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TOPIC PAPER

The basics of transition in congenital lifelong urology

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Abstract

Purpose Transition in urology is defined by the process that allows an adolescent or a young adult with a congenital or acquired urogenital anomaly to assume increasing responsibility for their own health care and to become the primary decision maker in their care.

Methods A review of the literature regarding transitional care for lifelong urologic congenital anomalies was performed with the aim of reporting expert opinion when data are non-existent. This review focuses on special considerations for adolescents and young adults with spina bifida, bladder exstrophy, anorectal malformations and differences of sexual development.

Results Urologic goals during the transition from childhood to adulthood continue to include attention to the preservation of renal function and optimization of lower urinary tract function. Additional concerns include care to decrease long-term surgical complications (especially after augmentation cystoplasty), to monitor for malignancy, to prepare for sex activity and fertility, and to help the adult patient in decision making. Transition aims to maximize quality of life and independence by ensuring uninterrupted appropriate care through a multidisciplinary approach which varies by geographical location and healthcare setting. Barriers include patient and family factors as well as provider and system related factors. A dedicated team is an important element of successful transition.

Keywords Transitional care · Urology · Adolescent · Young adult · Spinal dysraphism · Bladder exstrophy · Spina bifida · Myelomeningocele

Abbreviations

AC	Augmentation cystoplasty
ARM	Anorectal malformations
CAKUT	Congenital anomalies of the kidney and urinary tract
BEEC	Bladder exstrophy epispadias complex
CIC	Clean intermittent catheterization
DSD	Differences of sexual development
ESRD	End stage renal disease
FU	Follow-up
LUT	Lower urinary tract
NP	Nurse practitioner
PUV	Posterior urethral valves
QoL	Quality of life
RBUS	Renal and bladder ultrasound
SB	Spina bifida
TRAQ	Transition readiness assessment questionnaire
UTI	Urinary tract infection
VPS	Ventriculoperitoneal shunt

Introduction

Improvements in therapy for children with congenital diseases have led to an increase in survival. Transition to adult care is emerging as a priority. Adolescence is a time of physical, psychological and social change. Individuals with special health care needs face additional challenges. The process of transitioning to adult services has been shown to be associated with health deterioration. The goal of the transition process is to maximize lifelong functioning through uninterrupted health care services as the patient progresses from adolescence to adulthood [1, 2].

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The genitourinary tract can be affected by birth defects and abnormalities of the central nervous system that impair bladder function throughout life. Primary urologic goals are to: (1) preserve renal function, (2) improve urinary continence, (3) prepare for sexuality and fertility, (4) monitor sequelae of neurogenic bladder or interventions, e.g., malignancy, calculi and (5) help the adult patient in decision-making.

Congenital anomalies of the kidney and urinary tract (CAKUT) that require adult care are enumerated in Table 1 [3–6]. Acquired conditions in childhood, such as spinal cord injury, may also necessitate lifelong urology care.

Goals of transition

The main goal of transition is the development of the child's ability to make their own decisions and assume responsibility for their own healthcare. Transition aims at maximizing quality of life and independence by ensuring uninterrupted and developmentally appropriate care as the patient transitions into adult life. Explicit goals in areas of independence and self-care should be part of the routine discussion with the patient and family.

Renal function

Collaboration between urologists and nephrologists is required. In posterior urethral valves (PUV), one-third will progress to end stage renal disease (ESRD) [7]. In people with spina bifida, even if the kidneys appear normal on renal and bladder ultrasound (RBUS) at birth and even if bladder management is optimized, chronic kidney disease is higher in adults when compared to unaffected individuals [8, 9]. Renal function in people with congenital urologic anomalies often deteriorates during puberty as this period is when patients are at greatest risk to being lost to medical care. In a series of children with decreased renal function from PUV, 43% began to have deterioration after puberty. Surveillance with RBUS, identification of proteinuria and high blood pressure, and assessment of renal function are therefore recommended [3, 6]. Early intervention with ACE inhibitors may delay progression of renal disease [10].

