Treatment of the Occult Tethered Spinal Cord for Neuropathic Bladder: Results of Sectioning the Filum Terminale

P. D. Metcalfe,* T. G. Luerssen, S. J. King, M. Kaefer, K. K. Meldrum, M. P. Cain, R. C. Rink and A. J. Casale

From the Departments of Pediatric Urology (PDM, SJK, MK, KKM, MPC, RCR) and Pediatric Neurosurgery (TGL), James Whitcomb Riley Hospital for Children, Indiana University School of Medicine, Indianapolis, Indiana, and Pediatric Urology, University of Louisville (AJC), Louisville, Kentucky

Purpose: Occult tethered cord syndrome applies to patients with signs and symptoms consistent with a caudal spinal cord malformation despite normal neuroimaging. Although several reports of successful surgical treatment exist, controversy remains with respect to patient selection and efficacy. We present a large series with excellent clinical followup, neuroimaging and urodynamic characterization.

Materials and Methods: We present our experience with 36 patients at a single institution with preoperative clinical findings, neuroimaging and urodynamics available. Postoperative outcomes were assessed clinically and with urodynamics. We determined predictive parameters to improve patient selection.

Results: Approximately 0.04% of pediatric urology clinic visits resulted in neurosurgical referral for the potential of an occult tethered cord. They occurred after failure of a mean of 2 years of aggressive medical management. Daytime urinary incontinence was present in 83% of patients and 47% had encopresis. Preoperative urodynamics were markedly abnormal in all patients with mean bladder capacity 55% of expected capacity. Clinical improvement in urinary symptoms was seen in 72% of patients with resolution of incontinence in 42%. Bowel symptoms improved in 88% of cases, including resolution of encopresis in 53% within 3 months of surgery. Urodynamic improvements were demonstrated in 57% of cases. We were unable to determine preoperative factors that were more likely associated with surgical success.

Conclusions: In a highly select population with severe urinary and fecal dysfunction sectioning a normal-appearing filum terminale can result in significant improvement. We were unable to identify factors that may increase the chance of surgical success.

Key Words: bladder; bladder, neurogenic; neural tube defects; urodynamics

Spinal cord tethering occurs if the caudal spinal cord is prevented from normal upward migration during development or prevented from free movement during spinal motion. The clinical syndrome includes various orthopedic, neurological, cutaneous or urological symptoms and signs generally referred to as tethered cord syndrome.[1] When neuroimaging in these patients reveals an abnormally low lying conus medullaris, surgical release of the tether is often beneficial.[2]

Recently it was suggested that tethered cord syndrome may occur with normal anatomy on MRI.[3–7] Patients with this syndrome have been reported to improve after section of an apparently normal filum terminale. This entity has been referred to as occult tethered cord syndrome.[8] It has been suggested that some patients with DES actually have tethered cord syndrome, and enuresis and daytime incontinence improve after filum section.[3,6,9–13] Surgical release may relieve tension and improve cranial spinal cord migration.[2] Unfortunately definitive guidelines for treatment have not been developed and patient selection is widely variable.

At our institution sectioning of the filum terminale has been offered to rare patients with a normal-appearing MRI when they meet stringent clinical and urodynamic conditions. These patients are our most severe, medically refractory patients with DES and they must meet urodynamic criteria to be considered for surgery. We reviewed our data to determine short-term and long-term outcomes, and predictors of successful surgical management.

METHODS

This study was approved by the Indiana University-Purdue University Indianapolis Institutional Review Board. A retrospective chart review was performed in 18 male and 18 female patients who underwent section of the filum terminale for DES from 1997 to 2004. Average patient age at surgery was 8.3 years (range 1.2 to 15). Mean followup was 49 months and 25 patients had greater than 3 years of followup.

