Spina Bifida:

Your Guide to a Healthy Life

Neurosurgery

For ages 0-18+

Adapted from Guidelines for the Care of People with Spina Bifida, 2018

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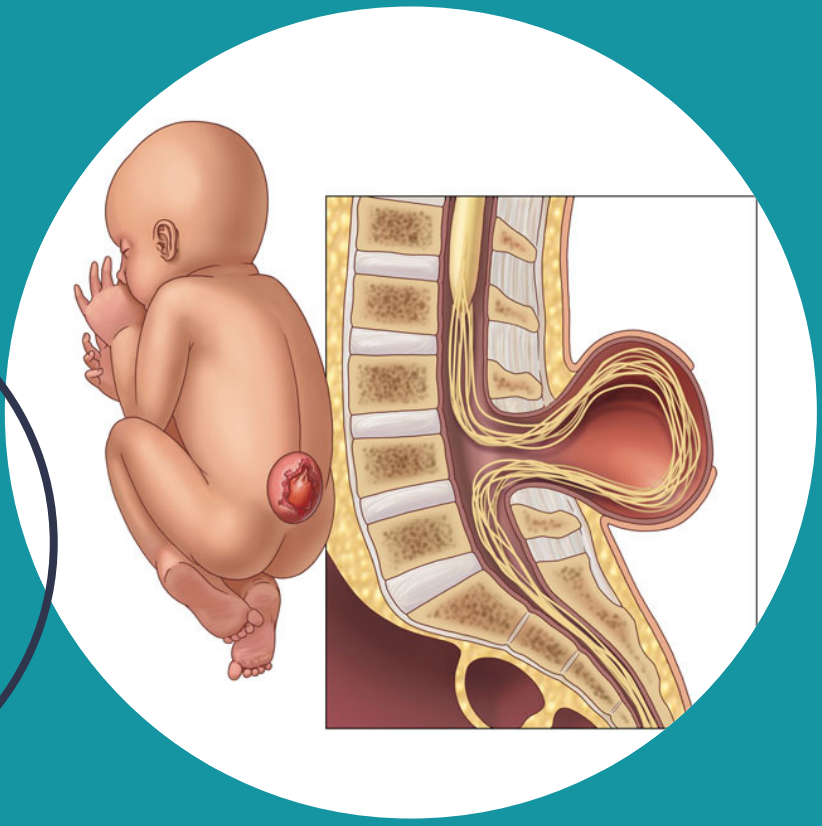
SPINA BIFIDA ASSOCIATION

Introduction



Myelomeningocele is the most common form of Spina Bifida and the most serious birth defect of the human nervous system that is compatible with long term survival. It occurs very early in a pregnancy and results in a variety of neurological problems. With myelomeningocele, the spinal cord is open and exposed, resulting in loss of function in the lower part of the body. In addition to myelomeningocele, there are other types of Spina Bifida. With these types, known collectively as occult spinal dysraphism, the spinal cord is covered by skin and the effects are less severe. Typically, the term “Spina Bifida” is used to refer to open myelomeningocele.

Fetal Surgery

Open myelomeningocele is typically closed by a neurosurgeon within 48 hours of birth. However, a prenatal surgery to correct open myelomeningocele in the fetus is an increasingly available option. Beginning in 2003, a randomized trial of this fetal surgery, known as the Management of Myelomeningocele (MOMs), began. This study showed that fetal surgery resulted in some improved outcomes in patients with Spina Bifida when compared to patients who underwent postnatal closure surgery. Improved outcomes include:[1]

- **Reduced need for ventricular shunts *(82% in postnatal surgery vs. 40% in prenatal surgery).***
- **A reduction in occurrence of the Arnold-Chiari II malformation, which can lead to problems with breathing and swallowing.**
- **Better function in lower extremities.**
- **Improved brain development.**

However, the MOMs study found that the surgery also led to increased complications for the mother, a higher incidence of premature delivery, and increased risk for complications in later pregnancies. Physicians have been working to improve techniques to reduce these complications, which is important as more sites have begun offering fetal surgery for myelomeningocele. Another important point is that the MOMs study had strict participation requirements, so it is not known if the results would be as positive for patients that don't meet those requirements.

