



PRESENTATION, MANAGEMENT, AND OUTCOME OF POSTERIOR URETHRAL VALVES IN WESTERN RAJASTHAN

Urology

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ABSTRACT

Introduction: Posterior urethral valve (PUV) is the most common cause of bladder outlet obstruction and renal insufficiency in male children. The aim of this study was to evaluate the presentation, management, challenges, and outcome of the disease in a Western Rajasthan tertiary health institution. **Methods:** In a retro-prospective study, 26 male children were included from November 2017 to March 2020 at Dr. S.N. Medical college, Jodhpur. Diagnosis was confirmed by voiding cysto-urethrography (VCUG). **Results:** A total of 26 patients were managed for PUV. The mean age of presentation was 3.45 years with 65.8% of the patients presenting after 1 year. Voiding dysfunction noted in 23(88.4%) patients was the most common presentation. Most common finding on physical examination was palpable bladder 15(57.6%) while UTI noted in 15 (57.6%) patients was most common complication. Abdominal sonography revealed dilated posterior urethra in 10(38.4%) cases, while VCUG revealed a dilated posterior urethra in 19 (75%) cases. The creatinine value at presentation ranged from 0.4-4.5 mg/dl with a mean of 1.10 ± 0.95 mg/dl. Valve ablation with a diathermy bugbee electrode & holmium laser provided relief of obstructions in the 23 (88.4%) patients who underwent the procedure without immediate complication. The period of follow-up ranged between 1 weeks to 2 years with a mean of 8.2 months. There was sustained improvement in urine stream, reduction in the mean creatinine concentration and incidence of UTI during follow-up. **Conclusion:** Many patients with PUV presented late within the reviewed period. Valve ablation provided relief of obstruction in most of the cases. Efforts at improving awareness among general population and early diagnosis and referral among the health team should be encouraged. There is a need to counsel parents on the need for long-term follow-up.

KEYWORDS

Posterior urethral valve, ablation, VCUG

INTRODUCTION

Posterior urethral valve (PUV) is a congenital anomaly of proximal urethral development that is characterized by the presence of abnormal obstructing urethral membranes. It occurs in male children, leading to obstruction in the outflow of urine with varying degree of urinary tract dysfunction.[1]. It is believed to arise mainly as a result of an anomalous insertion of the mesonephric duct into the primitive fetal cloaca.[1,2]

The disease has an incidence of 1:5000-1:8000 male births. It is the leading cause of bladder outlet obstruction and renal insufficiency in male children.[1,3] PUVs are classified in three types: Valves representing folds extending inferiorly from the verumontanum to the membranous urethra (Type 1), Valves as leaflets radiating from the verumontanum proximally to the bladder neck (Type 2.) and Valves as concentric diaphragms within the prostatic urethra, either above or below the verumontanum (Type 3). The most common type is type 1. [4,5,6] The first clinical case of PUV was reported by Young in 1913. The clinical features of the disease include poor stream and dribbling of urine after birth. The patients may also present with complications of the disease such as urinary tract infections, sepsis, impaired renal function, anemia, and failure to thrive.[1,3,7]

Diagnosis of the condition may be suspected with an antenatal ultrasound scan in the presence of fetal proximal urethral dilatation, hydronephrosis (usually bilateral) and oligohydramnios. However, diagnosis is, usually, confirmed postnatally with voiding cysto-urethrography (VCUG) which shows the classical posterior urethral dilatation and bladder trabeculation.[3,7,8]

The standard management of the condition is endoscopic valve ablation.[9] This leads to the establishment of satisfactory urine stream in most cases; however, patients need to be on long-term follow-up for assessment of renal and bladder function.[10] Indications for diversion in PUV are: prematurity, small body size, severe urinary infection/septicemia, and high grade renal insufficiency.[11] Vesicostomy is an approach which protects upper urinary tract complications.[12]

PUV has detrimental effects on kidney, bladder and the entire urinary system. This result from persistent and unrelieved pressure on the bladder leading to bladder diverticular, hydronephrosis and chronic to

end-stage renal diseases (ESRD). The anomaly is associated urosepsis, overflow urinary incontinence, chronic kidney diseases (CKD), hypertension, chronic anemia, failure to thrive, poor quality of life and even death.[13,14].

AIM AND OBJECTIVE

The aim of this study is to describe the epidemiologic, diagnostic and therapeutic aspects of PUV seen in our institute in Western Rajasthan.

MATERIALS AND METHODS

A retro-prospective study of 26 consecutive children managed for PUV between November 2018 and March 2020 were carried out at Dr. S. N. Medical college, Jodhpur.

Information on the age at presentation, clinical features, and anthropometric measures were noted. Blood urea nitrogen, serum creatinine, blood gas analysis and urine routine microscopy and cultures were done. Initial imaging included abdominal ultrasonography followed by a voiding cystourethrogram. VCUG showing dilated elongated posterior urethra and radiolucent valve cusps was considered diagnostic of PUV.

