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European Association of Urology

Review – Reconstructive Urology

Lifelong Congenital Urology: The Challenges for Patients and Surgeons

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Abstract

Context: Patients born with complex congenital genitourinary anomalies (including bladder exstrophy, cloacal exstrophy, epispadias, neurogenic bladder, hypospadias and posterior urethral valves) often require major reconstructive surgery in childhood. These conditions, their treatment and sequelae require lifelong follow-up. This has created the need for adult urologists to provide care as these patients grow into adults.
Objective: To evaluate current strategies for transition and provide a current position statement with examples of the challenges faced by patients and their health care teams as a result of these conditions and their treatment.
Evidence acquisition: Each of the authors was asked to provide a 500-word synthesis, based on current literature; to highlight the challenges faced in an area of their expertise.
Evidence synthesis: The authors assembled in March 2018 to form a consensus based on the data gathered. The aforementioned sections were reviewed and following the consensus discussion the paper was formulated and reviewed.
Conclusions: Lifelong care of congenital problems is challenging and essential for many but not all. Expertise is needed to provide the best care for patients and make the best use of resources. Specialist centres appear to be the most effective and safe model. In the long term it would be ideal to establish an evidence base focused on the common long-term problems with these conditions to ensure excellent care with appropriate expertise.
Patient summary: Patients born with complex congenital anomalies of the genitourinary system require specialist care in childhood. Many will need lifelong care to manage their condition and the treatment of it. There is growing interest in this area of medicine and this consensus statement addresses the need for lifelong care in this group. The aim is to ensure that all patients that need care at any age are able to find what they need.

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1. Introduction

We are among the first generation of surgeons and patients to see the long-term implications of congenital urogenital anomalies and the consequences of surgery to improve them. Current data are poor, and the potential for improvement lies in better measures of outcome, standardization, and patient satisfaction. Patients who have been treated under the care of paediatric urologists in the past 40 yr are now appearing with long-term problems. There is a lack of expertise in this area and a rudimentary understanding of outcomes. The conditions and patients can be medically, surgically, and psychologically challenging, with urological, nephrological, gynaecological, orthopaedic, and psychological issues. Unfamiliar practitioners will unsettle patients, jeopardizing the doctor-patient relationship and increasing the risk that patients will drop out of care [1]. Patients need guidance and structure about who should look after them in their adult life. In a recent survey of European paediatric urologists, almost 300 responded and estimated that between 10% and 20% of their treated patients will need long-term follow-up, and numbers of patients are expanding [2].

Our consensus process had three clear aims:

1. To recognize and state the importance of specialized long-term follow-up for patients born with and treated for congenital urological anomalies.
2. To educate practitioners about the challenges that arise in these complex patients.
3. To attract greater interest from adult urologists in this fascinating field of urology.

2. Data acquisition

This consensus was commissioned and supported by the European Association of Urology (EAU). All authors were selected on the basis of experience in this field and from a range of areas and health systems across Europe. A framework of topics was designed by the chairman and each author was given one topic and asked to write a section. All contributions were collated and circulated to the whole group. All of the authors were then invited to comment on all topics and a consensus meeting was held to examine the text. The principal messages were presented and open for comment at the 2018 EAU meeting in Copenhagen, Denmark. Thereafter the text was further modified. Consensus views were sought, the paper was formulated and submitted for peer review.

3. Data synthesis

There are two major elements of care: the first is maintaining a patient-centred approach and the second is the model of care to deliver that approach.

3.1. A patient-centred approach

Transition is the process that allows an adolescent to assume responsibility for their own health care and become the primary decision maker in their care. Effective transition into adult health care is paramount in complex conditions, and in other specialties there have been improvements in long-term function, respiratory outcomes, and survival [3–6]. Although this requires investment in teenagers, the long-term aim is to reduce the impact of their condition on individuals and the cost to health care systems.

