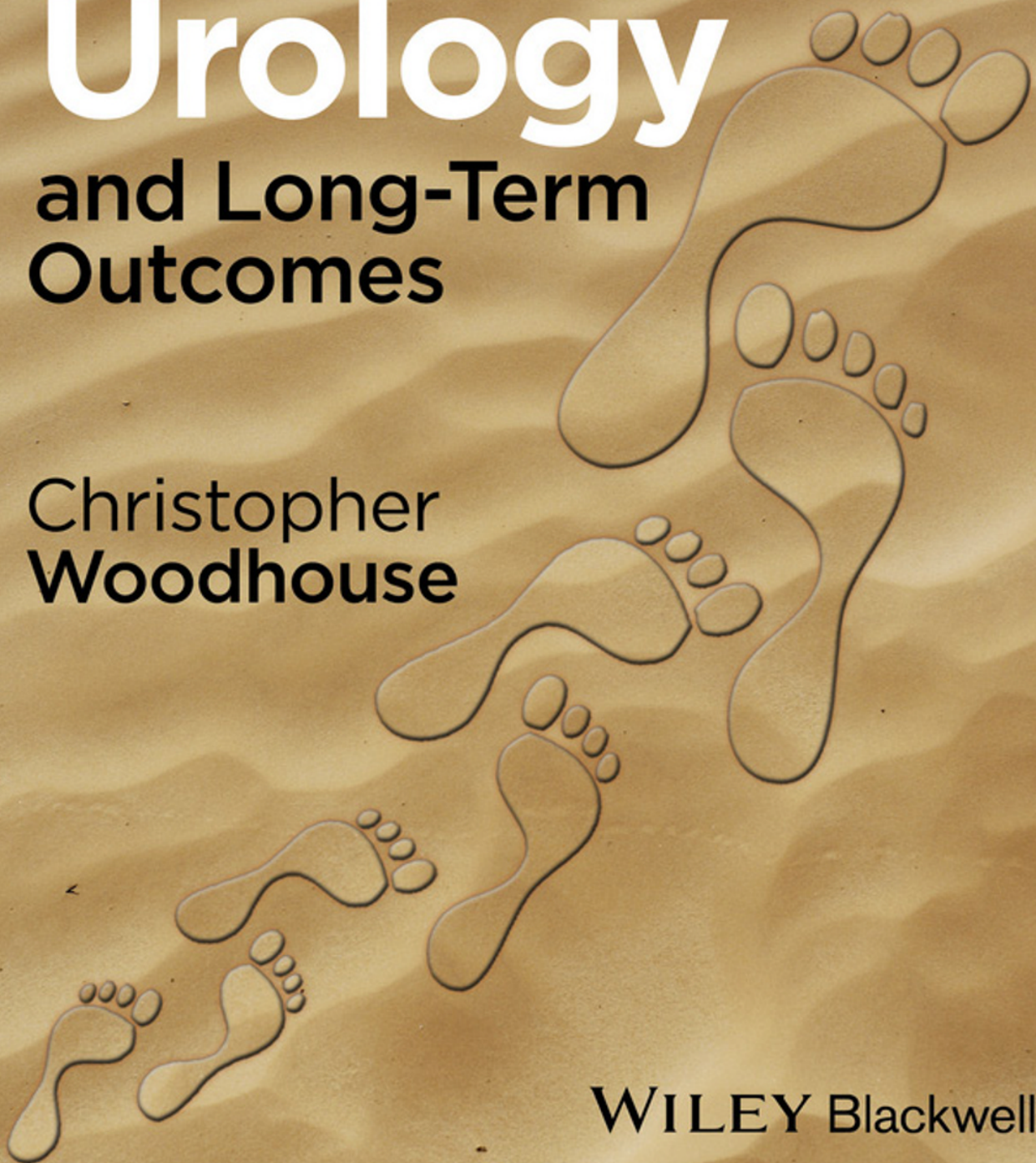
Adolescent Urology

and Long-Term Outcomes



Christopher **Woodhouse**

WILEY Blackwell

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To Anna, my wife, who has supported this project and my entire career.
Hers was the inspiration for the cover design.

Adolescent urology and long-term outcomes

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Preface

There are several surgeons who could be called "the father of pediatric urology." Sir David Innes Williams (1919–2013), surgeon at the Hospital for Children, Great Ormond Street (GOSH) and the Institute of Urology, has a good claim. He was the first to make a logical classification of congenital urological anomalies and his writings are still widely quoted, even though he retired in 1978. The operations that he described form the backbone of pediatric urology.

He never discharged his patients from follow up but transferred them to his adult clinic in the Institute of Urology. It was this "adolescent" practice that I inherited in 1981. Thereafter, the pediatric urologists at GOSH passed their patients on for lifelong follow up.

The good aspect of this arrangement was that the children, although attending an adult clinic, were in familiar buildings, seeing familiar staff, and maintaining a continuous medical record. Friendships that had been formed with other children were continued and occasionally patients married each other. Although this was not the first adolescent clinic in the world (the earliest that I can identify was in Boston Children's Hospital and started in 1954), it probably stimulated other specialties to set up the many adolescent clinics that now exist in University College Hospital in London (UCLH).

Unfortunately, there was a weakness in this apparently ideal system. There was no need for a formal process of "transition" from the children's clinic to the adult one. The hospitals were small; the staffs were partly the same; the buildings, clinical notes and supporting departments were the same; clinicians conducted joint clinics and meetings. The process of "transition" was, therefore, continuous and understood by staff and families.

Sadly, this happy state could not withstand the modernization of medical practice. Medical departments were moved into newer and larger buildings; children were separated from adults; clinicians became more specialized and drifted away from each other. My "adolescent clinic" became a part of UCLH while the children remained, for the large part, within GOSH, though adolescents also come from many other pediatric units in the United Kingdom and from elsewhere.

