

Overview of spina bifida and the nervous system

Chapter 3: Spinal cord tethering

Tethering of the spinal cord is an insidious cause of worsening incontinence that requires urgent medical assessment and possibly surgical intervention to prevent any further deterioration. Clinicians monitoring young people and adults with spina bifida need to maintain a high degree of suspicion to ensure the early detection of tethered cord syndrome to help promote and maintain independent living.

Key issues for clinicians

- Clinicians need to consider spinal cord tethering as a possible cause of change in continence patterns.
- Any suspicion of spinal cord tethering should be referred to a specialist centre for urgent assessment.
- Earlier surgical intervention in clinically demonstrated spinal cord tethering is more likely to result in an improved long term outcome.

Tethered cord syndrome — an insidious condition

Tethering or stretching of the spinal cord in young people and adults results from fixation of the spinal cord to inelastic structures.¹ Spinal cord tethering is a major source of morbidity in spina bifida and clinicians need to be familiar with its presentation and maintain a high degree of suspicion when monitoring patients with spina bifida.

In the past, spinal cord tethering was thought of as mainly a condition affecting only children, especially during growth spurts, but the condition can occur in people with spina bifida at any age.

Spinal cord tethering is a particularly insidious complication of spina bifida that can have a major adverse impact on independence through its effect on mobility and continence.

Many causes but consistent presentation

The fixation of the spinal cord in spinal tethering is due to fibrous or lipomatous tissue. In spina bifida this can be due to a myelomeningocele, lipomyelomeningocele, scar tissue, a fibroadenomatous filum terminale or many other conditions.²

Clinical presentation of tethered cord syndrome

Symptoms and signs

Tethered cord syndrome typically causes a progressive loss of function at or below the level of the spinal cord defect, with or without lower lumbar pain.

Regardless of the particular mechanical cause of spinal tethering, the spectrum of clinical presentations of tethered cord syndrome are consistent and should alert the clinician to the need for IMMEDIATE neurological and neurosurgical referral.

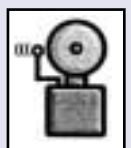


Table 3. Characteristics of pain in spinal cord tethering

- Pain in lower back and legs exacerbated by physical activity, especially any which involves flexion and extension of the lumbosacral area
- Leg pain is often in medial, lateral, anterior, or posterior aspects of thighs or legs
- Groin pain or pain in the genitorectal area is common
- Pain is not less when lying supine (as opposed to disc disease)
- Straight leg raising causes no difference to pain
- In adults, pain governed by three 'B' signs (usefulness will depend upon any pre-existing neurological and orthopaedic disabilities):
 - inability to sit with legs crossed (like Buddha)
 - difficulty in bending slightly at the waist with activities such as washing dishes
 - holding a baby or light material (< 2.5 kg) at the waist level while standing

Table 4. Common findings in tethered cord syndrome

- Pain (*Table 3*)
- Gait problems, often quite subtle
- Progressive urinary incontinence; symptoms include urgency, frequency and enuresis
- Progressive faecal incontinence including urgency, frequency, and encopresis
- Any motor or sensory deficits in lower extremities
- Altered sensation in genital regions and during sex
- Impotence
- Muscle weakness
- Muscle atrophy
- Hyporeflexia, especially any change in pre-existing signs
- Faecal incontinence
- Scoliosis/lordosis
- Foot deformities
- Skin abnormalities — herald marks

Tethering may occur in young adults at times of growth spurts, when lengthening of the spinal column can increase spinal cord tension, but it can occur at any adult age.

Key diagnostic issue — progressive loss and change

The key diagnostic issue in spinal cord tethering in young people and adults with spina bifida is a progressive deterioration in neurological function at or below the level of the defect. This includes urinary and faecal incontinence.

Spinal tethering needs to be excluded in the presence of progressive deterioration of urinary or faecal incontinence.

Neurological deficits in spina bifida are usually not progressive. Any change in signs requires immediate specialist assessment.

Spinal cord tethering can also occur in adults with no known past history of spina bifida.

Patchy distribution of clinical findings of tethered cord syndrome

The pattern of clinical findings of spinal cord tethering often fails to follow strict dermatomal patterns like those due to compression of one or two nerve roots or a particular level of spinal cord injury. Weakness, pain and

other signs of spinal cord tethering may have a patchy distribution below the level of the lesion, rather than a strict neurotomal pattern.

Pain in spinal cord tethering

Regardless of the cause, the characteristics of the associated pain are often suggestive of spinal tethering (*Table 3*).

While other causes of back pain, such as disc herniation, need to be excluded, clinicians still need to organise urgent neurosurgical referral and assessment to ensure that spinal cord tethering is not missed.

Other common findings in tethered cord syndrome

As highlighted above, the findings need to be considered in the context of any pre-existing neurological abnormalities, but clinicians should have a low threshold for specialist referral when there are any changes in pre-existing clinical findings (*Table 4*).

