What Does That Mean:
My Baby has Spina Bifida?

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Abstract: Spina bifida is an incomplete closure of the neural tube anywhere along the spine and affects over 1,600 newborns each year (Centers for Disease Control and Prevention, 2018a). There is evidence that a low level of folic acid increases the risk of spina bifida. The presentation depends on the type, level of the spine, and spinal nerves involved. Early and aggressive treatment is associated with the most favorable outcomes. The role of the childbirth educator revolves around prevention and education.

Keywords: spinal bifida, birth defects, myelomeningoceles, meningoceles

Pregnancy is a time of joy, anticipation, and imagining what one's newborn will look like. It is when parents start pondering over baby names, shop for cute baby clothes, and begin to decorate the nursery for their new arrival. Most parents are focused on having a healthy baby and not thinking of the possibility of birth defects. In the United States, babies born with costly and critical birth defects occur every 4-1/2 minutes (Centers for Disease Control and Prevention [CDC], 2018b). That translates into approximately 1 in 33 or 120,000 babies per year being born with birth defects in the United States (CDC, 2018b). Spina bifida, otherwise referred to as a neural tube defect (NTD), is a birth defect that can occur. NTDs affect over 1,600 newborns each year, are more prevalent in Hispanic and Caucasian female babies, and result in more than $1 billion dollars in hospitalization costs annually (CDC, 2018a). This paper will discuss the incidence, prevalence, types, treatment, complications, and nursing implications surrounding spina bifida for the childbirth educator.

Background

Spina bifida comes from the Latin words "split spine" which refers to an incomplete closure of the neural tube anywhere along the spine (Benaroch, 2017; Burns et al., 2017; Piatt, 2016). The severity depends on the size, location, and whether or not a portion of the spinal cord and nerves are exposed (CDC, 2018d). About 75% of NTDs are located in the lumbosacral area (Burns et al., 2017). Spina bifida cases fall into two categories; spina bifida aperta (open) and spina bifida occulta (hidden). Spina bifida aperta encompasses myelomeningoceles and meningoceles. The protrusion of the spinal cord nerve roots and membrane layers that cover the spinal cord and brain are referred to as a myelomeningocele (Burns et al., 2017). About 10-20% of myelomeningocele cases contain only the meninges and the other 80-90% of cases involves both the meninges and nerve roots (Burns et al., 2017). This is the most serious type of spina bifida because a sac is protruding through the infant's back containing damaged spinal cord portions and nerves (CDC, 2018d). A meningocele involves a sac protruding from the spine but does not contain the spinal cord in the sac, which results in less nerve damage than a myelomeningocele (CDC, 2018d). Myelomeningoceles and meningoceles can also protrude through the skull filled with cerebral spinal fluid causing hydrocephalus that may or may not cause neurologic damage (Burns et al., 2017). Spina bifida occulta is the mildest type of NTD and results from the vertebral arches failing to close (Burns et al., 2017; CDC, 2018d). This type of NTD may go unnoticed or not be diagnosed until childhood or adulthood since it does not typically cause any disabilities (CDC, 2018d).

During the third and fourth weeks of gestation (about 28 days) the neural tubes will close (Burns et al., 2017). If the neural tubes fail to close, it results in a form of spina bifida. This can be diagnosed while pregnant or after delivery (CDC, 2018d). During pregnancy the diagnosis can be made from an elevated alpha-fetoprotein (AFP) blood level around 16 to 18 weeks gestation, visualization of the defect on ultrasound at 18 to 20 weeks gestation, or with a sample of the

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amniotic fluid revealing high levels of AFP (CDC, 2018d). Testing has a high accuracy of 85-90% (Copp et al., 2015).

About 1 in 2,000 U.S. births results in an NTD, which is thought to be linked to genetics and environmental factors (Burns et al., 2017). There is sufficient evidence demonstrating that low levels of folic acid (vitamin B9) increases the risk of spina bifida (Phillips, Burton, & Evans, 2017; Williams et al., 2015). Therefore, in 1992 the U.S. Public Health Services issued a recommendation for all women who are capable of childbearing to consume 400 micrograms of folic acid daily to help prevent NTDs (CDC, 2018a; Williams et al., 2015). It is estimated that the folic acid fortification has prevented over 1,300 NTDs (Williams et al., 2015). In 2004, spina bifida cases began to level off despite the use of folic acid (Burns et al., 2017). Theories surrounding the leveling include decreased serum folate levels secondary to increasing obesity rates, low carbohydrate diets, increase in consumption of whole-grain breads, and maternal diabetes (Burns et al., 2017). Other risk factors associated with spina bifida include a high fever during pregnancy and taking valproic acid or Carbamazepine while pregnant for seizure control (Phillips et al., 2017; Piatt, 2016).