The timing of transition will vary with maturity and independence. Preparation of the child and family should probably begin from the age of 11–12 yr [7]. A number of transitional models have evolved to suit a range of different health care environments and varying patient needs [8]. Adolescents require focus on their needs and the security of a lifelong plan. If this is absent, the patient is more vulnerable to problems such as malignancy, renal impairment, incontinence, sexual issues, and a low health-related quality of life [9]. A dedicated key worker such as a nurse, social worker, or doctor needs to monitor, coordinate, and act as a focal point for care and/or advice when needed [10–12].

It is estimated that a population of 4 million would provide enough work for one adolescent urologist [10]. Evidence of transition is limited, with only four small, short randomised controlled trials in any disease area. These studies showed that better knowledge of their condition improved the self-efficacy of individuals and their confidence in their health care system. It appears that those transitioning around the age of 18 yr fare better, while individuals with spina bifida are the most reluctant to transition [13–18].

Therefore, good preparation, a clear plan, and education are vital elements [19]. Patient education must include the importance of follow-up and establishing confidence to ask for health care or address problems of concern to individuals.

3.2. Potential models of care

Recognition of the growth in patient numbers and their complexity may require examination of more creative solutions. These might include hub-and-spoke working, regional or national multidisciplinary teams, and the potential use of technologies such as telemedicine. Some potential models of care are laid out below.

3.2.1. Lifelong care from a paediatric urologist when paediatric and adult urology departments are combined

Paediatric urologists form part of the wider department of urology. This allows absolute continuity and removes the anxiety for patients having to move to another team. The paediatric urologist can monitor their own results in the long term; they must have training in adult reconstructive urology but will be able to engage further subspecialist expertise (eg, stones and endourology, andrology, and oncology) when required. There will be greater potential for flexibility including joint clinics and operating.

A potential difficulty is that the paediatric urologist's career will be shorter than the lifelong care needed for their patients, so patients will have to move to a new doctor at some point. Lifelong care in a paediatric environment is not healthy for either an individual or for other surrounding (paediatric) patients [8].

3.2.2. *Lifelong care from a paediatric urologist who integrates with a separate adult urology department*

This maintains much of the continuity of the first model. The paediatric urologist is, of course, fully aware of the original diagnosis and treatment. It is important that they maintain the links and the insight to hand over patients to other, appropriate experts when it is in the patient's best interest. A further challenge for paediatric urologists looking after adult patients is the need to integrate with adult specialists outside urology.

3.2.3. *Transition from a paediatric urologist to an adolescent/adult specialist with an interest in this area*

This model of care requires an interested and appropriately trained "adult" urologist. The practitioner must understand the paediatric diagnosis and treatment. They must have spent time working in a specialist paediatric centre and be trained in reconstructive urology. This urologist must be focused on the overall care of the patient and cooperate effectively with specialists including nephrology, radiology, psychology, endocrinology, gynaecology, reconstructive surgery, endourology, and andrology [20].

Transition needs careful management and requires close cooperation in terms of preparation and handover. A joint clinic and good patient information are important. Views will differ, but allowing patients to meet their new carers in the paediatric environment and to agree on an appropriate timing for transfer (ie, what is most familiar to the patient) seems ideal [10–12].

3.2.4. *Direct transfer from a paediatric urologist to adult urology*

This is the riskiest model if it does not include a transition plan, and may result in patients becoming lost to follow-up or only presenting in an emergency [21]. Anecdotally, this may be something that patients unwittingly introduce for themselves. As these individuals grow, they may wish to go to university or work in another city or country. They may not wish or be able to travel to see their original urologist and it can be difficult to find a comparable level of expertise for them.

There is clearly a need for training in this area and for cross-working between paediatric and adult colleagues, possibly from an earlier stage than is seen in many environments. It does not matter what the origin of a trainee is (whether paediatric or adult) as long as they have the expertise required.

It appears on the basis of experience that many urologists have no interest in looking after patients born with spina bifida or other neurological problems, and can be intimidated when confronted with a patient who has undergone surgery for exstrophy or epispadias, severe hypospadias, differences in sex development, reflux, or other obstructive uropathy. We highlight some of the challenges below.

