

# SPINA BIFIDA

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## SPINA BIFIDA

(also called myelodysplasia or neural tube defect)

### Condition Description

Spina bifida occurs in the first month of fetal life as the result of a failure of part of the spine to fully form due to an unknown cause.

Maternal nutritional state, exposure to radiation, drugs and chemicals as well as a genetic predisposition may act together to cause neural tube defects. The embryonic cells that form the neural tube and eventually form the spine do not fully develop. The spinal cord therefore lacks a bony covering due to defects in the vertebra bone formation. The nervous system tissue of the spinal cord can also be affected. This can occur anywhere along the length of the spinal column. Those that occur in the spine are categorized under the term "spina bifida", and 80% of these are located in the lumbar and sacral areas of the spine. The three forms of spina bifida range from very minor to severe:

1. Spina Bifida Occulta (**SPI**-nuh . **BIF**-id-uh . oh-**KULT**-uh): This is a very common occurrence that typically causes no problems. Probably 20-30% of North Americans have undetected spina bifida occulta because the under-developed gap in the vertebrae is very small and the spinal cord is normal. A small hairy patch, or a soft skin colored mass called a lipoma, or a small dimple in the skin may overlie the affected bony malformation and be the only external sign of spina bifida occulta.
2. Meningocele (muh-**NING**-go-**SEEL**): Unlike spina bifida occulta, a spinal meningocele rarely occurs (4% of neural tube defects). The defect in the vertebra is very small and fluid-filled meninges protrude

through the opening in the bone. The spinal cord and nerves are intact.

3. Myelomeningocele or meningocele: This is the most common type of spina bifida and it is typically severe in nature. Some of the spinal cord's neural tissue is exposed through a bony defect in one or more vertebrae. There is an absence of muscle and skin that would normally protect the spinal cord, and nerve damage is permanent, resulting in varying symptoms.

### Prevalence

Spina bifida is the most common of the severe birth defects, occurring in one of every 1500-2000 live births. The most severe variant, myelomeningocele occurs in one of every 4000 live births. There is a definite genetic predisposition in that it is more common in families of Hispanic and European heritage. However, 95% of infants with spina bifida are born to families with no family history of the disorder. There is a risk of recurrence of 1 in 400 in subsequent pregnancy and a considerably higher risk for women with diabetes or taking seizure medications (1:100).

Folic acid deficiency has been implicated in causing neural tube defects. If mothers took 4 micrograms daily of Vitamin B folic acid at least 1 month before and during pregnancy, 50-70% of neural tube defects could be prevented, even among mothers who have previously had a child with a neural tube defect. From 1996 to 2001 there was a 24% decline in spina bifida, probably due to the introduction of folic acid for women of child-bearing age.

### Common Associated Conditions

Symptoms vary depending on the location of the myelomeningocele. The overall mortality



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rate is 10-15% before the age of 4 years. Paralysis of the lower limbs is common. Neurogenic bladder and fecal incontinence are frequent complications because the nerves to the bladder and bowel do not function as they should. Hydrocephalus (excessive cerebral spinal fluid) occurs because the circulation of this protective fluid is blocked.

### Short-term Treatment and Outcomes

Significant spina bifida can be detected prenatally by an elevated level of alpha-fetoprotein in the amniotic fluid and by the unusual appearance of the spine on fetal ultrasound. Because the spina bifida occulta form is typically very benign, no treatment will be offered unless and until neurologic symptoms occur. There is no cure for the myelodysplasia type of spinal bifida because the nerve tissue cannot be replaced or repaired. However, surgery can assist these children in many ways.

An experimental prenatal surgery is being performed in a few medical centers to see if intrauterine surgery would be beneficial for the baby and reduce some of the severe complications. However this new technique brings new risks for both the mother and the baby and its benefits have not yet been clearly demonstrated.

