Abstract and Introduction

Abstract

Recurrent tethered cord syndrome (TCS) can lead to significant progressive disability in adults. The diagnosis of TCS is made with a high degree of clinical suspicion. In the adult population, many patients receive inadequate care unless they are seen at a multidisciplinary clinic. Successful detethering procedures require careful intradural dissection and meticulous wound and dural closure. With multiple revision procedures, vertebral column shortening has become an appropriate alternative to surgical detethering.

Introduction

Tethered cord syndrome is a condition that appears most commonly in the pediatric population. It results from stretching of the spinal cord between 2 fixation points, and treatment entails untethering the spinal cord. Results can vary depending on the time period between presentation and the initiation of treatment. If treated early, patients can recover from their neurological deficits. Chronic or excessive stretching of the spinal cord can lead to permanent disability. Currently the literature suggests that metabolic derangements and alterations in oxidative metabolism contribute to the reversible symptoms associated with TCS. Once neuronal damage occurs, however, partial neurological deficits can persist despite untethering.

For some patients, TCS can present de novo in adulthood or can impart recurrent clinical manifestations that extend into adulthood. Adult TCS may be neglected without the multidisciplinary care provided by a pediatric spina bifida clinic familiar with the natural history, diagnosis, and management of TCS. Given the fact that a majority of patients with spina bifida now live into adulthood, it is increasingly important to diagnose and appropriately treat TCS in adults. A lack of understanding or enthusiasm in treating adult TCS can lead to significant deterioration and
permanent disability.

Epidemiology

The incidence of neural tube defects is 0.17–6.39 per 1000 live births worldwide.\[^2\] There has been a decline in incidence over the past 20 years largely due to folic acid supplementation, termination of pregnancy after prenatal diagnosis, and other unknown factors. At centers experienced with myelomeningocele, the 20- to 25-year mortality rate is 24%.\[^1\] Significant improvements over the past couple of decades in the care of patients with myelomeningocele at centers for excellence have led to a growing population of these patients reaching adulthood. However, spinal cord tethering is a problem in this population. It occurs in 23% of patients between 1 month and 23.3 years after surgery. Twenty-nine percent of these cases occur in patients who have undergone > 1 tethered cord release procedure.\[^3\]

Clinical Presentation

In dealing with patients with TCS, a multidisciplinary team consisting of neurosurgeons, urologists, physiatrists, neurologists, orthopedic surgeons, and physical therapists is preferred. Patients with symptomatic TCS present with back pain, worsening motor weakness, or new bowel or bladder symptoms. Of these presenting symptoms, pain is the most common. Back pain occurs in 73% of patients presenting with TCS, whereas leg pain or sciatica is the presenting symptom in 56% of patients.\[^11\] Muscular weakness and bladder dysfunction occur as the presenting symptom in 78% and 71% of patients, respectively.\[^11\] Adults with a tethered cord tend to present in a delayed fashion compared with their pediatric counterparts.\[^12\] Adults may be in denial of any significant symptoms and unwilling to see their practitioner, and thus delaying their diagnosis and increasing their chances of disability. Progressive scoliosis, foot deformity, leg length discrepancy, or muscular atrophy diagnosed by a multidisciplinary team can also suggest TCS. Untreated, TCS can progress in 27.5%, 40%, and 60% of cases at 1, 2, and 5 years after diagnosis, respectively.\[^16\]

Diagnostic Workup

The determination of TCS is a clinical diagnosis. Potential shunt malfunction should be assessed in any patient with a ventriculoperitoneal shunt and undergoing evaluation for TCS before any treatment of a tethered cord. Head CT scanning and/or a shunt tap can be performed to determine shunt malfunction. Baseline manual motor test results from the physical therapist can be recorded and monitored on routine visits to the clinic. Routine urodynamic tests can detect any subtle changes in bladder function. Standing scoliosis radiographs should be obtained to monitor the progression of scoliotic curves as progressive scoliosis can indicate tethering. Once the diagnosis of TCS is made in conjunction with these tests, imaging is performed to identify potential areas of adhesions. Although MR imaging can visualize a low-lying cord, fatty filum terminale, or lipomyelomeningocele for the initial presentation of TCS, it is less useful for localizing adhesions. Comparing the location of the conus on previous MR images can help to support the diagnosis of TCS. Computed tomography myelography has been found to be more helpful in detailing the regions of adhesions, which prevent the flow of contrast through the thecal sac. More importantly, this imaging procedure provides an understanding of the bony anatomy that will be useful during exposure. Electrophysiological tests, such as SSEP recording and EMG, used in conjunction with imaging can also provide useful information. Somatosensory evoked potentials can provide information regarding the severity and extent of tethering.\[^17\] Somatosensory evoked potentials obtained preoperatively can be compared with postoperative measurements to confirm the benefits of surgery.\[^9\]

