Transitional Care

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1. Introduction

This chapter is dedicated to the adult longitudinal care of people born with congenital genitourinary anomalies. The field is known by many names, including Transitional Urology and Adult Congenital Urology. There is much in a name and we won't solve that controversy here; however, the name should convey the difficulties and high risk of complications and delayed care in the early transition phase (see AUA Core Curriculum **Pediatric Urology: Neurogenic Bladder**) as well as the significant effort that is required to follow these people longitudinally through all phases of their adult lives. By nature of the topic, this chapter cross references with several other chapters in both the pediatric and adult sections of the Core Curriculum. In many cases we leave the description of the disease epidemiology and pathophysiology to the referent pediatric chapters and focus our discussion on the nuances inherent to adult care of the condition. Potential models for transition and the delivery of optimal care are beyond the scope of this work but do require significant attention.

Further, by focusing on some of the most common and acute conditions we see in our clinical experiences, we necessarily leave out several other congenital conditions that can demand the attention of the adult urologist such as secondary malignancies after pediatric chemoradiation, **differences of sexual differentiation**, muscular dystrophy and other rare inherited neuromuscular disorders.

Many of the conditions discussed are treated surgically early in life to help correct the anomaly and to prevent further injury to the urinary tract. However, the sequelae of such diseases require lifelong follow-up. This curriculum highlights the need for long-term care and the medical and surgical challenges of this growing and complex patient population.

2. Spina Bifida

2.1 Epidemiology

Spina Bifida (SB) or myelomeningocoele occurs in approximately 3 per 10,000 live births in the U.S.,[1] with a current U.S. prevalence of over 70,000.[1,2] Excellence in pediatric SB care has led to improved survival. Approximately 85% of pediatric SB patients now live into adulthood; in fact, it is

estimated that there are currently more adults than children living with SB. [3]

2.2 Presentation

As people with SB age into adulthood, their parents age too and become less able to care for them. Many are unemployed and on Medicaid; [4] they may live independently, in a group home or face food and housing security problems. Even in the best-case scenario of a well-established transition clinic, only about two-thirds will have a well-developed transition plan. [5] Approximately one-third of patients will undergo surgery within a year of transition (most commonly for urinary stones), reflective of delayed care. [6] Although we focus here on the urologic sequelae of adults with spina bifida, it is important to assess the entire patient for comorbid conditions: obesity, obstructive sleep apnea, congestive heart failure, decubitus ulcers and neurogenic bowel are all common ailments in this population.

2.3 Evaluation

Spina Bifida Association (SBA) guidelines for urologic care suggest that adults with spina bifida should undergo annual surveillance with renal/bladder ultrasound and serum creatinine (or other measure of renal function in those with low muscle mass). [7] Cystoscopy should not be performed routinely in those with prior ileal bladder augmentation. Surveillance cystoscopy is not cost-effective and is associated with a high false positive rate and unnecessary invasive confirmatory testing such as bladder biopsy.[8] Higuchi et al. demonstrated that in 65 patients with congenital bladder pathology who were 10 or more years post-bladder augmentation, annual cystoscopy and cytology yielded no bladder tumors over a follow-up period of 15 years; but testing did lead to 26 false positive cytologies and 4 false positive cystoscopies with negative confirmatory biopsy. [9] Cystoscopy of ileal augments should be reserved for those with gross hematuria, bladder pain, recurrent UTIs, or concerning findings on ultrasound such as hydronephrosis or bladder stone. [7] Patients with colon augmentations should undergo routine cystoscopy after the age of 50 as per suggested guidelines for colon cancer screening. [10] Cystoscopy is recommended for those with gastric augmentations due to the high risk of malignancy in this population.

In contrast to cystoscopy, the best timing for urodynamics is not so clear. Although spina bifida is a fixed spinal cord lesion (as opposed to a progressive disease like multiple sclerosis), the bladder dynamics change over time. One cannot assume that just because an adult with lumbosacral SB had a low-pressure large capacity bladder in childhood, they will always have a safe bladder; compliance can, and often does, decline with age, sometimes due to spinal cord tethering and sometimes just due to chronic bladder pathology. [11] We recommend performing intake urodynamics at the time of transition to adult care (particularly if no pediatric augmentation was done) and then only performing it in the future for cause: recurrent UTIs, bladder pain, new hydronephrosis or new/worsening incontinence.

