Evaluation and Lifetime Management of the Urinary Tract in Patients with Myelomeningocele

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KEYWORDS

• Spina bifida • Urology • Transitional care • Myelomeningocele • Neurogenic bladder

KEY POINTS

- Goals in the lifelong management center around renal preservation as a priority, achieving some degree of continence, and optimizing quality of life.
- Management is multimodal and can be complex; although management techniques may vary throughout a child's lifetime, the goals remain constant.
- Complex medical needs become more challenging as patients become older and need to transition to adult providers faced with incorporating these patients into their practice.
- Continued evaluation of development and implementation of transition plans is imperative to promote smooth and successful transitions in the future.
- Different phases in life offer new challenges and barriers that require vigilant health care interactions and patient dedication.

INTRODUCTION

Spina bifida (SB) is a neural tube defect that is one of the most common causes of congenital lower urinary tract dysfunction. Although disease manifestation can be variable from patient to patient, it commonly affects bladder and bowel function, cognition, and the neuromusculoskeletal system.[1] The most frequent form is myelomeningocele (MMC), characterized by the extrusion of the spinal cord into a sac filled with cerebrospinal fluid.[2]

Epidemiology

SB affects all racial and ethnic groups with an overall prevalence from 2004 to 2006 of 3.5 per 10,00 in the United States when adjusted for maternal race and ethnicity.[3] Using the Kids' Inpatient Database, Lloyd and colleagues[4] concluded that the prevalence of the disease has been stable from 1997 to 2009. The worldwide incidence of neural tube defects has been cited at 0.3 to 4.5 per 1000 births.[5]

According to data from the Centers for Disease Control and Prevention, the prevalence declined nearly 20% among infants born to non-Hispanic black mothers and remained constant in infants born to Hispanic mothers and mothers of non-Hispanic white ethnicity between 2000 and 2005.[6] In the analysis by Lloyd and colleagues[4] using the Kids' Inpatient Database, there was a higher prevalence of SB in the Hispanic cohort as well. Over the past, century survival rates for babies born with SB have improved drastically, and this can be attributed to improvements in

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medical and surgical management with the introduction of the ventriculoperitoneal shunt (VPS), antibiotics, and intermittent catheterization.[7] Today, 85% to 90% of children are surviving into adulthood and, compared with those born in 1975, twice as many newborns with SB survived in the United States in 1995.[7]

Management Goals

Primary goals in the lifelong management of SB patients centers around renal preservation as a priority, achieving some degree of continence, and optimizing quality of life. Although we can rely on specific markers of renal preservation, such as imaging and functional studies, continence measures can be more subjective and variably defined. Although the International Children's Continence Society guidelines define incontinence as "uncontrollable leakage of urine," this leaves room for a variable interpretation. Furthermore, no general consensus exists on specific measures of continence for research purposes. Lloyd and colleagues[8] reviewed the literature to assess the degree of standardization of urinary incontinence definitions in children with SB. They found that out of 105 articles that met inclusion criteria, only 57% of studies had a clear definition of continence. The most commonly used definition was "always dry," which was used in 24% of studies.

Other variables, including presence of a VPS, can also have a significant impact on quality of life. Ramachandra and colleagues[1] evaluated health status and demographic variables as potential factors in patient- and parent-reported health-related quality of life. The study found that patients with shunted hydrocephalus had worse perceptions of their physical health than those without; however, this perception seemed to improve in older patients. Other studies have found that spinal lesion level and the number of shunt revisions was associated with poorer quality of life.[9]

In the United States, there are currently an estimated 25,000 children ages 0 to 19 years and about 166,000 of all ages currently affected by SB.[10] The management of patients with SB is often multimodal and can be complex; although management techniques may vary throughout a child's lifetime, the goals remain the same. These goals center around the upper urinary tract, prevention of urinary tract infections (UTIs), and establishing some degree of acceptable continence. As noted by the International Children's Continence Society, continence is usually addressed as the child reaches school age; however, upper urinary tract damage secondary to elevated detrusor pressure and/or reflux, and chronic UTIs are continuously addressed and may evolve over time.[11] Additional considerations such as the development of urolithiasis and the associated burden are highlighted in the adolescent years and into adulthood. These complex medical needs become more challenging as pediatric patients become older and need to transition to adult providers who are faced with incorporating these patients into their adult practice.

PRENATAL DIAGNOSIS

SB can be diagnosed as early as the first trimester on prenatal ultrasound imaging. Antenatal ultrasound findings suggest that insults to both the central and peripheral nervous systems may be progressive and sequelae may worsen during gestation.[2] Damage to the spinal cord and peripheral nerves usually is evident at birth and is irreversible despite early postnatal surgical repair.[2] Given these findings and that long-term survivors usually have major disabilities, including paralysis and bowel and bladder dysfunction, the Management of Myelomeningocele Study (MOMS) sought to evaluate whether prenatal repair of MMC resulted in improved neurologic function.

Management of Myelomeningocele Study

The MOMS trial randomized eligible women to undergo either prenatal surgery before 26 weeks of gestation or standard postnatal repair. The study found the primary outcome (fetal or neonatal death or the need for a cerebrospinal fluid shunt) was reduced in the prenatal surgery group (relative risk, 0.7). In the prenatal group, 40% of patients still required shunting, however, this was compared with 82% in the postnatal intervention group. Prenatal surgery was associated with increased risk of preterm delivery and uterine dehiscence at delivery. Furthermore, although prenatal surgery decreased the need for VPS and improved lower extremity outcomes, the prenatal group required more procedures for delayed spinal cord tethering.[2] Based on these outcomes, it was concluded that prenatal closure seemed to improve neuromotor function and decrease the need for VPS. The long-term follow-up available on children who have undergone prenatal closure include those who underwent closure before the MOMS trial. Danzer and colleagues[12] reported on results in patients at a median age of 10 years supporting persistent improvement in neurofunctional outcome after fetal MMC repair (79% community ambulators; 9% household ambulators; and 14% wheelchair bound). Despite these improvements, behavioral abnormalities were