



A Guide to
Understanding

Spina Bifida

Center for Spina Bifida
at Gillette Children's Specialty Healthcare

Our Mission

Gillette Children's Specialty Healthcare meets the special health-care needs of people — primarily children and teens — who have physical and functional disabilities. Our mission is to help those we serve realize greater well-being, independence and enjoyment in life.

We combine medical, nursing, therapeutic, technical, psychosocial and other expertise in family-centered programs. We provide services at our clinics, in our hospital and throughout the region, in response to community needs and often in collaboration with other organizations. We seek to build community partnerships that help continue our mission and enhance care for people who have disabilities.



Spina Bifida

Gillette's Center for Spina Bifida treats infants, children, teens and adults with spina bifida. Because spina bifida is a complex condition that affects each person differently, we provide a wide range of services to meet our patients' needs. This booklet contains information to help patients and families learn about spina bifida, its associated health issues, and the specialists who treat such issues at Gillette.

Braeden's Story

Michelle and Brian say they were shocked when an ultrasound revealed that their unborn baby had spina bifida and hydrocephalus. After Braeden was born, Michelle and Brian took him to Gillette, where he receives comprehensive services. Braeden has had more than 20 surgeries at Gillette, including several shunt surgeries to treat his hydrocephalus. Gillette's assistive-technology team designed a custom wheelchair for Braeden, and Gillette's therapeutic recreation specialists helped him find fun activities for people who have disabilities. Today, Braeden participates in many of the same activities that other boys his age enjoy, including T-ball, video games and horseback riding.



Braeden

Spina Bifida

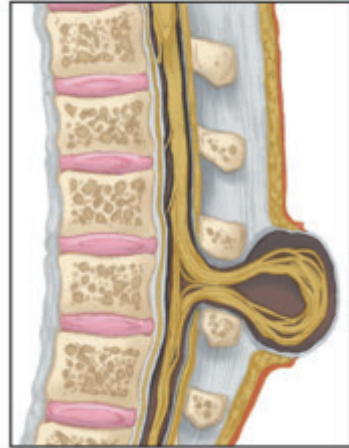
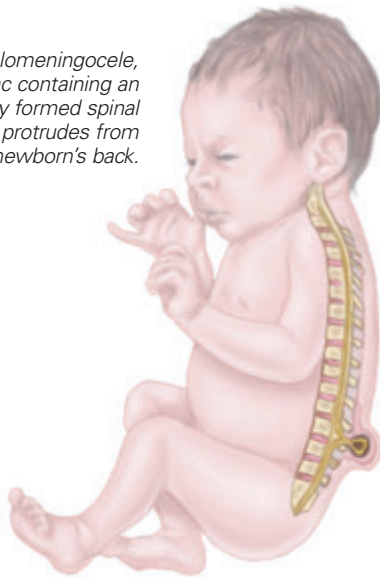
Spina bifida is a birth defect that occurs when an embryo's brain, spinal cord, surrounding nerves and/or spinal column fail to develop normally during the first 28 days of pregnancy. There are many types of spina bifida. The most common are:

- **Myelomeningocele:** In myelomeningocele — the most severe form of spina bifida — a sac containing an abnormally formed spinal cord protrudes from an opening in a newborn's back. The nerves at and below the affected area are damaged, causing paralysis and bowel and bladder complications. Babies need surgery shortly after birth to repair the affected area and prevent further injury or infection. The nerve damage, however, can't be reversed.
- **Meningocele:** In meningocele, a baby is born with a sac protruding from an opening in the spinal column. The condition differs from myelomeningocele in that the sac doesn't contain the spinal cord and fewer nerves are affected. Babies need surgery shortly after birth to repair the affected area. Most babies born with meningocele grow up without complications. Some have minor disabilities.
- **Spina bifida occulta:** In spina bifida occulta, the bones around a baby's spinal cord fail to develop normally. The nerves of the spinal cord usually are normal, as is the skin on the back. Sometimes, however, there will be a dimple, hair patch or red discoloration on the skin over the affected area. Babies born with spina bifida occulta usually don't experience complications. Sometimes, however, patients experience problems such as tethered spinal cords (see page 10).

Causes

Spina bifida occurs in one of every 1,000 births. Although the cause is unknown, a combination of genetic and environmental factors might be involved. Once a woman has a child with spina bifida, her chance of having another child with spina bifida is one in 20.

