SPINA BIFIDA
COALITION OF CINCINNATI

About Spina Bifida

The following information is provided solely for informational purposes. The information is not intended to be medical advice on the management or care of a person with Spina Bifida. Although every effort is made to assure that information is accurate and current, knowledge in the field of Spina Bifida is growing rapidly and all data is subject to change without notice. The SBCC does not make any warranty concerning the accuracy of any information found on or exchanged through this system. Please consult your primary care physician or developmental pediatrician for specific information regarding your personal diagnosis and treatment plan.

Spina Bifida is the most frequently occurring, permanently disabling, birth defect in the United States, affecting approximately 1,500 babies each year. Spina Bifida results from the spine's failure to close properly during the first month of pregnancy. There are three forms of Spina Bifida; Occulta, Meningocele and Myelomeningocele.

In Myelomeningocele, the most severe form of Spina Bifida, the spinal cord and surrounding membranes are open to the skin surface. This opening, or lesion, is normally repaired within 24 hours after birth leaving a visible and often large scar on the back. This surgery does not "cure" the patient, but minimizes the risk of infection and preserves the existing function in the spinal nerves. Meningocele is characterized by the meninges (protective covering of the spinal cord) protruding through the lesion. There is usually no damage to the nerves. In Spina Bifida Occulta there is no open lesion, but a malformation of the bones of the vertebrae. In fact, this condition often goes undiagnosed until adulthood. There may be a hairy patch or dimple at the bony defect along the spine. In Spina Bifida Occulta and Meningocele, problems such as back pain, incontinence and muscle weakness may develop as the person ages. Lipomeningocele is a related disorder that occurs when a fatty tumor develops over part of the spine. There may be some nerve damage which usually affects the function of the bowel and bladder.

The cause of Spina Bifida is not fully understood, but is thought to be associated with both genetic and environmental factors. Recent studies have found that taking 400 micrograms of the B-vitamin folic acid prior to and during the first weeks of pregnancy may reduce the risk of neural tube defects, including Spina Bifida by as much as 70%.

Fetal Surgery Some women who are carrying a child diagnosed with Spina Bifida may be eligible to have the lesion repaired in utero. The Management of Myelomeningocele

Study (MOMS) published in 2011 showed that significant benefit can be gained from closing the lesion before the baby is born. MOMS showed that babies who had the repair between 19 and 26 weeks gestation, often just after diagnosis is made, were less likely to develop hydrocephalus and need a shunt and had improved chances of walking without crutches by age 2 1/2.The surgery, however, requires family commitment, as the mother is on bed rest after the surgery, and at risk for uterine rupture during the pregnancy. Some babies who have had the surgery are also born pre-term. These risks must be discussed with your physician. Follow up studies continue to look at the long-term benefits or risks to the children who had fetal surgery to repair their lesion sites.

More information on fetal surgery for Spina Bifida repair can be found on the Cincinnati Children's Medical Center Web site.

Arnold Chiari II Malformation is a condition commonly associated with Spina Bifida. In Arnold Chiari II Malformation, the cerebellum (responsible for balance and coordination) and brain stem are positioned further down into the spinal column than normal, compressing the tissue. Symptoms of Arnold Chiari II Malformation are stridor (noisy breathing), difficulty swallowing or gagging, sleep apnea and muscle weakness. If symptoms are problematic, a surgery may be required to remove part of the skull and relieve the pressure on the cerebellum.

Hydrocephalus can result from the Arnold Chiari II Malformation blocking the flow of cerebrospinal fluid surrounding the brain and spinal cord. It occurs in about 80% of individuals diagnosed with Spina Bifida. This blockage causes excess fluid to back up into the ventricles (cavities) of the brain, causing pressure on the brain itself. Children with pronounced hydrocephalus usually require a special device called a "shunt" to by-pass this blockage and drain away the excess fluid. A shunt is a long, narrow tube with a one-way valve. Inserted under the skin, it effectively drains the spinal fluid to a convenient location for absorption by the body - usually the abdominal cavity. This surgery is often performed before a newborn goes home from the hospital.

Shunts can be very reliable, but blockages or malfunctions can occur which may require replacement or revision. Headaches, vomiting, seizures, sleepiness, poor performance at work/school or emotional changes can be signs that a shunt has failed. However, symptoms of shunt malfunction can be deceiving, and any unexplained changes in health or behavior should be discussed with a neurosurgeon.

Neurogenic Bladder and Bowel are disorders related to Spina Bifida. The nerves that carry messages to the brain that tell the body to hold or release urine and stool are

impaired at or below the Spina Bifida lesion site. Incontinence, high bladder pressure, bladder infections, urine reflux into the kidneys, stool leakage and chronic constipation are some of the problems that result from incomplete nerve messages to the brain. These symptoms create difficult social and medical problems for affected individuals. This is a complex area to manage, and surgery, medications and toileting techniques may all be needed to achieve a successful continence program.