2.4 Medical Management

Most SB patients transition to adult clinic already on CIC.[6] Medical management of **detrusor overactivity and low bladder compliance are similar to in childhood**; one starts with oral anticholinergics and/or beta-3-agonists. Botulinum neurotoxin injection is reserved for medication refractory cases; it is injected cystoscopically every ~6 months and reduces detrusor overactivity and can sometimes improve compliance. [12,13]

2.5 Surgical Management

Augmentation cystoplasty is generally reserved for medication-refractory cases. There is some evidence that the rates of pediatric reconstructive bladder surgery (including augmentation) are declining over the last 20 years and that those children who are receiving surgery are undergoing it in their teen years. One hypothesis is that improved medical management (including more widespread use of onabotulinum neurotoxin therapy) may be delaying augmentation until later in life. The techniques of ileal or colonic cystoplasty and Mitrofanoff or Monti creation are the same in adulthood as in pediatrics. The difference is that adults with SB tend to be more obese and many have had their appendix used for a prior failed Mitrofanoff or for a Malone antegrade continence enema procedure. These factors often lead us to need a double Monti, spiral Monti or continent cutaneous ileocecocystoplasty (CCIC) in adult SB patients. Further, Husmann has shown that obesity is present in 50% of adults undergoing continent channel creation and that obesity is associated with higher rates of channel stenosis in Monti stomas than in tapered ileal stomas. [14] For these reasons, we often prefer the CCIC in adult SB patients who require both an augment and a catheterizable channel.[15]

The variations of the Monti are well-described **elsewhere**. The technique of CCIC is similar to that of the right colon pouch (i.e., Indiana pouch), otherwise known as an Indiana augment.[16] The bowel segment is harvested based on the ileocecal pedicle including about 15cm of cecum and about 10cm of terminal ileum. The cecum is detubularized and used as the augment. As the cecum is rotated into the pelvis, the ileocecal valve naturally rotates to the dome of the augment and serves as the continence mechanism. The ileum is staple-tapered over a 12-16F catheter and trimmed to the length needed to make a non-redundant channel as it is matured to the umbilicus or abdominal wall.

Bladder outlet procedures to help achieve continence include bladder neck artificial urinary sphincter, bladder neck sling and bulking agents and are well described elsewhere. See AUA Core Curriculum: **Spina Bifida and Neurogenic Bladder**.

2.6 Complications

Finally, surveillance is particularly important in patients with prior surgical reconstruction of the lower urinary tract. Complications include bladder perforation, urinary tract infections (average of 1 to 3 per year),[17,18] urinary stones (~10%), new incontinence per urethra or stoma, and channel stenosis, stricture or false passage (~20-70%). Aggressive daily bladder irrigation with at least 240cc helps prevent urinary tract infections and stones in those with an augment.[19] Dilation of the stenotic channel should be done cautiously and only to the minimum size necessary to resume catheterization. Aggressive dilation can be associated with injury to adjacent bowel and fistulization. Use of an

L-stent overnight can avoid operative revision in those with stomal stenosis.[20] Revision of the channel for stricture, stenosis or incontinence is challenging and should be referred to centers of excellence. [21] We recommend annual follow-up with renal bladder ultrasound, basic metabolic panel, and vitamin B12 level (after year five) for all patients with a history of bladder augmentation.

3. Cerebral Palsy

3.1 Epidemiology and Background

Cerebral palsy (CP) is the most common disability of childhood; it is 10 times more common than spina bifida. CP is a motor and spasticity disorder caused by neonatal or infantile insults to the motor cortex of the brain. CP severity is highly variable; physical manifestations range from a nearly imperceptible limp to spastic quadriplegia. Up to 50% of people with CP have cognitive dysfunction. One-third of children with CP will have neurogenic bladder (NGB) with associated urinary incontinence. [22] Approximately 95% of children born with CP live past 10 years of age and survival rates continue to improve, largely due to advances in care for those most severely affected .[23]

3.2 Differences between pediatric and adult presentation and management

The degree of NGB is often correlated with the severity of CP. Pediatric urinary symptoms include urinary tract infections, urinary frequency, and incontinence. Urodynamics reveal reduced cystometric capacity and neurogenic detrusor overactivity (NDO), but post-void residuals are low and bladder compliance is usually preserved. [22] These symptoms and urodynamic findings are consistent with those one would predict from a **suprapontine neurologic injury** like CP. Mild symptoms can be managed with anticholinergics or onabotulinum neurotoxin therapy (BoNT); children with more severe incontinence typically remain in diapers. Adults with mild CP can become continent with the help of anticholinergics and detrusor BoNT, similar to pediatric management. However, in adults with severe CP, a combination of decades of NDO plus pelvic floor muscle spasticity leads to a complex clinical and urodynamic picture.