With myelomeningocele, a sac containing an abnormally formed spinal cord protrudes from a newborn's back.



Prevention

Folic acid is a B vitamin that can decrease chances of a woman having a baby with spina bifida by as much as 70 percent. Because spina bifida occurs before a woman knows she's pregnant, women of childbearing age should talk to their doctors about taking 400 micrograms of folic acid daily.

Treatment

The affected area on the spine of a child with spina bifida requires neurosurgery within 24 to 48 hours of birth. A neurosurgeon (a doctor who specializes in brain and spinal-cord surgery) removes the sac and places the spinal cord in as normal a position as possible, protecting any functioning nerve tissue from injury and infection. The damage to the spinal cord, however, can't be reversed. People who have spina bifida — particularly myelomeningocele — require ongoing treatment for problems stemming from damage to the spinal cord.

Associated Problems

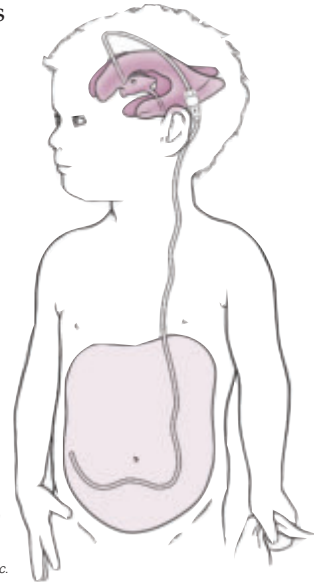
In myelomeningocele, the nerves at and below the affected area on the spine don't function properly, causing a variety of health problems.

Paralysis

People who have myelomeningocele can't control muscles below the affected area of the spine. Usually, the legs are totally or partially paralyzed and skin sensation is abnormal. The chart on page 7 shows the typical amount of leg function possible for people whose myelomeningocele is located at different levels.

Hydrocephalus

In babies with myelomeningocele, the brain also typically protrudes into the spinal canal (a position called an Arnold Chiari malformation). The brain's position blocks the flow of cerebral fluid, which builds up in the skull and causes hydrocephalus (sometimes called water on the brain). Between 80 and 90 percent of children with myelomeningocele need surgically implanted shunts, which drain excess cerebral fluid into another area of the body. A neurosurgeon performs shunt surgery. If severe hydrocephalus is present at birth, a neurosurgeon might place a shunt during the surgery to close the opening in the back. Otherwise, surgery is performed when necessary, usually within the first few weeks of life. People who have shunts typically need surgeries throughout their lives to replace or repair their shunts.



Surgically implanted shunts drain excess cerebral fluid from the ventricles to another area in the body, such as the abdomen.

Illustration courtesy of Medtronic, Inc.

**Myelomenin-
gocele Level**

**Amount of
Leg Function**

Thoracic 12
and above

Children typically use wheelchairs for most activities and continue using wheelchairs throughout their lives.

Lumbar 1

Children typically need special braces and/or crutches for walking. Teens and adults might walk only for exercise but use wheelchairs most of the time.

Lumbar 2 – 3

Children typically need crutches and long-leg braces up to the thigh or waist. Teens and adults might walk for exercise but use wheelchairs most of the time.

Lumbar 4

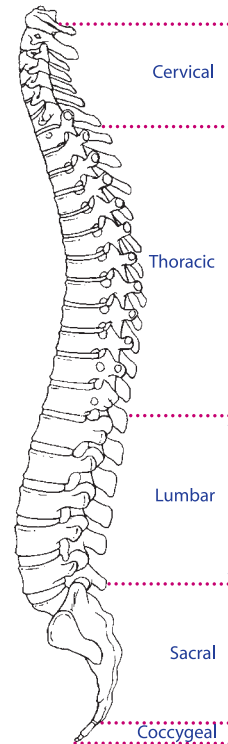
Children typically need short- or long-leg braces to walk. Teens and adults might use wheelchairs.

Lumbar 5 – Sacral 1

Children typically walk, but might need short-leg braces and crutches or canes.

Sacral 2 – 4

Children typically walk without braces or crutches but might need shoe inserts.



