



Spina Bifida and Neuromuscular Disorders

Medical Staff and Volunteer Training



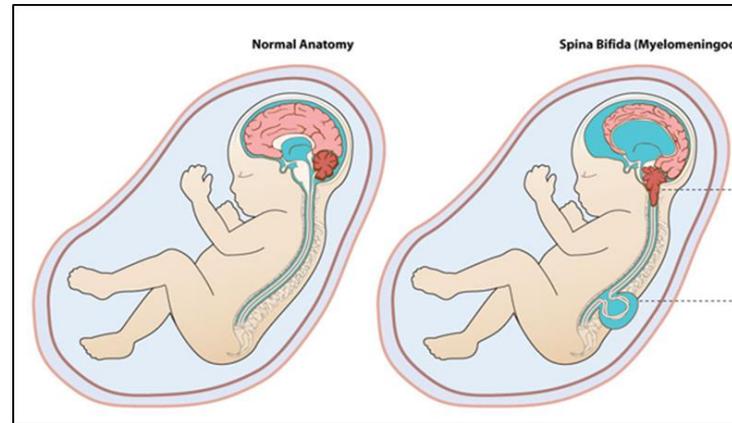
Spina Bifida



Spina Bifida

Spina bifida means “split spine.”

Spina bifida is the most common permanently disabling birth defect in the U.S., occurring in about 8 babies born each day, or 1,500 each year according to the CDC.



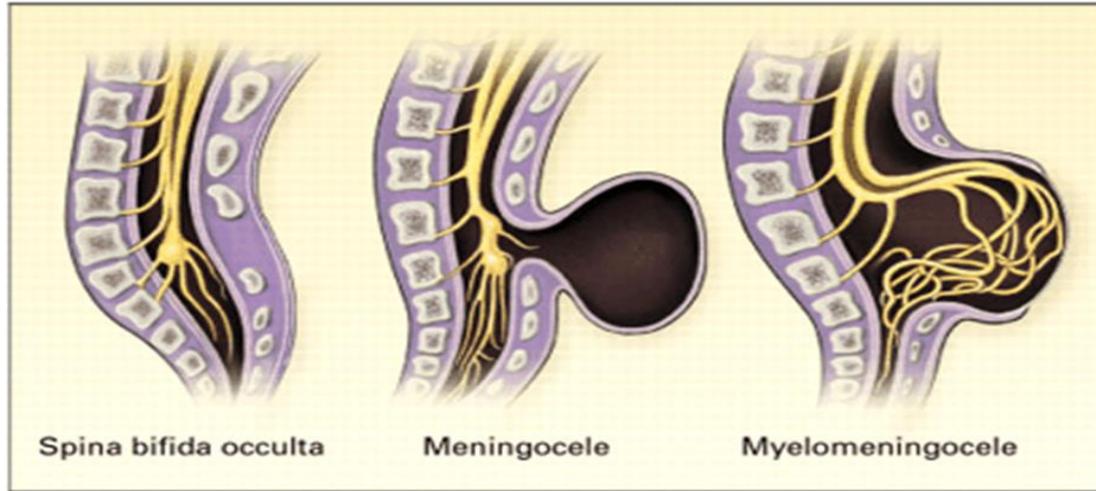
Spina bifida occurs when the spinal column does not close completely as it forms. This causes damage to the spinal cord and nerves.

There is no specific known cause for SB, although folic acid supplementation during pregnancy is known to be preventative for neural tube defects, which include spina bifida.



Spina Bifida

The 3 most common types of spina bifida are:



Spina Bifida Occulta

- No damage to spinal cord and nerves
- Often undiagnosed, "hidden," often asymptomatic
- Tethered cord risk

Meningocele

- Spinal fluid pushed into a sac outside the body
- Surgery to correct after birth
- Usually no nerve damage
- May have minor disabilities

Myelomeningocele

- Most severe form
- Spinal fluid, nerves, and parts of spinal cord protrude in sac
- Surgical correction after birth
- Hydrocephalus is present in 70-90%

Spina Bifida

Level of spinal cord injury from SB affects function.

Campers with SB have varying levels of function, but many cases of SB occur in the lumbar and sacral region.

This means they are likely to have age appropriate cognitive function and upper body control, but limited lower body strength and bowel/bladder control.

