



STUDY OF CLINICAL PRESENTATIONS, INVESTIGATIONS, AND MANAGEMENT OF POSTERIOR URETHRAL VALVE CASES AT A TERTIARY HOSPITAL

Paediatric Surgery

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ABSTRACT

Background: Posterior urethral valves (PUVs) are a congenital abnormal obstructing membrane that is located in posterior urethra. It is the commonest cause of bladder outlet obstruction in boys. The goal of this study is to study clinical features, laboratory, imaging parameters and surgical intervention in children with PUV presenting to our center. **Material and Methods:** Present study was single-center, prospective, longitudinal, interventional study, conducted in male children < 12yrs, diagnosed patients of posterior urethral valve. Results: 25 cases of PUV were studied. In the neonatal period 2 (4 %) were confirmedly diagnosed. 13 (52 %) patients were diagnosed in infancy after the neonatal period, Fever was seen in 11 (44%) while voiding symptoms were the presenting symptoms in 21 (84%) patients. In 13 cases (52%) pyuria was detected by urine microscopy. Urine culture was thus done for all cases of which 7 (28%) had significant growth of organisms having the most common being E. coli. USG demonstrated hydronephrosis but no hydroureter in 8 (32%), hydroureters but no hydronephrosis in 2 (8%), hydro-ureteronephrosis in 12 (48%), thick-walled urinary bladder in 10 (40%). Review of the MCU showed that 22 (88%) had gross dilatation of the posterior urethra, 13 (52%) had bladder trabeculations, 8 (32%) had bilateral vesicoureteric reflux, 4 (16%) patients had unilateral vesicoureteric reflux and 2 (4%) patients had significant post-void residue. DMSA scan was done in 9 (36 %) which showed subnormal function in 2 (8%) and 3 (12 %) of right and left renal units respectively. In addition, 2 renal units had scars in 2 different patients. 1 (4%) patient was catheterized. 18 (72%) patients fulgurated at 4 & 7 o'clock position. 5 (20%) underwent ureterostomy (R3, L2). 1 (4%) patient had undergone vesicostomy. Conclusion: Diagnosed patients require primary cystoscopy with primary valve fulguration/ablation of posterior urethral valves.

KEYWORDS

posterior urethral valve, antenatal USG, cystoscopy, bladder outlet obstruction

INTRODUCTION

Posterior urethral valves (PUVs) are a congenital abnormal obstructing membrane that is located in posterior urethra. It is the commonest cause of bladder outlet obstruction in boys.¹ Incidence of PUV ranges from 1 in 8,000 to 1 in 25,000 live male births and it constitutes about 10% of prenatally diagnosed hydronephrosis.^{2,3}

PUVs cause urinary outflow obstruction at the posterior urethra. Back pressure leads to proximal urinary tract dilatation (bilateral or unilateral hydronephrosis, hydroureter) and hypoplasia/dysplasia of developing kidneys. Bladder outlet obstruction may also result in a thickening and hypertrophy. These patients are then susceptible to incontinence, infection, and progressive renal damage manifesting as uremia, metabolic acidosis and hypocalcaemia.^{2,3}

Voiding cystourethrography (VCUG) or micturating cystourethrogram (MCU) is most useful for diagnosis of PUV.^{4,5} It permits assessment of vesico-ureteric reflux; which is present in almost 50% of the patients, along with visualisation of dilated and elongated posterior urethra.⁴ Renal scan (DMSA) is useful in assessing the functioning of kidneys and for detecting scars if any. The goal of this study is to study clinical features, laboratory, imaging parameters and surgical intervention in children with PUV presenting to our center.

MATERIAL AND METHODS

Present study was single-center, prospective, longitudinal, interventional study, conducted in department of urology, at XXX medical college & hospital, XXX, India. Study duration was of 2 years (August 2011 to August 2013.). Study approval was obtained from institutional ethical committee.

INCLUSION CRITERIA

- Male children < 12yrs, diagnosed patients of posterior urethral valve, parents willing to participate in present study

EXCLUSION CRITERIA

- Distal urethral obstruction

- Hypospadias
- Children above 12 years

Study was explained to parents in local language & written consent was taken for participation & study. Detailed history & clinical examination were performed in each case. Under strict aseptic precautions urinary bladder was catheterized by an infant feeding tube (size 5 Fr or 7 Fr) to relieve bladder outlet obstruction. Urine was collected and sent for routine examination microbiology and culture and antibiotic sensitivity test.

