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The Valve Bladder Syndrome

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Posterior urethral valves (PUV) are the most common cause of obstructive uropathy involving the lower urinary tract in children. Although endoscopic valve ablation is effective in resolving the anatomical obstruction, the sequelae on the bladder and kidneys after valve ablation remain (and sometimes deteriorate) during childhood and through adolescence. In 1988 Parkhouse *et al* reported that the prognosis for renal function is poor, with a third of the patients developing renal failure by early adulthood.[1] The etiology of this late onset renal failure remains unclear but urinary tract infection, nephron hyperfiltration, persistent vesicoureteral reflux, puberty and, probably most important, bladder dysfunction have been suggested as possible mechanisms.[2] Bladder dysfunction reported in upto 75% of boys with posterior urethral valves is established during gestation as a response to urethral obstruction and remains throughout childhood and adolescence even when obstruction is removed during early infancy.[2,3,4]

It has been nearly 30 years since Mitchell coined the term valve bladder syndrome to emphasize the relationship of altered bladder function, progressive hydronephrosis and renal function deterioration. This term describes clinical findings that can continue and progress long after an obstruction caused by posterior urethral valves has been relieved. The features of this syndrome include the persistent dilation of the upper urinary tracts, a thick-walled, noncompliant urinary bladder, urinary incontinence, and polyuria secondary to nephrogenic diabetes insipidus.

Pathophysiology

In general, bladders affected by valve–bladder syndrome are found to be thickened, poorly compliant and hypertonic. Histologic studies of these bladders have shown an altered ratio of collagen to smooth muscle deposition, which determines the compliance of the bladder.[5]

The fetal bladder is formed by the time of 21 weeks gestation, but cycling is required in order for it to mature and refine its cellular make-up. As the bladder begins to store urine, the ratio of collagen to smooth muscle in the bladder decreases, along with the ratio of type III to type I collagen, thus increasing the storage ability and overall compliance of the bladder.[5]

In obstructed bladders, the overall amount of collagen and muscle increases via both hypertrophy and hyperplasia, in an attempt to preserve function via increased compliance and contractility. If obstruction is prolonged, however, the bladder will decompensate, as fibrosis decreases compliance and myogenic failure leads to reduced contractility. In valve–bladder syndrome, the bladder outlet obstruction is alleviated by surgery, but the pre-existing molecular changes in the detrusor remain, and continue to cause bladder dysfunction.

Renal function in the neonate varies greatly, according to the degree of prenatal bladder obstruction in the fetus. It is important to remember that the kidney is instrumental not only in toxin clearance, but also in body-fluid homeostasis. Bladder obstruction distorts or alters homeostasis by causing a lifelong concentration defect or nephrogenic diabetes insipidus. Severe obstruction leads to impaired medullary development with a paucity of collecting-duct formation, further contributing to the development of nephrogenic diabetes insipidus. The concentration defect can be profound (especially in children who survive severe congenital obstruction), with urinary outputs of several litres per day. A renal concentration defect can be present at birth, or be seen later, as the first sign of renal insufficiency. The large volumes of urine produced as a result of the renal concentration defect alone would only cause frequent urination, but in association with a bladder that has poor sensation, contractility, and compliance, these large volumes of urine can lead to a cycle of chronic bladder overdistention, and valve–bladder syndrome. By these varied means, the upper and lower urinary tracts influence each other long after the obstruction has been alleviated.[6]

Effect of Primary Therapy on Bladder Function

The effect of primary surgical treatment (fulguration vs diversion) on subsequent bladder function continues to be debated. Differing opinions exist regarding the effect of defunctionalisation of the bladder.

In 1952 Veenema *et al* observed that suspension of vesical function by diversion produced a small capacity bladder. Nesbit observed 11 years later that vesical hypertonicity resulted from the bladder being "a prolonged empty space."[7] In 1969 Schmaelzle *et al* reported that soon after supravesical diversion in dogs bladder capacity was reduced to 12.5% of pre-diversion capacity. However, once the dogs were undiverted capacity returned to 88% of pre-diversion levels.

Duckett, Tanagho, and Close *et al* all suggested that the dysfunction present in the bladders of some patients with PUVs is more frequent and severe in those children who were defunctionalized in their first months of life by vesicostomy or ureterostomy after they had not responded well to valve ablation.[8] These authors all agree that collagen will infiltrate a defunctionalized bladder, causing it to contract and making later rehabilitation difficult. Nevertheless, there are studies showing that normal bladders do regain normal function after a period of defunctionalization.