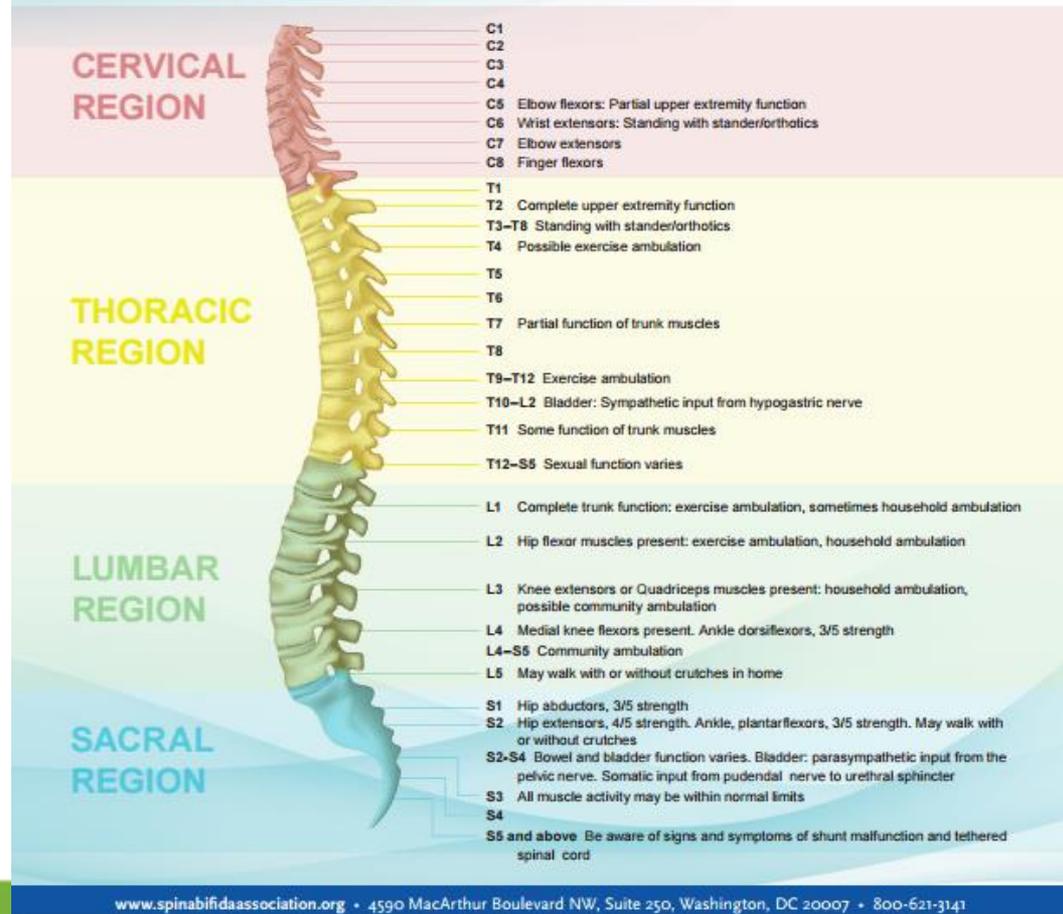
Spina Bifida Association. (2015, July 28).

<http://spinabifidaassociation.org/infosheets/sb-and-the-spine/>

SBA National Resource Center:
800-621-3141



Level of SB Function

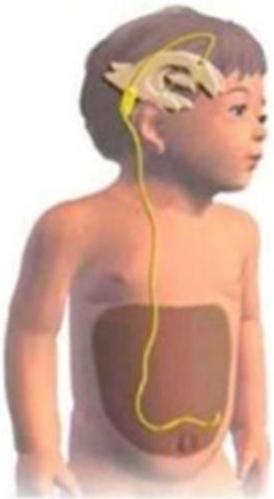


Spina Bifida

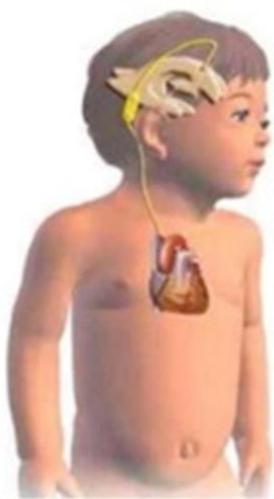
Hydrocephalus is present in 70-90% of babies born with myelomeningocele SB.

