Bladder augmentation and continent urinary diversion in boys with posterior urethral valves

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KEY WORDS

urinary bladder ◗ valve bladder ◗ bladder augmentation

ABSTRACT

Posterior urethral valve (PUV) is a condition that leads to characteristic changes in the bladder and upper urinary tract. Dysfunction of the bladder such as a hyperreflective, hypertonic, and small capacity bladder as well as sphincter incompetence and/or myogenic failure should be adequately treated. Poor compliance/small bladder could be treated with anticholinergics, but bladder augmentation will probably be indicated. Although bladder reconstruction with gastrointestinal segments can be associated with multiple complications, including metabolic disorders, calculus formation, mucus production, enteric fistulas, and malignancy formation, enterocystoplasty is still the gold standard. In contrast to a neuropathic or exstrophic bladder, augmentation of the valve bladder allows spontaneous voiding without significant residual urine in the majority of cases, but some require CIC (clean intermittent cathterization). Augmentation cystoplasty is also an efficient approach in those children who will require kidney transplantation in the future.

Posterior urethral valve (PUV) is a condition that leads to characteristic changes in the bladder and upper tract. The bladder develops hypertrophic changes including hypertrophy and hyperplasia of the detrusor muscle along with increased connective tissue. The ratio of muscle to connective tissue is the same as in the normal bladder but the type of collagen is different. The effects are visible as wall thickness, trabeculation, and diverticula. This results in poor sensation, hyper-contractility and low compliance of the bladder, and may contribute to poor emptying and incontinence. Almost all patients with PUV have severe hydroureteronephrosis at the time of diagnosis with associated reflux in 50-70% of cases. If there is no reflux the ureter and kidney are protected from the complete force of the bladder contraction, but if reflux occurs the entire pressure of the thickened and hyper-contractile bladder is transmitted directly to the upper tract with severe consequences. Obstructive uropathy involves both glomerular and tubular injury. Glomerular injury occurs when high pressure results in decreased renal perfusion and filtration. It is partially reversible with pressure reduction. Tubular damage results in failure to concentrate and acidify the urine. It worsens with age despite early relief of obstruction; the resultant high urine volumes contribute to the deterioration of renal and bladder function in late childhood.

The primary endoscopic ablation of the valves followed by a wait-and-see attitude is the most efficacious management of posterior urethral valves. Valve ablation in a neonate with significant reflux and a markedly trabeculated bladder can remodel itself remarkably within the first year of life. The persistence of hydronephrosis, bladder wall thickening, and trabeculation, as well as persistent elevation of serum creatinine can all be the manifestation of persistent bladder outlet obstruction (BOO), so urethroscopy with repeated valve ablation is necessary. But what do you do if the obstruction is not anatomic? Carr and Snyder consider the point at which a functional obstruction occurs and which management is reasonable [1]. They concluded that dysfunctions of the bladder such as a hyper-reflective, hypertonic, and small capacity bladder, as well as sphincter incompetence and/or myogenic failure should be adequately treated.

Myogenic failure with overflow incontinence and incomplete bladder emptying should be treated with time voiding, double voiding, α-blockers, and intermittent catheterization.

Detrusor hyperreflexia with urinary frequency and urge urinary incontinence (UUI) are usually managed with anticholinergics.

Poor compliance/small bladder could be treated by anticholinergics, but most probably will need bladder augmentation.

The place and timing of augmentation cystoplasty in a "valve bladder" have not yet been well established. In 1995, Kajbafzadeh et al. reported their experience with augmentation cystoplasty in 20 boys with previously treated PUV [2]. Urodynamic studies confirmed poorly compliant, unstable bladders with low functional capacities, which had failed to respond to anticholinergic treatment in all patients. The bladder was augmented with ileum in nine, stomach in seven, colon in two, and ureter in two cases. A Mitrofanoff channel was fashioned in six cases. Upper tract dilatation improved in 17 patients and remained stable in three. Of the patients, 17 are dry day and night. Eleven patients void spontaneously without significant residual urine, seven are on CIC for residual urine of greater than 50 ml, and two are completely dependent on catheterization. They concluded that augmentation cystoplasty is a safe and effective method to achieve continence in boys with a low capacity, poorly compliant bladder after valve ablation that do not respond to medical management. In contrast to the neuropathic and exstrophy bladder, the augmented valve bladder allows spontaneous voiding without significant residual urine in the majority of cases. Early intervention in these patients may prevent deterioration in renal function.

In reality, most of patients with "valve bladder" who qualified for augmentation cystoplasty presented with renal insufficiency.

