticipated in a patient with a true myelomeningocele. Therefore, both of these cases may pertain to patients with meningoceles and, therefore, may not be useful to evaluate the surgical outcome in postrepair myelomeningocele.

As expected, the outcomes following untethering and repair of the meningoceles appeared to be quite good in the aforementioned cases. In additional cases, Yamamura, et al., reported that patients experienced improvements in pain, gait, and sensory function, although urinary incontinence remained unchanged. Klekamp, et al., described the outcome in four adult patients with meningoceles. Three of four patients demonstrated progressive symptoms and underwent surgical untethering. At the follow-up examination (mean 19 months), the conditions of two of the three patients had improved without recurrence of symptoms. The other patient whose condition deteriorated was treated conservatively and observed. Pang, et al., reported two cases concerning patients with "occult myelomeningocele" that were probably meningoceles. The outcome of these two patients were combined with the totals for this series, in which all types of dysraphic patients experienced similarly good outcomes. Another possible discrepancy in terminology is that patients who were thought to harbor meningoceles may have been patients with postrepair myelomeningocele. Mourarij, et al., reported on two patients with postrepair meningoceles and Yamamura, et al., described a patient with a lumbosacral scar in whom imaging studies revealed spina bifida from L4–S1 with a low and elongated spinal cord. Both of these reports may involve patients with postrepair myelomeningocele; however, the clinical data presented were insufficient to make a final determination. Cleared descriptions of TCS in adult patients with postrepair myelomeningocele were found in the literature and are listed in Table 3. Of the 23 patients reported by Pang and Wilberger one was determined to be a patient with postrepair myelomeningocele because of a scar and intraoperative findings consistent with a repaired myelomeningocele. The authors did not report the outcome for this patient specifically, although the overall surgical outcome for the patient discussed was reported to be good with regard to improvements in pain and motor weakness but disappointing in terms of the resolution of bowel and bladder dysfunction.

Tamaki, et al., reported a series of pediatric patients with postrepair myelomeningocele that included patients older than 18 years of age. The exact number of older patients and their individual outcomes were not provided; however, overall improvement was reported in 71% of all patients. Oi, et al., described eight patients with postrepair myelomeningocele. Five patients who were younger than 15 years of age presented with motor deficits. The three patients who ranged in age from 15 to 20 years old presented with pain and motor deficit and all of these patients were treated by untethering, although the authors did not disclose which patients had undergone surgery. The authors did note that there was no overall improvement after untethering. One 20-year-old patient in that series suffered recurrent pain 1 year after untethering. The results of the other patients were not disclosed.

Filler, et al., compared two adult patients with TCS from postrepair myelomeningocele with 12 adults with tethering of another origin. Precipitous loss of lower-extremity motor function occurred in the two adult patients with postrepair myelomeningocele, and both exhibited spondylolisthesis of unrecorded grade. The authors reported a good outcome for both patients after surgical release of the tethered cord. Dexmethasone was used preoperatively in one patient and improvement in neurological function was reported. The condition of this patient deteriorated 1 year later and then improved following the administration of steroid medication; this episode is attributed to trauma rather than re-tethering. Although the presence of degenerative spondylolisthesis is implicated as the cause for symptom progression, the impact on clinical symptoms that resulted from the spinal instability secondary to spondylolisthesis was not disclosed.

Selber and Dias reviewed the long-term outcome of 46 adult patients (mean age 23 years) who had suffered from sacral-level myelomeningocele. In 13 patients signs of TCS were evident, and 12 patients were untreated. The authors report that all patients improved postoperatively; however, results are not provided for individual patients or stratified by presenting symptoms.