This divergence has been managed by the establishment of a "transition clinic" jointly staffed from GOSH and UCLH. Transition has become an expanding research field and is needed for all of the chronic conditions of childhood. Adolescence is a state instantly recognizable by anyone who has been the parent of a teenager. The characteristics that parents note in their offspring also translate into specific medical needs that are different both from those of children and adults. This produces a further dichotomy: should "adolescent" specialties exist in their own right and separate from the adult equivalent, or be the beginning of the relevant adult specialty?

It will be clear from this book that I believe it should be the latter. The seven ages of man, represented by the footsteps on the cover, are not independent but gradually evolve from one into the next. Although the needs of the "*infant, mewling and puking*" are different from those of the "*lover sighing like furnace*"* the medical needs are linked throughout life by the legacy of the original congenital anomaly. There should be an active process of transition out of childhood and into a specialty that can continue to supervise care indefinitely. A specialty that covers only the five or so years of growing up seems too narrow. Furthermore, much of the time in this period would be spent in preparing the adolescent for another move to yet another specialty. Although there are many difficulties in managing such a system, I believe that it is worth the effort and in the best interests of the patients.

The chapters in this book describe the long-term outcomes of the major anomalies of the genito-urinary tract. It should be particularly noted that almost none of the anomalies have an adult equivalent. This means, for example, that a patient with exstrophy or a posterior urethral valve is unlikely to get expert care in a general urology clinic. Apart

*Shakespeare, *As You Like It*: Act II, scene 7.

from describing the medical consequences, I hope they will show that even major congenital anomalies are perfectly compatible with a very successful adult life.

The development of adolescent urology described in the book would not have been possible without the support of my consultant colleagues and generations of registrars and nurses on whom the greater medical burdens always fall while the boss takes the credit! Many of them are recognized in the references to their work. I am most grateful to them and to the colleagues who have provided the critiques for each chapter.

I have been greatly helped by the staff of John Wiley & Sons, Ltd., and particularly the editors. Oliver Walter was kind enough to support the original proposal. Claire Brewer and Angela Cohen gave invaluable support and ensured that writing kept to schedule. Jayavel Radhakrishnan arranged the printing and accepted my many corrections without complaint. I suspect that the greatest burden fell to David Michael who had to check every word and every reference. His succinct queries arrived at regular intervals, including weekends, to produce the polished book.

Above all, I have learnt everything that I know about "adolescent urology" from the patients themselves. I am eternally grateful to them and full of admiration for their courage and for their enthusiasm for defeating their medical problems.

Christopher Woodhouse

CHAPTER 1
Adolescent and transitional urology—an introduction

Definition

The term "adolescent urology" means the care of patients with congenital or acquired conditions of the genitourinary tract from around puberty until death.

It is clearly not a very satisfactory term. "Adolescent" ought to refer only to the transitional period between childhood and adulthood. Unfortunately, no other term has so far emerged to cover the specialty.

Adolescent *medicine* is a specialty in its own right. However, it is predicated on the knowledge that the conditions are unique to the adolescent period, or that management of a chronic condition of childhood is altered by adolescence, but then returns to a more familiar pathway in adulthood. The urological conditions of childhood, as we shall see, do not have an obvious destination in adult urology.

Background

Historically the majority of children with the major congenital anomalies would have died young. There were almost no survivors of those born with spina bifida beyond infancy. Fifty percent of children with exstrophy died before the age of 10. There was no treatment for end-stage renal failure until chronic dialysis came along in the 1960s so about a half of those born with posterior urethral valves died before adulthood. There were no survivors amongst those born with cloacal exstrophy until 1960. Good antibiotics for gram-negative infections were discovered in the 1970s, before which gram-negative septicemia, especially in association with renal stones, was frequently fatal.

Although there were some long-term survivors, there was little need for long-term urological care until, perhaps, the 1970s. Figure 1.1 shows the number of papers listed in Pubmed with the search terms 'adolescent' and 'urology'.

With improving survival rates in the second half of the twentieth century came the realization that many children with major congenital anomalies had holistic and medical needs that would continue for the rest of their lives. The problem, then, was to identify clinicians with the knowledge and interest to take on such care. The needs are the same everywhere, although at present the solutions are limited. Gradually, special units have appeared, particularly in Europe and Australia, to provide this service. Adolescent wards are being built, mainly in pediatric hospitals.

However, there is a further difficulty: children do not become adults overnight or on achieving an arbitrary birthday. Adolescence is a phase of passing from childhood to adult life that is easily recognizable and creates unique problems in the management of chronic illness. It has a variety of formal definitions. In the *Oxford English Dictionary* it is described as "between childhood and manhood (14 to 25 years old) or womanhood (12 to 21)." The Department of Health in the United Kingdom is imprecise about its beginning, but rules that its end is the 19th birthday, which means that all care must be transferred at once to standard adult clinics. Neither of these definitions is wholly satisfactory, if only because children mature into adulthood at variable rates. For example when patients with

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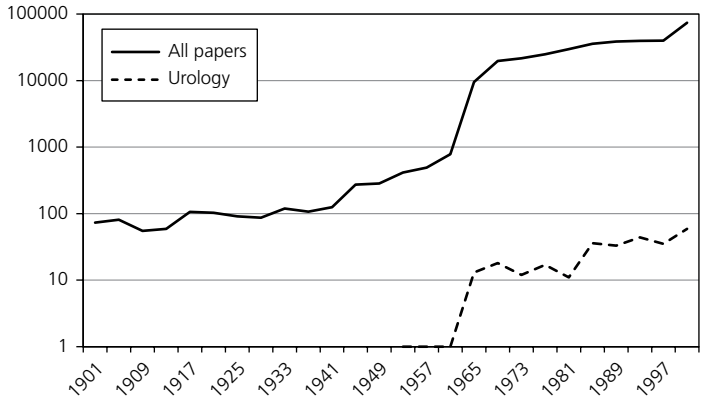

Figure 1.1 Graph with logarithmic scale to show the number of papers listed in PubMed with the search terms "adolescent" (solid line) and "adolescent urology" (broken line), by year. The data are inaccurate in about the first 50 years as there were no key words in most papers. On checking a random sample from the early years, most had no relevance to urology.

