A Tribute: Prof. David Thomas

FROM THE GUEST EDITOR

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Prof. David Thomas has retired from the National Health Service after an illustrious career in Paediatric Urology spanning over 25 years. He is the only Professor in Paediatric Urology in the United Kingdom holding a personal chair following the University of Leeds recognising his contributions.

He has been at the forefront of developments in Paediatric Urology in the UK and around the world. He is well respected in the paediatric surgical and urological scene from his association with the British Association of the Paediatric Surgeons (BAPS), the Royal College of Surgeons in England and as the Chairman of the Specialist Training Authority in Paediatric Surgery. He has published extensively, written chapters, and edited his own text book in Paediatric Urology which is one of the essential reads for postgraduate trainees and young consultants alike.

He has developed a special relationship with colleagues in North America, regularly attending the AAP annual meetings and has travelled to various centres in the USA as visiting professor. His efforts are well appreciated throughout Europe and the European Society of Paediatric Urology (ESPU), serving as a member of the Joint Committee of Paediatric Urology (JCPU). He was awarded the honorary medal by the ESPU at its annual congress in Amsterdam in 2009 where he delivered the John Duckett lecture on “Prenatally Detected Urological Anomalies and Their Long-term Outcome.”

He has been actively involved in basic research in collaboration with University of York looking at composite cystoplasty with tissue engineered urothelium, holding several grants and supervising many fellows through their PhD programs.

I am incredibly fortunate to have worked with David for the last eight years and am honoured to have been able to organise a symposium on the bladder problems in children held in Leeds in July 2010. The theme of the symposium was on “Transitions in Paediatric Urology: What we have achieved in the last 20 years and what’s on the horizon for the future.” This edition of the Dialogues in Paediatric Urology contains contributions from all the distinguished speakers at the symposium and is dedicated to Prof Thomas’s career and his contributions and achievements. We in the Paediatric Urology scene are indebted to him for all his efforts. I wish him the very best in his retirement and good times with his lovely wife, Marilyn, who has supported him for over 30 years.

FROM THE EDITOR

Anthony A. Caldamone, M.D.

It would be an exhausting list indeed to attempt to document the academic contributions of David Thomas – both in the clinical and basic science arenas. One would need to devote a considerable amount of time, just to read the titles of his significant manuscripts. However, what I would consider his most important contribution is the role he has played in the development of pediatric urology as a respected international specialty. His efforts in establishing standards of training notwithstanding, I see his ambassadorship for our specialty as one of the necessary stepping stones in the growth of pediatric urology. His credibility across related subspecialties, his ability to collaborate effectively, his willingness to participate regularly in international gatherings of pediatric urologists all helped to develop an infrastructure of communication and sharing of science that is needed in a nascent subspecialty. As I mentioned at the dinner honoring his career, he put Leeds on the map as a center of Pediatric Urology, at a time when there were very few centers around the world. David, for all of this the pediatric urologic community will be forever indebted.
The introduction of enterocystoplasty into Pediatric Urology in the early 1980s was the key factor in the development of lower tract reconstruction in children. Enterocystoplasty provides a reliable and effective means of increasing bladder capacity and compliance. However, it is also associated with significant potential morbidity and longer term complications which are almost entirely attributable to the fact that the structure and physiological function of intestinal epithelium is unsuited to prolonged contact with urine. The ideal material for bladder augmentation or substitution would, therefore, combine the functional properties of smooth muscle with a lining comprised of urothelium.

This contribution to Dialogues in Pediatric Urology is a brief personal perspective on the current status and future direction of the different research strategies aimed at achieving this goal.

Acellular Biomatrices

This concept envisages augmentation with an “off the shelf” acellular graft or matrix serving as a scaffold for tissue regeneration by cellular ingrowth from surrounding native bladder tissue. If successful, this approach would obviate the requirement for tissue-cultured cells. Favorable results in experimental animals have been reported from Oklahoma and Indiana using small intestinal submucosa (SIS) and bladder acellular matrix graft (BAMG) respectively. But the outcome in experimental animals may prove difficult to replicate in man. Firstly, unlike the materials used in experimental studies, the biological properties of materials currently approved for clinical use have been considerably altered during the manufacturing and sterilization process. To overcome this deficiency, a major European study is underway to explore the possibility of restoring biologically active agents such as growth factors into the acellular graft material prior to its use in surgical reconstruction. The second (and possibly insurmountable) problem relates to the dimensions of the acellular scaffold. The ratio of perimeter to area is almost certainly a critical determinant of the effectiveness of tissue regeneration by cellular ingrowth from surrounding native tissue. Typically, the area of an augmenting ileal segment exceeds 200cm² - an order of magnitude greater than the area of acellular grafts described in experimental models. Progressing this strategy to the clinical domain is likely to be dependent upon developing methods of inducing angiogenesis to promote vascularisation of the regenerating stroma and smooth muscle.

Tissue Engineered Bladder

Pioneered in Boston and Wake Forest this approach entails harvesting and isolating autologous urothelial and detrusor cells which are then seeded into an acellular collagen-polyglycolic acid ‘construct’. The cellularised construct is incubated in a bio-reactor for a number of weeks before implantation for bladder substitution.

Having piloted the technique in a canine model, the Boston group were granted approval to proceed to clinical trials, initially treating 9 young myelomeningocele patients (although data on only 7 patients were published because 2 were lost to follow up). This impressive scientific achievement attracted widespread media interest. However the published urodynamic data indicate that in only one patient was bladder capacity substantially increased post-augmentation, whereas capacity was only modestly increased, unchanged or reduced in the remaining 6 patients. The results of a second, Phase II study have yet to be published, but data presented to the 2009 ESPU meeting highlighted the importance of post-operative mechanical distension of the neobladder (‘cycling’) in promoting and maintaining adequate capacity and compliance.

Some questions, therefore, surround the functionality of tissue engineered smooth muscle when reintroduced into the patient. It remains to be seen whether the functional phenotype of smooth muscle cells is maintained over time or whether there is a tendency to de-differentiation into myofibroblasts - leading in turn to collagen formation and contraction.

Composite Cystoplasty

This strategy, developed in Leeds and York, is less ambitious since the tissue engineered component is limited to a single cell type, namely urothelium. Composite cystoplasty is not dependant upon tissue engineering to generate the smooth muscle component, since this is provided by a vascularised seromuscular segment of de-epithelialised native intestine.

