



## The Transition of Young Adults With Lifelong Urological Needs From Pediatric to Adult Services: An International Children's Continence Society Position Statement

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**Introduction:** Children with urinary tract disorders managed by teams, or individual pediatricians, urologists, nephrologists, gastroenterologists, neurologists, psychologists, and nurses at some point move from child-centered to adult-centered health systems. The actual physical change is referred to as the transfer whilst the process preceding this move constitutes transition of care. Our aims are twofold: to identify management and health-service problems related to children with congenital or acquired urological conditions who advance into adulthood and the clinical implications this has for long-term health and specialist care; and, to understand the issues facing both pediatric and adult-care clinicians and to develop a systems-approach model that meets the needs of young adults, their families and the clinicians working within adult services. **Methods:** Information was gleaned from presentations at an International Children's Continence Society meeting with collaboration from the International Continence Society, that discussed problems of transfer and transitioning such children. Several specialists attending this conference finalized this document identifying issues and highlighting ways to ease this transition and transfer of care for both patients and practitioners. **Results:** The consensus was, urological patients with congenital or other lifelong care needs, are now entering adulthood in larger numbers than previously, necessitating new planning processes for tailored transfer of management. Adult teams must become familiar with new clinical problems in multiple organ systems and anticipate issues provoked by adolescence and physical growth. During this period of transitional care the clinician or team assists young patients to build attitudes, skills and understanding of processes needed to maximize function of their urinary tract—thus taking responsibility for their own healthcare needs. Preparation must also address, negotiating adult health care systems, psychosocial, educational or vocational issues, and mental wellbeing. **Conclusions:** Transitioning and transfer of children with major congenital anomalies to clinicians potentially unfamiliar with their conditions requires improved education both for receiving doctors and children's families. Early initiation of the transition process should allow the transference to take place at appropriate times based on the child's development, and environmental and financial factors. *NeuroUrol. Urodynam.* 36:811–819, 2017. © 2016 Wiley Periodicals, Inc.

**Key words:** adolescent psychology; adolescent urology; congenital anomalies; lifelong urology; sexual development; transitional care

Abbreviations: AAFP, American Academy of Family Practitioners; AAP, American Academy of Pediatrics; ACEI, angiotensin converting enzyme inhibitors; ACP, American College of Physicians; ADHD, attention deficit hyperactivity disorder; CAKUT, congenital anomaly of the kidney or urinary tract; CDH, congenital heart disease; CF, cystic fibrosis; CIC, clean intermittent catheterization; Cr-EDTA, chromium labeled ethylene diamine tetra-acetic acid; DMSA, dimercaptosuccinic acid; eGFR, estimated glomerular filtration rate; HCG, human chorionic gonadotropin; ICCS, International Children's Continence Society; ICS, International Continence Society; LUTS, lower urinary tract symptoms; MAG-3, mercaptoacetyl triglycine –3 renal scan; SCD, sickle cell disease; SB, spina bifida; UT, urinary tract; GP, general practitioner.

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## INTRODUCTION

Children with complex health problems often have multiple organ system involvement. This is particularly true in urology where children with UT disorders are managed by teams comprised of individual pediatricians, urologists, nephrologists, gastroenterologists, neurologists, physiotherapists, psychologists, and nurses. At some point adolescents or young adults with on-going care needs are moved from child-centered to adult-centered health systems. Ideally, this process takes into account physical and emotional issues associated with passage through adolescence.

Long term care of any chronic childhood disease has major financial implications.<sup>1</sup> There are no figures to show how many “adolescent urologists” are required to adequately care for young people with lifelong urological needs. One estimate suggests a total population of about four million would provide enough work for one such urologist. This figure must be treated cautiously since the incidence of major anomalies is not accurately known and selective pregnancy terminations have become commonplace, reducing the incidence of individuals with complex care needs. This is juxtaposed against children with complex conditions who are living longer and well into adulthood.

Our aims are twofold: to identify health-service problems related to children with congenital or acquired urological conditions who advance into adulthood and the clinical implications this has for long-term health and specialist care; and, to develop a systems-approach model that meets the needs of older adolescents and young adults, their families and clinicians working within adult services. Delivery of clinical care to adolescents with urological needs is not the focus of this manuscript; however, Appendix 1 details specific multi-system care requirements.

## METHODS

A group of clinicians, allied health professionals, and researchers from a variety of specialties and parent representatives, gathered in Toronto in June, 2013 at an ICCS meeting in collaboration with the ICS, to discuss and cross-pollinate ideas related to the problems of transfer and transitioning children with congenital urologic issues into adult services. Experience in the small number of existing adolescent urology programs and plans for development in different health care systems were reviewed. This manuscript results from contributions of several of those in attendance and provides a framework identifying the needs of patients, the limitations of adult providers and the barriers for transferring care from a pediatric setting to an adult facility.