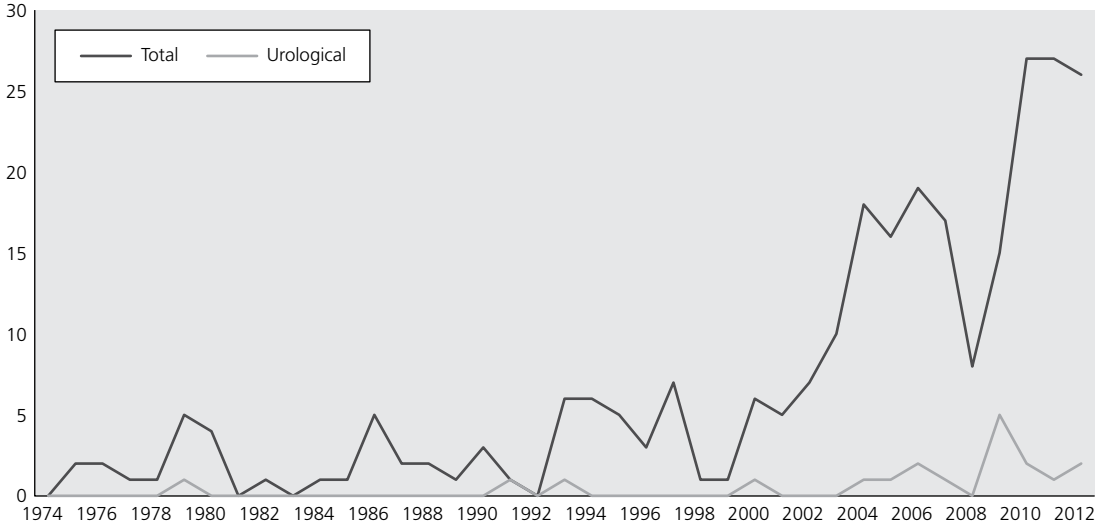

Figure 1.2 Papers listed in PubMed under "transitional care" (dark line) and "transitional care, urology" (light line) from 1974, when the first paper appears, to 2012.

congenital bladder disorders (mean age 20 years) were asked at what age they felt able to act independently from their parents, the mean answer was 17 years but the range was 11 to 25 years [1].

Although the need for a service to help the transition from pediatrics to adult medicine has been recognized in some specialties for 70 years or more, the main contribution in the twenty-first century has been the establishment of "transition clinics." The first that I can identify was founded by Dr. J. Roswell Gallagher in Boston Children's Hospital in 1954 [2]. Despite this, provision in the United States, and elsewhere, for long-term care remains limited [3]. Most subspecialties of pediatrics see the need for transition clinics but the provision is sparse and for many chronically ill or disabled children adult care is only available on sufferance in the pediatric hospital or in a general urology clinic. The lack of facilities for long-term care is reflected in the paucity of literature on the subject. Figure 1.2 shows the number of publications in PubMed on transitional care.

The purpose of a transition clinic is to prepare children and their families for continuing medical and

holistic care in the adult environment. It is predicated on the assumption that there is an adult unit to which such care may be transferred. In some chronic conditions of childhood, there is an obvious adult equivalent. For example, children with type I diabetes can eventually go to an adult diabetic specialist, although they will have a need for adolescent care in the interim. Even so, good transition is frequently unavailable [4].

In urology (and several other areas), there is no adult equivalent—there is no adult exstrophy, prune-belly syndrome (PBS) or disorder of sex development (DSD). Even the similarities that neural tube defects and posterior urethral valves (PUV) have with acquired adult conditions are deceptive. Children with major urological conditions need care that continues beyond adolescence and it is this which has to be planned in the transition clinic.

The prevalence of chronic illness in children in the United Kingdom is 17–19%. Data from other countries are limited. In the United States, in 1992, it was estimated that 31% of children had a chronic condition, however only 5% were considered to be severe and 29% moderately severe [5]. Even higher figures were recorded by Bethell *et al.* in 2011, but the increase was almost entirely accounted for by obesity. Excluding obesity, the prevalence of chronic disease in 12 to 17 year olds was 34.4% of which 50% were deemed to be severe. Although no urological condition appeared in the commonest 20 conditions considered, it is clear that long-term care is major numerical problem [6].

The process of transition

Barriers to transition

Perhaps the most obvious barrier to the movement of children with urologic anomalies into adult care is that there are very few clinics dedicated to receiving them! This makes transition difficult.

There are no figures available to show how many "adolescent urologists" are required. From the figures in Table 1.1 a total population of about 4 million would provide enough work for one urologist specializing in long-term care. This figure must be treated with caution. The incidence of the given anomalies is not accurately recorded. The incidences are falling and will continue to do so as selective pregnancy terminations become more common. Nonetheless, the figure does illustrate that it is a small specialty. If there is an even distribution of age and a working life of 30 years (both doubtful suppositions), there would only be a vacancy in the United Kingdom in alternate years. In the United States there would be about 80 adolescent urologists.

The practical aspects of geography have to be considered. There will be far fewer adolescent urologists than there are pediatric urologists. If there is a "long-term urologist" in the same city, a smooth transition should be possible. The more remotely the child lives, the more difficult will be the problem unless the family has the means and inclination to travel regularly.

In countries with some form of universal or socialized health system, it may be presumed that hospital funding would not be a problem. However, travel costs may not be refunded. In insurance-based systems, adolescents with long-term health problems are a poor commercial prospect.

The transition process

Children and their families are accustomed to the holistic care received in a pediatric setting. There is a specific focus on the anomaly, but education, social care, family support and other aspects are routinely covered. Patients feel particularly vulnerable as they approach adulthood [7]. Indeed, the strong attachment of the child to the pediatric staff has been shown to be a major barrier to transition [1].