Van Leuwen, et al., evaluated 57 consecutive adult patients with tethered cords resulting from varying origins; 12 were patients with postrepair myelomeningocele, and their outcomes were measured using the mRS and the motor-sum score during a follow-up period of at least 2 years. An independent examiner was used to eliminate observer bias; therefore, the results can be considered Level 2b evidence. Improvement or stabilization of motor symptoms was observed in 93% of patients. Three percent experienced postoperative motor deterioration. In 13 patients signs of TCS were evident and 12% of patients suffered rapid preoperative motor loss; however, changes for the motor-sum score were not significant for all patients (p = 0.069) or for those in the postrepair myelomeningocele and closed dysraphic subgroups. The mRS score improved in three (25%) of 12 patients with postrepair myelomeningocele and TCS, but this change was not significant.
TABLE 3

<table>
<thead>
<tr>
<th>Author &amp; Year</th>
<th>Description of Study</th>
<th>Evidence Class</th>
<th>Conclusions</th>
<th>Grade</th>
</tr>
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<tbody>
<tr>
<td>Pang &amp; Wiberg, 1982</td>
<td>retrospective case series of 23 adult patients w/ TCS, 1 patient w/ postrepair myelomeningocele reported</td>
<td>IV</td>
<td>postop outcome reported by symptom, not by cause of TCS; improvement in pain &amp; sensorimotor deficits; poor outcome for bladder function</td>
<td>GPP</td>
</tr>
<tr>
<td>Tamaki, et al., 1988</td>
<td>retrospective case series of 7 children &amp; undiscovered number of young adults; MRI for surgical outcome &amp; predictive nature of tethering; case report of adult w/ reported post-repair myelomeningocele</td>
<td>IV</td>
<td>postop outcome for young adults not disclosed; 71% of all patients improved</td>
<td>GPP</td>
</tr>
<tr>
<td>Logan, et al., 1989</td>
<td>prospective correlation of level of conus determined by MRI &amp; late deterioration in 8 of 74 children &amp; adults; untethering was performed in 5 of the 8</td>
<td>III</td>
<td>patient fared well; unclear whether myelomeningocele or a meningocele caused TCS</td>
<td>C</td>
</tr>
<tr>
<td>Ot, et al., 1990</td>
<td>prospective review of 2 patients w/ post repair myelomeningocele &amp; 12 adults w/ closed dysraphism</td>
<td>III</td>
<td>abrupt onset of symptoms noted in the 2 patients w/ myelomeningocele; patients improved postop, 8 of 12 patients w/ closed dysraphism improved</td>
<td>C</td>
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<tr>
<td>Filler, et al., 1995</td>
<td>retrospective review of 46 patients w/ sacral-level myelomeningocele; mean age 23;12 patients underwent tethered cord release</td>
<td>IV</td>
<td>good outcome reported in all patients</td>
<td>GPP</td>
</tr>
<tr>
<td>Selber &amp; Dias, 1998</td>
<td>prospective cohort study of 57 adult patients w/ TCS of various origins; outcome in motor-sum &amp; mRS scores were compared between 12 patients w/ post-repair myelomeningocele &amp; patients w/ closed dysraphism</td>
<td>IbB</td>
<td>early neurological deterioration in only the patients w/ lipomyelomeningocele; motor-sum score changes not significant in either group; improved mRS scores not noted in the postrepair group but were significant in the patients w/ closed dysraphism</td>
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(p = 0.059). In contrast, 80% of the patients with a closed dysraphism and TCS had improved scores that were statistically significant (p < 0.0001). The findings of this study reveal a better disability outcome for the patients with a closed dysraphism and TCS than for those with TCS resulting from postrepair myelomeningocele.

Complications Associated With Untethering

The mortality rate is low, with only one death reported in a diabetic patient due to meningitis from a CSF leak. Morbidity rates range from 11 to 36%; the most common lesions are superficial wound infection, wound dehiscence, and CSF subcutaneous pseudomeningocele. Significant morbidity resulted from intradural abscess of the cauda equina, which required exploration and drainage to relieve pain and led to paralysis and urinary incontinence.

The possibility of retethering remains a significant long-term problem. Filler, et al. observed retethering in 25% of the operated patients that occurred at intervals ranging from 1 to 9 years postoperatively. Huttman, et al., reported that 80% of patients in whom untethering was incomplete experienced recurrent symptoms within 5 years, Cochrane, et al., demonstrated that 55% of patients with postrepair myelomeningocele develop symptoms of retethering. Improvements in surgical technique may help reduce complications such as CSF pseudomeningoceles and retethering.