3.3. *The challenges of revision reconstructive surgery*

There is a clear imperative for long-term follow-up of complex diagnoses and surgical cases. A number of patients will need revision surgery, but we rarely have an accurate denominator to understand the precise risk. The risks of surgery for adolescents and adults with complex conditions and multiple prior surgeries can be significant. In adult neurogenic patients the overall risk of complications is 91.5% [22].

Some of the specific medical and surgical problems to be addressed in adolescent and adult life are as follows: (1) male or female fertility and sexual issues [23,24], (2) change in behaviour of bladder or reservoirs with age, (3) urological problems during pregnancy following reconstruction [25], (4) revision surgery, (5) anastomotic strictures (ureteric, urethral, and ureteroenteric), (6) small bowel obstruction (10%), (7) stomal stenosis (10%); and (8) incontinence (urinary 10–20%, faecal 20%) [26].

In any arena and at any age, primary surgery is easier than revision surgery; thus, at any age it must be clear that there is a defined surgical imperative for operating. In later life, abdominal adhesions, fibrosis, redundant tissue, and impaired blood supply will add to the complexity of revision surgery. There may be significant technical considerations such as renal access to deal with kidney stones with neobladders or conduits [27].

3.4. *Hypospadias: who to follow up and clinical concerns*

Hypospadias accounts for a substantial proportion of paediatric urology cases, and assessment and treatment require high-level expertise. Evidence increasingly suggests that this condition is best treated in a specialist centre [28,29].

Management can be complex and lacks standardisation of surgical technique. More than 300 different methods with a wide variety of modifications are described [28,30]. Centralisation of care has improved our understanding of hypospadias reconstruction. Complications may take decades to appear [31,32]. Pubertal growth may significantly affect the final outcome. In addition, psychosexual development and sexual function are important but can only be evaluated in adulthood [28,33–36].

Long-term aesthetic results need careful follow-up, as they become increasingly important to adult patients [37,38]. Genital and reproductive functions significantly affect the quality of life of adult patients with congenital penile anomalies [35]. Many studies only report short-term results for prepubertal patients; very few examine outcomes in adulthood [39].

Hypospadias patients have higher incidences of spraying, postvoid dribbling, and urinary stream deviation that worsen with the severity of hypospadias, leading to greater dissatisfaction. Dissatisfaction with sexual function and penile appearance is also more prevalent in adult patients

Table 1 – Overview of long-term hypospadias outcomes [36,38–40]

Outcome	Result (%)
Lower urinary tract function	
Spraying	10–63
Postvoid dribbling	20–40
Stream deviation	14–28
Lower urinary tract symptoms	3–85
Fistulae	0–25
Stricture	0–8
Cosmesis	
Patient dissatisfaction (mostly size)	7–81
Surgeon satisfaction	80–97
Psychosexual	
Sexual satisfaction	77–100
Curvature	5–23
Erectile difficulties	0–73
Ejaculation problems	5–36

than in controls. Table 1 summarises published data from the past decade. In these studies, results for lower urinary tract symptoms, psychosexual function, and quality-of-life score are equivalent, while outcomes for cosmesis and penile length are worse for patents with more severe hypospadias [36,38–40].

In conclusion, long-term outcomes of mild hypospadias repair are good and these patients usually need shorter follow-up. Patients with severe hypospadias will need long-term follow-up focusing on urinary and sexual function, fertility, and psychosexual support.

3.5. Neurogenic bladder in adolescence and adulthood

The majority of adolescents with neurogenic bladder have congenital spinal abnormalities requiring medical care in childhood. Neurogenic bladder may occur alongside other conditions including anorectal malformations, urogenital sinus, and cloacal exstrophy. Preservation of the upper tracts and maintaining a safe, compliant, and continent bladder are key [41]. Recent data show better survival without significant renal impairment [42]. However, the onset of puberty brings physical, psychological, and social development that can affect renal and bladder function [43].