Pseudodysynergia describes spasticity of the pelvic floor musculature including the external urinary sphincter that leads to outlet obstruction. It is termed pseudodysynergia rather than **detrusor-sphincter dyssynergia** (DSD) because it is due to a suprapontine injury rather than a suprasacral spinal cord injury. Pseudodyssynergia can describe any etiology of pelvic floor muscle spasticity (e.g., associated with chronic pelvic pain syndrome) but the pseudodysynergia associated with CP is unique in that it is neurogenic, the spasticity is tonic rather than intermittent, and the intensity of the spasticity is so great that even urethral catheterization is difficult. While the degree of pseudodysynergia varies, it is the driving force that differentiates NGB due to CP from NGB due to other disorders.

3.3 Chronic urinary retention, myogenic failure and CIC challenges

While about half of adults with advanced CP will present with symptoms typical of suprapontine NGB and urodynamics will confirm NDO, at least a quarter will present with urinary retention due to

pseudodysynergia and urodynamics will show a large capacity acontractile bladder .[24] Patients in retention may void only once or twice a day and will have high post-void residuals; their voids will be such high volume that they overflow their diaper and “flood” their clothing, wheelchair or bedsheets. A typical urologic response is anticholinergics and CIC; or when that fails then placement of an indwelling urethral or suprapubic catheter. However, pseudodysynergia (and often lower extremity spasticity) makes CIC painful and technically difficult; it distresses the caregivers and reduces patient quality of life (personal observation). We avoid CIC whenever possible. Instead, we permit urinary retention as long as the patient voids at least twice a day, their flooding is not problematic, urinary tract infections are <3 per year and there is no hydronephrosis or bladder stones on annual renal-bladder ultrasound. With this algorithm and very close follow-up we have only had to resort to CIC or indwelling catheter in 25% of our advanced CP population (7% for hydronephrosis). [25] Others have confirmed that pseudodysynergia is a risk factor for upper tract deterioration; so, close follow-up is key. [26] We also frequently use **BoNT injection to the external sphincter** (100-200U in 2-4cc NS) as has been described for DSD in patients with multiple sclerosis.

When attempts at preserving spontaneous voiding have failed and CIC per urethra is impossible, we offer suprapubic tube placement or catheterizable channel creation (See AUA Core Curriculum **Pediatric Neurogenic Bladder section 9.3**). For patients with advanced CP the catheterizable channel eases CIC by solving two problems at one time: it avoids catheterization through the spastic external sphincter, and it facilitates catheterization in a wheelchair. Of course, many patients with advanced CP cannot catheterize themselves due to severe limb spasticity. In those cases, it is important to confirm that there is adequate assistance at home to catheterize the patient. Interestingly, because it is difficult to initiate CIC per urethra pre-operatively, the pre-operative urodynamic findings can be misleading: the large capacity acontractile bladder seen pre-operatively can convert to a small capacity, low compliance bladder with NDO post-operatively once the bladder is allowed to decompress with routine CIC via the catheterizable channel. [27] One should strongly consider routine augmentation cystoplasty at the time of catheterizable channel creation for adult CP patients who are in chronic retention, regardless of urodynamics findings.

4. Hypospadias

4.1 Epidemiology

*Hypospadias occurs in about 0.5% of newborn males. Historically, success rates of pediatric distal **hypospadias repair** are very high; however, the few studies with long-term follow-up present a more sobering picture. A review by Tourchi et al[28] chronicles the micturition, cosmetic and sexual outcomes across 19 studies that used various hypospadias repair techniques in a total of 859 men. Though this represents only a select sampling of mostly young men with a history of pediatric penile surgery (the mean age of men was in the 20’s for the majority of studies) it is important to recognize the potential for long-term genital dissatisfaction in these patients.*

