Spina Bifida

DEFINITION
Spina bifida is part of a group of birth defects called neural tube defects. The neural tube is the embryonic structure that eventually develops into the baby's brain and spinal cord and the tissues that enclose them.

Normally, the neural tube forms early in the pregnancy and closes by the 28th day after conception. In babies with spina bifida, a portion of the neural tube fails to develop or close properly, causing defects in the spinal cord and in the bones of the backbone.

Spina bifida occurs in various forms of severity. When treatment for spina bifida is necessary, it's done through surgery, although such treatment doesn't always completely resolve the problem.

(Source: Reprinted from the MayoClinic.com article “Spina bifida: Definition” http://www.mayoclinic.com/health/spina-bifida/DS00417)

SYMPTOMS
Spina bifida occurs in three forms, each varying in severity:

Spina bifida occulta
This mildest form results in a small separation or gap in one or more of the bones (vertebrae) of the spine. Because the spinal nerves usually aren't involved, most children with this form of spina bifida have no signs or symptoms and experience no neurological problems. Visible indications of spina bifida occulta can sometimes be seen on the newborn's skin above the spinal defect, including:

- An abnormal tuft of hair
- A collection of fat
- A small dimple or a birthmark
- Skin discoloration

Many people who have spina bifida occulta don't even know it, unless the condition is discovered during an X-ray or other imaging test done for unrelated reasons.

Meningocele
In this rare form, the protective membranes around the spinal cord (meninges) push out through the opening in the vertebrae. Because the spinal cord develops normally, these membranes can be removed by surgery with little or no damage to nerve pathways.

Myelomeningocele
Also known as open spina bifida, myelomeningocele is the most severe form — and the form people usually mean when they use the term "spina bifida."

In myelomeningocele, the baby's spinal canal remains open along several vertebrae in the lower or middle back. Because of this opening, both the membranes and the spinal cord protrude at birth, forming a sac on the baby's back. In some cases, skin covers the sac. Usually, however, tissues and nerves are exposed, making the baby prone to life-threatening infections.

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Neurological impairment is common, including:

- Muscle weakness, sometimes involving paralysis
- Bowel and bladder problems
- Seizures, especially if the child requires a shunt
- Orthopedic problems — such as deformed feet, uneven hips and a curved spine (scoliosis)


CAUSES

Doctors aren't certain what causes spina bifida. As with many other problems, it appears to result from a combination of genetic and environmental risk factors, such as a family history of neural tube defects and folic acid deficiency.


RISK FACTORS

Although doctors and researchers don’t know for sure why spina bifida occurs, they have identified a few risk factors:

- **Race.** Spina bifida is more common among whites and Hispanics.
- **Family history of neural tube defects.** Couples who've had one child with a neural tube defect have a slightly higher chance of having another baby with the same defect. That risk increases if two previous children have been affected by the condition. In addition, a woman who was born with a neural tube defect, or who has a close relative with one, has a greater chance of giving birth to a child with spina bifida. However, most babies with spina bifida are born to parents with no known family history of the condition.
- **Folate deficiency.** Folate (vitamin B-9) is important to the healthy development of a baby. Folate is the natural form of vitamin B-9. The synthetic form, found in supplements and fortified foods, is called folic acid. A folate deficiency increases the risk of spina bifida and other neural tube defects.
- **Some medications.** Anti-seizure medications, such as valproic acid (Depakene, Stavzor), seem to cause neural tube defects when taken during pregnancy, perhaps because they interfere with the body's ability to use folate and folic acid.
- **Diabetes.** Women with diabetes who don't control their blood sugar well have a higher risk of having a baby with spina bifida.
- **Obesity.** Pre-pregnancy obesity is associated with an increased risk of neural tube birth defects, including spina bifida.
- **Increased body temperature.** Some evidence suggests that increased body temperature (hyperthermia) in the early weeks of pregnancy may increase the risk of spina bifida. Elevating your core body temperature due to fever or the use of saunas or hot tubs, has been associated with increased risk of spina bifida.
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If you have known risk factors for spina bifida, talk with your doctor to determine if you need a larger dose or prescription dose of folic acid, even before a pregnancy begins. If you take medications, tell your doctor. Some medications can be adjusted to diminish the potential risk of spina bifida, if plans are made ahead of time.


COMPLICATIONS

Spina bifida may cause no symptoms or only minor physical disabilities. Frequently, it leads to severe physical and mental disabilities.

Factors that affect severity

The severity of the condition is affected by:

- The size and location of the neural tube defect
- Whether skin covers the affected area
- Which spinal nerves come out of the affected area of the spinal cord

Range of complications

Complications may include:

- Physical and neurological problems. This may include lack of normal bowel and bladder control and partial or complete paralysis of the legs. Children and adults with this form of spina bifida might need crutches, braces or wheelchairs to help them get around, depending on the size of the opening in the spine and the care received after birth.
- Accumulation of fluid in the brain (hydrocephalus). Babies born with myelomeningocele also commonly experience accumulation of fluid in the brain, a condition known as hydrocephalus. Most babies with myelomeningocele will need a ventricular shunt — a surgically placed tube that allows fluid in the brain to drain as needed into the abdomen. This tube might be placed just after birth, during the surgery to close the sac on the lower back, or later as fluid accumulates.
- Infection in the tissues surrounding the brain (meningitis). Some babies with myelomeningocele may develop meningitis, an infection in the tissues surrounding the brain, which may cause brain injury and can be life-threatening.
- Other complications. Additional problems may arise as children with spina bifida get older. Children with myelomeningocele may develop learning disabilities, including difficulty paying attention, problems with language and reading comprehension, and trouble learning math. Children with spina bifida may also experience latex allergies, skin problems, urinary tract infections, gastrointestinal disorders and depression.