Posterior urethral valves: The value of high diversion

A. E. MacKinnon

Department of Paediatric Surgery, Sheffield Children's Hospital, Western Bank, Sheffield S10 2TH, UK

Correspondence: AE MacKinnon, Consultant Paediatric Urologist, Department of Paediatric Surgery, Sheffield Children's Hospital, Western Bank, Sheffield S10 2TH, UK. E-mail: emackinnon@sch.nhs.uk

ABSTRACT

The management of posterior urethral valves remains a challenge to the paediatric urologist. Many cases diagnosed early may be managed by endoscopic surgery. However, where there is a gross upper tract dilatation, consideration needs to be given to temporary diversion to allow recovery of renal function and the elimination of infection. Western literature leans strongly towards nondiversion. However, in those countries, most patients present early, and often by antenatal diagnosis. Countries not having universally, highly developed medical services encounter more of the complicated cases. Evaluation of the role of upper tract diversion in such circumstances has not been conclusively established, but literature review indicates a role in selected cases.

KEY WORDS: Posterior method values, Ureterostomy, Bladder dyafnnichimi

The development of neonatal endourethral instruments for ablation of posterior urethral valves (PUV) approximately 25–30 years ago dramatically changed the direction of management of patients with PUV. The advent of detailed antenatal ultrasound studies provides early warning of abnormality, although it can be difficult to decide how significant minor degrees of upper tract dilatation are. Today, most cases of urethral valves are managed under vision but where such instrumentation is not available one form or another of diathermy hook such as the Whitaker hook can be used with fluoroscopic control. Before this development, a difficult approach via a perineal urethrostomy employing an auroscope was the main procedure employed. This had at times to be postponed because of the patient's condition or because it was considered safer to allow the child to grow before attempting surgery. In such instances, urinary diversion was necessary either by ureterostomy or vescostomy and these operations continued to be practised for fear of not overcoming high pressure in the upper renal tracts. Fear of bladder neck obstruction due to detrusor hypertrophy, which was thought at one time to be the level of obstruction and indeed rarely may be a factor, also encouraged diversion. Outcome reviews in the mid 1980s therefore still reported diversion with apparent good results. Casale in 1990 reviewed the theories of the causation of progressive deterioration in some patients. He noted the fears of obstructive uropathy complicating renal dysplasia and this led the surgeons to aim to maximise effective drainage of the kidneys. However, critical comparison of outcomes of upper diversion led to the belief that there is no place for diversion in cases of PUVs and indeed some authors believed that the outcome was worse. To interpret recent reports, it is important to remember that diversion has only been performed of late in the most severe cases, who inevitably will have more severe renal dysplasia, and therefore, in general, the outcome will inevitably be worse.

There have been no controlled trials of large groups of patients, and that would be a formidable task, correcting for all variables. The difficulty in evaluating the outcome is due to the fact that undoubtedly most cases undergoing early valve ablation have an outcome determined by the established degree of renal damage, rather than ongoing high pressure with or without reflux. Clinical practice in countries with universally well-developed medical services, have seen a progressively earlier time of presentation even antenatally, but other countries may see a different spectrum. Most reported series have a relatively few cases of gross upper tract dilatation that does not resolve following valve ablation. My interest in the role of upper diversion was originally stimulated by forming a personal opinion in the mid 1970s that ureterostomy played no part in the management of urethral valves on the basis of disappointing experience of ureterostomy in
those infants whose renal function failed to improve following valve ablation. I said as much when lecturing in Delhi in 1981 but was kindly taken aside by a friend in the unit and told that ureterostomy was still necessary in cases who rarely presented before the age of 4 years and with gross sepsis and renal failure. At that time, attempts at valve ablation as the primary procedure had almost always been fatal. This made me reconsider whether or not there is a place for upper diversion for specially selected indications. The same unit now sees earlier presentation of these patients and uses valve ablation as the first line treatment of urethral valves with successful outcome. It is interesting to review recent literature and find that there still is controversy, although most authors consider any diversion to be contraindicated.

Then why has there been this change and is there no place for diversion at all? The dilemmas remain: persisting significant dilatation, recurrent infection, incomplete voiding, and high-pressure bladder, irrespective of capacity.

