MIDWEST NEUROSURGERY & SPINE Adult & Pediatric SPECIALISTS

Spina Bifida

Spina bifida is the most common, permanently disabling birth defect. It affects about one out of every 1,400 to 1,500 newborns in the United States. The Spina Bifida Association conservatively estimates that there are 70,000 people living in the United States with the condition. The prevalence appears to have decreased in recent years due in part to preventative measures followed by expectant mothers prior to and during pregnancy, as well as prenatal testing.

Spina bifida occurs during the third and fourth weeks of pregnancy when a portion of the fetal spinal cord fails to properly close. As a result, the child is born with a part of the spinal cord exposed on the back. Although scientists believe that genetic and environmental factors may act together to cause spina bifida, 95 percent of babies with spina bifida are born to parents with no family history. Women with certain chronic health problems, including diabetes and seizure disorders (treated with certain anticonvulsant medications), have an increased risk (approximately 1/100) of having a baby with spina bifida.

Types of Spina Bifida

There are three types of spina bifida:

Occulta is often called hidden spina bifida, as the spinal cord and the nerves are usually normal and there is no opening on the back. In this form of spina bifida, there is only a small defect or gap in the small bones (vertebrae) that make up the spine. In many cases, spina bifida occulta is so mild that there is no disturbance of spinal function at all. Most people are not aware that they have spina bifida occulta unless it is discovered on an x-ray which they have for an unrelated reason. However, one in 1,000 individuals will have such neurological deficits or disabilities as bowel or bladder dysfunction, back pain, leg weakness, or scoliosis.

Meningocele occurs when the bones do not close around the spinal cord and the meninges are pushed out through the opening, causing a fluid-filled sac to form. The meninges are three layers of membranes covering the spinal cord, consisting of dura mater, arachnoid mater, and pia mater. In most cases, the spinal cord and the nerves themselves are normal or not severely affected. The sac is often covered by skin and may require surgery. This is the rarest type of spina bifida.

Myelomeningocele accounts for about 75 percent of all cases of spina bifida. This is the most severe form of the condition, in which a portion of the spinal cord itself protrudes through the back. In some cases, sacs are covered with skin, but in other cases, tissue and nerves may be exposed. The extent of neurological disabilities is directly related to the location and severity of the spinal cord defect. If the bottom of the spinal cord is involved, there may be only bowel and bladder dysfunction, while the more severe cases can result in total paralysis of the legs with accompanying bowel and bladder dysfunction.

A baby born with spina bifida needs to have the exposed part of the spinal cord repaired to prevent further injury and infection. A neurosurgeon places the neural tissues back in the spinal canal and then closes the muscle and the skin. A plastic surgeon may get involved if there is a large area that is difficult to close. This procedure used to be considered a medical emergency, done a few hours after birth. Surgery is now most often completed within the first 48 hours of the baby's life.

About 80 to 90 percent of children with spina bifida develop hydrocephalus. Hydrocephalus is a condition in which excess cerebrospinal fluid (CSF) builds up within the ventricles (fluid-containing cavities) of the brain and may increase pressure within the head. Most of these children will require a ventricular shunt to control the build-up of spinal fluid. The shunt will remain in place throughout the individual's life, but usually needs to be replaced several times.

Prognosis

If a child is paraplegic (has no movement in the legs from the hips down), he or she will need a wheelchair. If a child is born with movement of the thigh muscles and feeling down to below the knees, the chances are good he or she will be able to walk with some sort of brace support. When there are no brain abnormalities, the child may have average or above average intelligence, even if there is advanced hydrocephalus at birth.

Fortunately, with proper medical care, many children with spina bifida can lead active and productive lives. Twenty year follow-up studies of children with spina bifida show that they enter college in the same proportion as the general population, and many are actively employed. With recent advancements in medical care for these children, their outlook continues to improve.

Tethered Spinal Cord

At birth, the spinal cord is normally located opposite the disc between the first and second lumbar vertebrae in the upper part of the lower back. In a baby with spina bifida, the spinal cord is still attached to the surrounding skin, preventing it from ascending normally, so the spinal cord is low-lying or tethered. Although the skin is separated and closed at birth, the spinal cord stays in the same location after the closure. As the child continues to grow, the spinal cord can become stretched, causing damage and interfering with the blood supply to the spinal cord. This can result in back pain, leg pain, changes in leg strength, progressive or repeated muscle contractions, orthopedic deformities of the legs and scoliosis, and bowel and bladder problems. A definitive diagnosis of a tethered spinal cord is made through diagnostic tests.

The shunt is checked through a computed tomography (CT or CAT scan) or magnetic resonance imaging (MRI). In some cases, the shunt is tapped to assess shunt function. If there is any question about shunt function, the neurosurgeon may explore or revise the shunt, before considering operating on the spinal cord.

If the shunt is working well, usually an MRI of the spine is done to exclude other problems. Additional studies such as a Manual Muscle Test (MMT) and special bladder studies (urodynamics) may be prescribed. These will be compared with prior studies to assess changes and to give a baseline against which to compare after the surgery. Untethering is generally performed only if there are clinical signs or symptoms of deterioration.

The surgery involves opening the scar from the prior closure down to the covering (dura) over the myelomeningocele. Sometimes a small portion of the bony vertebrae (the laminae) are removed to obtain better exposure or to decompress the spinal cord. The dura is then opened, and the spinal cord and myelomeningocele are gently dissected away from the scarred attachments to the surrounding dura. Once the myelomeningocele is freed from all its scarred attachments, the dura and the wound are closed.

The child usually can resume normal activities within a few weeks. Recovery of lost muscle and bladder function depends upon the degree and length of preoperative implications. The combined complication rate of this surgery is usually only 1 to 2 percent. Complications include infection, bleeding, damage to the spinal cord or myelomeningocele, which may result in decreased muscle strength or bladder or bowel function. Many children require only one untethering procedure. However, since symptoms of tethering can occur during periods of growth, 10 to 20 percent require repeated surgery.

Prevention

Women of childbearing age can reduce their risk of having a child with spina bifida by taking 400 micrograms (mcg) of folic acid every day. Because it is water soluble, folic acid does not stay in your body for very long, and needs to be taken every day to be effective against neural defects. Since half of all pregnancies in the United States are unplanned, folic acid must be taken whether a woman is planning a pregnancy or not. Research has shown that if all women of childbearing age took a multivitamin with the B-vitamin folic acid, the risk of neural tube defects could be reduced by up to 70 percent