A Booklet on Spina Bifida

BC Children's Hospital™

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Table of Contents

Section 1:
What spina bifida means for your child

What is spina bifida?

Spina bifida is a condition that affects the **nervous** system. The nervous system is made up of the brain, spinal cord and nerves throughout the body. Babies with spina bifida are born with a portion of their spinal cord exposed. The exposed spinal cord is at risk for injury even before the baby is born.

Different amounts of the spinal cord may be exposed. **Myelomeningocele** is another word for spina bifida where the spinal cord and protective membranes are exposed. **Meningocele** is another word for spina bifida where only the protective membranes are exposed. There are other types of spina bifida as well, such as a lipomeningocele or occulta.

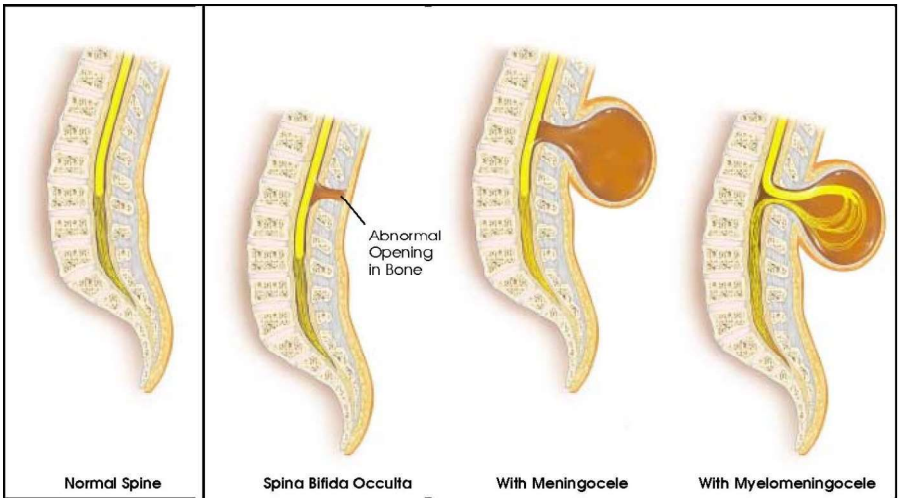

Figure 1: A normal spinal cord compared to three variations of spina bifida

Different regions of the spinal cord may be exposed. A **thoracic** opening is higher up on the spine. A **lumbar** opening is in the middle of the spine. A **sacral** opening is at the base of the spine.

The type of injury to the nervous system depends on amount of the spinal cord exposed and the region that is exposed.

- A myelomeningocele affects the nervous system more than a meningocele.
- A thoracic opening affects the nervous system more than a lumbar opening.
- A lumbar opening affects the nervous system more than a sacral opening.

What does spina bifida mean for your child?

Since the spinal cord is affected in babies with spina bifida, several body systems can be affected. The spinal cord is the connection between the brain and the rest of the body. The body sends sensory messages to the brain through the spinal cord and the brain sends back messages to the muscles of the body through the spinal cord.

Some of the areas commonly affected are the bladder, bowel, the legs and circulation of cerebrospinal fluid. See Section 3 for more details about the challenges of spina bifida.

How does spina bifida happen?

A baby's spinal cord is developed by 28 days of gestation. Experts are not completely sure how the spinal cord develops. From the information we have, spina bifida seems to be the result of a spinal cord that does not develop completely rather than a spinal cord that develops completely and then opens up.

Spina bifida can often be seen on a prenatal ultrasound at 20 weeks gestation. Some other signs on a prenatal ultrasound that a baby may have spina bifida are the *banana sign* — abnormal shape of a portion of the brain — and the *lemon sign* — flattening of frontal bones of the skull. Not all spina bifida defects are detected on ultrasound and not all spina bifida show the banana or lemon sign.

Spina bifida is a multifactorial condition, which means it is caused by **both genetic** and **environmental** factors. The combination of the two factors explains why you usually do not see spina bifida "running" in a family like other purely genetic conditions. However, because genetic factors do play a role, having one baby with spina bifida changes the chances of having another baby with spina bifida. The chances change from 1/1000 (0.1%) to approximately 1/40 (~3%). So there is still an approximately 97% chance that the next pregnancy will be healthy.

The most important point to remember is that **spina bifida is nobody's fault**. Nobody does anything to cause their baby to have spina bifida.