Prior to reconstruction, autologous urothelium is harvested and expanded in vitro to form robust sheets of urothelium which are then attached to the de-epithelialised seromuscular segment of intestine at the time of bladder augmentation. Proof of principle was established in a porcine model using de-epithelialised uterine smooth muscle as a surrogate for colon. Further laboratory work was then undertaken to enhance the functional integrity and barrier properties of the urothelium by promoting fuller differentiation and stratification prior to surgery. In a second porcine experimental study, sheets of autologous tissue engineered urothelium were combined with seromuscular segments of colon which had been created by a technique of extra-mucosal dissection similar to that first reported from Recife. After three months the augmenting segments were viable with no evidence of fibrosis or contraction. At a cellular level all the composite augmentations were lined by confluent urothelium which expressed normal differentiation markers. Importantly there was no evidence of colonic mucosal or crypt regrowth. Although this strategy has yet to make the transition into the clinical domain, the feasibility of using de epithelialised bowel segments is already well established in clinical surgery - for example, in the context of seromuscular cystoplasty.

Has the pursuit of a tissue engineered urothelial - lined augment already been sidelined by advances in medical management or recent innovations, notably intravesical Botulinum Toxin A?

To date, published experience with BTX-A in children is very limited and the probable requirement for repeated injections over the duration of a young patient’s lifetime is a major deterrent. Aggressive anticholinergic therapy hardly constitutes a therapeutic breakthrough. On the basis of their extensive experience of the pediatric neuropathic bladder, the Indiana group recently observed ‘despite the best medical management, there are still patients who develop a small-capacity, poorly compliant bladder and in this scenario, we would still recommend enterocystoplasty as the gold standard.’

In conclusion, the development of a safe and effective urothelial-lined alternative to conventional enterocystoplasty remains one of the most important and relevant goals in Pediatric Urology.
New Insights into the Fetal Development of the Lower Urinary Tract

Fetal Bladder Wall Development

In paediatric urology the understanding of bladder function and development has been tremendously advanced during the last decades. This was, in particular, achieved through a deeper insight into special features of functional voiding patterns, technical achievements of ultrasonography allowing precise, repeatable readings and substantial discoveries in basic research that elucidated the interplay of numerous genes and signalling molecules. Clinical studies on the characteristics of primary vesicoureteric reflux in male and female infants have highlighted gender-related differences. Severe reflux appeared more common in male infants than in female and was associated with thicker bladder walls. A gender specific transient urethral obstruction during early fetal development in males has been proposed. To assess detrusor muscle thickness, several authors provided ultrasound measurements and evaluated its reliability in children. At present, it is indisputable that antenatal obstruction of the bladder outlet results in significant changes in bladder morphology, due to measurable smooth muscle hypertrophy. Anatomical studies of human fetal bladder development revealed smooth muscle bundles in the bladder detrusor about the 12th week of gestation. At that time outgrowths of the prostate gland arise in male fetuses, whereas in female a comparable outgrowth does not occur.

Current literature reveals that fetal bladder wall development occurs at 9 weeks of gestation and takes place uniformly in both genders. Bladder wall thickness increases significantly with gestational age irrespective of the related sex. (Fig. 1a, 1b)

Figure 1a
A significant increase in bladder wall thickness from the 13th week of gestation (Fig. 1a) until the newborn period (Fig. 1b) is clearly visible.

Figure 1b

Fetal Development of the Ureterovesical Junction

The ureterovesical junction represents the boundary area between the low pressure in the upper urinary tract and the highly variable pressure of the lower urinary tract. (Fig. 2) The mechanism of ureteric motility as well as detailed physiology of the normal valve mechanism of the ureterovesical junction has been examined in some depth. Mackie and Stephens postulated that in congenital anomalies of the kidney and the urinary tract the initial ureteric budding occurs at an abnormal position of the Wolffian duct (mesonephric duct). The end of the ureter fuses with the urogenital sinus by day 37, the following caudal growth remains indefinite, mainly the distension and intravesical submucosal enlargement which is supposed most responsible for the antireflux mechanism. Migration of the ureteral orifice in the bladder is described in a cranial and lateral direction with the final position of the orifice at risk for reflux.

Two main anti-reflux mechanisms - one passive and the other active – are discussed in the literature. Sampson described the theory of a ureterovesical lock mechanism. Determining factors in passive valve function are the diagonal course and the length of the submucosal portion of the ureter; this intravesical course of the ureter is considered to be of functional importance in maintaining the ureterovesical closure preventing reflux.

The active valve function of the human ureter is thought to consist of a “ureterovesical sphincter” which contracts in response to vesical contraction and relaxes following contraction of the external urethral sphincter. Since a histologically definable sphincter could not be iden-
While King considered the ureterovesical valve to be activated by the intrinsic muscle of the ureter, Tanagho attributed this function to contraction of the muscle of the superficial trigone of the bladder. This theory of trigonal attachment of the ureteric function was weakened by Hannan, who excised the canine trigone except for the mucosal layer, and found reflux in only one specimen of his series.

Regardless which of these models of valve function is correct, the effectiveness of both depends on the functioning ureteric muscle. Thus, it seems clear that the preconditions of a patent ureterovesical junction is a timely and complete development of the smooth muscle coat of the intravesical ureter. It can be doubted that the length and course of the intravesical ureter is of sole importance in the prevention of reflux.

Initial ureteric budding occurs out of the Wolffian duct and the distal end of the ureter fuses with the urogenital sinus by day 37 of gestation. Further on intravesical ureter wall thickness increases throughout fetal development with a linear growth profile showing a linear relationship starting with a thickness of 131.77 mm at week 9 of gestation and increasing to 451.5 mm at week 39. There is an increase in ureter wall thickness of 11.25 mm per week throughout fetal development.

Concerning the intravesical ureter length, there is a linear growth profile with a positive linear relationship as well starting at week 9 of gestation with a length of 271.7 mm and resulting in a length of 3017.2 mm at week 39. Thus results in a growing of 97.4 mm per week during fetal development. (Fig. 3)

When looking at the ratio of intravesical ureter wall length to intravesical ureter wall thickness it is evident that there is no so-called tunnel during the early fetal period since the ratio is 1:1 till the 18th week of gestation. At birth the ratio increases to 2.23:1. Therefore, the ratio is obviously much lower than assumed so far.

One may conclude that the passive antireflux mechanism may not be as important as previously thought or alternatively that a shorter tunnel may be sufficient as well to prevent reflux. On the other hand there are certainly other factors that play a crucial role in the whole antireflux mechanism. The active valve mechanism, the muscular coat of the distal ureter, the connection with the trigone and the posterior urethra as well as bladder function has to taken into account.

