

# Spinal Cord Tethering

A common cause of deterioration in Spina Bifida.

## What is spinal cord tethering?

Tethering of the spinal cord is a condition in which the spinal cord becomes attached to the spinal column via surrounding structures. Normally, the spinal cord hangs loose in the canal, freely moving up and down with growth, bending and stretching. A tethered cord does not move. It is pulled tightly at the end, reducing blood flow to spinal nerves and causing damage to the spinal cord from both the stretching and the decreased blood supply.

Tethering can happen before or after birth in children and adults; and most often occurs in the lower (lumbar) level of the spine. All forms of SB can be accompanied by spinal cord tethering; but it rarely occurs with Spina Bifida Occulta (SBO). In children, a tethered cord causes the spinal cord to stretch as the child grows. In adults, the spinal cord will stretch during the course of normal activity which like bending and stretching. If a symptomatic tethered cord (tethered cord syndrome) is left untreated, it can lead to progressive, permanent spinal cord damage.

## What is tethered cord syndrome?

Tethered cord syndrome is the presence of several clinically recognizable signs (observed by a physician), or symptoms (reported by the patient) that occur together as a result of the tethering. These signs and symptoms can include: sensory disturbance, significant muscle weakness (as determined by neuro assessment), pain, and incontinence.

## How does tethering occur in Myelomeningocele?

During the early stages of a pregnancy, the spinal cord of the fetus extends from the brain all the way down to the coccygeal (tailbone) region of the spine. As the pregnancy progresses, the bony spine grows faster than the spinal cord, so the end of the spinal cord appears to rise relative to the adjacent bony spine. By the time a child is born, the spinal cord is normally located opposite the disc between the first and second lumbar vertebrae, in the upper part of

the lower back. In a baby with Spina Bifida, the spinal cord is still attached to the surrounding skin, and is prevented from ascending normally.

The spinal cord at birth is low-lying, or tethered. Although the myelomeningocele is surgically separated from the skin and closed at birth, the spinal cord, which has grown in this position, stays in roughly the same location after the closure, and quickly scars to the site. As the child (and the bony spine) continues to grow, the spinal cord can become stretched; damaging the spinal cord both by directly stretching it, and by interfering with the blood supply to the spinal cord. The result can be progressive neurological, urological, or orthopedic deterioration.

## How does tethering occur in milder forms of Spina Bifida?

Tethering, usually in adults with milder forms of SB may be related to the degree of strain placed on the spinal cord over time, and may appear or be significantly worsened during physical activity, injury, or pregnancy. It may also be caused by narrowing of the spinal column (stenosis) or bony spurs.

A tethered cord may go undiagnosed until adulthood. Delayed presentation of symptoms can be insidious, meaning that symptoms come on slowly over time, but can be complex and severe. Back pain, brought on or worsened by activity and relieved with rest, can be a sign of tethering.



Sometimes back pain is also associated with leg pain, even in areas that have decreased or no sensation. Changes in leg strength, deterioration in gait (walking), progressive or repeated muscle contractures, orthopedic deformities of the legs, scoliosis, and changes in bowel or bladder function may be signs of tethering.

### How is a tethered cord diagnosed?

If a child with myelomeningocele and shunted hydrocephalus presents with clinical worsening, the first issue is to determine whether or not the shunt is working, as shunt malfunction can appear the same as a tethered cord. So, always check the shunt first! Accordingly, the first test is usually a Computed Tomography (CT) or Magnetic Resonance Imaging (MRI) scan of the brain. Once the shunt is found to be working, or for those who do not have a shunt, an MRI of the spine is performed. It is important to know that virtually every child with Spina Bifida has evidence of tethering on the MRI. Untethering is therefore performed only if there are clinical signs or symptoms of deterioration.

The MRI is done to show the neurosurgeon the anatomy of the tethering, and to exclude other abnormalities such as a syringomyelia (syrinx), split cord malformation, or a dermoid cyst (small tag of skin in the area around or within the spinal cord). Additional studies may include spine X-rays or CT scans of the spine to look for other bony abnormalities, or to follow the progress of scoliosis. Other functional studies may be done, including Manual Muscle Testing (MMT) and urodynamics. Both are compared with previous studies to document changes, and to provide a pre-surgical baseline.

### When is surgery performed?

After reviewing the diagnostic studies, the neurosurgeon may decide to untether the spinal cord. The decision to untether requires some clinical judgment. The neurosurgeon considers the patient's symptoms, signs, and the results of the tests. Virtually every child with Spina Bifida has evidence of tethering on the MRI. Untethering is generally only done if there is clinical evidence of deterioration, progressive or severe pain, loss of muscle function, deterioration in gait, or changes in bladder or bowel function. The longer deterioration continues, the less likely it is that function will return.

### What happens after surgery?

Recovery in the hospital is generally about 2-5 days, and the patient often returns to normal activity within a few weeks. Some surgeons require the patient to remain flat in bed for a couple of days to minimize the risk of spinal fluid (CSF) leakage from the wound. Pain is often not severe,

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because there is usually some degree of numbness in that area anyway. Recovery of lost muscle and bladder function is variable, and depends on both the degree and length of the neurologic losses before the surgery. Untethering is designed primarily to prevent further deterioration, not to improve spinal cord damage that has already occurred. Modern microsurgical equipment and techniques have made untethering a relatively routine surgical procedure, in the hands of an experienced neurosurgeon

### Is there any treatment other than surgery available for a symptomatic tethered cord?

Although medications, alternative therapies, rest, and physical therapy "may" provide temporary relief from pain, the only successful intervention is untethering surgery.

### What are the complications of untethering?

Untethering is generally a safe procedure. However, the scar can make dissection difficult; and the abnormal anatomy can be confusing for any surgeon. Complications are few, but may include: 1) infection, 2) bleeding, 3) damage to the spinal cord and myelomeningocele, resulting in worsening of muscle, bladder, or bowel function. The combined complication rate of surgery is usually only 1-2%. Although some have suggested that shunt malfunction may occur secondary to untethering surgery, it is more likely that an occult or unrecognized shunt malfunction was the original cause of the deterioration in the first place.

### Is repeat untethering necessary?

Although most children require only one untethering procedure, some (perhaps 10-20%) require repeated untethering operations as the child continues to grow. The most problematic period is the pre-adolescent period (7-12 years). Since all children grow, it is not clear why some develop symptoms and signs of tethering, while others don't; perhaps some children's spinal cords tolerate a greater degree of stretching than others. In adulthood, clinical deterioration from tethering becomes much less frequent (although it can still occur). In adulthood, re-tethering may occur from scar tissue, spinal deterioration, or injury.

### Can tethering be prevented?

Many techniques have been tried to prevent or minimize tethering, but only surgical untethering has been successful in long term studies. Research continues into this important area. With close observation, it should be possible to diagnose this condition early and untether the cord before progressive and permanent damage occurs.