## RESULTS

The document summarizes how children receive care as youngsters and what they and their parents perceive as stumbling blocks to transferring their care from pediatricians and pediatric specialists to adult providers who may have limited experience with these conditions. Models for identifying and improving delivery of care, overcoming barriers that exist, including health systems and current fiscal policies are proposed, with the final segment outlining timetables for implementation, including targets for achieving successful outcomes.

### Patients

Children with chronic illness and their families are accustomed to receiving holistic care in a pediatric setting. There is

specific focus on the anomaly, but education, social care, family support and other aspects of management are routinely addressed. The service model can be likened to a hub and spokes, where the child sits at the convergence of various disciplines.

The focus of transitional care involves moving from one service to another, but it must be underpinned by acknowledging the changing developmental needs of adolescents and young adults. Adolescent transition could be defined as: “a purposeful, planned process that addresses medical, psychosocial, educational and vocational needs of adolescents and young adults—learning to live with their lifelong health condition”.<sup>2</sup> Pediatricians should start preparing their patients and parents (caregivers) for this transfer of care at an early stage so ultimately young patients become effective partners in their own transition. General guidelines already exist, but remain to be validated. Once in a new environment, the young person is able to accept responsibility for his/her own disease management as parents “step away” from overseeing care. This has been found to be a complex process, particularly as parents and children are apprehensive about change.<sup>1,2</sup> The child is gradually empowered to become the decision maker and supported, especially if conflicts arise in decision making which differ from those of his (her) parents.

Young people with chronic illness are significantly more likely than healthy peers to develop emotional issues requiring psychological support. The psychological difficulties with which young people present are intimately related to their physical condition and often their genital structures. By definition, the primary anomaly involves the genitourinary tract so the most consistent follow-up in adolescence and adulthood will be with urology. The challenge for urologists is they may not be the best specialists to act as “team” leader. Nonetheless, in practice they have often had to adopt this role, directing these individuals to appropriate colleagues to attain maximum support for their overall health. The specialty of adolescent medicine has emerged in several countries to fill this void for patients from puberty to a set termination in late adolescence or young adulthood (ranging from 19 to 25 years), but without an equivalent adult counterpart. Young people often worry about their most intimate body parts, about how they look and function. Such worries can lead to withdrawal, low self-esteem, depressed moods and anxiety. In certain situations, limited mobility has repressive effects on their ability to feel equal to peers.

For many, these worries only surface as the reality of a future with limited potential becomes tangible. From mid to late adolescence the reality of leaving home and developing independent lives can impact overall adjustment and well-being. Many childhood services provide a degree of psychological support; however, adult services often deal only with urological issues encountered by older individuals who already have support systems and relationships in place. Thus, the transition process frequently results in young people moving to services that are unprepared, under-resourced, and unable to offer the necessary care for a range of unplanned needs.

### Continuity of Care Barriers

The four most important parties to successful transition—the patient, their family, their pediatric specialist and adult consultant—may actually serve as barriers. Table I outlines the main issues faced by young people and their families when moving from family based care to independent management. This transition occurs at a critical time coinciding with other major life changing events, specifically, establishing independence and

**TABLE I. Issues Faced By the Young Person and Their Family When Moving From a Family Based Care Structure to Independent Management**

The young person	<p>Transition coincides with other major life changes</p> <p>Greater opportunity to participate in risk taking behavior</p> <p>Complete neurodevelopment may be limited by underlying condition</p> <p>May have poor health literacy → difficulty implementing treatment regimens</p> <p>Not likely to prioritize care for medical condition</p> <p>May not be ready to assume responsibility for good urological care</p> <p>Many sources of potentially conflicting information</p> <p>Often geographically distant from clinician and medical team</p>
The family	<p>Parents may no longer have a direct relationship with the clinician</p> <p>Limited recognition of legal status within the doctor-patient relationship</p> <p>Dancing between independence and responsibility for young person</p> <p>Conflict between care-giving and promoting young person's autonomy</p> <p>Advocating for young person versus impeding confidentiality</p> <p>Guilt feelings of responsibility for their child's condition → over-protectiveness</p>

autonomy, potential changes in living arrangements and moving from full time-education to work or higher education. Neurodevelopment is often not complete until the third decade,<sup>3</sup> meaning risk-taking behavior is common.<sup>4</sup> Patients with chronic disease commonly prioritize “normal” behaviors and body image above appropriate care for their medical condition.