Challenges with prenatal surgery include the following:

- It is a highly costly procedure and there is still limited availability for patients that may benefit from it.
- Long-term studies are not yet available to determine whether the positive outcomes for children who undergo fetal surgery are long lasting (or are not offset by other problems later on). Some studies show that improvements in hydrocephalus, Arnold Chiari II malformation, motor function, and learning remain. There may be a higher incidence in tethered cord among infants who have undergone fetal surgery, and neurologic loss from tethered cord has the potential to reduce the improvements in motor function and bladder control noted in the MOMs study patients. Fetal surgery did not result in a decreased need for catheterization in the most recent follow up from the MOMs study.
- Premature births following the surgery have been reduced but not eliminated, and the surgery makes future pregnancies riskier.

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[1] Adzick NS, Fetal myelomeningocele: natural history, pathophysiology and in-utero intervention. Semin Fetal Med 2010; 15: 9-14.

Prenatal Counseling

Neurosurgeons are an important part of the prenatal counseling team for families with a diagnosis of Spina Bifida. Neurosurgeons with experience caring for patients with Spina Bifida are uniquely qualified to discuss with families their realistic long-term expectations and challenges. *(Prenatal Counseling Guidelines)*. How a baby with myelomeningocele is delivered (vaginal delivery vs. cesarean, also known as a c-section) is a controversial issue, but there is no significant evidence favoring one route of delivery over another. Mothers who undergo fetal surgery must deliver via c-section.

Hydrocephalus

Following the post-natal closure of the lesion on the back, or following the birth of a baby who had fetal surgery, neurosurgeons determine whether the baby also needs to be treated for associated hydrocephalus. This often involves following the infant to observe for signs of hydrocephalus such as rapid head growth, leakage from the back wound or the development of a breathing abnormality called stridor. Ventricular shunts are the most common treatment, but there is currently research into 1) when to place a shunt, and 2) the use of an alternate treatment called endoscopic third ventriculostomy with choroid plexus coagulation (*ETV/CPC – see below*).

Around 80% of patients with myelomeningocele require treatment of hydrocephalus with a shunt, but the frequency of problems with shunts has led some neurosurgeons to allow ventricles to get bigger before placing a shunt. By allowing ventricles to grow larger, several experienced centers have reduced shunt rates to 55-65%. The long-term effect on the cognitive abilities of these patients is unknown, but they are spared repeated shunt operations and infections. The best available early data suggest that children with bigger ventricles are doing well. More time will be required to understand this more clearly.

ETV/CPC [1] is a promising alternative to shunts for treating hydrocephalus. Trials in the United States and internationally showed success in 70-75 percent of cases of children with hydrocephalus from Spina Bifida. Use of this procedure has expanded rapidly and research is ongoing to assess results.

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[2] ETV is a minimally-invasive procedure that creates an opening in the floor of the third ventricle in the brain. This allows cerebrospinal fluid (CSF) trapped within the brain's ventricles to escape into its normal pathway. CPC is a procedure that reduces the choroid plexus (tissue that produces CSF) in two of the four ventricles inside the brain. This decreases the amount of fluid produced and may also reduce the strength of pulses that can cause the ventricles to enlarge. This makes the ETV procedure more likely to succeed in a child's brain. (Boston Children's Hospital, http://www.childrenshospital.org/conditions-and-treatments/treatments/etv-cpc-procedure)

Arnold Chiari II Malformation

By definition, every child with myelomeningocele has a Chiari II Malformation, where the rear part of the brain (the cerebellum and posterior fossa) elongate and extend into the spinal canal. Effects can range widely, from no symptoms to problems with breathing and swallowing. Traditionally, decompression surgery has been done to address these symptoms, but the rate of these surgeries has declined recently due to:

- **Awareness that decompression is not always effective in alleviating Chiari II symptoms.**
- **The fact that symptomatic Chiari II is frequently caused by hydrocephalus and shunt failure.**
- **The recognition that some children's Chiari II malformations will not respond to decompression surgery.**

Tethered Spinal Cord

Tethered spinal cord is another important neurosurgical issue in Spina Bifida. This occurs when the spinal cord is scarred to tissue around the spine, most commonly at the base of the spine. As a result, the spinal cord can't move freely within the spinal canal. This can cause the cord to stretch as the spine grows, leading to neurologic decline, pain, and other symptoms. Ongoing research efforts have focused on understanding when to intervene and on improving surgical procedures to reduce re-tethering. This problem may require more attention as children undergoing fetal surgery have the potential for increased risk of a tethered cord.