Infants with meningoceles or myelomeningoceles should be delivered by caesarian section. X-rays and scans will be done right after birth to determine the extent of the defects. Within 24 hours the baby will have surgery to enclose the nerves and spinal cord within the canal to the extent possible and to cover the delicate spinal cord and nerves with muscle and skin to prevent infection and trauma. If the opening is large, a silicone patch might be used to cover defect until skin grafting can be done. Close observation for hydrocephalus and infection is essential. The baby usually requires a ventriculo-peritoneal

shunt to drain cerebral spinal fluid into the abdomen, thus preventing pressure from building up in the brain that could cause brain damage (seizures, blindness).

### Long-term Treatment and Outcomes

Spina bifida occulta does not pose problems in childhood. However, later in life there may be some neurological deterioration that needs treatment because fatty tissue can expand into the spinal column and cause pressure on nerves with resulting symptoms of weakness, numbness or loss of function. Not all children have paralysis of their lower limbs but if the defect is in the mid-sacral area they will typically have flaccid legs and lack reflexes and sensation.

Children with spina bifida will require ongoing assessment of neurologic function by specialists in a variety of areas. Orthopedic care will be required long-term, and they will need physical therapy and perhaps braces for their lower limbs to permit mobility and to prevent contractions. Other children will use wheelchairs for mobility or crutches for balance.

Bowel training and a high-fiber diet are essential to prevent constipation and stool impaction. Bladder expression or self-catheterization of urine is necessary to prevent kidney infection and damage. Children must have life-long coordinated interdisciplinary care from neurology, physiatry, urology and orthopedics. The child will need to be referred to appropriate adult providers in late adolescence. Later in childhood or adulthood there can be traction on the spinal cord causing neurologic symptoms (called a tethered cord), requiring surgical correction.

### Common Complications

Infection is an immediate concern and meningitis can spread very easily to the brain

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from the opening in the spine, and also through the bloodstream causing sepsis (blood-stream infection) in the newborn period. Most children (70-90%) with meningomyelocele will have hydrocephalus requiring drainage into the abdomen through a shunt. Seizure disorders are slightly more common in children with spina bifida compared to unaffected children. Eye disorders such as strabismus and amblyopia are also common and require ongoing assessment and care.

Many complications can occur such as mobility problems, bowel and bladder complications, latex allergy due to exposure to medical equipment containing this substance, tendonitis, obesity, skin breakdown, gastrointestinal disorders, learning disabilities, depression, social isolation and sexual issues.

### Implications for Children's Development

The outlook for children with spina bifida has greatly improved due to new surgeries and specialized teams that know how to care for these children and support their families. Most children (90%) live well into adulthood and are in school with children their own age; 80% are of normal intelligence, and 75% take part in recreational and sport activities. They will require frequent psychological and social evaluations to be sure that they are coping with their condition and receiving maximum help to live full and independent lives.

Mobility is a high priority in order to ensure independence and normal developmental progression. Children can become independent in managing their own bowel and bladder habits by bowel training and self-catheterization, and this should be done at an early age to normalize a child's life as much as possible. Children can self-catheterize to empty their bladder of urine in the nurse's office at school and have bowel training to empty their bowel daily before going to school. Parents, school and health professional staff all

must work together to be sure that systems and support are in place to allow children with spina bifida to be independent, healthy and well-educated. Within the limits of safety they can participate in activities with their non-disabled peers and be responsible for their own health needs. Educational evaluation with careful consideration of class placement and special services is essential, and it is very important to remember that there is a huge range of disability among children with spina bifida, from very mild to complex.

The research on outcomes long-term for children with spina bifida indicates:

- Typically normal intelligence, often with higher verbal than math skills
- Perceptual-motor difficulties that affect eye-hand coordination for handwriting and other fine motor activities
- Learning disabilities for some children, perhaps related to memory, comprehension, attention, impulsivity, sequencing, organization and reasoning.

Each child's strengths and areas of difficulty will differ and change as years go by, making the individual education plan (IEP) extremely important. Because of the many systems involved, children and parents may need encouragement in independent living and self-care; over-dependence on parents and others can limit child's self-esteem, long-term options and motivation. Therefore a strength-based approach from earliest infancy is essential for the child and family. Transition planning to encourage independent decision-making within safe limits must begin in the childhood years, including finding adult health care providers when pediatric services are no longer appropriate.

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