Transition presents difficult challenges to the family as well. During childhood, parents acting as primary stewards have a direct relationship with clinicians. As their child moves into adulthood they find their position relegated to less prominent roles. In many jurisdictions there is limited recognition of their legal status within the doctor-patient relationship. Parents face an obligatory transfer of care for their child from a unit with whom they have established trust and understanding. Families face conflicts between care-giving and advocating for their family member versus potentially impeding a patient's developing autonomy and violating patient confidentiality.<sup>5</sup> Parents recognize that the young person is becoming more independent but may have difficulty relinquishing responsibility for oversight of care, particularly when adult teams and safety net practices are poorly understood or unknown. Occasionally, the medical condition requires increased levels of intervention at this time. As such families and young people, are often keen to postpone transfer to unknown adult services.

Table II lists areas of difficulty experienced by both pediatric and adulthood physicians/surgeons. Whilst many issues relate to systems-based communication, other problems directly result from specialized education targeting only adults.

Inertia in handing over such patients to adult services is a key factor, particularly if there are unresolved therapeutic problems. The preparation necessary for transition is onerous.

The effective transfer of such patients requires much more work than simply discharging the individual to an adult service. Even if the transfer entails only a letter of referral, it is a nearly impossible to encapsulate many years of care into a few descriptive pages. Unfortunately, pediatric clinicians are frequently unaware of all documentation required for adequate transition<sup>6,7</sup> and the process is often initiated too late.

There are many obstacles to achieving good care facing clinicians who initiate moving patients from a pediatric service. When patients have particularly difficult problems, their care may be transferred to someone with whom they may have little or no relationship, resulting in decisions being made before trust has been established.<sup>6</sup> Paradoxically, good relationships with pediatric teams often foster reluctance by patients, families and pediatricians, to “let-go”.<sup>7-9</sup>

When medical records are unavailable, teams may be unaware of relevant past history while facing difficult clinical problems. Given scarcity of expertise in managing childhood urological conditions in adulthood,<sup>10</sup> adult clinicians may, but often don't concentrate in areas where they can access skills of other specialists to help care for patients with multi-organ system conditions. Given the paucity of adult medical specialists involved with multi-system, complex care patients, there may be no equivalent general provider who can oversee these patients' needs; thus, patients and families must seek out someone who can assume this role for them.

Sub-specialisation in post-graduate medical training is effectively separating paediatric from adult urology practice. Adult urologists are, therefore, unable to assume care of complex paediatric problems of which they have little knowledge. The numbers of children with complex childhood conditions is still quite small; thus, unless a sub-specialty is developed each standard adult urologist will only see one or two cases in their career. This is neither consistent with delivery of good care or with adequate feed back to paediatric urologists regarding outcomes of their management.

#### Models of Transitioning Young Adults From Child to Adult Services in Other Specialties

It is obvious young adults with urological conditions need appropriate and effective transition. However, the process is often poorly managed and too focused on service transition and transfer.<sup>11</sup> There is a need to improve transition so it becomes a holistic process of moving to adulthood and independence.

The transition period occurs during high-risk times for young adults. Whereas successful transition empowers independence in young adults and the ability to manage their disease-specific condition, failed transition may negatively affect health outcomes, leading to increased hospital admissions and non-adherence to treatment recommendations.<sup>12</sup>

Surrogate markers evaluating efficacy of transition services include patients lost to follow-up, inappropriate admissions and medication non-adherence rates.<sup>13</sup> One example is the impact of transition of glycemic control and complications in diabetics, using hospital data.<sup>13-15</sup> Decreases in diabetes care visits were noted following structured transition when specific patient education programs, and a transition care coordinator with oversight at joint pediatric/adult clinics was in attendance.<sup>16</sup> Young people surviving malignant disease in childhood have been described as “the lost tribe,” with an increased risk of loss of follow-up. This is attributable to wide variability in models of transition, if transitioning occurs at all.<sup>17-19</sup> In patients with SCD, older age at the time of transition and a greater travel distance to new services were associated with unsuccessful transitioning.<sup>19</sup> Similar difficulties have been observed in other chronic illnesses, particularly ADHD.<sup>11</sup> Without improved continuity of care within adult services and adherence to medication, adolescents and young adults are at greater risk of academic, social and vocational difficulties, as well as behavioral problems.<sup>11</sup>

Many medical societies suggest developing transitioning programs with combined schedules, printed and web-based

TABLE II. Healthcare System Barriers to Seamless Transition of Adolescents With On-Gong Urological Needs From Childhood Services to Adult Management

