

Approximately 1,500 babies are born each year in the United States with spina bifida (Centers for Disease Control and Prevention [CDC], 2014a). Spina bifida is a birth defect that occurs when the vertebrae do not form properly around the spinal cord. It is the most common defect of a group called neural tube defects. The “neural tube is the embryonic structure that eventually develops into the baby’s brain and spinal cord and the tissues that enclose them” (Mayo Clinic, 2014c). In a normal pregnancy, the neural tube forms by the 28th day after conception, which is often before a woman knows she is pregnant (March of Dimes, 2014).

Spina bifida occulta, often called hidden spina bifida, is the mildest and most common form of this condition. As many as 15% of healthy people may have this condition and not know it (Spina Bifida Association of America (SBAA), 2014d). This type occurs when there is a small separation or gap in one or more of the vertebrae of the spine. The majority of people with spina bifida occulta do not experience symptoms and therefore do not require any treatment. In fact, the only indication of this condition may be an abnormal tuft of hair, a collection of fat, a small dimple or birthmark, or discoloration of the skin over the affected area of the spine (Mayo Clinic, 2014e).

Meningocele, the rarest form, is a condition where the protective membranes around the spinal cord (meninges) push out through the opening in the vertebrae. However, the spinal cord usually develops normally and the membranes can be removed with surgery with little or no damage to the nerves (Mayo Clinic, 2014e).

Myelomeningocele is the most severe type of spina bifida and often causes considerable nerve damage and severe disabilities (SBAA, 2014d). In myelomeningocele a segment of the spinal nerves push out of the spinal canal. Sometimes the skin covers the sac that is formed by the nerves, but most of the time tissues and nerves are exposed increasing the chance for life-threatening diseases (Mayo Clinic, 2014e).

Medical problems that occur with spina bifida myelomeningocele and the severity of these complications are determined by the size and location of the neural tube defect, whether the skin covers the affected area, and whether spinal nerves come out of the affected area of the spinal cord (Mayo Clinic, 2014a).

- About 70% to 90% of patients develop **hydrocephalus**, or water on the brain, which is caused by circulation issues and can result in intellectual and developmental disabilities if not treated. When the fluid collects around the brain, the head can become enlarged and usually requires surgically inserting a tube called a shunt that drains the excess fluid (SBAA, 2014d).
- **Chiari II malformation** is where the “lower part of the brain sits in the upper part of the neck. Some babies with this condition have hydrocephalus (fluid buildup in the brain), weakness in the upper body (like in the arms and hands) and/or breathing or swallowing problems” (March of Dimes, 2014).
- **Tethered Spinal Cord**. The spinal cord, which normally slides up and down freely, is held in place with soft tissue. The cord

stretches as the baby grows causing nerve damage. Back pain, scoliosis, leg and foot weakness, changes in bladder or bowel control are potential issues. This condition is treatable with surgery (CDC, 2014c; March of Dimes, 2014).

- **Mobility impairments.** Depending on the severity of spina bifida and the location on the spinal column, the use of leg braces, crutches, and/or wheelchairs may be necessary (CDC, 2014c).
- **Nerve damage** also leads to bowel and bladder problems often requiring the use of urinary catheters and urinary tract infections are a common problem (CDC, 2014c).
- **Latex allergy** is a high risk, probably resulting from frequent medical interventions at an early age (SBAA, 2014b).
- **Secondary conditions** include learning disabilities, depression, obesity, skin breakdown, tendonitis, psychological and sexual issues (SBAA, 2014c).

Risk Factors

The exact cause of this birth defect is unknown, but many doctors and scientists believe that environmental and genetic factors play a role (CDC, 2014c). However, 95% of babies with spina bifida are born to parents with no family history of the disorder (SBAA, 2014c).

- **Folic acid deficiency** is one significant factor that seems to increase the chance of having a baby with spina bifida. Folic acid is essential for the functioning of the human body. However, *in order to prevent spina bifida and other neural tube defects, folic acid must be consumed before a baby is conceived.* Studies have shown that if all women in the United States took enough folic acid prior to and during early pregnancy, up to 70% of neural defects could be prevented (SBAA, 2014a).
- **Other risk factors** include:
 - Race – there is a higher incidence of spina bifida among Hispanics and whites
 - Sex – girls are affected more often
 - Previous pregnancy with spina bifida
 - Obesity in the mother
 - Maternal diabetes

- Some medications, primarily anti-seizure medications
- Exposure to high temperatures/increased body temperature, i.e. high fever or use of sauna

(Mayo Clinic, 2014d)

Early Detection

Spina bifida and other neural tube defects can be detected before birth. The primary prenatal test for birth defects is a blood analysis to check for certain high levels of a protein, called alpha-fetoprotein (AFP screening test), which is elevated for women having fetuses with spina bifida. After an ultrasound to check for gaps in the fetus' spine and to assess severity, the mother might be advised to have an amniocentesis to analyze the level of AFP present in the amniotic fluid (Mayo Clinic, 2014f).

