ABSTRACT:
Spina bifida comes from the Latin word “divided spine”, is a group of neural tube defects that involves the brain and the spinal cord and/or meninges. The first recorded case was in 1085 AD in Arabia. It occurs when the neural tube does not close during the baby’s development. There are three major types of spina bifida. Spina bifida occulta, meningocele, and myelomeningocele. Meningocele is a rare form of neural tube defect in which the spinal cord develops normal but the meninges protrude from a spinal opening. This group of congenital anomalies of the CNS results from failure of the neural tube to close spontaneously. Normally the closure of the neural tube occurs around 28 days after fertilization. Myelodysplasia refers to any malformation of the spinal canal and cord.

KEY WORDS: Spina Bifida, occulta, meningocele, myelomeningocele.

INTRODUCTION
Spina bifida is a birth defect that happens when a baby’s backbone (spine) does not form normally. As a result, the spinal cord and the nerves that branch out of it may be damaged (Potts 2000 et al.). The term spina bifida comes from Latin and literally means "split" or "open" spine. This defect happens at the end of the first month of pregnancy, when a baby’s spine and spinal cord (a bundle of nerves that runs down the center of the spine) are developing. Depending on the severity of the defect and where it is on the spine, symptoms vary. Mild defects may cause few or no problems, while more severe defects can cause serious problems, including weakness, loss of bladder control, or paralysis. Children with an exposed opening on the back will need surgery to close it (Lowdermilk 2004 et al.).

CASE STUDY OF MASTER X
Master X, 4 years old male, presented with history of increasing head circumference and diagnosed as Meningocele T4 – T6 with Syringomyelia T4 – T9, Obstructive Hydrocephalus Secondary to Chiari II Malformation. During the pregnancy our patient’s mother always complies on the pre-natal check up, she had her immunizations such as tetanus toxoid. She never took any medications that are harmful to her pregnancy and eats foods that are good to her and to the baby. At the first month of her pregnancy she was noted to have frequent emesis gravid arum and UTI. Sometimes she was also expose to stress due to her work, which is a "labandera", and the lack of taking supplementary vitamins. After giving birth to our patient they noticed that there is a mass growing at the upper back. They seek medical attention and they were advised to have a surgical intervention but due to financial problems they refused and went home so that they could save some money for the operation. Patient X had completed his immunizations. He has no known allergy to foods and drugs and has only caught minor diseases such as colds, fever, and cough. At the age of 2 years old he had a convulsion; he was rushed to the hospital and was treated. But at the age of 3 years old, they noticed a slight change in patient X’s behavior. They noticed that he has a short temper and often cries or having a temper tantrums; they also started to notice that his right eye and right area of his jaw cannot move, tolerable headaches and a slight increase in the head circumference. History of Present Illness Three months before admission, patient X was having his check-up because of cough and colds. As days pass, patient X was complaining of headaches, pain at the back, and they noticed that his head is larger than any other child his age. They went to Hospital to seek for medical intervention and they were advised to admit their patient for VP shunting and he was diagnosed
Spina Bifida with Non–Communicating Hydrocephalus (emedicine.medscape.com).

Spina bifida is a birth defect that involves the failure of the osseous (bony) spine to close. The term spina bifida comes from Latin and literally means "split" or "open" spine. It has been reported that in 1000 live births 1-2 babies have this kind of condition worldwide (Pilliteri and Adele 2003).

### Table 1. ETIOLOGY OF SPINA BIFIDA

<table>
<thead>
<tr>
<th>Book Picture</th>
<th>Patient Picture</th>
</tr>
</thead>
<tbody>
<tr>
<td>Genes but in most cases there is no familial connection. (Asnwal 1999 et al.)</td>
<td></td>
</tr>
<tr>
<td>A high fever during pregnancy may increase a woman’s chances of having a baby with spina bifida.</td>
<td>UTI</td>
</tr>
<tr>
<td>Folic acid deficiency</td>
<td>lack of taking supplementary vitamins</td>
</tr>
<tr>
<td>Radiation during pregnancy</td>
<td></td>
</tr>
<tr>
<td>Maternal malnutrition</td>
<td></td>
</tr>
<tr>
<td>Women with epilepsy who have taken the drug valproic acid to control seizures may have an increased risk of having a baby with spina bifida</td>
<td></td>
</tr>
</tbody>
</table>

### TYPES OF SPINA BIFIDA

The two forms of spina bifida are spina bifida occulta and spina bifida cystic (Potter 2004 et al.).