Experts have found that taking 1 mg folic acid daily 3 months before a pregnancy significantly decreases the chances (~70%) of having a baby with spina bifida; however, it cannot prevent it completely. Basically it is recommended that **all women who are capable of having a baby should be taking folic acid supplements.**

Section 2:
What spina bifida means for parents

Changes for a parent

Parents have dreams and expectations for their children. A parent who has a child with spina bifida can still have dreams and expectations but these may or may not be the same dreams and expectations they had in the beginning.

It is normal for parents to cope differently with the news that their dreams and expectations may have to change. Parents may have different feelings and these feelings may surface at different times as well. Several different types of feelings are going to surface. The important point is that all feelings such as anger, grief, confusion, and sadness are normal and all of them should have a chance to be expressed. **Communication is important.**

Changes for a family

Parents need to think about the amount of time a child with spina bifida may need. A child with spina bifida will require more of a parent's time than most other children. In a big family this does not necessarily mean taking time away from other children. It may just mean some re-organizing. In fact, it is important that a child with spina bifida have as much independence as possible. There are many tasks that can be learned.

Parents need to remember that they need to take care of themselves too. There are services available, such as respite care (someone can look after you child while you take a break).

Section 3: Some of the challenges

Hydrocephalus, shunts & other neurological challenges

Hydrocephalus is the buildup of **cerebrospinal fluid (CSF).** This is a problem because a baby's brain only has so much room to grow in the skull. The extra CSF surrounding the brain takes up space, resulting in less room for the brain to grow.

Hydrocephalus is caused by a difference in the position of a baby's brain. The brain sits lower in the skull than it normally does and blocks the fluid from circulating into the spinal cord. This position of the brain is known as the **chiari II malformation.**

Hydrocephalus can develop at any time in a person's life. Often it develops near birth. One of the first signs of hydrocephalus includes a larger head size.

Hydrocephalus cannot be cured, but it can be treated. A device called a **shunt** creates a passageway for the CSF to drain from the space around the brain. A shunt is placed inside the body with surgery. It usually remains inside for the length of someone's life. Enough tubing is placed inside to grow with a person.

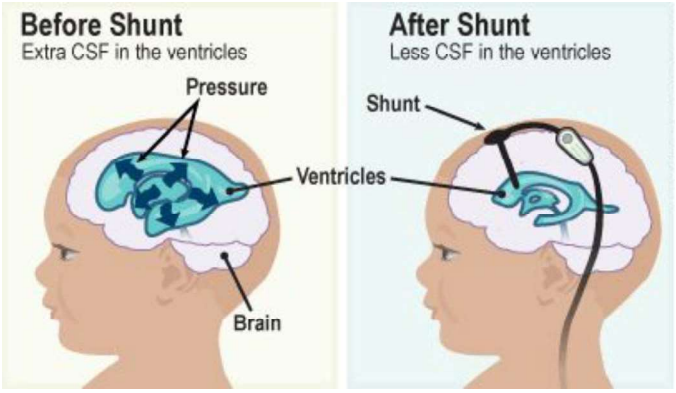

Figure 2: Examples of what a shunt may do to help a child with excess CSF

There are a few reasons for why a second surgery may be needed. There may be a mechanical failure of the shunt or an infection. A second surgery would be required to fix the shunt or replace it in those cases.

Untreated hydrocephalus can affect intelligence and the development of the brain. If hydrocephalus is caught early enough a baby's brain can develop normally and intelligence usually develops within the normal range. The normal range for IQ is between 80 and 120. Children with spina bifida tend to score lower in the normal range but within the normal range. This is regardless of whether they have a shunt or not.

Other neurological challenges may include the development of a **tethered cord, syringomyelia** or **seizures**:

- A **tethered cord** occurs when tissue from the spinal cord becomes attached to the bones of the spine causing the brain to sit lower than it should.
- **Syringomyelia** is the build-up of CSF in the spinal cord.
- **Seizures** are the result of abnormal changes in brain activity and can be caused by several different factors independent of spina bifida.

These other neurological challenges may occur alone, together or not at all.

Mobility & mobility equipment

One of the most common questions parents ask is if their baby is going to walk. This question is also one of the most difficult to answer, especially in the prenatal period. On a prenatal ultrasound the level of the spina bifida is usually visible. The **level** of the spina bifida can give clues as to whether a baby is going to walk.

