Spina Bifida

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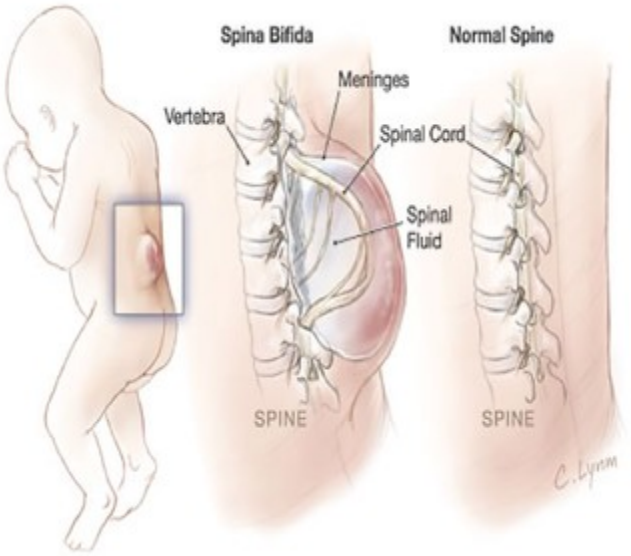
Definition and introduction to the population or specific disability

Spina Bifida literally means “split spine.” Spina Bifida happens when a baby is in the womb and the spinal column does not close all of the way. Every day, about eight babies born in the United States have Spina Bifida or a similar birth defect of the brain and spine.

Spina bifida, which literally means “cleft spine,” is characterized by the incomplete development of the brain, spinal cord, and/or meninges (the protective covering around the brain and spinal cord). It is the most common neural tube defect in the United States—affecting 1,500 to 2,000 of the more than 4 million babies born in the country each year. An estimated 166,000 individuals with spina bifida live in the United States.



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What are the different types of Spina Bifida?

1. *Occult Spinal Dysraphism* (OSD)

Infants with this have a dimple in their lower back. Because most babies with dimples do not have OSD, a doctor has to check using special tools and tests to be sure. Other signs are red marks, hyperpigmented patches on the back, tufts of hair or small lumps. In OSD, the spinal cord may not grow the right way and can cause serious problems as a child grows up. Infants who might have OSD should be seen by a doctor, who will recommend tests.

2. *Spina Bifida Occulta*

It is often called “hidden Spina Bifida” because about 15 percent of healthy people have it and do not know it. Spina Bifida Occulta usually does not cause harm, and has no visible signs. The spinal cord and nerves are usually fine. People find out they have it after having an X-ray of their back. It is considered an incidental finding because the X-Ray is normally done for other reasons. However, in a small group of people with SBO, pain and neurological symptoms may occur. Tethered cord can be an insidious complication that requires investigation by a neurosurgeon.

3. *Meningocele*

A meningocele causes part of the spinal cord to come through the spine like a sac that is pushed out. Nerve fluid is in the sac, and there is usually no nerve damage. Individuals with this condition may have minor disabilities.