Blood sample was collected on admission for estimation of hemoglobin, total and differential leucocyte count, serum potassium, serum creatinine and calcium profile. Blood gas analysis was performed in each case. Ultrasound of abdomen & kidney, mentioning kidney size, hydronephrosis, bladder capacity pre & post catheterization, was performed in all patients. Details of micturating cystourethrogram (MCU) were recorded with respect to followings findings on the MCU: thickened trabeculated bladder, a dilated or elongated posterior urethra, hypertrophied bladder neck, diverticulae or vesicoureteral reflux (VUR). Diagnosis was confirmed by Micturating cystourethrogram (MCU) & cystoscopy. Special radionuclide scans like Tc-dimercaptosuccinic acid (DMSA) were done when appropriate.

Patients were treated with primary cystoscopy with primary valve fulguration/ablation of posterior urethral valves by pediatric resectoscope/Bugby electrode under general anesthesia. Data was collected and compiled using Microsoft Excel, analysed using SPSS 23.0 version. Statistical analysis was done using descriptive statistics.

RESULTS

25 cases of PUV were studied. In the neonatal period 2 (4 %) were confirmedly diagnosed. 13 (52 %) patients were diagnosed in infancy after the neonatal period, 16 (64%) patients after infancy and before the age of 2 years and 7 (28%) patients after the age of 2 years. 18 (72%) of the patients developed symptoms for the first time before their second birthday.

Table 1: Distribution of cases by age at presentation(n=25)

Age group	No (%)
<1 month	2 (8%)
1 -6 months	6 (24%)
6 – 12 months	5 (20%)
1 yr - 2 yr	5 (20%)
2 yr - 5yr	3 (12%)
>5 yr	4 (16%)

Maternal antenatal ultrasound was done in all 25 (100%) cases. Of these 16 (64%) had abnormal USG. 12(75%) out of 16 patients developed H/o of Respiratory distress & NICU admissions after birth.

Table 2- H/o respiratory distress & NICU admission in Abnormal ANC USG (n=16)

H/o respiratory distress & NICU admission after birth	Abnormal ANC USG
Present	12(75%)
Absent	4(25%)

Fever was seen in 11 (44%) while voiding symptoms were the presenting symptoms in 21 (84%) patients. The voiding symptoms were characterized by straining and/or excessive crying/irritability during micturition in 19(76%), poor urine stream noticed by the mothers in15(60%), dribbling of urine in12(48%) and abdominal distension/palpable mass noticed neither of patient (16%). 12 (48%) patients were had posterior urethral valve obstruction after investigation for urinary tract infections.

Table 3: Clinical presentation

Symptoms	No (%)
Straining/crying during micturition	19 (76%)
Poor urinary stream	15 (60%)
UTI in past	12 (48%)
Frequency	12 (48%)
Dribbling	12 (48%)
Fever	11 (44%)
Urinary retention	7 (28%)
Failure to thrive	6 (24%)
Pain in abdomen	4 (16%)
Pulling at penis	2 (8%)

13 (56%) had turbid urine. Hypertension and tender abdomen were observed in 3 (12%) and 4 (16%) of patients respectively. 3 cases (6%) presented with hematuria.

Table 4: Clinical examination & urine investigation.

Signs	No (%)
Turbid urine (pus cell in urine)	13(52%)
Hypertension	3(12%)
Tender abdomen	4(16%)
Gross hematuria	3(12%)
FTT	4(16%)

Urine microscopy was done for all subjects. In 13 cases (52%) pyuria was detected by urine microscopy. Urine culture was thus done for all cases of which 7 (28%) had significant growth of organisms having the most common being E. coli.

Table 5- Urine microscopy & culture

Characteristics	No (%)
Urine Microscopy – Pyuria	13 (52%)
Urine culture	
No Growth	18 (72%)
E. coli	5
Enterococcus	2
Klebsiella	2
Enterobacter	1
Streptococci	1

Serum creatinine levels were elevated in eight (32%) patients at presentation while in 16 (64%) patients, the venous bicarbonate level was <15 mEq/l. 9 (36%) patients were anaemic.