Watching for Shunt Problems

Talk to your doctor about the signs and symptoms of shunt problems, including:

- Return of original symptoms of hydrocephalus, such as headaches, vomiting, irritability and fatigue
- Signs of infection, including fever and redness or swelling along the shunt tract

If you notice any of these symptoms, call our nurse triage line immediately at 651-229-3890.

Bone and Muscle Problems

Because myelomeningocele causes paralysis, children with myelomeningocele are more likely to develop foot deformities, dislocated hips, bowed legs and scoliosis (an abnormal curvature of the spine). Orthopaedic surgeons (doctors who specialize in treating bone and muscle problems) provide ongoing treatment for such problems. Children might also need braces, specially designed seating systems, and other adaptive equipment.

People with myelomeningocele who can't stand, and therefore don't typically bear weight on their legs, are at greater risk of developing fractures. Because they have little or no sensation, they might be unaware of injuries. Watch for warning signs, including swelling, redness, increased warmth or bone deformities. Report any such signs to an orthopaedic surgeon.

Kidney and Bladder Problems

In most people who have myelomeningocele, the kidneys work normally, but the bladder doesn't. Normal bladder control usually isn't possible for people who have myelomeningocele, because they can't feel when their bladders are full and they're unable to empty their bladders on their own. As a result, they might experience frequent urinary tract infections. Sometimes, urine pushes into the kidneys, a condition called reflux. Reflux can damage the kidneys.

Joel Hutcheson, M.D., urologist, discusses a renal (kidney) ultrasound with a young patient and her mother. People who have spina bifida require regular appointments with urologists to help detect and treat kidney and bladder problems.



Maintaining a healthy bladder and kidneys requires regular checkups with a urologist (a doctor who specializes in bladder and kidney problems). Urologists can test bladder and kidney function and detect infections. Some people need to use catheters to empty their bladders. Parents or caregivers manage catheterization for babies and young children. Some people who have spina bifida need medication or surgery to stay dry and keep their kidneys and bladders healthy.

Bowel Problems

Myelomeningocele also affects the nerves that control the bowel. As a result, people with myelomeningocele typically have no control of the anal sphincter, the muscle that controls the passage of stool from the rectum. They can't feel stool in the rectum, tighten the sphincter to hold in stool, or loosen the sphincter to have a bowel movement. Bowel accidents can be common. Because stool moves through the intestine more slowly in people with myelomeningocele, constipation also is a frequent problem.

Early intervention by a pediatrician familiar with myelomeningocele or a gastroenterologist (a doctor specializing in problems of the gastrointestinal

tract) is important. Doctors might prescribe a high-fiber diet, adequate fluid intake, regular toileting, and a regimen of suppositories, laxatives or enemas to establish and maintain a successful bowel program. In some cases, people need bowel surgery.

Tethered Cord

Tethered cord occurs when the lower end of the spine becomes tethered, or stuck, to the dura (the tough membrane covering the spinal cord). When the spinal cord becomes tethered, it can't move freely. As a result, the cord pulls or stretches. When that happens, people can experience:

- Difficulty walking
- Bladder and bowel problems
- Increased spasticity
- Worsening scoliosis

Although some people don't develop symptoms and don't require treatment for tethered cord, about 40 percent of people with tethered cord develop significant symptoms. They require detethering surgery performed by a neurosurgeon. Watch for the symptoms listed above.

Seizures

Approximately one in 20 people with spina bifida develops seizures. Those at highest risk are people who've had shunt problems, central nervous system infections, or a history of apnea (temporary absence of breathing). A pediatric neurologist (a doctor who treats brain and nervous system problems) typically manages seizures with medicines called anticonvulsants.

Eye Problems

Children with spina bifida are at greater risk of developing a condition called strabismus (sometimes called lazy eye). In addition, hydrocephalus can put pressure on the optic nerve and cause vision problems. Discuss vision problems with a pediatrician or primary-care physician, who might refer you to an ophthalmologist (a doctor who specializes in vision and eye problems).



Laura Gueron, occupational therapist, teaches an adult patient how to use a long-handled mirror to inspect her feet for skin problems, such as blisters, cracks or swelling.

Latex Allergies

Children with spina bifida are at greater risk of developing allergies to natural rubber or latex products. Allergic reactions can be life-threatening. Reactions also might be limited to mild itching, a runny nose, watery eyes and/or a skin rash. People with spina bifida should avoid exposure to latex products, including latex catheters, gloves, rubber balloons and toys. Discuss concerns with a pediatrician or primary-care physician.

