Care of the Adolescent with Spina Bifida

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KEYWORDS

• Spina bifida • Adolescence • Chiari • Hydromyelia
• Tethered cord syndrome • Scoliosis • Club foot
• Latex allergy

Teenagers with spina bifida cystica have survived significant challenges to this stage of development. Unlike other many chronic conditions, myelodysplasia with attendant spinal dysraphic disorders can be and often is progressive. In addition to the neurologic deficit present since the prenatal period, hydrocephalus, Chiari malformations with hydromyelia, tethered cord syndrome, or combinations of one or all lead to changing functional ability and progressive deformities. Orthopedic deformities, such as scoliosis, foot and ankle deformities, dislocated hips, and knee flexion and valgus deformities, often appear. These new deformities are frequently a warning about a symptomatic tethered cord, hydromyelia, or shunt failure. Many of these deformities can be prevented from appearing or becoming destabilizing if the subtle signs are picked up and early intervention is provided. This has been one of the great advances in the care of patients who have myelodysplasia that was fostered over the past two decades by Dr. David McLone of Children's Memorial Hospital in Chicago. Dr. McLone[1] remembered the deformed teens who were afflicted by severe scoliosis, lower extremity rigid deformities, and bladder and bowel incontinence, all leading to a miserable life of restricted confinement. With the new attention to the neurologic conditions, the teens now may often be walkers with straight spines, interacting with the world in far more effective ways than was expected 30 years ago. Advances in urologic surgery have also provided better control of continence in addition to reduced infections and renal destruction. Adolescence is a time for continued monitoring of the adolescent for signs of deterioration and early but aggressive intervention so that more severe deformities and disabilities are prevented. Researchers have also been studying the educational and social development of these adolescents so as to obtain some idea of prognosis for adult function. Many studies are on patients who did not have special attention paid to their neurologic condition; thus, the research, educational, and medical communities need to undertake further study to see if they can improve the prognosis for these adolescents as they become adults.

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This article reviews the monitoring and treatment considerations that have led to such a significant improvement in outcomes in patients who have myelodysplasia.

NEUROLOGIC DISEASE PROGRESSION

Neural tube development is disrupted by maternal folic acid deficiency. Spina bifida is also related to maternal use of valproic acid or carbamazepine.[2–4] Maternal diabetes or pregnancy by the same partner with offspring who are also afflicted is also associated with neural tube deformities. Once present, these deformities can be progressive and lead to further loss of function and spine and lower extremity deformities. The adolescent is especially prone to these problems because of rapid growth, especially in the case of tethered cord, deterioration in the case of hydromyelia or Chiari malformation, or a failed ventriculoperitoneal (V-P) shunt. Understanding and management while the symptoms are early and mild can prevent more severe loss of function and deformities.

Hydrocephalus

Hydrocephalus, which occurs early in patients who have spina bifida, is usually treated with a V-P shunt within the first month of life, often before the patient is discharged from the hospital after birth. Although most shunt failures seem to happen in the preteen years, teenagers still can experience rapid or delayed symptoms of shunt failure. Rapid shunt failure presents with the familiar symptoms of headaches, nausea, photophobia, and lethargy. Rapid shunt failure can be life threatening and requires emergent repair or replacement.

Slow or delayed shunt failure presents as a subclinical picture. This might be the teen who has a recent change in a club foot or metatarsal adductus deformity, especially if the change is unilateral. Shunt failure may also present as a new level or location of weakness, again in a unilateral presentation. The newly formed deformities may lead to pressure sores on the foot, sacral, or pelvic area. The physical therapist or orthopedic surgeon is usually the first to see these changes. It is important that the whole treatment team identifies that these subtle changes may represent shunt failure and initiates evaluation by repeat CT or MRI scanning compared with baseline studies. Repair and replacement of the shunt may reverse the process; however, that is not always the case.[5] Orthopedic procedures may be required to restore function and to prevent or allow healing of pressure sores secondary to the new deformity.

CHIARI MALFORMATION AND HYDROMYELIA

This malformation is believed by McLone[6] to represent the growth of the cerebellum as the last structure of the brain to develop but within an already crowded ''box,'' the skull. The reduced rostral volume is believed to be a result of reduced hydrostatic pressure secondary to the lower spina bifida defect. This leads to a smaller than required skull after the cerebellum develops fully. The result is herniation of the tonsils of the cerebellum through the foramen magnum and dorsal to the upper cervical cord. The Chiari malformation often results in abnormal cerebral spinal fluid dynamics and the development of hydromyelia, also called a syrinx, of the spinal cord. Except for bulbar symptoms, which can occur from direct pressure on the brain stem and upper cord, many of the problems are caused by hydromyelia. The cysts, which have several suspected theories of pathogenesis,[7,8] can be of varying size and range along the spinal cord. They often reach holocord dimensions with just a rim of spinal cord around the fluid-filled cyst. The hydromyelia is associated with scoliosis[9] and with lower extremity deformities and weakness.[10] If the multispecialty team is alert to these findings