HYDROCEPHALUS CANADA
Championing Spina Bifida & Hydrocephalus Communities

Understanding Spina Bifida

What is Spina Bifida?

Spina bifida is a condition that people are born with and affects them throughout their lives. There is no cure but people with the condition can live happy, productive, and fulfilling lives.

Approximately 26 in 100,000 babies in Canada are born with some form of the condition. It is not a disease and cannot be spread like an infection or illness.

Spina bifida is classified as a neural tube defect (NTD). The neural tube develops into the brain, the long bundle of nerves that make up the spinal cord, and the tissues that enclose them. In most babies, the neural tube forms by the 28th day after conception.

With spina bifida, a portion of the neural tube does not develop or close properly. Bone, muscle and skin cannot form around the spinal cord where the tube is open. This incomplete formation of the spinal cord creates an opening through which nerves and spinal fluid may protrude and creates a sac covered by skin or a thin membrane on the unborn baby's back called a cele. Nerves around the cele may by damaged or improperly formed.

The location of the cele will determine the number of spinal nerves involved. The higher up the cele is formed, the more damage is likely. The impact on a person can range from mild to severe and may include:

- partial or total paralysis
- loss of sensation in the legs
- challenges with bowel and bladder function
- difficultly with hand skills, vision and hearing
- learning challenges

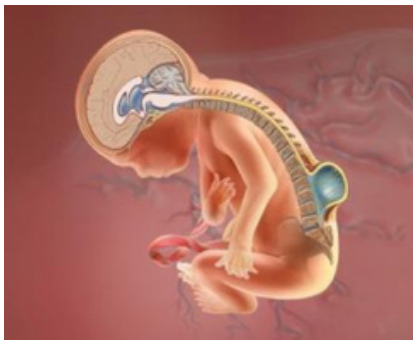
What Causes Spina Bifida?

The exact cause of spina bifida is not known. Although medical research has confirmed a link between women not getting enough folic acid before and during early pregnancy, no single factor is believed to be responsible for the condition.

Development of the condition is likely the result of a number of factors. These may include genetics of both parents, the mother's diet, and a number of yet unidentified environmental factors present before and during conception and early pregnancy. There may also be other unknown factors that contribute to development of the condition.

Breakthrough Treatment

Babies born with spina bifida are usually transferred to a specialized children's hospital. If the skin is not completely closed over the spinal cord and nerves, neurosurgeons repair the sac during the first few days of life. This surgery reduces the risk of infection and further damage to the spinal cord and nerves. It is important



Credit: The Children's Hospital of Philadelphia

to note that surgery cannot repair the nerves that are already malformed. This damage is permanent.

One of the most recent and exciting developments in the history of treatment for spina bifida is the introduction of in-utero fetal surgery. In some babies diagnosed before 25 weeks gestation, surgery to close the opening in the baby's back while they are still in the womb may be an option. Because spinal cord damage can worsen during the baby's development, repair of the opening while the baby is still in the womb may prevent further damage and may offer significantly better results than traditional postnatal repair. It may also reduce the need to divert fluid from the brain (hydrocephalus), improve mobility and improve the chances that the individual will be able to walk independently.

This surgery recently became available in Canada. If you are given a prenatal diagnosis of spina bifida, talk to your health care provider for more information about your options as soon as possible.

Types of Spina Bifida

Spina bifida is a life-long condition with ongoing medical challenges. Each of the four main types of spina bifida, listed below, affect individuals differently.

Occulta

This is a form of spina bifida where a small hole in the lower segment of the spine and the bones are not properly closed. It may involve an abnormality of the vertebrae or vertebrae and spinal cord. Many people have this condition and only become aware of it if they experience unexplained incontinence, chronic back ache, or changes in the muscles of their legs.

Lipomyelomeningocele

In lipomyelomeningocele, there is a protrusion of abnormal fatty tissue through a defect in the vertebrae that forms in the buttocks or lower spine. Damage to the nerves may occur from compression of the nerves by the fatty mass or abnormal formation of the spinal cord.

