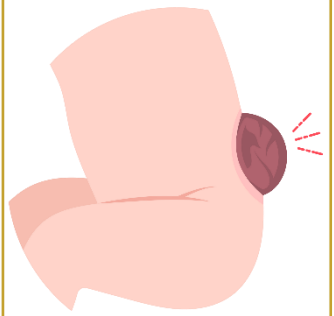
جامعة الإمام عبدالرحمن بن فيصل
IMAM ABDULRAHMAN BIN FAISAL UNIVERSITY

مستشفى الملك فهد الجامعي
King Fahad Hospital The University

Spina Bifida



What is Spina Bifida?

Spina bifida is a congenital deformity that affects the spinal cord and the spinal column of the fetus. This deformity occurs due to the failure of the lower part of the neural tube to close during fetal development. This results in the formation of an opening in the spinal column, exposing the spinal cord to contamination and pressure, leading to serious health complications.

What are the causes and risk factors of spina bifida?

The neural tube forms during early stages of pregnancy and closes after approximately four weeks from the beginning of pregnancy. Spina bifida occurs when there is a defect in the formation of a part of the neural tube or when it fails to close properly, resulting in abnormalities in the spinal cord and spinal bones.

What are the causes and risk factors of spina bifida?

However, the exact cause of this deviation is still unknown. Like many other health problems, spina bifida may be the result of genetic and environmental factors:

- Previous history of pregnancy affected by spina bifida.
- Use of certain antiepileptic medications such as valproic acid and carbamazepine.
- Pre-existing diabetes before pregnancy, not gestational diabetes.
- Insufficient intake of folic acid.
- Obesity before pregnancy.
- Maternal exposure to high temperatures during early stages of pregnancy

What are the spina bifida types?

1 Occult Spina Bifida (Closed Spina Bifida):

It is the mildest and most common type of spina bifida. It results in a separation or non-fusion of the spinal vertebrae and usually does not affect the nerves or spinal cord. It may be discovered incidentally during imaging tests for unrelated reasons and often does not cause disabilities. However, signs can be observed on the skin of newborns above the spinal defect, such as:

1. The presence of a small abnormal area on the baby's back with hair.
2. Small dimple.

What are the spina bifida types?

2 Meningocele:

It is a rare type of spina bifida where there is a sac containing cerebrospinal fluid protruding from an opening in the spinal column. The nerves and spinal cord within the sac are not affected, and it may cause mild difficulties in bladder and bowel functions.

3 Myelomeningocele (Open Spina Bifida):

It is the most severe type of spina bifida.

It is characterized by a gap in the spinal column that exposes the spinal canal and leads to the formation of a sac on the baby's back containing the spinal cord and nerves.

What are the spina bifida types?

3 Myelomeningocele (Open Spina Bifida):

This sac may come into direct contact with the external environment. It can cause moderate to severe disabilities, such as loss of sensation in the legs or feet.

Signs of Myelomeningocele:

1. The spinal canal is open along the length of the spinal vertebrae.
2. A sac containing the spinal cord or nerves protrudes.

What are the spina bifida complications?

1. Walking and mobility problems: as there may be weakness in leg muscles and loss of movement, depending on the location of the defect in the spinal column and the size of the neural opening.
2. Bone problems: such as scoliosis, foot deformities, hip dislocation, bone diseases, and joint issues.
3. Intestinal and bladder problems: as there may be a lack of normal control over the bowels and bladder.
4. Accumulation of fluid in the brain: also known as hydrocephalus.

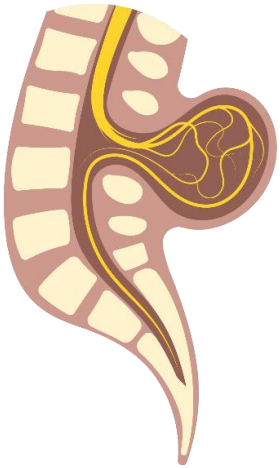
What are the spina bifida complications?

5. Shunt malfunction: which is the disruption or malfunctioning of the shunt placed in the brain to treat hydrocephalus or due to infection.

6. Some warning signs of shunt malfunction include headaches, vomiting, drowsiness, irritability, and swelling or redness along the shunt.

7. Gastrointestinal complications, including feeding difficulties, gastroesophageal reflux, and constipation.

8. Neurological complications, such as paralysis, muscle weakness, and developmental delays.



How to diagnose spina bifida?