Lower urinary tract (LUT) function

LUT symptoms may affect renal function and quality of life (QoL) by increasing urinary tract infections (UTIs) and incontinence. In PUV, post-pubertal bladders tend to have decreased contractility and overdistention [11]. In individuals with myelomeningocele, bladder capacity and detrusor leak point pressure increased with puberty [12]. Men with corrected hypospadias reported LUT symptoms twice as often as controls [13]. For adults with myelomeningocele and women with bladder exstrophy epispadias complex (BEEC), decreased QoL associated with urinary incontinence has been demonstrated [14–16].

Long-term surgical complications

Children with bladder dysfunction from myelomeningocele, BEEC, and PUV will often proceed to augmentation cystoplasty (AC) [17]. Hyperchloremic metabolic acidosis is the main complication whose severity depends on length of bowel used, bladder management habits, and baseline renal function and may require supplementation with potassium citrate or sodium bicarbonate [18]. The risk of vitamin B12 deficiency increases over time despite surgical preservation of at least 15 cm of distal ileum [18, 19]. More than one third of patients will require revision surgery for bladder perforation, stomal complications, urinary stones, or bowel obstruction [20, 21]. Bladder perforation is a life-threatening complication which occurs in about 3–4% of these patients. One of the risk factors is to have lower adherence with clean intermittent catheterization (CIC) [18].

Table 1 Childhood urologic conditions with potential for long-term urologic adverse outcomes

	Renal function	Lower urinary tract function	Urologic malignancy risk	Sexual and reproductive functions
Posterior urethral valves	×	×		×
Exstrophy-epispadias	×	×	×	×
Vesicoureteral reflux	×	×		×
Prune belly	×	×	×	×
Myelomeningocele	×	×	×	×
Hypospadias		×		×
Differences of sexual development		×	×	×
Cloacal malformation	×	×	×	×
Undescended testicle			×	×

Malignancy

The association is between cryptorchidism and testicular cancer is well known with a 3.5–8 relative risk [22]. This risk is also elevated of gonadal tumors in differences of sexual development (DSD). This risk depends on the presence of a defined gonadoblastoma region on the Y chromosome [23]. Colorectal cancer risk is increased by 100-fold after ureterosigmoidostomy that mixes the urinary and fecal stream. This requires regular surveillance sigmoidoscopy beginning ten years after surgery [3, 4]. Furthermore, individuals with neurogenic bladder with or without AC are at increased risk for bladder cancer [24]. Age at presentation is younger, symptoms are less common, and cancer stage is higher. Routine screening with annual urine dipstick, RBUS and cystoscopy has not been shown to be effective [18].

Sexual / reproductive function

Ideally sex education should address: self-esteem and body image, public and private body parts, emotional and physical changes of puberty, menstruation, physical mechanics of sex and reproduction, sexually transmissible infections, safe sex practices and birth control [25].

With hypospadias and BEEC, dissatisfaction with phallic appearance is reported in 20–40% of males and is commonly related to penile size [13]. Dorsal curvature is the norm. Adults with failed hypospadias repair, BEEC, and dysraphism may be more likely to have erectile problems [26, 27]. Ejaculatory dysfunction is also prevalent and the semen is often of poor quality [5, 28, 29].

Young women with cloacal anomaly and DSD may undergo vaginal reconstruction and uterine surgeries to facilitate menstruation and to create a vagina adequate for sexual intercourse. Vaginal stenosis and pelvic organ prolapse are unfortunately common [30, 31]. The Female Sexual Function Index can evaluate domains of desire, arousal, lubrication, orgasm, satisfaction and pain, with significantly lower scores. Bladder and bowel incontinence are an obstacle to sexual function [32, 33]. Additionally, fertility can be impaired [34].

Transition process

The process of transition needs to begin early and must allow sufficient time for an individual to feel prepared. Additionally, the process may vary based on patient readiness. When approached as a quick switch, it leads to understandable feelings of abandonment [35].

Goals of transition

Transition aims to maximize health, QoL and independence by ensuring uninterrupted appropriate care. These goals require a multidisciplinary approach to improve the individual experience of health care and the health of populations and to reduce the associated costs [36]. Education of the adolescent should aim to avoid health deterioration and decrease emergency visits [37, 38].