All patients were referred for neurosurgical evaluation by the referring urologist only after a prolonged attempt at medical management had failed. This included anticholinergic therapy, aggressive bowel management with polyethylene glycol, biofeedback training and prophylactic antibiotics, when appropriate. Patients were managed in concert

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* Correspondence: Riley Hospital for Children, 702 North Barnhill Dr., Suite 4230, Indianapolis, Indiana 46202 (e-mail: pmetcalf@iupui.edu).

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TABLE 1. *Significant pathological findings in our cohort despite maximal medical therapy**

Symptom	% Prevalence
Daytime urinary incontinence	83
Nocturnal enuresis	78
Recurrent urinary tract infection	36
Constipation	69
Encopresis	50

* Including maximal anticholinergic therapy, an aggressive bowel program and biofeedback training.

with the neurosurgeon, urologist and family. All except 3 neurosurgical consultations and procedures were performed by a single neurosurgeon (TGL).

Voiding symptoms and the degree of improvement were assessed by the urologist, as reported by the patient and family, while neurological symptoms and physical examination findings were determined by the neurosurgeon. Bowel symptoms were categorized only by the subjective presence of constipation and encopresis. Any improvement in symptoms was only considered a result of surgery if present at the 3-month followup clinic appointment and they were excluded if improvements were first documented greater than 1 year after surgery. Filemaker Pro 7.0, version 2 (Filemaker, Santa Clara, California) was used for data collection and statistical significance was determined using chi-square analysis.

RESULTS

During the study period approximately 90,000 patients were seen at our pediatric urology clinics. Therefore, these 36 patients comprise 0.04% of all clinic visits. Mean time from original urological consultation to initial neurosurgical evaluation was 23 months.

There were no neurological complications from surgery. There were 2 superficial wound infections, which responded well to bedside débridement and antibiotics.

Preoperative Findings

Clinical symptoms. DES symptoms were severe and disabling (table 1). Only 1 patient was not on anticholinergic therapy because he was being treated for urinary retention. Nonurological symptoms were elicited by the neurosurgeon in 20 patients (table 2). Significant physical findings were seen in 16 patients (table 3).

MRI was done preoperatively in all patients and there were no pathognomonic abnormal findings. Of the 36 patients 27 had a completely normal MRI with a conus located above the L2 level, and a normal-appearing conus medullaris and cauda equina. The remaining 9 patients had subtle findings considered by our team to be within the realm of normal. If considered in isolation, these indications would not be an indication for surgery (table 4).

TABLE 2. *Symptoms elicited by neurosurgeon that were not detected by urologist*

Symptom	No. Pts
Leg or foot pain	7
Back pain	6
Increased pain with exercise	6
Decreased perineal sensation	3
Lower extremity clumsiness	3
Total	25

There were 20 patients with these 25 extra-urinary symptoms.

TABLE 3. *Subtle physical findings elicited by neurosurgeon that were not detected by referring urologist*

Physical Finding	No. Pts
Abnormal patellar or Achilles reflex	7
Asymmetric feet	4
Asymmetric gluteal crease	2
Scoliosis	2
Lumbar venous plexus	1
Abnormal lumbar pigmentation	3
Total	19

There were 16 patients considered to have an abnormal examination, accounting for these 19 findings.

Urodynamic data. Preoperative urodynamics were performed using standard techniques with room temperature water infused at a rate of 10% expected capacity per minute. These studies were markedly abnormal in all 36 patients tested. Unstable contractions were present in 25 of 36 patients (69%) and the mean amplitude of these contractions was 40 cm H_2O.

Maximum cystometric capacity was less than expected for age, as determined by the equation, age + 2 in years $\times$ 30 ml, in 32 of 36 patients (89%). Mean capacity was 55% of that expected. Of the 36 patients 21 (58%) had less than 50% of expected capacity for age. Volume at first sensation was also markedly decreased at a mean of 98 cc with a mean expected bladder capacity of 33%. First desire to void occurred at a mean volume of 136 cc, equivalent to 47% of expected capacity.