In 1996, Smith *et al.*, based on a study of 100 patients with PUVs, of whom 74% underwent valve ablation, 13% vesicostomy, and 9% high diversion, argued that high diversion produced bladders with a smaller capacity and worse compliance. Close *et al.*, in a study of 31 patients with PUVs (eight underwent high diversion and 23 valve ablation), 19 of whom had had urodynamic studies, concluded that the bladders of patients initially treated with valve ablation had a larger capacity than did those of patients treated with high diversion, but the number of urodynamic studies was somewhat limited in this study.[8] Podesta´ *et al* found that those patients treated with vesicostomy for a mean of 34 months before closure had bladders that were smaller and less compliant after closure than those after primary valve ablation alone.[9]

Unlike the authors of all these arguments, Jayanthi *et al.*, Khoury *et al.*, and Kim *et al.* found that neither vesicostomy nor ureterostomy have negative effects on later bladder function, but their conclusions are also affected by the limited number of patients in their studies.[10] In addition, Jaureguizar *et al* compared 2 groups of patients with valves who had a similar status before initial therapy of cutaneous pyelostomy or ureterostomy or primary valve ablation. They found no significant difference in bladder function between these 2 treatment groups and concluded that neonatal supravesical urinary diversion had no adverse effect on bladder function. They concluded that poor bladder function is probably a consequence of detrusor damage in utero and has little to do with the mode of primary treatment. Puri *et al* correlated bladder dysfunction with the initial surgical treatment in 67 patients with PUV and concluded that primary valve ablation is associated with better bladder function than vesicostomy and should be the treatment of choice in PUV.[11]

However, the problem with comparing the primary ablation group with the diversion group in all such studies is the assumption that the diverted group started out worse than the primary ablation group. The patients who require high diversion definitely have a more severe form of disease with greater changes in the bladder and upper tracts at presentation. The outcome in these patients, therefore, cannot be compared.

Bladder Dysfunction

The reported incidence of bladder dysfunction in patients treated for PUV is 13%–38%.[12,13,14] Persistent bladder dysfunction has been implicated as a cause of deterioration of the upper urinary tract and kidney function, and this has lead to the urodynamic investigation of boys with a history of PUV as part of their regular followup.

Urodynamic studies have been used to categorize bladder dysfunction into three patterns: hyperreflexic (unstable) and hypertonic (non compliant) and myogenic failure (over distended). These three patterns have a prevalence of about a third each, but also significantly overlap. Bauer *et al* reported their findings in 8 boys with a history of posterior urethral valves who had abnormal voiding and found that only 1 had a normal urodynamic study. They divided the bladder findings into myogenic failure, high voiding pressure, uninhibited contractions and small capacity. Peters *et al* reviewed the urodynamic findings of Bauer *et al* in 41 boys with valves (35 studied for incontinence, 3 for frequency, 2 for hydronephrosis and 1 for urinary tract infection). They divided abnormal urodynamic patterns into the 3 types of hyperreflexic, small hypocompliant and myogenic failure. Parkhouse and Woodhouse reported urodynamic studies in 42 consecutive boys after valve ablation, of whom 75% had abnormal studies.[2] They used the corresponding terminology of instability, hypocompliant and acontractile. It is not clear whether the terms myogenic failure and acontractile bladder are synonymous.

Although the three patterns of bladder dysfunction overlap considerably, myogenic failure, with true bladder atony or unsustained voiding contractions (hypocontractility) seems to be the predominant pattern.

Therefore, it has been suggested that myogenic failure is the likely end-stage of bladder dysfunction in adolescent boys with PUV. Holmdahl *et al.* correlated the patients' age with these three classical patterns to

determine whether bladder dysfunction changed during infancy, childhood and adolescence. They suggested, in the first urodynamic study reported in infants with PUV, that the fairly uniform pattern of initial hypercontractility and low bladder capacity may change during the first year of life, with resolving hypercontractility and increasing bladder capacity. Subsequently, the same authors reported urodynamic studies in boys with PUV aged <15 years, comparing them with those in postpubertal patients.[15] They found patients had a changing urodynamic pattern with instability decreasing with time, increasing bladder capacity and commonly an unsustained voiding contraction causing emptying difficulties. Postpubertal boys had high capacity bladders with low contractility. Based on these observations, Holmdahl *et al.* suggested that the three urodynamic findings are variations of the same basic pattern that change with time, toward detrusor decompensation. De Gennaro et reported similar results and concluded that the early detection of 'covert' hypocontractility and the possibility of early bladder rehabilitation might be helpful in preventing emptying difficulties secondary to functional obstruction, and the progression of hypocontractility to patent detrusor myogenic failure and overdistension after puberty.[16,17]

