REVIEW ARTICLE

POSTERIOR URETHRAL VALVE

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Introduction:

Posterior urethral valve (PUV) is one of the most common causes of lower urinary tract obstruction in male neonates. Although not precisely known, its prevalence is reportedly1/8000 to 1/25000 live births.(1,2) PUV has been observed exclusively in boys,(3) but several reports in adults have been published.(4-7) The definitions of many of the disease manifestations have been changed in recent years. The primary pathology is a mucosal membrane in the prostatic urethra, although secondary complications of this membrane result in injuries in the kidneys and the urinary bladder, which determine the fate of the children with this primary urethral membrane.(8-11)

PUV was first described in 1515 and subsequently observed at autopsies. In 1802, the first definition for PUV was written and presented in an article on lithotomy.(12) The first report in British journals is found in the Lancet, in which Dr Budd reported a PUV in a 16-year old boy who had died of renal failure.(13) He stated that severe dilation of the kidneys and the urinary tract, as well as renal failure, had all been due to the obstruction caused by the PUV.In 1913, Young reported the first clinical case of PUV, before which all the cases had been diagnosed postmortem.(14) In 1919, he published a report of 36 cases from the papers of that time, 12 being his own patients and the other 24 from various other papers.(15) It was in this paper in which he presented a classification for the PUV. The numbers of case reports or case series of the patients continued to grow from the early 20th century, so that by 1949, there were 207 published cases of PUV worldwide.(16-18) In the last 10 years, several hypotheses have

been proposed regarding bladder function and its relationship with renal function followingcorrection of the primary obstruction, as well as several urodynamic studies and their relation with renal function and treatment options, all of which has led to major progress in the field. Still,after 300 years since the initial diagnosis of this disease, more than one third of the affected children develop renal failure.

Types of posrerior urethral valves:

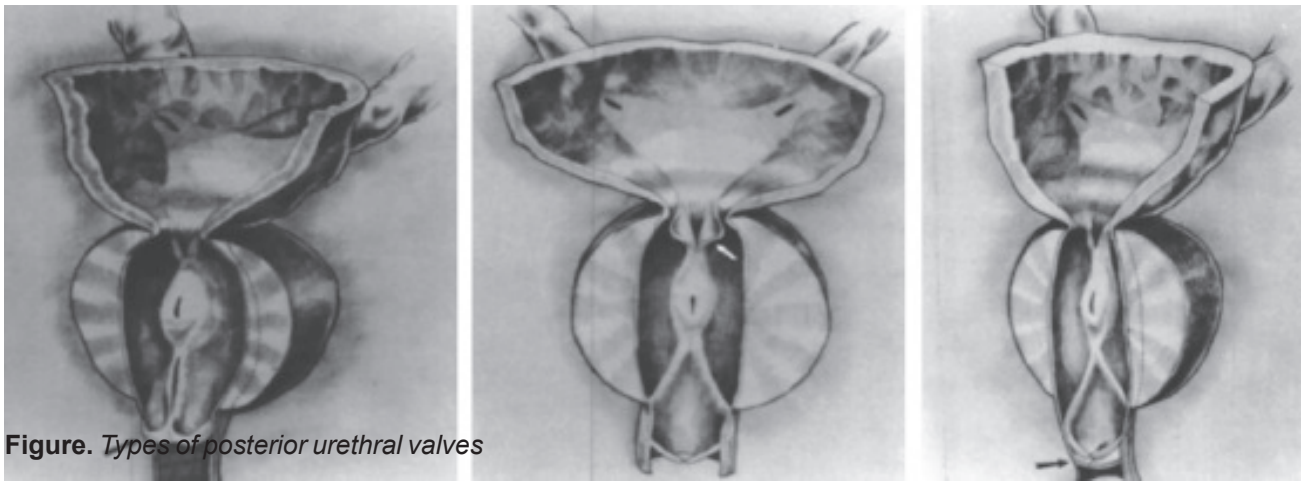

Figure. *Types of posterior urethral valves*

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The original classification of the types of posterior urethral valves proposed by Young in 1919 is still in use today. (A) Type I posterior urethral valves (arrow) are mucosal folds extending anteroinferiorly from the caudal aspect of the verumontanum, often fusing anteriorly at a lower level. They are derived from the plicae colliculi and constitute the vast majority of valves. (B) Type 2 posterior urethral valves (arrow) are mucosal folds extending anterosuperiorly from the verumontanum toward the bladder neck. A rare occurrence, they are probably an effect rather than a cause of bladder obstruction. (C) Type 3 posterior urethral valves (arrow) are disc-like membranes located below the verumontanum and unrelated to it. They constitute a small percentage of posterior urethral valves.

Pathophysiology of Posterior Urethral Valves

Posterior urethral valve (PUV) is the most common cause of obstructive uropathy leading to renal failure in male neonates [19]. Although the true incidence of PUVs is not precisely known, PUV is estimated to occur in 1: 5000 live births [1]. The normal male urethra is anatomically divided into the prostatic and membranous portions (posterior urethra) and the spongy or anterior urethra. The urethral crest is a mucosal ridge that gives a specific form to the posterior urethra, and on either side of the ridge is the prostatic sinus. The urethral crest continues below the verumontanum and coalesces in a small midline bridge. This membrane, extending laterally and downward, eventually vanishes [20].

The classic form of PUV is found in the prostatic urethra, below or proximal to the verumontanum. Although the precise embryologic mechanism of PUV remains unknown [21], four theories have been proposed to explain their development and include hypertrophy of the urethral mucosal folds, persistence and continuation of the urogenital membrane [22], abnormal development of the Wolffian or Mullerian duct [23], and fusion of the verumontanum or the posterior urethral roof epithelium [24].

Pathogenesis of Renal Dysfunction

There is no single genetic mutation or biologic model that reproduces the phenotype of posterior urethral valves or congenital bladder outlet obstruction. In early work with fetal sheep, surgical obstruction caused hydronephrosis within one week and resulted in dysplastic changes at term [10]. Further studies confirmed the presence of dysplastic changes and altered nephrogenesis in kidney's exposed to outlet obstruction during development [11–13]. Altered cellular differentiation, phenotypic changes in glomerular cells [14], apopotosis, and increased oxidative stress [15] contribute to decreased nephrogenesis and renal dysplasia. A key early event appears to be distention and mechanical stretch of the dilated collecting system, leading to activation of the apoptotic and inflammatory cascades [16].

The decreased number of total nephrons present at birth leads to hyperfiltration injury, exacerbation of the underlying inflammatory process, renal fibrosis, and ultimately renal failure [17, 18]. The early impact of altered differentiation and activation of the apoptotic cascade clearly illustrates that the molecular mechanisms and alterations in renal architecture which underlie progressive renal failure are established in utero.

Surgical Care

Surgical care of the patient with PUV varies according to age, bladder status, and renal status. Antenatal surgery has been reported in patients diagnosed with PUV with the goal of improving postnatal outcomes. Antenatal hydronephrosis is detectable only after renal development has occurred and urine production has started.

With improvement in antenatal ultrasonography, the hope was that earlier intervention with vesicoamniotic shunting would improve postnatal renal function. However, identification of those patients who may benefit from early intervention remains elusive. To date, improvement in renal function has been difficult to demonstrate, and antenatal intervention remains experimental.

Urinary drainage

Urinary drainage may be accomplished by means of postnatal primary valve ablation, vesicostomy, or cutaneous ureterostomies.

Postnatal primary valve ablation

Ideal treatment involves transurethral incision of the PUV during the first few days of life. Current infant resectoscopes are available in 8 French and smaller sizes. The valves can be incised at the 12-, 5-, and 7-o'clock positions, with either a cold knife or an electrocautery. Some surgeons prefer to leave a catheter in place for 2-3 days after the procedure. The timing of the postoperative VCUG varies and ranges from several days to several months.