Concerning the very distal part of the ureter there is a strong significant positive linear relationship between gestational age and both intravesical ureteral wall thickness of the mesenchymal and smooth muscle wall and Intravesical ureteral length. Additionally, there is no development of an intravesical tunnel until late fetal period with a low ratio of the intravesical ureter length to the ureteral diameter (2.23:1) in newborns. Consequently, the passive antireflux mechanism (the so-called intravesical tunnel) might not be as important as previously thought.

Fetal Development of the Prostate

The fetal development of the prostate starts in the 10th gestational week while its regulation is androgen dependent. First morphologic changes indicating prostate development occur with the outgrowth of the urogenital sinus epithelium into the surrounding mesenchyma forming small epithelial buds which develop into the main prostatic ducts. During this branching morphogenesis the ducts elongate and form a complex, widely ramified system. During this development the columnar, basal and neuroendocrine cell compartments of the prostate's epithelium become visible. The stroma of the mesenchyma consists of fibroblasts, smooth muscle cells, neural and vascular elements as well as an extracellular matrix. In previously examined fetal specimens we have already assumed an accelerated growth of the prostate and a remarkably low bladder-prostate ratio reminding of benign prostatic hyperplasia in adults. A pathophysiological link between these two periods may be the hormonal “imprinting” during the fetal and neonatal period as well as in growing adult prostates upon aging. It has been shown that fetal infravesical obstruction in male fetuses can be correlated with the prenatal enlarged prostate, which in consequence may

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be responsible for the transient higher micturition pressures in male newborns and infants. This hypothetical infravesical obstruction may be a possible cause for the higher incidence of VUR in male newborns; in addition, the high reflux resolution rate within the first two years of life could be explained by the decrease of testosterone levels after the 5th postnatal month.

Starting at the beginning of the 2nd trimester fetal prostate growth as well as prostate volume increase exponentially. A correlation with the increase of maternal testosterone level is apparent. In comparison with the comparative small fetal bladder, an infravesical obstruction may be assumed. This may be an additional reason for the higher incidence of vesicoureteral reflux in infant males. The decrease in postnatal testosterone levels is associated with the quiescence of prostatic growth leading to a shift of the size relation between the bladder and the prostate. In this context the higher resolution rate of high grade reflux in boys is reasonable and might serve as an explanation of the high rate of spontaneous reflux cessation in male infants within the first two years of life. (Fig. 4)

**Fetal Development of the Female Spincter**

In 1809, Wilson was probably the first to describe an individual striated muscle surrounding the urethra in both sexes. Within proceeding literature, the existence of this individual muscle was denied by many authors and it was thought to be a part of the so called “urogenital diaphragm”. The discovery that striated fibers of the sphincter musculature surrounding the urethra were significantly smaller than those of other pelvic muscles gave new support for the theory of an independent external urinary sphincter muscle. From that point, morphological descriptions were mainly dedicated to the whole muscle aspect while the spatial extent and configuration remained subjects of discussion. Meanwhile it is widely accepted that the external urinary sphincter is built from a smooth (lissosphincter) and a striated (rhabdosphincter) component. In addition to that, several authors suggested that both the female rhabdosphincter and the lissosphincter extend as a single unit along the entire urethra. (Fig. 5) More recent concepts of the external urethral sphincter system in the female were drawn in regard to urethra sparing cystectomy. In contrast to previous studies it was shown that the rhabdosphincter was predominantly located in the distal two thirds whereas the lissosphincter layers were abundant only in the proximal two-thirds of the urethra. Consequently both sphincter components are overlapping only in the middle third of the urethra corresponding to

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the point of maximum urethral closure pressure. Furthermore, as in the male, the rhabdosphincter in the female is supposed to have an omega-like shape in transverse sections due to a posterior connective tissue raphe, nevertheless some groups have described a nearly ring-shaped, circular configuration with posteriorly crossing muscle fibers.

Up to now, the extent and the configuration of the female urethral sphincter continue to be controversial issues in the literature. Embryological studies of the external sphincter apparatus of the female urethra have already been conducted in order to elucidate and understand its adult anatomy and function. Nevertheless, it still remains a subject of confusion, since different configurations such as omega-, ring-, or elliptical-shape are described. In addition to that, the extent of the sphincter components to either the proximal and/or the distal third of the urethra is characterized differently.

According to current literature the female rhabdosphincter and the lissosphincter have a common primordium (SUP) which consists of a myoblast plate located in the anterior aspect of the urogenital sinus. The SUP is already present before the beginning of the development of the vagina. Both the lissosphincter and the rhabdosphincter acquire an omega-shaped configuration during fetal development due to a constant posterior connective tissue raphe. The maximum thickness of the external sphincter complex is achieved in the middle third of the urethra corresponding to the point of peak urethral closure pressure on urodynamic evaluation. The development of the vagina leads to modifications of the configuration and the spatial extent of both the smooth muscular as well as the striated part of the external sphincter apparatus. Alterations of the rhabdosphincter architecture are limited to the late fetal period.

**References**


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tively obstructing the bladder outlet may bring short-term benefits of control but that these are finite over a lifetime.

When faced with an operation that delivers poor results the surgeon has only two possible explanations – either he’s the wrong surgeon and unable to perform the procedure properly or it’s the wrong operation that cannot deliver the result that both he and the patient require. Although many excellent and meticulous surgeons have continued to try to replicate the best results reported for the modern staged repair, others have now accepted that new techniques are needed. Two main approaches have emerged – the complete neonatal reconstruction proposed by Mitchell and the Kelly operation. Since the advent of these procedures, negative campaigning by proponents of the Modern Staged Repair has sought to overshadow their promising early results. We are still denied an objective, scientific comparison that can inform the next generation of surgeons as how best to treat their patients.

Twelve years ago, when he spent some time in our centre, Justin Kelly shared his premise that in bladder extrophy “normal structures are wrongly orientated and if correctly reconstructed can regain physiological function”. This fundamentally different approach set an appealing outcome measure - long-term physiological voiding – but offered also considerable technical challenges. Reopening the extrophy bladder and performing bilateral ureteric reimplants is technically demanding and full mobilisation of the penile corpora from the inferior pubic rami by freeing the pudendal pedicles from their course beneath the pelvic floor is not for the faint hearted. The latter important and characteristic manoeuvre does allow the base of the penis to come together in the midline, resulting in greater external penile length. Moving the soft tissues from the bone allows reconstruction of the penile base, bladder outlet and proximal urethra without the need for routine pelvic osteotomies, whose role becomes simply to allow tension free closure of the midline in a minority of patients with either a wide pubic diastasis, delayed primary closure or revision bladder closure. The bladder neck closure in this procedure is loose and its position about half way between the veru montanum and the ureteric orifices. The proximal urethra is brought ventrally between the penile corpora to restore its usual angle and at this point the muscle of the pelvic diaphragm can be wrapped around it as a notional external sphincter. If the urethral plate is short it is completely lifted from the glans and a hypospadiac urethral opening made on the ventrum of the penis. In some cases however a Cantwell-Ransley urethral repair can be incorporated, obviating the need for later distal urethral reconstruction.

