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EDITORIAL

# Prone to error, or enlightenment?

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Stamates and colleagues report a moderately large series of patients undergoing evaluation for tethered spinal cord by using MRI to measure ventral excursion of the conus medullaris in the prone position.[6] Based on the widely held theory that spinal cord tethering reflects distal traction on the cord, the authors propose that tethering results in a statistically significant reduction in ventral excursion. The study was meticulously conducted by an expert group and represents a very important contribution to the literature. Nevertheless, its value is limited by a number of factors that are extremely common in published studies of tethered cord (including my own): retrospective analysis, heterogeneous dysraphic anatomy, inadequate or inappropriate controls, mixed age groups, a combination of symptomatic and asymptomatic patients (the latter generally young children with external gluteal abnormalities), and most importantly, a lack of reference to validated outcomes. In fact, Stamates and colleagues define the study's dependent variable, spinal cord tethering, by their own decision to operate rather than by observed natural history, proven response to treatment, or other objective finding.

Stamates and colleagues acknowledge in their abstract that "imaging sensitivity and specificity for tethered cord can be low." In actuality, imaging sensitivity and specificity for tethered spinal cord are impossible to quantify because this entity's diagnostic boundaries are so uncertain. Sensitivity and specificity are meaningful only in the context of the population investigated. Because patient selection for tethered cord surgery on clinical grounds is so variable, and indications for surgery so controversial, sensitivity and specificity measures derived from a moderately sized single-institution study have limited meaning in general practice. As the authors themselves state, "complaints such as back pain or subtle decreases in gait or bladder function are nonspecific and can be difficult to objectively assess clinically."

The positive predictive value and the negative predictive value of a positive or negative test result are probably the most useful descriptive variables for patient counseling and clinical decision making. In this case, the positive predictive value of impaired ventral cord excursion in the prone position is perfect (100%), and the negative predictive value of normal cord excursion in the prone position is very high (91%). In other words, prone MRI in these authors' hands perfectly predicts the occurrence of surgery for tethered or retethered cord, and correctly excludes the diagnosis of tethered cord 91% of the time. This pattern is especially favorable because the authors propose the use of prone MRI for high-stakes surgical decision making. If generalizable, these results suggest that prone imaging will never falsely indicate an unnecessary intervention and will prevent a needed intervention less than 10% of the time. Unfortunately, this only tells us whether prone imaging alone would have accurately indicated detethering surgery in comparison to these authors' own clinical judgment.

Previous studies of children with a significantly low conus and a terminal lipoma suggest that prone imaging does not provide additional diagnostic information to standard MRI.[5,10] Of note, there is relatively little clinical controversy about indicating untethering in these children.[4] By contrast, there is considerable controversy regarding indications for surgery in children with a normal-level conus medullaris and a normal-caliber filum terminale: so-called imaging-negative or occult tethered cord syndrome (OTCS).[3] Prone MRI might therefore be more useful in patients with OTCS. Using prone MRI, Nakanishi and colleagues demonstrated perfect discrimination between patients with OTCS and those with normal spinal cords who were used as a reference group.[1] By contrast, Stamates and colleagues describe highly variable ventral conus motion in a handful of patients with OTCS who were excluded from their principal analysis. Further study is needed to definitively assess the utility of prone MRI in these patients.

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Most experts believe that there is no radiological distinction between clinically well patients and those with retethering after previous repair of major dysraphic abnormalities such as lipomyelomeningocele (other than the occasional appearance of a new syrinx). Defining an MRI parameter that could identify retethering in patients with major dysraphism would therefore be of considerable interest. Unfortunately, in a study of 17 well children performed after recent untethering of mostly major dysraphic defects, Vernet and colleagues observed ventral cord excursion in only 24%.[10] Stamates' patients with clinical retethering (Group 2) also included a large majority with major dysraphic abnormalities (88%). As expected, ventral conus excursion in these patients was significantly reduced compared to reference patients with normal results on lumbar MRI (Group 3). Unfortunately, as the authors acknowledge, this comparison is not meaningful for the relevant clinical question of retethering in a child with major dysraphism.

By contrast, only 2 symptomatic patients in Group 2 had previously undergone filum transection (13%). Although filum retethering has been reported in up to 9% of patients,[11] it is a controversial entity. Again, the surgeon's individual definition of tethered cord recurrence is the true underlying independent variable. Given that the stump of the proximal filum generally retracts beyond the extent of the closed surgical durotomy, at face value the likelihood of filum retethering appears to be quite low. In 513 filum untethering procedures over 17 years, I have performed re-do untethering in only 1 patient (unpublished data).