4.2 Presentation

38-80% of adult men with previous hypospadias repair (AMWPHR) are dissatisfied with their urination. Common urinary complaints include spraying and post-void dribbling.[29,30] Few studies report long-term flow rates; those that do generally show good results. There is limited data regarding urethral calibration, cystoscopy or urethrogram outcomes in AMWPHR. Several studies have examined cosmetic satisfaction and the results are inconsistent; some show no difference in satisfaction with genital appearance compared to controls while others show a difference. One consistent finding is that men born with more proximal hypospadias are less satisfied than men born with distal hypospadias. Similarly, reports of erectile function are inconsistent but **ejaculatory dysfunction** is a consistent complaint.[28]

When AMWPHR present to the adult reconstructive urologist they have already undergone, on average, three prior attempts at repair.[31] Complaints can include stricture, fistula, hair in the urethra, stone in the urethra, recurrent urinary tract infections, pain, penile curvature and/or shortening.

4.3 Evaluation

Like any stricture patient it is important to collect uroflowmetry, patient reported outcome measures and to understand the patient's goals of surgery. One should make every attempt to retrieve old operative notes so as to understand the altered anatomy during reoperation. Finally, one should have a low threshold to perform the cystoscopy (using a narrow scope) and retrograde urethrogram under anesthesia. Unlike the average stricture patient, most AMWPHR have been traumatized by multiple repairs and embarrassing exams since childhood. [30] Performing the assessment under anesthesia provides better information and is patient-centered.

4.4 Management

Occasionally strictures can occur proximal to the prior hypospadias repair. These may be iatrogenic due to prior catheterization, urethral dilation or perineal urethrostomy. If these are far enough from the prior repair (i.e., mid to proximal bulbar urethra) then they can be corrected like any other **bulbar stricture**, except that it makes sense to avoid urethral transection or extensive mobilization so as not to compromise the blood supply to the fragile distal urethra.

Most strictures, though, occur at one of the ends of the prior repair or throughout the prior repair. Depending on the patient's goals, meatal stenosis can be managed with dilation, meatoplasty, or 2-stage repair. [32] Strictures throughout the repair or in the proximal end of the repair should be reconstructed in 2 stages due to prior disruptions in the native blood supply and a poorly supported spongiosum. Buccal mucosa is often quilted to the corpora cavernosa in the first stage and a tunica vaginalis flap is used to protect the urethral closure in the second stage. Stricture-free rates are approximately 80%[30] but it can take more than two operations to achieve success. Depending on the patient's goals, age and length and location of the stricture, alternatives may include urethral dilation or perineal urethrostomy.

5. Posterior Urethral Values

5.1 Epidemiology

Posterior urethral valves (PUV) are the most common form of congenital bladder outlet obstruction in boys. Today, most boys with **lower urinary tract (LUT)** obstruction are diagnosed by prenatal ultrasound. PUV are estimated to occur in 1.6 to 2.1 per 10,000 live male births.[33] There is a spectrum of severity of disease associated with PUV from delayed continence to significant pulmonary hypoplasia from severe oligohydramnios. Up to 30% of PUV patients will progress to end stage renal disease in their lifetime. [34]

5.2 Presentation

Bladder, renal and sexual dysfunction in adulthood may occur as a sequela of the structural and functional changes of the LUT obstruction and surgical and medical therapies for obstruction.

5.3 Evaluation

Once obstruction has been relieved surgically, boys with PUV require lifelong therapy to optimize bladder function and to maintain renal health. Standard guidelines regarding follow-up care of men with PUV do not exist. Attention to renal function is critical and yearly nephrologic follow-up with attention to proteinuria and polyuria is recommended. Long-term attention to bladder function is also important as bladder function may change over the course of childhood and adolescence.

LUT dysfunction may be related to the abnormal bladder or bladder neck, residual valve tissue, stricture, polyuria and upper tract dilation. Lower urinary tract symptoms including hesitancy, weak stream, incomplete emptying and straining are of increased prevalence but not severity in men with PUV. [33] Variable urodynamic findings are reported including detrusor overactivity, reduced compliance and hypocontractility.[35] Boys may have worsening upper tract dilation, reduced sensation of bladder fullness and poorly compliant bladders known as valve bladder syndrome.[36] Additionally, a poorly compliant bladder may decompensate to a hypocontractile bladder over time (myogenic failure). New or worsening incontinence or hydronephrosis should prompt urodynamic evaluation.

Changes in voiding may also be indicative of stricture disease. A recent study of stricture disease in boys, found 5.6% to be related to valve fulguration.[37] Urethral stricture is likely iatrogenic related to injury to corpora spongiosum as a consequence of poor technique, size mismatch between scope and urethra, and monopolar current.[38,39] “Dry” urethras after valve ablation may also result in stricture.