Major complications should be the most compelling reasons for abandoning a reconstructive operation and none is more disastrous in exstrophy surgery than the loss of the whole or part of the penis. These cases tend to migrate to major exstrophy centres for their salvage procedures and we have seen patients with this complication following modern staged repairs, complete neonatal repairs and penile disassembly techniques or even ‘simple’ primary closures. The standardised Kelly procedure has now been performed in over 100 male cases in our centre over the past 10 years and thus far without penile, corporal or glanular loss. Having started with one of our four surgeons, it is now the standard approach for all to stage the procedure with a simple neonatal bladder closure and then the Kelly operation usually in the second year of life. Steps that help to minimise risk to the penis include avoiding proximal dissection medial to the corporal bodies above the veru montanum; maintaining a bridge of glanular tissue between the two corporal bodies where possible (i.e., no complete disassembly) and avoiding any sutures that may directly or indirectly constrict the corporal bodies proximally. Moreover, the ability to mobilise the corpora fully has proved very useful for penile salvage and male cloacal exstrophy cases - in the latter almost eliminating the need to change sex of rear ing.

Just over 11 years ago, around the same time that we introduced the Kelly operation, an historic agreement amongst UK paediatric urologists concentrated the management of bladder exstrophy in two centres (Great Ormond Street and Manchester Children’s Hospital) with central funding by the National Health Service. This arrangement has now been in place for 11 years and the large numbers (between 8 and 16 new cases per annum) have provided an opportunity to study this new procedure, with routine prospective data collection on every patient and regular reporting of results and complications to both the NHS commissioners and the British Association of Paediatric Urology (BAPU).

From the beginning, a standardised operative technique has been applied to consecutive patients, regardless of the initial size of their bladder plate or early bladder capacity development after the primary closure. To understand continence outcomes a simple classification was developed and this has proved both practical and easy for both patients and their carers to understand (outlined below). Regular independent assessment by our clinical nurse specialists, who have always been very objective about our results, has been a very important part of the follow up routine.

A practical classification of continence after surgery in bladder exstrophy

<table>
<thead>
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<th>Level</th>
<th>Description</th>
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<tr>
<td>0</td>
<td>Dribbles urine the whole time with no control</td>
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<tr>
<td>I</td>
<td>Able to retain urine with a ‘dry interval’; some control but still wearing protection</td>
</tr>
<tr>
<td>II</td>
<td>Sufficient dry intervals by day; in underwear and not needing protection; wet at night</td>
</tr>
<tr>
<td>III</td>
<td>Dry by day and night; no protection or accidents; “normal” child</td>
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The first review of 20 cases revealed that most (18/90%) developed a dry interval quite soon after the operation (level I - demonstrated using a nappy alarm). Being able to hold on for longer and get out of nappies by day (level II) required the development of bladder capacity. Only 17% followed for less than two years attained this, although amongst 14 patients with longer than 2 years follow up 9 (64%) were dry by day and of these 5 (36%) were also dry at night, attaining level III. At the 50 patient milestone 71% of patients followed for more than 2 years were at level II and 25% at level III. Of those followed
more than 5 years, 80% had achieved daytime continence. So continence develops after this operation in line with increasing bladder capacity, itself following the provision of outlet resistance.

This gradual attainment of continence suggests physiological function and the lack of post-operative upper tract dilatation, commonly seen in the days of classical bladder neck reconstruction, seems to confirm we are not causing obstruction. We regularly perform uroscopy and can see the coaptation of the bladder outlet and its opening by the irrigation jet from the cystoscope. Video-urodynamic studies performed so far in a few patients show not only physiological pressure flow dynamics but also suggest appropriate anatomical orientation of the bladder outlet and its complete emptying.

The same surgical approach was used initially in the management of epispadias, believing that as a more minor variant of the exstrophy-epispadias complex the results should be better. Although 5 out of 6 females became dry by day (83%), only 9 of 18 males achieved this (50%) after a Kelly operation. On reflection, two important determinants of a minor variant – that it occurs more commonly and the results of treatment are better – are not fulfilled for male epispadias patients. Moreover the observation of small and unusually tight ureteric orifices (often calibrating less than 3Fr), unusual trigonal morphology and thin bladder neck muscle in these patients suggests that bladder neck dysplasia is a dominant influence. We have changed our approach to the management of these patients.

In conclusion, physiological voiding is now the goal of bladder extrophy management and the Kelly operation is definitely an advance but may not be the final solution. In our unselected patients it does provide the hope of daytime continence in over 70% of cases with classic bladder extrophy with a low complication rate and may also offer significant benefits to the penis. We badly need a standard assessment tool for continence in children which enables the results of various approaches to be understood and compared. Epispadias in males is not just a minor variant of the exstrophy-epispadias complex.

Search for a Standardized Quantitative Evaluation of Lower Urinary Tract Symptoms in the Pediatric Population

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Lower urinary tract dysfunction without neurogenic origin, which usually present with nocturnal / diurnal wetting and urinary tract infections, is a common clinical entity in pediatric urology, with a reported prevalence of 2% to 7% in children ages 4-7 years. When lower urinary tract symptoms are minor, parents and children may not find it worthwhile for a consultation. However, when the wetting and other lower tract symptoms including urinary tract infections become bothersome, it turns into an important clinical problem and require further investigation and treatment. Although the pathophysiology and presentation of this clinical entity are well described, there is not yet a generally accepted method of quantitative and standard evaluation of clinical symptoms. Since the nature and the extent of such symptoms vary greatly, such evaluation would be very useful to quantify the severity and to define the nature of the symptoms in a standardized way and will be helpful to monitor the response to treatment. Since early 2000 we aimed to investigate and attempted to develop a questionnaire to measure the severity of lower urinary tract symptoms using scientifically validated methods.

We empirically designed a symptom scoring system - a questionnaire - that was composed of items regarding daytime symptoms, nighttime symptoms, voiding habits, bowel habits and quality of life. The initial study compared two groups of patients using this questionnaire: Group 1 was composed of 86 patients who were seen in the outpatient clinic with complaints of day and/or night time wetting, and Group 2 was asymptomatic normal primary school students. Both groups were asked to fill out the questionnaire and in Group 1 all patients underwent diagnostic testing, such as video-urodynamic studies. The questionnaire was designed primarily for parents and parents were asked to fill it in with their children.