A shunt allows this extra fluid to drain from the ventricles into the peritoneal space (VP) or atrium (VA), reducing pressure on the brain.

VP SHUNT



VA SHUNT



In a camper with a shunt, we are concerned about:

- Change in level of consciousness (extreme agitation, behavior change, extreme/unusual fatigue)
- Headaches
- Vomiting



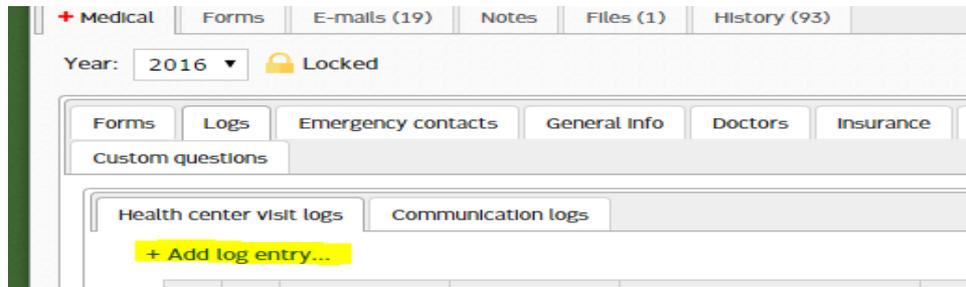
Spina Bifida at Victory Junction

If you have concerns about a camper's shunt, notify the provider on call and the full time medical staff.

For example, if a camper with a VP shunt has vomited twice in an hour and has a headache:

1. notify the provider on call and full time medical staff
2. assess the camper for any other changes or potential causes
3. document your assessment and any interventions in a log note

in CampSite



The screenshot shows a web interface for a medical record system. At the top, there are tabs for '+ Medical', 'Forms', 'E-mails (19)', 'Notes', 'Files (1)', and 'History (93)'. Below these is a 'Year: 2016' dropdown menu and a 'Locked' status indicator with a padlock icon. A horizontal menu contains 'Forms', 'Logs', 'Emergency contacts', 'General info', 'Doctors', and 'Insurance'. Underneath, there are sections for 'Custom questions', 'Health center visit logs', and 'Communication logs'. A yellow highlight is placed over a '+ Add log entry...' button.

If you are concerned...

Call for help!



Spina Bifida at Victory Junction

Campers with SB will have decreased mobility, strength, and sensation in their lower bodies.



Campers with SB often use mobility aids, such as crutches, AFOs, or walkers.

Additional concerns for campers with SB: skin checks and pool safety!



Spina Bifida at Victory Junction

Campers may also use a wheelchair for mobility.

Wheelchair safety at camp:

- Every should wear closed-toe shoes.
- Brakes on!
- Don't encourage racing.



A camper's wheelchair is an extension of him/herself.

Wheelchair etiquette at camp:

- Allow independence, do not push a camper's chair without asking.
 - Avoid leaning on a camper's wheelchair.
 - Get on the camper's level.

As a medical volunteer, you can help reinforce this etiquette with counselors and volunteers.



Spina Bifida at Victory Junction

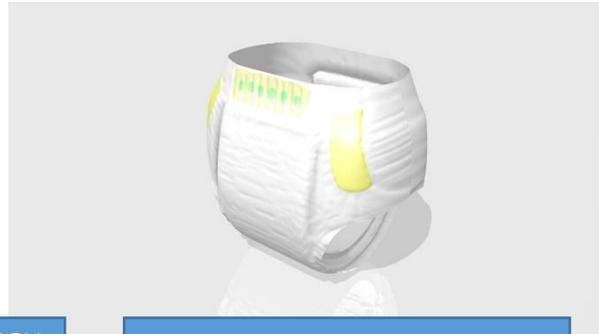
In addition to decreased strength, mobility, and sensation in the lower body, damage to the spinal cord in SB affects bowel and bladder function.

Neurogenic bladder and decreased nerve function lead to leakage and incontinence, and a default “off” setting that requires intervention for draining urine.



Anticholinergic medications may help encourage complete emptying and decrease bladder filling pressure, which allows the kidneys to drain and prevents infection.

side effects include reduced ability to sweat



Campers may use absorbent pants/pads to manage leakage and incontinence.

Counselors will assist with these ADLs.



Intermittent catheterization to drain the bladder is common in campers with SB. This may be done through the urethra or a stoma on the abdominal wall.