Postoperative Results

Clinical symptoms. Patients and families reported urinary symptom improvement in 26 of 36 patients (72%) and constipation improved in 23 of 25 (92%) (table 5). Of the patients 22 (63%) were able to discontinue anticholinergic medication by the 1-year postoperative visit. In 4 patients bowel function improved without concomitant urinary improvement. There were no instances of clinical deterioration after improvement was noted at the 3-month followup appointment.

Urodynamic data. Of the patients 28 had preoperative and postoperative urodynamics available and improvement in urodynamic parameters was noted in 16 (57%) (table 6). Seven of the 8 remaining patients had an excellent response to surgery and invasive examination was believed to be

TABLE 4. *MRI findings*

MRI Finding	No. Pts
Conus L2–L3	2
Hydromyelia	1
Minimal fatty infiltration	6
Total	9

Our team of neurosurgeons, urologists and radiologists considered these results a variation of normal and in isolation they would not be considered an indication for intervention.

TABLE 5. *Patients who reported any degree of improvement in urinary symptoms*

Symptom	% Overall Improvement	% Cure	
		At 3 Mos	Within 1 Yr
Urinary symptoms	72		
Daytime incontinence	68	26	40
Urgency	64	45	73
Frequency	40	40	50
Nocturnal enuresis	36	21	40
Constipation	92	32	40
Encopresis	83	50	61

Patient or caregiver subjectively defined improvement, urgency, frequency and constipation with incontinence results based on the presence or absence as reported by the caregiver.

unnecessary or it was refused. Six patients demonstrated a decreased capacity of 7 to 88 cc. Unstable contractions resolved in 8 of 25 patients (32%). However, 5 of the 17 patients (29%) without cessation of urodynamic instability had improvement in symptoms.

Comparison of Symptoms, Physical Findings and MRI Results

There were 20 of 36 patients (55%) who had nonurological neurological symptoms that might suggest a tethered spinal cord (table 3). These symptoms were not predictive of a positive response to surgery. Patients with voiding dysfunction and additional neurological symptoms had an 80% rate of clinical improvement (16 of 20) compared to 62.5% (10 of 16) in those without additional neurological symptoms ($p = 0.24$).

Of the patients 16 (44%) were considered to have an abnormal physical examination (table 4). Any of these physical findings was not predictive of a positive response to treatment since 12 of the 16 patients (75%) in this group improved with surgery, while those with a normal examination had a 70% (14 of 20) improvement rate ($p = 0.74$).

Although no MRIs demonstrated any overt pathological condition, 9 patients had potentially significant findings (table 1). Seven of these 9 patients (78%) improved with filum section, while those with normal MRIs responded with a success rate of 70% (19 of 27) ($p = 0.19$).

There were 11 patients who presented with voiding dysfunction who had a completely normal history, physical examination and MRI. These patients improved at a rate of 54% (6 of 11) after surgery. This is not statistically significantly different from the 72% rate (18 of 25) in patients with voiding dysfunction who had any additional symptoms, neurological findings or variation on MRI ($p = 0.30$, table 7).

TABLE 6. *Improvement in urodynamic parameters in whole cohort*

Urodynamic Parameter	Mean Vol Increase (cc)	Mean Increase (% expected vol)
Bladder capacity	53	16
To first sensation	36	12
To first contraction	42	14
To first desire to void	34	11

These values reflect a demonstrable change despite the exclusion of 7 patients who reported excellent clinical results and refused followup urodynamic testing.

TABLE 7. *Outcome differences in patient groups*

Preop Parameter	% Improvement		p Value
	Normal	Abnormal	
History	63	80	0.24
Physical examination	70	75	0.74
MRI	70	78	0.23
History, physical examination + MRI	54	72	0.30

There were no significant differences in outcomes between patients with no vs subtle findings on neurological history, physical examination and/or MRI, and patients with subtle neurological symptoms with subtle symptoms, physical findings and MRI seemingly improved more than those without such findings but the differences were not statistically significant.