Symptoms of lipomyelomeningocele may include a pinkish patch, dimple or skin tag in the lower spine area, muscle weakness of legs, foot deformities, loss of sensation in patches on the lower legs, feet and buttocks, back pain, and bladder and bowel incontinence.

Meningocele

In meningocele, the bones do not close fully around the spinal cord. The meninges (coverings of the spinal cord) are pushed out through the opening to form a sac containing cerebrospinal fluid (CSF). No portion of the spinal cord is pushed out into the sac – the spinal cord remains in the spinal column and the nerves may not be as severely affected. The sac is often covered by skin. There may be motor or sensory changes after the sac is surgically repaired.

Myelomeningocele

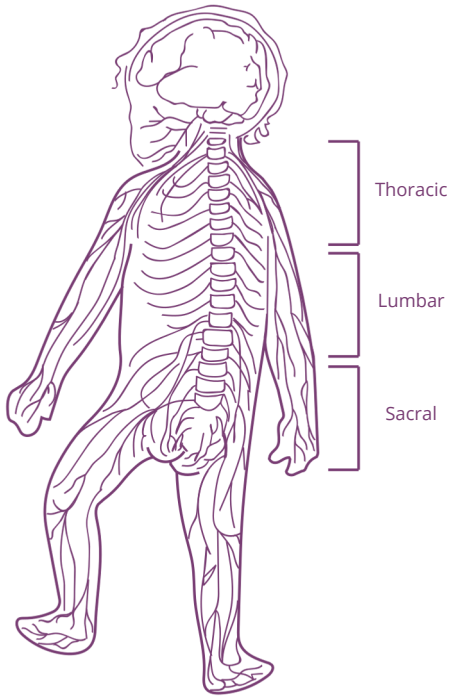
Myelomeningocele is the most severe form of spina bifida whereby the bones fail to close around the spinal cord. The meninges and the spinal cord protrude through the opening in the spine to form a sac. This sac contains cerebrospinal fluid (CSF). This sac is usually transparent and not covered by skin. The spinal cord fails to develop properly and spinal nerves are damaged.

Folic Acid May Help Prevent Spina Bifida

While there is still no single known cause for the development of spina bifida, folic acid has been proven to reduce the risk of neural tube defects (NTDs), by as much as 70%. Women of child-bearing age, who are sexually active, should take a daily multi-vitamin which contains at least 0.4 mg of folic acid, three months prior to conception and during the first 3 months of pregnancy. Any woman with a family history of NTDs, a previous pregnancy affected by an NTD, a personal or family history of NTDs (or your partner has such a history), insulin-dependent diabetes, who uses certain anti-seizure medications, is clinically obese, abuses alcohol or is of Celtic, Northern Chinese, Cree or Sikh heritage is at increased risk and should consult a physician about folic acid.

Understanding How Location Impacts Nerve Damage

Spinal nerves	Area of the spinal cord where the nerves come from:	Spinal nerves carry messages to and from the muscle in the:
Thoracic nerves T10 - T12	The upper back	Chest, back and stomach
Upper lumbar nerves L1 - L3	The mid back	Hips and front of thighs
Low lumbar nerves L4 - L5	The lower back	Knee and front of lower leg
Sacral nerves S1 - S3	The lowest part of the spinal cord	Ankles, feet, calves, buttocks, bladder and bowel



Additional Health Challenges Associated with Spina Bifida

Syringomyelia or Syrinx

Sometimes an abnormal pocket of cerebrospinal fluid (CSF) forms inside the spinal cord. This causes pressure within the central canal of the spinal cord and may result in increased scoliosis and changes in sensation such as “pins and needles” and/or weakness of the hands, arms.

Chiari II Malformation

For people with spina bifida, the brain stem (lower part of the brain) is lower than usual and compresses the upper part of the spinal cord in the neck. Most individuals with myelomeningocele have chiari II malformation. A small percentage of these individuals may experience aspiration (food and liquid enter the lungs), apnea (stop breathing for more than 5 to 10 seconds), hypersensitivity to objects in the mouth, light and loud sounds, gagging, choking, vomiting, stridor, tightness and/or weakness of arms, arching of head backwards, and/or a weak suck when bottle or breast feeding in babies.

