12. Posterior Urethral Valves

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Introduction

Posterior Urethral Valves is the most common infravesical obstruction in boys (1). It comprises 10% of all prenatally diagnosed uropathies and occurs approximately 1:4000 live male births. Each year another 10 cases with severe prenatal urethral obstruction in the same referral centre (Leeds Teaching Hospitals) opt for elective abortion (2).

Posterior Urethral Valves are rarely associated with other congenital anomalies, except undescended testicles, which occur in 12% of valve patients (3).

Valve anatomy

The male urethra can be divided into two parts the anterior and the posterior. The posterior urethra is composed of prostatic and membranous parts. The former contains verumontanum with the ejaculatory ducts and the latter corresponds to the external urethral sphincter (Image 1).

In 1919 Young classified the congenital obstruction of the posterior urethra in three types (4). The vast majority of valves fits to the description of
Young’s Type I: two leaflets that arise from the distal base of the verumontanum follow oblique course, traverse the external urethral sphincter at its anterolateral aspects and fuse onto the anterior urethral wall just distally to the sphincter (Image 2).

Young’s type II is now completely discounted. It essentially describes the mucosal folds located between the bladder neck and verumontanum. These folds are proved to be non-obstructive.

Young’s type III valve is a membrane located at the verumontanum. In 1992 Dewan and Duffy made more precise description of this membrane. In fact they found it in the majority of patients with uninstrumented urethra. They describe a single membrane, arising from the distal base of the verumontanum, extending through the external urethral sphincter and attached to the anterior urethral wall. There always is a small eccentric aperture in the middle of the membrane. These authors believe that all valves are initially type III valves and the type I results either from the partial intrauterine rupture of the membrane or from the urethral catheterization (5).
Pathophysiology and long-term outcome

An abnormal confluence of the mesonephric duct with the urogenital sinus membrane during the 7th week of gestation, results in the valvular urethral obstruction of the posterior urethra.

The earlier the obstruction the more severe is its consequence. This implies that there is a spectrum of clinical severity in patients with posterior urethral valves. At the extreme end of this spectrum is a severe obstruction, with arrested renal development, oligohydramnios and pulmonary dysplasia, which in clinical practice translate into spontaneous abortion or neonatal death. Neonates with underdeveloped kidneys but well-developed lungs, who survive the neonatal period, still have 20-30% risk of progression to the end stage renal failure in the first decade of life (6).

At the opposite extreme of the spectrum are the essentially normal boys with normal renal function, mild or absent lower urinary tract symptoms.

The majority of posterior urethral valve patients however, belong to the middle of the spectrum. The consequence of obstruction and the long-term prognosis in this group is determined by interplay of the severity of renal dysplasia, urinary bladder abnormalities and the clinical course of the patient. Recent studies show that the long-term renal function outcome in this group has been considerably improved. This improvement is believed to emanate from prenatal diagnosis, from a heightened awareness of posterior urethral valves, from optimal neonatal urological care and nephrological support (7).
The majority of PUV patients of all groups have a degree of incontinence. This is initially related to the uninhibited detrusor contractions and reduced bladder capacity.

Later in life, particularly after puberty, when the bladder capacity increases, the incontinence can be related to the hypocontractile detrusor and large post void residuals (8).

**Presentation**

**Prenatal**

The diagnosis of posterior urethral valve in prenatal period is made by ultrasonography. Up to 80% of patients are diagnosed prenatally. The most severe obstruction with dilatation of the urinary tract may be detected as early as the 14th week of gestation. Half of the PUV foetuses are picked up before the second trimester scan.

The heightened awareness of the condition implies that virtually any male foetus with bilateral hydronephrosis assumed to have posterior urethral valves until proven otherwise. In addition to this concept there are some ultrasonographic features of posterior urethral valves that help in diagnosis and prediction of the severe cases.

The persistently distended foetal urinary bladder with the keyhole sign (image 3) is highly suggestive of PUV (9).

The gestational age at the detection of PUV before the 24th week, the thickened bladder wall, the hypergenicity of the renal parenchyma and oligohydramnios are all features predictive of poor renal function with an early onset of renal failure (2).
Neonatal

The neonatal presentation can be related to the effects of impaired renal function: poor feeding, listlessness, and irritability. Less commonly parents or careers pick up a poor urinary stream. Even less common is the urinary ascites that develops as a pop off mechanism in severe form of valvular obstruction.