Skin Problems

Because myelomeningocele affects sensation, skin problems can develop from prolonged pressure on a particular area of the body. Abrasions, burns, frostbite and other injuries also can be more severe and heal more slowly in people with myelomeningocele. To avoid a problem, people with myelomeningocele should perform regular skin inspections — especially of the feet and buttocks — and learn good skin-care practices. Report such symptoms as sores, irritation, blisters, cracks, peeling, swelling, and red, black or purple discolorations to your doctor. Early treatment can prevent serious infections of the skin and underlying bones.

Weight Problems

As children with spina bifida grow older and larger, their ability to walk often decreases. As their activity decreases and appetite increases during adolescence, weight gain can further decrease walking ability. Children with spina bifida are typically shorter than other children of the same age, contributing to the risk of obesity. It's important to establish good eating habits and a regular exercise program early in childhood to avoid obesity later. Discuss concerns with a pediatrician or primary-care doctor. Gillette also has registered dietitians and therapeutic recreation specialists who help patients develop good nutritional habits and exercise programs.

Learning Problems

People with spina bifida can experience problems with eye-hand coordination, difficulty understanding spoken language, hyperactivity, poor attention spans, poor memories and difficulty with organization and problem-solving. Discuss problems with teachers and pediatricians. Special-education services and/or medicines might be helpful.

Common Tests

People who have spina bifida see a wide variety of specialists who might perform tests to help diagnose and treat problems. This section explains a few of the most common tests associated with spina bifida.

Computerized tomography (CT) scan: A CT scan uses X-rays to produce detailed pictures of structures inside the body. Doctors use CT scans of the head to check the size of the brain's ventricles. Enlarged ventricles indicate hydrocephalus.

Cystometrogram/electromyogram (CMG/EMG): These tests help doctors evaluate the ability of the bladder to store and release urine. Testing involves filling the bladder with normal saline. Machines record the pressure inside the bladder and the activity of the rectal muscle.

Magnetic resonance imaging (MRI): This test uses a powerful magnetic field and radio signals to create detailed pictures of structures in the body, including the brain, spinal cord and joints, including the joints of the spine.

Manual muscle testing: Physical therapists perform manual muscle tests using devices that can detect forces produced by muscles. The tests also check for muscle weakness.

Shunt series: This test involves X-rays that show two views of the head and one view of the chest. The images allow doctors to view shunt tubing.

Ultrasonography: Ultrasonography uses high-frequency sound waves to outline structures in the body. With ultrasonography, doctors often can diagnose spina bifida and hydrocephalus before a baby is born. Doctors also can use ultrasonography to view structures in the brains of infants whose fontanels (soft spots) are open. Urologists use ultrasonography to evaluate the kidneys and bladder.

Voiding cystourethrogram: This X-ray of the bladder enables doctors to rule out reflux (urine backing up into the kidneys). Before taking the X-ray, the provider uses a catheter to fill the bladder with contrast solution.

Center for Spina Bifida

As the largest spina bifida program in the five-state area, Gillette’s Center for Spina Bifida leads the region in providing comprehensive specialty health care. Our specialists work together to provide services tailored to each patient’s and family’s needs. Moreover, we show our commitment to lifelong care by offering transition-planning and health-care services for teens and adults who have spina bifida.

Our Team Approach

Gillette’s Center for Spina Bifida includes experts in:

- Assistive technology and seating
- Casting
- Child life and therapeutic recreation
- Endocrinology
- Gastroenterology
- Genetic counseling
- Neurology
- Neurosurgery
- Nursing
- Occupational therapy
- Ophthalmology
- Orthopaedics
- Pediatrics
- Physical therapy
- Psychology
- Rehabilitation medicine
- Social work
- Urology

Lifelong Needs, Lifelong Care

The effects of spina bifida require lifelong management and care. We often begin seeing patients as infants and continue providing age-appropriate care throughout their lifetimes. As our patients approach adulthood, we support families by discussing independence and the transition to adult-focused health care.

Gillette Lifetime Specialty Healthcare continues Gillette’s tradition of excellence with services tailored to the specific needs of adults. In addition to specialty physician services, the clinic offers rehabilitation therapy, assistive



Gillette's Center for Spina Bifida brings together a caring team of professionals from a wide variety of disciplines. We work together to offer comprehensive care for every patient.

technology, counseling, nutrition services and a fully accessible radiology suite for adults who have disabilities. The clinic accepts patients 16 and older. For more information about our services for adults, call 651-636-9443.