These results seem to confirm that bladder dysfunction in boys with PUV changes from the unstable/hypercontractile bladder found in infants to hypocontractility in childhood, which may deteriorate with age, leading to a true myogenic detrusor failure after puberty. Other factors, *e.g.* the high renal output caused by renal impairment and the development of the prostate gland, may contribute to the bladder overdistension.

Incontinence

Incontinence is seen in 19% to 81% of 5-year-old boys with a history of a posterior urethral valve. It was formerly assumed that incontinence in these patients was the result of some injury to the external sphincter, produced either after valve ablation or during sectioning of the bladder neck.

The first to suggest that urinary incontinence might signal bladder dysfunction in these patients was Bauer *et al* who noted abnormal urodynamic parameters in those boys with incontinence. Glassberg *et al* found that all patients with persistent upper tract dilatation to be incontinent, which they believed was secondary to the combination of loss of compliance and a large urine output secondary to acquired nephrogenic diabetes insipidus.[18]

Johnston and Kulatilake found that incontinence always disappeared with puberty and believed that the incontinence disappeared secondary to growth of the prostate as it filled the widened canal.[19] Smith *et al* found that incontinence in boys with valves decreases with age, being present in 81% of 5-year-old, 46% of 10-year-old and 1% of 20-year-old patients. Holmdahl *et al* thought that instability was the major cause of the incontinence and that incontinence decreases with age because instability decreases with age. Incontinence was not noted in the postpubertal boys.They also found that compliance also improves with age and often over improves with capacity increasing on average to twice normal. In some patients myogenic failure develops as well. Boys who were incontinent when younger were those most likely to have myogenic failure when older. DeGennaro *et al*, who also observed the bladder in boys with valves as a function of increasing age, also found that bladder capacity and bladder compliance improve with age.[20] They believed that myogenic failure represents decompensation secondary to chronically elevated pressures when patients are younger.

Boys with a history of PUV also have poorly sensate bladders. This theory originated from the observation that boys with PUV can unknowingly have distended abdomens, and be in a state of overflow incontinence. A further complication is the common finding of polyuria secondary to the inability to fully concentrate their urine. Thus, these boys will often have no knowledge of their full bladder and dilated upper tracts, which, even with timed voiding, will fill quickly because of both an elevated PVR and copious urine production, and require frequent voids.[6]

Fertility and Sexual Function

The information about fertility and sexual dysfunction in men born with posterior urethral valves is sparse. Woodhouse *et al* concluded that paternity is possible,[21] which is in agreement with the findings of Holmdahl *et al.* They concluded that the ability to father children appears to be more dependent on renal function and not the valve as such, as none of the men in their study with ESRD had children, while the same thing applied to all but 2 of the men without uremia.[22]

Persistent Hydroureteronephrosis

Persistent upper tract dilatation on followup investigation has been of concern.[23] Lyon suggested that persistent upper tract dilatation was secondary to large urine output, a result of renal tubular damage. Of 22 boys two-thirds were unable to concentrate urine to a specific gravity of greater than 1.013. Lyon believed that these children were at greatest risk from persistent HUN, especially without diversion. He believed that dilatation impeded kidney function as well as individual growth rates.

Persistent upper tract dilatation in patients with PUV following adequate valve ablation should not be dismissed as merely the residual stretching of a previously obstructed system. Most if not all patients with persistent HUN have associated abnormal bladder dynamics when carefully evaluated. The high intravesical pressure generated by some of these dysfunctional bladders, particularly in those with poor compliance, will distort the anatomy of the trigone and the ureterovesical junction and may result in obstruction and reflux.[24] Clinical and experimental studies have demonstrated that elevated intravesical pressure makes urine drainage at the level of the ureterovesical junction more difficult.[25] Polyuria, secondary to impaired urinary concentrating capacity, can significantly affect upper urinary tract and renal function. In the presence of decreased bladder compliance, the increased urinary volume associated with polyuria can increase hydrostatic pressure on the kidneys and lead to progressive renal injury. The same occurs in boys with PUVs and massive vesicoureteral reflux, which will produce constant bladder refilling with large amounts of refluxed urine. Urinary stasis and increased risk of urinary tract infection would accompany this situation of prolonged dilatation and poor drainage at the level of the ureterovesical junction. These ideas would explain the persistence of upper urinary tract dilatation that exists despite adequate valve ablation.