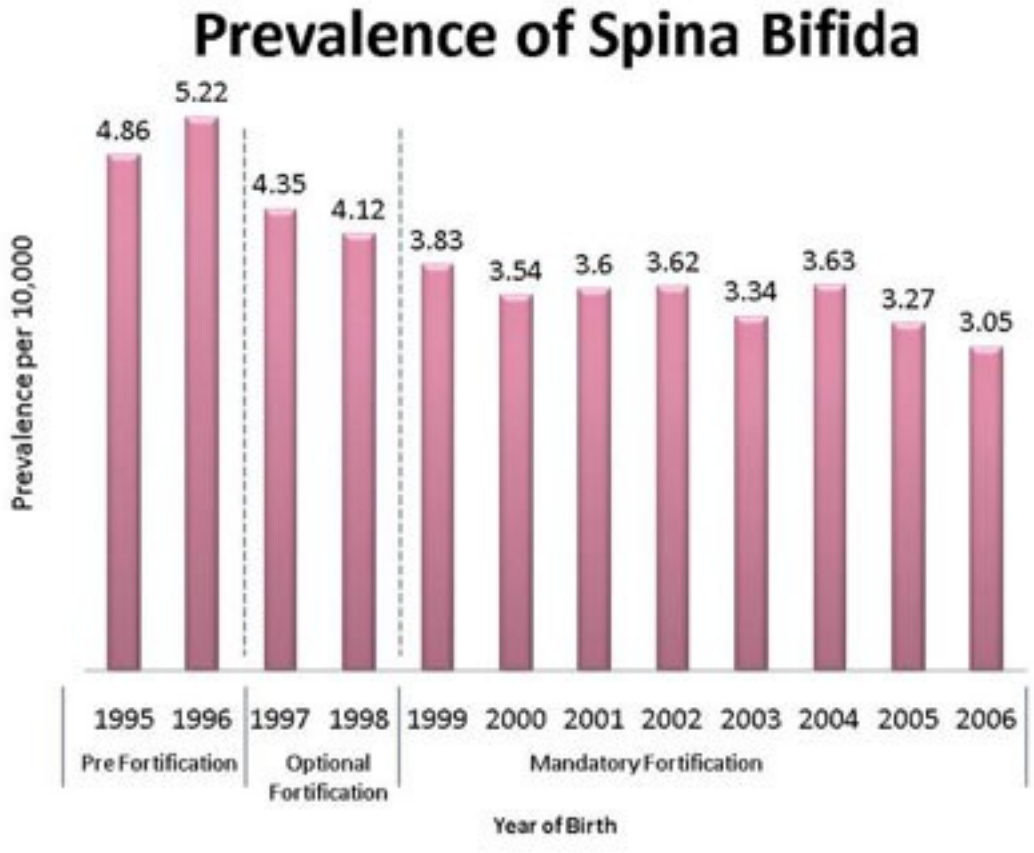
4. *Myelomeningocele* (Meningomyelocele), also called Spina Bifida Cystica

This is the most severe form of Spina Bifida. It happens when parts of the spinal cord and nerves come through the open part of the spine. It causes nerve damage and other disabilities. Seventy to ninety percent of children with this condition also have too much fluid on their brains. This happens because fluid that protects the brain and spinal cord is unable to drain like it should. The fluid builds up, causing pressure and swelling. Without treatment, a person's head grows too big, and may have brain damage. Children who do not have Spina Bifida can also have this problem, so parents need to check with a doctor.

Random Facts about Spina Bifida:

- Each year, about 1,500 babies are born with spina bifida.
- Hispanic women have the highest rate of having a child affected by spina bifida compared with Non-Hispanic White and Non-Hispanic Black women:
    - Hispanic: 4.17 per 10,000
    - Non-Hispanic Black or African-American: 2.64 per10,000
    - Non-Hispanic White: 3.22 per 10,000
- In 1992, the U.S. Public Health Service recommended that all women of childbearing age consume 400 micrograms (mcg) of folic acid daily to reduce the risk of having a pregnancy affected by neural tube defects (NTDs), such as spina bifida.
  Subsequently, the U.S. Food and Drug Administration mandated adding folic acid to all enriched cereal grain products by January 1998.
    - The prevalence rate of spina bifida declined 31% from the prefortification (1995–1996) rate of 5.04 per 10,000 to the post-fortification (1998–2006) rate of 3.49 per 10,000.

- An estimated 1,000 more babies have been born without an NTD each year since fortification began.



Diagnosis, including specific characteristics, symptoms, prognosis, progression, etc.

The exact cause of this birth defect isn't known. Experts think that genes and the environment are part of the cause. For example, women who have had one child with spina bifida are more likely to have another child with the disease. Women who are obese or who have diabetes are also more likely to have a child with spina bifida.

Diagnosis

During pregnancy, you can have a blood test (maternal serum triple or quadruple screen) and an ultrasound of the developing baby. These tests check for signs of spina bifida and other

problems. If test results suggest a birth defect, you can choose to have an amniocentesis. This test helps confirm if the baby has spina bifida.