Regular monitoring can be challenging if the patient chooses not to comply; support from the team and those around them are vital to try and keep these individuals safe. Annual consultation, an ultrasound scan, and measurement of serum creatinine are important. Further investigations such as radionucleotide scanning of kidneys, formal measurement of glomerular filtration rate, and urodynamic investigations may be performed in cases of deterioration in renal function, altered bladder function, loin pain, or recurrent urinary tract infection [44,45].

Sensitive discussion is needed regarding the type, practicality, and visual appearance of continence aids [46]. Minimising the use of indwelling catheters and continence pads is ideal. With physical maturity, alternative continence solutions such as fascial slings [47,48] and artificial urinary sphincters may be appropriate [49].

Sexual function needs to be considered, as concerns regarding urinary and faecal incontinence are heightened when patients become sexually active. Joint management of pregnancy (including preparation for conception with folic acid) with urologists and obstetricians is important.

Spina bifida patients deserve additional attention; many of them have a degree of cognitive impairment and will struggle with executive function, complex tasks (eg, clean intermittent self-catheterisation) and decision-making, and many depend on the input and supervision of caregivers.

These children are often treated in multidisciplinary paediatric neuropathic teams. Both parents (caregivers) and patients contribute to this way of working, which can change in an adult setting. There has to be cautious preparation for the increase in responsibility for their own care. There is a balance in creating a safety net that facilitates transition and allows independence (avoids rebellion and loss of engagement) but prevents harm.

3.6. Posterior urethral valves

Posterior urethral valves are the most common cause of male congenital lower urinary tract obstruction, with an incidence of 1 in 5000 live births. Posterior urethral valves cause renal dysplasia, severe reflux, chronic hydronephrosis, bladder dysfunction, and prenatal oligohydramnios resulting in pulmonary hypoplasia. Up to 20% of patients suffer with end-stage renal failure; some authors have suggested better long-term outcomes as a result of prenatal diagnosis [50].

Early recognition (ideally antenatally) and aggressive management, improvements in endourological instruments, nephrological management, neonatal care, and paediatric renal transplants have improved the initial poor prognosis.

In the longer term, patients with posterior urethral valves achieve day- and nighttime urinary continence significantly later than their healthy peers [51]. Adult valve patients with ongoing incontinence report more sleep disturbance and regard themselves more physically disabled, while those with renal insufficiency report lower quality of life in several domains [52].

3.6.1. Bladder dysfunction

In adulthood, the occurrence and bother of most lower urinary tract symptoms are doubled in posterior urethral valve patients [53]. Adult clinical phenotypes range from detrusor overactivity with poor compliance to myogenic failure with a significant postvoid residual [54]. Regular noninvasive bladder assessment is necessary.

3.6.2. Renal impairment

The lifetime risk of end-stage renal disease is approximately 28%, although a creatinine nadir of 1 mg/dl during the first year of life is a good long-term prognostic factor [55–58]. Progress to end-stage renal disease during or after puberty is unpredictable [59]. Proteinuria is a significant warning sign of deterioration and needs to be monitored.

Polyuria may increase postvoid residual volumes, causing progressive uropathy and a deterioration in the concentrating ability of the renal medulla that further

compounds polyuria, creating a cycle of decline. Good bladder emptying is vital; in extreme cases, overnight bladder drainage may delay renal deterioration and can improve sleep for patients with polyuria [60].

3.6.3. *Preparing for a renal transplant*

Patients in end-stage renal disease needing dialysis will have to be prepared for renal transplantation. Before transplantation, a full bladder and voiding assessment, including urodynamics, is necessary. Some patients will undergo bladder augmentation before renal transplantation. Urinary tract infections may be a factor for many reasons, but as long as the bladder is emptying these will not result in impaired graft function [61].

The outcome of augmentation is similar before or after kidney transplantation, and thus it may be acceptable to postpone cystoplasty as patients will be closely monitored [62].