Pediatricians should start to prepare their patients at an early stage. There can be no fixed time, but just the introduction of the idea that the doctor has a long-term plan is reassuring. General guidelines already exist, but remain to be validated [8, 9]. Viner has identified three elements to ensure safe effective transition:

- A cultural shift in staff attitudes and training. In particular, medical practice must adapt to the needs of the adolescent and not vice versa.
- Systems must change to ensure that all pediatric chronic illness and disability services have effective transition programs in place.
- Young patients must become effective partners in their own transition

Although he was writing about the National Health Service in the United Kingdom, the principles are universally applicable [10, 11].

The first formal step, perhaps at 11 or 12 years old, would be an assessment by the pediatric team of the long term needs. At about 13 or 14 years of age, the

Table 1.1 An estimate of the number of adolescent urologists required in the United Kingdom. Births per year are based on published incidences.

	Births per year	Total adults in population[a]	Total consultations at two per patient/year	Urologists needed at 1/1600 consultations
Exstrophy	14	840		
Posterior urethral valves	70	4200		
Spina bifida	105	4200		
Others (estimate)		2760		
Totals		12000	24000	15 = 1 per 4 million population

Notes: [a]It is presumed that all except those with spina bifida will have an adult life of 60 years, but only 40 years for those with spina bifida.

child should start to attend a clinic with both the pediatric urologist and members of the team who will be taking over. This allows mutual introductions and the beginning of a medical record for adult care. It has been shown that a key factor in successful transition is continuity of care [12]. At least four to five visits are required for the adolescent to build the same level of trust with the new physician that was experienced with the pediatrician [13]. With this level of continuity it has been shown that, at least in adolescent diabetics, the number of admissions can be reduced by up to 23% compared to those with no transitional arrangements [14].

For the adolescent urologist, it is essential to establish the status of the genito-urinary tract. This can be a complex process and in a few cases even the main diagnosis may have been lost in the mists of time. The basic requirements are shown in Table 1.2.

There then follows a transfer phase in which the consultations and decisions are made predominantly by the adolescent/adult clinicians. The pediatricians remain involved. Great care must be taken in the arrangement of joint clinics. Adolescents do not like the experience of facing a bank of clinicians together. There is also a danger that "management by committee" will lead to unsatisfactory decisions. A possible solution used in the transition of adolescents with DSD has worked well in the author's experience. The patients are discussed by a large multidisciplinary team before the patient visit. Two or three clinicians, well known to the patient (usually including a nurse specialist), then conduct the consultation. This allows a consideration of the surgical, endocrine, psychological, gynecological and sexual issues without a large cohort of people in the room.

Even this is subject to the difficulties of geography if the participants are all from different cities. To receive such specialist care, somebody has to travel. Different models are emerging. In the United Kingdom, most pediatric urology units have identified an "adult urologist" to continue the care. In the South Island of New Zealand, the relevant adult urologist travels to outlying pediatric units on a regular basis for transition and adult clinics; patients needing surgery or multidisciplinary care have to travel to the central hospital in Christchurch. In Sweden, a large country with a small population, the plan is to have two or three centers to which the patients must travel (personal communications).

Beyond childhood, clinicians should provide the same comprehensive care, but focused on adult life.

Consequences of transition failure

There are two very unsatisfactory possibilities—the child is "dumped" without any preparation, or continues as a cuckoo in the pediatric nest.

Personal experience suggests that "dumping" has a poor outcome. In the well structured transfer system in the author's practice, many patients were still lost. Those who returned in later life had sad accounts of their management in nonspecialist units. Intuitively it would be thought that a patient with a condition such as exstrophy would pose a challenge to a general urologist. However, there may well be patients who never returned and were happy with their management. In one series 49% of potential transferees felt that they could arrange their own urologic care (although only 24% knew what to expect) [1].

If such patients continue their care in a pediatric hospital, it will soon cease to be pediatric—humans with

Table 1.2 Details required for successful transfer of children to the adolescent urology clinic. Not all are required for all patients.

Kidneys	Bladder/reservoir	Genital tract—females	Genital tract—males	General
Glomerular filtration rate	Tissues of construction	Chromosomes	Chromosomes	Family history
Differential function	Outlet	Ovaries	Gonads: • Type • Location • Descent	Co-morbidity
Blood pressure	Continence system	Tubes	Endocrinology	Knowledge of diagnosis
Urinary protein	How emptied Infection pattern Metabolic consequences	Uterus Vagina Endocrinology		Schooling

a normal lifespan spend three-quarters of their lives as adults. In a pediatric hospital in Rio de Janeiro with no transition arrangements, adolescents and adults made up 19.8% and 2.7% of outpatients respectively [15].

In a survey of academic pediatric units in the United States, 2% of admissions from 1999 to 2008 were of patients between 18 and 21 years of age and 0.8% were over 21 years. This gave 60 000 inappropriate admissions in the 10-year period and there was a 6.9% annual increase [16].

Training requirements

There is no title, at present for the specialist who looks after children from pediatric urology when they grow up. For want of anything better, the term "adolescent urologist" is used and understood by pediatricians and urologists. However, the role goes well beyond the care of patients in their time between puberty and adulthood. There seems no point in having two transitions: childhood to adolescent care and then on to adult care. Once the patient leaves childhood, it is adult physical and emotional problems that predominate.

There are three broad requirements: knowledge of the relevant anomalies in pediatric urology and their management, an understanding of the emotional and physical changes that occur in adolescence, and a broad training in adult urology. The majority of the patient requirements are in adult medicine.

The breeding ground of the adolescent urologist will, therefore, be in standard adult urology. Some training, perhaps a year, is needed in pediatric urology. The problem of training in adolescent medicine is unresolved. The specialty of adolescent medicine is, itself, new and much of the training experience has been in family practice. Online training is available at http://www.e-lfh.org.uk/programmes/adolescent-health/ (accessed October 15, 2014).

The adolescent clinic

The age at which a patient transfers to the adolescent clinic must depend on the individual's level of maturity. As such clinics, at least in urology, are looking after people for the rest of their lives, the huge majority will definitely be adult. The clinical environment can be made appropriate for an adolescent, but the hospital or clinic will be adult. In some countries, there is a legal age at which patients can be treated in an adult environment. In the United Kingdom it is 16 years old. Patients with DSD should be seen in a separate clinic.

