

UNDERSTANDING SPINA BIFIDA

CARMEN AND JOHN THAIN CENTER FOR PRENATAL PEDIATRICS

What is spina bifida?

Spina bifida is the most common neural tube defect (NTD), a birth defect of the brain and spinal cord. The most severe form of spina bifida, known as *myelomeningocele*, results in a sac-like lesion along the spine where the spinal cord and the nerves are open and exposed; there is no protective skin or backbone covering. The less serious forms of *spina bifida* are *meningocele*, in which the spinal cord itself is not damaged but its protective covering is exposed, and *spina bifida occulta*, in which only the bones of the spine are malformed but the nerves and spinal cord are normal and covered by skin. Throughout this information sheet, the discussion of spina bifida will be largely focused on *myelomeningocele*, the most severe form of the condition.

The damage caused by spina bifida varies, depending on its location along the spinal column and to what extent the spinal nerves are affected. About 80% of spina bifida lesions are on the lower back.

Spina bifida can be surgically repaired after birth, but the nerve damage is permanent, and can result in paralysis, sensory loss, learning disabilities, difficulty with bowel and bladder control, orthopedic problems, and *hydrocephalus*, an accumulation of the fluid that is produced by the brain.

How common is spina bifida and what causes it?

1 in every 2,000 babies born every year in the United States has spina bifida. It is more common in Latina and white women. Diabetic mothers, folate deficiency and those with seizure disorders treated with certain medications also have a higher chance of giving birth to a baby with spina bifida.

Spina bifida results from an error in development very early in pregnancy, around 28 days post-conception. The spinal cord is formed from a sheet of special cells that fold together to form a structure known as the *neural tube*. Spina bifida results when the neural tube does not form correctly and fails to close completely.

How is spina bifida detected during pregnancy?

Many women first learn of their chances to have a baby with spina bifida through a routine blood test given to them between 15 and 22 weeks



Top: There are three forms of spina bifida. Each type varies in severity (from left to right) myelomeningocele, meningocele and spina bifida occulta. **Bottom:** Newborn with spina bifida.

of pregnancy known as the “AFP” test (also known as the “triple” or “quad” screen). It will determine if a woman has an increased risk for a baby with spina bifida. This test is not diagnostic, and many women who have an abnormal result deliver perfectly healthy babies.

Ultimately the best detection of spina bifida is through an ultrasound (sonogram) to check the baby’s anatomy and development, generally performed between 18 and 20 weeks of pregnancy. Ultrasound can detect certain brain features characteristic of spina bifida and determine the location of the open sac along the spinal column. Fetal MRI may be recommended to help determine the severity of the defect.

Amniocentesis can be performed to analyze the AFP in the amniotic fluid; elevated levels are diagnostic of spina bifida. It will also check the baby’s chromosomes. Some studies suggest that babies with spina bifida have an increased chance to have a chromosomal abnormality.

How will my pregnancy be managed now that spina bifida has been detected?

Through the Carmen and John Train Center for Prenatal Pediatrics you will have a thorough ultrasound to examine the anatomy and development of the baby. If you have not had an amniocentesis, you will also be offered that test. You may also have a fetal MRI for a detailed look at the baby’s spine and to determine how serious the spina bifida may be. (continued on next page)

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How will my pregnancy be managed? (continued)

Your prenatal care will be handled by an Obstetrician with special training and expertise in high-risk pregnancy, one of the doctors from Maternal-Fetal Medicine (MFM). Once the diagnosis of spina bifida is confirmed, you will have an opportunity to meet with a variety of pediatric subspecialists who routinely see patients with this diagnosis and work together as a team to best monitor your pregnancy and prepare for the birth of your baby. Your Clinical Care Coordinator will make appointments for you to meet with the following specialists: Genetics, Pediatric Neurosurgery, Pediatric Neurology, Pediatric Urology, Pediatric Orthopedics, and Neonatology. Each one of these specialists will work with you and your MFM to determine the best course of action for your delivery and for the baby's care after birth. As your delivery date approaches, your MFM will discuss with you the delivery plan; in some cases, a planned cesarean section may be the best option. In other cases, elective cesarean section has not been shown to benefit the baby or mother. You and your MFM will decide what is best for you.

What treatment will my baby have after birth?

Surgery to close the spine is performed shortly after birth, generally within the first three days, to best avoid additional nerve damage or infection. The neurosurgeon will place the exposed spinal cord and nerves in the appropriate place and then cover them with muscle and skin.

The accumulation of fluid on the brain (hydrocephalus) is not always obvious before the surgery, over 90% of babies with severe spina bifida will also need a shunt surgically placed to drain the fluid from the brain to the abdomen and keep it from building up in the head. Since paralysis can be a problem for babies with spina bifida, physical therapy begins shortly after surgery to prepare the limbs for possible future walking with braces and special crutches. While many children with spina bifida will need to use a wheelchair to move around (about 50% by age 20), 70% of kids with spina bifida are able to walk with crutches or braces.

What is the long-term outlook for babies with spina bifida?

The degree of handicap and other long-term considerations are largely dependent on where along the spine the nerves were damaged, the size of the lesion, what other types of birth defects are present, and how aggressively the spina bifida is treated. In general, the lower the lesion is along the spine, the better the outlook.

80% of children with spina bifida are able to control their urine output by performing regular self-catheterization (using a tube to drain urine from the bladder). Up to 70% of children with spina bifida have a latex allergy. Around 10% of children with spina bifida develop curvature of the spine (scoliosis) that requires surgical repair. In general, children with spina bifida also have a higher rate of obesity, urinary tract disorders, and learning disabilities. Approximately 80% of children finish high school

and about 80% live at home as adults. Women with spina bifida are able to have children, though their pregnancies are considered high-risk and they have an increased chance of having a baby with spina bifida also.

What are the chances I could have another baby with spina bifida?

Most babies with spina bifida are born in families with no other affected members. Generally it is an isolated condition with no particular inheritance pattern. If you have a baby with spina bifida, the chances for another affected baby are about 1 in 40. If you are thinking of having more children, you should take 4 milligrams of folic acid daily, beginning at least one month before conception and throughout the first trimester of pregnancy. This large dose of folic acid is believed to reduce the chances for another affected pregnancy by 70%.

What can I expect from the specialists at Morgan Stanley Children's Hospital?

NewYork-Presbyterian Morgan Stanley Children's Hospital/Columbia University Medical Center is well-equipped to handle your pregnancy and the baby's care after birth. Morgan Stanley Children's Hospital has consistently been ranked one of the best pediatric hospitals in the country; our pediatric neurosurgeons have extensive experience with spina bifida, and our MFM team is among the largest and most experienced anywhere. Our NICU is one of the most advanced in the United States, and has been cited several times for its excellence and dedication to patient care.

The well-being of you and your baby are extremely important to everyone involved in your care. Together we are all dedicated to giving you the best pregnancy and healthiest outlook for your child.

About the Carmen and John Thain Center for Prenatal Pediatrics

Complex pregnancies receive better care when specialists collaborate. The Carmen and John Thain Center for Prenatal Pediatrics is dedicated to helping pregnant women and their families when a birth defect or genetic syndrome is detected before the baby is born. The Center offers sensitive, complete, up-to-date information and testing, and an integrated approach to care that begins in the prenatal period and continues after birth with pediatric follow-up. A collaborative, coordinated program of care is created among specialists in perinatology, neonatology, genetics, pediatric cardiology, pediatric surgery and all pediatric subspecialties.