Embryology and pathology
It has been postulated that every male fetus develops a urethral membrane that normally is resorbed. Failure to resorb may lead in the least severe instance to a degree of upper tract dilatation even without any evidence of residual valves. The other extreme is residual obstruction continuing throughout development with severe renal damage leading to oligohydramnios and early neonatal death from respiratory failure. Histological evaluation of nephrectomy specimens and animal studies have shown that early obstruction leads to dysplasia of the mesoderm, while later obstruction causes cystic changes in the kidney. With PUVs, the main histological changes are those of dysplasia and scarring indicating early fetal damage which must happen shortly after the kidneys commence to produce urine before 12 weeks. It is accepted that such renal damage is irreversible. Attention in managing the effects of urethral valves must therefore be concentrated on the pathophysiology of bladder and ureteric function, and the possible development of infection. We know from experiments on pigs that the combination of high-pressure reflux and infection causes severe renal damage and approximately one-third of patients with valves will have no reflux, one-third have unilateral, and one-third bilateral reflux. In those cases with unilateral reflux, it is recognised that the affected side may act as a ‘pop-off’ valve, thus, providing some protection to the opposite kidney. However, because such severe renal insult happens early in fetal life, it is not surprising to find that any protection is partial and the ‘protected’ kidney is also at risk of later deterioration.

New avenues of study that may lead to better understanding of the progressive nature of renal damage may come from the study of substances such as urinary transforming growth factor b (1) [TGF b (1)]. This factor is a profibrotic inflammatory mediator, which has been shown to be increased in experimental models of urinary obstruction. The TGF b (1) is increased in urine in certain cases of glomerular disease. MacRae et al have shown that it is increased in patients with PUV aged between 3 and 15 years. The levels did not correlate with GFR, nor with bladder wall thickening, or with previous diversionary surgery. Agents such as angiotensin-converting enzyme inhibitors may slow the progress of renal disease and might offer hope for future reduction in the progress of renal damage. Estimation of such factors may provide an indicator of those patients who are at risk of late deterioration needing life long surveillance.

Bladder dysfunction
The term ‘valve bladder’ encompasses a wide range of abnormalities, which include detrusor instability, poor compliance, detrusor-sphincter dyssynergia, sphincter weakness, and ureteric reflux. These factors, usually in combination, may lead to incomplete bladder emptying, infection, and possibly uretero-vesical obstruction. Hyper-reflexia and poor compliance have been shown to persist in a significant number of boys and this correlates with renal impairment. The trend is, in the early years, the bladder tends to be hypercontractile, but it changes in adolescence to hypocontractility. These authors studied two groups of boys who had undergone valve ablation and were free of voiding symptoms at ages 1–4 and 5–13 years with urodynamics. They underwent a further study at least 3 years later. Where the initial study demonstrated high pressure with or without loss of compliance there was a trend towards normal or low contractility. A similar reduction in detrusor activity was found with some boys, who initially had normal results. The same results were found in both the younger and in the older patients. They claimed that there is a loss of detrusor function with time. Furthermore, these results indicate that the finding of normal urodynamics does not mean that the patient is safe. They postulated that initial detrusor overactivity may cause detrusor failure in the course of time. Hypocontractile bladders are at risk of incomplete emptying, and with the development of the prostate gland at puberty this is likely to become more serious.

Close and colleagues reviewed the outcome in 31 boys, of whom 23 underwent valve ablation in the first year of life and compared them with eight boys who had
a high diversion but without giving clear indications for the choice of procedure. Progressive renal failure was not prevented in either group, which implies that this is determined by pre-existing damage. Valve ablation led to the improvement in the appearance of the bladder on cystography not seen with diversion, but in the latter the bladder had been defunctioned leading to the loss of normal cycling. However, Kim et al found the reverse when comparing urodynamic results.\[13\] They found that patients undergoing initial vesicostomy or pyelostomy followed by valve ablation and reconstruction after an average of 25 months had larger functional capacity, better compliance, and less detrusor instability when compared to primary valve ablation. Their theory is that resting the bladder permits recovery of the detrusor muscle. The incidence of progressive renal failure was similar in the two groups, but there is no indication as to why one or other treatment route was chosen.