For each question within the questionnaire, odds ratios have been obtained by comparing the replies from parents of patients (group 1) with the normal group (group 2). The value of odds ratio was used to evaluate the strength of each question to differentiate the patient population from the normal group. According to the odds ratio, each question has been given a score ranging from 1 to 3 and the total score of the questionnaire has ranged from 0 to 35. The questions with low odds ratios were considered to be inadequate to differentiate and removed and the total items were modified to 13 questions and 1 quality of life question at the end of the study. Among the 86 patients in group 1 mean score was obtained from the questionnaire was 18.56 while among the 265 controls in group 2 the mean score was 2.88. At the end of the evaluation, within the confidence interval of 96.2%, the patients with threshold scores of 8.5 or greater (not including quality of life score) had clinically significant lower tract dysfunction with a sensitivity of 90% and a specificity of 90%. When mean symptom scores for different sex and age groups were investigated, we were not able to find a marked difference between groups. Therefore, we have not only validated the questionnaire but also found that it could be used for all gender and age groups.

We have also used the same questionnaire for the siblings of the children diagnosed with lower urinary tract dysfunction. While the mean score of the patient group (males n: 26 and females n: 40 / average age 7.7) was 22.7 for females and 28.1 for males, the siblings (males n:42 and females n: 36 / average age 9) have received a score of 4.02 and 3.34 respectively. Although slightly higher than normal controls siblings also scored within the normal range (below 8.5: the cut-off point). Thus, this questionnaire has been validated once again. This time the same questions were actually asked to the same parents and this helped to reduce the error of interpretation by different parents.

Another questionnaire has been developed and being used by the Toronto group. This is mainly a modification of the International Prostate Symptom Score (IPSS) for adult benign prostatic hyperplasia pa-
tients to evaluate their lower tract symptoms. They have used a similar approach in validating their questionnaire and tested on different patient groups. We found this questionnaire a little more difficult to interpret by the child age group as they are asked to quantify their symptoms as half the time or more than half the time. In our questionnaire the majority of the questions are much simpler and the replies are generally in a yes or no format.

We have recently compared the questionnaire with the results of two day voiding charts and looked at the concordance of two investigations. The study included 40 children (31 girls and 9 boys; average age 7.2) presenting with various lower tract symptoms. The results of the questionnaire and voiding charts were compared and we have found the results defining voiding frequency and urgency were highly concordant while the results defining severity of wetting and night time symptoms were not concordant. The results of questionnaires were more sensitive for monitoring the response to treatment. In that sense the questionnaire did not actually give all the information that one can receive from voiding charts, yet we think that it could be used as a very valuable and more practical adjunct.

The first step in the management of patients with voiding problems should be the stratification and grading of the clinical problem. Therefore, the history of the patient should be taken in a structured way. Selection of the therapeutic approach and assessment of the response to the treatment may be done by questionnaires. Further detailed and invasive investigations must be reserved for selected and more complex patients. The scoring systems have the ability of obtaining a systematic patient evaluation, grading the severity of symptoms, assisting for the probable treatment decision and the monitoring the response to treatment. The questionnaires may also be useful standard tools for clinical research in lower urinary tract dysfunction and reflux. We have been able show that this kind of a symptom scoring system, with the 90% sensitivity and specificity it possesses, can be created and is feasible as it is in the adult practice such as IPSS for lower urinary tract symptoms.

Further reading:

Lower Urinary Tract Reconstruction - Some Reflections

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The Appendix

It is accepted that intermittent catheterisation per urethra is the most problem-free method of emptying an abnormal or reconstructed bladder. It is also accepted that the appendix is the “best channel” for intermittent catheterisation through an abdominal wall stoma into the abnormal or reconstructed bladder. Other tissues have been utilised but in the long term they are specific problems of increased elongation and flexibility; both these issues are creating difficulty with catheterisation. There is accepted variety internationally concerning the position of the appendix channel, viz the abdominal wall stoma may be in the right iliac fossa or may be placed at the umbilicus.

There is a possibility that catheterisation through the umbilicus may not provide reliable complete bladder emptying and may lead to increased urinary infections or calculi. There were initial problems with protrusion of the appendix channel at the abdominal wall stoma. This has been rectified by ingenious abdominal wall skin flaps and the principle utilised reconstruction is the “VQZ” stoma.

There are some variations to these particular skin flaps – none have been demonstrated to be an improvement on the initial technique.

The appendix has also been used as a channel to perform large bowel washouts. The procedure described disconnecting the appendix from the caecum, maintaining its blood supply and reconnecting the channel to the caecum in a non-refluxing manner. This technique has been modified and in most institutions, the appendix is mobilised onto the abdominal wall by laparoscopic or open techniques without formal disconnection from the caecum and reversal of position. This does not appear to have created problems of faecal or gas leakage, but it will remain to be seen if this becomes a long term problem. The appendix now may be positioned within the ascending colon or if the bowel pathology is mainly in the descending element, it may be positioned in a manner to create only a partial bowel washout.

Bladder Augmentation

Bladder reconstruction mainly with small bowel is now a routine procedure. The complications are well documented – mucus production, urinary infection, bladder calculi and possible electrolyte imbalance. The long term issue with the use of small bowel and other parts of the gastrointestinal tract such as colon or stomach is cancer. The number of patients affected within a period of 15-19 years after reconstruction is very small. The question which has not yet been answered is what to do as routine surveillance to detect cancer early and when to commence routine surveillance.

Bladder enlargement, as stated, is commonly performed with intact bowel. Proposals to provide alternative solutions are:
1. Remove the mucosal lining of the bowel and place on it patches
of urothelium harvested from the bladder or place the demucosalised bowel on in situ urothelium after division of the detrusor muscle.

2. Create a patch of tissue with cells obtained from the in situ bladder or stem cells and grow these on a biomatrix scaffold which may consist of varying structures – collagen, elastin, polylactic glycolic acid.

3. Grow in the laboratory a “bladder” consisting of 2-3 layers.

The treatment options have not yet replaced the common use of intact bowel, created into a non-peristaltic patch or pouch and placed onto the bladder.

