Tethering Spinal Cord

Spinal cord tethering is a common cause of deterioration in a child with Spina Bifida. Although the exact frequency with which it occurs isn't entirely known, it is estimated that from 20-50% of children with Spina Bifida will, at some time, require surgery to untether the spinal cord, making this operation the second most common operation (behind shunt operations) in these children. This article will address some of the common questions that have been raised about spinal cord tethering.

What is spinal cord tethering, and why is it bad?
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, or ascend, 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 about 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 therefore low-lying, or *tethered.* Although the myelomeningocele is 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 usually quickly scars to the site of the surgical closure. As the child (and the bony spine) continues to grow, the spinal cord can become stretched; 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 orthopedic deterioration.

What are the symptoms and signs of spinal cord tethering?
Children with spinal cord tethering may develop many different symptoms and signs. Conversely, many of the symptoms and signs of tethering can be caused by other problems, and the neurosurgeon needs to sort out what are the likely causes of the signs and symptoms in each case. Back pain, typically brought on or worsened by activity and relieved with rest, can be a sign of tethering. Sometimes the back pain is also associated with leg pain, even in areas that are numb. Changes in leg strength, or deterioration in gait (walking) can be signs of tethering. Manual Muscle Testing (MMT) of muscle strength is usually performed by physical therapists experienced in performing these tests, and can detect muscle weakness; these tests should ideally be performed on at least an annual basis, or whenever there is a change, to document changes before they become severe. The MMT has been shown to be a very reliable test of muscle strength if performed by experienced therapists. Progressive or repeated muscle contractures or orthopedic deformities of the legs, and scoliosis, may be signs of tethering as well. Finally, changes in bowel or bladder function can be signs of tethering; urodynamic studies, which provide an objective test of bladder function, can be very helpful in determining whether the changes are significant, and can sometimes detect bladder changes before they become apparent clinically.

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 cause *any* of the signs or symptoms discussed above – one should therefore *always check the shunt first!* Accordingly, the first test is usually a Computed Tomography (CT) or Magnetic Resonance Imaging (MRI) scan of the brain. Although an increase in the size of the ventricles of the brain (that contain the cerebrospinal fluid) suggests a shunt malfunction, it is important to know that as many as 10-15% of children with shunt malfunction may have little or no change in the size of the ventricles. In some cases, the shunt is tapped (by inserting a needle through the skin into a 'tapping chamber' for this purpose) and the flow of cerebrospinal fluid from the shunt, and the shunt pressures, are measured as another means of assessing shunt function. If there is any question about shunt function, the neurosurgeon may explore or revise the shunt at surgery before considering an untethering operation.

Once the shunt is found to be working, 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 for the reasons discussed above; untethering is therefore generally only performed if there are *clinical signs or symptoms of deterioration.* The MRI is obtained both to show the neurosurgeon the anatomy of the tethering, and to exclude other abnormalities such as a syringomyelia - or syrinx for short (a fluid filled cavity within the spinal cord); diastematomyelia, or split cord malformation (in which the spinal cord is split into two halves over a part of its length, with a bony spur between the two halves of the spinal cord); or a dermoid cyst (in which a small tag of skin that was enclosed within the area around or within the spinal cord). Although MRI is the imaging study most commonly used, additional studies may include spine X-rays or CT scans of the spine, to look for various other bony abnormalities, or to follow the progress of scoliosis. Again, other functional studies may be done, including MMT and urodynamics, both to compare with previous studies to document a change, and to give a baseline against which to compare after the surgery.

When and how is surgery performed?
After all of the diagnostic studies have been performed, the neurosurgeon may want to untether the spinal cord. The decision to untether requires some clinical judgment on the part of the neurosurgeon, who must take into account both the patient's symptoms and signs and the

results of the pre-operative studies. Unfortunately, since the MRI almost always shows tethering radiographically, the decision usually relies on the neurosurgeon's judgment as to what is causing the patient's symptoms and signs. A child with mild back pain who is otherwise stable might reasonably be watched or managed without surgery as long as they remain stable and the pain is manageable. On the other hand, progressive or severe pain, loss of muscle function or deterioration in gait, or changes in bladder or bowel function usually require an operation to prevent further deterioration. Timing of surgery is important, as the longer deterioration is allowed to continue, the less likely function will return to its baseline with surgery; the timing depends upon the magnitude and rapidity of the changes.