Pediatric team	<p>Long relationship with patient and understanding of developmental care</p> <p>Responsible for training of young person to self-report, evaluate condition and articulate needs</p> <p>May have paternalistic approach → the patient and family reluctant to “letting-go”</p> <p>If young people are not engaged in self-management, likely to have poor treatment compliance</p> <p>May have limited contact with adult primary provider and be unfamiliar with community resources</p> <p>May not be competent with issues of confidentiality and privacy</p> <p>Must take responsibility for effective transfer</p> <p>May not view transition as a shared responsibility</p>
Adult physician/ surgeon	<p>Large patient load; adolescents with complex needs form small portion</p> <p>Time required; trust takes upwards of four visits to establish</p> <p>May lack all relevant previous clinical information</p> <p>May have minimal expertise in managing childhood conditions in adulthood</p> <p>Lack of focused training on adolescent healthcare</p> <p>May expect collaborative and reactive relationship with young people who have a history of care-giver directed and family oriented consultations</p> <p>May have poor tolerance for health risk behaviors and immaturity</p> <p>May not recognize a young person's unmet psychosocial needs that influence self-management</p> <p>May not be familiar with overlay of learning difficulties or mental health issues associated with complex needs</p> <p>Care for various needs may be fragmented—urologist may need to compensate for lack of this comprehensive support</p> <p>Will need to discuss sexual issues, including fertility and safe sex practices</p> <p>May not consult from a youth-friendly service</p> <p>May be disincentivised by lack of insurance payment/reimbursement</p>

materials, and transition-dedicated physicians, nurses and psychologists. Disease specific pioneering programs in place already, include CF, developmental disability, diabetes and CHD.<sup>20–23</sup> In 2011 the AAP, AAFP, and ACP jointly authored a clinical report providing an algorithm to implementing “best practices” for transitioning patients both with and without special health care needs.<sup>24</sup> No single model will suit all clinical conditions and flexibility is required.

Transition should begin early in life and continue over a period of years. Employing a dedicated coordinator who guides a young adult and his/her family optimizes collaboration between childhood and adult services. No patient should be transferred to an adult unit without having discussed a plan. Transfer settings should be age and developmental specific, to accommodate individual variation. Although 16 years is generally viewed as an appropriate time to start preparing for transfer, many studies suggest the actual time of transfer should be decided upon by considering age and readiness.<sup>25</sup> Reports describe cultural differences between pediatric and adult care as responsible for significant barriers to continuity of care. Close collaboration between children's and adult services is crucial to bridging the gap and for successful transition planning. Cohort studies with comparison groups involving CF showed clinic attendance was improved in patients who were introduced to the adult specialists before transition.<sup>26</sup> Patients showed improved resilience when they had a good network of friends and other psychosocial support.

Lugasi describes issues which facilitate transition: (i) presenting transition positively, (ii) revealing and adjusting young adults' expectations about the transition process, (iii) empowering patients in early teenage years to become increasingly autonomous, (iv) involving young adults in planning and preparations for transition, and (v) having a specific person coordinating the transition process.<sup>27</sup>

Recently, transition of young patients with congenital heart disease from the perspectives of all stakeholders offered practical recommendations for developing transition programs.<sup>5,28</sup> Findings supported the need for multidisciplinary teams (nurses, psychologists, social workers, and transition coordinators) to address transition-related concerns and barriers. Cost benefits accrue from successful transfer while

poor outcomes are reduced (e.g., decreased morbidity and less emergent medical care). Inadequate transition may result in multiple medical, psychosocial, work-related and health insurance difficulties.

#### Health Service and Fiscal Implications

The question that arises regarding transition of any young person with lifelong urological needs is point of care, for example, identifying an ideal place to care for patients in their early adult years. Specifically, should transition be seen in a disease or age-specific context?

Support for chronic health conditions in childhood, for example, cardiac congenital anomalies, being retained indefinitely within pediatric facilities hinges on technical expertise of the pediatric healthcare workers and specialists as well as established trust and comfort for the patient and family.<sup>29</sup> In the United States, it is estimated approximately 500,000 “youths with special needs” reach 18 every year<sup>30</sup>; an estimated 4.5 million or 18.4% of the American population aged 12–18 years have special health care needs.<sup>31</sup> Over the last decade the number of adults (>18 years of age) with childhood conditions admitted to children's hospitals has increased. Such adults have disproportionately higher resource utilization, as shown by longer hospital stays and higher charges compared to pediatric counterparts.<sup>32</sup> Caring indefinitely for childhood chronic conditions, in pediatric settings is unsustainable.<sup>33,34</sup>

Using adult SB populations as examples, securing resources to create a successful adult program is challenging. Difficulties include engaging policy makers on behalf of patients who lack self-advocacy.<sup>34</sup> Significant burdens of healthcare costs with unplanned hospital admissions has been the basis for negotiating funding for establishing support teams for adult SB patients. For single-payer public healthcare systems (e.g., Canada, United Kingdom), there are significant challenges in securing adequate funding for care of adolescents with urological multi-system needs over the course of their adult lives.