- Babies with **thoracic level** spina bifida may walk when they are younger, but often use a wheelchair for their main method of transport.
- Babies with **lumbar level** spina bifida may walk when they are younger as well, but their main method of transport will really depend on the individual. Some will be able to get around on crutches while others may use wheelchairs.
- Babies with **sacral level** spina bifida may need to use crutches to move around, but may use a wheelchair for longer distances.

Children with spina bifida may not have the mobility other children have to develop strong bones and muscle. Working with a **physiotherapist** can help develop the bones and muscles needed for getting around. A physiotherapist is an individual trained in working with different muscle groups.

Children with spina bifida, with different ranges of mobility, can participate in most physical activities other children are involved in. Many children learn to swim, dance and participate in team sports like wheelchair baseball or basketball. Use caution when considering physical activities with direct physical contact such as ice/floor hockey, football or wrestling. Direct physical contact may affect the function of a shunt.

Orthotic equipment can help children with spina bifida move around. **Casts** can be used to correct the position of feet. **Braces** can be used to preserve the joints. There are several types of braces commonly used. **Ankle Foot Orthotics (AFOs)** are used to brace the ankle joint. **Knee Ankle Foot Orthotics (KAFOs)** are used to brace the ankle and knee joints. There are also braces for standing only. Other types of mobility equipment include **canes**, **crutches**, **walkers** and **wheelchairs**. Sometimes surgery may also be necessary to help with mobility.

Bladder management

This body system is usually affected in children with spina bifida. A **neurogenic bladder** does not know that it is full and needs to let the urine out or that it needs to hold the urine until an appropriate time to empty.

Most children will have to use a **catheter** to empty their bladder. Catheterizing means that a thin tube is placed into the urinary tract each time the bladder needs to be emptied. Some babies need to be catheterized, while some babies will not need it until they are older. Many children are capable of catheterizing themselves. Using a catheter is usually painless, since most children with spina bifida do not have sensation in their urinary tract.

Sometimes using a catheter may not be effective for bladder management. There are medications that can relax the bladder muscles or tighten the bladder muscles. Bladder surgery may also be needed to help with bladder control. Bladder surgery to create a **Mitrofanoff stoma** allows a person with spina bifida to catheterize through the abdomen instead of through the urinary tract.

Proper bladder control is important. Children with spina bifida are at greater risk for **urinary tract infections (UTIs)** as well as other urological problems such as **urinary reflux** and **hydronephrosis**.

- **Urinary reflux** occurs when the bladder is too full and urine is pushed back towards the kidneys. Urinary reflux can lead to hydronephrosis.
- **Hydronephrosis** is the name of the condition where the kidneys retain urine. Hydronephrosis can cause damage to the kidneys.

Proper bladder control is also important for social reasons. Clothing wet by urine can make it difficult for children to make friends.

Bowel management

Similar to the bladder system, the bowel system is usually affected in children with spina bifida. A **neurogenic bowel** does not know when it needs to empty and whether or not it is an appropriate time to empty. Achieving **bowel continence**, the ability to empty bowel movements voluntarily, is important in avoiding constipation or diarrhea. There are several programs that can help your child learn and maintain bowel continence. These programs involve:

- Potty training;
- Learning about a diet that promotes soft stools; and
- Medications such as enemas, suppositories and laxatives.

Bowel surgery may also be an option. A **cecostomy** stoma allows an enema to be used from the abdomen instead of through the rectum.

Bowel continence (control of bowel movements) is important for social reasons. Full bowel continence may be not achievable, but achieving "social" continence may be the goal. Social continence is the concept of controlling bowel movements to the extent that a child can participate in social activities with other children. Achieving bladder and bowel continence also establishes a sense of independence for a child **with spina bifida.**

Other Medical Challenges

As children with spina bifida get older, often they are **less mobile** and gain weight easily. The extra weight makes it more difficult for a child to move causing a cycle of weight gain. It is important to help your child maintain a healthy diet and support an active lifestyle.

Some children with spina bifida have **eye challenges** such as **strabismus** and **nystagmus.**

- **Strabismus** is the inability of both eyes to line up properly for normal vision. It can usually be treated by patching the eye that does not line up properly
- **Nystagmus** describes an eye that moves rhythmically and rapidly on its own, Currently there is no approved effective treatment for nystagmus in children. Nystagmus, however, occurs less commonly than strabismus.