Barriers to transition

It is not surprising that barriers to transition have been identified as variable, depending on geographical location and healthcare setting [38, 39].

Patient- and family-level barriers

Barriers may include personal preferences about care, education, self-management, and support [38]. An individual's willingness to engage in transition is the ingredient for success regardless of medical complexity [37, 40]. Not visiting the adult clinic before transition was noted as a barrier [41].

Provider-level

One of the most significant barriers is the lack of transition clinics [3, 42]. Other potential limitations include lack of formalized training opportunities in adult congenital care and lack of standardized care guidelines for patients with congenital diseases in adult life.

System-level

The complexities of issues experienced by young adults should not be underestimated and include health insurance, fragmented care, and difficultly navigating administrative bureaucracy [38, 43]. Failing to address financial needs associated with transportation, supplies, office co-pays and out of pocket expenses, as well as lost income from time away from work is also a concern [43].

Transition process

Six core elements of transition have been defined: transition readiness and planning, transfer of care, transfer completion, and transition tracking and monitoring [44]. Tools and resources associated with transition can be found at www.gottransition.org. Three additional elements have been postulated: active patient involvement, clinics must be responsive to adolescent needs, and systems to ensure attention to all aspects of chronic illness during transition [45]. Transition readiness encompasses five components: time devoted

to the process, knowing and understanding the process, level of interest in learning about transitioning, areas of concern, and acknowledgment of importance of life-long follow-up (FU). Anticipatory planning should include formally presenting the adolescent with the transitional policy at about 12 years old. However, the process should be flexible and individualized [43].

Tracking and monitoring

If the goal is to ensure that adults transition successfully, both success and failure must be precisely defined. Two studies have previously defined successful transition simply as attending a single transitional urology appointment within 18 or 24 months of the last pediatric urology appointment [37, 38]. It must be noted that despite a well-planned transition process, as few as half of patients will still be lost to FU [3, 37]. Finally, data should be collected using validated tools like the Transition Readiness Assessment Questionnaire (TRAQ) [46].

The transition team

In an ideal state, the Lead Clinician will have full knowledge and understanding of the background diagnosis, treatment received to date, ongoing problems/challenges, and the future needs. The Nurse Practitioner (NP) or similar level provider is useful and should be present at transition clinic appointments and be integral in care planning for each young person. They may serve as the first-line provider in urgent or chronic ongoing health care needs. A Transition Coordinator plays an important role in keeping transition moving ahead at an appropriate pace. This includes ensuring that appointments are made at convenient times for the young person and their family to minimize missed appointments and maximize the effectiveness of the transition process. An efficiently managed transition/transfer system is greatly enhanced by the involvement of an Administrative Assistant who works closely with the team. A Social Worker can offer enormous support and may serve to help the patient and family navigate the often-disjointed health services organizations.

Transitional care multidisciplinary teams

Spina bifida (SB)

Multidisciplinary SB transitional clinics serve as the prototype for transitional care in urology. Improvements in early medical and surgical management of neurogenic bladder, hydrocephalus, and neurosurgical closure have successfully decreased pediatric mortality.

Urologist

Urologic care begins at birth with a focus on aggressive bladder management in the interest of renal preservation, UTI prevention, and later, continence. This continues into adolescence and adult life with management of concerns around sexual function, fertility, and pelvic organ prolapse. Furthermore, urologists aim at detecting bladder cancer and at monitoring urinary diversion with its complication. Bowel program can be managed by urologists by performing antegrade enema procedure (MACE). Poor surveillance of patients can often lead to irreversible outcomes [47, 48].

Sexual health, fertility, and pregnancy

Matters surrounding sexuality, fertility, and pregnancy become more relevant as these patients age. Genetic counseling prior to consideration of parenthood is recommended for men and women. During pregnancy, bowel and bladder care can be affected and an indwelling catheter for bladder drainage may be required [49]. Ventriculoperitoneal shunt (VPS) obstruction during pregnancy has also been reported. Hydronephrosis requiring percutaneous drainage and UTIs are common [49]. An obstetrician with specialty training in high risk pregnancy should play a central role in determining which delivery method is safest. However, a urologist should be available to assist in Caesarian deliveries in a reconstructed patient.

