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Surgical Management of the Tethered Spinal Cord - Walking the Fine Line

James M. Drake, F.R.C.S.C.

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Abstract and Introduction

Abstract

The tethered spinal cord is a fascinating yet controversial condition seen frequently in neurosurgical practice. Treatment decision making is made difficult by the variety of lesions and clinical presentations comprised by this condition and the absence of high-quality clinical outcome data to provide guidance. Clinical presentations may be divided into four general categories or typical scenarios: 1) significant dysraphic abnormality, clear clinical deterioration; 2) significant dysraphic abnormality, clinically normal or stable deficit; 3) incidentally discovered abnormality, other problem; and 4) tethered spinal cord symptomatology, normal imaging. The author provides case examples to illustrate potential treatment approaches and suggests balancing the risks and benefits for each general category.

Clearer diagnostic and treatment strategies for the tethered spinal cord will only result from high-quality clinical and basic research. Until the results of such research are available, surgeons should endeavor to maximize benefit and reduce risk for patients who may have a tethered spinal cord, walking the fine line between over- and undertreatment.

Introduction

Few topics in pediatric neurosurgery generate as much controversy, debate, or angst as the diagnosis of tethered spinal cord. As is often the case in surgery, the controversy is fueled by plausible explanations and strong convictions based largely on anecdote, and its intensity is all the greater because of a lack of good data. Patients with alleged tethered spinal cords are fairly common in pediatric neurosurgical practice. Walking the fine line between over- and undertreatment seems a rational approach until better information becomes available. As previously discussed,^[3] these cases, united by a single pathogenetic concept--tethering of the spinal cord--are a very diverse group clinically and radiologically. Rather than discuss each possible abnormality, it seems reasonable to address the common presentations individually, beginning with the most severe, where surgery is least questioned, moving to the "almost" normal cases--in which surgery might almost be regarded as an alternative therapy joining the ranks of craniosacral therapy, acupuncture, and "cupping."

Significant Dysraphic Abnormality, Clear Clinical Deterioration

Patients with a significant abnormality, such as a lipomyelomeningocele, and clear clinical deterioration--progressive leg weakness, urinary incontinence--would seem to be the most straightforward surgical candidates. In this scenario, the surgeon and the parents or patient usually agree that the potential benefits outweigh the risks. Figure 1 shows images obtained in such a patient. Born with an obvious lipomyelomeningocele, the patient was apparently neurologically normal. An early imaging study, performed when the patient was 3 months old, demonstrated the typical features of a lipomyelomeningocele, with extension of the lipoma into the left buttock suggesting that untethering might be technically difficult (Fig. 1A,B). The patient's condition was routinely monitored in both neurosurgery and urology clinics. Subtle deterioration over the next 3 years--repeated bladder infections culminating in the recommendation for intermittent catheterizations, in-turning of the left foot, and loss of anal tone--prompted a repeated imaging study, which demonstrated an enlarged syrinx in the distal spinal cord (Fig. 1B). The child underwent surgery and the cord was completely untethered. Surgery was complicated by a CSF collection requiring a subcutaneous peritoneal shunt. The patient's condition improved rapidly following surgery with respect to foot position, anal tone, and bladder function. Follow-up imaging showed reduction in the size of the syrinx and

lipoma component, but the lipoma was apposed to the overlying dura (Fig. 1C.)