**Spina bifida occulta** is the mildest form of spina bifida (occulta means hidden). It occurs most frequently in the lumbar-sacral area (L5 and S1). Spina Bifida Occulta: A bony defect in the vertebral column that causes a cleft that remains covered by skin. Treatment is usually not required and spinal cord is left unaffected. -Mildest form. It may not be apparent unless there are associated cutaneous manifestations or neuromuscular disturbances.

**Spina bifida cystic** refers to a visible defect with an external sac like protrusion. It includes two types of spina bifida (Luxner 2005):

1. Meningocele involves the meninges and spinal fluid but no neural elements. If the meninges push through the hole in the vertebrae (the small, ring-like bones that make up the spinal column), the sac is called a meningocele.

2. Myelomeningocele (meningomyelocele) is the most severe form of spina bifida which contains meninges, spinal fluid, and nerves. Most babies who are born with this type of spina bifida also have hydrocephalus, an accumulation of fluid in and around the brain. The condition may be asymptomatic or if the defect is large, severe neurological abnormalities may result.

### PATHOPHYSIOLOGY

The malformation that causes neural tube defects occurs during embryogenesis, typically no later than 26 days after fertilization. Two different processes lead to formation of the CNS. The first is primary neurulation, which refers to the invagination of the neural plate into the neural tube, and subsequently the embryonic brain and spinal cord. Secondary neurulation refers to the formation of the lower spinal cord, which gives rise to the lumbar and sacral elements. Any disruption that occurs when the neural plate begins its first fold and fuses to form the neural tube (days 17-23) can cause craniorachischisis, the most severe form of neural tube defect. Closure of the rostral neuropore occurs between days 23 and 26. Disruption during this phase of embryogenesis results in anencephaly. Myelomeningocele results when the closure of the caudal neuropore is disrupted during days 26 to 30 (Wong 2005 et al.). (Table 2)
Spina Bifida- A Case Study

Handly, J.

Table 2. CLINICAL FEATURE

<table>
<thead>
<tr>
<th>BOOK PICTURE</th>
<th>PATIENT PICTURE</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Spina bifida cystica</strong></td>
<td>Kyphosis</td>
</tr>
<tr>
<td>Sensory disturbances usually parallel motor dysfunction below second lumbar vertebra:</td>
<td>Head is slightly larger than normal with 52 cm in diameter. With Ventriculo-Peritoneal Shunt at right side of the occipital area, with pinkish scar noted at the left side of the occipital area.</td>
</tr>
<tr>
<td>• Flaccid, partial paralysis of lower extremities</td>
<td></td>
</tr>
<tr>
<td>• Varying degrees of sensory deficit</td>
<td></td>
</tr>
<tr>
<td>• Overflow incontinence with constant dribbling of urine.</td>
<td></td>
</tr>
<tr>
<td>• Lack of bowel control</td>
<td></td>
</tr>
<tr>
<td>• Rectal prolapse (sometimes)</td>
<td></td>
</tr>
<tr>
<td>• Below third lumbar vertebra:</td>
<td></td>
</tr>
<tr>
<td>• No motor impairment</td>
<td></td>
</tr>
<tr>
<td>• May be saddle anesthesia with bladder and anal sphincter paralysis.</td>
<td></td>
</tr>
<tr>
<td><strong>Joint deformities (sometimes produced in utero):</strong></td>
<td></td>
</tr>
<tr>
<td>• Talipes valgus or vagus contractures</td>
<td></td>
</tr>
<tr>
<td>• Kyphosis</td>
<td></td>
</tr>
<tr>
<td>• Lumbosacral scoliosis</td>
<td></td>
</tr>
<tr>
<td>• Hip dislocations</td>
<td></td>
</tr>
<tr>
<td><strong>Spina bifida occulta</strong></td>
<td></td>
</tr>
<tr>
<td>• Frequently no observable manifestations</td>
<td></td>
</tr>
<tr>
<td>• May be associated with one or more cutaneous manifestations:</td>
<td></td>
</tr>
<tr>
<td>• Skin depression or dimple</td>
<td></td>
</tr>
<tr>
<td>• Port-wine angiomatous nevi</td>
<td></td>
</tr>
<tr>
<td>• Dark tufts of hair</td>
<td></td>
</tr>
<tr>
<td>• Soft, subcutaneous lipomas</td>
<td></td>
</tr>
<tr>
<td>• May be neuromuscular disturbances:</td>
<td></td>
</tr>
<tr>
<td>• Progressive disturbance of gait with foot weakness</td>
<td></td>
</tr>
<tr>
<td>• Bowel and bladder sphincter disturbances (Luxner 2005).</td>
<td></td>
</tr>
</tbody>
</table>

