Main Points

- The strongest evidence exists for MRI accurately diagnosing tethered spinal cord and studies show moderate to high sensitivity and specificity (moderate strength of evidence).
- A small number of existing studies indicates 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 and the strength of evidence was low.
- A larger body of evidence documents various treatment of symptomatic patients, in particular for surgical detethering. Studies report improvement of neurological status (moderate strength of evidence) but it is 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, but the strength of evidence is low for all outcomes.

Background and Purpose

Tethered spinal cord 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 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 for Neurological Surgeons (CNS) clinical practice guidelines.

Methods

We followed methods outlined in the AHRQ Evidence-based Practice Center Program Methods Guidance where applicable.[1] For this update, we searched PubMed (biomedical literature), EMBASE (pharmacology emphasis), CINAHL (allied nursing), Web of Science (technical innovation), and SCOPUS (general research). We also searched US and international research registries (clinicaltrials.gov, ICTRP) from inception to December 2023 (the search will be updated during public review) to capture all relevant data regardless of the publication status. In addition, we searched the same databases used for primary research plus the Cochrane Database of Systematic Reviews and PROSPERO to systematically identify existing research syntheses. We also systematically searched for existing clinical practice guidelines, using the ECRI repository, G-I-N, MagicApp, and ClinicalKey to inform this protocol.

Based on their clinical and methodological expertise, researchers were assigned to abstract data from each of the eligible articles. One researcher abstracted the data, and a second reviewer checked for accuracy and completeness. Disagreements were resolved by consensus.

Summary tables include ratings for individual strength of evidence domains (i.e., risk of bias, consistency, precision, directness) based on the totality of underlying evidence across studies.

Results

Searches identified 5,827 citations, 1,537 were obtained as full text. In total, 100 studies met inclusion criteria, with an additional 333 case series providing additional information published in 441 publications..

Although multiple diagnostic modalities have been suggested for the diagnosis of tethered spinal cord, we did not identify studies reporting on diagnostic accuracy or the comparative impact of the test with the exception of MRI, ultrasound, myelogram, and evoked potential. The strongest evidence exists for MRI and ultrasound. While MRI produces consistently moderate to high sensitivity and specificity (moderate strength of evidence), ultrasound produces more variable results (low strength of evidence).

A small body of evidence exists that evaluates prophylactic surgery. Studies reported benefits, for example in motor function and neurological function (low strength of evidence). However, prophylactic surgery was associated with post operative complications such as surgical site infection.

A larger body of evidence documents treatment of symptomatic patients, with the most evidence being available for surgical detethering. Across studies, surgical detethering improves neurological status (moderate strength of evidence). Earlier surgery may have more neurological benefits (low strength of evidence). Combined scoliosis and TSC surgery may improve sensory deficits and lead to a more complete recovery (low

strength of evidence). Intraoperative monitoring may improve neurological status, may be associated with fewer post operative complications, and may reduce the need for repeat surgery (low strength of evidence). Surgical detethering may improve urinary function (low strength of evidence). Surgical detethering or spine shortening surgery may improve pain (low strength of evidence). Surgical detethering is associated with postoperative complications such as cerebrospinal fluid leakage (moderate strength of evidence). Combined surgery versus staged surgery for different spine conditions may be associated with fewer post operative complications (low strength of evidence). Maintaining patients flat after detethering surgery may not prevent cerebrospinal fluid leakage (low strength of evidence). The need for repeat surgery may be reduced with combined surgery versus staged surgery for different spine conditions (low strength of evidence.

A very small body of evidence indicates that neurological status benefits may be associated with revision detethering and spinal column shortening for repeat surgery (low strength of evidence). Spinal column shortening may be associated with improved bladder or bowel functioning and pain (low strength of evidence). There is a lack of evidence on other 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. Despite the documented research volume on tethered spinal cord diagnosis and treatment 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.

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

[to be inserted in the final report]