The new medical team has a difficult tightrope on which to walk! It is most important for the new physician to remember who is now the patient and not become a go-between in a parent/adolescent battle. A common problem arises when a symptom that is not life threatening, for example poor urinary continence, is seen by the parents as devastating and by the adolescent as unimportant.

In most societies, even when patients have not reached the legal age of consent, they are deemed competent to make medical decisions providing they understand the consequences of their actions (in the UK this is called

Gillick competence after the legal case that established the precedent).

A more difficult, but fortunately rare problem arises when an adolescent refuses life-saving treatment. Such cases will always require involvement of mental health teams and often legal ones as well.

There is no easy way of transferring responsibility from parents to the adolescent. I find it helpful, right from the start, to converse directly with the patient. Even when the parents ask a question, I prefer to address the answer to the patient. There will usually be an opportunity to examine the patient and the parents can be asked to wait outside. The physician can then ask if there are any private concerns that need discussion.

Most patients want to discuss their sexuality but are afraid to ask. Most physicians are afraid that they will ask! It is not usually a good idea for the physician to ask outright if sex is on the agenda. If the patient (or parents) broaches the subject, it must be addressed in a truthful and straightforward manner. Some families will have been given unrealistic expectations. If it is not raised at the first meeting, the subject must be introduced gently and sensitively in the subsequent ones.

Most of the conditions are urological in origin. The main exception is spina bifida (SB). In the United States most children with SB are looked after in dedicated hospitals such as Shriners (http://www.shrinershospitalsforchildren.org, accessed September 19, 2014). These are multidisciplinary units that provide an ideal environment for the management of a very difficult condition. Unfortunately, there are few replications of this elsewhere in the world or for adolescents and adults. As so many of the problems in children are with the bladder and kidneys, the long-term care is often based in urology.

As the urological patients grow through adulthood, they have needs that go beyond the genito-urinary system. It is necessary to have a group of clinicians who can share the care. Ideally, they should have an interest in the long-term care in their own fields, but this is not always possible. It is essential that they understand the idiosyncrasies of congenital urological problems. All the conditions require the support of radiologists and nuclear medicine specialists who understand the anomalies.

It is impractical to have multidisciplinary clinics with all the specialties, but so many have renal problems that a nephrologist is virtually essential for the running of an adolescent urology clinic. Table 1.3 shows the other specialists who may be needed.

Table 1.3 Associated specialists required for support in an adolescent clinic by urological diagnosis. This list is not exclusive. There is a universal need for specialist radiologists and nuclear medicine specialists. Psychologists are often required for any of these diagnoses.

Condition	Specialists required
Renal anomalies	Nephrologist
	Physician in hypertension
	Transplant team
Spina bifida	Nephrologist
	Orthopedist
	Neurologist
	Neurosurgeon
	Podiatrist
	Gynecologist/obstetrician
	Geneticist
	Plastic surgeon
Intestinal reservoirs	Gynecologist/obstetrician
	Biochemist
	Stone surgeon
	Stoma therapist
Exstrophy	Gynecologist/obstetrician
	Orthopedist
	Psychologist
	Oncologist
Posterior urethral valves	Nephrologist
Disorders of sex development	Andrologist
	Endocrinologist
	Biochemist
	Gynecologist
	Geneticist
	Sex therapist/psychologist
	Plastic surgeon
	Fertility specialist
Prune-belly syndrome	Nephrologist

There is a particular problem with psychology. Pediatric hospitals are usually well served with this specialty but, even so, the demand outstrips the supply. Many surgical papers point out the need for such support [17]. Adolescent psychology is a well defined subspecialty but is absolutely confined to the management of adolescents up to the age of 19 or 20. The period of true adolescence is crucial for the long-term physical and mental health of the individual. There is a change in the perceived health and social priorities, which require parental, medical and psychological support [18].

Beyond this is a huge void in the provision of psychological expertise for the chronically ill. One of the difficulties is in understanding the relationship between the strictly physical problems of an uncorrectable congenital anomaly and its psychological effects.

Geographical factors are a particular problem when arranging psychological care. The management needs frequent visits, which may not be possible if the patient lives a long way from a major medical center.

A very important use of follow up in a dedicated clinic is to record the outcomes. Some are known or predictable from knowledge of the original condition, such as renal failure in boys with posterior urethral valves. Others are linked to the original syndrome, but only identifiable as such when many are seen in a rare condition, such as bladder cancer in exstrophy. Finally, in perhaps the majority of cases, there are those diseases that all humans experience and are nothing to do with the original congenital anomaly.

Who can be discharged without long term follow-up?

Dischargeable patients

Children who have had inguino-scrotal surgery, successful pyeloplasty, surgery for undescended testis, and a few other conditions, may be considered cured and should be discharged. There is a small risk of neoplasia in boys with undescended testes, of which parents should be warned.

Fetal ultrasound has identified large numbers of babies with hydronephrosis. The commonest diagnosis is vesico-ureteric reflux, over 80% of which is in males. There is a high rate of spontaneous resolution even in high grades, with up to 40% being normal by 2 years old. Most will be discharged in childhood. Regardless of the age at diagnosis, long-term follow up is only required if there is significant renal failure or for the development of hypertension, the latter probably in the community.

The second commonest finding in the fetus with hydronephrosis is an abnormality at the pelvi ureteric junction (PUJ), which may or may not be obstructing. The management of this problem remains a matter of debate. Of the PUJ abnormalities, 90% of mild cases and 28% of severe cases resolve in childhood [19]. Those who are left with normal kidneys can be discharged.

Unresolved follow-up problems

Pelvi-ureteric obstruction

It is relatively easy to define children who have had surgical treatment and who can be considered cured. It is much more difficult to know what to do with those who have been managed "conservatively" and whose abnormality is still present. There is an obvious attraction for parents in finding that surgery is not necessary. At what point, however, can they be told that their "abnormality" is really a variation of normal and not a "disease"? This is not only of medical importance but is relevant to the mundane aspects of adulthood such as obtaining life or health insurance.