A nadir creatinine of less than 0.7ng/dL in the first year of life portends good long-term kidney function, however up to 100% of boys with a creatinine greater than 1 ng/dL will require renal replacement therapy.[40] Progression to ESRD in adolescence and young men is not as predictable but may be related to high grade VUR and untreated or persistent bladder dysfunction.[41] In one series a third of ESRD cases presented after the age of 17 years. [42]

Most men treated for PUV in childhood have good erectile function, with International Index of Erectile Function (IIEF) scores similar to the general population.[43] However, sexual function and fertility may be adversely affected by associated chronic kidney disease (CKD) and ESRD. [44] Small

studies have found overall fertility in these men is similar to the healthy population.[35,43] However, low or normal sperm counts have been reported with some men having immotile sperm, increased liquefaction times, abnormal sperm agglutination, pyospermia and retrograde ejaculation.[45,46,47] High rates of **cryptorchidism** (up to 10%) are also found in boys with PUV.[44] Scarring of the posterior urethra and reflux into the seminal vesicles and ejaculatory ducts may occur causing epididymo-orchitis and potential negative effect on semen parameters.[48] Retrograde ejaculation may occur as a result of alpha blocker therapy or prior bladder neck incision.

5.4 Medical and Surgical Management

The bladders of boys and adult men with PUV are often overactive and thick resulting in upper tract dilation and poor bladder emptying. Poor bladder emptying coupled with high urine output from nephrogenic diabetes insipidus in some may result in a need for double voiding, intermittent catheterization (CIC) or overnight catheterization. Alpha-blockers may facilitate bladder emptying. Antimuscarinics may improve bladder overactivity, diminished compliance and elevated storage pressure. Antibiotic prophylaxis may become necessary in those with vesicoureteral reflux (VUR) and/or poor emptying. VUR spontaneously resolves in most boys following valve incision and therapy for bladder dysfunction. [49]

Men with a history of PUV may have normal voiding, detrusor overactivity, poor compliance or myogenic failure requiring catheterization. Polyuria may develop as a result of deterioration of the kidneys' concentrating ability. Unchecked polyuria may worsen bladder function as in myogenic failure and may require frequent urination or overnight catheterization.[50] Men with valve bladder syndrome may also require catheterization.

Renal replacement therapy with dialysis or renal transplantation may become necessary in up to 36% of PUV patients by the age of 30 years.[51] Prior to transplantation, bladder dynamics should be evaluated, and treatment should be customized to the individual. Graft survival in valve patients with good adherence is comparable to non-affected patients requiring transplant.[52]

Men with PUV should be counseled on potential need for reproductive assistance if difficulties achieving conception arise.

6. Cryptorchidism

6.1 Epidemiology

Cryptorchidism affects approximately 3% of full-term boys and is more common in premature infants.[53] **Orchiopexy is recommended** between 6 and 18 months of life. Testicular examination is recommended as part of the annual physical examination by pediatricians as testicular position may change during childhood, and in rare cases ascend.

6.2 Presentation

Two significant concerns for boys with a history of undescended testicles as they reach adulthood

are subfertility and testicular malignancy. Poor testicular position following orchiopexy may be a consideration for some men.

6.3 Medical/Surgical Management and Complications

6.3.1 Fertility

Fertility is adversely affected by cryptorchidism. There is a six-fold increased risk of infertility in previously bilateral cryptorchid men (38%) when compared to matched controls (6%).[54] The paternity rate in formerly unilateral cryptorchid men is 89.7%.[54] Additionally, time to pregnancy is almost three times longer for bilateral cryptorchid men when compared to unilateral cryptorchids and controls (33.9 months vs. 11.1 months).[55] In a long-term study following young adults who had orchiopexy in childhood with initial testicular biopsy, lack of germ cells correlated to a 75 to 100% risk of infertility. Lack of germ cells on biopsy of previously unilateral cryptorchid boys was associated with approximately 33% risk of later infertility.[56,57,58] Attempts at paternity, particularly in previously bilateral cryptorchid men, may require assisted reproductive techniques.

