Profile of boys with posterior urethral valves from a tertiary care center in a developing country

Ashish Jain, Mukta Manthan, Nisha Kumari, Srijan Singh

From Department of Neonatology, Maulana Azad Medical College, University of Delhi, Central Health Service, Government of India, New Delhi, India

Correspondence to: Dr. Ashish Jain, Department of Neonatology, Ward 38, LNJP and Allied Hospitals, Gate No 2 LNJP Hospital, 2 Bahadur Shah Zafar Marg, New Delhi – 110 002, India. EXT: 1245. E-mail: neoashish2008@gmail.com

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ABSTRACT

Introduction: Posterior urethral valves (PUVs) are the most common cause of obstructive uropathy in boys. While most patients in developed countries are diagnosed in the antenatal period, our patients often present late which adversely affects their outcome. This retrospective study was aimed to study the clinical profile of boys with PUV at follow-up. **Methods:** Records of 45 boys with PUV who were in follow-up for at least 1-year were retrieved between February and December 2015. Age of presentation, signs and symptoms, anthropometry, radiological findings, surgical interventions, and biochemical investigations were recorded in a pre-structured pro forma. The data were later analyzed on an Excel spreadsheet. **Results:** The mean age at the diagnosis of PUV was 16±2.8 months, and the mean age at enrollment was 70.6±53.5 months. The condition was detected antenatally in 5 (10.9%) subjects only. At the time of data collection, the mean height and weight standard deviation scores were −1.7 and −1.5, respectively. Hypertension was present in 29% of patients. 44.5% of subjects had moderate-to-severe malnutrition and 42.2% had short stature. Mean GFR of the subjects was 66.3 ml/min/1.73m². Bilateral hydronephrosis was present on ultrasound in 26 (57.8%) subjects. Mean anteroposterior diameter on ultrasound was 22.1 mm for the right kidney and 20.8 mm for the left kidney. Significant post-void residue was detected in 54.6% of patients. Bilateral reflux was seen in 21.7% of subjects on micturating cystourethrogram. Urodynamic studies were available for 16 patients, and 31.3% had significant post-void residual urine. Detrusor instability was present in 18.8% of subjects. 35.6% of subjects underwent valve fulguration alone as the surgical procedure while 19 (42.2%) had a diversion procedure (vesicostomy/ureterostomy) along with valve fulguration. **Conclusions:** Most of the patients with PUV are still diagnosed postnatally and have significant renal damage at presentation. Urinary diversion procedures are required in almost half of these patients. All these contribute to a poor long-term outcome.

Key words: *Posterior Urethral Valves, Tertiary*

Posterior urethral valve (PUV) is the most common cause of obstructive uropathy in male infants leading to end-stage renal disease. Its incidence in India is not known, but the overall incidence published is around 1:5000–1:8000 male births [1]. Most of the patients in the developed countries are diagnosed during the antenatal period. However, in developing countries, these are rarely diagnosed early. Most of the patients present after birth with either urinary stream abnormalities or with an episode of urinary tract infection. The presence of obstruction, especially in the antenatal period, compromises on the growth of the kidneys and the urinary tract during organogenesis. Besides, due to obstruction, the bladder wall is exposed to high pressures that stimulate increased collagen deposition and also causes muscle hypertrophy. Both these processes thicken the bladder wall which compromises the bladder compliance. Relieving the bladder outlet obstruction at the earliest is the best treatment.

There are few studies from India describing the long-term outcome of the children with PUV, but they mostly describe the surgical aspects [2-4]. A recent study looked at the outcome of all children diagnosed in the antenatal period and concluded that early diagnosis improves the outcome [5]. The present study was conducted to study the epidemiology, clinical profile, management, and long-term outcome of boys with PUV that presented to our center.

