Spina Bifida

What is spina bifida?

Spina bifida means cleft spine, which is an incomplete closure in the spinal column. A closed defect is characterized by spinal tissue that is covered by skin. An open defect is characterized by spinal tissue that is not covered by skin. The 4 types of spina bifida are

- **Spina bifida occulta:** One or more of the vertebrae (bones) of the spinal column has an opening without damage to the spinal cord.
- **Occult spinal dysraphism (OSD):** The child has a minor abnormality of the skin overlying the lower spine, such as a hairy patch, a pigmented area, or a small opening (sinus). The spinal cord below this abnormality is at high risk for injury as the child grows.
- **Meningocele**
  - Meninges, the protective covering around the spinal cord, have pushed out through the opening in the vertebrae in a sac called the *meningocele.* However, the spinal cord remains intact.
  - This form of spina bifida can be repaired with little or no damage to the nerve pathways.
- **Myelomeningocele:** This form of spina bifida is the most severe: a portion of the spinal cord itself protrudes through the back.

How common is it?

- The most severe forms occur at approximately 0.3 out of every 1,000 births.
- Of these newborns, the majority (94%) has myelomeningocele, and the rest (6%) have meningocele and OSD.

What are some common characteristics of children who have spina bifida or of spina bifida as children present with it?

- **Muscle weakness:** Children with myelomeningocele usually have muscle weakness or paralysis below the area of the spine where the incomplete closure (or cleft) occurs. Children may need wheelchairs or may be able to use crutches or walkers.
- **Sensation disturbances:** Children may not feel sensation in their limbs or body parts below the cleft.
- **Bowel and bladder problems:** Children with spina bifida usually do not develop normal bowel and bladder control, because this control depends on sacral nerve-root function. Many children with myelomeningocele need special assistance to learn to manage their bowel and bladder functions. Most require catheterization, specifically the insertion of a tube to permit the passage of urine. Many require medications to try to optimize bladder and bowel functions.
- **Latex allergy:** All children with spina bifida should avoid latex because of risk of allergy.
- **Hydrocephalus:** In addition, this condition may cause an accumulation of fluid in the brain, or hydrocephalus. (See Hydrocephalus and Shunts Quick Reference Sheet [page 151] for more details.)
  - It is estimated that 70% to 90% of children born with myelomeningocele have hydrocephalus.
  - The higher the abnormality is on the spine, the greater the risk for hydrocephalus.
  - Hydrocephalus is controlled by a surgical procedure called *shunting,* which relieves the fluid buildup in the brain.
  - Before shunting was available, most children born with a myelomeningocele died shortly after birth. Now that surgery to drain spinal fluid and protect children against hydrocephalus can be performed early in life, children with myelomeningocele are more likely to live.
  - Often, however, they must undergo a series of operations, including shunt revisions, throughout their childhoods.
- **Vision problem:** Children with spina bifida may have problems with their eyes or vision.
- Approximately 10% to 15% of children with spina bifida also have seizure disorders.

Who might be on the treatment team?

- In addition to the primary health care professional who ensures that the child receives routine preventive care services, many pediatric specialists are involved in the care of children with spina bifida. Pediatric developmental and rehabilitation specialists, neurosurgeons and orthopedic surgeons, neurologists, gastroenterologists, and urologists often work together in a *multispecialty center* to address and coordinate the medical needs of these children.
- Many children with spina bifida benefit from *physical therapy, occupational therapy,* or *speech-language therapy* to learn adaptive skills and how to function with their peers.
- Children who are younger than 3 years (ie, 36 months) may receive these therapies through *early intervention* services. Early intervention is a system of services to support infants and toddlers with disabilities and their families.
- For children 3 years and older, most have learning disabilities that require support. *Special education and related services* are available through the public school to provide the therapies and educational supports necessary for school achievement.

Spina Bifida (*continued*)

What are some elements of a Care Plan for children with spina bifida?

- Care Plans for children with spina bifida often include *intermittent catheterization,* a procedure during which a tube is placed into the bladder and urine is drained, usually at least 3 times per day. Care Plans also need to provide support for fecal incontinence, which is as common as urinary incontinence in children with spina bifida.
- The Care Plan may also incorporate physical or occupational therapy exercises into a daily routine.
- These plans may include mobility aids such as wheelchairs, walkers, splints, braces, communication devices, and adapted toys to help children be more active, participate more, and have fun while they are working their bodies.
- Exposure to latex (eg, rubber-containing toys, bandages) should always be limited to prevent development of latex allergy and managed in those who have already developed sensitivity to it.
- A written plan called the *Individualized Family Service Plan* will be provided for children in early intervention programs.
- An *Individualized Education Program* will describe an older child's unique needs and the services available to address them.
- These children should have at least one visit with a qualified pediatric ophthalmologist (an eye specialist).

What adaptations may be needed?

Physical Environment and Other Considerations

- Many children learn self–intermittent catheterization at an early age, sometimes as young as 5 years. A private area in which this insertion can be done is helpful.
- A successful bladder and bowel management program can be incorporated into the school day or child care program.
- Architectural factors need to be considered when caring for a child with spina bifida. Ramps, ground-floor entrances, and wheelchair-accessible areas may be needed. In addition, schools should help ensure that handicapped-accessible playgrounds and activities are available, so the social interaction between children with spina bifida and typically developing children can occur.
- Children with spina bifida have varying physical capabilities and limitations. Work with the child's family and therapists on goals to improve the child's mobility without increasing child or family frustration with the child's limitations.

What should be considered an emergency?

- If the child has a seizure, follow emergency guidelines for seizures (see Seizures, Febrile, Quick Reference Sheet [page 177]).
- Notify parents/guardians of
  - Fever
  - Severe headache, lethargy, irritability, or new eye crossing
  - Numbness or tingling in the limbs
  - Any loss of function (eg, weakness in the legs)
  - Inability to obtain urine with catheterization
  - Any sign of allergic reaction, especially if latex exposure is suspected
- Children with spina bifida may need extra time, supervision, or transport in case of an emergency such as a fire.
- Any critical adaptive equipment and supplies would also need to be brought in an evacuation.

What types of training or policies are advised?

- Catheterization
- Care Plan specifics
- Safe transfer or transportation
- Children with spina bifida are more likely to miss school because of medical visits and medical emergencies or because of care issues. School providers should work to ensure that children and adolescents with spina bifida are provided appropriate supports to address these necessary absences.

What are some resources?

Spina Bifida Association:
www.spinabifidaassociation.org, 1-800-621-3141

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