After stabilizing the patients by controlling infection, correction of fluid, electrolyte balance, definitive valve ablation was done. After valve ablation, a urethral catheter was left in situ for 3–5 days to allow the oedema to subside and to enable measurement of urinary output. Patients were then subsequently discharged on antibiotic prophylaxis and followed up in an outpatient clinic with voiding history, abdomino-pelvic ultrasonography, and serial serum creatinine estimation. The period of follow-up ranged between 1 weeks and 2 years with a mean of 8.2 months, however only in five patients was this period >6 months while rest of patients didn't turn up for follow up visits.

All data in relation to the study objectives were recorded and analyzed. Ethical clearance was obtained from the ethical committee and the institutional review board of Dr. S.N. Medical College, Jodhpur.

RESULTS

A total of 26 males was managed for PUVs during the period. Their ages at presentation ranged between 1 day and 14 years with a mean of

3.45 years. 3 (11.5%) patients presented before the age of 1 month, 6 (23.2%) between the ages of 1 month and 1 year, while 8 (30.7%) presented in the age group of 1 year-5 years and 9 patients presented with age above 5 years [Table 1]. Prenatal diagnosis with the aid of a maternal ultrasound scan was made in 2 (7.6%) patients. Poor stream/dribbling of urine noted in 23(88.4%) patients was the most common mode of presentation while hematuria recorded in 2 (7.6%) was the least [Table 2].

Table 1: Age at diagnosis

Age group	Number of patients	Percentage
Neonate	3	11.5
1 month-1 year	6	23.2
1 year-5 years	8	30.7
>5 years	9	34.6

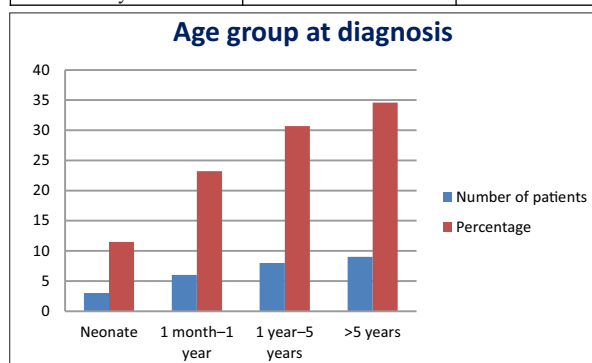
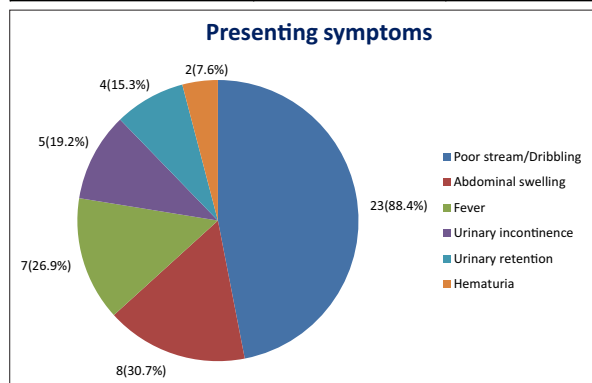


Table 2 : Presenting symptoms in the patients

Symptoms	Number of patients	Percentage
Poor stream/Dribbling	23	88.4
Abdominal swelling	8	30.7
Fever	7	26.9
Urinary incontinence	5	19.2
Urinary retention	4	15.3
Hematuria	2	7.6



The most common finding on physical examination was a palpable bladder in 15 (57.6%) patients while the least was generalized edema in 2 (7.6%) patients. (Table 3)

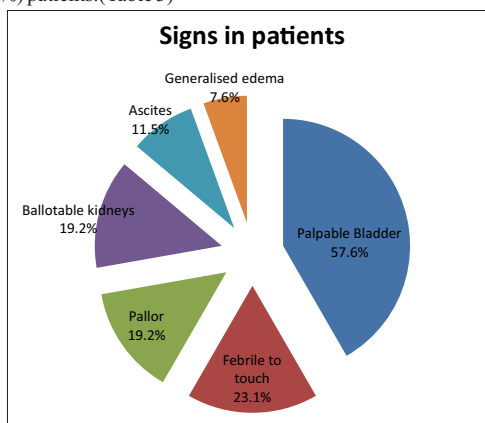


Table 3: Signs in the patients

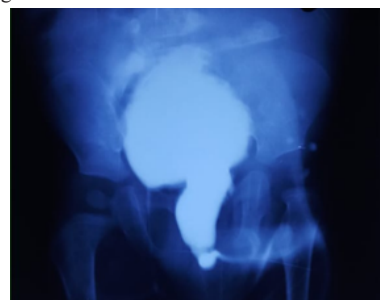
Sign	Number of patients	Percentage
Palpable Bladder	15	57.6
Febrile to touch	6	23.1
Pallor	5	19.2
Ballotable kidneys	5	19.2
Ascites	3	11.5
Generalised edema	2	7.6

Urinary tract infection noted in 15 (57.6%) patients was the most common complication of the disease followed by anemia (30.7%), Septicemia (11.5%), Renal failure (11.5%), and Hypertension (7.6%). (Table 4)

Table 4: Complications of posterior urethral valves in the patients

Complications	Number of patients	Percentage
Urinary tract infections	15	57.6
Anemia	8	30.7
Septicemia	3	11.5
Renal failure	3	11.5
hypertension	2	7.6

Abdominal ultrasonography revealed dilated posterior urethra in 10 (38.4%) patients, bilateral hydronephrosis in 20 (76.9%), bilateral hydroureter in 5 (19.2%), and thickened bladder wall in 18 (69.2%). Micturating cystourethrogram revealed a dilated proximal urethra in 19 (75%) cases, diverticulum in 3 (11.5%) and vesicoureteric reflux in 5 (19.2%) (3 bilateral, 2 unilateral). There were 16 positive urine cultures, with *Escherichia coli* noted in 12 cases being the most common organism isolated.