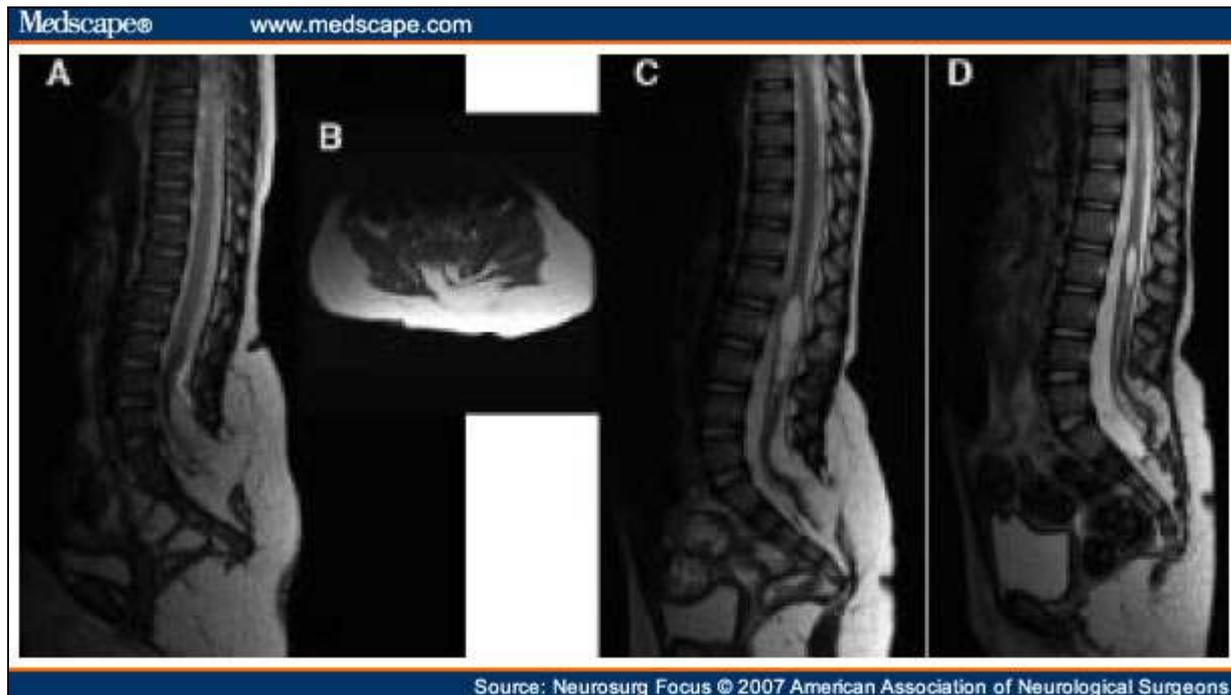


Figure 1. Magnetic resonance images obtained in a patient with lipomyelomeningocele. A,B: Sagittal (left) and axial (right) images obtained when the patient was 3 months of age. Note extension of the lipoma into the left buttock. There was a very small syrinx of the distal spinal cord, not well demonstrated on these images. C: Sagittal image obtained when the patient was 3 years old showing progressive enlargement of the syrinx. The patient had experienced concomitant clinical progression. D: Postoperative follow-up MR image showing a decrease in the size of the syrinx and apposition of the remaining lipoma to the overlying dura.

Although this case represents only a single anecdote, it nevertheless provides evidence that progressive deterioration occurs in some patients and illustrates the kind of early improvement that can occur following surgery. On a larger scale, it is important to note that there is literature that supports the notion that clinical progression in patients with lipomyelomeningocele is predictable based on the anatomy of the lipoma^[2] and that surgery may not prevent this progression over the long run.^[8]

There are several caveats about surgery for these lesions. Lipomyelomeningoceles manifest significant variability in their configuration and the technical difficulty of treatment. While the lesions are classically divided into dorsal, transitional, and terminal, typically lipomyelomeningoceles are transitional, and vary most in terms of their relationship to the spinal cord and surrounding nerve roots. In cases involving lipomyelomeningoceles that are quite asymmetrical, and particularly those that have large lipomatous components exiting either out nerve root sheaths or into the sacral epidural fat, it can be very difficult--in some cases, impossible--to perform a complete untethering procedure safely. In such scenarios, it is probably better to perform only a partial untethering rather than risk an immediate atonic bladder, the most common serious complication. Intraoperative monitoring can be used to identify partially obscured roots or detect an early neurological injury, manifest by evidence of persistent spontaneous muscle activation, but it has not been convincingly proven to improve the outcome of surgery.