The method of bladder augmentation is open to discussion. Enterocystoplasty is the most popular but ureterocystoplasty seems to be ideal. In 2007, Youssif et al. presented eight boys (mean age 5 years) with valve bladder syndrome after successful valve ablation [3]. When conservative treatment failed, ureterocystoplasty was scheduled. The entire ureter was folded and used in four boys after nephrectomy for a non-functioning kidney. The lower dilated ureter was used to augment the bladder, a transureteroureterostomy (TUU) was used in two patients, and reimplantation of the remaining ureter were performed in another two patients. Bladder capacity and compliance were significantly

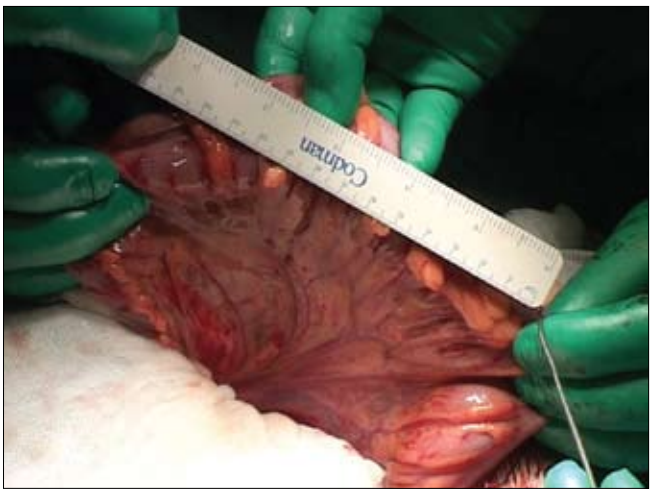

Fig. 1. A 15 cm long sigmoid segment is isolated.

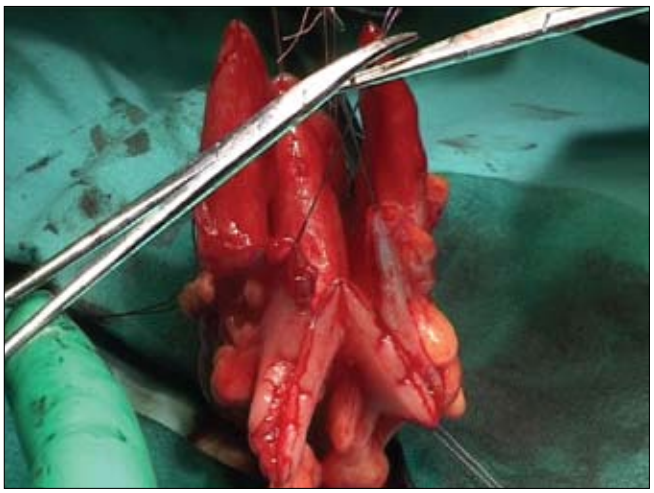

Fig. 2. Demucosalized sigmoid segment is folded in W-shape.

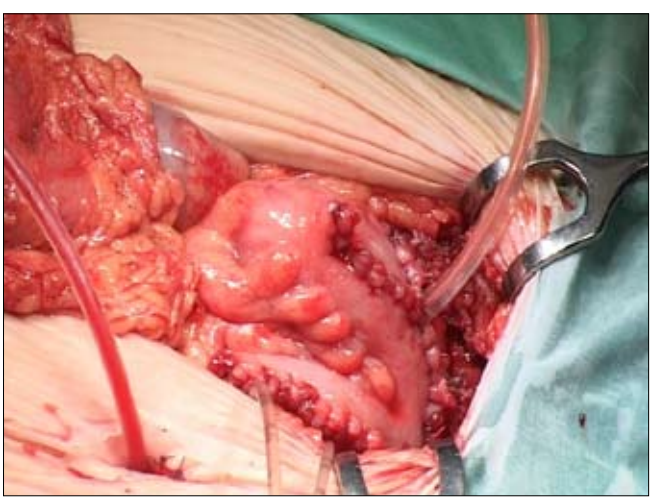

Fig. 3. Formed bowel is anastomized with the wide open bladder after continent stoma creation (appendico-cutaneostomy according to Mitrofanoff).

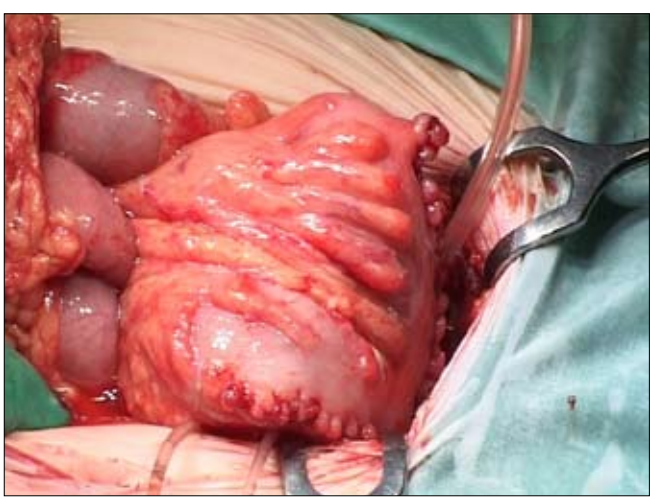

Fig. 4. Reservoir is continent with adequate capacity.

