Comparative Effectiveness Review
Number 274

Diagnosis and Treatment of Tethered Spinal Cord

Executive Summary

Main Points

- The strongest evidence exists for MRI accurately diagnosing tethered spinal cord and studies showed moderate to high sensitivity and specificity (moderate strength of evidence).
- A small number of existing studies indicated benefits of prophylactic surgery for motor function and stability of neurological status over time, but it is also associated with complications such as surgical site infection (low strength of evidence).
- A larger body of evidence evaluated various treatments, primarily surgical detethering, for symptomatic patients. Studies reported improvement of neurological status with surgical detethering (low strength of evidence) but it was also associated with post operative complications such as cerebrospinal fluid leakage (moderate strength of evidence).
- A very small body of evidence exists for revision detethering and spinal column shortening for repeat surgery (low or insufficient strength of evidence for all outcomes).

Background and Purpose

Tethered spinal cord is a condition in which the lower end of the spinal cord is abnormally attached to the surrounding tissues, which prevents the spinal cord from moving to keep up with the lengthening of the spine as individuals grow. As a result, there is abnormal tension on the lower spinal cord and associated nerves that leads to neurological injuries and pain in patients. It is most commonly caused by spinal dysraphism, including myelomeningocele, lipomyelomeningocele, diastematomyelia, dermal sinus tract, and thickened/fatty filum terminale. Tethered cord syndrome is a clinical disorder associated with excessive spinal cord tension that leads to motor and

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sensory deficits involving the cauda equina and spinal cord. Many patients initially present in childhood, adolescence, or early adulthood due to the congenital nature of spinal dysraphism disorders. Nevertheless, patients with tethered cord syndrome can present in adulthood and later in life when there is an occult tethered cord with delayed presentation or when patients develop recurrent tethered cord syndrome after prior surgical treatments.

The review will summarize the evidence regarding diagnosis, prophylactic treatment, symptomatic treatment, and repeat surgery of tethered spinal cord. With funding from the Patient Centered Outcomes Research Institute (PCORI), the Agency for Healthcare Research and Quality (AHRQ), commissioned this work to synthesize the findings on the diagnosis and treatment of tethered spinal cord. The systematic review will support the Congress of Neurological Surgeons (CNS) clinical practice guidelines.

Methods

We followed methods outlined in the AHRQ Evidence-based Practice Center Program Methods Guidance.[1] Key informants and a technical expert panel provided input. We searched five databases, two research registries, and four guideline repositories from inception to March 2024. We reference-mined reviews and contacted primary research authors for additional data. Eligibility screening was supported by machine learning. One researcher abstracted the data, and a second reviewer checked for accuracy and completeness. We assessed the risk of bias, strength of evidence (SoE), and applicability.

Results

Searches identified 6,285 citations of which 2,005 were obtained as full text. In total, 103 studies met inclusion criteria, and an additional 355 case series (in 469 publications) provided additional information.

Although multiple diagnostic modalities have been suggested for the diagnosis of tethered spinal cord, we only identified studies reporting on diagnostic accuracy of MRI, ultrasound, myelogram, and evoked potential. None reported on comparative impact of using the test. The strongest evidence existed for MRI and ultrasound. While MRI produced consistently moderate to high sensitivity and specificity (moderate strength of evidence), ultrasound produced more variable results (low SoE).

A small body of evidence evaluated prophylactic surgery and indicated benefits for motor function and neurological function (low strength of evidence). However, prophylactic surgery appeared to be associated with post operative complications such as surgical site infection (low SoE).

A larger body of evidence documented treatment of symptomatic patients, with the most evidence being available for surgical detethering. Surgical detethering may improve neurological status, urinary function, and pain outcomes (low SoE). Earlier detethering surgery may have neurological benefits (low SoE). Surgical detethering was associated with postoperative complications such as cerebrospinal fluid leakage (moderate SoE).

Combined scoliosis and TSC surgery may improve sensory deficits and lead to a more complete recovery (low SoE). Intraoperative monitoring may improve neurological status and may be associated with fewer post operative complications (low SoE). Combined surgery versus staged surgery for different spine conditions may be associated with fewer post operative complications (low SoE). Maintaining patients flat after detethering surgery may not prevent cerebrospinal fluid leakage (low SoE).

A very small body of evidence exists for revision detethering and spinal column shortening for repeat surgery but SoE was low or insufficient for all outcomes, including adverse events.

Limitations

The systematic review identified a large number of studies, but the body of evidence is limited due to the lack of controlled studies and limited reporting of outcomes for diagnostic modalities other than MRI and initial treatments other than surgical detethering. Multiple key outcomes could not be addressed due to insufficient evidence. Despite the large volume of existing research on tethered spinal cord diagnosis and treatment evaluated in this review, better reporting and controlled studies are urgently needed to advance the evidence base for this important clinical condition.

Implications and Conclusions

The evidence base for the diagnosis and treatment of tethered spinal cord is limited with few exceptions (use of MRI or ultrasound for diagnosis, surgical detethering improving neurological status in symptomatic patients, complications associated with open detethering surgery) and would benefit from stronger study designs (tool evaluations reporting diagnostic performance and treatment studies with concurrent comparator).

References

1. Agency for Healthcare Research and Quality (AHRQ). Methods Guide for Effectiveness and Comparative Effectiveness Reviews. Content last reviewed March 2021. Effective Health Care Program. Rockville, MD: 2021. https://effectivehealthcare.ahrq.gov/products/cer-methods-guide

Full Report

Hsieh P, Apaydin E, Briggs RG, Al-Amodi D, Aleman A, Dubel K, Sardano A, Saint-Val J, Zhang D, Blythe K, Sysawang K, Yagyu S, Motala A, Tolentino D, and Hempel S. Diagnosis and Treatment of Tethered Spinal Cord. Comparative Effectiveness Review No. 274. (Prepared by the Southern California Evidence-based Practice Center under Contract No. 75Q80120D00009.) AHRQ Publication No. 24(25)-EHC031. PCORI® Publication No. 2024-SR-04. Rockville, MD: Agency for Healthcare Research and Quality; October 2024. DOI: https://doi.org/10.23970/AHRQEPCCER274. Posted final reports are located on the Effective Health Care Program search page.