

Posterior Urethral Valves with Severe Unilateral Vesicoureteral Reflux in a 3-year-old Boy
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SUMMARY

In patients with posterior urethral valves (PUV), severe unilateral vesicoureteral reflux (VUR) is one of the three conditions associated with preservation of renal function. Others are urinary ascites or urinoma in newborns and large congenital bladder diverticula. These conditions most likely provide a pop-off mechanism preventing the development of high intravesical pressure. Only 5% of patients with PUV and an associated pop-off mechanism will develop renal failure as opposed to 40 % of patients with PUV without a protective factor.

We present a 3-year-old boy with posterior urethral valves and a severe right unilateral vesicoureteral reflux. He had PUV ablation with Mohan's valvotomy and made satisfactory post operative recovery with indication on follow-up of preservation of renal function.

Keywords: *Posterior Urethral Valves, Unilateral Vesicoureteral Reflux, Hydrocalycosis*

Introduction

Congenital anomalies of the lower urinary tract are a significant cause of morbidity in infancy. Radiologic investigation is an important source of clinical information in lower urinary tract disorders but should not inconvenience the patient, expose the patient to unnecessary radiation, or delay surgical correction. In pediatric patients with suspected underlying urologic structural anomalies, screening ultrasonography is commonly the initial diagnostic study. If dilatation of the urinary tract is confirmed, micturating cystourethrography (MCUG) is performed to determine the presence of vesicoureteric reflux (VUR) and other causes of upper tract dilatation.¹

Posterior urethral valves (PUV) are by far the most common congenital obstructive lesion of the urethra, occurring mainly in phenotypic boys.¹ Young et al initially classified posterior urethral valves into three types, but it is now clear that there is only one type (formerly called type I).² MCUG is the best imaging technique for the diagnosis of posterior urethral valves.¹ VUR is the abnormal flow of urine from the bladder into the upper urinary tract. In the majority of cases, it occurs as a result of a primary maturation abnormality of the vesicoureteral junction or a short distal ureteric submucosal tunnel in the bladder that alters the function of the valve mechanism.³ VUR may be associated with PUV.⁴ Unilateral reflux, may occur in up to 35% of boys with PUV, and has been linked with protected renal function.⁵

This is a case of PUV with a severe unilateral VUR. The severity and unilateralism of the VUR prompted this report.

Case Report

A 3-year-old male was referred to the University College Hospital (UCH) Ibadan where he presented

with poor urinary stream since birth, urinary frequency and abdominal distension of 2 months duration. His parents first noticed his difficulty with micturition at 3 months of age; he usually strained at micturition with a poor urinary stream and terminal dribbling. These symptoms were associated with recurrent fever and failure to thrive.

Patient had groin surgery at 18 months of age, presumably a hydrocelectomy at a private hospital on account of scrotal swelling with no significant improvement.

He was later transferred to a mission hospital where he had an ultrasound examination for the first time which showed bilateral hydronephrosis and distended urinary bladder; an impression of obstructive uropathy with urinary retention from a possible PUV was made. He was later catheterized to relieve obstruction and referred for definitive management.

Examination revealed a young child, chronically ill-looking, wasted, pale and mildly febrile, on continuous urinary catheter drainage, with no pedal oedema. There were crepitations in both lung bases. The pulse rate was 160 beats per minute, with a haemic murmur. Both kidneys were ballotable. Patient's packed cell volume (PCV) was 21%, White Blood Count was 23,300/mm³. Urine microscopy and culture yielded *Pseudomonas* which was sensitive to Ciprofloxacin but resistant to Ceftriazone.

Abdominal ultrasound showed both kidneys to be enlarged and containing multiple communicating cysts, the right (Fig.1) was greater than the left and showing significant cortical thinning of the renal parenchyma in keeping with bilateral asymmetric hydrocalycosis, the ureters were also differentially dilated in favour of

the right. The urinary bladder showed a thickened and irregular wall and outline.

MCUG (Fig.2 and 3) revealed a dilated posterior urethra, irregular bladder outline with trabeculations, tortuous dilated right ureter and grade V vesicoureteral reflux.

Electrolyte and urea examination showed mild hyponatremia and acidosis. He was placed on Ceftriazone which was later changed to Ciprofloxacin based on culture results.