DISCUSSION

Tethered cord syndrome is a clinical syndrome of pain, progressive weakness, orthopedic deformity, and neurogenic bladder and bowel dysfunction as a result of a caudal spinal developmental malformation. Some groups have suggested that the spinal cord can be tethered by an inelastic filum, which creates the clinical entity of tethering without associated MRI findings, and surgical intervention may be beneficial.[8,14,15]

These reports have been sufficiently encouraging to question what the normal position and appearance of the conus medullaris is and whether MRI is able to differentiate normal from pathological conditions.[15] Therefore, the clinician is left with a scenario in which a patient has a debilitating problem and the potential for improvement with a limited and well-tolerated neurosurgical procedure. Controversy arises because of the lack of definitive imaging or functional studies for selecting patients.

The group at our institution has only sought neurosurgical consultation for our most refractory voiding dysfunction cases or those with associated neurological symptoms. We compiled a large series of carefully selected patients with extended followup, which provides insight into this entity and its treatment.

In this study we emphasize the infrequency with which we consider neurosurgical intervention for refractory voiding dysfunction. We think that this entity is rare and spinal surgery should only be considered as a last resort after the failure of prolonged and intense medical management. With 5 pediatric urologists and a full-time nurse practitioner at our tertiary referral center we treat the most difficult patients in the area. Therefore, we believe that our surgical intervention rate of 0.04% strongly reinforces our belief that neurosurgical intervention is an uncommon end point. Unfortunately we were not able to compile data on the number of patients referred who did not undergo surgery but we estimate that the proportion is less than 50%. We firmly believe that our close working relationship with our neurosurgical colleagues is invaluable for ensuring accurate and prudent patient selection.

Given the refractory nature of these cases, we think that the overall clinical improvement of 91% with respect to bowel symptoms and 72% subjective improvement in urinary symptoms represent a satisfactory outcome. This is similar to the urinary improvement seen by others, who noted clinical improvement seen in 71% to 97% of patients.[3,4,7,8] Wehby et al also reported a similar 97% rate of fecal improvement.[8]

Resolution of long-standing incontinence at a rate of 53% for fecal incontinence and 26% for urinary incontinence within 3 months of filum section is consistent with a true surgical effect.

The more subjective declaration of clinical improvement may reflect a reporting bias or a delayed effect of medical intervention but after such prolonged and exhaustive efforts we think that this is less likely. Because the durability of improvement after filum section has been questioned by some groups,[4] we are encouraged by the sustained results in our patients.

We quantified our results with urodynamics since we strongly believe that urodynamic information is mandatory for diagnosing an occult tethered cord. We would never consider neurosurgical intervention without significantly abnormal functional studies. We observed that all of our patients had significant deviations from the norm with average unstable contractions to 40 cm H_2O and the mean cystometric capacity of 55% expected according to the age of each patient. Also impressive was that first sensation to void was at 33% of expected capacity, implying that the sensory component may have a significant role in voiding dysfunction.

Postoperative urodynamic improvements demonstrated improvement in 59% of cases, which is less than the overall rate of clinical improvement. Noguiera et al also noted this effect since they were unable to show a 1:1 correlation with urodynamic and clinical improvement.[16] Four of our 6 patients in whom recorded capacity decreased (range 7 to 88 cc) reported clinical improvement in symptoms. This may speak again to more of a sensory component to the pathophysiology. Therefore, increased volume to first sensation (mean 36 cc) may be a more significant effect than increased cystometric capacity. However, our results may be significantly biased because the 7 of 8 patients who were not tested postoperatively had a good response to surgery.