3.7. *Transitional care in bladder exstrophy*

Classic bladder exstrophy (CBE) is part of the bladder exstrophy-epispadias complex that also includes cloacal exstrophy, male epispadias, female epispadias, and other rare variants. The CBE incidence is one in 30 000–40 000 live births and the male/female ratio is 1.5–5:1 [63]. In some European countries, centralisation of CBE care has led to focused expertise and better organization of care [64]. However, a demographic study established that only 12 out of 116 units receive more than six BE or epispadias referrals per year [65].

Management of BE in infants is well established and continence outcomes in specialist centres are good [66,67]. Adolescent and adult care has never been formally centralised, but the challenges are complex. In addition to urinary function, sexual and reproductive functions can be impaired in both sexes; in terms of impairment of sexual function, dorsal chordee occurs in up to 49% of men and vaginal stenosis in some 31.8% females [23–25,68].

Uterine prolapse among females with BE is much higher than among those not affected [69]. This highlights the need for well-supported multidisciplinary care combined with an expert urologist.

Any patient who has had major reconstruction needs advice about what to do in an emergency. Giving contact details for the reference centre for use by patients or less experienced staff is helpful, especially in emergencies.

3.8. *Discussion*

This paper sets out a current position and understanding of what is needed for patients born with congenital urological anomalies. Subspecialisation within paediatric urology seems to have shown benefit; while evidence to support this is improving, it remains lacking overall.

Transition represents a difficult stage in the treatment of patients with complex congenital malformations of the kidney or genital or urinary tract who will require lifelong specialist follow-up. A multidisciplinary service is important, with a range of specialists working together to manage these complex patients from cradle to grave. More often than not, the paediatric urologist will act as the “team leader” and coordinator of care within the multidisciplinary team.

Joint working can be achieved in a variety of ways, as discussed earlier in this paper. The objective is to guarantee the best possible and most reliable continuity of care for these patients. The decision taken by the European Society for Paediatric Urology and the EAU to initiate a close collaboration is important.

One of the main difficulties has always been the lack of “adult” specialists in urology dedicated to the treatment of patients affected by rare and complex congenital diseases. There is a need for adult urology to recognize the expanding group of these patients. In the first instance there need to be settings in which “adult” urologists can learn about paediatric care and effective management of patient transition. Formal training for dedicated specialists with the correct professional competences, for example, a fellowship in lifelong congenital urology, would represent an important step to generate interest in the field.

3.9. *Future research*

All the authors of this consensus have participated in and contributed to research looking at long-term outcomes for congenital urological problems. All acknowledge the shortcomings of the work to date. There needs to be a concerted effort to improve the research and thus the evidence available. This will further improve outcomes for patients, support clinicians, and demonstrate the importance of this work. Prospective shared data to produce larger series and more robust outcome measures are necessary. National data sets with full numerical information about procedures would provide clear denominators.

We need to provide a range of validated tools for standardised measures of both patient and surgical outcomes. These may involve patient-reported outcome measures or disease-specific quality-of-life measures. The evolution of specialist centres in collaboration with other allied centres and focused training and fellowships will further drive research.

There have been some good examples of outcome data in hypospadias, posterior urethral valves, and neuropathic bladder that have given insights into what we can learn from long-term care.

4. Conclusions

Lifelong care of congenital problems is challenging and essential for many but not all. Expertise is needed to provide the best care for patients and make the best use of resources. The political environment may significantly influence care for these patients. Specialist centres appear to be the most effective and safe model. In the long term it would be ideal to establish practice guidelines focusing on the common long-term problems of congenital urological conditions. The ultimate goal would be a structure whereby

all these patients will have access to excellent care with appropriate expertise.

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Study concept and design: Wood.
Acquisition of data: Wood, Baird, Carmignani, De Win, Hoebeke, Holmdahl, Manzoni, Nijman, Taylor, Tekgul.
Analysis and interpretation of data: Wood, Baird, Carmignani, De Win, Hoebeke, Holmdahl, Manzoni, Nijman, Taylor, Tekgul.
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