Review Article

Update on Urological Management of Spina Bifida from Prenatal Diagnosis to Adulthood

Devon C. Snow-Lisy, Elizabeth B. Yerkes and Earl Y. Cheng*

From the Ann and Robert H. Lurie Children's Hospital of Chicago, Northwestern University Feinberg School of Medicine, Chicago, Illinois

Abbreviations and Acronyms

BNP = bladder neck procedure

CIC = clean intermittent catheterization

CKD = chronic kidney disease

MACE = Malone antegrade continence enema

MOMS = Management of Myelomeningocele Study

QOL = quality of life

UTI = urinary tract infection

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* Correspondence: Division of Urology, Ann and Robert H. Lurie Children's Hospital of Chicago, Northwestern University Feinberg School of Medicine, 225 E. Chicago Ave., Box 24, Chicago, Illinois 60611 (telephone: 312-227-6340; FAX: 312-227-9412; e-mail: ECheng@luriechildrens.org).

Purpose: We review the current literature regarding urological management of spina bifida from prenatal diagnosis to adulthood.

Materials and Methods: We searched MEDLINE®, EMBASE® and PubMed® for English articles published through December 2014 using search terms "spina bifida," "spinal dysraphism" and "bladder." Based on review of titles and abstracts, 437 of 1,869 articles were identified as addressing topics related to open spina bifida in pediatric patients, or long-term or quality of life outcomes in adults with spina bifida. We summarize this literature to inform clinical guidelines and create a framework for disease management.

Results: The birth prevalence of spina bifida in the United States has recently plateaued at approximately 30 per 100,000. With improved management more individuals are surviving to adulthood, with an economic impact of $319,000 during the lifetime of an individual with spina bifida. Recent advances in prenatal surgery have demonstrated that prenatal closure of spina bifida is possible. To assess safety and efficacy, the National Institutes of Health sponsored Management of Myelomeningocele Study was undertaken, in which subjects were randomized to prenatal or postnatal closure. Until the urological results of this trial are published, the impact of prenatal intervention on future bladder function remains unclear. Controversy continues regarding the optimal use and timing of urodynamic studies, and the indications for initiation of clean intermittent catheterization and anticholinergics in infants and children. Many favor expectant management, while others argue for a more proactive approach. Based on the current literature, both approaches appear to protect the child from renal injury, although delayed intervention may increase rates of bladder augmentation. The current literature regarding this topic is difficult to interpret and compare due to heterogeneity of patient populations, variable outcome measures and lack of reporting of quality of life outcomes. Surgical intervention is indicated for those at risk for renal deterioration and/or is considered for children who fail to achieve satisfactory continence with medical management. Traditionally surgery concentrates on the bladder and bladder neck, and creation of catheterizable channels. For those with a hostile bladder, enterocystoplasty remains the gold standard for bladder augmentation, although use of bowel for augmentation remains suboptimal due to secondary complications, including increased risk of infections, metabolic abnormalities, neoplastic transformation and risk of life threatening perforation. Recent advances in tissue engineering technology may provide an alternative to traditional augmentation. However, recent results from phase II trials using current techniques to augment the bladder with engineered bladder tissue are disappointing. Catheterizable channels to the bladder and ascending colon further facilitate continence measures and promote independent

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care. While surgical reconstruction is clearly successful in improving continence, recent outcome studies have questioned the true impact of this type of surgery on quality of life. With improved survival transitional care issues, including health related independence, sexual health needs and development of a support system, are increasingly important. Transitional care remains a significant issue for which few public health measures are being quantitatively evaluated.

Conclusions: Despite consensus regarding early urological involvement in the care of patients with spina bifida, controversy remains regarding optimal management. Major reconstructive urological surgeries still have a major role in the management of these cases to protect the upper urinary tract and to achieve continence. However, future studies are needed to better clarify the true impact on quality of life that these interventions have on patients and their families. Transition of urological care to adulthood remains a major avenue for improvement in disease management.

Key Words: disease management, meningomyelocele, spinal dysraphism, urinary bladder

WITH a birth prevalence in the United States of approximately 30 per 100,000, spina bifida is the most common nonchromosomal birth defect, resulting in severe disability of multiple organ systems.[1] The economic impact of spina bifida is significant. Estimated total lifetime medical expenses adjusted for inflation have increased from $236,000 to $319,000 in just 20 years.[2] These estimates constitute only a part of the total economic impact of spina bifida.

Spina bifida is caused by a failure of the caudal neural tube to fuse normally in early development. A variety of neurological deficits can be seen, depending on the severity of the fusion abnormality and location of the lesion. Myelomeningocele, in which the spinal cord and neural elements are exposed, is the most common and clinically severe of the open spina bifida defects. Variable impact on the somatic, parasympathetic and sympathetic innervation of the bladder affects the ability to store and empty urine, and can ultimately cause chronic kidney disease due to poor bladder dynamics. Urological issues can be a significant source of morbidity and mortality, and are implicated as a cause of death in almost a third of patients with open spina bifida followed long term.[3]

We examine the data driving current urological management of children with isolated spina bifida. We expound the data behind the current controversy regarding expectant vs proactive management. The challenges of integrating QOL outcomes to medical and surgical outcomes are reviewed. In addition, the issues of transitioning care into adulthood are briefly addressed. Spina bifida occulta, tethered cord and issues related to renal transplantation are outside the scope of this article.

METHODS

We searched MEDLINE, EMBASE and PubMed with the terms “spina bifida” or “spinal dysraphism” and “bladder.” These results were then narrowed to English language publications and human studies. We excluded case reports, expert opinions, editorials and review articles, except to identify other potentially relevant primary articles. Abstracts were reviewed to identify articles on the management of children with spina bifida from diagnosis to adulthood, excluding articles on renal transplantation, spina bifida occulta and cord tethering/retethering. A total of 437 articles of the 1,869 retrieved were identified for potential inclusion based on review of titles and abstracts.

RESULTS

Prenatal Diagnosis and Fetal Surgery

Accurate prenatal diagnosis of various forms of spinal dysraphism by ultrasound with determination of severity and prognostic factors is vital, due to a potentially high termination rate (up to 65%).[4] Ultrasound screening improves prenatal diagnosis and can also characterize the anatomical nature of the lesion, allowing identification of the level of the lesion, with low spinal lesions being associated with increased bladder dysfunction.[5] Although expensive, fetal MRI differentiates unique anatomical features.

Besides influencing the decision regarding termination, prenatal diagnosis also allows for consideration of prenatal closure of the defect. Prenatal vs postnatal closure was evaluated in MOMS, a multicenter, randomized, controlled trial.[6] Initial results revealed a decreased need for ventriculoperitoneal shunting and improved lower extremity motor outcomes. However, these benefits were partly offset by an increased incidence of preterm delivery and uterine dehiscence in those who underwent prenatal intervention.

While awaiting the urological results of MOMS, several groups have evaluated the results of prenatal intervention on urological outcomes using case-control methodology and patient cohorts that were mostly closed before MOMS.[7,8] They have noted no