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Presentation, management, and outcome of posterior urethral valves in a Nigerian tertiary hospital

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ABSTRACT

Background: Posterior urethral valves (PUV) remain 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 Nigerian tertiary health institution. **Patients and Methods:** Retrospectively, medical records of male children with a diagnosis of PUVs over a 10 year period (2003-2012) were retrieved. All data in relation to the study objectives were recorded and analyzed. **Results:** A total of 44 patients was managed for PUV within the period. The mean age of presentation was 3.95 years with 56.8% of the patients presenting after the age of 1 year. Voiding dysfunction noted in 40 (91.0%) patients was the most common mode of presentation. The most common finding on physical examination was a palpable bladder while urinary tract infection noted in 23 (52.3%) patients was the most common complication noted. Abdominal ultrasonography revealed dilated posterior urethra in 16 (36.4%) cases, while micturating cystourethrogram revealed a dilated proximal urethra in all 35 cases in which it was done, diverticulum in 6 and vesicoureteric reflux in 9. The creatinine value at presentation ranged between 0.4 mg/dl and 4.0 mg/dl with a mean of 1.02 ± 0.93 mg/dl. Urethroscopy in 37 patients confirmed type I and type III PUV in 35 and 2 patients, respectively. Valve ablation with a diathermy bugbee electrode provided relief of obstructions in the 37 patients who underwent the procedure without any significant immediate complication. The period of follow-up ranged between 2 weeks and 3 years with a mean of 10.2 months. There was sustained improvement in urine stream, reduction in the mean creatinine concentration and incidence of UTI during follow-up. However, patients with significantly impaired renal function had a poorer outcome. **Conclusion:** Many patients with PUV presented late within the reviewed period. Valve ablation provided relief of obstruction in most of the cases. There is a need to counsel parents/guardians on the need for long-term follow-up.

Key words: Management, outcome, posterior urethral valves, presentation

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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] The exact embryology of the condition is not completely understood, but 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 which has an incidence of 1:5000-1:8000 male births is the leading cause of bladder outlet obstruction and renal insufficiency in male children.[1,3] Its presentation depends on the degree of urinary obstruction which is determined by the severity and orientation of the valves. The clinical features of the disease include poor stream and dribbling of urine after birth. Older children may present with urinary incontinence. The affected patients may also present with complications of the disease such as urinary tract infections, sepsis, impaired renal function, anemia, and failure to thrive.[1,3,4]

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 a micturating cystourethrogram which shows the classical posterior urethral dilatation and bladder trabeculation.[3-5]

The standard management of the condition is endoscopic valve ablation.[6] 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.[7] The study retrospectively evaluates the cases of PUVs seen in our unit over a 10 year period.

PATIENTS AND METHODS

All patients with a diagnosis of PUVs in the urology unit seen over a 10 year period (2003-2012) were retrospectively studied. After approval by the Hospital ethics committee, information regarding patients' biodata, clinical features, radiological and laboratory findings, management, and its outcome were retrieved from the patients' hospital records and analyzed using Microsoft excel. The results were presented in the form of means/standard deviation and percentages.

RESULTS

A total of 44 males was managed for PUVs during the period. Their ages at presentation ranged between 1 day and 14 years with a mean of 3.95 years. Eight (18.2%) patients presented before the age of 1 month, 11 (25.0%) between the ages of 1 month and 1 year, while 25 (56.8%) presented after the age of 1 year. Prenatal diagnosis with the aid of a maternal ultrasound scan was made in 6 (13.6%) patients. Poor stream/dribbling of urine noted in 40 (90.9%) patients was the most common mode of presentation while hematuria recorded in 2 (4.5%) was the least [Table 1]. The most common finding on physical examination was a palpable bladder while the least was generalized edema [Table 2]. Urinary tract infection noted in 23 (52.3%) patients was the most common complication of the disease while pulmonary hypoplasia noted in 1 (2.3%) was the least. Other associated congenital anomalies present were Down syndrome in one patient, bilateral pelviureteric junction obstruction in 1 and inguinoscrotal hernia in 1.

Abdominal ultrasonography revealed dilated posterior urethra in 16 (36.4%) patients [Figure 1], bilateral hydronephrosis in 34 (77.3%), bilateral hydroureter in 7 (16.0%), and thickened bladder wall in 32 (72.7%). Micturating cystourethrogram was done in 35 (79.5%) patients. This revealed dilated posterior urethra in all patients, diverticulum in six, and vesicoureteric reflux in 9 (6 unilateral and 3 bilateral). There were 26 positive urine cultures, with *Escherichia coli* noted in 12 cases being the most common organism isolated. The creatinine concentration at presentation ranged between 0.4 mg/dl and 4.0 mg/dl with a mean of 1.02 ± 0.93 mg/dl. Seven (16.0%) patients had creatinine value ≥1 mg/dl.