Our Evaluation Guidelines

The chart on pages 16 and 17 provides general guidelines for when to seek evaluations with various specialty providers. It's important for patients with spina bifida and their families to actively seek out needed services and maintain a regular schedule of visits. If you'd like assistance coordinating your appointments, contact Gillette's care coordinator at 651-602-6889.

Specialty Care Evaluation Guidelines

Specialty	First Visit	Up to 12 Months	1 to 6 Years
Pediatrics and Developmental Pediatrics	As needed	As needed	As needed
Neurosurgery	By 6 weeks	Every three months	Every six months to one year
Orthopaedics	By 6 weeks	Every three months	Every six months to one year
Urology	By 6 weeks	At 3, 6 and 12 months	Every six months
Rehabilitation Medicine	By 6 weeks	Every year	Every year
Genetic Counseling	As needed		
Ophthalmology	At 9 months		Before school begins
Physical Therapy	By 6 weeks	Every six months	Every year
Occupational Therapy	By 6 weeks	Every six months	Every year
Therapeutic Recreation	Between 4 and 6 years		Once between 4 and 6 years
Speech Therapy	At 3 years		Language testing at 3 years
Psychology	At 5 years		At 5 years
Social Work	Once during teen years, if not sooner	As needed	As needed
Assistive Technology	As needed	As needed	As needed

Depending on the results of each evaluation, patients might need additional visits to receive services. In addition to these evaluations, be sure to see your primary-care provider regularly.

7 to 12 Years	13 to 16 Years	16 Years Through Adulthood
As needed	As needed	Not applicable
Every year	Every year	Every year
Every six months to one year	Every six months to one year	Every year
Every six to 12 months	Every year (discuss sexuality concerns)	Every year (discuss sexuality concerns)
Every year	Every year	Every year
		As needed
As needed	As needed	As needed
Every year	Every year	Every year
Every year	Every year	Every year
As needed	Once during the teen years	As needed
As needed	As needed	As needed
As needed	As needed	As needed
As needed	Once, by referral, to discuss transition to adult-focused health care and services	Every year (includes screening for mental-health needs)
As needed	As needed	As needed

Information & Referrals

For information about Gillette's Center for Spina Bifida, call 651-229-3878 or 800-719-4040 (toll-free). To refer a patient, call 651-229-3944. For information about Gillette Lifetime Specialty Healthcare for teens and adults, call 651-636-9443.

Additional Resources

- Spina Bifida Association of America
800-621-3141 (toll-free)
www.sbaa.org
- The Hydrocephalus Association
888-598-3789 (toll-free)
www.hydroassoc.org

LEARN MORE!

For more information
about spina bifida,
visit our Web site at
www.gillettechildrens.org.



Centers of Excellence at Gillette

Treating people who have disabilities and complex medical conditions requires a team approach. At Gillette, our doctors, surgeons, nurses, therapists, psychologists, social workers and other specialists work together to care for patients. Throughout our centers of excellence, we offer leading-edge medical treatments tailored to the unique needs of each patient.

Gillette's centers of excellence:

- Center for Cerebral Palsy
- Center for Craniofacial Services
- Center for Gait and Motion Analysis
- Center for Pediatric Neurosciences
- Center for Pediatric Orthopaedics
- Center for Pediatric Rehabilitation
- Center for Pediatric Rheumatology
- Center for Spina Bifida



Gillette Children's
Specialty Healthcare

St. Paul (Main) Campus
200 University Avenue East
St. Paul, MN 55101
651-291-2848
800-719-4040 (toll-free)

Burnsville Clinic
305 East Nicollet Boulevard
Burnsville, MN 55337
952-223-3400
866-881-7386 (toll-free)

Duluth Clinic
Lakewalk Center
1420 London Road
Duluth, MN 55805
218-728-6160
800-903-7111 (toll-free)

Minnetonka Clinic
6060 Clearwater Drive
Minnetonka, MN 55343
952-936-0977
800-277-1250 (toll-free)



Gillette Lifetime
Specialty Healthcare

New Brighton Clinic
550 County Road D
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www.gillettechildrens.org