In multiple-payer systems (e.g., US), the process is also daunting. The AAP has issued specific recommendations about the needs for transition services and financing of pediatric care, stating that youths with special healthcare needs should

receive high-quality, developmentally appropriate health services that continue in an uninterrupted fashion from adolescence to 26 years of age.<sup>35,36</sup> However, AAP advocacy that the ideal age to begin transition at 18–21 years, is too late to insure complete continuity of coverage.<sup>37</sup> In one study 20.8% of young adults between 19–23 years with special health care needs were uninsured.<sup>38</sup> Only 35% of practitioners discussed implications relating to insurance coverage continuity while shifting to an adult health care provider.<sup>31</sup> With gradual and eventual full implementation of the Affordable Health Care Act, most young Americans will have acceptable health care coverage, which should continue lifelong, and not end at 26 years old.

In summary, regardless of the model (single or multiple, public/private payer) and notwithstanding the experience gained over the last decade in creating successful transition programs, from a health systems perspective, the process is still evolving. Learning from examples of adult SB population, recommendations can be summarized as: (i) all young adults should be transitioned to adult health care providers; (ii) investment in large networks of support services will lead to fewer hospitalizations; (iii) young adults benefit from multi-disciplinary diagnostic care resulting in prevention or early management of severe complications that are associated with substantial morbidity and high costs; (iv) since patients require lifelong health care services after transition, access to and maintenance of adequate insurance coverage must be integral to the transition process.

#### Setting Targets for the Transition of Young Adults With Lifelong Urological Needs

Current management of young adults with on-going urological care needs is often two dimensional, where the clinician and the patient interact. In reality, a third and fourth dimension are needed. We propose adopting a model that integrates the patient's family and the adult care team along a continuum that places over-riding importance on building readiness skills for transitioning children from an early age. This process facilitates transition and ultimate transfer of care from family centered, developmental models (paediatrics) to individual-centered, problem-based ones (adults).<sup>28,29</sup> It must be remembered that adult physicians, and particularly urologists, rarely have had comprehensive paediatric training. It is essential, therefore, patients and their families be introduced to the realities of adult medical life before leaving the child-centered environment. This necessity reinforces the need for a transition process that includes paediatric and adult input.

Planning for care transition and eventual transfer is a tailored process involving facilitating self-management and independence in the young person. From 8 years onward pediatric teams can incorporate opportunities for young patients to build attitudes, skills and understanding of processes needed to optimize their urinary tract function. This takes the form of systematically building skills itemized in a readiness checklist. For example, a child learns to articulate his/her illness to someone else and to understand what sort of treatment is needed to stay healthy. Whilst such checklists can be re-visited semi-annually by allied health team members, clinical consultations incorporate opportunities for children to practice both self-reporting their condition and framing questions related to their care. From early adolescence onward, many clinicians should include short periods of time alone with patients; this builds independence skills and provides opportunity for patients to raise confidential issues.

Addressing needs of all parties triangularly during childhood incorporates the role played by families of young people with on-going care needs. Making the young person, not their disorder, the centre of transition assists families in progressively relinquishing certain tasks and responsibilities. The young person must ultimately become the "CEO" of his/her own condition; in reality, the family is likely to become a lifelong "second in command" player. Childhood clinics could incorporate programs that build expectations in parents so handing over of care is normal and a positive progression for best long-term management. This begins with monitoring the child as he/she begins to assume some responsibilities that carry through to adult clinics providing surveillance and intervention. Parents play a pivotal role in preparing their children for assuming whatever level of self-care their condition supports.

Adolescent medicine incorporates all the following: psychosocial, educational and vocational issues, risky behaviors, mental wellbeing, sexual function and support structures. Whether within pediatric or adult systems, at some stage the care team is responsible for addressing these factors and their ramifications for a young person's condition throughout normal adolescent milestones. Since these issues extend into adulthood we propose adult teams build expertise in this aspect of adolescent care. Linkage between chronic disease populations can create opportunities for young persons to access meaningful peer groups and professional support for the full spectrum of pertinent issues.