Table 1: Presenting symptoms and signs in the patients

Symptoms/signs	Number of patients (n = 44)	Percentage
Symptoms		
Poor stream/dribbling	40	90.9
Abdominal swelling	18	40.9
Fever	15	34.1
Urinary incontinence	7	15.9
Hematuria	2	4.5
Signs		
Palpable bladder	17	38.6
Febrile to touch	12	27.3
Pallor	10	22.7
Ballotable kidneys	10	22.7
Ascites	6	13.6
Generalized edema	2	4.5

Table 2: Complications of posterior urethral valves in the patients

Complications	Number of patients (n = 44)	Percentage
Urinary tract infection	23	52.3
Anemia	13	29.5
Failure to thrive	12	27.3
Septicemia	4	9.1
Renal failure	4	9.1
Hypertension	2	4.5
Pulmonary hypoplasia	1	2.3

Preoperatively, urethral catheterization was done in 31 (70.5%) patients, including 2 (4.5%) who had percutaneous nephrostomy. Six 13.6%) patients had vesicostomy prior to valve ablation [Figure 2]. Four (9.1%) patients died shortly after presentation, two as a result of overwhelming sepsis, one due to end stage renal disease and one due to pulmonary hypoplasia. The parents of three patients requested referrals to other centers after presentation during a brief period of instrument failure in the unit. The other 37 patients had endoscopic valve ablation with the aid of a bugbee electrode. Of these, 35 (94.6%) had type I PUVs while 2 (5.4%) 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 2 weeks and 3 years with a mean of 10.2 months, however, only in nine patients was this period >6 months. One patient died from progressive renal deterioration 2 months after surgery. Six weeks post ablation there was no clinical or microbiological evidence of UTI in 16 out of the 23 patients that presented initially with this

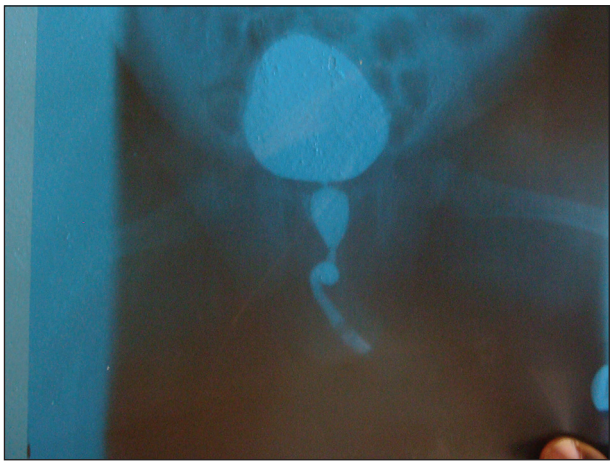

Figure 1: MCUG showing dilated posterior urethra

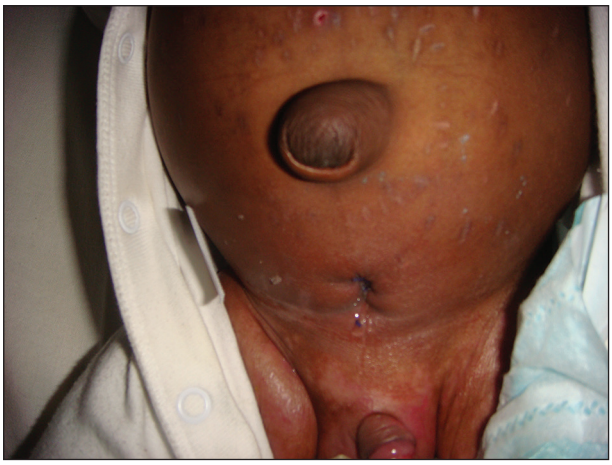

Figure 2: Child with vesicostomy prior to valve ablation

complication. The mean creatinine concentration after 3 months was 0.7 ± 0.46 mg/dl. Ultrasonography showed persistence of hydronephrosis up to 3 years after valve ablation in two cases. Repeat MCUG as well as urodynamic studies to assess disorders in bladder function were not done in the course of follow-up.

DISCUSSION

Posterior urethral valves remain a significant cause of bladder outlet obstruction and renal insufficiency in male children.[3,4] The study aimed at evaluating the clinical presentation, management, and outcome of the disease in our institution.