Treatment for spina bifida can begin even before the baby is born. There is ongoing research surrounding prenatal surgery as an effective treatment. The first successful repair of spina bifida in a fetus was reported in 1998. A randomized trial of fetal surgery versus the standard treatment after birth, begun in 2003, was closed in 2011 “because the efficacy of fetal therapy –the fetal therapeutic approach – had been demonstrated. Babies who were treated before birth did better in virtually all aspects of their neurologic function. Fewer needed a ventricular peritoneal shunt for hydrocephalus. The ability to walk occurred twice as frequently in the fetal surgery group compared to the postnatal surgery group. Finally, the effects of hindbrain herniation were much less marked in the fetally repaired group” (Adzick, 2012, p.1). The procedure carries a variety of risks including premature birth, uterine scarring that can break down during the pregnancy and the need for the mother to have a cesarean delivery for this and all subsequent pregnancies (Adzick, 2012).

“Cesarean birth may be part of the treatment for spina bifida. Many babies with myelomeningocele tend to be in a feet-first (breech) position” (Mayo Clinic, 2014g). Researchers have also determined that caesarean section is the best way to deliver a normally presented (head down) spina bifida baby with a large cyst.

In cases of spina bifida occulta, no further treatment is usually required. However, if the child has spina bifida myelomeningocele, surgery will be performed within 24-48 hours after birth in order to correct the protruding nerves and tissues. A shunt to drain cerebrospinal fluid may also be inserted at the same time. Ongoing specialist care is usually required to assist children who have problems resulting from nerve damage (Mayo Clinic, 2014g).

It is estimated that every baby born with spina bifida will have an average lifetime medical cost of \$635,000 (Case and Canfield, 2009).

Social and Emotional Issues

Children with spina bifida can often lead relatively long and normal lives with the help of today’s technology. “About 90 percent of babies born with spina bifida now live to be adults, about 80 percent have normal intelligence and about 75 percent play sports and do other fun activities” (SBAA, 2014d). Children with spina bifida can walk short distances with the help of a cane, brace or walker, and can be very independent. While most children with this condition have normal intelligence, sometimes early intervention is needed. Encouraging children with spina bifida to engage in activities with their peers, while keeping in mind their physical limitations, is key to having an emotionally balanced child (Mayo Clinic, 2014b).

Prevalence in Texas

From 1999-2011 there were 1,872 children born with spina bifida in Texas, 206 of whom were in Region 8 which includes the KCF counties of interest (Texas Department of State Health Services, 2015). The following table presents the most recent data for the number of spina bifida births in the Kronkosky counties of interest over a three-year period:

Number of Spina Bifida Births				
County	2004	2005	2006	Total
Bandera	0	0	0	0
Bexar	14	4	13	31
Comal	1	0	0	1
Kendall	1	0	0	1
Total	16	4	13	33

(Texas Department of State Health Services, 2014)

Adults and children with spina bifida can find themselves suffering from a variety of health, social, and financial difficulties. Over the past several years resources have become more readily available to families dealing with these special challenges. Medical researchers continue to look for a cure, but until then, caregivers can only promote prenatal health while improving the quality of life for those already living with this condition.

Spina Bifida Texas is one of three Spina Bifida support organizations in Texas and the only one supporting the Central and South regions of the state (Spina Bifida HQ, 2013). It provides a variety of services to families coping with spina bifida including:

- *New parent support* – information packets detailing medical and community resources, meetings with veteran parents, and support groups in both English and Spanish
- *Resource library*
- *Lending closet* of medical supplies and equipment
- *Self advocacy support* – help with eligibility for special programs and educational needs
- *QOL & Scholarship opportunities* including sponsorship to attend camp and participate in sports and arts programs
- *Independence training* to help teach children about personal hygiene and self-catheterization skills
- *FashionABLE* –pairs high school fashion design students with spina bifida models where designers create a garment modified to help the child in daily life
- *SB Conference* – full day conference covering medical, independence, learning disabilities, and physical fitness
- *Prevention Awareness*—Folic acid awareness outreach

(Spina Bifida Texas, 2012)

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