Perhaps the greatest dilemma is with the symptomless hydronephrotic kidney, especially those found on fetal ultrasound. After birth, most babies are managed conservatively. Some will resolve spontaneously and others will be cured by surgery. There remain about 20% of children whose hydronephrosis remains stable at least to the age of 16 and possibly to 22 years old. It is possible that late deterioration will occur in those who have been managed conservatively in childhood. At present there is inadequate evidence to decide whether children who have stable hydronephrosis at 16 years old can be discharged (see Chapter 2).

Hypospadias

It is well documented that the results of hypospadias repair are not as good as surgeons often report. Some surgeons have advocated long-term follow up of children to detect poor results and correct them [20,21].

The psychological problems encountered in children and adults with hypospadias may have been underestimated. In infancy there is evidence that surgery between 6 and 15 months of age minimizes psychological damage [22]. A good surgical result does produce a happier adult, even allowing for the disagreement between surgeons and patients over the definition of "good result." The worse the surgical result and the more severe the original hypospadias, the worse is the psychological outcome [23].

The dilemma is whether long-term follow up alters the physical or mental outcome for adults. There are no data to help, nor would it be easy to obtain them. It might be thought that an annual visit to the surgeon for the discussion and examination of a penis, which the boy thought to be normal and about surgery, of which he had no recollection, might generate a

Table 1.4 (a) The results of a questionnaire undertaken in the adolescent clinic at University College London Hospital over an 8 week period in 2010. All patients were asked to name their current occupation, which was recorded against their principal diagnosis. No patient refused to answer. (b) Examples.[a]

(a)

	n	Professional	Administrative	Skilled	Unskilled	Student	Housewife	Unemployed
Exstrophy	65	19	7	21	6	5	3	4
MMC	20	10	1	2	1	3	1	2
Ano/rectal	11	2	1	2	2	2	0	2
Ureteric	8	3	0	2	1	1	0	1
PUV	6	2	2	0	1	1	0	0
Other	16	7	0	0	2	5	1	1
TOTALS	**126**	**43**	**11**	**27**	**13**	**17**	**5**	**10**

(b)

	Profession	Administrative	Skilled	Unskilled	Not earning
Examples	Doctor	Social work	Farmer	Telesales	Student
	ITU nurse	Practice manager	IT tech	Decorator	Housewife
	Teacher	Book keeper	Footballer	Receptionist	Too ill
	Bomb disposal		Comedian	Caretaker	
	Film director		Electrician	DJ	
	Pilot		Shoe maker	Chauffeur	
	IT Academic				
Number	43	11	27	13	32
%	34	9	22	10	25

Note: [a]Woodhouse, C. R. J., Neild, G. H., Yu, R. N. and Bauer, S. (2012) Adult care of children from pediatric urology. *Journal of Urology* **187**(4), 1164–1171.

psychological problem that would not otherwise exist (see Chapter 15).

Acquisition of patients in adolescence

The huge majority of patients requiring long-term adolescent care will have one of the major congenital anomalies. Statistically, PUV and SB should account for 35% each, exstrophy about 5% and others about 25%. However, the workload will be more a reflection of the complexity of the cases, than the original diagnosis.

There are a few patients who develop a urological disease in adolescence, such as pelvic cancer or a late Wilms tumor. There are occasional late diagnoses of a congenital anomaly, such as DSD (especially the androgen insensitivity syndromes) or a PUV.

Drug abuse is an unfortunate fact of life in adolescence. Most of those that are commonly used have some genito-urinary effects. The most serious is ketamine (see Chapter 11).

Outcome measures

As far as urology is concerned, the first adolescent literature dates from the 1960s. To report on the outcome in young adulthood of patients treated as children, the follow up is a minimum of 18 to 20 years. Many of the congenital conditions are rare, so the span of follow up, to achieve any meaningful numbers, may be as long as a decade. This means that reports published in 2013 are of children who were born between 1983 and 1993. As pediatric urology is in a constant state of development,

great care must be taken in assessing long-term results, which may be irrelevant to children born in 2013.

Adult urologists should be aware that, despite many physical and emotional difficulties, adolescents with major congenital anomalies have an overwhelming desire to be normal, to be treated as normal, and to become normal adults. Their success may be measured from a survey of occupations they achieve (Table 1.4) [24].

In urological terms, renal function, continence, sexuality and fertility are the obvious goals. They are dealt with in the relevant chapters. Nonetheless, patients view their medical care not as an end in itself, but as a pathway to a normal life. Their priorities include education and training, employment, development of partnerships and family life. In other words, they have the same aspirations as their "normal" contemporaries.

Commentary

Russell Viner, University College London

Woodhouse provides us with an excellent and scholarly overview of a complex area. He begins by immediately grappling with one of the central dilemmas of modern specialist medicine with the observation that "adolescent urology" is not a very satisfactory term for the adolescent and adult management of congenital or early acquired urological problems.

This is a common problem in many specialty areas, and of course results from the historical origins of the way we organize medical specialties around our interests rather than the needs of the patients. Perhaps one of the closest analogies for urology comes from cardiology, where there have been a long history of disasters resulting from adult cardiologists (experts in coronary arteries!) managing a new caseload of adults surviving with univentricular hearts and peculiar anatomies that arise from repaired congenital heart disease. Pediatric and adult cardiologists have banded together, not without drama, to try to solve this issue by spawning a new subspecialty called "grown-up congenital heart disease" or GUCH. Woodhouse argues convincingly that a similar setup is needed for survivors of congenital urological anomalies.

He moves on to examine the history of healthcare for adolescents across the second half of the twentieth century and immediately meets a second set of similar challenges around defining adolescence, particularly its beginning and its end. He then focuses his discussion around the process of transition, its barriers, the difficulties of the process itself, training issues and the practicalities of how to run transitional clinics. In each of these he moves easily back and forward between urology and general observations.

