

CHAPTER 93

URETHRAL VALVES

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Introduction

Posterior urethral valves (PUVs) are a rare malformation in the subcollicular region of the male urethra. They cause varying obstructions of the lower urinary tract. The resulting secondary pathological conditions extend from mild—in part, clinically unmanifested—urinary dysfunctions to severe, bilateral changes in the upper urinary tract and associated renal failure. The literature puts the incidence between 1 in 5,000 and 1 in 8,000 male births. No clear hereditary association has been demonstrated, although a certain hereditary aspect is probable.¹ An incidence in identical twins and, very rarely, a familial cluster have been described in the literature. There are usually no concomitant malformations outside the genitourinary tract.

The modern literature on PUVs had its origin in the publications of Young et al. in 1919.² Their differentiation of three types of valves is a subject of debate, but is still used regularly. From the perspective of paediatric surgery, type I and type III urethral valves are of particular significance. In these, the flow of urine is opposed either by mucous membrane folds projecting “backdrop-like” into the lumen in lithotomy position from 7 to 5 o’clock directly below the seminal colliculus (type I), or by an aperture-like stenosis (type III).

A more recent description of the valve structures with the designation COPUM (congenital obstructive posterior urethral membrane) subsumes Young et al.’s type I and II valves into one class. Endoscopic observations agree well with this classification, when it is taken together with the different structure known as “Cobb’s collar”, which is clearly distal of the seminal colliculus.³

Aetiology

The embryological aetiology of PUVs is not clear. The physiological and morphological changes do not fit easily into the accepted idea of the development of the urethra out of parts of the embryonic cloaca, the urogenital sinus, and parts of the wolffian duct. Further, dysplastic changes in the kidneys in severe cases of urethral valves suggest a dysontogenetic aspect in the development of the wolffian duct derivatives beyond the purely secondary changes resulting from the distal obstruction. Histologically, it is possible to find parallels here to findings obtained in cases of multicystic diseased kidney (MCDK).⁴

In addition to PUVs, so-called anterior urethral valves in the more distally located part of the male urinary tract occasionally have been described. These may represent a separate condition; they appear extremely rarely and have not been described uniformly in the literature. Therefore, they will not be covered in this chapter.

Pathological Features

In regions with a well-developed prenatal diagnostic infrastructure, suspicious findings pointing to urethral valves can be obtained early in pregnancy. Severe secondary changes, such as megacystis and bilateral hydronephrosis, are already visible in the 10th to 12th week of gestation.⁵ For the most part, however, the pathological findings are obtained during the second trimester screening ultrasound (US) after the 20th week of pregnancy (Figure 93.1). At this point in utero, the kidneys

are largely developed. As a result, intrauterine measures to protect the foetal kidneys are of doubtful value. The complete sonographic urethral valve picture with megacystis (keyhole bladder), bilateral hydronephrosis, and oligohydramnios need not be present, even in the case of a pronounced finding of urethral valves. More frequently, one finds—in addition to a conspicuously large, rarely emptying bladder (see Figure 93.1)—a single-sided hydronephrosis. In some cases, the subvesical obstruction causes a backup of urine that is relieved over a rupture, usually spreading out into the fornix area of the kidney, or more rarely of the urinary bladder. These cases become conspicuous because of an associated urinoma or an associated ascites.⁶ This “pop-off” mechanism can bring about protection of the kidneys. If, as a result of the inadequate foetal excretion of urine, anhydramnios develops, the child shows the renofascial dysplasia known as Potter sequence. These children are not viable. In the case of oligohydramnios, as well, lung development and maturation of the foetus can be disturbed, which results in a disruption of postnatal adaptation.



Source: Courtesy of Kokila Lakoo, Oxford Children’s Hospital, Oxford, UK.

Figure 93.1: Foetal US (left) showing PUV and foetal magnetic resonance imaging (MRI; right) showing large bladder.

Neither the sonomorphology alone nor the point in time at which the secondary pathologies are determined are reliable predictors for the outcome of the patient. In addition, investigations of the foetal urine do not result in additional useful indications. The creatinine and urea of the foetus are excreted through the placenta through the mother’s kidneys, so only after the first day of life can the retention parameters of the child be taken as an expression of the child’s own renal functioning.