Neurosurgeon

Many patients have hydrocephalus, Chiari II malformations, and syringomyelia. Eighty-two percent of shunted patients have sequelae of hydrocephalus including motor, cognitive, and behavioral deficits, epilepsy, vision loss, and endocrine disorders. Recognition of shunt failure is not always straightforward [50, 51].

Orthopedist, physiatrist, physical therapist

Common conditions include joint contractures, foot deformities, and hip subluxation. Progression of kyphoscoliosis can lead to restrictive lung disease. Immobility and obesity can exacerbate these conditions.

Gastrointestinal health

Eighty percent of these patients will require a bowel program [52]. Fecal incontinence is known to correlate with

poor QoL [53]. When conservative measures fail, surgeries including MACE or end colostomy may be required.

Social work

Patients commonly report difficulties with insurance, transport, finances and employment opportunities, and these can be essential elements to successful navigation of the adult health care system and patient compliance.

Nutrition

Obesity is common in non-ambulatory adults with SB and complicates nearly all aspects of care including CIC, hygiene/skin care, pulmonary function, and any surgical intervention [54]. It also increases the risk of diabetes and metabolic syndrome with the associated cardiovascular sequelae.

Wound care

Skin breakdown is a common concern for older patients with SB [55]. Immobility, obesity, insensate skin, wheelchair use, lymphedema, and urine leakage are among the contributing factors. Patients and families should be educated on skin health as it is much easier to prevent skin breakdown than to treat it. Lack of access to medical assist devices, orthotics, and physiatry services can impair timely replacement of ill-fitting braces and chair and promote wound issues.

Mental health

As children become older and develop self-awareness, the psychosocial challenges of adolescence and adulthood come to the forefront. Poor adherence with medical routine, particularly CIC, is common. Substance abuse, depression and anxiety are well described [3]. Disability itself places patients at risk for depression.

Bladder exstrophy epispadias complex, cloacal exstrophy

Orthopedics and neurosurgery

All patients with BEEC have pelvic diastasis resulting in abnormalities of the pelvic ring increasing the risk for pelvic floor prolapse. Additionally, long-term studies have suggested a higher incidence of hip and lower back pain [56]. In cloacal exstrophy, spinal anomaly and scoliosis may exist and may affect lower extremity function and mobility. Those with hydrocephalus and VPS will need long-term neurosurgical surveillance.

Reconstructive surgeons and obstetricians

The combination of a stenotic, shortened vagina and prior reconstructive surgery make pregnancy high risk [57]. Pregnancies in women with BEEC could be successful and should include specialized obstetricians [49, 57]. Caesarean section is most commonly preferred either due to maternal health concerns (e.g. prolapse, hydronephrosis, preeclampsia) or fetal concerns (e.g. breech presentation), both of which are more common in the pregnant woman with BEEC. Moreover, up to half of adult women with BEEC develop prolapse. Outcomes are reported to be best with transabdominal approaches such as sacrocolpopexy [58].

Gastroenterology

The prevalence of fecal incontinence in patients with cloacal exstrophy is difficult to assess as many will have either a permanent ostomy or are in need of a chronic bowel program if they underwent a coloanal pull-through. Parastomal hernias and issues with appliance fit often occur later in adulthood and the expertise of a good wound-ostomy care nurse may be needed. Alternatively, they can be followed in a multidisciplinary clinic that is managing patients with SB since many cloacal exstrophy patients also have SB. Patients with classic BEEC who have a urinary diversion with a ureterosigmoidostomy have a need for GI cancer surveillance and require referral to a gastroenterologist.

Mental health

There is a high rate of clinically significant anxiety disorders in female patients with BEEC and concerns about urinary continence, sexual function, body image, and medical interventions. Males have been reported to demonstrate increased externalizing behaviors [59]. Importantly, there is a higher risk for depression and suicidal behaviors in these patients. It is recommended that they be under the care of a pediatric psychologist during childhood and should be transitioned to an adult mental health provider [59].

