

FACTSHEET

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Tethered Cord and Spina Bifida

What is Tethered Spinal Cord Syndrome?

For people living with Spina Bifida their spinal cord at birth is usually low-lying or tethered. This is due to the nature of the development of nerves and spinal cord.

The initial surgical repair by the neurosurgeon to close the skin after birth means the myelomeningocele is surgically separated from the skin. The spinal cord will more or less stay in the same position and often scar tissue will form around it. In some cases the spinal cord is still attached to the skin and is prevented from ascending normally.

A tethered cord does not move. It is pulled tight, whereas normally the spinal cord hangs loose in the spinal canal. The spinal cord can become stretched as a person living with spina bifida grows. This damages 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 orthopaedic deterioration that leads to loss of regular function.

Tethered cord syndrome can occur at any stage through life. It usually presents with a combination of symptoms which can include:

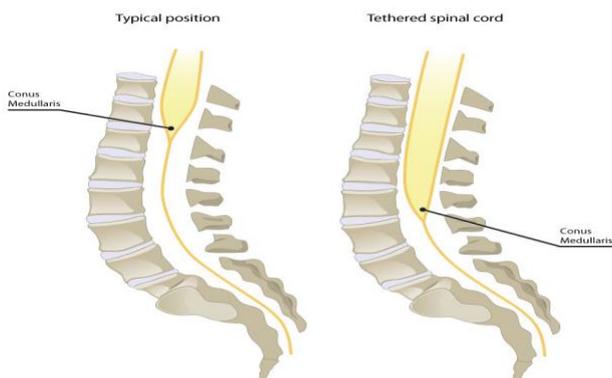
- Pain (back pain or pain radiating into the legs)
- Sensory changes
- Motor changes such as:
 - Progressive weakness
 - Changes in the shape of the feet
 - Progressive scoliosis
 - Loss of bowel and bladder control.

These symptoms are related to the degree of strain placed on the spinal cord over time. They may be worse during sports or pregnancy. The symptoms may also be due to narrowing of the spinal column (spinal canal stenosis) or bony spurs that develop with age.

Tethered cord symptoms can also occur in people without the presence of Spina Bifida. This happens when the cord for unknown reasons has become stuck within the spinal canal. Signs on the skin (stigmata) that could suggest a tethered cord can include:

- Tuft of hair in the midline over the spine
- Discoloration of the skin (birthmark)
- Dimple
- Small lump of fatty tissue on the back.

In people with these stigmata an ultrasound scan or plain x-ray of the spine could be performed to check for underlying incompletely fused vertebrae (a gap or opening in the spine) and abnormally low lying spinal



cord. Further imaging with an MRI is usually done to get more detailed information about the spinal cord.

Can anything be done to prevent tethering?

Many methods have been tried to prevent or reduce tethering, but none have been as successful as surgery. The tethered cord is a common condition and is treatable.

Close monitoring including regular medical reviews and assessments by the physiotherapist (manual muscle testing) means it should be possible to pick up symptoms early. Giving a chance to untether the cord before progressive and permanent damage occurs. The decision to de-tether the spinal cord is made based on presenting symptoms. Most people with spina bifida will have a tethered cord on the MRI. The decision to perform surgery to untether the cord requires clinical judgment. After reviewing the studies, the neurosurgeon will consider the patient's symptoms in combination with the results of tests. Untethering is usually only done if there is clinical evidence of loss of motor function, significant or progressive pain, or deterioration to bladder or bowel function. Discuss any symptoms and how they affect your function with your spina bifida clinic prior to seeing a neurosurgeon.

People living with shunted hydrocephalus need to have shunt malfunction (blocked shunt, broken tubing) ruled out first. Usually a CT scan of the brain will be done. For people without a shunt, or if the shunt is found to be working normally an MRI of the spinal cord is usually done. This is to check other reasons why someone could be having functional or neurological changes.

What are the complications of untethering?

Most people living with Spina Bifida will show some tethering on an MRI (radiological tethering). Untethering surgery will only occur if there are physical signs and symptoms of change (symptomatic tethering).

Risks of surgery are low, but may include:

1. Infection
2. Bleeding
3. Damage to the spinal cord and myelomeningocele. This could result in worsening muscle, bladder, bowel and sexual function.
4. Cerebrospinal fluid (CSF) leak.

Chances of full recovery from any deterioration reduce the longer this condition is left untreated. Untethering surgery aims to prevent further deterioration.

Is repeat untethering necessary?

Symptomatic tethering can occur at any time through life. Symptoms from tethering can often occur during periods of rapid growth. Since all children grow, it is puzzling as to why some children develop symptoms and signs of tethering, while others don't.

Most children require only one untethering procedure. A minority, perhaps 10-20%, require repeated untethering operations as they continue to grow.