The Bladder Outlet

Bladder neck reconstruction remains a most challenging operation. Many procedures have been described and outcomes vary. In the bladder extrophy group, the results are related to several variables which include (a) the type of operation, (b) the type of tissue available to reconstruct, and (c) the status of the bladder which is to be the container. A new approach is evolving which challenges all the previous operations by performing increased mobilisation of tissues with release of the corpora and the crura from the inferior pubic rami. There must be meticulous attention to preserve the neurovascular bundle supplying the penis. This provides increased mobility of tissues but the question remains as to whether the function of the outlet mechanism will be improved and there is concern about the performance of the organ which has been disconnected from the pubic rami.

Within the area of bladder neck reconstruction, a place of particular interest to me is how the anatomy can be exposed more clearly, as in most operations it is a struggle to work on the bladder neck behind the symphysis pubis. Therefore I raise the possibility of incising and opening the symphysis pubis to permit improved exposure. There has been no functional problem with the pelvic ring in the small number of patients on whom I have used this technique.

The artificial sphincter has had popularity in the paediatric age group in the 80s and 90s. However, the insertion of this device is now not performed so commonly in most paediatric urological units. It is possible that many paediatric urological surgeons come from the paediatric surgical background and have not, in their training, been exposed to this procedure. Insertion of this device requires very specific expertise and in many adult urological units the sphincter is not inserted into patients until after puberty.

In the neuropathic bladder with abnormal sphincter, there has been renewed interest in urethral dilatation. This was a procedure used in the adult population for several decades. There is a possibility this simple procedure in the female neuropathic lower urinary tract may improve bladder compliance and capacity with subsequent decreased dilatation and vesico-ureteric reflux. There are not many publications concerning this issue.

Thank you for allowing me to give you some reflections on developments within the field of paediatric urology and I wish Professor David Thomas, who has made a major contribution to paediatric urology in the clinical arena, scientifically and, within the UK, politically to the development of paediatric urology, ALL THE VERY BEST IN HIS RETIREMENT.

Endoscopy

Endoscopy of the urethra and bladder is a common procedure. Endoscopic surgery of the abdomen for the “older” paediatric urologist is a new approach to urological abnormalities of the upper and lower urinary tract. However, laparoscopy will continue to replace open surgery. The development to review is whether the robotic machine can be modified further to enable increased use in the paediatric patient.

Posterior Urethral Valves

The posterior urethral valve patient is generally being managed very well in the modern paediatric urological era. However, there is, and if there is not, there should be, controversy concerning the management of the “valve bladder” in utero. There is a reluctance to aggressively approach the valve bladder in utero when function, as indicated by normal volume of amniotic fluid, and moderate dilatation of the upper tracts would suggest at the early stages a safe situation. It is postulated that if there is a reliable diagnosis of obstructive posterior urethral valves, a large bladder, with dilatation of the upper tracts and a normal amniotic fluid detected at 18-20 weeks, it is at this stage the obstruction should be relieved by in utero insertion of a suprapubic catheter. This has not been widely accepted and the treatment options remain: observation with obvious deterioration of renal function, or insertion of a suprapubic catheter in utero at a time when kidney function has clearly deteriorated and has now been shown not to improve. A trial of early insertion of a suprapubic tube will only take place when paediatric urologists actively participate in the management of these patients within the scenario of a large obstetric unit and regular urological antenatal clinics. The lack of progress to test the “early” treatment is related to the natural hesitancy of obstetricians to interfere in a pregnancy when apart from dilated tracts, there is no other obvious problem with the foetus.

Botox

Botox is an interesting chemical now being inserted into the bladder to improve bladder function. It has gained great popularity in the adult world and is being used in paediatric patients. Early reports indicate it has an initial good effect. However, the effect is temporary and there is a need for repeat injections. Only the long term results will indicate whether repeat injections will have the same effect. The technique is non-invasive with minimal early complications.

Thank you for allowing me to give you some reflections on developments within the field of paediatric urology and I wish Professor David Thomas, who has made a major contribution to paediatric urology in the clinical arena, scientifically and, within the UK, politically to the development of paediatric urology, ALL THE VERY BEST IN HIS RETIREMENT.
The Valve Bladder

It was a pleasure to be asked to participate in the COPU 2010 Course and the academic day honoring the pediatric urologic career of David Thomas. David’s contributions to pediatric urology involved all aspects of our discipline. It is said, when David Thomas spoke we listened.

Congenital obstruction due to posterior urethral valves was first described by Hugh Hampton Young in a report of a 17 year old boy who had problems with voiding since birth. He stated in a publication in the Journal of Urology in 1919, “The upper aspect of the verumontanum was bifurcated and from it were continued two thick folds of tissue…Considerable pus was seen issuing from this.” He subsequently reported 12 cases; age ranging from 11 days to 42 years which he added to 23 previously reported cases in the literature. Of those 23 cases reported in the literature, 22 were post-mortem and 1 was treated by catheterization. None of the previous 23 reported cases had the valve treated directly. Early in the evolution of our thinking on posterior urethral valves we concentrated on the effects of the bladder outlet obstruction on renal development and focused on hydrenephrosis, reflux, and congenital renal dysplasia. D.I. Williams was one of the first to be concerned about the persistence of hydrenephrosis after valve ablation. He reported on 35 cases from Great Ormond Street, 14 of which he had operated on, and noted that besides the persistent dilation following valve incision, many of these patients had reduced sensation of bladder fullness. As he reported in the British Medical Journal in 1954, “the bladder may be atonic….and if free reflux into the bladder leading to urinary incontinence. In addition, the abnormal reflux is that of nocturnal bladder drainage. He theorized that when the bladder was refilled to nearly its pre-void volume. This would be a cycling phenomenon. The term “pseudo residual” urine is urine that was in the bladder following refilling from reflux urine, where as pseudo-pseudo residual is that of refilling of the bladder from dilated upper tracts in the absence of reflux. Nocturnal catheter drainage, however, would modulate the upper tract dilation by keeping the bladder

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The debate raged in the literature for some time trying to define whether the long-term outcome of boys with posterior urethral valves was due to two independent variables that of bladder function and that of kidney function, or were these variables somehow related.

The prevailing theories of late onset renal failure include the idea that children outgrow their renal reserve and the medical demands of puberty are not able to be met by their kidney function. However, deterioration of renal function will often precede somatic growth. Nephron hyperfiltration injury has also been implicated; however, the association with renal failure has been quite vague. Finally reflux and renal damage in the absence of infection we know requires elevated bladder pressures. Ghanem and associates reported 116 children with posterior urethral valves who had urodynamic evaluations. Thirty-five percent had functional renal impairment. Eighty percent of these patients had abnormal bladder dynamics including poor compliance, detrusor overactivity, or a combination of both of those with normal voiding dynamics 4 of 23 went on to renal failure. Here again was evidence that bladder dysfunction is correlated with renal failure, however, normal bladder dynamics does not necessarily preclude renal deterioration.