Table 3. DIAGNOSTIC EVALUATION

<table>
<thead>
<tr>
<th>BOOK PICTURE</th>
<th>PATIENT PICTURE</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Prenatal tests</strong></td>
<td>Hemoglobin 12.6 g/L</td>
</tr>
<tr>
<td>The alpha-fetoprotein (AFP) test, performed between the 16th and 18th weeks of pregnancy. If the amount is high, the test is repeated because in many cases, high AFP readings are false. If the second result is high, other tests will be done to double-check and confirm the diagnosis.</td>
<td>White Blood Cells 7.4 10^6 g/L</td>
</tr>
<tr>
<td></td>
<td>Neutrophils .31</td>
</tr>
<tr>
<td></td>
<td>Lymphocytes .60</td>
</tr>
<tr>
<td></td>
<td>Eosinophils .09</td>
</tr>
<tr>
<td></td>
<td>Hematocrit .35 0.40</td>
</tr>
<tr>
<td></td>
<td>Protrombine time 14.0</td>
</tr>
<tr>
<td></td>
<td>APTT 34.5 secs</td>
</tr>
<tr>
<td><strong>MRI, ultrasound, CT, and myelography</strong></td>
<td>Cranium CT Scan - Contiguous axial - Non communicating images of the brain</td>
</tr>
<tr>
<td><strong>Amniocentesis</strong></td>
<td></td>
</tr>
</tbody>
</table>

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hydrocephalus were obtained. There is a moderate degree of dilation of both lateral and 3rd ventricles. The 4th ventricle is normal in size. The gray-white matter interface is maintained. There is no evidence of acute intracerebral hemorrhage. There is no midline shift. The cortical sulci, cisterns, sella and CP angles are normal for patients stated age. The visualized paranasal sinuses and mastoid air cells are pneumatised. The visualized cranium is intact.

CSF ANALYSIS Color - Colorless Transparency - Cloudy.

Colorless Cloudy CSF indicates an infection or an increase in the WBC.

Differential Count: Lymphocytes 92 60 – 70 % Increased. Indicates infection

CULTURE AND SENSITIVITY: No growth after 72 hours of incubating.

TREATMENT OF SPINA BIFIDA

BOOK PICTURE

Children with spina bifida occulta seldom need treatment.

In cases of spina bifida cystica, treatment depends on the type of spina bifida and its severity.

Babies with meningocele usually have an operation during infancy in which doctors push the meninges back and close the hole in the vertebrae. Many will have no other health problems later unless there is nerve tissue involved with the sac.

Babies with myelomeningocele need more immediate attention and often have surgery within the first 1 to 2 days after birth. During this first surgery, doctors push the spine back into the vertebrae and close the hole to prevent infection and protect the spine. Management of these children with receives support from a medical team that may include several doctors (such as neurosurgeons, urologists, orthopedic surgeons, rehabilitation specialists, and general pediatricians), a nurse practitioner, physical and occupational therapists, and a social worker.

A baby who also has hydrocephalus will need an operation to place a shunt in the brain. The shunt is a thin tube that helps to relieve pressure on the brain by draining and diverting extra fluid. In addition, some children need subsequent surgeries to manage problems with their feet, hips, or spine.

The location of the gap in the back often dictates what kind of adaptive aids or equipment a child with myelomeningocele will need. Those with a gap high on the spinal column and more extensive paralysis often need to use a wheelchair, while those with a gap lower on the back may be able to use crutches, leg braces, or walkers (www.enurse-careplan.com).

Treatment of renal problems includes:

• Prompt and vigorous treatment of infections
• Some type of regular emptying of the bladder, such as clean intermittent catheterization (CIC)
• Medications to improve bladder storage and continence, such as oxybutynin chloride
• Surgical procedures such as vesicotomy (stoma created on the abdominal wall for urinary drainage)
• And augmentation enterocystoplasty (increases bladder capacity and reduces high bladder pressures (Spina bifida 2017).