Individuals with Spina Bifida usually require clean intermittent catheterization (CIC) performed at regular intervals throughout each day to help keep the bladder and kidneys healthy and individuals continent. A catheter is a straw-like device inserted into the bladder to effectively drain urine. Sometimes, catheterization and medication alone are not successful in alleviating high bladder pressure. In this case, a bladder augmentation may be performed to increase the size of the bladder so that it may hold urine at a safe pressure. In this procedure, tissue from the stomach or intestine is sewn onto the bladder, creating a larger reservoir for urine. Sometimes the best solution for a neurogenic bladder is a Mitrofonoff surgery, in which an opening to the bladder is created in or near the navel. This opening (stoma) allows the bladder to be emptied without leaving a wheelchair.

Bowel function is also likely to be impaired in people with Spina Bifida. They may not sense when it is time to use the toilet or have limited control over when the bowel will empty. They also must work hard to prevent constipation through diet and medications. This aspect of Spina Bifida requires personal commitment and care. The Continent Cecostomy, also called the ACE or MACE procedure, surgically creates a channel to access the colon and flush it clean of stool using a regular irrigation or ante grade enema program. This is becoming increasingly popular as a treatment for bowel incontinence when other measures are not successful.

Tethered Cord occurs when the spinal cord is attached to scar tissue or fatty growths at the lesion site, preventing the cord from ascending normally. As a child grows, the spinal cord can become stretched, causing damage and interfering with its blood supply. This can result in back, neck or leg pain, changes in leg strength, progressive or repeated muscle contractions, bone deformities of the legs and back (such as scoliosis or lordosis), as well as new bowel and bladder problems. Surgical repair of tethered cord may be performed if significant symptoms are present. This may not restore lost function, but should stop further loss of ability.

Lower Limb Paralysis and orthopedic abnormalities can occur when the nerves are damaged at and/or below the point at which the lesion occurs. Generally, the higher the lesion is located on the spine, the greater the level of paralysis to the lower

limbs. Clubfoot, dislocated hips, spinal curvatures, and contractures (tightness) in the hips, knees and/or ankles are often found in individuals with Spina Bifida. Most people with Spina Bifida will need leg braces or more involved orthotics or assistive devices to walk. Braces can help to support and protect weak muscles or joints and assist with proper alignment of the bones. Physical therapy may be indicated to stretch muscles and keep them in balance. Various orthopedic surgeries may also be necessary to facilitate correct positioning. Many people with Spina Bifida use walkers, forearm crutches or wheelchairs for mobility to help them keep up with their able-bodied peers.

Skin sensation is often absent or irregular in the same area as the paralysis. People with Spina Bifida must be diligent in checking their skin for abrasions, pressure sores and burns, which can be dangerous, if left untreated. Maintaining a healthy weight, moving positions often and regular exercise can help to prevent pressure sores.

Learning Challenges are also associated with Spina Bifida. Individuals often have difficulty with initiation and executive function: the ability to make a plan and carry it out. Because of this, a person with Spina Bifida may be misdiagnosed as lazy or uncooperative. People with Spina Bifida can also have trouble with fine motor skills (such as handwriting and manipulating buttons or fasteners), mathematics, reading comprehension, sequencing, attention and memory. Many of these difficulties mirror a learning disability called Non-verbal Learning Disorder (NLD).

Although people with Spina Bifida generally have normal intelligence, these academic challenges can lower their performance on the job or in the classroom. Verbal step-by-step instruction, highlighting important reading content, using graph paper to organize and align math problems and giving background information about the text can be useful tools for a student. Adults with Spina Bifida may find a planner or hand-held computer device helpful in getting through daily activities. Individuals with Spina Bifida may also have trouble reading non-verbal cues (body language) or picking up subtle inferences. This can create problems in relationship building, especially as they become teens and adults.

Latex Precaution, the avoidance of products containing natural rubber, is advised for all individuals with Spina Bifida. Research has shown that they have an increased potential to become allergic to latex. Allergic responses include watery eyes, wheezing, hives, rash, swelling and in severe cases, anaphylaxis (life-threatening restriction of the air passages). These responses can occur when items containing latex touch the skin, mucous membranes (such as the mouth), genitals, bladder, rectum, wounds or surgical openings or the bloodstream. Some common products that may contain latex include balloons, rubber bands, erasers, Koosh balls, gym balls, and medical and cleaning

gloves. A complete listing of items containing latex can be found on the website of the Spina Bifida Association, www.spinabifidaassociation.org.

Outcome: It takes a team of dedicated parents, teachers, therapists and medical support provided by comprehensive Spina Bifida Clinics like the Spina Bifida Clinic at Cincinnati Children's Hospital Medical Center, as well as the commitment of individuals with Spina Bifida themselves, to achieve their maximum potential. Adults with Spina Bifida have a wide range of abilities, need varying degrees of intervention and experience unique and medical complications throughout their lives. However, many can expect to be employed, drive, live independently and have full, productive lives.