Fortunately Chris Woodhouse avoids the classic trap that lies in wait for those enlarging on transitional issues, which is to focus on transition between pediatric and adult health care systems. Instead, Woodhouse shows an acute understanding of the needs of the patients by recognizing that specific clinics for young people need to be strongly established rather than merely a transitional process.

Woodhouse is clearly a master of his subject and I would have little argument with any of his pronouncements, except to challenge his statement that "it is not usually a good idea for the physician to ask outright if sex is on the agenda." I would, of course, bow to the knowledge of an experienced urologist but it is my experience and that of many colleagues that in the modern world we often need actively to raise such issues as part of holistic care.

Woodhouse ends by reminding us of the long perspective necessary to judge the outcomes of surgical intervention for complex anomalies. Yet he provides hope in showing that 43% of his patients end up in professional or managerial occupations, with achieving financial independence and vocational satisfaction surely one of the main end results of the tasks of adolescence.

Comment by Christopher Woodhouse

I think this is a very good review and just what I was hoping for. I agree that sex really should be on the agenda. It is the subject about which the patients are usually most anxious and clinicians are least anxious to discuss! A dialogue on sexuality and fertility should be an integral part of a good Transition Clinic. The problem arises if patients move directly from a pediatric environment to an adolescent one without any preparation. It is then very difficult to for a clinician to broach the subject of sex at the first meeting unless the patient or family raises the issue.

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PART I
The kidney

CHAPTER 2
Anatomical and cystic anomalies of the kidney

Pelvi-ureteric junction (PUJ) obstruction

Introduction

The problem of the dilated renal collecting system drained by a normal ureter has been of great interest to pediatric urologists since the anomaly was first identified. It was the recognition that not all such kidneys were obstructed that led to the invention of the Whitaker test and to the conclusion that dilatation of the kidney was not synonymous with obstruction.

Ante natal presentation

Fetal ultrasound has identified large numbers of babies with hydronephrosis. The second most common finding is an abnormality at the pelvi ureteric junction, which may or may not be obstructing. The management of this problem remains a matter of debate. Of the PUJ abnormalities, 90% of mild cases and 28% of severe cases resolve in childhood [1]. Those who are left with normal kidneys can be discharged.

When investigation after birth shows that renal function is good and there is no demonstrable obstruction, the children are monitored. If the kidney deteriorates, the pelvi-ureteric junction is reconstructed. It is difficult to know what to do with those who have been managed "conservatively" (that is without surgery) and whose abnormality is still present. Parents may be glad that surgery can be avoided; unfortunately we do not know when a dilated upper tract can be defined as an insignificant variation of normal.

On Society of Fetal Urology grading, all with grade I are likely to remain stable. The more severe the hydronephrosis, the more likely is it that surgery will be performed, so that all of grade IV and about three-quarters of grade III will have been operated by the age of seven, with the latest at ten. About a half of all patients managed conservatively will have complete resolution of the hydronephrosis.

There remain about 20% of children whose hydronephrosis does not disappear but is stable to the age of 16 years [2]. Babies in the large series from Great Ormond Hospital for Children (GOSH) have now been followed for up to 28 years. The prenatal and postnatal anteroposterior diameter of the renal pelvis measured in the transverse plane of the kidney has been found to be a very useful predictor of the need for surgery. Data are complete up to 16 years old when patients are transferred to adult follow up with notification of annual results back to GOSH. To date, no patient with renal pelvic dilatation < 25 mm has required late surgery (Dhillon, H. K. 2014 unpublished data quoted with permission).

Patients presenting with PUJ obstruction in adulthood are presumed to have had it since birth, and provide some limited evidence. In asymptomatic adults with renal pelvic dilatation confirmed *not* to be obstructed by renography, Whitaker test or both, deterioration is rare unless a complication such as a stone occurs. However, follow up seldom extends beyond 5 years [3]. In a series of asymptomatic or minimally symptomatic patients, 10 of 50 patients deteriorated within 2 years but none thereafter up to a mean of 4.5 years. Only two of 19 completely asymptomatic patients deteriorated [4].

Adolescent Urology and Long-Term Outcomes, First Edition. Christopher Woodhouse.

At present, however, there is inadequate evidence to decide whether silent deterioration will occur later or how monitoring in adulthood should be arranged when the renal pelvis remains dilated. The matter will never be resolved unless arrangements are made to follow up the patients with continuing but stable renal dilatation.

Adult presentation

The classic presentation is with renal pain, sometimes quite severe, provoked by fluids. In adolescents, the rapid fluid intake that is characteristic of socializing may uncover a PUJ obstruction that has been present since birth. With the free availability of ultrasound, many cases now are found by chance or on investigation of abdominal symptoms that have nothing to do with the kidney. Urinary tract infections that may have progressed to pyonephrosis are sometimes seen with an end-stage kidney, especially in the elderly. In children, infection in PUJ is rare unless there is coincident reflux.

There is a general presumption that PUJ obstruction is congenital. However, I have seen one case that was acquired in middle age. Figure 2.1(b) shows the CT scan of a woman with a classic history of pain on drinking, which had begun about two years earlier. Isotope scan confirmed obstruction. Figure 2.1(a) shows her intravenous urogram (IVU), made about 6 years earlier by her gynecologist because of recurrent UTI with intercourse and she had no upper tract symptoms. The IVU appears normal. At surgical repair there was only a single renal artery and the typical appearance of a PUJ obstruction. Histology of the excised junction was normal. Although it is usually foolish to draw major conclusions from one case, it does seem as though PUJ obstruction can occasionally be acquired.