*Fiber supplements, laxative suppositories, or enemas aid in producing regular evacuation.

MANAGEMENT OF MASTER X

V.P shunting was done in patient
COMPLICATIONS

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.

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) (Bannink F 2016 et al.).

Other Complications: 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 (Spina bifida 2017).

Master X developed non-communicating hydrocephalus. (Table 4)

<table>
<thead>
<tr>
<th>Needs and Concerns</th>
<th>Interventions</th>
</tr>
</thead>
</table>
| **1. Defective closure of vertebral column** | • Keep infant in prone position, hips slightly flexed, and legs abducted, with pad between knees.  
• Turn prone-to-side or side-to-side every 2-4 hour.  
• Place infant on sheepskin to reduce pressure.  
• Follow physicians orders for feeding position.  
• Keep dressings over the sac moistened with antibiotic solution or ointment, as ordered.  
• Protect sac from contamination by urine and feces.  
• Observe, record, and report any leaks from the sac, irritation of sac, signs of infection (redness, odor, fever, lethargy, poor color, vomiting, full fontanels, uncial rigidity). |
| **2. Development of increased intracranial pressure** | • Check vital signs, including blood pressure and pupil reaction, every 4 hour.  
• Measure and record head circumference daily.  
• Observe, report, and record signs of increased intracranial pressure (early-lethargy, irritability, poor feeding, vomiting; late: change in pupil size and equality, widening of pulse pressure, decreasing pulse, irregular or decreasing respiratory rate, seizures, bulging fontanels). |
| **3. Urinary tract infection** | • Keep perineal area clean.  
• Express urine from bladder every 3 hour (Credemaneuver)  
• Observe for signs of infection (foul-smelling urine, excoriation of perineum, fever, lethargy)  
• Provide good fluid intake. |
| **4. Constipation** | • Record number of stools per day; note consistency  
• Glycerin suppository as needed (emedicine.medscape.com). |

**POSTOPERATIVE**

A. Physiologic

1. Repair of defect | • Place infant in prone position  
• Keep the incision line clean and dry.  
• Keep infant on abdomen or side until incision is healed.  
• Check vital signs, including blood pressure, every 4 hour.  
• Protect the incision from urine and fecal contamination.  
• Observe for signs of infection and increased intracranial pressure.  
• Record, describe, and report drainage from incision site. |
Hold infant for feedings as soon as possible.
Resume preoperative care as appropriate.

B. BEHAVIOURAL

2. Possible delayed developmental milestones.
   - When feeding or bathing provide tactile, visual, auditory stimulation.
   - Attach toys and brightly colored objects at eye level.
   - Move objects in front of infant’s eyes (vertically first, then horizontally).
   - Place toys within infant’s reach.
   - Provide parents with age-appropriate infant-stimulation programs.

C. FAMILIAL

3. Parental anxiety, guilt feelings
   - Provide opportunities for parents to talk and ask questions.
   - Reassure parents that nothing they did caused the defect.
   - Focus on parent’s concerns about infant.

4. Care by parent
   - Explain what you are doing and why. Demonstrate care.
   - Ask parent to assist while you provide care.
   - Have parent provide care while you assist.
   - Support interdependent care by parent.
   - Teach home management skills—skin care, care of incision or care of myelomeningocele, bladder and bowel care, signs of increased intracranial pressure, nutrition, safety precautions, developmental stimulation.

D. ENVIRONMENTAL/COMMUNITY

Community services
   - Inform about Crippled Children Services and Social Security benefits.
   - Inform about local parents’ groups.
   - Refer to infant-stimulation program if available.
   - Make public-health nurse referral if needed.

SUMMARY

Master X was cooperative with health personnel. Although his symptoms were well responding to treatment, it was recurring. But he did not further complications during the hospital stay.

CONCLUSION

Prevention of disease is of fundamental importance. When prevention of disease is not possible prevention of further complication is a priority. Children with Spina bifida need encouragement to participate in activities with their peers and to lead independent lives, within their physical limitations and capabilities (www.enurse-careplan.com). It may be helpful to remember that these children have never known what’s accepted as normal function and often adapt to their condition in remarkable ways. If child has spina bifida, parents may benefit from finding a support group of other parents who are dealing with the condition. Talking with others who understand the challenges and rewards of living with spina bifida can be helpful.

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www.cureresearch.com

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