A CSF leak remains the other main complication and can be quite difficult to prevent or treat. The defect in the thinned-out dura following lipoma excision can be significant and may require patching either with autologous fascia or dural substitute and sealing with any of various fibrin preparations. Leaving the subcutaneous lipoma as a biological graft (for subsequent elective removal by liposuction) or inserting a lumbar drain intraoperatively (impossible to do percutaneously postoperatively) are other unproven strategies to prevent CSF leak.

Although clinical deterioration can be fairly rapid, and occasionally acute, in cases in this category, it may more often be described as "glacial," especially in cases involving older children. For this reason, continued clinical

monitoring, with careful documentation of motor, sensory, and urodynamic function, is important. Deterioration that is extremely slow, but persistent, often makes decision making regarding surgery, particularly in terms of timing, difficult. Parents and patients themselves may find that the pace of deterioration is so slow that surgery should be delayed until a certain point is reached, although that point is usually ill defined.

Significant Dysraphic Abnormality, Clinically Normal or Stable Deficit

Into this fairly common group usually fall the asymptomatic lipomyelomeningoceles, spina bifida occulta with abnormal MR imaging findings (such as a lipoma of the filum terminale, Fig. 2), diastematomyelia, caudal regression syndromes (such as imperforate anus), and dermal sinus tract (not a normal sacral dimple). The true dermal sinus tract, usually in the lower lumbar area and sometimes discharging detritus or pus, is clearly of some risk to the patient, and should be investigated and treated expeditiously (Fig. 3). Episodes of meningitis are not uncommon, and many patients harbor an intraspinal dermoid. This dermoid can often be completely excised, de novo, but following recurrent episodes of infection, and particularly an intraspinal abscess, an entangled mass of cyst, nerve root, and scar may develop—impossible to completely remove and almost certain to redevelop after incomplete removal.



Figure 2. Axial (left) and sagittal (right) MR images obtained in a patient with a fatty filum and a normal-level conus.