Investigation in adults

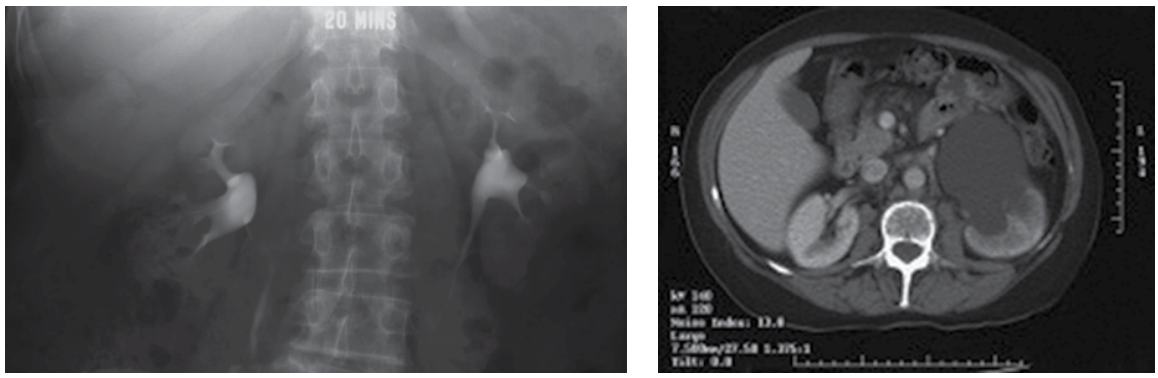
The standard investigations are well known and will not be discussed here. There are occasional cases in adolescents where intermittent flank pain appears to be renal in origin but renography is normal or equivocal. A Whitaker test may be helpful but since the widespread use of isotopes, few have the skill to perform it. Furthermore, in a unit where it is used regularly I have found that the result may also be equivocal.

It has been useful to try to provoke the pain by controlled fluid loading, usually intravenously and with furosemide, and then to image the kidneys. Either MAG3 renogram or a standard IVU can be used. If the kidneys look normal in the presence of pain, PUJ obstruction is not the diagnosis and vice versa.

The insertion of a double J ureteric stent as a diagnostic trial is seldom useful because it causes its own pain, which the patients cannot distinguish from their original pain.

Surgery

The dismembered pyeloplasty has a number of variations. The Anderson–Hynes operation was originally described in 1949 for the management of the retro-caval ureter. It has been in regular use ever since. The results of the open operation have been successful in 90% to 100% of children [5].



(a) (b)

Figure 2.1 (a) Intravenous urogram of a middle-aged female with uncomplicated recurrent cystitis and no upper tract symptoms. (b) CT scan of the same woman about 6 years later when she presented with a 2-year history of recurrent left flank pain associated with fluid intake.

The operation may be done through a flank or a dorsal lumbotomy incision. It has also been described as a laparoscopic or a robot assisted procedure [6, 7]. The outcome for all of these in terms of success and complications is the same. The virtues of one over the others are debated for this, as they are for many other operations. In a recent Dutch study on children the hospital stay was reduced from 6.7 to 1.2 days for laparoscopy and success as judged by renographic drainage was 98% versus 83%, when compared to lumbotomy [8].

For patients presenting as adults, the same techniques are used. Laparoscopic pyeloplasty is now a day case procedure in some hospitals. The results on follow up to 5 years are durable. The open operations are cheaper than the laparoscopic ones which are themselves cheaper than the robotically assisted ones [9–11].

A less invasive procedure of endoscopic pyelotomy has been described. It is inappropriate if there is a lower pole crossing vessel and it requires a stent for 6 weeks. There is a successful outcome in about 80% of children, but there is no long-term follow up and it has been largely replaced by the laparoscopic operations. It may be useful to salvage a failed formal pyeloplasty in adults [12].

Outcomes

The success of the operations is conventionally measured by relief of symptoms and as such is excellent. The appearances on standard imaging do not usually return to normal but are improved in up to 90% and stable on follow up at least to 5 years [5].

Functional improvement is less satisfactory in symptomatic children with only about a third showing renographic improvement even when the symptoms have been cured [13]. Those diagnosed by ante natal ultrasound do better. At least 50% with initial function of <40% have improved function after pyeloplasty. Improvement is greatest when the initial function is 30% to 39% (Dhillon, J., 2014, unpublished data quoted with permission). In these, there is an increase in the relative renal function of about 5% to 10% [14]. In babies who are initially managed conservatively but whose relative function deteriorates, surgery will return the function to the previous level in 95% of cases [15].

In adults, there is no renographic improvement with any level of preoperative function although, interestingly, the concentrating ability can improve [16, 17].

The long-term results, at least in children, are durable. The renographic function is unchanged at follow up at least to 5.5 years. These data have been confirmed in a group of 49 adults from 578 patients who were originally diagnosed by antenatal ultrasound. Pyeloplasty was performed at a mean age of 4.7 months and the 49 patients were followed at least to 18 years of age. No patient deteriorated beyond ten years of follow up and specifically none during or after puberty [14].

In the 6% or so or children who do require further surgery for a failed pyeloplasty, there is a tendency to persist with "conservative" procedures for too long. Some have two or even three procedures. The best chance of salvage is with conventional open surgery [18]. The reoperation rate on patients operated as adults is said to be 8% [19].

Conclusions

Successful pyeloplasty is almost always curative at whatever age it is done and follow up is not required. Babies diagnosed with a dilated renal pelvis by antenatal ultrasound and which resolves spontaneously can be discharged. However, if the renal dilatation does not resolve, the outcome is not yet certain. Current data make it clear that deterioration beyond 10 years old and up to 28 years old is very rare. The probability is that patients can safely be discharged at 16 and told that the kidney has a normal variant structure. However, every effort should be made to follow such patients from large pediatric research cohorts indefinitely in order to determine the natural history.

Horseshoe kidney

Horseshoe kidney (HSK) is the commonest structural abnormality and is found in about 1 in 400 people. As an isolated anomaly it is of no clinical significance unless a complication occurs. There is no increased risk of renal failure at least up to 30 years [20]. It may be associated with other major and minor congenital anomalies.

The HSK may develop any of the conditions that occur in normal kidneys. Whether any of them are more common in HSK is uncertain. To determine an increased risk of any acquired condition, it would be necessary to know the incidence of HSK in the normal