Spina bifida can be diagnosed through various prenatal and postnatal tests, including:

- Maternal serum alpha-fetoprotein (MSAFP) screening: This blood test measures the levels of a protein called alpha-fetoprotein, which is produced by the fetus and can indicate the presence of neural tube defects.
- Ultrasound: This imaging technique allows visualization of the fetus and can detect structural abnormalities, including spina bifida.
- Amniocentesis: A procedure in which a small amount of amniotic fluid is extracted and tested for genetic and chromosomal abnormalities.

How to treat spina bifida?

- There is no direct cure for spina bifida, but treatment focuses on symptom relief and prevention of complications.
- Treatment may involve surgery to close the opening in the spinal column and repair neural deformities.

How to treat spina bifida complications?

Potential complications of spina bifida are treated on an individual basis according to their type and severity. This may include surgical treatment, rehabilitation therapy, medical support, and neurological treatment.

How to prevent spina bifida?

- Taking folic acid supplements before and during the early months of pregnancy is considered an important preventive measure against spina bifida.
- Maintaining a healthy weight before pregnancy and avoiding obesity.
- Controlling diabetes before pregnancy.
- Avoiding exposure to high temperatures during pregnancy.

What is Neurosurgery role in spina bifida?

First: Neurological assessment:

a. Motor Function: Spina bifida patients are evaluated for motor function. The neurological examination assesses muscular strength, tone, and coordination in the lower extremities. Spina bifida patients may have weakness, spasticity (increased muscular tone), or trouble with balance and walking as a result of nerve damage to the lower extremities.

What is Neurosurgery role in spina bifida?

First: Neurological assessment

b. Sensory function: is a crucial component of neurological assessments. The evaluation evaluates the capacity to feel touch, pain, temperature, and position awareness (proprioception) in the afflicted portions of the body, especially below the level of the spinal lesion. Each dermatome's pain perception, temperature using tubes, Tuning fork vibration, proprioception, light touch from a cotton wool wisp, Stereognosis and two-point discrimination.

c. Reflexes: such as the knee-jerk reflex (patellar reflex) and ankle reflexes. The existence, absence, or abnormalities of reflexes can reveal information about the spinal cord and nerve pathways' integrity.

What is Neurosurgery role in spina bifida?

Second: Examination

Spine: Inspect the spine for any abnormalities, noting the location and size of any lesions or deformities.

Neurological Examination:

- Conduct a thorough neurological assessment to establish a baseline.
- Measurements: Measure the infant's head circumference.
- Reflexes: Assess the baby's cry, sucking reflex, anal sphincter function, and urinary stream.
- Motor Function: Evaluate muscle bulk, spontaneous movements, muscle tone, and responses to stimulation.
- Sensory Function: Conduct a full sensory examination.
- Check for any foot or hip deformities.

What is Neurosurgery role in spina bifida?

Third: Complications

The obstruction of cerebrospinal fluid leads to hydrocephalus, which is the abnormal accumulation of fluid surrounding the brain. This condition is commonly found in individuals with myelomeningocele but is not typically associated with other forms of spina bifida. The fluid buildup can exert harmful pressure on the brain.

Hydrocephalus is often treated by surgically placing a plastic shunt, a hollow tube, into the brain to redirect the excess fluid to the abdomen, where it can be absorbed by the body. Another treatment method is an endoscopic third ventriculostomy (ETV), which establishes a new pathway for fluid drainage.

What is Neurosurgery role in spina bifida?

Third: Complications

Meningitis, an infection of the membranes surrounding the brain, can sometimes occur in patients with ventriculoperitoneal (VP) shunts. This infection may result in brain damage and can be life-threatening.

Meningitis can be caused by various bacteria, viruses, fungi, and parasites, with many infections being transmissible from person to person. A small number of cases arise from injuries, cancers, or medications.

Bacterial meningitis is the most serious type and can become fatal within 24 hours. While effective treatments and vaccines are available for some of the leading bacterial causes of meningitis, it remains a significant global health threat.

What is Neurosurgery role in spina bifida?

Third: Complications

The four primary causes of acute bacterial meningitis are:

1. Neisseria meningitidis (meningococcus)
2. Streptococcus pneumoniae (pneumococcus)
3. Hemophilus influenzae
4. Streptococcus agalactiae (group B streptococcus)

Fourth: Clinical Features

The clinical picture of meningitis can differ based on the underlying cause, the progression of the disease (acute, subacute, or chronic), the involvement of the brain (such as in meningoencephalitis), and any systemic complications like sepsis. Common symptoms include fever, confusion, neck stiffness, or changed in mental status, vomiting, nausea, and headache.

What is Neurosurgery role in spina bifida?