Presentation in infancy

The presentation of PUV in the first year of life is mostly related to the urinary tract infections and urinary sepsis. Other signs include abdominal distention, palpable bladder or kidneys, poor urinary stream, urinary dribbling, failure to thrive or poor feeding.

Late presentation

Fifty per cent of boys with PUV over five year of age present with urinary incontinence or enuresis (10).

Diagnosis

History

The history of the patient with posterior urethral valves depends on the age of presentation.

In neonates the prenatal history based on ultrasononographic information should be obtained. As mentioned above the onset of the obstruction, the echogenicity of the renal parenchyma, constantly distended urinary bladder and oligohydramnios are not only suggestive of the diagnosis, but also predict the early onset of renal failure. In infants, there usually is a history of urinary tract infections or urinary sepsis. Symptoms such as poor weight gain, poor feeding can be suggestive of a degree of renal insufficiency.

Parents or caregivers can sometimes report on a poor urinary stream or urinary dribbling. The latter, lower urinary tract symptoms can also be elicited from the history of the majority of older boys with urinary incontinence due to posterior urethral valves.

Ultrasonography

Both in prenatal and postnatal period, posterior urethral valves can be suspected in the presence of bilateral hydronephrosis with ureteric dilatation associated with increased thickness of the bladder wall, hyperechoic renal
parenchyma and increased post void residuals (in older boys). All boys with suspected posterior urethral valves should undergo micturating cystourethrogram.

**Micturating cystourethrogram**

This remains a gold standard for the diagnosis of posterior urethral valves (Image 4).

On cystourethrogram the posterior urethra is always dilated and elongated. The more severe is the obstruction the greater the dilatation of the posterior urethra.

Some patients with severe obstruction regurgitate the contrast into the ejaculatory ducts and spermatic vesicles. The bladder neck is usually clearly demarcated. The urinary bladder wall is thickened with different degree of trabeculation and pseudodiverticulae.

27-55% of patients with PUV also have vesico-ureteric reflux (10).

Usually this is a gross secondary vesico-ureteric reflux into a much dilated ureter, renal pelvis, calyces and renal parenchyma.

A high grade unilateral vesico-ureteric reflux, a giant diverticulum of the bladder, the perinephric urinoma with subsequent urinary ascites are considered to be a "pop off" mechanism, which protects the renal parenchyma of the contralateral kidney by relieving the high pressures in the urinary tract.
**Differential diagnosis**

Obstruction of the male urethra can also be caused by rare conditions, such as anterior urethral valves, urethral diverticulum and syringocele.

Anterior urethral valves usually cause only lower urinary symptoms. However, there have been severe cases with a detrimental effect on renal and bladder function.

Urethral diverticulum is located at the level of peno-scrotal junction and presents with post micturitional dribbling or urinary tract infections.

Cowper's gland cyst or syringocele is usually presented with post micturitional dribbling in older boys.

Generally, saccular lesions of the male urethra may occur anywhere along the penile urethra including the fossa navicularis. Their presentation varies from mild obstructive symptoms to recurrent urinary tract infections and sometimes stone formation within the diverticulum.

**Treatment**

**Foetus**

Currently there are three possible ways to treat the antenatal bladder outlet obstruction in foetus:

1. In cases with an early, severe obstruction with marked dilatation of the urinary tract and oligohydramnios termination of pregnancy is undertaken.
2. A progressive dilatation of previously normally appeared kidneys in association with progressive oligohydramnios warrants prenatal urinary diversion (shunting).

A JJ stent is inserted under ultrasonographic guidance through the abdominal wall into the foetal bladder. The foetal urine is diverted into the amniotic cavity. The benefit of this treatment in terms of renal function remains controversial. Furthermore, there are significant complications associated with antenatal shunting such as shunt malfunction, damage to nearby structures and abortion. Despite this, properly selected for shunting midterm foetuses will certainly benefit from this intervention in terms of pulmonary development and some of them may even preserve their renal function in long term.