Table 6: Hematological and biochemical Parameters

Parameters	Deranged (%)
S. Creatinine	8(32%)
S. Potassium	6(24%)

Anemia	9(36%)
Metabolic acidosis	16(64%)

Ultrasonography was done for all children. USG demonstrated hydronephrosis but no hydroureter in 8 (32%), hydroureters but no hydronephrosis in 2 (8%), hydro-ureteronephrosis in 12 (48%), thick-walled urinary bladder in10 (40%).

Table 7: Postnatal USG Findings

USG findings	Cases (%)
Hydronephrosis (HN)	8(32%)
Thick-walled bladder	10(40%)
Hydroureter (HU)	2(8%)
Hydroureteronephrosis (HUN)	12(48%)

All children underwent Micturating Cysto Urethrogram (MCU). Review of the MCU showed that 22 (88%) had gross dilatation of the posterior urethra, 13(52%) had bladder trabeculations, 8 (32%) had bilateral vesicoureteric reflux, 4 (16%) patients had unilateral vesicoureteric reflux and 2 (4%) patients had significant post-void residue.

Table 8: Micturating cystourethrogram (MCU)

MCU findings	No (%)
Dilated posterior urethra	22(88%)
Bladder trabeculations	13(52%)
Significant post void residue	2(8%)
Reflux bilateral	8(32%)
Reflux unilateral	4(16%)

DMSA scan was done in 9 (36 %) which showed subnormal function in 2(8%) and 3(12 %) of right and left renal units respectively. In addition, 2 renal units had scars in 2 different patients.

Table 9: Renal Scan Findings 9 (36%)

DMSA scan done	9(36 %)	
Function	Right	Left
Normal	6	7
Subnormal	2	3
Scars Present	1	2

In present study, all patients underwent cystoscopy. 1(4%) patient was catheterized. 18 (72%) patients fulgurated at 4 & 7 o clock position. 5 (20%) underwent ureterostomy (R3, L2). 1 (4%) patient had undergone vesicostomy.

Table 10: Operative procedure.

Intervention	Number (%)
Catheterization	1(4%)
Cystoscopy with primary valve Fulguration	18(72%)
Cystoscopy with primary valve Fulguration with vesicostomy	1(4%)
Cystoscopy with primary valve Fulguration with Ureterostomy	5(20%)

DISCUSSION

Posteriorure thralvalve is the commonest structural cause of urinary out flow obstruction in boys with a broad spectrum of clinical severity and sequelae It is also the most common type of obstructive uropathy leading to childhood renal failure.⁶

PUVs represent a spectrum of severity, ranging from disease incompatible with postnatal life to disease that is minimal with urinary tract infection or symptoms of obstruction and may not manifest until later in life. Most patients present with obstructive symptoms like straining, poor stream, dribbling and with features of urinary tract infection.² Abdominal and pelvic USG is required for assessment of renal parenchyma, evidence of renal collecting system dilatation, bladder wall thickness and postvoid urine.⁴

Patients with the most severe form of the disease often die immediately after birth and do not present for treatment.⁷ In addition, upto 26% of patients treated in the neonatal period subsequently develop end stage renal failure,. There is an intermediate group that does not require renal transplantation but also does not have enough renal function to permit growth and development.⁷ More recent reviews have demonstrated increased presentation during the neonatal period and this early presentation is attributed to greater awareness about the condition, and better antenatal diagnosis by routine prenatal USG.^{7,8} The incidence of PUV in a study by A N Gangopadhyay was 3.5 per 1000 pediatric

surgical OPD cases.⁹

The normal bladder is imaged on maternal transabdominal ultrasound around 16-18 weeks. A normal antenatal scan does not always exclude valves as in majority cases the ultrasound scan below 24 weeks failed to identify upper tract dilatation.¹⁰

In a study by Elder et al.,¹¹ only 10 % of antenatally diagnosed hydronephrosis were PUV while antenatal diagnosis of hydronephrosis was recorded in only 5 cases (5.5%) in a study by Choudhary et al.⁷ 20 % of patients were diagnosed antenatally in a study by Imaji et al.¹² This discrepancy in incidence is attributed partly to timing of antenatal ultrasounds can and partly to terminology used to define hydronephrosis.