The untethering procedure usually involves opening the scar from the prior closure; occasionally, an incision may be made perpendicular to the original scar, particularly if the original closure was horizontal on the back. The scar is dissected down to the covering (dura) over the myelomeningocele; often the dissection includes the more normal covering just above the scarred area to obtain landmarks and orientation. Sometimes a small portion of the bony vertebrae (the laminae) are removed to obtain better exposure or to decompress the spinal cord. The dura is then opened, and the spinal cord and myelomeningocele are gently dissected away from the scarred attachments to the surrounding dura. There are many methods for doing this including scissors, scalpels, and various lasers; one way is not necessarily better than the others, and the surgeon usually has his or her own preference based upon their experience. Once the myelomeningocele is freed from all of its scarred attachments, the dura and the wound are closed.

Recovery in the hospital is generally about 2-5 days; some surgeons require that the child remain flat in bed for a couple of days to minimize the risk of spinal fluid leakage from the wound. Pain is usually not severe, as the child usually has some degree of numbness in that area anyway. The child is usually back to fairly normal activities within a few weeks. Recovery of lost muscle and bladder function is variable, and again depends upon both the degree and length of the pre-operative losses. Although we hope for improvements, it is important to understand that untethering is designed primarily to prevent *further* deterioration, rather than to improve deterioration that has already occurred.

What are the complications of untethering?
Untethering is generally a very safe procedure in experienced hands; however, the scar can make dissection difficult and the abnormal anatomy can be confusing at times, even to the experienced neurosurgeon. Complications are few, but include 1) infection, 2) bleeding, 3) damage to the spinal cord and myelomeningocele resulting in worsening 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 probably more likely that an occult or unrecognized shunt malfunction was the original cause of the deterioration in the first place.

Is repeat untethering necessary?
Symptomatic tethering can occur at any time in the child's life, although the most common time is in the early pre-adolescent period (7-12 years) and extending into mid-adolescence. Symptoms from tethering can often occur during periods of growth, as might be expected, or during growth hormone treatments for short stature. Since all children grow, it is puzzling as to why some children develop symptoms and signs of tethering, while others don 't; perhaps some children's spinal cords are more lax early in life, or tolerate a greater degree of stretching than others.

Although most children require only one untethering procedure, a minority (perhaps 10-20%) require repeated untethering operations as the child continues to grow. Those who undergo untethering very early in life (as toddlers or young children) may more frequently require additional untethering procedures later, as they continue to grow. Fortunately, once the child stops growing, and their ‘adult, height is reached, clinical deterioration from tethering becomes much less frequent (although it can still occur).

Can anything be done to prevent tethering?
Many techniques have been tried to prevent or minimize tethering, but none has met with unqualified success in long term studies. Surgeons have placed grafts of various substances such as Dacron, Teflon, and other materials around the myelomeningocele hoping to prevent scarring to the surrounding dura. Some of these have actually produced more scarring. Others have cut off the myelomeningocele (cordectomy) if the child has no leg function, hoping to eliminate the scar from the myelomeningocele (however, this technique carries a risk of worsening bladder function and obviously eliminates any hope of recovering any functions if something in the future were to allow this). Most neurosurgeons now bring the edges of the flat myelomeningocele ‘placode' together during the initial closure, and sew the edges together to re-create the spinal cord in order to minimize formation of scar. Although this makes subsequent untethering easier and less risky should it need to be done, neither this nor any other technique have been proven to reduce the frequency of subsequent tethering in the long run. Research continues into this important area.

In conclusion, although the tethered cord is a common condition requiring surgery, it is also very treatable. Modern microsurgical techniques and the availability of such techniques as the operating microscope and lasers have made this a relatively routine surgical procedure in the hands of an experienced neurosurgeon. With close observation, it should be possible to diagnose this condition early and untether the cord before progressive and permanent damage occurs.