MCUG showing dilated posterior urethra

The creatinine concentration at presentation ranged between 0.4 mg/dl and 4.5 mg/dl with a mean of 1.10 ± 0.95 mg/dl. 8 (30.7%) patients had creatinine value ≥ 1 mg/dl.

Preoperatively, urethral catheterization was done in 15 (57.6%) patients, while 2 (7.6%) patients had percutaneous nephrostomy. 2 (7.6%) patients had vesicostomy prior to valve ablation. 23 patients had endoscopic valve ablation with the aid of a bugbee electrode and holmium laser. Of these, 22 (95.6%) had type I PUVs while 1 (4.4%) had type III. Improvement in urine stream was recorded in the above patients after valve ablation, although one required a repeat surgery to achieve satisfactory urine stream. No significant immediate postoperative complications were noted.

The period of follow-up ranged between 1 weeks and 2 years with a mean of 8.2 months, however, only in 5 patients was this period >6 months. Six weeks post ablation there was no clinical or microbiological evidence of UTI in 12 out of the 15 patients that presented initially with this complication. The mean creatinine concentration after 3 months was 0.8 ± 0.48 mg/dl. Ultrasonography showed persistence of hydronephrosis up to 2 years after valve ablation in two cases. Repeat MCUG study was done in the course of follow-up after 3 months.

DISCUSSION

Posterior urethral valve remains a significant cause of bladder outlet obstruction and renal insufficiency in male children. [3,7] The study aimed at evaluating the clinical presentation, management, and outcome of the disease in our institution.

The mean age of presentation (3.45 years) suggests that patients with this condition present late in our environment. More than half of the patients managed within the period presented after the age of 1 year.

Similar findings were also noted in previous studies in southwestern and eastern Nigeria in which 57.1% and 52% of patients, respectively, presented after the age of 1 year as well as in study by malik et al., but in contrast to study by choudhary et al.[4,7,15,16] Factors which may have contributed to delayed presentation include suboptimal prenatal ultrasound scan utilization, poverty, ignorance on the part of parents, and paucity of specialist care periphery areas.

Voiding dysfunction was the most common reason for presentation. Abdominal distension noted was due to the presence of ballottable hydronephrotic kidneys and urinary ascites while the presence of fever was mostly due to complications such as UTI and septicemia. UTI was the most common complication. This is similar to studies by *Nasir et al.*, *Mirshemirani et al.*, *Orumuah et al.*, *El-sherbiny et al.*, *Tambo et al.*[1,19-22]

Urinary drainage by means of urethral catheterization was done to improve renal function and to allow optimization prior to surgery. Two of such patients also had percutaneous nephrostomy on account of severe hydronephrosis. Vesicostomy was done in patients whose urethra could not accommodate the available telescope, in those who presented during a brief period of instrument failure and as a way of diversion. Because of the availability of bugbee electrode and laser fiber and small size cystoscope, our diversion rate was low, 15.3% same as other studies like *Orumuah et al.* and *Sudarsanan et al.*[20,23] but in contrast to various Indian studies like *Choudhary et al.*, *Lal et al.*, *Shittu et al.* where diversion rates were very high.[16]

Endoscopic valve ablation, the gold standard in the definitive management of PUV was done in our patients with the aid of a diathermy bugbee electrode and holmium laser. This provided relief of obstruction and improvement in urine stream of the patients without any significant sequelae. However, one patient required a repeat valve ablation to attain satisfactory passage of urine. Complications such as urinary retention, urethral bleeding and urethral stricture have been reported to be associated with the procedure.[9] However, these were not recorded in our patients within the period of follow-up for which further long follow up is needed same as *orumuah et al.*[20]

Most of the patients were lost to follow-up. It may be due to the reluctance of parents to continue hospital visits in the presence of satisfactory urine stream. The absence of obvious complications, long distance from the hospital and the financial burden of repeat investigations may have also led to the short mean period of follow-up.

Limitation

The applicability of our study may be limited by relatively small sample size of 26 patients. We recommend a large multicentre case-control or cohort study to assess the relationship of most of the variables associated with PUV.

CONCLUSION

Patients of PUV presented late to our institute during the period reviewed. VCUG is still the gold-standard imaging modality for documenting PUV. The factors like renal dysplasia and UTI have their role in final outcome. Valve ablation was very helpful in establishing satisfactory urine flow in affected patients. However, there is a need to further educate parents on the symptoms of the disease and the benefits of long-term follow-up of patients.

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