Comparison of the posterior urethral diameter with anterior urethral diameter can provide an objective measure of valve ablation. In most patients, the posterior

urethra is markedly dilated. Postincision diameter should decrease if the incision is successful. The normal posterior-to-anterior urethral ratio is approximately 2.3. Approximately two thirds of patients have successful valve ablation with one procedure, manifested by a postincision ratio of 3.1 or less. [13] One third of patients require a second incision to achieve this level of posterior urethral reduction.

Because approximately one third of patients will require a second valve incision, some authors recommend routine surveillance cystoscopy 1-2 months after the initial incision to evaluate and treat any residual valvular obstruction. [14]

In a study by Shirazi et al, factors significantly associated with a higher incidence of obstructive remnant leaflets after valve ablation for PUV included the following [15] :

- Younger age at the time of surgery
- Hyperechogenicity of renal parenchyma
- Presence of vesicoureteral reflux (VUR)
- Grade 4 or 5 reflux preoperatively

Vesicostomy

When urethral size precludes safe valve ablation, a communicating channel between the bladder and lower abdominal wall (ie, vesicostomy) can be created to provide bladder drainage. Generally, an 18- to 20-French stoma is created approximately midway between the pubis and the umbilicus in the midline. Take care to bring the dome of the bladder to the skin and to limit the stomal size to prevent prolapse of bladder urothelium through the vesicostomy. Formation of too small a stoma results in stomal stenosis and inadequate bladder emptying; formation of too large a stoma allows for bladder prolapse. Vesicostomy use has decreased because most patients can be safely drained and can undergo valve ablation.

Cutaneous ureterostomies

Bilateral cutaneous ureterostomies can also be placed to provide for urinary drainage. Techniques for cutaneous ureterostomy include the following:

- End stomalureterostomy
- Loop ureterostomy
- Y-ureterostomy (in which the ureter is divided and one end is brought to the skin and the other is reanastomosed in a ureteroureterostomy)
- Ring ureterostomy

Potential complications of cutaneous ureterostomies, all of which are rare, include the following:

- Ureteral devascularization
- Inadequate drainage
- Stomal stenosis

Secondary bladder surgery

Secondary bladder surgery takes the form of augmentation cytoplasty or continent appendicovesicostomy.

Augmentation cystoplasty

Indications for bladder augmentation include inadequately low bladder storage volumes and high bladder pressures despite anticholinergic medication and clean intermittent catheterization. The ileum is most commonly used; however, the large bowel, stomach, and ureter are also used, depending on clinical conditions and surgeon preference.

Before an augmentation procedure is undertaken, the implications of bladder augmentation should be carefully reviewed with parent and family. Augmentation should only be offered to patients willing to commit to lifelong intermittent catheterization.

Potential complications include the following:

- Bladder rupture (~10% of patients)
- Electrolyte disturbances, which may be worsened by the placement of intestinal mucosa in contact with urine, especially in those with a serum creatinine greater than 2 mg/dL
- Mucus production, which can be a source of catheter blockage and may be a nidus for stone formation

The future risk of neoplasia has not yet been defined in these patients, but several cases of malignant degeneration in augmented bladder have been reported. Augmentation cytoplasty does not appear to have an adverse effect on overall renal outcome in PUV patients who undergo kidney transplantation, though it is associated with a higher incidence of recurrent urinary tract infection (UTI). [16]

Despite these risks, augmentation can significantly improve patient lifestyle in those who have intractable incontinence as a consequence of poor compliance and bladder overactivity. By lowering intravesical pressures, the upper urinary tract may also be protected.

Continent appendicovesicostomy

Also called the Mitrofanoff technique, continent appendicovesicostomy involves placing a nonrefluxing tubular conduit for catheterization between the bladder and skin to provide an alternative channel for catheterization. In children with PUVs, institution of intermittent catheterization through a sensate urethra can be difficult. In addition, some patients may have a highly dilated proximal urethra that may not be easily catheterized. The stoma often can be hidden in the umbilicus to provide acceptable cosmesis. The appendix, ureter, and tubularized bowel can be used for formation of this channel.

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