3. In cases where oligohydramnios occurs in later pregnancy elective preterm delivery with induction of lung maturity can be
recommended. In this cases a planned Caesarian Section should be performed in a referral centre with adequate neonatal, nephrological and paediatric urological support.

Neonate and infant

Certain steps are followed in a newborn with known posterior urethral valves before the definite resection is undertaken:

1. Bladder drainage with 6-8 Fr feeding tube inserted per urethra. Balloon catheters should be avoided due to the possibility of balloon distention within the posterior urethra instead of the bladder.
2. Immediate venous access. Blood sample for full blood count, serum urea, creatinine, potassium and sodium with subsequent monitoring.
3. Monitoring of the renal function with a record of fluids given by mouth and intravenously, monitoring catheter output and daily baby weighing
4. Aggressive treatment of electrolyte and fluid imbalance
5. Prevention and treatment of the urinary tract infection or urinary sepsis (antibiotics)
6. Valve ablation

Older child

Usually all that is needed in this group is valve ablation.

Valve ablation

In neonates the valve ablation should be undertaken promptly. This aims to minimize the risk of electrolyte disturbances and urinary tract infection. In an infant with previously treated urinary sepsis, sterility of the urine and stabilization of the fluid and acid-basis balance should be achieved prior to the operation.

Endoscopic valve ablation

The procedure is performed under general anesthetic. Valve leaflets should be first visualized. For this purpose 7-8 Fr cystoscope is used. The ablation is performed with the use of 8-9 Fr resectoscope armed with a specially designed cold knife or a right-angled diathermy hook. Valve leaflets incised from their free margin to the base at the 5 and 7 o'clock position, additional incision can be made at 12 o'clock position.
An alternative endoscopic approach in boys with very narrow urethra is achieved with 3FR Bugbee or Button electrode, which is inserted via 7Fr cystoscope. In this case valves are carefully cauterized using monopolar low cutting current.

Irrigation medium used during procedure should ideally be non-electrolyte and isosmotic. Although glycine solutions satisfy this criterion, in the majority of referral centres 0.9% NaCl is still widely used for the purposes of valve ablation.

The bladder overdistention should be carefully avoided. If the endoscopic technique is correct, bleeding is usually negligible. Foley’s balloon catheter is inserted postoperatively for 24-48 hours. Second look cystourethroscopy with or without additional valve ablation is performed two to four weeks after the initial surgery.

**Urinary diversion**

This mode should be reserved to particular rare situations, such as very small babies, patients with renal failure not improving after resolution of urethral obstruction and patients with refractory urinary sepsis.

Surgical techniques for urinary diversion in posterior urethral valves include bladder dome vesicostomy and low uni- or bilateral ureterostomy.

Posterior urethral valve ablation should not be combined with urinary diversion. A resulting "dry urethra" is responsible for the increased rate of postoperative urethral strictures.

**Follow-up and long term management**

The main and the most important parameter of the success in valve surgery is the clinical course of the patient. Good weight gain, continuous decrease in the serum creatinine and the absence of urinary tract infections are all suggestive of satisfactory pressure relieve.

In a majority of patients with vesico-ureteric reflux a postoperative MCUG, performed 2-3 months after valve ablation, shows complete resolution of reflux (11).

Independently on the severity of the initial condition, all boys with posterior urethral valve should be carefully followed-up through their adolescence into the early adulthood. Combined nephro-urological care should include regular height, weight and blood pressure measurement, ultrasonography of the urinary tract, urinalysis and urinary culture, determination of serum urea electrolyte and creatinine. Depending on the progress and symptoms additional tests are performed to monitor the lower urinary tract and renal function. These test include uroflowmetry, filling and
voiding cystometry (urodynamics), static or dynamic renal scintigraphy (DMSA or MAG3 scan), and estimation of the glomerular filtration rate.

**Scintigraphy**

DMSA and MAG3 renal scans aim to the determination of the renal function and scaring. MAG3 dynamic renography is less useful in the setting of PUV and is usually requested in patients with persistent upper tract dilatation.

**Urodynamics**

Voiding chart, uroflowmetry, video filling and voiding cystometry are particularly useful for follow-up in patients with PUV, after the age of acquisition of urinary and faecal control.

**References**