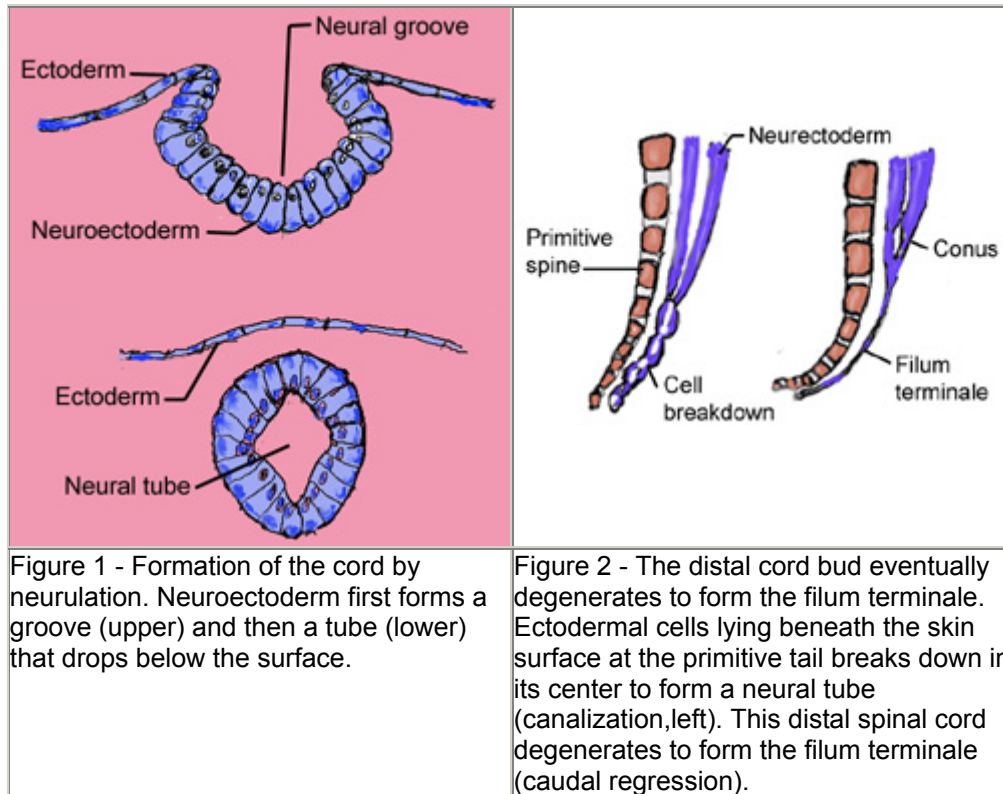


# Laminectomy for Tethered Cord

A tethered spinal cord is characterized by an abnormal attachment of the spinal cord to the surrounding tissues. The lower end of a normal spinal cord is found in the upper lumbar spinal canal. The attachment to the spinal cord usually causes the spinal cord to end lower in the lumbar or sacral spinal canal. In the majority of cases it is congenital (the patient is born with it).

## Developmental Anatomy

- In the developing fetus the spinal cord is formed by a complex process. It forms from the same layer of cells that later form the skin (called the ectoderm)
- Beginning around the 18th day of development and extending to the 22nd day, the spinal cord is formed by a process called neurulation
  1. The ectodermal layer thickens in the midline of the back forming neuroectoderm that first forms a groove and then a tube that drops below the surface to be later protected by bone and muscle (Figure 1)
  2. As the tube drops below the surface, skin closes over the tube
  3. The tube begins to close first in the thoracic (chest) region and the closure spreads toward the head (to eventually form the brain) and towards the upper lumbar region
- The lowest part of the spinal cord is formed by a different process called canalization and caudal regression during the 28-48th day of development (Figure 2)
  1. A group of ectodermal cells lying beneath the skin surface at the primitive 'tail' begins to break down in its center to form a neural tube (canalization)
  2. This tube then fuses with the neural tube formed by neurulation
  3. This distal spinal cord degenerated to form the filum terminale (caudal regression)
  4. This process is less precise and therefore more liable to defects than neurulation. Failure of degeneration creates the entity of tight filum terminale
- Beginning about 45th day, a third process called regression occurs and extends into the first year of life
  1. The lower neural tube then forms into the lowest part of the spinal cord (conus medullaris), the nerve roots that go to form the nerves to the legs (cauda equinae) and a fibrous cord from the conus medullaris to the sacrum (filum terminale)
  2. By the process of regression and a greater growth of the bony spine than the spinal cord, the conus medullaris eventually ends up at the level of the second lumbar vertebrae