6.3.2 Malignancy

There is a significantly increased incidence of **testicular cancer** in men with a history of cryptorchidism varying from 0.05% and 1%.[59,60] While prepubertal orchiopexy decreases the risk of testicular cancer, the incidence does not decrease to that of normal controls.[61,62] Because of this, monthly testicular self-examination is recommended and should be explained to men with a history of cryptorchidism.[63] Men with previously intra-abdominal testes, and/or cryptorchidism in conjunction with abnormal external genitalia such as hypospadias or those with an abnormal karyotype are at highest risk for cancer.[64] In a review by Wood and Elder, men with bilateral undescended testicles and in-situ intraabdominal testicles are at higher risk for tumors than those with inguinal testicles. Seminomatous tumors were more common in testicles that remained cryptorchid while those that were surgically positioned in the scrotum were more likely to develop non-seminomatous germ cell tumors.[65]

6.3.3 Post-Pubertal Cryptorchidism

Multiple studies have confirmed that cryptorchid testes in postpubertal men do not contribute to fertility and have increased risk of testicular cancer. Therefore, orchiectomy has been recommended for healthy post pubertal men between the ages of 12 and 50.[65,66,67] After age 50 the risk of death from orchiectomy exceeds the risk of death from testicular cancer.[68]

7. Vesicoureteral reflux (VUR)

7.1 Epidemiology

VUR is a common condition in children with an estimated prevalence of 0.4–1.8% in the general pediatric population and almost 30% of children who have had a febrile UTI.[69] Despite the high rate with which VUR resolves (spontaneously or by surgical correction) rare serious long-term effects

may occur as 15-30% will develop permanent renal scarring and reflux nephropathy.[70,71,72]

7.2 Presentation

VUR is most commonly diagnosed after evaluation for a febrile UTI in childhood. In older children and adults VUR may be found during evaluation of recurrent UTI, hypertension, proteinuria or voiding dysfunction.

7.3 Medical/Surgical Management and Complications

Patients with reflux nephropathy are at increased risk for hypertension (HTN), CKD and ESRD.[73] Reflux nephropathy is the fourth leading cause of chronic renal insufficiency, dialysis and pediatric renal transplantation.[74]

Despite the lack of specific follow-up protocols, the **AUA guidelines on VUR** recommend annual blood pressure monitoring, growth assessment and urinalysis to evaluate for proteinuria and UTI through adolescence in those with abnormal kidneys by ultrasound or DMSA scan. If a person with resolved VUR has a febrile UTI, evaluation for recurrent VUR and bowel and bladder dysfunction are recommended. Older patients and families should be counseled on the increased risk of HTN, CKD, recurrent UTI and increased risk of VUR in offspring.

Pre-pregnancy counseling is important in women with reflux nephropathy. Maternal morbidity is considerably higher when renal scarring is present. Increased risk of UTI and complication during pregnancy appear to be related to the presence of renal injury associated with VUR as women with normal kidneys and a history of low grade VUR are not considered at increased risk.[75,76] In a series of 87 women (175 pregnancies) with a history of VUR, 64% had pregnancy complications. During pregnancy, women with renal scarring had higher rates of hypertension (33%), proteinuria (40%), and UTI (42%) compared to women without scarring. Fetal complications including extreme prematurity (less than 32 weeks gestation), prematurity (less than 37 weeks gestation), low birth weight and intrauterine growth retardation occurred in 13% of women with a history of VUR. There was no significant difference in complications in those women with a history of VUR and renal scarring or proteinuria during pregnancy.[77] Based on a review of the literature, Hollowell found that the primary risk factor of maternal morbidity during pregnancy was the finding of renal scarring, and that this risk was independent of the presence or absence of VUR. As such, there is currently no evidence that ureteral reimplantation for low grade VUR decreases the risk of UTI during pregnancy. [75,77]

8. Solitary Kidney/Solitary Functioning Kidney

8.1 Epidemiology

Children may have a solitary functioning kidney (SFK) for a variety of reasons including unilateral renal agenesis (incidence 1 in 500-1000 births), multicystic dysplastic kidney (incidence 1:4300 births) as a result of nephrectomy, e.g., for tumor or poorly functioning kidney with recurrent UTI.[78]

8.2 Presentation

With antenatal ultrasound, most SFK are diagnosed prior to delivery. Despite the widespread use of prenatal ultrasonography, some individuals may be found to have SFK only after diagnostic imaging for urologic or non-urologic symptoms later in life.