After birth, a doctor can usually tell if a baby has spina bifida by how the baby's back looks. If spina bifida is suspected, the doctor may do an X-ray, an MRI, or a CT scan to see if the defect is mild or severe.

Spina bifida symptoms will depend on how severe the defect is. Most children with the mild form of spina bifida don't have any problems from it.

In many cases, children with meningocele don't have any symptoms.

Children with the most severe form of spina bifida often have spine and brain issues that cause serious problems. They may have:

- Little or no feeling in their legs, feet, or arms, so they may not be able to move those parts of the body.
- Bladder or bowel problems, such as leaking urine or having a hard time passing stools.
- Fluid buildup in the brain (hydrocephalus). Even when it is treated, this may cause seizures, learning problems, or vision problems.

A curve in their spine, such as scoliosis.

How is it treated?

Most children with the mild form of spina bifida don't need treatment. Children with meningocele may not need treatment either. But children with the most severe form usually need surgery. Sometimes surgery to correct severe spina bifida can be done before a baby is born.

A child who has hydrocephalus will need surgery to put in a drainage tube called a shunt. It relieves pressure on the brain by draining excess fluid into the belly. This keeps the swelling from causing more damage to the brain.

Experts such as physical therapists and occupational therapists work with children who have severe spina bifida. The work starts soon after the child's birth. These therapists can teach parents and caregivers how to do exercises and activities with the child.

Some children may need a brace, a wheelchair, or other aids. Children with bladder control problems may need help using a catheter each day to prevent infection and kidney damage. To help prevent bowel problems, parents usually begin working with the doctor or nurse on managing bowel care as soon as the child starts eating solid food. As children with severe spina bifida grow, other treatments and surgeries may be needed to manage problems that arise.

Spina Bifida Treatment

Treatment for spina bifida depends on the severity of the condition.

- Most people with spina bifida occulta require no treatment at all.
- Children with meningocele typically require surgical removal of the cyst and survive with little, if any, disability.
- Children with myelomeningocele, however, require complex and often lifelong treatment and assistance. Almost all of them survive with appropriate treatment starting soon after birth. Their quality of life depends at least partially on the speed, efficiency, and comprehensiveness with which that treatment is provided.

Before Pregnancy tips

Before and during pregnancy, a woman can help prevent spina bifida in her child.

- Get plenty of folic acid each day, both before you get pregnant and during pregnancy. All foods made from grains and sold in the United States have folic acid added. Foods rich in folic acid include fortified breakfast cereals and breads, spinach, and oranges. Your doctor may recommend that you also take a daily vitamin with folic acid or a folic acid supplement.
- If you take medicine for seizures or acne, talk with your doctor before you get pregnant. Some of these medicines can cause birth defects.
- Don't drink alcohol while you are pregnant. Any amount of alcohol may affect your baby's health.

Don't let your body get too hot in the first weeks of pregnancy. For example, don't use a sauna or hot tub. And treat a high fever right away. The heat could raise your baby's risk for spina bifida.

Tips for parents with children with Spina bifida

There are many ways you can support your child:

- Go to all scheduled doctor visits.
- Help your child be active, and encourage him or her to be as independent as possible.
- Encourage your child to drink plenty of fluids and eat foods high in fiber, such as whole grains and fruits. This helps prevent constipation.
- Check your child's skin each day for cuts, bruises, and pressure sores. Children who have little or no feeling in their legs and feet may get hurt and not know it. And that could lead to an infection.
- Be sure to get your child's vision checked regularly. Children with spina bifida often have weak eye muscles.
- Keep your child away from latex products if he or she has a latex allergy.
- Watch for learning difficulties, and talk to your child's doctor or teacher if you have any concerns.
- When your child is ready to start school, talk with teachers and other school workers. Public schools have programs for people ages 3 through 21 with special needs.

Remember that your needs are important too. Take good care of yourself so you can stay healthy and have the energy to enjoy your child. Make time for activities you like, even if it's just for a short while each day. And reach out to family, friends, and support groups when you need help.

How do they surgically fix it in utero?

