Seminars in Pediatric Surgery 24 (2015) 73–78

Contents lists available at ScienceDirect

Seminars in Pediatric Surgery

journal homepage: www.elsevier.com/locate/sempedsurg

Transitional care in pediatric urology

Sarah M. Lambert, MD[a,b,*]

[a] Department of Urology, Columbia University, New York, New York
[b] Division of Pediatric Urology, New York Presbyterian Hospital, Morgan Stanley Children's Hospital, New York, New York

ARTICLE INFO

Keywords:
Neurogenic bladder
Transition of care
Pediatric genitourinary tumors
Posterior urethral valves
Bladder exstrophy
Long-term outcomes

ABSTRACT

The transition from childhood to adolescence and into adulthood occurs as a natural component of human development. As children progress through school and gain independence, health care practitioners must facilitate a parallel transition from pediatric to adult providers. Modern medicine has succeeded in extending the life expectancy for many children with complex conditions, and adult providers are participating in their medical care through adulthood. Transitioning pediatric urology care to adult urology care is unique to every individual and his or her underlying condition, while the transition process is universal. The objectives of all pediatric urologists include preservation of the kidneys and lower urinary tracts, safe urine storage, safe urine drainage, urinary continence, fertility, sexual function, and genital cosmesis. For some children, these objectives can be attained during childhood, while other children require lifelong maintenance and management. Children with posterior urethral valves, exstrophy–epispadias complex, cloaca, vesicoureteral reflux, neurogenic bladder, disorders of sex development, cancer, hypospadias, nephrolithiasis, undescended testes, varicoceles, ureteropelvic junction obstruction, solitary kidney, and upper tract anomalies all require long-term evaluation and management. The obstacles of altering a patient and caregiver paradigm, locating adult urologists with special expertise, coordinating care with other adult specialties such as nephrology, and navigating the adult health care environment can impede the transition process.

The transition from childhood to adolescence and into adulthood occurs as a natural component of human development. As children progress through school and gain independence, health care practitioners must facilitate a parallel transition from pediatric to adult providers. According to the American Academy of Pediatrics, "optimal health care is achieved when each person, at every age, receives medically and developmentally appropriate care."[1] Multiple organizations and agencies are currently focused on improving health care transition, including the American Academy of Pediatrics, American Academy of Family Physicians, and American College of Physicians. The U.S. Department of Health and Human Service's Maternal and Child Health Bureau also promotes comprehensive services for the transition of care period.[2]

Transition of care is essential but not without difficulties, limitations, and frustrations. The key components of transition include the patient, his or her caregivers, the pediatric providers, the adult providers, adult and pediatric support staff, adult and pediatric hospitals, insurance companies, and the health care system. The objective of this article is to highlight specific pediatric urologic conditions that require long-term care and transition.

Transition of care is not unique to urology, but it is common across all pediatric disciplines. While diseases such as diabetes and asthma are familiar to adult providers, congenital anomalies and pediatric disease historically associated with decreased life expectancy have no parallel in adult medicine. Modern medicine has succeeded in extending the life expectancy for many of these children with complex conditions, and adult providers become responsible for those who survive childhood. Cardiologists and pulmonologists are among leaders in training providers in transitional medicine that encompasses pediatric and adult care through specialized fellowship programs.

Pediatric urology encompasses many genital and urinary conditions. These conditions typically present prenatally or during childhood, but some conditions emerge in adolescence as well. These conditions range from mild to life-threatening. The objectives of all pediatric urologists include preservation of the kidneys and upper tracts, safe urine storage, safe urine drainage, urinary continence, fertility, sexual function, and genital cosmesis. For some children these objectives can be attained during childhood while other children require lifelong maintenance and

* Correspondence address: Division of Pediatric Urology CHN-11, New York Presbyterian Hospital, Morgan Stanley Children's Hospital, 3959 Broadway, New York, NY 10032.
E-mail address: sml72@columbia.edu

http://dx.doi.org/10.1053/j.sempedsurg.2015.01.004
1055-8586/

management. While all children deserve an organized transition of care, it is the latter group of children in which direct and seamless transition of care is mandatory from a urologic perspective. These children often require subspecialty care from other disciplines as well, such as orthopedics, general surgery, neurosurgery, endocrinology, nephrology, oncology, cardiology, and gynecology. Adolescents with complex urologic disease often also utilize non-physician provider services, such as catheterization nurses, stoma and ostomy nurses, social workers, psychologists, and child life specialists. The transition into late adolescence and adulthood also translates to insurance status changes and changes to caregiver relationships. These changes can impact confidentiality, informed consent, patient responsibility, transportation, and the physician–patient relationship.