8.3 Medical Management and Complications

People with a SFK may have glomerular damage related to hyperfiltration injury with resultant hypertension, albuminuria and even ESRD. Because of this increased risk, lifelong surveillance is recommended. [79] Data on individuals with SFK since childhood are more sobering than those who lost a kidney later in life. Studies have found that a SFK may result in renal injury before adulthood in over 50% of those affected and that 20-40% may require renal replacement therapy by age 30.[80] True risk assessment for children with SFK is made difficult by limited data – retrospective studies are limited by bias, as patients with no renal dysfunction are less likely to be seen in follow up, and prospective studies require decades of follow up.[81] Prospective and retrospective studies have identified risk factors for renal deterioration, including small renal size, and congenital anomalies of the remaining kidney or urinary tract. [78,81]

Pre-conceptional counseling of women with SFK is advised. Pregnancy normally results in glomerular hyperfiltration that may further increase the risk for kidney injury. In studies of women with a solitary functioning kidney, maternal and fetal risks including increased risk of gestational HTN, diabetes, proteinuria, and preeclampsia have been reported. [82,83]

Additionally, family members of a person with a solitary kidney may be at increased risk of a urogenital anomaly and should be counseled of this potential. [84]

9. Bladder exstrophy epispadias complex

9.1 Epidemiology

The incidence of classic bladder exstrophy in the United States is approximately 1 in 50,000 live births with a male: female ratio of approximately 2.3:1. [85] Epispadias and cloacal exstrophy are less common and seen in approximately 1:100,000 and 1:300:000 live births respectively.[86]

9.2 Presentation

Bladder exstrophy and epispadias are most often diagnosed at birth with only 10-32% detected on prenatal ultrasound.[87,88,89]

Most anomalies associated with bladder exstrophy and epispadias are surgically corrected in childhood. However, continued attention to urinary function is necessary across the lifespan. Most affected individuals will require multiple surgeries to achieve continence, optimize sexual health, and to improve cosmesis. As children mature, attention to sexual and reproductive function becomes increasingly important.

9.3 Evaluation/Follow-up

Proposed yearly follow-up should include an assessment of:

1. renal function
2. bladder cancer risk evaluating for pain, hematuria, recurrent infection and transplant status
3. urinary continence including methods of emptying, changes in continence and satisfaction with continence
4. sexual function and fertility goals
5. psychological health

Serum studies should include serum chemistries and vitamin B12 levels if patients have had ileocystoplasty or other reconstruction using bowel. Other diagnostic tests such as KUB, ultrasound, cross-sectional imaging, urodynamics and cystoscopy should be performed when clinically indicated based on changes in continence, functional status, hematuria, or increased frequency of UTI.

In addition to urologic follow-up, a multidisciplinary approach may be necessary to address behavioral and mental health, gynecologic and reproductive health, kidney health and physical function. [90]

9.4 Medical/Surgical Management and Complications

9.4.1 Renal function

Up to 40% of adults with exstrophy will have some level of renal dysfunction.[91,92,93,94] This may be related to VUR, recurrent infection or poor emptying of the reconstructed bladder, augmented bladder or bladder substitute (conduit, pouch or ureterosigmoidostomy).

9.4.2 Continence and bladder health

The exstrophied bladder will always be abnormal, regardless of how and when it was closed and/or reconstructed. Urinary continence is rarely achieved with bladder closure alone and may require bladder neck reconstruction, augmentation and/or continent diversion. Describing the steps and relative successes of these procedures is beyond the scope of this work and is poorly defined in the literature given the lack of a standardized definition of continence in this patient population.

Despite varying continence rates, attention to changes in continence is essential, as this may indicative of changes in bladder dynamics, poor catheterization technique, incomplete drainage of the augmented bladder or diversion, bladder calculus, or breakdown of the surgically created continence mechanism. A tailored evaluation should be performed including urodynamics, voiding or emptying diaries, renal bladder imaging, and/or endoscopy of the catheterizable channel or bladder.

Adults with bladder exstrophy have a risk of bladder cancer that is, at low estimate, 27 times higher than the general population. Malignancies can be of colorectal origin, from previous augmentation, or from the bladder itself, thought to be related to dysplasia from the exposed and irritated urothelium

(Smeulders N, Woodhouse CRJ. Neoplasia in adult exstrophy patients Jan 2002. BJUI 87(7):623-628). In 2008, Husmann and Rathbun reported 3/38 patients with exstrophy and an augmented bladder developed multifocal adenocarcinoma of the bladder suggesting an inherent risk of bladder cancer in this population. [95] Other complications of augment are similar to those noted in previous sections.