Clinical Manifestations

The presentation depends on the type, level of the spine, and which spinal nerves are involved (Benaroch, 2017). Meningoceles present with a visible fluid-filled sac that may or may not be covered with a thin layer of skin that protrudes through the spine or skull (National Institute of Neurological Disorders and Stroke, 2018). Myelomeningoceles present like meningoceles but typically the sac involved is not covered with a thin layer of skin which exposes the spinal cord tissue and nerves (National Institute of Neurological Disorders and Stroke, 2018). Clinical presentation may include weakness, absent deep tendon reflexes and pain sensation, loss of bladder or bowel control, enlarged head with bulging fontanel, abnormal shaped feet, uneven hips, scoliosis, seizures, trouble breathing, difficulty swallowing, and paralysis (Benaroch, 2017; Burns et al., 2017; Piatt, 2016). More severe symptoms tend to occur when the opening is higher on the spine (Piatt, 2016). Spina bifida occulta presents with either no findings or an abnormal tuft of hair or small dimple (birthmark) at the site of the spinal malformation (National Institute of Neurological Disorders and Stroke, 2018). Many individuals with spina bifida occulta find out by accident when they obtain an x-ray for other reasons (Benaroch, 2017).

Treatment

Early and aggressive treatment is associated with the most favorable outcomes for meningoceles and myelomeningoceles. The goal is surgical closure of the NTD within 24 to 48 hours after birth to prevent compromise to the spinal cord and nerves (Phillips et al., 2017). This involves placing the membrane sac, tissue, nerves, and spinal cord back into the baby's body and closing the opening (Benaroch, 2017; Burns et al, 2017). If surgery is not performed within one week of birth these infants may die by one year of age from meningitis or sepsis (Burns et al., 2017). Otherwise, a favorable prognosis is 85-90% if treated early (Burns et al., 2017). In some cases, surgery may even be performed intrauterine before 26 weeks gestation; however, this can be perilous and may cause preterm labor (Benaroch, 2017; Phillips et al., 2017). Occasionally these infants may need a ventriculostomy (hole in third ventricle of brain) or a shunt (hollow tube that connects to stomach internally) placed in the brain to drain fluid, which should also be done at 24 to 48 hours after birth (Benaroch, 2017; Piatt, 2016).

Complications

Spina bifida requires a multifaceted approach with comprehensive care from the pediatric provider, orthopedics, urology, neurology, dermatology, ophthalmology, and habilitation (Phillips et al., 2017). As many as 20-50% of patients with myelomeningocele will experience progressive tethering or tethered cord syndrome (NIH, 2018). This is when the spinal cord adheres to the immovable membranes and vertebrae causing abnormal spinal cord stretching during growth (NIH, 2018). Symptoms include back pain, leg pain, hypertonia, spasms, hyperreflexia, decreased sensation, weakness, gait deterioration, constipation, worsened scoliosis, and loss of bladder function (Phillips et al., 2017). A large number of infants will have a lesion between S2 and S4 that presents with normal kidney function at birth that eventually can lead to vesicoureteral reflux, hydronephrosis, recurrent urinary tract infections, and kidney damage or failure (Phillips et al., 2017). Patients with hydrocephalus typically require additional surgeries to replace the shunt as they grow or to unclog or remove infection (NIH, 2018). They may also experience learning delays in school requiring services (Piatt, 2016). Moreover, since the brain is abnormally positioned this can cause a Chiari II malformation that can interfere with breathing, vocal cord function, swallowing, and upper extremity strength (Phillips et al., 2017). Surgeries may be needed to correct feet, hips, or the spine. If the malformation is high on the spine a patient may be wheelchair bound from paralysis. If the malformation is lower on the spine then the patient may need crutches, leg braces, or walkers. Bowel

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and bladder training should start after birth with measures such as catheterization (NIH, 2018). Babies with spina bifida are also prone to skin sores, blisters, burns, and calluses on their feet, ankles, and hips secondary to decreased sensation below the level of the spinal malformation (CDC, 2018c). However, with the proper medical care and self-care, most infants and children can lead a normal active life.