Age at presentation was suggested as a predictor of renal function in children with PUV.¹³ In a study by Ziyilan et al, renal function was significantly impaired in patients who presented late (age greater than 5 years) compared to that in patients with early diagnosis (48% vs. 13.7%).¹⁴

Clinical presentations of posterior urethral valves in our study are comparable to some of other studies. In neonates, obstructive symptoms (straining, poor stream, dribbling and palpable bladder) may be seen in 77% of cases and many of them present with features of renal failure.^{15,16} Study by Gangopadhyay et al.,⁸ at had 75% of cases with obstructive symptoms (14) while it was 73% in a study by Kalhor et al.,¹⁷ The later presentations are indicative of milder forms of valvular obstruction and their presentation may be delayed into adulthood.

In the study by Osama et al.,¹⁶ 30 patients (25%) had UTI while Bhaumik et al.,¹⁸ had 51 (65%) patients with UTI, E. coli being the most common organism grown. 91 % patients had elevated serum creatinine levels. In a study by K Bhaumik et al.,¹⁸ In a study by Imaji et al.,¹² serum creatinine value ranged from 0.8mg/dl to 10.6 mg/dl, while in our study it ranged from 0.2 to 5.6 mg/dl.

In the context of posterior urethral valves; USG demonstrates hydroureteronephrosis, thick-walled bladder, dilated posterior urethra only hydronephrosis and only hydroureter. Normal renal echogenicity and corticomedullary differentiation are useful to predict a good outcome while CMD loss and increased echogenicity are relatively insensitive and poorly specific. The finding of at least one kidney with good CMD on diagnostic USG in boys with PUV is associated with a good prognosis.¹³ In the study by Osama et al.,¹⁶ 40% patients had bilateral hydronephrosis on USG. Hydronephrosis was seen in 69 cases (87.3%) and hydroureter in 46 cases (58.2%) in a study by Bhaumik et al.,¹⁸

MCU is the gold standard in diagnosis of a case of posterior urethral valve. It permits assessment of vesicoureteric reflux in almost 50% patients.¹⁹ Of these, 20% will have renal dysplasia.^{4,20} In 15%, there is spontaneous resolution of reflux and in the remaining 15% reflux persists and needs surgical correction.²⁰ The posterior urethral changes in MCU vary from very minimal to severe dilatation.

Study by Osama et al.,¹⁶ revealed unilateral VUR in 36.5% and bilateral in 42.5%. In our series, MCU was done under antibiotic cover. All children underwent MCU. Review of the MCU showed that 22(91.6%) had gross dilatation of the posterior urethra, 13 (52%) had bladder trabeculations, 4 (16%) had unilateral vesicoureteric reflux, 8(32%) patient had bilateral vesicoureteric reflux. 2 (8%) patients had post void residue.

Primary valve ablation has become the mainstay of management of PUV. Availability of neonatal resectoscopes has changed the initial mode of treatment in favour of per urethral fulguration of valves. Primary valve fulguration was done in 90 patients (91.1 %) in a study by Chowdhary et al.⁷

In our study, all patients underwent cystoscopy. Out of which 18 had posterior valve fulguration only. As these 18 patients were presented with creatinine levels within normal ranges but their kidney functions were not yet compromised and MCU was showing dilated posterior urethra with refluxes but their DMSA were showing normally functioning kidney units so we done in our study check cystoscopy and valve fulguration only.

The surgical management changes in children labelled bad prognosis.

These patients who diagnosed HN/HU, dilated posterior urethra, thickened bladder wall and post void residue and nadir creatinine is raised (>1mg) even after catheterization, renal dysplasia may require temporary high urinary diversion procedures like vesicostomy as emergency procedure and ureterostomy, pyelostomy or nephrostomy for patients poorly functioning renal units on DTPA study.^{16,17}

CONCLUSION

Increased antenatal USG screening will be required so as to increase the early detection rate of PUV. Diagnosed patients require primary cystoscopy with primary valve fulguration/ ablation of posterior urethral valves. Acute management with long term follow up is required while dealing with posterior urethral valve.

CONFLICT OF INTEREST: None to declare

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