No significant difference in bladder function was found in 59 children reviewed by Jaureguizar half having had valve ablation and the others pyelostomy.\[14\] Normal bladder function was found in 46.6% (14) of the former group, and 38% (11) were normal after diversion. The main abnormalities were instability (10, 12) followed by poor compliance (4, 5), and myogenic failure (2, 1). This was the same conclusion drawn by Jayanthi et al in a wider group of clinical condition, where ureterostomy had been performed.\[15\]

Puri et al\[16\] looked at urodynamic studies in a series of 67 patients treated between 1985 and 2000. Thirty-eight had primary fulguration of the valves and these had essentially normal urodynamic parameters in terms of capacity, compliance, and detrusor activity. Twenty-five patients had an initial vesicostomy and they were found to have a small capacity hyper-reflexic bladder. Only four patients had ureterostomies and all of them had a bladder function similar to those patients undergoing fulguration.

The only paper that discusses the technique of ureterostomy was presented by Liard et al\[17\]. In their series, they evaluated 17 boys out of a total of 120 who underwent Sober-en-T ureterostomy. This involves division of the ureter with re-anastomosis of the distal limb to the side of the proximal limb. This is in contrast to other papers in which a loop ureterostomy is reported, if the details of surgery are included. Liard et al\[17\] point out that loop ureterostomy is known not to provide assured free drainage, whereas the ‘T’ procedure does. At a mean of 13 months, they found no deterioration in bladder function and an immediate lowering of serum creatinine levels in all cases. However, they were unable to confirm or refute any long-term improvement in renal function.

Renal function
Farhat et al on the other hand were unable to support this,\[18\] Studying 50 cases, not surprisingly, they found that 22 patients born with normal upper tracts with normal biochemistry remained normal at 5 years following valve fulguration. Of those with normal renal function but upper tract dilatation and or reflux, 50% had normalisation of the upper tracts. However, they do not specify their management for those where normalisation did not ensue. Fifteen patients had azotaemia with ureteric dilatation and or reflux. Renal function returned to normal in six of seven of those treated by fulguration, but in only one of eight treated by diversion. Their intention was to treat all of them by fulguration and this was performed primarily in 44. Two of these later underwent high diversion for persistent sepsis, but their outcome is not documented. They concluded that the outcome is determined more by renal dysplasia and infection rather than the detail of management of the valves. Their results are not surprising because the diverted patients appeared to have suffered from renal dysplasia, but we are left wondering about the 50% with abnormal upper tracts but normal biochemistry in whom there was no anatomic improvement.

Smith et al from Philadelphia\[19\] noted that in various historical series, good results of treatment of urethral valves had been obtained with diversion. They proposed that the same results could be achieved by fulguration alone. They looked at the follow up of 100 patients at a median of 11.2 years treated before 1985. Seventy-four were treated initially by fulguration, and of these four of them later had a vesicostomy but only one improved. Three patients initially having fulguration had high diversion without benefit later. Their conclusion was that there is little or no benefit to be gained from diversion as opposed to ablation and little benefit from diversion, as a rescue procedure.

Perhaps one of the most convincing reports on the efficacy and safety of ureterostomy is that of Parag et al.\[20\] In a series of 200 cases, only 26 underwent bilateral loop ureterostomy for persistently high-serum creatinine levels, urosepsis, or urinary ascites. After a period of 1 year, the median creatinine levels had fallen from 2.5 to 0.6 mg/dl. At the time of reporting, undiversion had been performed in 12 boys without complication but three had died. Documented ureteric reflux was treated surgically in only 1 of 14 cases.
Urinary-vesical junction

There has been a debate as to the existence of an obstruction in the urinary-vesical junction in cases of urethral valves. Tietjen et al. aimed to answer this question by studying 26 patients with raised serum creatinine levels following 1 week of bladder decompression by an indwelling catheter. All proceeded to upper tract diversion with delayed reconstruction and valve ablation. In the first 12 months following diversion, the serum creatinine level steadily fell from a mean of 2.5 to 1.0 mg/dl. At reconstruction, a Whitaker test was performed in all 52 renal units and in only two was there evidence of obstruction. Renal biopsy in nine revealed dysplasia as was to be expected and at a median of 11 years post-treatment 11 patients had a renal failure. However, this study was conducted after a significant period of diversion during which time it could be argued that any obstructive element may have resolved with the detrusor being ‘rested’.

Rare reports exist of the outcome of apparent urethral-vesical obstruction. Noe and Jerkins described two infants in whom there was rapid upper tract dilatation following valve ablation. These infants were treated conservatively and the dilatation remitted.