Guidelines for Neurosurgical Care

0-11 Months

Prior to birth

- Meet with a neurosurgeon and others who are part of the prenatal counseling team after receiving a diagnosis of Spina Bifida, to discuss the impact of Spina Bifida on your child and to learn about newborn care. The neurosurgeon can provide more information about outcomes you may expect for your child and explain the need for long-term multidisciplinary care. Your team of healthcare providers will review options for continuation versus termination of pregnancy, fetal surgery, and provide information on newborn care management. *(Prenatal Counseling Guidelines)*

- If desired, learn more from the neurosurgeon about surgical centers that can evaluate whether your child is a candidate for fetal surgery. The neurosurgeon can provide information on what outcomes you can expect with fetal surgery.

- Consult with a multi-disciplinary team prior to birth to establish a joint plan for delivery and care. This could involve coordination of care with local and regional medical centers for delivery, immediate care, or to transfer to centers with the specialists necessary to provide the best possible early care.

Delivery and after

- If you've undergone fetal surgery, your baby will be delivered via c-section. For infants that will have closure of the back after birth, many physicians prefer cesarean delivery despite the lack of evidence that it is the better approach.

- Your baby's back closure site will be covered with clean, moist dressings, with the aim of having surgery to close the lesion within 48 hours of birth.

- Watch for symptoms of hydrocephalus that could lead to the need for a shunt or ETV/CPC, including:
  - accelerating head growth and/or splitting of the sutures on the infant's skull
  - bulging soft spot (fontanelle)
  - increasing irritability
  - loss of appetite and/or vomiting
  - eyes that look downward or "sun setting eyes"
  - change in behavior
  - high-pitched breathing (stridor); muscle spasms causing arching of the back (opisthotonus); silent cry; inability to swallow saliva; slow or temporary stopping of breathing (hypopnea/apnea); and leaking of fluid from the back wound

- If your child has Chiari II symptoms, surgery may be necessary. Typically, this surgery is only performed if symptoms persist after a shunt is placed or ETV/CPC is performed and is working properly.

- You are encouraged to develop a relationship with a multidisciplinary Spina Bifida clinic, where your infant should be followed at three- to four-month intervals.

1-2 Years

1. Be aware of the signs of acute shunt failure (headache, vomiting, and lethargy/sleepiness) and chronic shunt failure (accelerated head growth or failure to meet developmental milestones such as using words or understanding commands). Your neurosurgeon can instruct you on what to do if you suspect a shunt failure.

2. Keep an eye out for Chiari II symptoms that might occur in this age range (inability to swallow saliva, swallowing dysfunction, stridor, and difficulty acquiring language).

3. Be aware of symptoms of tethered cord (back pain and declining lower extremity sensation and function).

4. Your child will likely be followed at a multidisciplinary Spina Bifida clinic at six-month intervals. Neurosurgeons may use studies such as MRIs, CT scans, and sleep and swallow studies as needed to make decisions about your child's care.

3-5 Years

1. Be aware of the signs of acute shunt failure (headache, vomiting, and lethargy/sleepiness) and chronic shunt failure (low grade recurring headache and neck pain, loss of developmental milestones) in your child. Your neurosurgeon can instruct you on what to do if you suspect a shunt failure.

2. Keep an eye out for Chiari II symptoms that might occur in this age range (inability to swallow saliva, swallowing dysfunction, stridor, and difficulty acquiring language).

3. Be aware of symptoms of tethered cord (back pain, declining lower extremity sensation and function, and urologic dysfunction).

4. Keep an eye out for signs of syringomyelia (syrinx or cyst in the spinal cord) such as back pain and sensory changes in hands.