## Pathology

- Any event, which interferes with the development of the spinal cord and cauda equinae, can lead to the ingrowth of other tissues like fat and skin, which creates an abnormal attachment, or tethering of the spinal cord
- Tethering of the spinal cord interferes with the normal regression process causing damage to the spinal cord as it is stretched and placed under abnormal tension
- The tension injures the spinal cord and may cause symptoms
- Other entities can be associated with tethering including tumors, cysts, tracts and spinal cord malformations. There are several distinct entities forming a spectrum of diseases. Each may occur separately or grouped together in the same patient
  1. Dermal sinus is a tract lined by skin cells that leads from the skin to anywhere along the back of the spine. The tract may end anywhere to just below the skin to the spinal cord. In the latter situation tethering can occur. This connection can be well developed and lined by skin posing a risk of infection of the nervous system
  2. Lipoma or lipomyelomeningocele (lipo, fat; myelo, spinal cord; meningocele, thin ballooning out of the coverings of the spinal cord) are malformations in which a fatty tumor (lipoma) under the skin is fused to the back of the lower spinal cord. The degree of abnormality can vary from a simple lipoma attached to the cord to a more complex connection. The lipoma may extend to just under the surface of the skin and present as a soft lump in the back. The fat may engulf nerve roots and extend deep within the cord making complete removal extremely difficult or impossible. They are usually slow growing but growth rates can vary.

3. Epidermoid or dermoid cyst is similar in formation as a dermal sinus but a tumor is formed somewhere along the tract. The dermoid cyst is a benign cystic tumor formed from skin elements. Epidermoids are filled with a pearly white material called keratin
  4. Diastematomyelia (split spinal cord malformation) involves the upper portions of the cord. A diastem is bony or cartilaginous spicule, which divides the spinal cord in two. There are a number of varieties of split cord malformations. In some cases there are two separate spinal cords (diplomyelia)
  5. Tight filum terminale syndrome is characterized by a thickened filum terminale over 2 mm. in diameter, a low lying conus medullaris (below L2) and no other cause for the tethering
  6. Neurenteric cyst is a cyst lined by tissue similar to the gut or airway. It has a connection to the spinal cord, vertebrae or both. The spinal tumor may connect via a stalk to mesentery or gut
  7. Myelocystocele is a complex malformation of the distal tethered spinal cord in which the end of the spinal cord is ballooned into a cystic and associated with syringomyelia. It is often associated with malformations of the bladder and anus
  8. Syringomyelia is a dilatation of the central portion of the spinal cord. The dilatation may involve a segment of the cord, the whole cord or be multiple
  9. Retether following myelomeningocele repair. A myelomeningocele is a form of spina bifida (split spine) in which a segment of the spinal cord is exposed along the midline of the back and requires repair. Frequently the spinal cord gets tethered by scarring to the area of repair
- The skin over the tether is intact and usually the surrounding bone of the spine is incomplete (spina bifida occulta). It is called an occult spinal dysraphism (OSD). The opposite is true in the open spinal dysraphic state or myelomeningocele
  - Spinal cord tethering may be either primary or secondary. Primary is typically a form of OSD while secondary usually occurs following a myelomeningocele repair or other type of spinal cord surgery