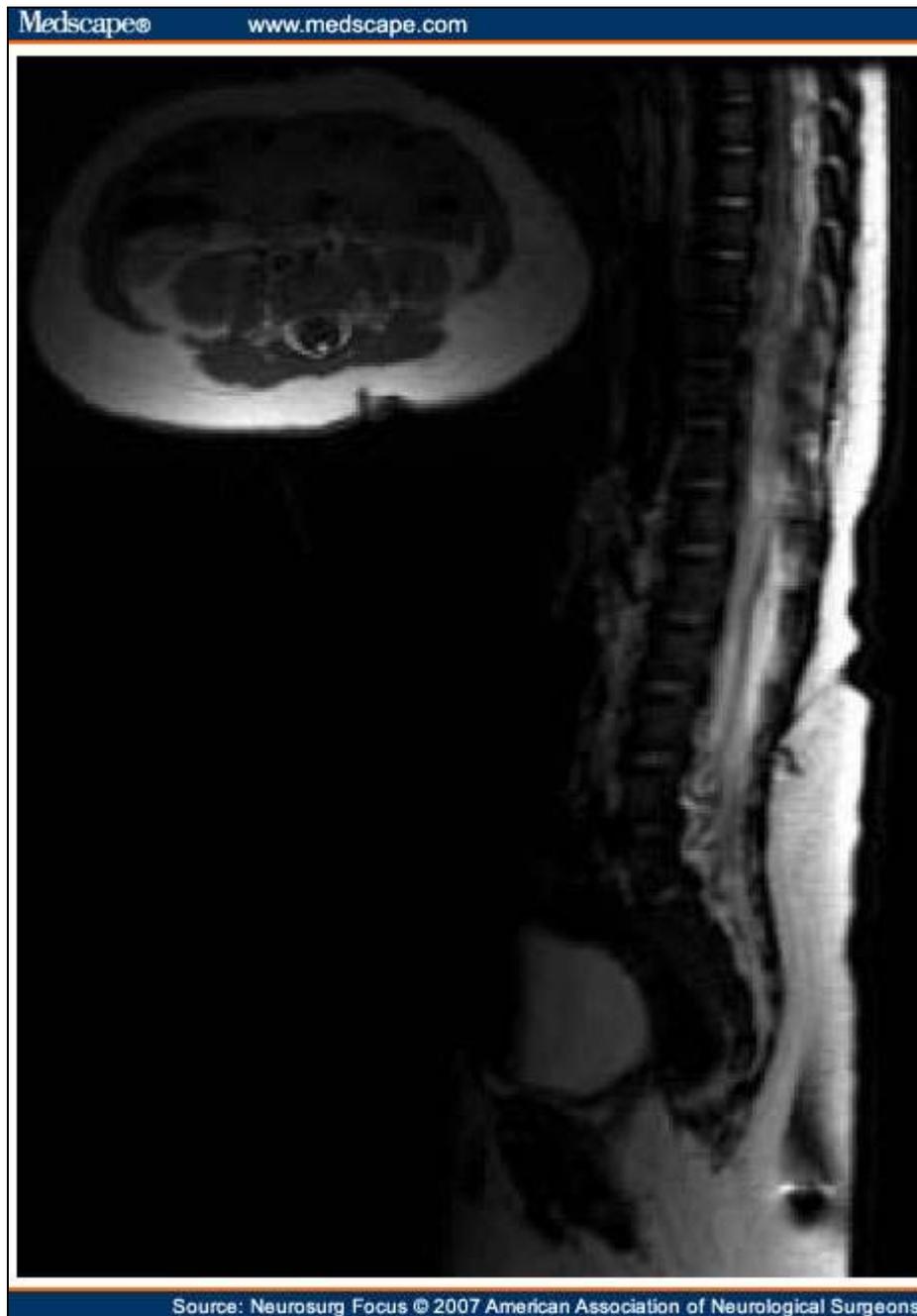


Figure 3. Axial (left) and sagittal (right) MR images obtained in a 3-month-old patient with a lumbar dermal sinus tract. There is an intraspinal lipoma of the filum but no inclusion cyst.

The indications for surgery for the other lesions are less clear. While it has been held that clinical deterioration in cases of lipomyelomeningocele, in particular, is inevitable, and therefore that treatment should be undertaken prophylactically,^[5] as discussed previously, this tenet is not universally held. Complications in neurologically normal patients are particularly distressing to surgeons as well as patients. In patients with preexisting deficits, for whom some possible improvement might be entertained, complications seem somewhat more tolerable. The worst of the most common complications is a neurogenic bladder; patients who experience this complication can expect a lifetime of incontinence, drug therapy, and intermittent catheterizations.

There is even less evidence for surgical treatment of split cord malformations than for surgery in cases of lipomyelomeningocele. Split cord malformations are quite rare and may take various forms, with the most significant probably being a bone septum dividing separate dural sacs.

In cases involving neurologically normal patients without a true dermal sinus tract, I think a reasonable approach is

careful observation, with a plan to intervene should deterioration occur. One must be aware of "glacial" deterioration as discussed above. It is useful to obtain a baseline measurement of bladder function with a voiding cystourethrogram, and in the presence of incontinence or recurrent bladder infections urodynamic testing is an important method of monitoring. Subtle changes in the results of urodynamic testing alone, however, do not appear to warrant intervention as there is considerable variability in the tests and their interpretation.^[18] One should be prepared to regard changes that could reflect maturation of the nervous system or could be due to extremity growth (progressive Achilles tendon shortening) as not being mandatory indications for surgery. This is clearly a difficult task, and the need to be careful in interpreting test results when monitoring a patient's condition cannot be overemphasized.