Long-term Renal Function

Nearly 30% of boys with a severe PUV develop renal failure before adolescence. Renal insufficiency (RI) can develop very early in some of these patients, and, in these cases, it is probably the result of renal dysplasia. In other boys, loss of renal function progresses slowly over the years. In these cases, the late onset of renal failure could be at least partially the result of a combination of abnormal bladder function, metabolic demands of puberty, and hyperfiltration injury.[26]

In their study of 22 boys diagnosed with PUV during their first year of life and followed for an average of 5.8 years, Warshaw *et al.* found that over half of their patients had RI at the end of the study. They distinguished two groups of patients: those who developed RI in the first months of life and those who did so in adolescence or later. They concluded that RI was the result of renal dysplasia in the first group, and the triggering factor was bladder dysfunction in the second group.[27] Parkhouse *et al.*, in a study of 114 boys with PUVs, showed that patients over 5 years of age who still suffered diurnal incontinence had worse renal function than those who did not.[1] About 26% of these patients had chronic renal failure and all, except one, had normal renal function during their infancy. Their findings implicate bladder dysfunction as being responsible for the patients' incontinence and worsened outcome. Based on these and other studies, one could argue that the patients' bladder dysfunction was clearly related to their eventual worsened renal function.[28]

Some authors have attempted to relate loss of renal function to a specific urodynamic pattern and, thereby, identify which types of bladder dysfunction could be associated with the worst prognosis for long-term renal function. Nijman *et al.* studied 65 patients with PUVs. They demonstrated a close relationship between some types of dysfunction (instability and poor compliance) and a higher risk of progressive renal failure. In a long-term urodynamic evaluation of 59 boys with PUVs, López-Pereira *et al.* found that 22 were in end-stage renal disease (ESRD); they found a significant relation between types of bladder dysfunction and long-term renal function. In this study, 89% of the patients with poor compliance, 66% of those with myogenic failure, and only 23% of those with instability were in ESRD. The bladders with poor compliance had the worst outcome in terms of renal function as well as the youngest average age for entering ESRD.

Bajpai *et al* reviewed the outcome of renal function in 58 children with PUV. The choice of therapy in each case primary valve fulguration, vesicostomy, or high ureterostomy—was individually decided on the basis of the response to initial catheter drainage of the bladder. The authors concluded that serum creatinine at presentation is not predictive of subsequent renal function, but the values after a period of urinary-tract decompression are prognostically more useful; delay in diagnosis results in a poor outcome of renal function; and for optimal recovery of renal function, the choice of the primary procedure varies from case to case and can be determined by a systematic, stepwise approach (stepladder protocol).

Renal Transplants and Augmentation

PUV can have a profound effect on the bladder and upper urinary tract. Some groups reported that ESRD develops in 13% to 28% of children, requiring dialysis or transplantation. Others reported a 42% rate of ESRD and a 58% rate of chronic renal insufficiency. The 2006 annual report of the North American Pediatric Renal Trials and Collaborative Studies listed obstructive uropathy as the second most common cause for transplantation, accounting for 1,424 of 8,990 transplant cases (15.84%) since 1987.[29]

The results of renal transplant survival in patients with PUVs who progress to ESRD have been mixed. Several studies show a higher risk of renal transplant loss and increased creatinine in patients with vs without a history of PUVs.[30,31] Recent studies revealed no difference in these outcomes.[32] Patients thought to be at highest risk for renal transplant loss are those with lower urinary tract dysfunction. It is unclear whether the initial valve intervention in children with PUVs has an effect on the development of bladder dysfunction and subsequent renal graft failure. If one considers that bladder dysfunction, consequent to PUV, contributes to the slow deterioration of kidney function, it would be logical to think that, if untreated, the same dysfunction, particularly poor compliance, would also affect the outcome of transplanted kidneys.