Although the authors state that patients in their study "underwent operation according to standard criteria that excluded prone imaging," it seems unlikely that imaging obtained during the course of routine care in a retrospective study without investigational consent did not influence clinical decision making. Reference Group 3 may not therefore represent an entirely appropriate comparison, raising concern that the high specificity and sensitivity seen here at least in part reflect circular logic with regard to the definition of tethered spinal cord.

Although this study is inherently flawed, it describes carefully collected and meticulously analyzed information regarding the diagnosis of tethered cord, and represents some of the best information available regarding prone imaging. Nevertheless, ruthless application of logic to the problem of defining tethered cord suggests that an entirely different approach will eventually be needed, with reference to validated long-term outcomes that compare natural history and surgical intervention. A prospective controlled study of filum transection for OTCS failed to achieve funding in the US or to meet enrollment goals in Canada.[7] Modern registry science offers a legitimate alternative approach, with the potential to rapidly enroll relatively large numbers of patients for limited expense. Challenges include the availability of validated disease-specific outcome instruments for tethered cord, enrollment of a matched comparison cohort, and adequate length of follow-up.

In the coming era of population health- and value-based care, all physicians will be called on to justify the efficacy of any intervention based on outcome. Invasive, expensive, and/or risky interventions, such as intradural surgery, will be subject to especially rigorous scrutiny. Although technically simple and very rarely associated with serious complications,[8] filum terminale transection is nevertheless a major intervention. Stamates and colleagues provide us with new and promising data about a relatively convincing way to confirm our own clinical suspicions about the occurrence of symptomatic tethered cord.[6] What we need most, however, is a more reliable tool to predict what will happen to patients with or without surgery. Whether prone MRI will provide such a tool is, at this point, speculation.

We know that indicating spinal cord detethering on clinical grounds alone is based largely on Class III medical evidence.[2,4,9] We also know that patients selected for surgery based on clinical indicators and formal urodynamic studies harbor anatomically[3] and histologically[9] abnormal fila terminale. We also believe that such patients improve as the result of surgery at a rate of approximately 90%, again based on Class III evidence collected largely without the use of validated outcome instruments.[3] Prone MRI obtained to evaluate ventral conus excursion, based on Stamates' results, clearly provides an additional important method that we should subject to prospective, outcomes registry–based analysis.[6] Prone MRI appears to be less likely to add value in the setting of major dysraphism or retethering, and most promising in the setting of minimal or borderline indications for tethered cord surgery, such as OTCS.

I congratulate the authors for an important contribution that, like all good work, clearly identifies the next important questions and opportunities for advancement.

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### Disclosures

The author reports no conflict of interest.

## Response

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Our deepest appreciation goes to Dr. Selden for his thoughtful commentary on our article and his review of controversies surrounding tethered cord in general. We agree wholeheartedly that "further study is needed to definitively assess the utility of prone MRI...." Controversy exists in the diagnosis, definition, and natural history of 2 particular groups: those with OTCS, and those suffering recurrence of symptoms. We aimed to investigate how expanding MRI technology could specifically aid in describing and defining these entities.

Current attempts in the literature to define a diagnosis of OTCS have used anatomical[2] and radiographic[1] comparisons. As it stands, OTCS is defined as a "normal"-level conus with no identifiable spinal abnormality besides spina bifida occulta. The natural history may be a premature question, because patients who underwent tethered cord treatment with filum sectioning for OTCS in a recent randomized, controlled pilot study seemed to show a clinical course similar to patients undergoing medical treatment alone for abnormal urodynamics.[3]

When considering "retethering," or our Group 2 patients, the natural history varies widely. We believe that this largely speaks again to an issue of establishing a firm diagnosis. Speaking strictly on filum sectioning (and excluding lipomyelomeningocele or myelomeningocele), Dr. Selden's rate of retethering is almost nonexistent (0.2%), whereas other authors have noticed enough retethering to classify patients into "early" and "late" subsets.[4] This difference may be attributed to varying levels of diagnostic certainty prior to a repeat untethering procedure, surgeon-specific technical modifications, or perhaps differing characteristics in the populations themselves.

Given the challenges relating to diagnosis of OTCS and retethering, both of these populations would be greatly benefitted by a test that ideally would be simple to perform, noninvasive, cost-effective, and easily repeatable, and that provides 100% sensitivity and specificity. Does such a test exist? We recognize the need for vigorous justification of invasive procedures in an era of cost-conscious care. At our institution, we have found prone MRI to provide one piece of this difficult diagnostic puzzle; however, we have to prove to you, our esteemed colleagues, the merits of an additional radiographic study in the decision to treat or repeat treatment in patients with tethered cord symptoms while questions remain regarding the definition, diagnosis, and natural history of these entities. Our current efforts have now expanded to postoperative studies, and to using these as a baseline going forward as we gather long-term follow-up data in this challenging set of patients.

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