Ample evidence exists that care delivered jointly by members of pediatric and adult teams can be targeted to meet expectations and needs of the young person. Age appropriate environment, optimal access by public transport, flexible consultation hours, use of telemedicine and electronic communication are all-important issues. A pertinent question is whether these services should constitute a separate transition clinic. It is difficult enough for patients to leave an all-enveloping paediatric environment without the additional problems of moving on again from transition specialists to adult clinics after three or 4 years.

Individual specialties have a mandate to improve transfer of knowledge and skills from pediatric/developmental clinicians to adult care providers. Improving survival in chronic conditions with urological components must contain changes in specialist educational practices. Whilst joint clinics offer some progress towards this goal, the sub-specialty of adequately trained adolescent urologists must be encouraged. Experience in performing procedures should pre-date involvement in decision-making regarding health-care issues.

Along with joint care must come full access to a young person's complete medical records and individualized case discussion with childhood providers. Given that adult care relies heavily on community-based primary practitioners, teams should ensure that GPs have ready access to relevant information about a young person's condition and care plan. Investment by young people in understanding and advocating for their management, and red flag events or routine requirements causing them to seek care, is paramount.

The outcomes of paediatric management are usually reported with follow up data only to the end of childhood. From patients' perspectives this likely represents less than a quarter of his or her life. To appropriately measure adult outcomes at least another 20 years of data collection is needed. Without this, paediatric specialists may persist in management protocols that are less than optimal or lead to revisions without adequate supporting evidence. The essential role for long-term follow up is to collect such data and feed it back to paediatric

providers. This is unlikely to happen without subspecialty development for lifelong follow-up.

### SUMMARY

There is a pressing need to improve delivery of clinical services to adolescents and young adults with lifelong urological care needs. Research is urgently needed to provide evidence for the most effective change strategies. Investment in disease-specific health literacy and early preparation for the continuum of ongoing care will likely optimize overall patient's wellbeing. Effective collaboration between sectors of health-care, with particular emphasis on communication and access to medical records, is paramount. These changes are likely to improve both quality of life and health parameters in affected young people.

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## Appendix 1

### Care Requirements in Adolescence and Adulthood

**Renal care.** The safety of the upper urinary tract is paramount throughout pediatric and adult care for any patient who has CAKUT, and or who has undergone reconstruction. The major risks can be classified as:

1. Primary renal failure as part of an predicted deterioration.
2. Primary renal failure that is unpredictable.
3. Obstructive nephropathy.
4. Renal failure from other causes, for example, recurrent UTIs.

As part of a patient's transition into an adult care setting a baseline must be re-established. Overall function is important as increased body mass may lead to rising creatinine levels, necessitating a new baseline. eGFR is not reliable when bowel has been incorporated into urinary tract reconstruction; therefore, Cr-EDTA GFR is more appropriate. Split function tests are helpful—either DMSA, if a static picture is required, or MAG-3, if outflow obstruction is suspected.

A joint uro-nephrology clinic is ideal as it facilitates a common dialogue for care between clinicians. Patients require ultrasound surveillance, identification of proteinuria and annual review of blood pressure and renal function. Deteriorating renal function or worsening hydronephrosis should be investigated to exclude obstruction.<sup>10</sup> If new or worsening proteinuria is identified, renal protective therapy should be initiated to slow the decline<sup>39</sup>; ACEI is effective treatment.

The most difficult management occurs when a gradual deterioration in renal function occurs without a clear nephrological or urological cause. This may be attributed to recurrent UTIs, indicative of an underlying problem with upper urinary tract drainage resulting from critical bladder volume and pressure.<sup>40</sup>

In the transition phase where responsibility for care, intervention and decision-making will shift from parents to patient, the importance of maintaining clinic attendance and compliance with treatment recommendations must be emphasized. A young adult experiences many social changes: a shift to university or working life, independent living arrangements, new relationships and increased personal/fiscal responsibility. It is easy to imagine why someone who is asymptomatic would chose to delay visiting a urologist or nephrologist for routine surveillance, unless adequately informed of consequences of neglecting follow-up.

**Urological care.** In the care of complex congenital conditions urological and nephrological management needs to be cohesive to be effective.