Reinberg *et al.* compared 18 patients with PUV to unmatched controls to evaluate 5-year transplant survival and renal function. Graft survival at 5 years was 50% in those with PUVs while a vesicoureteral reflux group that went on to transplantation had 73% graft survival and a control group with ESRD due to nongenitourinary causes had 75% graft survival. From this they argued that the effects of the valve bladder may explain these findings since in comparison a vesicoureteral reflux bladder did not have these problems and was similar to nongenitourinary causes of renal transplant survival. Salomon *et al.* compared the outcome of renal transplantation in 66 children with PUVs and 116 with malformation uropathies; they found a statistically significant increase in serum creatinine at 10 years in children with PUVs but not in the control group.[33] Churchill *et al.* reported 5-year survival figures as low as 30% in patients with PUVs. Bryant *et al.* reported no significant difference in graft survival per se, but they did note a tendency toward worse kidney function in patients with PUVs that could eventually result in worse long-term graft survival. None of these authors report using urodynamic studies to diagnose the type of bladder dysfunction, and, consequently, no dysfunction was treated prior to transplantation.

When a bladder dysfunction is treated before transplantation, however, there are no significant differences at 5 years in either graft survival or function with a control group.[34] Fine *et al.* evaluated the impact of surgical approaches to posterior urethral valves on renal transplant survival and compared transplant survival in children with vs without posterior urethral valves. The authors concluded that of children who undergo renal transplantation boys with PUV are not at increased risk for graft failure. The initial treatment mode of PUV does not affect ultimate graft survival after transplantation or the rate of bladder dysfunction. The potential for bladder dysfunction and increased renal graft demise in PUV cases underlines the need for continued long term pediatric urology follow up, and individualized intervention and management. Indudhara *et al* reviewed renal transplantation records from May 1968 to November 1988 and found 54% and 41% 10-year survival outcomes in patients with and without a history of PUVs, respectively ($p=0.35$).

Any patient in ESRD secondary to a PUV should undergo urodynamic studies before being included in a transplant program. By identifying the dysfunction, it will be possible to treat it and thereby avoid any negative repercussions on the transplanted kidney.

Severe bladder dysfunction can be managed safely and effectively with continent urinary reconstruction. There is no doubt that a stable urinary reservoir with adequate drainage is essential for successful transplantation. There are several options including detubularized bowel, ureter, or stomach to perform cystoplasty. Ureterocystoplasty is an alternative when a dilated ureter is available and nephrectomy is performed.

There are, however, several concerns about this augmentation procedure including 1. risk of compromising peritoneal dialysis, common in children with end-stage renal disease; 2. need for clean intermittent catheterization (CIC) in some patients, with the associated risk of bladder rupture; 3. possible complications related to an augmented bladder left empty while waiting for a renal graft; 4. mucus production with potential stone formation; 5. metabolic disorders, primarily hyperchloremic acidosis; and 6. risk of malignant lesions. The importance and frequency of complications suggest that use of this procedure should be restricted to patients who really need bladder augmentation.

Table 1 outlines specific considerations in pre-transplant bladder augmentation. Gastrocystoplasty has an important role in this population. Stomach has been recommended as the preferred bowel segment for bladder augmentation in patients with renal insufficiency. The secretion of acid into the urinary tract offsets metabolic acidosis of chronic renal failure. The favorable metabolic parameters of gastrocystoplasty, namely aciduria, in

addition to decreasing the risk of systemic acidosis also reduces the incidence of calculi, bactiuria, and mucous production. Hematuria-dysuria syndrome secondary to acid secretion in the urine of these patients may be problematic. This symptom complex is exacerbated by oliguria and by an incompetent bladder neck because the urethra is most sensitive to the effects of aciduria. Consequently, bladder neck competence should be ensured whenever gastrocystoplasty is performed, and bladder neck reconstruction performed as necessary. Additionally, preemptive bladder irrigation and proton pump inhibition may prevent or minimize hematuria-dysuria symptoms.

TABLE 1. Considerations regarding pre-transplant bladder augmentation[35]

Augmentation type	Advantages	Disadvantages	Applications
None	Lowest risk of complication	Renal injury if bladder non compliant	Whenever possible
Autoaugmentation	1.No metabolic surface not amenable implant. performed 3.Limited clinical	1.Augmentation bladder surface for 2.Risk of perforation bladder capacity experience	1.Sufficient native 2.May be 2.Near normal consequences to ureteral implant extra-peritoneally
Ureterocystoplasty	1.No metabolic surface not amenable implant. performed extra-peritoneally	1.Augmentation bladder surface for 2.Requires presence of large dilated ureter	Sufficient native 2.May be consequences to ureteral implant
Gastrocystoplasty	1. Avoids acidosis/ infection /mucous 2. Facilitates tunnels for continence and antireflux 3. May potentiate growth in children	1. Hematuria-dysuria Syndrome 2. Rare alkalosis	Wide application
Ileocystoplasty	Technically simple surface not amenable unavailable	1.Augmentation bladder surface for 2. Acidosis/ infection/	1. Sufficient native to perform to ureteral implant implant 2.Stomach mucous
Colocystoplasty	1. Technically simple to perform 2.Ureteral implant possible	Acidosis/ infection / mucous	Stomach unavailable