5. Your child will likely be followed by a multidisciplinary Spina Bifida clinic at six- to 12-month intervals. Neurosurgeons may use studies such as MRIs, CT scans, and sleep and swallow studies if needed to make decisions about your child's care.

6-12 Years

1. You are encouraged to establish working relationships with your child's educational system including teachers and other educational professionals to ensure your child's success in school.

2. Work with the Spina Bifida clinic coordinator and/or social worker to identify any changes in cognitive function in your child and communicate them to the medical team so that needed resources can be identified. *(Neuropsychology Guidelines)*

3. Be aware of the signs of acute shunt failure (headache, neck pain, vomiting, and lethargy/sleepiness) and chronic shunt failure (recurring low-grade recurring headache and neck pain; loss of developmental milestones; cognitive, behavioral, or neurological decline; and orthopedic or urological regression) in your child. Your neurosurgeon can instruct you on what to do if you suspect a shunt failure.

4. Keep an eye out for Chiari II symptoms that might occur in this age range (inability to swallow saliva, swallowing dysfunction, stridor, and difficulty acquiring language).

5. Be aware of symptoms of tethered cord (back pain, declining lower extremity sensation and function, bladder or bowel control decline, and worsening orthopedic problems and/or scoliosis).

6. Keep an eye out for signs of syringomyelia (syrinx or cyst in the spinal cord) such as neck or back pain and sensory changes in arms and hands.

7. Your child will likely be followed by a multidisciplinary Spina Bifida clinic at 12-month intervals. Neurosurgeons may use studies such as MRIs, CT scans, and sleep and swallow studies if needed to make decisions about your child's care.

13-15 Years

1. Be aware of the signs of acute shunt failure (headache, neck pain, vomiting, and lethargy/sleepiness) and chronic shunt failure (recurring low-grade headache and neck pain, behavioral and/or cognitive changes, neurological decline, urological changes, and increasing orthopedic deformities and/or progressive scoliosis) in your child. Your neurosurgeon can instruct you on what to do if you suspect a shunt failure.

2. Keep an eye out for Chiari II symptoms that might occur in this age range (inability to swallow saliva, swallowing dysfunction, stridor, and declining language ability).

3. Be aware of symptoms of tethered cord (back pain, declining lower extremity sensation and function, urological changes, and worsening orthopedic problems and/or scoliosis).

4. Keep an eye out for signs of syringomyelia (syrinx or cyst in the spinal cord) such as neck or back pain and sensory changes in arms and hands.

5. Your child will likely be followed by a multidisciplinary Spina Bifida clinic at 12-month intervals. Neurosurgeons may use studies such as MRIs, CT scans, and sleep and swallow studies if needed to make decisions about your child's care.

6. Your child's neurosurgery team should assist you in learning about the transition to adult care and in identifying an adult neurosurgery provider. *(Transition Guidelines)*

18+ Years

1. Review information about transitioning to adult care, including:
- Knowledge and autonomy for personal health decisions.
- Awareness of own body symptoms/signs.
- Knowledge about predictors of good quality of life in adulthood. *(Transition, Self-Management and Independence Guidelines)*

2. Your neurosurgeon should help identify an adult neurosurgery provider and facilitate and support completion of transitional care. (*Transition Guidelines)*

3. Be aware of the signs of acute shunt failure (headache, neck pain, vomiting, and lethargy/sleepiness) and chronic shunt failure (recurring low-grade recurring headache and neck pain, and behavioral and/or cognitive changes). Your neurosurgeon can instruct you on what to do if you suspect a shunt failure.

4. Keep an eye out of Chiari II symptoms that might occur in adults (inability to swallow saliva, swallowing dysfunction, stridor, and declining language ability).

5. Be aware of symptoms of tethered cord (back pain, declining lower extremity sensation and function, and urological dysfunction).

6. Keep an eye out for signs of syringomyelia (syrinx or cyst in the spinal cord) such as neck or back pain and sensory changes in arms and hands.

7. You will likely be followed by a multidisciplinary Spina Bifida clinic at 12-month intervals. Neurosurgeons may use studies such as MRIs, CT scans, and sleep and swallow studies if needed to make decisions about your care.