The urethra: Many pediatric urologists recommend boys with hypospadias be seen during puberty. Important issues include what is “normal” voiding function—although less efficient in hypospadiac compared to unaffected men, few data exist to

define normal, in operated post-pubertal men. The most common complaints are spraying and post-micturition dribbling. Rynja et al have described LUTS in up to 38.7% and spraying in 43.4% of males with a history of corrected severe hypospadias.<sup>41</sup>

Adolescents with exstrophy and epispadias may incur or have ongoing difficulties with incontinence or bladder emptying, upwards of 80%, primarily related to urethral incompetence. Following bladder neck reconstruction there will inevitably be an element of fixed resistance. Poor emptying may be managed with urethral self-CIC. Incontinence due to sphincteric failure will likely require creation of a continent urinary diversion with closure of the bladder neck and formation of a Mitrofanoff or Monti channel for CIC. For some, bulking agents injected at the bladder neck, a simple outpatient procedure, provides an attractive alternative; however, it may produce only temporary relief of incontinence. Since both complete voiding and continence are more tenuous in adult life, and bladder function may deteriorate over time, urodynamics are essential before and after any endoscopic procedure.<sup>42</sup> It is important that any endoscopy be performed with an appropriate sized cystoscope, as dilation to allow insertion of an adult scope will likely cause damage.

Young adult males with posterior urethral valves may present challenges. They often feel well and don't perceive a problem, thus limiting engagement with the medical team. Renal, bladder and reproductive function are all difficult to predict necessitating surveillance through early adulthood and beyond. It is not uncommon for adolescents and young adults with prior PUV to progress to renal failure requiring transplantation, making surveillance mandatory.

The bladder: Bladder reconstruction in childhood creates a lifelong contract between the patient and his/her medical team. Long-term management involves surveillance of functional, symptomatic and metabolic/renal status.<sup>43</sup> Patients must be under the care of a multidisciplinary team capable of providing appropriate understanding, advice and support for potential concerns. It is common for issues like CIC and regimented bladder emptying to engender resistance in adolescents.

A full understanding of the reconstructed anatomy is paramount, particularly ureteric reconfiguration and bowel segment employment. Although no statistical difference exists regarding risk of malignancy compared to the normal population, augmentation cystoplasty, particularly stomach or intestinal segments, is associated with higher than expected risks of malignant transformation.<sup>44,45</sup> Other factors that may lead to malignant transformation include treatment related (immunosuppression) or linkage to underlying diagnosis (exstrophy or neurogenic bladder), or be secondary to inadequately treated chronic bacteriuria. Unfortunately, these cancers tend to be aggressive in nature and late in presentation.<sup>10</sup>

The consensus view is despite its very low detection rate, annual “malignancy screening” cystoscopy is inappropriate for both patients and healthcare providers.<sup>46–48</sup> Patients who have any change in symptoms, for example, new onset of infections, hematuria, abdominal bloating, or pain should seek advice,

imaging and endoscopic investigation. It is sensible to create a protocol for follow-up that will include ultrasonic imaging of the reservoir (full and empty) and the upper urinary tract. Metabolic assessment should include creatinine, urea and electrolytes, bicarbonate, chloride and vitamin B12 levels. Sometimes, it is difficult excluding use of the terminal ileum in the reconstructive surgery; not measuring (or not detecting) the irreversible neurologic consequences of subsequent low B12 levels, is indefensible in these patients.

It is important that patients and care providers outside a specialist centre (e.g., family practitioners) understand there is little adaptation of the bowel when it has been added to the genitourinary tract, that mucus is still produced and problematic, and metabolic consequences may be significant, necessitating life-long timely surveillance and possible reoperation. Patients with Mitrofanoff or Monti channels are commonly frustrated by leakage of even small amounts of mucus onto the skin, even though it is not urinary incontinence; there are comfortable dressings available to protect clothing and avoid embarrassment. When urine is stored in an intestinal segment, urine pregnancy tests often produce a false positive result (>50%), making serum HCG assay essential to confirm or rule out pregnancy.<sup>49,50</sup>

Bacteriuria is classified as symptomatic or asymptomatic, the former often do not need systemic treatment, although increased fluid intake and more frequent emptying are warranted. Symptoms such as fever, malaise or increased incontinence merit early evaluation and culture to allow tailored antibiotic management.<sup>51</sup>

**Andrological and gynaecological care.** It is completely normal for all adolescents to question themselves and others about sexual and reproductive function, and to seek clarification regarding the impact their condition and its treatment will have on this. Few prospective data have been collected about sexual development in the context of congenital urological anomalies.

**Genital function:** In males any condition involving penile reconstruction needs to be reassessed after puberty, given this period of maximal growth. This entails questioning sensation, erectile ability, orgasm and ejaculation. Assessment for chordee in men who have exstrophy or had hypospadias surgery as children is needed as complications occur late, with some data suggesting an incidence approaching 25%.<sup>52</sup> Standardized reporting to clarify differences between proximal and distal hypospadias is needed.