For pediatric urologists, the concept of transitioning long-term patients with complex conditions can be difficult and uncomfortable. Clinicians, patients, and caregivers can all experience hesitation, ambivalence, and fear. Adolescents with chronic bladder conditions develop confidence and trust with their pediatric care provider and may feel ill prepared to transfer to an adult provider. Conversely, these adolescents also reported that issues surrounding relationships, sexuality, and fertility were rarely discussed in the pediatric office.[3]

Adult providers may also be faced with management challenges when accepting care of these patients. Young adult patients with congenital urologic disease are complex, with multiple urologic and medical issues not easily addressed. This situation can be overwhelming and lead to frustration on the part of both patient and health care provider. However, successful management of this transition is critical, as failure to sustain long-term follow-up of chronic urologic problems places these patients at high risk for adverse health outcomes.[4] Young adults lost to surveillance can present with devastating conditions, such as renal failure or complications from urinary incontinence including decubitus ulcers and sacral osteomyelitis.[5] Development of a standardized transition-of-care program and collaboration with adult providers is essential to safe and successful transition.

In discussing transition of urologic care, Rink[6] very simply and eloquently asked, “What problems will I be leaving?” Devising a process to support seamless management of these problems is the crux of the transition process. Luckily, children with hypospadias, undescended testes, and varicoceles often require only adult urology subspecialty care, which streamlines the transition process. Children with more complex disorders, such as posterior urethral valves, vesicoureteral reflux, hypospadias, nephrolithiasis, undescended testes, varicoceles, ureteropelvic junction obstruction, solitary kidney, and upper tract anomalies all require long-term evaluation and management. Such patients often require adult urology, nephrology and, occasionally, transplant surgery management. Finally, the most complex patients, such as children with cancer, disorders of sexual development, neurogenic bladder, cloaca, and exstrophy–epispadias complex, often require multiple subspecialty care including general surgery, endocrinology, gynecology, neurology, neurosurgery, gastroenterology, oncology, nephrology, and urology.

Transition-of-care plans for these children should include standardized goals, but they must be tailored to each child. An understanding of these congenital anomalies and the long-term issues associated with the disease processes, surgical interventions, and resultant complications is key to providing lifelong care. In the following sections, these conditions will be briefly reviewed and the long-term issues highlighted.

Neurologic conditions

Neurologic conditions in children include myelomeningocele, tethered spinal cord, cerebral palsy, sacral agenesis, and other spinal dysraphisms. Hinman’s syndrome, or non-neurogenic neurogenic bladder, can be included in this category as well. Spina bifida as a broad category is the most common permanent birth defect in the United States.[7] These neurologic conditions can be associated with other comorbidities and syndromes or can be isolated. The management of neurogenic bladder has significantly improved since the initiation of clean intermittent catheterization and anticholinergic therapy. The treatment armamentarium also includes botulinum toxin, augmentation cystoplasty, catheterizable channel creation, antegrade continence enema creation, other urinary diversions, and vesicostomy. These children are unable to store urine safely or empty urine efficiently. Consequently, their care is lifelong, and requires monitoring as the clinical situation can deteriorate. Prior to clean intermittent catheterization, the life expectancy of children with myelomeningocele was severely limited. These children often died of urosepsis, stone disease, and renal failure. Currently, this population is one of the largest pediatric populations in need of complex transition of care.

Children with neurogenic bladders are at risk for infection, upper tract damage, nephrolithiasis, urethral stricture, and bladder cancer. These children must be followed up for changes in bladder dynamics with serial urodynamic evaluation. Serial renal and bladder ultrasounds must be performed to assess upper tracts for dilation and nephrolithiasis. Urethral trauma and stricture need to be addressed and often require reconstructive surgical intervention. Incontinence of urine and stool can be a continued problem that persists into adulthood. Throughout adolescence, children with a history of myelomeningocele with worsening urodynamic parameters should be evaluated for cord tethering.

Children with neurogenic bladder who undergo bladder augmentation or urinary diversion in childhood have additional long-term risks that are associated with intestinal or gastric reservoirs. Many children with bladder augmentation have catheterizable channels created for urinary drainage. These channels are at risk for stenosis and diverticulum. Bladder augmentation increases the risks for metabolic acidosis, bone mineral density abnormalities, vitamin B12 deficiency, augment perforation, and tumor formation within the augmentation. The re-augmentation risk at 10 years post initial augmentation has been reported as 3.7–5.9%.[8] A large clinical series from Indiana University documented a 34% risk of further operative procedures after augmentation. Approximately 9% of children required a laparotomy for bowel obstruction. Additionally, they reported bladder perforation in 8.6% of children. Additionally, 15% of children with a mean follow-up of 13.3 years required treatment for bladder stones. The incidence of malignancy was 0.6% and all patients presented with metastases and died of disease.[9]

A review of adult dedicated spina bifida clinics at the Universities of Minnesota and Utah documented that 85% of young adults reported an active issue at the time of initial adult evaluation. Urinary incontinence, urinary traction infection, catheterization difficulties, and calculi were the most common. Of these young adults, 34% required a surgical intervention.[10] These complications are long-term issues and therefore require lifelong surveillance and treatment throughout adulthood. In the adult population, fertility concerns become an issue. Male patients often require assistance with reproduction including electroejaculation.

Posterior urethral valves

Boys with posterior urethral valves also require long-term follow-up to protect renal function and prevent further damage to the bladder and upper tracts. Although primary valve ablation or urinary tract diversion typically occurs in infancy, the valve bladder syndrome is a progressive condition that continues to