Surgical Management

The surgical management of tethered cord remains controversial. Although treatment in a patient with TCS and
evidence of neurological decline is straightforward, the care of patients who are asymptomatic despite their condition remains controversial. As previously stated, the natural history of TCS suggests a progressive decline when managed conservatively. Thus, in a symptomatic patient, any change in routine urodynamic or manual motor testing should be closely monitored.

With surgery, the initial approach is aimed at dealing with the local pathology. Over time, however, recurrent tethering can produce symptoms. In the adult population, the rate of retethering has been quoted to be as high as 25%.[5]

Lee et al.[11] found that surgical untethering can improve back and leg pain in 78% and 83% of patients, respectively. However, motor weakness will stabilize or improve in only 27% and 64% of patients, respectively. Sensory deficits remained unchanged in 50% of patients. Urological abnormalities improved in 50% of patients undergoing untethering and remained stable in 45%.

**Surgical Untethering**

When obtaining informed consent regarding any surgical untethering procedure, it is important to discuss all scenarios that may be encountered. Dural closures may be particularly difficult, and a CSF diversion procedure, such as a lumbar drain or external ventricular drain, may be required. A gluteal flap may be considered if significant difficulties are expected when closing the wound.

The patient is positioned prone after intubation. If any instrumentation is planned, devices are placed on a Jackson table. Any previous surgical incisions are marked out accordingly. A wide area is prepped laterally and inferiorly to include the buttocks and lateral thigh in the event that a gluteal flap is needed. Dissection is performed along the old incision site. It is important to create a single dissection plane deep and near the spine to preserve the subcutaneous tissue for closure. Ideally, dissection should be initiated along the superior or inferior margin of interest where normal bony anatomy is present to prevent entering the intradural spaces without recognition. Under these circumstances CT myelography is particularly helpful to isolate where there may be a defect in the lamina. Once the scar tissue overlying the dura as well as surrounding bony margins are identified, intradural exposure is performed. It is important to note on imaging if the spinal cord is adherent to the dura posteriorly given that entry into the intradural space can cause injury to the spinal cord. Using a 15-blade scalpel, a midline incision in the dura is made over the region of the suspected adhesions as demonstrated on CT myelography or MR imaging. A Woodson dissector is helpful intradurally to dissect a plane for sharp dissection or opening the dura. Edges of the dura are tacked up to the surrounding tissue by using a 4-0 nonabsorbable suture. A microscope is brought in for better visualization. From here, the Rhoton instruments, microscissors, and an arachnoid knife are helpful in providing safe lysis of adhesions. On occasion, the CO₂ laser may be helpful as a means of the subtle lysis of adhesions when the risk of injuring or pulling the cord with sharp or blunt dissection is significant.[4] When bipolar electrocautery is used, Isocool (Codman/Johnson & Johnson Professional) or nonsticking bipolar tips are used as well. The lysis of adhesions should continue until the cord is visualized to fall freely from its attachments.

Lumbosacral nerve roots should be identified prior to microsurgical untethering. Microsurgical techniques should focus on the careful lysis of adhesions and the removal of significant pathology leading to tethering. Such techniques are performed in conjunction with intraoperative monitoring to prevent any significant morbidity from occurring.

During evaluation, careful attention should be directed at all lumbosacral nerve roots including the anal sphincter. Common modalities used to monitor include SSEP recording, spontaneous EMG, and triggered EMG. Somatosensory evoked potentials offer high specificity and low sensitivity for detecting damage to the nervous system. Conversely, continuous EMG provides high sensitivity and low specificity in detecting new neurological deficits.[15] Thus, neurological monitoring with SSEP recording and EMG provides high specificity and sensitivity in
detecting new neurological deficits. With untethering of the spinal cord, SSEPs have been shown to improve.\[9\] Spontaneous EMG is used to detect inadvertent manipulation of nerve roots. Triggered EMG allows for the stimulation of tissues for the purpose of differentiating important neural structures from adhesions or scar tissue when performing a dissection.