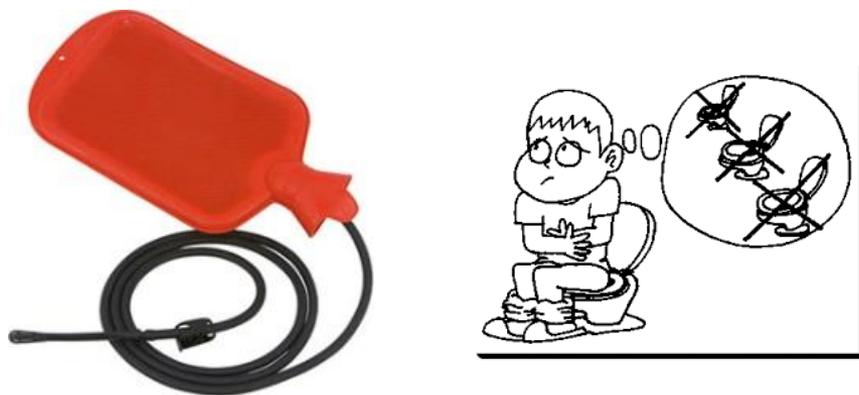
Nurses are responsible for performing all cathing (or assisting independent campers in self-cathing).

Spina Bifida at Victory Junction

Bowel routines, or “poop parties,” are common during this week at camp.

Like the bladder, a neurogenic bowel has a default “closed” setting and can have problems with leakage, incontinence, and decreased sensation.

Campers with SB will have a variety of routines.



Most bowel routines include filling the colon with a solution (through a MACE stoma or enema) and sitting on the toilet for up to 1-2 hours to empty.

Information about each camper’s bowel and bladder routine is found on the Medical Camper . You will update this during medical check-in with any changes.

Spina Bifida at Victory Junction

People with spina bifida are likely to have other medical complications, including:

- Latex allergy (Victory Junction strives to be completely latex free – including free of fruits with similar properties.)
- Learning disabilities
- ADHD (most often inattentive type)
- Precocious puberty
- Social challenges



Neuromuscular Disorders

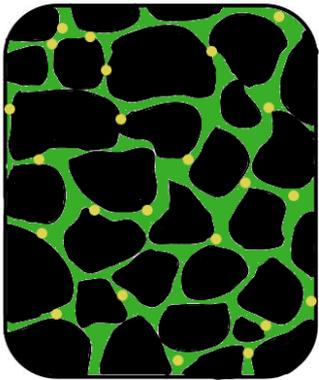


Neuromuscular Disorders

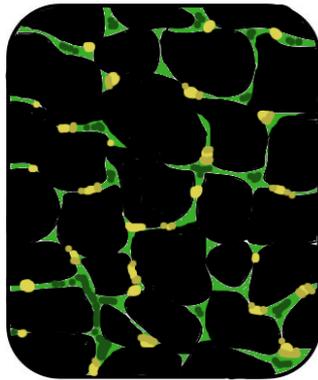
There are 9 types of muscular dystrophy, the most severe of which is Duchenne Muscular Dystrophy (DMD).

DMD is a genetic disorder that results in an absence of the protein dystrophin, which helps keep muscle cells intact, and is characterized by progressive muscle weakness and degeneration.