He improved significantly and had posterior urethral valve ablation with the Mohan's urethral valvotome under general anesthesia on the 12th day of admission. Post operative period was uneventful and patient was discharged home on urethral catheter to be followed up at the surgical out patient clinic.

He was later seen at the clinic with good urinary stream with no difficulty in micturition following removal of catheter 2 weeks post surgery. Follow-up abdominal ultrasound done 4 weeks afterwards showed a 1.3 cm reduction right renal length with persistent marked hydrocalycosis, the left however showed no significant change in renal size and with only minimal calyceal dilatation. Electrolytes, urea and creatinine values also obtained were within normal limits. An intravenous urogram (IVU) or a radionuclide renal imaging study was not requested to assess individual renal function.

The patient was subsequently lost to follow-up.

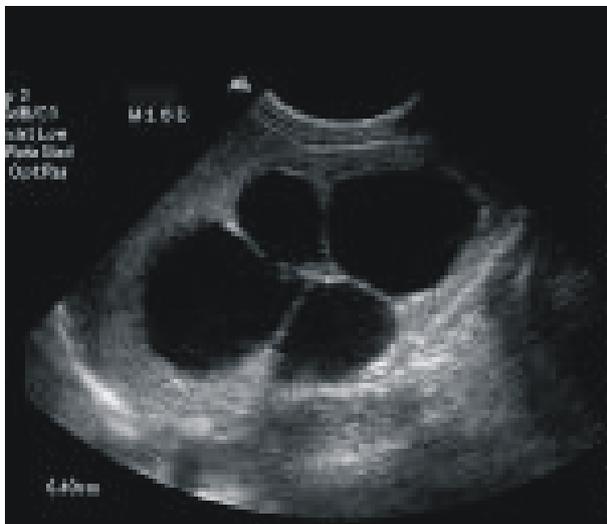


Fig.1 Ultrasound image showing the right kidney with markedly dilated calyces (c) and thinning of the renal parenchyma.



Fig. 2 MCUG image (antero-posterior view) showing a dilated posterior urethra (arrow), bladder trabeculations (*) and right vesicoureteric reflux. The dilated posterior urethra assumes a “spinning top”/ “electric bulb” appearance (arrow).



Fig. 3 MCUG image (antero-posterior view) showing massive reflux with tortuosity of entire dilated right ureter and ballooning of the calyceal system.

Discussion

Management of PUV remains a clinical challenge, requiring active management from infancy into adulthood to avoid progressive dysfunction and deterioration of both the upper and lower urinary tracts.⁶

PUV results from the formation of a thick, valve-like membrane from tissue of Wolffian duct origin that course obliquely from the verumontanum to the most distal portion of the prostatic urethra. In essence, the valve is a diaphragm, but because it is more rigid along its line of fusion, progressive distention during voiding causes it to become bilobed or sail-like.⁷

MCUG is the best imaging technique for the diagnosis of posterior urethral valves. Radiologic findings include dilatation and elongation of the posterior urethra and, occasionally, a linear radiolucent band corresponding to the valve.⁸ The bladder neck becomes hypertrophic and appears narrow in relation to the dilated posterior urethra. Any cause of bladder outlet obstruction such as PUV will cause bladder trabeculation or thickening of the wall (Fig 3). VUR with gross hydronephrosis, dysplastic kidneys, and urine ascites are common findings.⁸ VUR occurs in nearly 50% of patients. Bladder trabeculation, hypertrophy, and diverticula are also demonstrated at MCUG.^{7, 8}

Prenatal ultrasound is actually the usual method of detecting posterior urethral valves.⁹ Abnormalities are most evident when the valves cause severe obstruction and these may include oligohydramnios, bladder distention, and, occasionally, fetal posterior urethral dilatation.

PUV represents a spectrum of obstruction severity. The degree of obstruction caused by this abnormality varies considerably depending on the configuration of the obstructive membrane within the urethra. The morbidity of PUV is not limited merely to transient urethral obstruction; however, the congenital obstruction of the urinary tract at a critical time in organogenesis can have a profound and lifelong effect on kidney, ureteral, and bladder function. Diagnosis usually is made before or at birth when a boy is evaluated for antenatal hydronephrosis. Before the era of prenatal sonography, PUV was discovered during evaluation of urinary tract infection (UTI), voiding dysfunction, or renal failure. In the case presented even though symptoms were present since birth diagnosis was not made until much later after episodes of UTI, urinary retention, and hydronephrosis.⁶ While rare, adult presentation of PUV has been described in case reports,^{10, 11} with symptoms varying from obstructive voiding symptoms to post ejaculatory dysuria.