improved in all cases. Hydroureteronephrosis improved in six boys (75%). Self-CIC was performed routinely in all cases after surgery, which was weaned off from as deduced from the voiding pattern of the child. They concluded that ureterocystoplasty is an ideal option for augmenting the hypocompliant bladder in boys with valve bladder syndrome. The entire ureter or the dilated lower part can be used. The procedure avoids almost all the complications of enterocystoplasty.

In 2010, Fisang et al. confirmed the efficacy of ureterocystoplasty in bladder augmentation, but enterocystoplasty is still the method most frequently used while the ideal gastrointestinal segment remains controversial [4].

Ileocystoplasty seems to be the most common technique for augmentation [5]. This is because the ileum has been demonstrated as the least contracting bowel segment. However, using this bowel requires the isolation of a very long segment of ileum (usually 20-40 cm) depending on the volume needed.

Sigmoidocystoplasty allow reducing the length of bowel segment by even 15 cm, but strong unit contractions could come forward [6].

Metabolic complications related to the storage of urine within an intestinal segment is hyperchloremic acidosis. Mitchel and Piser noted that every patient after intestinal augmentation had an increase in serum chloride and a decrease in serum bicarbonate level, although full acidosis was rare if renal function was normal [7]. Hall confirmed that there is an increase in the urinary acid load with wasting of bony buffers even in the absence of frank acidosis [8]. Such wasting may result in bone demineralization and can cause retarded growth in children after augmentation cystoplasty.

Careful long-term follow up is obligatory because of metabolic disturbances and upper urinary tract changes as well as the potential risk of malignancy. Multiple cases of bladder cancer have been reported recently in young adults with a history of bladder augmentation in childhood, but the mechanisms of developing cancer in intestinal segments remain uncertain. The initiating event appears to occur soon after surgery. Epithelial proliferation at the healing anastomosis in the presence of promoter carcinogens may be a causing factor [9, 10].

Gastrocystoplasty is an alternative to intestinal augmentation. The use of gastric patch was promising because of the quite easily available tissue, lack of absorption of hydrogen ions, and bactericidal effect of acid secretion [11]. The wedge-shaped segment from the greater curvature became popular in children. Unfortunately, the secretory nature of gastric mucosa results in two serious complications: hypokalemic, hypochlochloremic metabolic alkalosis and hematuria-dysuria syndrome. Also, local lesions could appear as perforation of the gastric segment and skin injury if urinary continence is not sufficient to collect a big amount of urine diluting the gastric acid [12, 13].

Mucus production is another problem related to alimentary duct segment incorporated to the urinary tract. It is known, that

the gastric segment is the lowest producer and colonic segment produces more mucus than the ileal segment. Mucus could impede bladder drainage particularly during CIC with small-caliber catheter. Mucus collection may result in infection or stone formation, particularly if it remains in the bladder for a long period. To minimize these complications, daily irrigation of the augmented bladder is necessary. Also, the use of the demucosalized sigmoid segment is helpful [14]. In the author's experience, mucosa should be removed carefully using dry swabs (not scissors), to prevent submucosal layer injury [15]. This maneuver reduces mucus secretion and prevents shrinkage of the demucosalized sigmoid segment.

The goal of augmentation cystoplasty is to create low-pressure reservoir that allows storage of urine and assures continence with protection of the upper urinary tract. As it was mentioned before, the augmented valve bladder allows spontaneous voiding without significant residual urine, but some still require CIC. Catheterization via the urethra is possible, but if it is difficult or painful an appendicocutaneostomy is necessary, especially in those with a damaged posterior urethra and bladder neck. Bladder neck insufficiency with incontinence is the indication for continent reservoir creation or ureteroileocutaneostomy as noted by m. Bricker.

To create a continent reservoir, we used 15 cm of demucosalized sigmoid segment (Fig. 1). The bowel is folded in a W-shape to obtain the largest possible capacity (Figs. 2, 3, 4). Appendicocutaneostomy is created according to Mitrofanoff and Y-V plasty is done to prevent obstruction of the stoma. After appendix implantation, the bladder is cut out of the urethra. The dissection is done in two steps and the urethra is closed [15].

A continent reservoir assures dryness, but good cooperation by the patient is necessary with regular CIC and daily bladder washing should be recommended to prevent stone formation.