Body growth

In 1980, Krueger and colleagues reported improved body growth and renal function in patients diagnosed in the first year of life and treated by initial temporary supravesical diversion. However, Reinberg et al. reported different results on long-term follow up (mean 18 years), but body growth being maintained in spite of a higher rate of renal failure. Eight of twenty-five patients underwent diversion and of these five progressed to renal failure, six of 17 not diverted did so, and in all patients this was related to renal function in the first year after surgery. Interestingly, in spite of poorer outcome in terms of renal function, the mean percentage height at age 10 years was 61% of the norm for those diverted and 35% for those not diverted.

DISCUSSION

Where then do we find ourselves with regard to upper tract diversion in the management of PUVs? The factors that determine renal outcome are first and foremost the degree of pre-existing renal dysplasia followed by detrusor dysfunction and urine infection. The lack of controlled trials concerning management makes a decision regarding the place of diversion difficult, particularly with the conflicting evidence provided. A further confounding issue is that almost all discussion on ureterostomy has been based on the use of either loop or ring ureterostomies. Both these procedures may produce an obstructed system because it is easy to rotate the ureter, a problem avoided by the ‘T’ (or ‘Y’) ureterostomy.

I believe that it is inescapable in developed countries where the majority of cases present in the neonatal period, or at least in the first few months of life. The correct and successful first line management is valve ablation with antibiotic cover. The presence of ureteric reflux will demand that chemotherapy is continued up to the age of 5 years, or at the least up to 3 years. All patients should have urodynamic studies performed at perhaps the age of 3 months or as soon as reasonable following valve ablation if performed after this age. The likelihood of there being detrusor hyper-reflexia and limited compliance indicates that anticholinergic medication should be commenced. However, it should be withheld for 7 days before urodynamics to allow the natural situation to be identified. The finding of incomplete voiding indicates the need for intermittent catheterisation three times daily. Some authorities recommend that this is commenced in all infants because it is easier to introduce a catheter at this age, and then it becomes accepted by the child. From the current review, this action does not seem to be well founded. If improvement is gained in renal function and upper tract anatomy, there is no need for further surgery.

The management of patients in whom improvement does not take place demands a consideration of diversion on a temporary basis. Arguments against diversion include (a) the very limited benefit gained because renal function depends on the degree of dysplasia, (b) variable reported detrimental effects on bladder function, and (c) the necessity for serial operative procedures. Arguments in favour of diversion include (a) the rapid control of urosepsis, (b) alleviation of the ‘load’ on the detrusor muscle, and (c) possible improved body growth. The following scenarios may present themselves following adequate valve ablation:

1. Persistently raised creatinine levels with limited upper tract dilatation. This must be due to renal dysplasia for which diversion is of no benefit. Renal ultrasound may support the impression of dysplasia if both kidneys are seen to be bright, and likely small for the child’s age.

2. Persistently dilated upper tracts with improving biochemical values. It is unlikely that diversion will be of benefit.

3. Persistently dilated upper tracts without improve-
ment of biochemical values. Confirmation of adequate valve ablation is needed before aggressive management of bladder dysfunction. If this fails to make improvements then there could be a place for diversion.

4. Persistently dilated upper tracts with urosepsis not responding to aggressive chemotherapy. This situation seems to be ideally suited to management by diversion. Diversion provides rapid control of infection with maximal reduction in upper tract pressure.

5. Poor physical growth. It seems inappropriate to perform diversion for this alone.

It remains for us to decide what form of diversion should be performed if such surgery is considered. There seems to be sufficient evidence that vesicostomy, though often successful in overcoming infection, may lead to detrimental effects on bladder function. Percutaneous pyelostomy has been practised but is not a stable situation in the long-term, but has been advocated as a temporary procedure to determine the possible benefit of diversion. However, improvement may be slow and the drainage may easily be lost before proper evaluation is possible. Ureterostomy has been criticised for diverting urine from the bladder. However, if one of the ‘chimney’ types of anastomosis is performed this problem can be avoided and there is evidence that bladder function may normalise.

In conclusion, I believe that there is still an occasional indication for upper tract diversion in PUVs. The main indication is slowly responding urosepsis, but there may occasionally be an indication when there is persisting upper tract dilatation and failure to achieve improved renal function.

REFERENCES