Tethered Cord

In people with spina bifida, the spinal cord can get stuck at the site of the cele and is often stretched over time. This can lead to injury of the spinal cord, which will cause symptoms called Tethered Cord Syndrome. Symptoms may include bladder changes (increased urgency and frequency and urinary wetness, urinary infections), bowel changes (urgency and frequency in bowel soiling, changes in stool consistency), back pain, leg and/or foot changes and, scoliosis.

Bladder Challenges

Bladder function is controlled by the nerves at the bottom of the spinal cord. Almost all people with spina bifida have issues with bladder function. Nerve damage may limit bladder and sphincter muscle control and reduce sensation with the following conditions potentially developing and changing over time:
Small Spastic Bladder which occurs when the bladder is unusually small and holds little urine. This results in uncontrolled bladder muscle contractions that may lead to back up of urine to the kidneys, enlarged kidneys and wetness.
Large flaccid bladder which occurs when the bladder becomes unusually large with a relaxed sphincter. This results from an inability of the bladder muscles to contract properly. This may lead to frequent infections.

Bowel Challenges

In people with spina bifida, bowel muscle control and sensation varies greatly depending on the location of the cele. When the anal sphincter muscles are weak, or there is decreased sensation due to nerve damage, it may not be possible to “feel” the need to have a bowel movement and “accidents” or fecal incontinence may result. Conversely, when the muscles of the bowel are weak, waste material moves more slowly through the bowel and more water is absorbed by the body causing constipation which can be severe.

Hydrocephalus

An essential liquid called cerebrospinal fluid (CSF) delivers important nutrients and chemicals to, and removes waste products from, the brain. When too much CSF accumulates, a chronic neurological condition called hydrocephalus develops and can cause serious damage to the brain impacting its ability to function.

More than 85% of people with spina bifida also have hydrocephalus. Brain surgery to insert a shunt that diverts CSF to another part of the body or provides an alternate path for CSF to exit the brain are currently the only treatments.

Walking

Many children with spina bifida can stand and walk. The ability to walk will vary depending on the location of the cele and extent of the damage to the nerves.

Level of Spina Bifida

S2-S4	Usually walks. May need shoe inserts.
L5-S1	Usually walk with ankle foot orthoses (AFOs) or short ankle braces. May use crutches.
L4	Can usually walk with AFOs or knee ankle foot orthoses (KAFOs) or braces above knee. Often use wheelchair for long distances.
L2-L3	Usually need wheelchair for long distances. May walk with braces above knee or hip when younger.
T12-L1	Usually require wheelchair. May walk at home or for exercise with braces above hip when younger.

Planning for the Future

Spina bifida brings lifelong challenges. It is important to remember that these challenges can be managed.

Attitudes and expectations, assistive devices, community supports and a host of services all play important roles in the self-esteem of each individual's independence and quality of life.

Participation at school, work and the community as well as knowing when to ask for assistance are all important activities that can contribute to experiencing the best health and life possible.

About Hydrocephalus Canada

Our goal is to empower individuals impacted by spina bifida and hydrocephalus to experience the best life possible. We believe everyone affected:

- has the right to our attention, compassion and commitment
- has value and deserves to be treated with dignity
- requires and deserves access to safe, effective care
- benefits from, and offers benefit to, collaborative communities
- has the responsibility to help everyone understand the conditions

Our Current Efforts are Focused on:

- increasing general awareness of spina bifida and hydrocephalus
- advocating for solutions to support prevention; early accurate diagnosis; access to appropriate treatment; optimal outcomes; and ultimately a cure
- developing education and support tools for patients, caregivers, healthcare professionals, policy makers and media
- establishing supportive communities that encourage inclusive, proactive conversations, activities, and
- funding meaningful, breakthrough research

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This educational resource was created in partnership with the health care professionals in the Spina Bifida Service at Holland Bloorview Kids Rehabilitation Hospital.