A new study in the New England Journal of Medicine says that if a baby is operated on while still in the uterus, the most serious complications of the worst form of spina bifada, myelomeningocele, can be lessened.

A clinical trial of 158 women found that brain malformations were reversed in one-third of the fetuses and that almost half the babies could eventually walk without crutches.

"This is not a cure," said Dr. Scott Adzick, lead author of the study and the chief of pediatric surgery at the Children's Hospital of Philadelphia. But, he said, the results of this clinical trial show that prenatal repair surgery is a viable option.

The trial was run at three hospitals: Vanderbilt University Medical Center in Nashville, the Children's Hospital of Philadelphia and the University of California, San Francisco.

The study was stopped early, last December, because the results were so good, Adzick said.

Half the mothers had the surgery from the 19th through the 25th weeks of pregnancy. The other half carried their babies to term and the surgery was performed a few days later.

According to the study, about one in 3,000 babies are born with this type of spina bifida. The bone around the spine doesn't form properly, causing part of the spinal cord to stick out of the baby's back. About 10% of these babies die after birth.

Almost 90% of babies born with this type of spina bifida have hydrocephalus, excess brain fluid. The only way to alleviate the pressure is to insert a tube or shunt to drain it.

After one year, researchers found only 40% of babies in the prenatal surgery group had to get shunts, compared with 82% who had the surgery after they were born.

"The reduced risk of hydrocephalus is by the far the biggest benefit of this surgery," said Dr**.** Noel Tulipan, director of Pediatric Neurosurgery at Vanderbilt University Medical Center. "I see these patients all the time, and the thing that brings these patients back to the hospital over and over again are the shunts."

Prognosis

The prognosis for individuals with SB depends on the number and severity of abnormalities. Prognosis is poorest for those with complete paralysis, hydrocephalus, and other congenital defects. With proper care, most children with SB live well into adulthood.

Medical research has improved for people with spina bifida in the past few years and medical technology has improved the prognosis for them to experience a longer life expectancy than in the past. Depending on the severity of their ailments as well as any other complications they may have suffered, it is not unusual for those with spina bifida to live well into adulthood. Available treatment options also make it more likely they can live productive lives much longer than those with the ailment in the past few years.

The prognosis for those with spina bifida will depend largely on the type of illness they have. For example, those with spina bifida occulta will have few symptoms and a more positive prognosis than those with more severe types of the affliction. Even those with the most severe type of spina bifida will often suffer different levels of symptoms that will affect the overall prognosis on a patient-by-patient basis.

Some symptoms of spina bifida will indicate how the prognosis will affect the patient such as full paralysis. Other issues that have a negative effect will include incontinence, hydrocephalus and any nerve damage suffered from the beginning. The more mild the symptoms the patient exhibits the better the prognosis they can expect. With the wide range of symptoms and the many different degrees of severity it is difficult to develop a prognosis that will fit each person affected by spina bifida.

There will also be issues faced by adults with spina bifida that affect their prognosis. Some of the problems they may develop in later years that weren't present when they were young will have a negative effect on their individual prognosis. Since many will have limited activity obesity is one thing that affects those with spina bifida. It can lead to diabetes, high blood pressure and depression.

Additionally, those who have been using braces or other orthopedic devices can develop joint problems that will affect their future. As they grow, adjustments will need to be made to the devices to ensure a good fit. If they have been fitted with a shunt for water on the brain, it can change position and lead to many other health problems.

Medications, physical therapy, possible surgery and experienced health care will help improve their quality of life and ensure a more positive prognosis. However, when younger it will be their

parents and health care professionals that set the stage for their continued improvements as adults.

Specific needs

Quality of Life

- Many adolescents and young adults with spina bifida report a high level of satisfaction with their health-related quality of life, are entering and succeeding at college life, and are participating in sports and other recreational activities.
- Some adolescents and young adults are concerned about their future because of secondary health conditions they experience frequently.
- While many parents of adolescents and young adults with spina bifida are satisfied with their children's overall quality of life, they say their children face challenges in continence and getting around.

What are the complications of spina bifida?