In the presonography era, late presentation of PUV was considered a good prognostic sign suggestive of a lesser degree of obstruction. Diurnal enuresis in boys older than 5 years, secondary diurnal enuresis, voiding pain or dysfunction, and decreased force of stream

may indicate the presence of PUV. It is sometimes discovered during evaluation of abdominal mass or renal failure. Hydronephrosis or proteinuria found on examination for unrelated conditions may be the first sign of PUV.

VUR commonly is associated with PUV as demonstrated in this patient. In the majority of cases, it occurs as a result of a primary maturation abnormality of the vesicoureteral junction or a short distal ureteric submucosal tunnel in the bladder that alters the function of the valve mechanism.³ This patient had severe unilateral VUR, characterized by gross dilatation and tortuosity of the right ureter; gross dilatation of the renal pelvis and calyces; papillary impressions were no longer visible in the majority of the calyces, regarded as a Grade V, based on the International Reflux Committee Study.¹²

Its unilateral nature may be due to the preservation of the sphincteric tone and the function of the valve mechanism at the vesicoureteral junction on the left. Regardless of the age of the patient, the existence of bilateral reflux at the time of presentation implies a worse prognosis than does unilateral or no reflux.¹³

VUR associated with PUV, is generally secondary to elevated intravesical pressures. Recurrent UTIs are common in patients with PUV. Elevated intravesical pressures predispose patients to infection, possibly by altering urothelial blood flow.

Additionally, patients with PUV may have elevated post-voidal residual urine volumes, leading to stasis of urine. Dilated upper urinary tracts, with or without VUR, further elevate the risk of UTI. UTI management is usually directed at lowering bladder pressures (anticholinergic medication), lowering post-voidal residual urine volume (via clean intermittent catheterization), and, at times, administering prophylactic antibiotics.⁶ VUR predisposes to pyelonephritis because it carries bacteria from the bladder to the upper urinary tract.¹⁴ This was well demonstrated in this patient who had recurrent intermittent fevers and whose urine culture grew *Pseudomonas*. The majority of pediatric patients who develop renal scars after a urinary tract infection have VUR, and higher grades of reflux are associated with an increase in parenchymal scarring.¹⁵ Detection of VUR in neonates and infants is particularly important because these patients are more predisposed to the formation of renal scars than are older children.¹⁶

Reflux nephropathy is a common cause of renal failure; therefore, it is important that this condition be detected as early as possible to allow prompt prophylactic antibiotic treatment and hopefully reduce the risk of scarring and reflux nephropathy.¹⁷ Reflux is also the most common cause of antenatal hydronephrosis, being responsible for 40% of intrauterine cases.¹⁸

MCUG should be used as was done in this case to document the presence of VUR and to determine the

grade of reflux and whether reflux occurs during micturition or during bladder filling.⁶

Eklof *et al*,¹⁹ in a series of 65 male infants and children, all with the diagnosis of PUV, showed that preoperatively diagnosed impairment of kidney function and concomitant dilatation of the upper urinary tract, with some exceptions remained fairly unchanged at postoperative examinations. In the case of marked VUR, permanent kidney function annihilation was significantly commoner than with slight or no reflux. Our patient's post operative ultrasound assessment showed slight reduction in renal size and only minimal change in hydrocalycosis. The post operative clinical and biochemical follow-up status were however satisfactory in the short term, Shittu *et al* in the same setting had shown that following PUV ablation with Mohan's valvotome, complications are not common but may occur in the long term²⁰. In the absence of renal radionuclide imaging studies, it is difficult to conclude that the refluxing kidney has retained renal function however this is most likely considering the study of Donnelly *et al*⁵ in which all eight patients with unilateral reflux studied had normal renal function on long-term follow-up. This also gives credence to the assertion that unilateral VUR in PUV is associated with protected renal function. Their study also showed that apart from significant renal function being present in the refluxing kidney, the function may even increase with time⁵. These findings suggest that, contrary to previous reports^{21,22}, the refluxing kidney in patients with PUV is not always dysplastic.

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