After the lysis of adhesions is complete, closure of the dura is achieved with a running 4-0 locking stitch. Placing DuraGen (Integra LifeSciences) subdurally at the time of closure can be considered as it has been shown to reduce postoperative fibrosis.\[7\] A Valsalva maneuver is performed by an anesthesiologist to ensure no CSF leakage from the closed durotomy site. A dural sealant can be applied to the dural edges if leakage occurs around the margins. In certain circumstances in which primary dural closure is not achieved, a dural substitute can be used to close the wound. Subcutaneous wound closure is then performed. Note that a primary closure is desired. A subfascial Jackson Pratt drain can be left in place if necessary. When a drain is inserted, no or minimal bulb suction is applied without promoting CSF leakage into drain. In patients with a history of posterior operations, subcutaneous wound closure can be difficult. Achieving a tight fascia closure is ideal in the setting of a closed durotomy, and this can be done with a nonabsorbable suture instead of Vicryl sutures. After fascial closure, the subcutaneous tissue may be tense especially in regions where there is a significant amount of scar tissue. Fascial release both above and below the plane may provide adequate release of tension for closure of the subcutaneous suprafascial tissue. In certain circumstances in which there is not enough suprafascial tissue to provide for complete closure, plastic surgeons may be needed to perform a gluteal flap.

With multiple untethering procedures, arachnoid scarring around the nerve roots can lead to potential neurological deficits that may be difficult to treat. There is the risk of damaging spinal nerve roots while performing the dissection. This risk increases with each subsequent untethering operation. Multiple surgeries also increase the risk of wound dehiscence and wound infection.

**Postoperative Management**

The most common complication of surgical untethering is CSF leakage. The patient should be kept flat for 3–5 days to allow the durotomy site to heal. If it is believed that a CSF diversion procedure is needed, a decision needs to be made regarding the placement of a lumbar drain versus an external ventricular drain. For patients with myelomeningocele and shunted hydrocephalus, a functional shunt may need no additional intervention. Given a low-lying cord in most circumstances, a lumbar drain should be placed under fluoroscopic guidance. However, in a small portion of patients without a VP shunt in which a lumbar drain cannot be safely placed or if placement of the lumbar drain involves entering the surgical bed, an external ventricular drain is preferable. An MR image is obtained within 48 hours to establish a baseline position of the spinal cord.

**Vertebral Column Shortening**

Patients who have recurrent symptoms despite multiple detethering procedures may be candidates for a vertebral column shortening procedure. Vertebral column resection and pedicle subtraction osteotomy are 2 powerful techniques used in the correction of spinal deformity. These 3 column osteotomies allow the spine to be manipulated in both the coronal and sagittal planes. Both techniques effectively shorten the vertebral column.

In 1995 Kokubun\[10\] described a case report of spinal osteotomy for vertebral column shortening in a patient with a low-lying conus medullaris. Grande et al.\[6\] used a cadaveric model to examine the effect of vertebral column shortening at the thoracolumbar junction on the spinal cord, lumbosacral nerve root, and filum tension. They found that shortening the vertebral column by 15–25 mm significantly reduces tension on the spinal cord, nerve roots, and filum terminale. To achieve this same effect by a traditional detethering procedure would require the release of more than 90% of the neural elements.
Since Kokubun's original report, 2 other papers have been published in the English language describing vertebral column shortening procedures for the treatment of tethered cord.\textsuperscript{8,13} All procedures were performed at the thoracolumbar junction (4 at L-1 and 1 at T-12). There are several advantages in performing a VCR at these levels. Candidates for this procedure have often undergone several detethering procedures in the lumbosacral region. Typically, both intra- and extradural scar tissues are present, which can buckle during closure of the osteotomy and cause neurological injury.\textsuperscript{14} Vertebral column resection also avoids the reopening of friable dura, which poses a challenge to watertight closure and leads to CSF leakage. The thoracolumbar junction has minimal curvature, which makes approaching and closing the osteotomy technically easier. Furthermore, VCR at the thoracolumbar junction minimizes the loss of motion that would occur if the procedure were performed at lower lumbar levels.