Complications of spina bifida can range from minor physical problems with little functional impairment to severe physical and mental disabilities. It is important to note, however, that most people with spina bifida are of normal intelligence. Spina bifida's impact is determined by the size and location of the malformation, whether it covered, and which spinal nerves are involved. All nerves located below the malformation are affected to some degree. Therefore, the higher the malformation occurs on the back, the greater the amount of nerve damage and loss of muscle function and sensation.

In addition to abnormal sensation and paralysis, another neurological complication associated with spina bifida is Chiari II malformation—a condition common in children with myelomeningocele—in which the brain stem and the cerebellum (hindbrain) protrude downward into the spinal canal or neck area. This condition can lead to compression of the spinal cord and

cause a variety of symptoms including difficulties with feeding, swallowing, and breathing control; choking; and changes in upper arm function (stiffness, weakness).

Chiari II malformation may also result in a blockage of cerebrospinal fluid, causing a condition called *hydrocephalus*, which is an abnormal buildup of cerebrospinal fluid in and around the brain. Cerebrospinal fluid is a clear liquid that surrounds the brain and spinal cord. The buildup of fluid puts damaging pressure on these structures. Hydrocephalus is commonly treated by surgically implanting a shunt—a hollow tube—in the brain to drain the excess fluid into the abdomen.

Some newborns with myelomeningocele may develop meningitis, an infection in the meninges. Meningitis may cause brain injury and can be life-threatening.

Children with both myelomeningocele and hydrocephalus may have learning disabilities, including difficulty paying attention, problems with language and reading comprehension, and trouble learning math.

Additional problems such as latex allergies, skin problems, gastrointestinal conditions, and depression may occur as children with spina bifida get older.

Medical Issues in the Classroom

Self-care skills can be taught by the teacher in the classroom to help the child with spina bifida. One aspect of self-care for these children is to check their bodies for injuries such as sores or pressure areas. This is important because children with spina bifida have a loss of skin sensation below the spinal defect and therefore they may have sustained an injury of which they are not even aware. Korabek and Cuvo (1986) state that establishing a daily routine in which positive reinforcement is used is one effective way to teach children with spina bifida this important skill.

Muscle weakness below the spinal lesion affects the child with spina bifida in many ways including making it difficult to perform self-care skills. The muscle weakness can be more or less severe depending on where the lesion is located (Laurence and Beresford, 1976). Korabek and Cuvo (1986) describe that when the lesion is located in the neck or upper back there is

usually less paralysis in the lower extremities and legs. The bladder may also function normally. If the myelomeningocele lesion occurs in the high lumbar area which is between the ribs and pelvis around the twelfth thoracic vertebrae the damage usually involves some degree of paralysis of the lower trunk and thighs. However, when the lesion is in the lower lumbar region at the level of the third or fourth lumbar vertebrae, this will normally result in the paralysis of the legs, feet, and ankles as well as a loss of bladder control. If the lesion is at the lowest part of the back children may not have any leg paralysis but may have a lack of bladder control. To combat the effects of the muscle weakness, physical and occupational therapists often work to build the upper body strength of these children. This enables the children to be more effective in their daily living skills and activities (Korabek and Cuvo, 1986).

Another issue that teachers often focus on is the bowel and bladder incontinence of children with spina bifida. Korabek and Cuvo (1986) note that almost all children with spina bifida have some degree of bowel or bladder incontinence. Teachers who do not understand the medical reasons behind the malfunction often spend a great deal of wasted time trying to toilet train these students. The muscles of the bowel and bladder walls of these children are weak. This results in the child having the inability to control when they will release the contents of their bowel and bladder. Sometimes they also have a lack of sensation that would normally tell them when they have to go. Korabek and Cuvo (1986) state that teachers who understand these medical reasons find it more effective to habit train students. This is when the students learn to go to the restroom at regular intervals rather that relying on their body to tell them when they have to go.