The mean age of presentation (3.95 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.[4,8] Factors which may have contributed to delayed presentation noted in the region include a sub optimal prenatal ultrasound scan utilization,[9] poverty, ignorance on the part of parents, and paucity of specialist care.

At present, PUV can be suspected *in utero* when prenatal ultrasonography shows the presence of hydronephrosis, dilated proximal urethra, thickened bladder wall, and oligohydramnios[1,3] as was the case in six patients in the study. The sensitivity of a prenatal scan in detecting PUV varies between 40% and 60% with greater sensitivity in the third trimester.[10] The above patients did not have any form of intervention *in utero* because of lack of the required facilities and expertise. In more advanced centers, the use of serial vesicocentesis and vesicoamniotic shunts to relieved pressure in the urinary tract has been practiced.[11] This is aimed at preservation of renal function in severe cases prior to definitive treatment in the postnatal period. However, there is no clinical consensus on the overall long-term benefits of prenatal interventions.[11,12]

Voiding dysfunction was the most common reason for presentation. This is in keeping with findings from many other studies in the literature.[3-5] It is generally accepted that cases with severe obstructive symptoms are more likely to present at an earlier age. Abdominal distension noted was due to the presence of ballotable hydronephrotic kidneys and urinary ascites while the presence of fever was mostly due to complications such as UTI and septicemia. Two neonates in our series presented on account of hematuria. This uncommon symptom which has also been reported in earlier studies may be due to trauma on the hydronephrotic kidney during delivery.[13]

The presence of a palpable thick bladder was noted more in those patients who presented in the neonatal period perhaps due to the presence of severe obstructing PUV. UTI was the most common complication of the condition noted in the study. This is similar to findings from previous studies.[4,8] Factors which increased the risk of UTI included the presence of diverticulum, reflux, and upper tract dilatation. The patient with pulmonary hypoplasia died shortly after birth from respiratory failure. This severe complication of the disease occurs as a result of failure of normal lung development and expansion due to severe oligohydramnios. It requires urgent respiratory support in the form of endotracheal intubation, positive pressure ventilation, and extracorporeal membrane oxygenation which is not readily available in our setting. Even

in advanced centers, the mortality rate from this complication approaches 50%.[14]

The cases were not routinely subjected to screening tests to detect other congenital anomalies due to financial considerations and paucity of screening facilities. However, associated congenital anomalies noted included a case each of Down syndrome, bilateral pelvi-ureteric junction obstruction, and inguinoscrotal hernia. Previous studies have noted congenital defects in the cardiovascular, urogenital, gastrointestinal, and central nervous systems of males with PUV.[15,16]

Postnatal abdominal ultrasonography was helpful in the detection of a dilated proximal urethra in a third of the patients. Micturating cystourethrogram was not done in all cases due to the inability of some parents to afford the cost of the procedure and to poor clinical state of some patients.

Urinary drainage by means of urethral catheterization was done in the majority of the patients to improve renal function and to allow optimization prior to surgery. Two of such patients also had percutaneous nephrostomy on account of severe hydroureteronephrosis. Vesicostomy was done in patients whose urethra could not accommodate the available telescope and in those who presented during a brief period of instrument failure in the unit. This allowed bladder drainage in these patients without the potential risk of long-term indwelling catheterization. The above patients, however, subsequently had valve ablation in the unit. Other forms of urinary diversion like loop ureterostomies reported in the literature[5,17] were not noted in the study.

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. 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, urinary extravasation, urethral bleeding and urethral stricture have been reported to be associated with the procedure.[6] However, these were not recorded in our patients within the period of follow-up. Other means of valve ablation include the use of an endoscopic loop resectoscope, hook diathermy electrode, cold knife urethrotome, balloon catheter, and valvulotome.[6] The patients who died as a result of end-stage renal disease did not have the benefit of renal replacement therapy in the form of renal transplantation as this is not readily available and affordable within the region.

Most of the patients were lost to follow-up. The reason for this is not certain, however, 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. This suggests a need to further enlighten parents and guardians of affected patients in our setting on the benefits of long-term assessment of renal function and other clinical parameters. Urodynamic studies to assess bladder and voiding functions are also important in the follow-up protocol.[7] However, these were not done in our patients due to unavailability of the required facilities and expertise during the period reviewed.

CONCLUSION

Many patients with PUV presented late during the period reviewed. Provision of better and affordable prenatal health services may lead to early age of presentation in our setting. 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. There is also a need to improve on the availability and affordability of renal replacement therapy in our region.

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