The natural course of the pregnancy when urethral valves are present does not result in a relevant premature delivery. In a case of megacystis or ascites causing displacement, foetoamniotic shunting or serial puncture can be considered. Experience with vesicoamniotic shunting has not been good due to frequent dislocation of the shunt. Serial amniotic fluid instillation also does not improve the prognosis. Because intrauterine interventions, including foetoscopic ablation of the valves, have to be carried out late in pregnancy, the prognostic advantage for the baby has to be questioned. In addition, as is often the case in prenatal therapy, we lack reliable prognostic factors for establishing reliable indication for therapy for individual cases. It is probable that prenatal diagnosis

of PUV and the immediate postnatal therapy improve outcome only in cases with the most pronounced findings.

Given the situation of medical care in developing countries, both prenatal and, perhaps, postnatal sonographic diagnosis cannot be placed in the foreground.⁷ In one reported series,⁸ all diagnoses were based on postnatal US, micturiting cystourethrography, and urethrocystoscopy in older children and adolescents. The great majority of patients become conspicuous due to the symptoms of urethral obstruction. These are mainly difficulty in urinating, recurring urinary tract infections (UTIs), or a palpable intraabdominal mass. This means that there can be a considerable delay in making a relevant diagnosis. The presence of associated urinomas or an associated ascites is possible. Special attention should be paid to urinary irregularities and to performing further diagnostic tests after recurring UTIs in boys.

Diagnostic Work-Up

Transperineal US can be used to detect urethral valves. However, the diagnosis of urethral valves is made on the basis of a voiding cystourethrography (VCUG) (Figure 93.2). This shows the dilated proximal urethra; the deformed, thick-walled urinary bladder with pseudodiverticuli; and (usually) the vesicoureteral reflux (VUR) (Figure 93.3). In cases of pronounced pathology of the upper urinary pathways involving disruption of urinary transport (grades IV–V), VUR can be absent. Even in the case of unclear findings with involvement of the upper urinary tract, diagnostic/therapeutic cystoscopy is recommended. Repeated determination of the retention parameters is necessary as a basis for estimating the global functioning of the kidneys. Initial pathologically raised retention parameters (serum creatinine > 1 mg/dl) and, more significantly, raised retention parameters later than two weeks after treatment are prognostically unfavorable results.⁹ For the evaluation of single renal clearance and, if necessary, to determine the extent of the urinary transport disturbance, the authors perform, after therapy at no earlier than the age of 6 weeks (corrected for gestation age), a technetium-99m mercaptuacetyl triglycine (Tc-99m MAG3) scan with a furosemide (Lasix®) load (Figure 93.4).

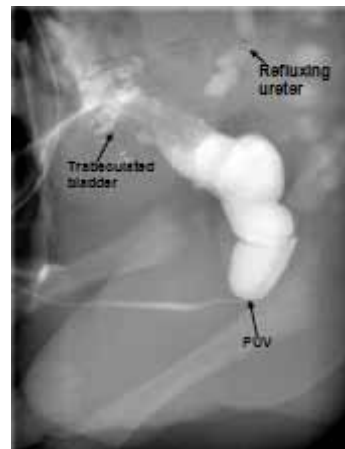
Treatment Protocol

On suspicion of urethral valves, treatment of the symptoms is more important than diagnostics. Symptomatic therapy generally consists of insertion of a transurethral or vesical catheter to drain off the urine. Normally, there is no difficulty in overcoming retrograde entrance of the valve structures with a catheter. If urine drainage is expected to last longer than 2–4 weeks, surgical urine drainage such as vesicostomy is preferable.

At every age, treatment consists of endoscopic ablation of the urethral valves.

1. Incision of the valve structures using a neodymium:yttrium aluminium garnet (Nd:YAG) laser over a 6 Fr cystoscope can be carried out even with the majority of prematurely born children. Fundamentally, laser ablation using the Nd:YAG laser in contact procedure with the bare fibre is preferred.
2. With endoscopic monitoring employing very precisely dosed energy, the valve structures are incised at 12, 5, and 7 o'clock with the patient in the lithotomy position.
3. A bladder catheter is inserted for 1 week. Before the patient is discharged, the result of surgery is checked once again cystoscopically.