The second aim of our study was to determine if there are other clinical or imaging predictors of successful intervention. We defined normal history as patients who had been screened by the urologist or nurse practitioner without any obvious neurological symptoms and the subtleties elicited by the neurosurgeon. This parallels the definitions of normal and abnormal physical examination. Neurosurgeons were able to elicit abnormal reflexes and detect minimal asymmetry in the feet, scoliosis or abnormal pigmentation, which were not obvious to the urologist. Nine of our patients had subtle MRI findings of questionable clinical significance. Our neurosurgeon believes that these variations are within the spectrum of normal. Without the associated urinary and bowel dysfunction they would not in and of themselves have been an indication for surgical intervention.

Unfortunately we were unable to definitively identify parameters to predict surgical success. Neurological symptoms demonstrated a trend but they could not predict statistically significant differences in surgical outcome. Likewise patients with subtle symptoms, physical findings and MRI results also trended toward significance but this was likely impaired by the limited number of patients in our study. Physical findings and MRI in isolation seemed least predictive of outcome.

CONCLUSIONS

Diagnosis and treatment of occult tethered cord syndrome will remain controversial until definitive treatment guidelines are established. We observed that in our extremely select population section of the filum terminale resulted in an acceptable outcome in most patients. This effect was seen in cases of urinary and bowel dysfunction with marked immediate and durable results. Unfortunately MRI results, clinical history and physical examination could not improve our ability to predict the patients who would benefit most from surgery. Most importantly we strongly believe that stringent patient evaluation, including urodynamics, is mandatory to identify patients with bowel and bladder dysfunction that is neuropathic in nature before any consideration of neurosurgical treatment. It is also necessary that all attempts at medical management must be exhausted before surgical intervention is considered.

Abbreviations and Acronyms

DES = dysfunctional elimination syndrome
MRI = magnetic resonance imaging

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EDITORIAL COMMENT

Incontinence refractory to behavioral and medical therapy represents one of the greatest challenges to the pediatric urologist. A surgical procedure with the potential for a permanent and drug-free cure is tantalizing for the child, their parents and those who care for them. When considering sectioning of the occult tethered cord as a treatment option pediatric urologists must ask whether it is efficacious and which patients should be referred to the neurosurgeon.

The subjects in this study were well evaluated with preoperative urodynamics in all and postoperative urodynamics in 78%. However, subjective assessment of incontinence and its postoperative improvement was insufficiently evaluated, and no objective measures of incontinence were used.

Inclusion of 9 patients with some degree of radiological abnormality, 20 with symptoms suggestive of tethered cord and 16 with potential signs of neuropathy dilutes the study population of greatest interest to the pediatric urologist, that is the child with refractory incontinence and no signs or symptoms of tethered cord and a completely normal MRI. Of these patients only 52% clinically improved and no urodynamic data were presented separately for this subgroup. The absence of a statistically significant difference in clinical improvement between this subgroup and patients with some abnormality (74%) may be more a function of an underpowered study than of physiology.

Finally, the inexact correlation between urodynamic data and clinical symptomatology is interesting. This finding highlights the reality that urodynamic testing, even in experienced hands, does not always provide an accurate assessment of bladder function in the real everyday world outside the urodynamics facility.

John S. Wiener
Duke University Medical Center
Durham, North Carolina

REPLY BY AUTHORS

We agree that our study was limited by the lack of prospective data collection and standardized questionnaires, and we look forward to further investigation in this manner. This may have compromised our correlation with the urodynamics but may also reflect a dominant sensory component to the disorder. One of the most difficult aspects of this entity is the definition of normal vs abnormal imaging. We cannot be definitely sure what constitutes a normal or clinically significant finding on MRI, or which signs and symptoms are pathognomonic for cord dysfunction (references 5 and 15 in article).[1] Therefore, we chose to consider all of these "shades of gray" in our study and did not find a difference. This result does not imply that we are able to define normal or abnormal, or which patients should be offered therapy but only that we were unable to accurately discriminate between normal and pathological. Therefore, we believe that we have shown that sectioning of the filum terminale is a viable option in the most severe dysfunction elimination syndrome case, regardless of whether the MRI, symptoms or physical findings are completely normal or have subtle variations.

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