Anorectal malformations (ARM) and cloacal anomalies

Patients with high anorectal anomalies and cloacal anomalies in females have high rates of CAKUT and spinal anomalies (vertebral anomalies, scoliosis, tethered spinal cord, sacral agenesis) leading to neurogenic bladder dysfunction [60]. Lifelong attention to bladder management and renal function is needed.

General or colorectal surgeon

Bowel incontinence remains an issue and many may ultimately opt for a permanent stoma [61]. Services of a stoma nurse specialist are important.

Gynecology and obstetrics

Females with ARM have a significant risk of abnormalities of their reproductive tract including duplication anomalies of the cervix and the uterus, vaginal septi and agenesis. Vaginoscopy in infancy may reveal anomalies. If noted, this information should be passed on to the patient and their adult providers as fertility issues may arise in post pubertal life. Some anomalies may become evident at puberty and the patient and providers should be aware of the possibility. Pregnancy is considered high risk in patients with ARM and cloacal anomalies. In those with urinary diversion or reconstruction, additional risks may be present including difficulty with CIC, recurrent UTIs and potential risks with vaginal delivery [49]. Preconception delivery planning is ideal, including a joint consultation between the obstetrician, the general surgeon, and urologist managing the patient. Delivery plans should take prior surgeries into account. Early referral to obstetricians specializing in reproduction/endocrine/infertility should be offered to all young women with ARMs.

Differences of sexual development

DSD encompasses a vast array of genital, gonadal, and chromosomal abnormalities. Multiple specialists are necessary to achieve hormonal, sexual, reproductive, and psychological health in these patients.

Urologist

The urologist addresses long-term genital and sexual function, fertility ad urinary symptoms. Urologists may work in concert with gynecologists and/or reconstructive surgeons to address gender-affirming genitoplasty in those who have not had surgery in childhood or in those presenting with gender dysphoria and seeking revision genitoplasty in later life.

Gynecologist

A gynecologist may be the first point of contact for DSD patients not diagnosed in infancy [62]. Phenotypically female patients can present later in life with amenorrhea, pubertal delay, virilization at puberty, vaginal shortening and sexual difficulties, or infertility. For older DSD patients who have not had reconstructive surgery, gynecologists may direct vaginal dilation or assist with vaginoplasty or gonadectomy. For patients who have undergone feminizing genitoplasty, long-term gynecologic complications such as vaginal stenosis requiring revision surgery may occur. Revision labioplasty or vulvoplasty may be requested to improve the cosmetic appearance of the vulva.

Endocrinologist

The role of the endocrinologist includes sex hormone supplementation and suppression, monitoring blood pressure, bone health and growth, and mineralocorticoid and glucocorticoid therapy [63]. In conjunction with surgeons and patient families, the endocrinologist also gives valuable input regarding timing of gonadectomy, gonadal function, gender correspondence, and risk of malignancy.

Fertility specialists

Genetic counseling is prudent prior to considering parenthood. Infertility in DSD can result from abnormal gonadal development, hormone imbalances, or structural problems with internal or external genitalia [64]. Fertility potential appears to be highest in patients with XX or XY congenital adrenal hyperplasia and is rare in pure or mixed gonadal dysgenesis, ovotesticular disorder, and XX males. Assisted reproductive technologies have broadened fertility options [63].

Psychologists

Psychological distress in the DSD population is more prevalent than among non-DSD peers, and early mental health support appears to decrease this distress [65]. Patients have complex body image, identity, and romantic concerns, and psychological distress arises from issues surrounding social stigmata of their diagnosis, sexual anxiety, gender dysphoria, and fertility.

Conclusion

Transition in urology remains a challenge and requiring collaboration and cooperation in a multidisciplinary clinic. Coordination and communication from all relevant stakeholders should be efficient. Conferences to share interdisciplinary knowledge and to allow discussion of complex cases should be built into the multidisciplinary clinic schedule. Team meetings are important to review successes and opportunities for improvement. A patient tracking system is recommended to monitor success, track patients who lapse from care, and seek feedback for quality improvement from patients throughout the transition process.

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