The alternative surgical option is cold incision of the structures by using the endoscopic resectoscope, which is larger in size. Electrocoagulation (fulguration) is not recommended for infants due to the risk of urethral strictures, which have not been observed after use of the laser. Technically simpler methods for destroying the valve structures are the use of a balloon catheter (either blind or with x-ray monitoring) or a transurethral incision by using the Mohan urethral valvotome.¹⁰ In developing and resource-poor countries, catheter balloon avulsion, open



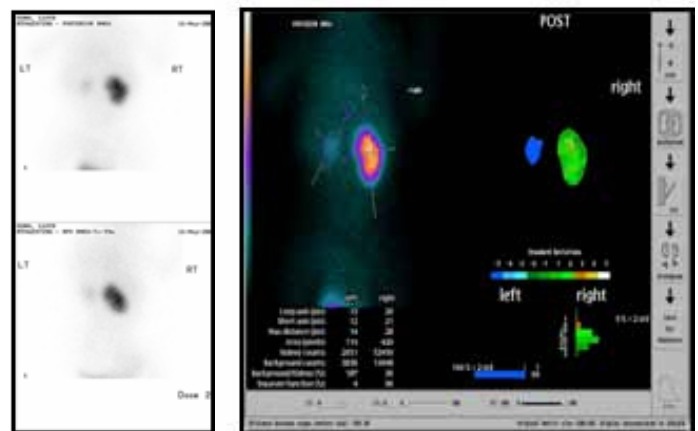
Source: Courtesy of Kokila Lakhoo, Oxford Children's Hospital, Oxford, UK.

Figure 93.2: Micturiting cystourethrography (MCUG) showing PUV and sequel.



Source: Courtesy of Kokila Lakhoo, Oxford Children's Hospital, Oxford, UK.

Figure 93.3: Postnatal US showing thick bladder wall.



Source: Courtesy of Kokila Lakhoo, Oxford Children's Hospital, Oxford, UK.

Figure 93.4: MAG3 scan showing 4% function in the left kidney and 96% in the right kidney.

(suprapubic) excision, and endoscopic transurethral fulguration of the valves remain the main modalities of treatment.

Endoscopic treatment of the valves is the only causal therapy. Restoration of unimpeded flow in the urethra is the necessary precondition for the physiological and functional rehabilitation of the urinary bladder and the upper urinary tract. However, successful treatment does not ensure that the secondary pathologies will be cured.

Since the obstruction was present and active prenatally, it leads to functional and structural changes in the urinary bladder and the upper urinary tract known as the valve-bladder syndrome.

After successful treatment, the authors have observed both a complete restoration of urine flow as well as chronic functional disturbances of the urinary bladder and the kidneys. In the authors' patient population, all boys with a continued pathological increase in the retention parameters 2 weeks after successful surgery developed a terminal renal failure before reaching the age of 11 years.⁹ The disorders in bladder function can be not only a lazy bladder, but also a hyperreflexive bladder of low capacity. The morphological correlates are a bladder with a large volume with high compliance and a significant amount of residual urine or a small bladder with a thick wall and low compliance. Urodynamic examinations of boys with a conspicuous pattern of excretion after the first year of life are conducted. An isolated enuresis nocturna as the sole symptom of a urethral valve disorder has not been observed. A subjective and objective reduced flow of urine requires clarification (sonography, VCUG, or possibly cystoscopy).⁸

If there is no possibility of a quick endoscopic treatment, surgical vesicostomy offers the possibility of an effective and technically simple temporary treatment.

From the perspective of maximal protection of the kidneys, a persistent restricted renal function can make introduction of a high diversion necessary. This can be achieved, for example, by means of a terminal cutaneous ureterostomy, loop ureterostomy, ureterostomy according to Sober, or pyelostomy. Catheter nephrostomy is suitable only for short-term relief due to the risk of infection. A vesicostomy *after* a successful treatment of the valves is not indicated and should be considered only in the case of nonrelevant, proximal disruption of urine transport.

Conclusion

In summary, in a case of conspicuous urethral valves—after immediate symptomatic treatment and carrying out the endoscopic procedure as quickly as possible following evaluation of the renal function—treatment of possible secondary pathologies such as VUR, renal flow obstruction, and voiding disorders should be undertaken in the first year of life, but as conservatively as possible. After the passage of a year, new examinations (sonography, VCUG, scintigraphy (if necessary), and urodynamics) are carried out to determine the course of the condition and whether there is need for a directed therapy. Any therapy here would focus on the individual primary conditions (ureteropelvic junction stenosis, ureteral orifice stenosis, severe vesicoureteral reflux, etc.) and be based on the criteria for their treatment. All children with initial restricted renal function—in particular, with persisting global renal restriction—have to be in nephrological/paediatric surgical (urological) outpatient care.

Studies have shown that, despite adequate surgical treatment, patients with an initial urethral valve symptomatology only rarely show no morphological or functional urinary tract pathologies on in-depth follow-up examinations. The prognosis of valve patients requiring transplants also depends on whether there are any persisting bladder dysfunctions. The bladder dysfunctions can be influenced therapeutically—for example, by medication. With this approach, it is possible to improve long-term prognosis in the treatment of posterior urethral valves.

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