While ureteral reimplantation can be successfully performed in a gastric segment or the tenia of a colic segment, reliable reimplantation is not possible into an ileocystoplasty, ureterocystoplasty or an autoaugmentation. an increased risk of mucous production, bactiuria, and calculi complicate ileocystoplasty as well as colocystoplasty. The clinical experience and applicability of autoaugmentation is limited. Finally, if a non-reconstructible native bladder is encountered, an intestinal conduit or a continent diversion may be considered. However, the risk of technical and infectious complications of these procedures is not insignificant.

Management

Treatment of bladder dysfunction in these patients is basically directed toward improving their renal function prognosis and avoiding the urinary incontinence that is present in some of these valve bladders.

Treatment is determined by bladder dysfunction. Post pubertal adolescent boys usually have a high capacity bladder with poor contractility, progressing towards myogenic failure. In these patients with myogenic failure and elevated postvoiding residues, obtaining efficient bladder voiding is absolutely necessary, and this is only possible with timed and frequent voiding, and double voiding in patients who can empty bladders efficiently and clean intermittent catheterization (CIC) in those who cannot.. Urethral catheterization can be quite difficult and is often painful in these patients. In this situation, a Mitrofanoff procedure would be justified to make bladder catheterization more bearable.[36] The role of ? 1-adrenergic antagonist in boys treated for PUV with secondary bladder neck obstruction and myogenic failure is also under evaluation.

Nocturnal bladder drainage by timed voiding or either intermittent or indwelling catheterization has been shown to alleviate the symptoms of valve–bladder syndrome and might slow down the progression to renal failure. Koff *et al.* postulated that the lack of a strictly timed nocturnal voiding regimen, as maintained during the day, led to chronic overdistention and subsequent treatment failure.[37] He, therefore, added a regimen of strictly timed or continuous nocturnal bladder drainage for 12 boys with a history of PUV. A marked improvement in hydroureteronephrosis was found in these patients, and most showed an improvement or stabilization in renal function, as well. Koff *et al.* concluded that nocturnal bladder drainage could be attempted in any disease that is complicated by chronic bladder overdistention due to severe polyuria or impaired bladder emptying.

Anticholinergic therapy with oxybutynin chloride has proven very effective in patients with instability. Side effects, however, may limit the clinical efficacy of this therapy and ultimately force alternative approaches. Intravesical oxybutynin has been effective in lowering bladder pressure but can only be used in cooperative patients. Other anticholinergics with fewer side effects and better tolerance, such as tolterodine tartrate and trospium chloride, have still not been widely used in children. However, anticholinergics are often not advisable and should only be used with great caution when there is a large bladder capacity and light or mild instability. Use of these drugs could further increase bladder capacity and decrease voiding detrusor pressure, which could result in incomplete bladder voiding that would thereby require clean intermittent catheterization (CIC). Indications for augmentation cystoplasty in boys with PUVs are similar to those for any augmentation; the main indication is a refractory poor bladder compliance that, despite anticholinergic therapy, produces upper tract dysfunction or unmanageable incontinence. Bladder augmentation will transform a high-pressure bladder to one with a large capacity and low pressure, thereby helping to avoid urinary tract deterioration. Boys with PUVs who have undergone bladder augmentation and, subsequently, become anuric must have their bladders irrigated daily before transplantation to remove bowel secretions and to maintain distensibility of the augmented segment.

Most patients with poor-compliance bladders of neuropathic origin require CIC after augmentation to void effectively; only half of the boys with PUVs who undergo augmentation will require CIC for voiding. Nevertheless, when considering bladder augmentation for a patient with a PUV, before surgery we must determine whether he will be able to undergo urethral CIC. Otherwise, bladder augmentation must necessarily be associated with a Mitrofanoff procedure to make it possible for him to endure CIC and void his neobladder to a satisfactory extent.

Conclusion

The importance of checking renal and bladder function throughout life in adolescents born with PUV cannot be over emphasized. Increasing attention to bladder dysfunction and its early treatment could probably improve the long-term prognosis.

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