Successful operation will increase the quality of life for most patients but: "does bladder augmentation stabilize serum creatinine in urethral valves disease?" [16]. It was a question asked by Bhatti et al. who evaluated the results of bladder augmentation in 19 boys with PUV. The mean serum creatinine at the time of augmentation cystoplasty was 2.11 mg/dl. The serum creatinine stabilized in 14, but failed to do so in five boys. A serum creatinine level of more than 2 mg/dl at the time of augmentation was associated with a significantly worse rate of success. They concluded that bladder augmentation has been beneficial in children with pre-augmentation creatinine level up to 2 mg/dl.

Augmentation cystoplasty is also an efficient approach in those children who will need kidney transplantation in the future. It has the advantage of restoring the lower urinary tract before immunosuppressive therapy, and supplies the best possible reservoir for a transplanted kidney [17].

Bladder augmentation is still a commonly performed reconstructive procedure for pediatric patients with severe bladder dysfunction. Although bladder reconstruction with gastrointestinal segments can be associated with multiple complications, such as metabolic disorders, calculus formation, mucus production, enteric fistulas and potential for malignancy, enterocystoplasty is still the gold standard [18]. In order to avoid those complications research has been conducted into using tissues other than bowel, which challenges the current tissue engineering technology [19].

REFERENCES

1. Carr MC, Snyder HM: *Urethral valves. Fate of the bladder and upper urinary tract.* Urologe A 2004; 43 (4): 408-413.
2. Kajbafzadeh AM, Quinn FM, Duffy PG, Ransley PG: *Augmentation cystoplasty in boys with posterior urethral valves.* J Urol 1995; 154 (2Pt2): 874-877.
3. Youssif M, Badawy H, Saad A, et al: *Augmentation ureterocystoplasty in boys with valve bladder syndrome.* J Pediatr Urol 2007; 3 (6): 433-437.
4. Fisang C, Hauser S, Müller SC: *Ureterocystoplasty: an ideal method for vesical augmentation in children.* Aktuelle Urol 2010; 41, suppl. 1: S50-2; Epub 2010 Jan 21. Surer I.
5. Ferrer FA, Baker LA, Gearhart JP: *Continent urinary diversion and the exstrophy-epispadias complex.* J Urol 2003; 169 (3): 1102.
6. Bhatanagar V, Dave S, Agarwala S, Mitra DK: *Augmentation colocystoplsty in bladder exstrophy.* Ped Surg Int 2002; 18 (1): 43.
7. Mitchell ME, Piser JA: *Intestinocystoplasty and total bladder replacement in children and young adults: Follow-up in 129 cases.* J. Urol 1987; 138: 579.
8. Hall MC, Koch MO, McDougal WS: *Metabolic consequences of urinary diversion through intestinal segments.* Urol Clin North Am 1991; 18: 725.
9. Filipas D, Stein R, Fisch M: *Orthotopic and nonorthotopic bladder substitution.* In Gearhart, Rink, Moruriquand: Pediatric Urology. Philadelphia, WB Saunders Company, 2001: 947.
10. Austin JC: *Long-term risks of bladder augmentation in pediatric patients.* Curr Opin Urol 2008; 18 (4): 408-412.
11. Leong CH: *The use of gastrocystoplasty.* Dialog Pediatr Urol 1988; 11: 3.
12. El-Ghoneimi A, Muller C, Guys JM, et al: *Functional outcome and specific complications of gastrocystoplasty for failed bladder exstrophy closure.* J Urol 1998; 160: 1186.
13. Mingin GC, Stock JA, Hanna MK: *Gastrocystoplasty: long-term complications in 22 patients.* J Urol 1999; 162 (3Pt2): 1122-1125.
14. Jednak R, Schimke CM, Ludwikowski B, Gonzalez R: *Seromuscular colosystoplasty.* BJU International 2001; 88: 752.
15. Baka-Ostrowska M: *Bladder neck closure with sigmoidocystoplasty and continent appendicostomy. Video-presentation of the technique accessible on the ESPU website* (www.espu.org) - "ESPU Members only area".
16. Bhatti W, Sen S, Chacko J, et al: *Does bladder augmentation stabilize serum creatinine in urethral valve disease? A series of 19 cases.* J Pediatr Urol 2007; 3 (2): 122-126.
17. Djakovic N, Wagener N, Adams J, et al: *Intestinal reconstruction of the lower urinary tract as a prerequisite for renal transplantation.* BJU Int 2009; 103 (11): 1555-1560.
18. Gurocak S, Nuininga J, Ure I, et al: *Bladder augmentation: Review of the literature and recent advances.* Indian J Urol 2007; 23 (4): 452-457.
19. Alberti C: *Metabolic and histological complications in ileal urinary diversion. Challenges of tissue engineering technology to avoid them.* Eur Rev Med Pharmacol Sci 2007; 11 (4): 257.

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