METHODS

In this retrospective study, records of all children, who presented with PUVs in the previous 6 years (2010–2016) and who were in follow-up for 1 year, were reviewed. Approval for the study was granted by the institute's ethical committee. Forty-five boys with PUV between the ages of 15 days and 12- years were enrolled for

the study. Information was collected from the patients' hospital records, including age of presentation, age at diagnosis, and signs and symptoms at initial presentation. The anthropometry of the subjects was recorded. The latest results of blood urea and serum creatinine were recorded. The reports of ultrasonography and micturating cystourethrogram (MCUG) done at initial workup and later in follow-up were recorded. The dimercaptosuccinic acid scan, diethylenetriaminepentaacetic acid scan, and urodynamic study findings were recorded in a pre-structured pro forma.

Antenatal ultrasound reports were recorded if reports were available either from the hospital records or from the original ultrasound report. The GFR was calculated using the Schwartz formula [6]. Subsequently, the patients were classified into stages 1–5 of chronic kidney disease (CKD). The presence of rickets, hypertension (systolic and/or diastolic blood pressure >95th centile for gender, age, and height), malnutrition (weight for age <−2 standard deviation [SD]), and short stature (height for age <−2SD according to the WHO classification) were recorded [7]. The surgical procedures performed on the subjects and the outcomes at last follow-up were recorded. The data were entered into an Excel spreadsheet and analyzed using descriptive statistics.

RESULTS

A total of 45 boys attending the outpatient services of the division of pediatric nephrology of the department of pediatrics were included in the study. The mean age at diagnosis was 16±2.8 months, and the mean age at enrollment into the study was 70.6±53.5 months. At the time of data collection, the mean height and weight SD scores were −1.7 and −1.5, respectively. The baseline characteristics of the study subjects are given in Table 1. PUV was detected antenatally in only 5 (10.9%) subjects. At the time of diagnosis, majority of the patients (22.2%) presented with dribbling of urine, 8.9% had urinary incontinence, and another 8.9% presented with urinary tract infection. Only 6.7% of subjects had oliguria and 8.9% had acute kidney injury. Remaining 33.5% had presented with multiple symptoms (dribbling of urine, urinary incontinence, urinary tract infection, oliguria, and acute kidney injury). At the time of data collection, 33% (15/45) of the boys had moderate-to-severe malnutrition, while short stature was present in 42% (19/45) of boys and 29% (13/45) were hypertensive.

The mean GFR of the study subjects was 62.6 ml/min/1.73m². On radiological investigations, bilateral hydronephrosis was present on ultrasound in 26 (57.8%) subjects at initial presentation. The mean anteroposterior pelvic diameter on ultrasound was 22.1 mm for the right kidney and 20.8 mm for the left kidney indicating moderate hydronephrosis. Significant post-void residual urine was detected in 53.3% of patients at initial assessment on sonography. Bilateral vesicoureteric reflux (VUR) was detected in 20% of subjects on initial MCUG. The urodynamic studies were available for 16 patients, and of these, 5 (31.3%) had significant post-void residues; detrusor instability was present in 3 (18.8%) subjects. The details of investigations are described in Table 2.

Table 1: Baseline characteristics of children with posterior urethral valves

Parameter	Values mean (SD)
Clinical	
Age at diagnosis	16 (2.8) months
Age of enrollment	70.6 (53.5) months
At the time of data collection:	
Weight mean (SD)	17.2 (11.4)
Height mean (SD)	103.7 (30.5)
Patients (%) with malnutrition	20 (44.4%)
Patients (%) with short stature	19 (42.2%)
Rickets	6 (13.3%)
Patients with hypertension	13 (29%)
Biochemical	
Serum creatinine	(0.86) mg/dL
Glomerular Filtration Rate	62.6 (42.4) ml/min/1.73m²
Stages of CKD	
Patients in CKD Stage 1 and 2	16 (35.6%)
Patients in CKD Stage 3, 4, and 5	29 (64.4%%)