Female sexual function also generates controversy when surgery is discussed. Outcome data for surgery are lacking. Some highly publicized reports have initiated a debate about early surgery, evaluating outcomes, questioning necessity of correction in infancy and potential benefits (or disadvantages) of early surgery.<sup>53,54</sup> This ultimately relates to potentially balancing parental wishes with patient consent for elective procedures. It is important that decisions surrounding genital surgery in all these patients, regardless of their age, be made as part of a multi-disciplinary team to ensure all options and ramifications have been considered.<sup>55</sup>

By adolescence and early adulthood it is important to ensure a conduit for menstrual flow, an introitus and vagina for sexual intercourse and normal sensation to allow sexual pleasure. In exstrophy, vaginal prolapse may be a concern that is difficult to treat.<sup>56</sup> In cloaca patients the incidence of internal genitalia abnormalities is high; a thorough investigation in all adolescent girls avoids acute abdominal problems.

Timely bladder emptying with catheterization, and evacuating the bowels with enemas or suppositories, will increase the

likelihood of “accident-free” sex. After intercourse, females must again empty their bladder to decrease risk of UTI. If substantial urinary leakage occurs during arousal or frequent UTIs afterwards, urinary retention should be determined.

Since sexual arousal may not always induce vaginal lubrication, a water-soluble lubricant is helpful. Some women have difficulty with intercourse due to physical restrictions from orthopedic deformities, joint mal-alignments and/or muscle atrophy, that affects movement of hips and legs, and thus need counseling about ways to improve their ability to achieve satisfactory sexual relations.

**Fertility:** In the largest reported series of women with exstrophy, 81% were sexually active but only 21% experienced spontaneous conception within a year.<sup>57</sup> Fertility potential in all young people with on-going care needs guides early treatment decisions. Advice and decisions must be based on diagnosis, treatment and the individual. As with many areas of patient care, expertise and support from within a multidisciplinary team is vital.

**Contraception:** Women with SB have similar options for birth control as the general population. Based on available information, a woman’s SB level does not dictate contraception choices, including birth control pills. Factors like age, smoking status, mobility, presence of uterine abnormalities (e.g., unicornuate uterus is more common in bladder exstrophy) and personal or family history of thrombo-embolism do impact on the risk of some birth control choices. If osteopenia or osteoporosis is present, Depo Provera should be avoided. Similarly, women on antiepileptics have limitations for using low dose combination hormonal drugs and progesterone only pills.

Due to the high incidence of latex allergy in SB, latex free condoms are mandatory unless the patient has tested negative for latex allergy. In men with epispadias, standard condoms are usually too large; made-to-measure condoms are available online.<sup>58</sup>

**Obstetric care.** Many young women with SB and/or hydrocephalus are able to become pregnant. The risk of having similarly affected offspring varies between 3% and 8%.<sup>59</sup> Taking folic acid can reduce this risk by about 70%. Advice from genetic counselors regarding these concerns should be sought. Prior to contemplating pregnancy current medications need reviewing, particularly those taken for bladder management, hypertension and seizure control, to ensure their safety during gestation. Because half of all pregnancies are unplanned, all sexually active woman with SB should take between 4 and 5 milligrams of folic acid supplementation daily before becoming pregnant, and throughout their first trimester.

Denervation involving pelvic nerves and muscles creates challenges for women with SB not applicable to the general population. The presence of neurosurgery and urology will help ensure the pregnancy is managed in a manner that does not adversely affect a shunt or renal function, or vice versa. Planning for delivery depends on multiple physical factors, including sensory level in cases of spinal cord dysfunction, ability to push using abdomino-pelvic muscles, size of the pelvis, and flexibility of the hips and knees.<sup>60</sup> The more conducive these factors are to the normal birthing process, the more likely a vaginal delivery will be possible.

If a safe vaginal delivery is not feasible, Cesarean section is recommended. The obstetrician should confer with the urologist regarding a woman’s current urinary tract anatomy, if she has had reconstructive surgery, to ensure a safe delivery. Having urology present or in abeyance when women with lower urinary tract reconstruction undergo a Cesarean section is obligatory. Women with SB have successfully and safely received epidural anesthesia for their delivery although



ultrasound guidance is needed to visualize spinal anatomy during placement. Successful epidurals have been conducted for most myelomeningocele lesion levels, with or without scoliosis. Due to altered anatomy these epidurals are trickier than in normal females; thus, it is best to arrange delivery at a facility experienced in high-risk pregnancies, where urology and anesthesiology are available.

Women with bladder exstrophy have a median gestation at delivery of 37 weeks, with 26% occurring before 37 weeks, and most have a planned Cesarean performed with a general surgeon or urologist present. Dean reported 12% of women experienced major birth-related complications, for example, ureteric transection, vesico-vaginal or urethra-vaginal fistula and postpartum hemorrhage.<sup>57</sup>