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

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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 hyperreflexive, 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 catheterization). 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, hypercontractility and low compliance of the bladder, and may contribute to poor emptying and incontinence. Almost all patients with PUV have severe hydronephrosis, hypertension, and hypercontractile bladder. Myogenic failure with overflow incontinence 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, Kakabazadeh 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 improved.
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

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 anastomosed with the wide open bladder after continent stoma creation (appendico-cutaneostomy according to Mitrofanoff).

Fig. 4. Reservoir is continent with adequate capacity.
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 ureterocutaneostomy 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].

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