Specific test

- During pregnancy you can have a blood test done
- Also during pregnancy you can have an ultrasound done
  - These test can check for signs of spina bifida and the overall health of the baby
- Also after the 15th week of pregnancy a mother can choose to have an amniocentesis
  - This test studies the amniotic fluid surrounding the baby which can give clues as to the babies health

Equipment

· Individuals with spina bifida may need a brain shunt to drain the spinal fluid build up

· Wheel chairs are also common to help with mobility due to spinal cord damage.

· Antibiotics are also commonly prescribed to help with infection due to the brain shunt that may get infected.

TR implications

Jean Driscoll--olympian with Spina Bifida: https://www.youtube.com/watch?v=OPXymHaN67A

Can children with Spina Bifida grow up and live full lives?

Yes. With help, children with Spina Bifida can lead full lives. Most do well in school, and many play in sports. Because of today's medicine, about 90 percent of babies born with Spina Bifida now live to be adults, about 80 percent have normal intelligence and about 75 percent play sports and do other fun activities.

There is no cure for spina bifida. The nerve tissue that is damaged cannot be repaired, nor can function be restored to the damaged nerves. Treatment depends on the type and severity of the disorder. Generally, children with the mildest form need no treatment, although some may require surgery as they grow.

- Because there is no known cure, depression and other mood disorders are not rare. As RTS we must work on helping those with Spina Bifida improve their quality of life and reach a level and realize that their limitations do not define them.

How is Spina Bifida managed? (notice the word choice, *managed* not "CURED")

As type and level of severity differ among people with Spina Bifida, each person with the condition faces different challenges and may require different treatments.

The best way to manage Spina Bifida is with a team approach. Members of the team may include neurosurgeons, urologists, orthopedists, physical and occupational therapists, orthotists, psychologists and medical social workers.

What conditions are associated with Spina Bifida? (RELATE EACH OF THESE AND HOW TR CAN HELP WITH THEM)

Children and young adults with Spina Bifida can have mental and social problems. They also can have problems with walking and getting around or going to the bathroom, latex allergy, obesity, skin breakdown, gastrointestinal disorders, learning disabilities, depression, tendonitis and sexual issues.

Resources (local, state, national, & international)

BRAIN
P.O. Box 5801
Bethesda, MD 20824
(800) 352-9424
http://www.ninds.nih.gov

National Institute of Child Health and Human Information Resource Center P.O. Box 3006 Rockville, MD 20847 NICHDInformationResourceCenter@mail.nih.gov http://www.nichd.nih.gov Tel: 800-370-2943 888-320-6942 (TTY) Fax: 301-984-1473 **March of Dimes** 1275 Mamaroneck Avenue White Plains, NY 10605 askus@marchofdimes.com http://www.marchofdimes.com Tel: 914-997-4488 888-MODIMES (663-4637) Fax: 914-428-8203 **Spina Bifida Association** 4590 MacArthur Blvd. NW Suite 250	

Washington, DC 20007-4266 sbaa@sbaa.org http://www.spinabifidaassociation.org Tel: 202-944-3285 800-621-3141 Fax: 202-944-3295 **National Dissemination Center for Children with Disabilities** U.S. Dept. of Education, Office of Special Education Programs 1825 Connecticut Avenue NW, Suite 700 Washington, DC 20009 nichcy@aed.org http://www.nichcy.org Tel: 800-695-0285 202-884-8200 Fax: 202-884-8441	

Works Cited:

- http://www.spinabifidaassociation.org/site/c.evKRI7OXIoJ8H/b.8277225/k.5A79/What_is_Spina_Bifida.htm
- http://www.ninds.nih.gov/disorders/spina_bifida/detail_spina_bifida.htm
- http://faculty.frostburg.edu/mbradley/EC/spinabifida.html
- http://www.cdc.gov/ncbddd/spinabifida/data.html
- http://www.webmd.com/parenting/baby/tc/spina-bifida-topic-overview?page=2
- http://www.rightdiagnosis.com/s/spina_bifida/prognosis.htm
- http://www.emedicinehealth.com/spina_bifida/page5_em.htm
- http://www.cnn.com/2011/HEALTH/02/09/surgery.spina.bifada/index.html
- http://www.spinabifida.net/prognosis.html