Miyakoshi and colleagues\textsuperscript{13} 3 cases were initially treated for TCS. Because of VCR's technical demands and relatively limited track record, it is our practice to reserve this resection as an option in patients with multiple retetherings who have been treated with traditional detethering. We also consider VCR in patients in whom traditional detethering has failed or the risk of neurological decline with further detethering is thought to be elevated. Further experience will determine optimal indications for VCR. The procedure has been highly successful in a limited number of patients thus far and may become a more common operation for TCS in the future.

**Illustrative Case**

**History and Examination.** A 37-year-old woman with a history of spina bifida and 4 previous tethered cord releases presented with the chief symptoms of back and bilateral leg pain, which occurred immediately after a child ran into her knocking her to the ground. Her symptoms progressively worsened with increased pain, numbness, and decreased gait and urinary function. Her surgical history was also notable for intrathecal pain pump placement at the time of the last detethering procedure. Neurological examination revealed 4/5 strength on left dorsiflexion, plantar flexion, and extensor hallucis longus muscle function. Sensory examination revealed numbness in L-5 and S-1 dermatomes with absent Achilles reflexes. She stood in neutral coronal and sagittal alignment and slightly favored her left leg on gait testing.

A review of radiographic images revealed multilevel midline defects in her lumbar spine consistent with laminectomy defects. Magnetic resonance imaging suggested ventral tethering of the cord at the L5–S1 level and dorsally at the L3–4 level with a degenerated disc at the L4–5 level (Fig. 1). Standing scoliosis radiographs showed normal coronal and sagittal alignment; no motion was noted on lumbar flexion and extension radiographs.
Figure 1. Magnetic resonance image of the lumbar spine revealing a low-lying conus and ventral tethering of the cord at the L5–S1 level.

After a thorough discussion of the risks and benefits of a traditional tethered cord release versus VCR, the patient elected to undergo a T-12 VCR with a T10–L2 posterior spinal instrumentation and fusion.

Surgical Technique. After inducing general endotracheal anesthesia, the patient was placed in a Mayfield head holder and carefully positioned prone on a Jackson frame. (As this patient population can have shunted hydrocephalus, take care not to injure any shunt apparatus with the Mayfield fixation.) All pressure points were carefully padded. After prepping and draping in a standard fashion, a midline incision was made. Her intrathecal catheter was identified and preserved during the operation. Subperiosteal dissection was performed from T10–L2. Radiographic verification of all levels was obtained. Pedicle screws were placed from T-10 to L-2 using a freehand technique. A T-12 and partial T-11 laminectomy was performed while saving bone for an autograft. The pedicles were then resected flush with the vertebral body. The T-12 rib was identified and the rib head was resected. The T11–12 disc space was identified. At this stage, 1 cm of the cancellous vertebral body was resected bilaterally using a drill, curettes, and pituitary rongeurs, while saving cancellous bone for use as an autograft. Temporary rods were placed for column stability while performing the osteotomy. A T11–12 discectomy and T-12 superior endplate resection was performed. The defect measured 20 mm at this point and was believed to be sufficient to achieve the surgical goals. The osteotomy was confirmed to be symmetric on both sides. Lateral vertebral body walls were resected to match the defect in the cancellous bone. A posterior vertebral body wall impactor was positioned ventral to the dura and used to disengage the posterior vertebral body wall, including the T11–12 disc. The posterior wall was impacted anteriorly into the osteotomy defect and was carefully removed. A Woodson elevator confirmed the neural elements were free of residual bone and disc material. The osteotomy was then closed using temporary rods, with careful attention paid to the amount of dural buckling. Somatosensory and motor evoked potentials were monitored throughout the osteotomy closure. No electrophysiological changes were noted. Permanent rods were then placed, and the osteotomy was inspected and confirmed to be bone on bone. Intraoperative CT scanning confirmed appropriate placement of the pedicle screws and bone-on-bone contact of the osteotomy. Posterior arthrodesis was performed. A large amount of locally harvested autograft from the osteotomy acted as the primary graft material. A subfascial drain was placed and the wound was closed in standard fashion.

Postoperative Course. Postoperatively, the patient noted improvement in her neurological status. Her leg pain and urological function returned to baseline. Standing scoliosis radiographs demonstrated bone-on-bone contact for the osteotomy and solid fixation of spinal instrumentation (Fig. 2).
Figure 2. Lateral plain radiograph showing bone-on-bone contact in the region of the T-12 VCR with posterior segmental instrumentation.

References

EMG = electromyography; SSEP = somatosensory evoked potential; TCS = tethered cord syndrome; VCR = vertebral column resection.

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