Management

Diagnostic imaging

Magnetic resonance imaging

Magnetic resonance imaging (MRI) is the best currently available technique for viewing the spinal

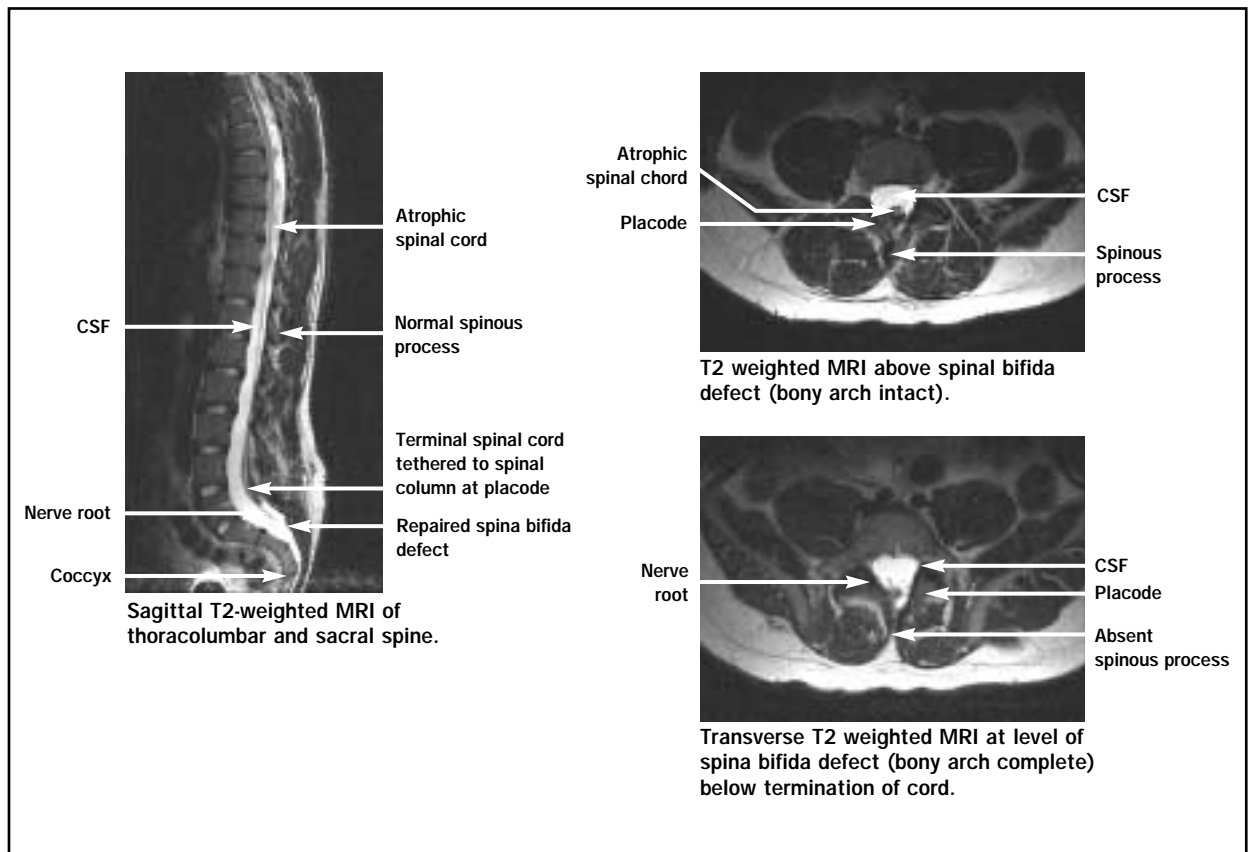


Figure 5. Surgically repaired spina bifida defect with absent spinous process and posterior spinal arch at low sacral level. Atrophic cord tethered to placode and spinal column. (Courtesy of Department of Diagnostic Imaging, Monash Medical Centre, Melbourne.)

cord. Most people with spina bifida will have some MRI findings suggestive of spinal tethering, but the decision to treat is based upon a combination of MRI and clinical findings. Ideally, a baseline MRI should be taken on all young people and adults with spina bifida to compare any changes with subsequent imaging, should symptoms of tethering arise (*Figure 5*).

Typical MRI findings in spinal cord tethering

Typical MRI findings in spinal cord tethering include

- thick filum terminale (>2 mm in diameter)
- presence of structures such as fibrolipomatous filum terminale
- obliteration of subarachnoid space suggesting caudal spinal cord or nerve root adhesion
- changes in the structure of the spina bifida lesion such as dermoid, epidermoid cyst, myelomeningocele, lipomyelomeningocele or other problem
- elongation of spinal cord
- posterior displacement of conus medullaris with the filum pressing against the thecal lining at or near L5, or when compared with previous films.

Surgical intervention

Indications for detethering the cord

Once the diagnosis of spinal cord tethering is made, the decision for surgical intervention is based on clinical evidence. Treatment is especially indicated in the presence of new or worsening symptoms.

In studies of adults, when performed by experienced neurosurgeons, spinal cord tethering has been demonstrated to be a well tolerated, effective intervention.

Ultimately, the neurosurgeon can only confirm the presence of spinal tethering at operation. The surgery performed depends upon the intraoperative findings.

Effect of surgical intervention on spinal cord tethering³⁻⁵

Timely surgical intervention of tethered cord syndrome can arrest, and in some cases improve neurological signs. The more long standing the neurological signs, the less the chance of resolution of symptoms and signs.

Pain improves in many people and is usually relieved within three months of the detethering, but improvements in neurological signs can take many more months.

Patients need constant monitoring after the operation to assess recovery and help to adjust to any lifestyle changes.

Case history: a 27 year old woman with worsening incontinence.

M is a 27 year woman with spina bifida. She is independently mobile, has never used calipers, has no hydrocephalus, is fully employed, and is in a steady relationship

M presents with 12 months of progressively worsening faecal incontinence. She is now freely incontinent of faeces and manually evacuates herself before going out to avoid humiliation.

On examination, the anal tone is grossly reduced and at the time of assessment, anal sphincter reconstruction was being considered.

She has also had mild urinary frequency and urgency for six months, as well as eight months of reduced vaginal sensation during intercourse.

Her MRI demonstrated the presence of a tethered cord with a large neural placode. M was referred to a neurosurgeon and detethering is now planned.

References

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