Muscular Dystrophy



Normal Muscle Tissue



Affected Muscle Tissue

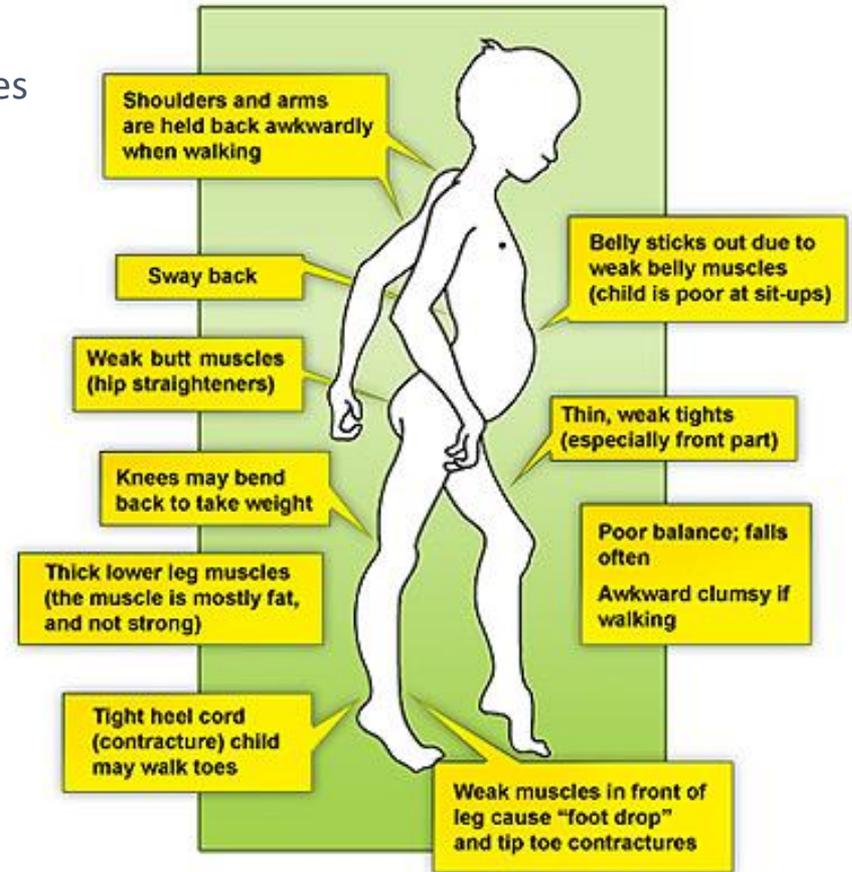
The genetic flaw that leads to DMD is an X-linked chromosome. The illness **primarily affects males.**

Females act as carriers of this gene, and can rarely be affected by the disease.



Duchenne Muscular Dystrophy

- Symptoms (often clumsiness) appear between ages 3 and 5.
- The first muscles affected are around the core (hips, thighs, shoulders), and lead to the gait changes shown in this image.
- Skeletal muscles of the arms and legs are affected next.
- Children often begin transitioning to using a wheelchair during the school age years (7-12).
- By the teen years, the heart and respiratory muscles are affected.
- Life expectancy used to be the teen years, but survival into the early 30s is now more common.



Neuromuscular Disorders

Major complications of Duchenne muscular dystrophy include:

- **Heart function**

- In the teen years, cardiac muscle may weaken and lead to cardiomyopathy
- Children with DMD should be followed closely by a cardiologist
- There is some evidence that cardiac decline can be slowed by beginning treatment with ACE inhibitors and/or beta blockers at early signs of change.

- **Respiratory function**

- Around age 10, the diaphragm and intercostal muscles begin to weaken, making inspiration and expiration less effective.
- Decreased ability to cough can lead to respiratory infections and pneumonia
- Headaches, poor concentration, poor sleep, and fatigue can indicate declining respiratory function

- **Neurological function**

- DMD doesn't affect the nerves, so smooth muscle functions are maintained (including bowel and bladder control)
- About 1/3 of children with DMD have a mild learning disability
- Muscle deterioration is not painful, but muscle cramps or other pain may occur

Neuromuscular Disorders

Treatment of DMD includes:

- **Mobility and positioning aids**
 - Use of splints, walkers, and wheelchairs is common to assist in maintaining mobility and independence as muscles deteriorate.
- **Management of physical variances (i.e. scoliosis, contractures)**
 - Surgery, AFOs, and other supports may be indicated
- **Physical therapy**
 - Some exercise is helpful for strengthening muscles, but too much may cause damage. PT can help maintain mobility and independence.
- **Medications**
 - Prednisone, or other steroids, may slow the deterioration process of DMD
- **Respiratory support**
 - Initially, non-invasive support (i.e. BiPAP, CPAP) while sleeping can improve respiratory status.
 - As the disease progresses, some people require mechanical ventilator support, often through a tracheostomy.

Things to Remember...

- Every child is unique. Do your best to get to know your campers and listen to them!
- We do our best to stick to campers' home routines. Medications and treatments should be done the way they are done at home as much as possible.
- As a volunteer, you are not expected to be an expert in everything. We welcome your knowledge and experience and aim to work as a team!
- The unit nurses, full time staff, and specialist providers are excellent resources and here to help. Always ask if you are unsure or concerned about a medication, treatment, or camper's condition.



Thank
you!



References

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