Nursing Education

The role of the childbirth educator revolves around prevention and education. First, anyone who is of childbearing age should be encouraged to take the recommended daily allowance of folic acid to help reduce the risk of NTDs. Additionally, foods high in folic acid such as eggs, citrus fruits, beets, legumes, asparagus, fortified breads, rice, and pasta, and dark green leafy vegetables should be consumed routinely before and during pregnancy. Any fever experienced during pregnancy should immediately be treated with acetaminophen. Sitting in hot tubs and saunas should be avoided as they can cause hyperthermia and increase the risk for NTDs. Counseling overweight or obese women to gain control over their weight before attempting to become pregnant may help reduce the risk of NTDs. The same advice should be given to patients with diabetes who want to become pregnant. Women with spina bifida, who have a child with spina bifida, or who have had a pregnancy complicated by an NTD, are five to ten times more likely to have another child with spina bifida and need to be encouraged to take 4,000 micrograms of folic acid at least one month prior to getting pregnant through the first three months of pregnancy (Benaroch, 2017).

Once spina bifida has been diagnosed, the role of the childbirth educator shifts to a more supportive one. The news can be overwhelming, and parents need to know where to turn for answers. Referrals to specialists, support groups, community resources, reliable websites, and spina bifida organizations are just a few ways childbirth educators can assist parents.

Conclusion

Spina bifida is a birth defect that can cause serious complications to the newborn and throughout life; therefore, education on prevention is crucial for all childbirth educators. Relaying best practices to all childbearing women needs to be enforced to help reduce the occurrence of spina bifida and the multitude of problems that can be associated with the diagnosis.

References

Benaroch, R. (2017). What is spina bifida? Retrieved from https://www.webmd.com/parenting/baby/spina-bifida#1

Burns, C. E., Dunn, A. M., Brady, M. A., Starr, N. B., Blosser, C. G., & Garzon, D. L. (2017). *Pediatric primary care* (6th ed.). St. Louis, MO: Elsevier.

Centers for Disease Control and Prevention. (2018a). Data & statistics on spina bifida. Retrieved from https://www.cdc.gov/ncbddd/spinabifida/data.html

Centers for Disease Control and Prevention. (2018b). Facts about birth defects. Retrieved from https://www.cdc.gov/ncbddd/birthdefects/facts.html

Centers for Disease Control and Prevention. (2018c). Living with spina bifida: Infants. Retrieved from https://www.cdc.gov/ncbddd/spinabifida/infant.html

Centers for Disease Control and Prevention. (2018d). What is spina bifida? Retrieved from https://www.cdc.gov/ncbddd/spinabifida/facts.html

Copp, A. J., Adzick, N. S., Chitty, L. S., Fletcher, J. M., Grayson, N., Holmbeck, G. N., & Shaw, G. M. (2015). Spina bifida. *Nature Reviews Disease Primers, 1*(15007). doi:10.1038/nrdp.20158.7

National Institute of Neurological Disorders and Stroke. (2018). Spina bifida fact sheet. Retrieved from https://www.ninds.nih.gov/Disorders/Patient-Caregiver-Education/Fact-Sheets/Spina-Bifida-Fact-Sheet

Phillips, L. A., Burton, J. M., & Evans, S. H. (2017). Spina bifida management. *Current Problems in Pediatric and Adolescent Health Care, 47*(7), 173-177. doi:10.1016/j.cppeds.2017.06.007

Piatt, J. H. (2016). Spina bifida. Retrieved from https://kidshealth.org/en/parents/spina-bifida.html

Williams, J., Mai, C. T., Mulinare, J., Isenburg, J., Flood, T. J., Ethen, M., … Kirby, R. S. (2015). Updated estimates of neural tube defects prevented by mandatory folic acid fortification – United States, 1995-2011. *Morbidity and Mortality Weekly Report, 64*(01), 1-5.

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