Another medical challenge is **skin care.** Children with spina bifida do not have a strong **sense of touch.** The sense of touch is a natural defense mechanism for the skin. When a child with spina bifida accidentally wets himself or herself, he/she may not be able to tell. Trapped moisture can cause the spread of infections. A child with spina bifida may not notice that the bath water is too hot, or that there are sharp objects on the ground. If a child with spina bifida is sitting in one position for a long time, he or she may not feel uncomfortable. Sitting in one position for a long time can cause a pressure sore to appear. It is important to take care of the skin as it is one form of protection against germs.

Section 4: The Spina Bifida Team

Here at BC Children's Hospital, we have a team of professionals that provide support to parents and children with spina bifida. The spinal cord clinic includes:

- **Pediatrician,** a doctor who specializes in the care of children.
- **Nurse Clinician/Coordinator,** provides counseling, teaching, emotional support and advocacy for your child, you and your family.
- **Neurosurgeon,** a doctor that specializes in the nervous system.
- **Orthopedic surgeon**, a doctor that specializes in the skeletal system.
- **Urology surgeon,** a doctor that specializes in the bladder system.
- **Physiotherapist,** specializes in assessing your child's motor (muscles and movement) and sensory abilities and your child's bracing and mobility needs.
- **Occupational Therapist,** helps your child adapt to activities of daily living
- **Social Worker,** helps your child, you and your family with the non-medical parts of your life.

Glossary

Banana sign	Abnormal shape of the brain as seen in a prenatal ultrasound that is a sign of a spina bifida.
Bowel continence	The ability to empty bowel movements voluntarily.
Catheter	A thin tube placed to empty the bladder. The catheter can be placed into the urinary tract or through a stoma (hole) surgically made in the abdomen.
Cecostomy	A stoma (hole) surgically made to allow an enema to be used from the abdomen instead of through the rectum.
Cerebrospinal fluid (CSF)	Fluid made by the brain to produce nutrients for the brain and spinal cord.
Chiari II formation	The position of the brain where it sits lower in the skull than it normally does and blocks the fluid from circulating into the spinal cord.
Clean intermittent catheterization	A procedure of inserting a small catheter/tube into the bladder. It is inserted through the urethra to completely drain urine.
Gestation	The time of pregnancy, between fertilization and birth.
Hydrocephalus	The buildup of cerebrospinal fluid in the skull. One of the first signs of hydrocephalus is an enlarged head circumference (distance around the head).

Hydronephrosis	Condition where the kidneys retain urine and can cause damage to the kidneys.
Lemon sign	Abnormal shape of the brain showing a flattening of frontal bones of the skull as seen in a prenatal ultrasound. This is a sign of a spina bifida.
Lipomeningomyelocele	A skin covered lesion containing a lipoma, or fatty tumor, usually located over the lumbar-sacral spine.
Meningocele	A type of spina bifida where only the protective membranes are exposed.
Meningomyelocele	A type of spina bifida where the spinal cord and protective membranes are exposed.
Mitrofanoff stoma	A hole that allows a person with spina bifida to catheterize through the abdomen instead of through the urinary tract.
Neurogenic bladder	When the bladder does not know that it is full and needs to let the urine out or that it needs to hold the urine until an appropriate time to empty.
Neurogenic bowel	When the bowel does not know that it is full and needs to have a bowel movement or that it needs to hold the bowel movement until an appropriate time to empty.
Nystagmus	A condition where the eye moves rhythmically and rapidly on its own.
Occulta	This is the mildest form of spina bifida.

Orthotic equipment	A device that is used to stabilize a joint or maintain alignment and position around the joint. This equipment is made of lightweight material. It is named for the joint that it supports (ankle orthotic or knee orthotic).
Seizure	Abnormal changes in brain activity causing a physical convulsion and other symptoms. They can be caused by several different factors independent of spina bifida.
Shunt	A device that creates a passageway for cerebrospinal fluid to drain from the space around the brain. A shunt is placed inside the body with surgery.
Strabismus	The inability of both eyes to line up properly for normal vision.
Syringomyelia	The buildup of cerebrospinal fluid in the spinal cord.
Tethered cord	Tissue from the spinal cord becomes attached to the bones of the spine causing the brain to sit lower than it should.
Urinary reflux	When the bladder is too full and urine is pushed back towards the kidneys.