Incidentally Discovered Abnormality, Other Problem

Not infrequently patients undergo an MR imaging examination as part of an investigation for some other problem, and imaging features compatible with a "tethered" cord are discovered. In some cases these findings are almost to be expected--for example, in cases of caudal regression syndrome or Currarino triad. Patients with these conditions frequently have dysfunction related to their abnormality and experience bowel and/or bladder incontinence because the innervation of these organs overlaps with the lower lumbar and sacral roots. Nevertheless, even in the presence of dysraphic components, including a low spinal cord and a thickened and fat-filled filum, progressive deterioration is not inevitable.^[17]

Other common scenarios include cases of scoliosis with what appears to be an idiopathic curve, in which the conus is reported as being low or "borderline low" at the inferior margin of L-2 with or without fat in the filum. The concerns in such cases are whether this finding suggests a causative mechanism for the scoliosis and whether--to prevent a spinal cord injury--the condition should be treated prior to or at the same time as the spine is straightened and instrumentation is placed. There is a normal distribution of position of the conus in healthy patients, and a position at one end of this range does not mean surgery is indicated. The pathogenesis of scoliosis is as poorly understood as that of the tethered spinal cord, so invoking one to explain the other really muddies the waters. In this setting I leave the filum alone.

Patients with formation and segmentation abnormalities of the spine, such as hemivertebrae, block vertebrae, or Klippel-Feil syndrome, often have spinal curves and may have an abnormally positioned conus. These congenital curves have a clearer mechanical mechanism: asymmetrical growth. Counting the vertebrae to assign conus position can be difficult, and the results may have little meaning. The significance of a conus that is low by one vertebral level, with "lumbarized" or "sacralized" vertebrae at the lumbosacral junction is unknown, and such a finding should probably be ignored, especially if the patients are normal neurologically (as are those under consideration in the current discussion.)

Tethered Spinal Cord Symptomatology, Normal Imaging Findings

This remains the most controversial of all the tethered cord scenarios. Even though the concept arose in my own institution,^[4] my position remains a skeptical one, as outlined in a recently published series of articles.^[3,12,13] There are several reports of symptoms of incontinence improving after division of what appears to be a normally positioned filum terminale,^[4,19,20] and in some of these cases abnormal urodynamic test results had been documented. Without revisiting the entire debate, I will simply point out that these are single-surgeon retrospective reviews, with poorly defined entry criteria, outcomes, complications, and so forth. It is worth remembering that recent randomized trials with a sham surgery arm demonstrated significant clinical improvement in the sham surgery group.^[6,7] Incontinence is so common in children as to be regarded as almost normal, and it is difficult to quantify, even with urodynamic testing.^[1,10,14,16,21]

The most problematic area of controversy is pathogenesis. Acute tension on the spinal cord has been shown to produce spinal cord dysfunction and deranged metabolism in animal experiments,^[15,23] but it is not known how these findings relate to the chronic situation typical of the patient with a tethered spinal cord. Superficial in vivo measurements of changes in cytochrome gradients^[22] or laser Doppler blood flow^[9] have been demonstrated in patients, but again the significance of these changes is not clear. Although increased tension on the conus from a

thickened or fibrous filum^[11] or asymmetric growth of the spine and spinal cord provides a plausible explanation for spinal cord dysfunction, plausibility does not equal causality. Developing in vivo methods of measuring spinal cord blood flow and metabolism would presumably shed a great deal of light in this area. There is a current prospective pilot study of patients with secondary incontinence, evidence of a neurogenic bladder, and normal imaging, randomized to either surgical division of the filum or best medical management, in which our centre is participating (Paul Steinbok, personal communication). A larger study of this nature is probably the only way that the role of surgery in this condition can be evaluated.

Conclusions

The tethered spinal cord syndrome remains a fascinating surgical enigma, surrounded by much debate, controversy, and uncertainty. Progress in resolving issues related to the myriad conditions that can masquerade as a tethered spinal cord will only come with better information from basic research and prospective clinical trials. Surgeons who advocate treatment "just in case it helps" should be sure they are doing more good than harm.

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Authors and Disclosures

James M. Drake, F.R.C.S.C.

Division of Neurosurgery, Hospital for Sick Children, University of Toronto, Toronto, Canada

Reprint Address

James Drake, F.R.C.S.C., Division of Neurosurgery, Hospital for Sick Children, Room 1504, 555 University Avenue, Toronto, M5G 1X8, Canada. email james.drake@sickkids.ca .

Abbreviation Notes

CSF = cerebrospinal fluid; MR = magnetic resonance.

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