CKD: Chronic kidney disease, SD: Standard deviation

Table 2: Radiological findings in the subjects

Investigations	Proportion n (%)
Ultrasonography	n=45
B/L hydronephrosis	26 (57.8)
U/L hydronephrosis	8 (17x. 8)
Significant post-void residue	24 (53.3)
MCUG	
U/L Vesicoureteric reflux	16 (35.6)
B/L Vesicoureteric reflux	9 (20)
Bladder back pressure changes	29 (64.4)
Dilated posterior urethra	16 (35.6)
Decreased bladder capacity	2 (4.4)
Urodynamic studies	(n=16)
Post-void residual	5 (31.3)
Detrusor instability	3 (18.8)

MCUG: Micturating cystourethrogram

Sixteen (34.8 %) subjects underwent valve fulguration alone as the surgical procedure while 18 (41.3%) had a diversion procedure (vesicostomy/ureterostomy) along with valve fulguration. Remaining patients had not undergone surgery at the time of presentation. At the last follow-up, 16 (35.6%) subjects were detected with CKD stages 1 and 2 while 29 (64.4%) were in stages 3–5.

DISCUSSION

Our study describes the clinical profile, management, and long-term outcome of 45 children with PUV presented to our hospital over a period of 5 years. Most of the patients in this study were diagnosed with PUV only after birth. This is similar to several studies published in the past from developing countries [6-8] where very few cases were diagnosed antenatally. Age at presentation is known to be an important prognostic factor, as those diagnosed

antenatally are seen to have preserved renal functions due to early intervention [9].

Majority of the children in our study presented with dribbling of urine followed by urinary incontinence, urinary tract infection, and acute kidney injury; and very few presented with oliguria and voiding dysfunction. This was comparable to numerous studies published in the past [10-12]. Nearly half of our patients had stunting and one-third had moderate-to-severe malnutrition according to the WHO classification. This was similar to Uthup *et al.* [13] and Tejani *et al.* [14] who have reported growth failure in 30–40% of children with PUV. Gangopadhya*y et al.* [15] from India have also reported height below 50th centile in 50% of their patients. Therefore, nearly 50% of children with PUV are known to have growth failure.

The presence of VUR in PUV patients is supposed to have a poorer prognosis as it causes postnatal renal damage in these children [15]. In our study, 20% of children had bilateral VUR [16]. VUR is also considered as an important risk factor for developing late-onset renal failure [17]. Its role is still controversial as there are numerous studies where no such correlation has been documented [18]. Of the 16 patients whose urodynamic studies could be done, detrusor instability was found in 18% of patients and significant PVR in 30% of children. Detrusor fibrosis causes decreased bladder compliance and detrusor instability, which leads to recurrent UTI and incomplete emptying of the bladder [19]. Therefore, bladder dysfunction is known to be one of the major factors contributing to long-term outcome of these children [20].

In our study, 35% of patients underwent valve fulguration alone as the surgical procedure while 41.3% also had a diversion procedure (vesicostomy/ureterostomy) along with it. The preferred treatment of PUV is direct fulguration of valves without prior diversion procedures [21,22]. Delayed presentation could be a major factor attributing to a high percentage of children undergoing diversion procedures and poorer long-term outcome.

The strength of our study was that we have described the epidemiology, clinical profile, and long-term outcome of PUV in Indian population. Some limitations of our study were retrospective collection of information for our patients and lack of long-term follow-up. PUV is one of the most common causes or renal failure in children. Majority are diagnosed after delivery with a very small percentage diagnosed antenatally in developing countries. Delayed presentations, elevated serum creatinine at presentation, presence of VUR, and detrusor instability are considered as poor prognostic factors. Urinary diversion procedures are required in almost half of these patients. All these contribute to a poor long-term outcome.

CONCLUSIONS

Most of the patients with PUV are still diagnosed postnatally and have significant renal damage at presentation. Urinary diversion procedures are required in almost half of these patients.

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