Conclusions

The literature regarding adult patients with postrepair myelomeningocele and TCS includes one Level 2b study, two Level 3 studies, and four Level 2 evidence studies. These patients often present with pain, sensorimotor loss, or bladder/bowel dysfunction that occur in a precipitous fashion. Magnetic resonance imaging is the imaging modality of choice, but CT scanning with or without myelography and plain radiographs may be helpful in defining associated lesions, spinal curvature, and spine instability. No clear role for ultrasonography has been established. A thorough urological evaluation, including urodynamic testing, is warranted to optimize urological outcome. The use of SSEP monitoring may be helpful in identifying compromised spinal cord function and predicting postoperative improvement in those patients with radiographic evidence of a tethered cord. Early aggressive surgical untethering and repair of the dysraphic lesion have been recommended but not sufficiently supported by outcomes and long-term compared with patients with postrepair myelomeningocele overall disability outcome. Currently 36%, and the mortality and CSF pseudomeningocele is the same entity, however, retethering postrepair myelomeningocele is the same entity, however, retethering is the same entity, however, retethering is the same entity, however, retethering.

Future prospective studies are needed to help determine the optimal timing of surgical untethering and repair myelomeningocele. Incorporating these studies will help determine whether myelomeningocele or a meningocele caused TCS. The researchers conduct a retrospective review of 2 patients w/ post repair myelomeningocele & 12 adults w/ closed dysraphism. Patients improved postop, 8 of 12 patients w/ closed dysraphism improved.

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Evidence-based outcomes and long-term outcomes compared with patients with postrepair myelomeningocele overall disability outcome. Currently 36%, and the mortality and CSF pseudomeningocele is the same entity, however, retethering postrepair myelomeningocele is the same entity, however, retethering is the same entity, however, retethering is the same entity, however, retethering.

Future prospective studies are needed to help determine the optimal timing of surgical untethering and repair myelomeningocele. Incorporating these studies will help determine whether myelomeningocele or a meningocele caused TCS. The researchers conduct a retrospective review of 2 patients w/ post repair myelomeningocele & 12 adults w/ closed dysraphism. Patients improved postop, 8 of 12 patients w/ closed dysraphism improved.
Evidence-based outcome in adult TCS

ciently supported by the available data. The operative outcomes and long-term results appear promising; however, compared with patients with closed dysraphism, the patients with postrepair myelomeningocele fared worse in overall disability scores and in improvement in neurological outcome. Complication rates ranged between 11% and 36%, and the most common complications were infection and CSF pseudomeningocele. Mortality rates are low; however, retethering is a significant long-term problem.

Future prospective studies, probably cohort controlled, are needed to help identify clinical, radiological, and electrophysiological predictors for adult patients with postrepair myelomeningocele and TCS. The researchers conducting these studies will need to determine whether adult TCS is the same entity in patients with closed defects as it is in those with postrepair myelomeningocele. These studies should be designed to delineate the natural history and address the optimal timing of surgery. Additionally, methods to prevent complications, such as CSF leakage, and strategies to improve poor outcome in symptom categories such as urological dysfunction need to be developed. Randomized trials to evaluate the role of prophylactic untethering and novel strategies to restore lost spinal cord function should be considered. An excellent resource for continued study of this group of patients is the National Institute of Child Health and Human Development’s Management of Myelomeningocele Study.

References


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In the majority of ambulatory patients with CP the clinical features of spasticity involve bilateral spasticity and, particularly in the lower limbs, a marked decrease in passive stretch and angular velocity of limb movements. Usually the knee is excessively flexed both at foot contact and in stance, whereas knee flexion is reduced and/or delayed in the swing phase. Typically reduced. Usually the ankle plantar flexion in swing occurs coupled with ankle plantar flexion in the swing phase. Usually the ankle plantar flexion in swing occurs coupled with ankle plantar flexion in the swing phase. Typically reduced.

Abbreviations used in this paper: CP = cerebral palsy; SDAI = Spasticity Disability Assessment Inventory; GMFCS = Gross Motor Function Classification System; GMFM = Gross Motor Function Measure; SD = standard deviation; SDR = selective dorsal rhizotomy.2,22