9.4.3 Sexual health and fertility

MALE

Boys with bladder exstrophy epispadias complex (BEEC) are born with a short, flat, broad, and upwardly tethered penis. The urethral opening in boys with isolated epispadias may be anywhere from the area of the bladder neck to the distal dorsal penis. The dorsal foreskin is insufficient or lacking. The goal of genital reconstruction is a dependent penis satisfactory for penetration. The shape, size, and chordee of the reconstructed penis may interfere with sexual function in some men. [96,97] Dorsal chordee is present in up to 49% of men.[98,99,100,101] Complications such as urethrocutaneous fistula, urethral stricture, and hypospadias may occur, affecting urinary and sexual function. Scar revision and lengthening phalloplasty may improve penile appearance in the flaccid and erect penis.[102] Radial forearm phalloplasty has also been performed to improve sexual function and quality of life in select men.[103,104]

As boys approach adolescence and adulthood there is increased concern for genital appearance and genital function.[105] Genital reconstruction in boys with BEEC is often completed early in life and may have extensive impact on the self-esteem, sexuality and body image.[106] Some small series report that up to 92% of boys with BEEC have an acceptable cosmetic result of their penile reconstruction, with higher rates for those having a Cantwell-Ransley epispadias repair.[98,107] Others report poor satisfaction with penile appearance.[100,108] Studies describing sexual satisfaction have reported variable rates of satisfaction ranging from the same as the general population to moderate sexual dysfunction. [109,110]

Abnormal ejaculation, diminished sensation, and erectile dysfunction may negatively impact sexual satisfaction.[111] Fertility may be thwarted by abnormal ejaculation and slow seminal emission. Additionally, semen parameters may be abnormal due to obstruction and recurrent infections of the genitourinary tract. Assisted reproductive techniques may become necessary. [110,111,112] Anejaculation has been reported in up to 24% of men.[96]

Anxiety may exist regarding sexual activity. Penetrative intercourse and ejaculation may adversely affect sexual satisfaction. This, in addition to the stressors associated with the disease, has led to the many to suggest inclusion of psychosocial experts as part of care teams.[113,114,115]

FEMALE

Similar to boys with BEEC, girls may have issues that adversely affect sexual and reproductive health as they grow – with concerns ranging from cosmetic to functional. Aesthetic concerns are prevalent – up to half of women with BEEC report some degree of dissatisfaction with genital appearance.[116]

Reconstruction to improve cosmesis may include vaginoplasty, re-approximation of the clitoral halves after initial reconstruction, vulvoplasty, monsplasty and rearrangement of hair-bearing skin on the lower abdominal wall for improved cosmesis. The vaginal orifice may be stenotic and require revision to allow for vaginal penetration or tampon use – in one study of 130 women with BEEC, over half required surgery for vaginal stenosis. [116] This may be accomplished with a flap vaginoplasty using a perineal skin flap.[117]

Uterine prolapse occurs more commonly and earlier in women with bladder exstrophy and is related to abnormal pelvic floor musculature and the space between the pubic bones.[85] Over 50% of women with bladder exstrophy will develop prolapse. This risk increases with pregnancy. Prophylactic suspension has been suggested by some authors.[118,119] Given the high rate of prolapse, long-term gynecologic care is recommended. Dyspareunia and anorgasmia have been reported and may be related to prior surgeries or prolapse.[96]

Maternity rates are reported to be between 25% and 68% with some requiring reproductive assistance.[96,120,121,122,123] Potential reasons for decreased fertility include functional anomalies of the Fallopian tubes or genitalia associated with surgical reconstruction. [110] Pregnancy in women with BEEC is high risk for fetus and mother and should include the support of high-risk obstetricians. Optimally, delivery would be scheduled to occur at a tertiary center with urologists available to provide assistance.[85,90,120] Cesarean section is advocated as the safest mode of delivery[124] and likely decreases the risk of both prolapse and injury to the surgically created continence mechanisms.

Videos

Continent Catheterizable Ileal Cecocystoplasty: University of Utah

Surgery: Continent Catheterizable Ileal Cecocystoplasty

Bladder Neck Placement of Artificial Urinary Sphincter

Presentations

Transitional Care Presentation 1

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