Steve Koff introduced the concept that if the bladder is completely empty for a prolonged period of time, the associated hydrenephrosis and potential effect on renal function might be lessened. His concept was that of nocturnal bladder drainage. He theorized that when the bladder emptied in the children, there was significant residual urine in the upper tracts, and, therefore, within several minutes after voiding the bladder was refilled to nearly its pre-void volume. This would be a cycling phenomenon. The term “pseudo residual” urine is urine that was in the bladder following refilling from reflux urine, where as pseudo-pseudo residual is that of refilling of the bladder from dilated upper tracts in the absence of reflux. Nocturnal catheter drainage, however, would modulate the upper tract dilation by keeping the bladder
empty for an extended period of time. This management was shown to be advantageous in other reports by Fumo and Nguyen.8,9

Glassberg argued, however, that nocturnal bladder drainage is not necessary if the valve bladder can be prevented with the use of anticholinergic and selective intermittent catheterization in a proactive manner.10 Work by Holmdahl showed us that bladder dynamics following posterior urethral valve ablation is in constant evolution.11 There is a progressive decrease in bladder instability over time with a successful increase in expected and mean bladder capacities. As time goes on the population of children with previously treated valves will be unable to sustain a voiding contraction and require straining in order to empty their bladders. This represents the myogenic failure component that one sees in valve bladders.

If we look into the future, therefore, rather than just simply concentrating on the effect of bladder outlet obstruction on kidney function, one must be cognizant of potential bladder effects on kidney function secondarily. Addressing these with aggressive management is the potential key to maintaining intrinsic renal function as long as possible. This can be done by using early urodynamic evaluation in addition to routine follow-up ultrasound and VCUG. Urodynamics at about a year of age, maybe helpful in predicting those bladders that will go one to develop noncompliance and eventually myogenic failure. A combination of early anticholinergic therapy with selective intermittent catheterization or nocturnal bladder drainage maybe the key to preventing valve bladders of the future.

Although there is no direct evidence at this time, one might wonder what the potential effect of a temporary vesicostomy or in-utero shunting of the bladder may have on development of normal bladder dynamics postnatally. There has been some evidence in the literature by Close et al. that primary valve ablation may be advantageous to early urinary diversion in improving bladder compliance and capacity as cycling of the bladder and work by the bladder is thought to be necessary for normal bladder development.12 Similarly work by Youssif et al. indicates that early valve ablation results in better bladder compliance and reduces instability which translates to easier toilet training and long-term continence.13 They noticed a reduction in bladder dysfunction as well as improved continence compared to children who were ablated greater than a year of age.

References

Bladder Neck Repair for Incontinence: The Road Ahead

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Bladder outlet insufficiency (BOI) in a paediatric urology practice commonly occurs in two primary groups of children: one with neurogenic bladder dysfunction such as spina bifida or dysraphism and the other with structural defects of the sphincter complex such as extrophy-epispadias complex.

Philosophy
The aim of management in children with BOI is to achieve social continence with voiding at will whilst preserving the upper tracts. There are a number of techniques to achieve continence depending on the surgeon’s choice dictated by his or her experience and technical ability. There is no clear favourite in terms of the procedure performed which illustrates the fact the ideal procedure is yet to be discovered. The real issue is that these procedures are often not performed independently partly due to coexisting bladder dysfunction in terms of compliance or capacity needing bladder augmentation and partly due to the risk of unsettling the bladder dynamics from the increased outlet resistance, in terms of storage at low pressure and voiding to completion. In the latter instance, however, this cannot be accurately predicted preoperatively and, therefore, most surgeons embark on adding a drainage procedure such as Mitrofanoff channel for CIC along with augmentation especially if they have performed an invasive bladder neck repair. Possibly a minimally invasive approach may alter this approach giving the surgeon a chance to evaluate bladder dysfunction and voiding once outlet resistance is increased.

Procedures
The range of procedures commonly performed fall into four groups:
- Slings - creating extrinsic resistance around the bladder neck
- Bladder neck repairs - formal repairs from within the bladder
- Bladder neck injections
- Bladder neck closures.

The first group classified as slings include fascial slings or SIS; artificial sphincter cuff and colposuspension. Bladder neck repairs include Salle procedure, Kropp–Salle procedure, Young – Dees repair, bladder neck wrap and Keel repair, although there are various modifications within the same theme of using bladder tissue from within the bladder neck. Bladder neck injections are performed either by the antegrade or retrograde technique and the materials used can vary from...

(continued on next page)
detransomer - hyaluronic complex to collagen or macroplastique. Bladder neck closures should be the last resort for intractable incontinence, but this procedure does have a place in management.

**Interpretation of Outcome Data**

The real issue is to assess the outcome data and make comparisons of the published literature. The lack of standard definition of success coupled by the fact that these procedures to correct incontinence are rarely isolated and are accompanied by a fairly high rate of concomitant bladder augmentation. There is no isolated bladder neck procedure described yet that can leave the patient dry for at least 4 hours with no detrimental effect on bladder volume and dynamics and thereby preventing damage to upper tracts by allowing a pop-off mechanism should bladder pressure rise. Other secondary characteristics for the ideal procedure are that it is technically simple to perform and train with long-term reliability and allow for easy catheterisation.

**Current Status**

**Catheterisable Channel (CC):** A CC is almost always performed as is recommended in literature to provide an alternate channel for safe bladder emptying and irrigation following bladder neck repairs or slings.

**Bladder augmentation (BA):** The fact, as evidenced in literature, is that the vast majority of children who have had bladder neck repairs or slings have had concomitant BA. Very few cases have had bladder neck repairs alone. Notable exception is the study by Snodgrass et al where they performed a 360 degree fascial sling with CC in 30 neurogenic bladders with low leak point pressure and stress incontinence with an initial success rate of 83%. They demonstrated that the bladder neck slings can be performed without the need for concomitant BA. However, closer evaluation shows that in the longer term, bladder compliance and capacity is decreased. The Toronto experience with long-term follow-up of isolated bladder neck procedures is not favourable with all needing BA eventually for worsening upper tract dilatation and renal function or incontinence.

Bladder neck closure remains an effective option with concomitant BA and CC in intractable urinary incontinence particularly in the case of poor quality bladders in exstrophy and severely damaged bladders where reconstruction is not an option or has failed on multiple occasions. The risk of bladder perforation or loss of renal function post-transsection procedure fortunately is low as documented in literature.