### History and Exam

- Tethered spinal cord is most commonly diagnosed in infancy by the discovery of a abnormality noticed on the skin of the back. This is called cutaneous stigmata and found in 70% of patients with OSD. These may include:
  1. Hypertrichosis is a hairy patch commonly seen with diastematomyelia and split cord malformations
  2. Capillary hemangioma (red patch made up of capillaries)
  3. Nevus (discolored area) (Figure 3) or nevoid areas (Figure 4)
  4. Atretic meningocele is a small silvery patch of skin overlying a rudimentary meningocele (Figure 5)
  5. Dermal sinus, which looks like a small dimple in the skin if the midline back. (Figure 6) (It must be distinguished from a sacrococcygeal pit or sinus tract located at the coccyx (lowest bones of the spine) and usually not associated with OSD.) (Figure 7)
  6. Subcutaneous lipoma (Soft fatty tumor under the skin usually associated with a lipoma extending to the spinal cord.) (Figure 8)
  7. Caudal appendages (skin tags, tails and pseudotails) (Figure 9)
- Later in life symptoms of pain and nerve deficit predominate
- Pain may be present either in the back or radiating down the legs

- Weakness or loss of sensation in the lower extremities is often associated with alteration in gait
- Urologic complaints of a change in or loss of bladder function and recurrent urine infections may be present
- Change in or loss of bowel function
- The neurologic examination often reveals foot abnormalities and at times asymmetry (Figure 10). Weakness or sensory loss may also be demonstrated
- The spine may develop an abnormal curvature called scoliosis

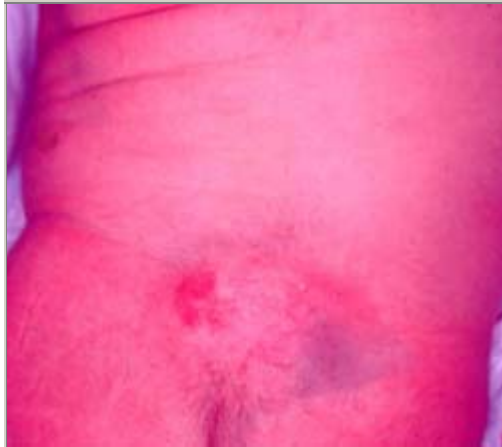


Figure 3 - Capillary hemangioma over the low back. Courtesy S. Schneider, MD



Figure 4 - Nevus. Courtesy S. Schneider, MD





Figure 7 - Sacro-coccygeal dermal sinus tract (arrow). Courtesy S. Schneider, MD

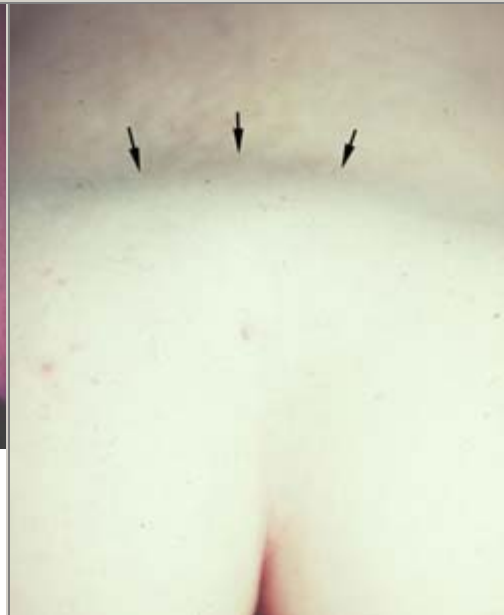


Figure 8 - Subcutaneous lipoma (arrows). Courtesy S. Schneider, MD



Figure 9 - Tail-like appendage. Courtesy S. Schneider, MD


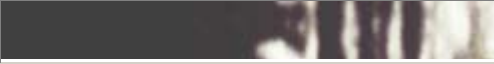




Figure 10 - Foot asymmetry. The left foot is smaller than the right. Courtesy S. Schneider, MD