Fourth: Clinical Features

Less common symptoms may include seizures, coma, and neurological deficits, which can manifest as hearing or vision loss, cognitive difficulties, or weakness in the limbs.

Fifth: Epilepsy and Seizures

Some people may experience seizures or epilepsy primarily as a result of spina bifida, hydrocephalus, or brain trauma. Tethering of the spinal cord can cause tension on the brain, which may lead to seizures once the tension is released. There are various factors that can contribute to the development of epilepsy or seizures in individuals with spina bifida. Drugs are available to help manage these seizures.

What is Neurosurgery role in spina bifida?

Sixth: Clinical Features

Babies born with spina bifida require surgical intervention to repair the exposed part of the spinal cord, preventing further injury and infection. Neurosurgeons carefully place the neural tissues back into the spinal canal and close the muscle and skin. In complex cases, plastic surgeons may assist in closure. This surgery is typically performed within the first 48 hours of the baby's life

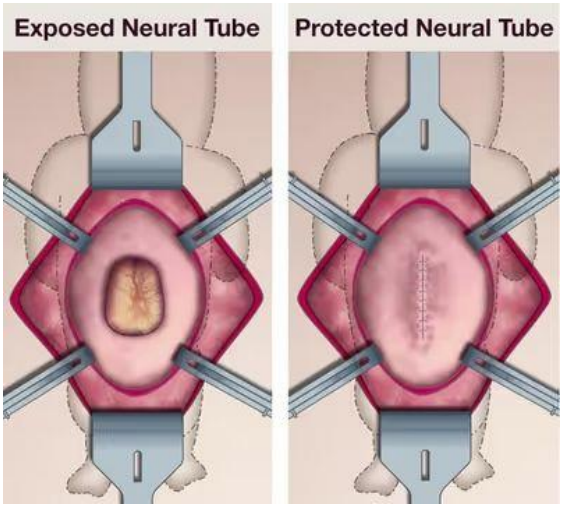


Image1: Neural tube defect before and after surgery

What is Neurosurgery role in spina bifida?

Seventh: Hydrocephalus management

Around 80 to 90% of children with spina bifida develop hydrocephalus, characterized by excess cerebrospinal fluid build-up in the brain and its cavities. These individuals often require a ventricular shunt to control fluid accumulation. Shunt replacements may be necessary throughout their lives.

Eighth: Prenatal repair of myelomeningocele (MMC):

It is a specialized surgical procedure performed while the fetus is in the womb. It involves opening the uterus and closing the opening in the baby's back to prevent further spinal cord damage.

What is Neurosurgery role in spina bifida?

Eighth: Prenatal repair of myelomeningocele (MMC):

Although not curative, prenatal repair has shown improved outcomes compared to postnatal repair, including reduced need for brain fluid diversion, enhanced mobility, and increased chances of independent walking. Follow-up care involves regular assessments, including baseline imaging, detailed neurological and muscular examinations, and prompt identification of treatable causes of deterioration.

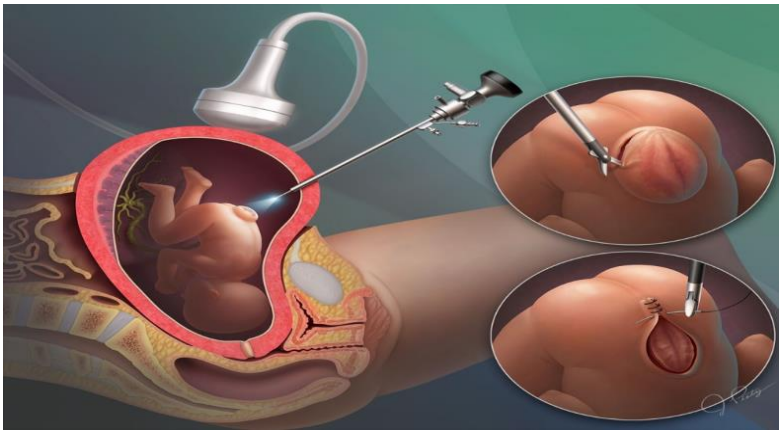


Image 2: Ultrasound guided Prenatal repair of (MMC)

Notes:

References and resources:

Saudi Ministry of Health

World Health Organization

American Association of Neurological Surgeons

Texas Children's Hospital, USA

Children's Hospital of Philadelphia, USA

University of California San Francisco, USA

Johns Hopkins Hospital

Illustrations from Canva

Review and audit

The content of this booklet has been reviewed by neurosurgery consultants at King Fahad Hospital of the University.

Neurosurgery Department

Health Awareness Unit

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