Bladder neck injections are generally not effective on their own as a primary procedure and are useful as an adjunct to increase the resistance achieved with previous bladder neck procedures. Repeated injections (more than 3 attempts) offer no additional benefit and the overall success rate is about 30% in most series. The CC channel offers the advantage of antegrade injection which seemingly is more effective although this is not proven.

**The Future**

As we look ahead, the challenge is to see if there are procedures on the horizon which may allow a less invasive approach to the bladder neck that allows closer follow-up without CC or BA. Two such approaches are worth mentioning and pursuing.

**Pneumovesicoscopy**

The minimally invasive intravesical approach is attractive in that the bladder neck repair is done with minimal access and the surgeon feels comfortable to not add a CC or BA at the first instance. This is certainly not the case with formal open procedures where the option of redo surgery for a subsequent bladder augment or CC is technically more demanding. The majority of the traditional approaches for bladder neck repair are performed on the posterior bladder wall whereby the ureters may have to be moved. The minimally invasive approach instead allows us to look at the anterior bladder wall thus leaving the ureters alone. I personally have switched to using the anterior bladder wall for bladder neck repair (Keel) in open surgery with CC and no augment for some time now with encouraging initial results (data analysed comparing anterior and posterior Keel repair and manuscript in preparation). The added advantage is that the sharp angulation provided by the repair improves the chances of success with continence. A number of girls in our series are voiding comfortably per urethra whilst being dry and using the CC to ensure adequate bladder drainage.

**Male Advance Sling**

In boys, The Male Advance Sling is an attractive option. This sling can be placed in as a day case procedure via the perineum and the boys followed closely with bladder dynamics and assessment of renal function and upper tracts. The advantage is that the access to the bladder and neck is untouched and allows further procedures in case of failures with the added benefit in that it allows easy self catheterisation through the native urethra.

The sling works as a hammock in peripubertal boys by elevating the bulbar urethra to the pelvic bone via the obturator foramen. The procedure has been successfully used in adult patients for incontinence post radical prostatectomy. The adult urologists say that this works only in patients with some residual sphincter function and by elevating the bulbar urethra, the sphincter complex is restored into its normal location to function effectively.

The Ghent experience (presented at the ESPU 2009 annual congress) suggests that it is effective in neurogenic bladders in children as well and possibly explains the more logical mechanical effect and residual function in the sphincter may not be necessary after all. I have started using this approach for peri-pubertal boys with sphincter deficieny and am encouraged with the initial experience. We will have to wait and see if this stands the test of time.

**Further Reading:**

David Thomas has been one of the leading and most highly respected Paediatric Urologists in the UK and internationally over the past 30 years (Figure 1). His contributions are enormous but one of his most significant legacies is the large number of Paediatric Urologists he has trained, who are now the leaders and opinion formers of our specialty. It is fitting that we are discussing another giant of Paediatric Urology, Paul Mitrofanoff, whose contribution is 30 years old this year, to celebrate David’s career.1

In 1972 Jack Lapides revolutionised the management of the neuropathic bladder with the development of clean intermittent catheterisation (CIC). Mitrofanoff made the application of CIC available to countless numbers of new patients with the development of a continent catheterisable abdominal conduit, The Mitrofanoff Procedure, which has stood the test of time over the past 30 years. There have been many modifications during this time and it is timely to examine what approach now represents “The ideal Mitrofanoff.”

The Conduit

Virtually every native tubular structure and many reconstructed tubes from multiple native tissues have been utilised over the years. However, there are only 2 that are regularly used in 2010, appendix and Monti tube reconstructed from small bowel.

A review of the published literature demonstrates continence rates of 90%-96% after the original procedure, rising to 98% after a single revision. There are no differences between the appendix and Monti in terms of continence. However, it is this author’s experience that with longer follow-up, catheterisation difficulties do develop in the Monti conduits, particularly if the longer spiral Casale technique is used.2

Therefore, current evidence would suggest that the appendix, single or split (if a simultaneous ACE is required) is the conduit of choice for the Mitrofanoff, but the Monti is a reasonable alternative if the appendix is not available.

How to connect it to the reservoir

There are a number of options here. The conduit may be implanted into the native bladder or the bowel segment augmenting the bladder. The implant may be performed intra- or extravescically, into the anterior or posterior aspect of the reservoir or high or low in the bladder. There is a single study examining continence rates with implantation into the bowel segment.4 This reported a continence rate of 87% (with an 18% revision rate). This does not compare favourably with the 90%-96% continence rates when the conduit is implanted into the native bladder.

There are no reported differences, in terms of ease of catheterisation, whether the conduit is implanted into the native bladder or augmenting bowel segment.

A single study has suggested that there is a significant increase in the incidence of urine infection and an insignificant increase in the incidence of bladder stones when the conduit was placed anteriorly in the bladder as opposed to a posterior placement.5 It was speculated that this could be as a result of increased difficulties in evacuating mucous.

Although there are no reports in the literature, it is this author’s experience that pain on catheterisation is a significant problem in senescent patients if the internal orifice is near the trigone. In this group it is probably best to site the conduit high in the bladder away from the trigone.

The stoma

There are a number of options: anastomose the enteric mucosa directly to the skin, tubularise a V-flap and bury the muco-cutaneous junction or employ a multi-flap technique to construct the stoma (VQZ/VQC). There is also the choice of placing the stoma in the umbilicus or on the abdominal wall. When the mucosa is anastomosed directly to the skin there are very low stenosis rates but the stoma is unsightly and clothing is stained with mucous discharge (Figure 2). The results reported suggest there are no differences in stoma outcome between the appendix or Monti conduits.

One study reported a stenosis rate of 45% for the tubularised V-flap, 24 % for the umbilical stoma and 0% for the VQZ stoma.6 Other studies do not match these results but evidence still suggests that stenosis rates are lowest when multi-flap stomas such as the VQZ are used.7 Another advance is the use of the ACE Stopper (Medicina, UK) for up to 6 months following surgery (Figure 3). In this author’s experience this significantly reduces the incidence of stoma stenosis.
The Ideal Mitrofanoff

The appendix implanted into the posterior wall of the native bladder with the internal orifice away from the trigone and the stoma constructed using a multi-flap technique followed by the use of an ACE stopper for 6 months post-operatively is the best combination for success.

After 30 years of the Mitrofanoff it can be stated that that it works, 95% of patients continue to use it long-term, although up to 50% have required some form of revision surgery. It would be fair to predict that the Mitrofanoff will still be here in 30 more years.

“Vive Mitrofanoff”.

References

Figure 2 - Unsightly stoma with exposed mucosa.

Figure 3 - The Medicina ACE stopper.