## Tests

- Plain x rays may demonstrate bony abnormalities of the spine including spina bifida and scoliosis (Figure 11)
- CT scans can also be utilized to delineate associated bony abnormalities especially diastematomyelia
- Ultrasound is only useful in the infant spine and is useful primarily as a screening examination ( Figure 12)
- MRI is the primary mode of diagnosis. This examination shows the location of the end of the spinal cord and whether or not tethering is seen. It can also disclose other associated anomalies such as dermal sinus tracts, split cord

- malformations and lipoma (Figures 13, 14 and 15)
- Urodynamics or tests of bladder function are useful as a baseline and to follow patients after surgery

	
Figure 11 - X-ray of the infant spine demonstrating bony defects (arrow).	Figure 12 - Ultrasound image of a tethered cord. Courtesy S. Schneider, MD
	
Figure 13 - MRI of a tethered spinal cord. A normal conus medullaris lies at the level	Figure 14 - MRI of a diastematomyelia showing the split spinal cord and tethering

The conus in this patient lies near the sacrum. Courtesy S. Schneider, MD

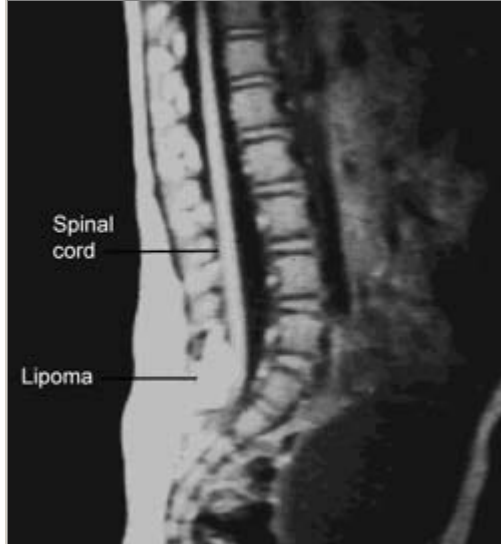


Figure 15 - MRI of a lipoma tethering the spinal cord. Courtesy S. Schneider, MD

### Indications/Contraindications

- There is little controversy regarding the management of a tethered cord that is symptomatic. These patients should be untethered prior to developing progressive deterioration
- All tethers associated with a dermal sinus tract should be untethered to prevent meningitis or potentially life threatening infection
- The management of the asymptomatic tether especially a lipoma is more controversial. At the current time most surgeons recommend a prophylactic (preventive) detethering
- The main goal of surgery is to stabilize neurologic progression, a proportion of patients improve as well as continue to progress despite untethering.
- Contraindications to surgery are relative, therefore, surgery should be deferred until the patient is in stable medical condition to tolerate general anesthesia

### Surgical Procedure

- The principle for tethered cord surgery is to release the end of the spinal cord from the surrounding adhesion and restore its location in the spinal canal
- The surgery is performed under general anesthesia in the prone (face down) position
- An incision is made over the area of the tether and muscle dissected off the back of the spine (lamina- [see Decompressive Laminectomy for anatomy](#))
- There is usually a defect in the lamina (spina bifida occulta) and the lamina removed (laminectomy)
- The dural tube (fibrous covering surrounding the spinal cord) is opened and all tissue attached to the end of the spinal cord is released and the spinal cord and nerve roots are separated from the surrounding tissues (untethered)
- Additional procedures may be required to manage some of the associated abnormalities as noted below:

1. Diastematomyelia and split cord malformation. The bony or cartilaginous spur dividing the spinal cord is removed and the dural tube reconstructed (Figure 16)
  2. Lipoma. The lipoma is debulked (removed piecemeal). It is usually not possible to remove the entire tumor as it blends into the end of the spinal cord. As much tumor is removed as can be done safely
  3. Epidermoid and dermoid cyst. Usually these are removed completely with removal of the dermal sinus tract and untethering of the cord
  4. Dermal sinus tract. The skin and involved tract are removed as they are traced down through the defect in the lamina (spina bifida occulta) to the cord (Figure 17)
  5. Tight Filum Terminale. This thick fibrous band is removed to untether the cord
  6. Myelocystocele. Often associated with malformations of the bladder and anus. The cyst is drained and the end of the cord released
  7. Neurenteric cyst. The cyst is carefully removed
  8. Syringomyelia. Syrinxes, which are usually large, are drained with a small tube inserted into the syrinx to keep it from reforming
  9. Secondary retether following myelomeningocele repair. Initially repaired at birth, retethering at a later time is the most common cause of secondary retethering. The site of the prior surgery is explored and scar tissue removed
- Any other associated abnormality is also repaired
  - Following the release of the tether and management of associated anomalies the dural tube is closed and the layers of muscle, fascia and skin closed

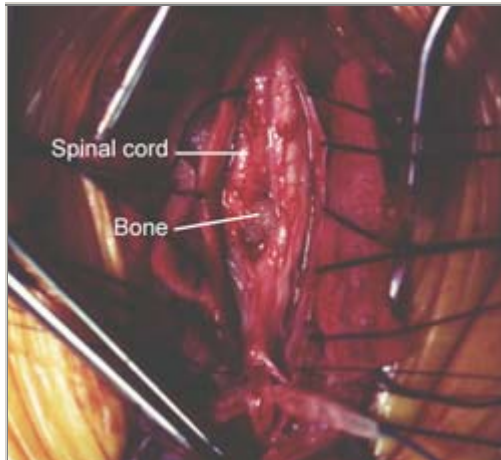


Figure 16 - Surgical photograph of a diastematomyelia showing the split spinal cord and bony spur. Courtesy S. Schneider, MD

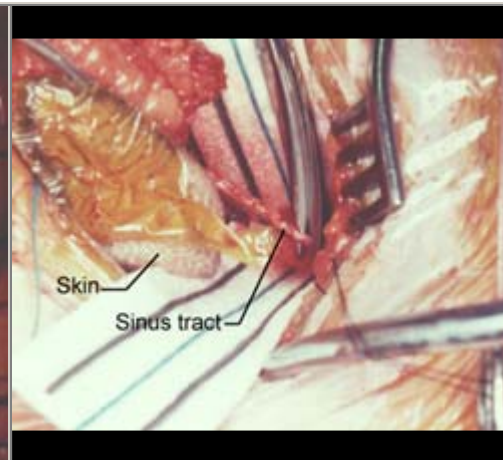


Figure 17 - Surgical photograph showing removal of a dermal sinus tract. Courtesy S. Schneider, MD

### Complications

- As with any major operation there are the usual risks of infection, bleeding and anesthesia
- Risks of transient or permanent loss of motor or sensory function and/or loss or alteration in bladder, bowel or sexual function
- Retethering requiring reoperation
- Tumor or cyst recurrence



- Cerebrospinal fluid (CSF) leakage. If CSF leak persists, reoperation may be necessary. (CSF is the fluid that bathes the brain and spinal cord)

### **Postoperative Care**

- Depending on surgical procedure the child may go to the intensive care unit or the general pediatric surgical floor
- Vital signs (pulse, blood pressure and respirations) are monitored
- A catheter may be inserted in an artery to monitor blood pressure and obtain blood for red blood cell counts and other blood tests
- A transfusion may be necessary
- A catheter is usually placed in the bladder to measure urine
- Medications are given which include antibiotics to prevent infection and steroids to decrease swelling
- Medications are also given to control pain
- A drain may be placed under the skin to drain fluid and is removed in a few days
- The child is checked to be sure that neurologic function is intact
- An MRI scan may be done to view the untethered spinal cord

### **After Care**

- The child is discharged when the blood count is stable, pain controlled and the infant is eating well. This usually takes several days
- Outpatient follow-up is necessary to ensure continued satisfactory growth and to monitor for the potential need for additional procedures (retethering, CSF leak, tumor recurrence, etc).