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Research Article

Posterior urethral valve clinical assessment and outcome

Pranaya Kunal¹,Sushant Kumar²,Priyanka³, Ahsan Ulla³

 ¹Associate Professor, Department of General Surgery, Mata Gujri Memorial Medical College & Lions Seva Kendra Hospital, Kishanganj, Bihar- 855107, India
 ²Professor, Department of General Surgery, Mata Gujri Memorial Medical College & Lions Seva Kendra Hospital, Kishanganj, Bihar- 855107, India
 ³Post Graduate Trainee, Department of General Surgery, Mata Gujri Memorial Medical College & Lions Seva Kendra Hospital, Kishanganj, Bihar- 855107, India
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Abstract

Background: Posterior urethral valves are the most common cause of infra-vesical obstruction in the male child. A posterior urethral valve (PUVs) represents the most common etiology for congenital urethral obstruction and the most common congenital cause for bilateral renal obstruction and somatic growth retardation. It is also the most common cause of obstructive uropathy leading to childhood renal failure. With an estimated incidence of 1/5000 to 1/8000 male births, they account for 10% of all prenatally diagnosed urinary obstructions. The aim & objective of present study is to find out the incidence, age & modes of clinical presentation relevant investigations and different management in cases with diagnosed posterior urethral valve and its short term outcome. **Materials & Methods**: Study was carried out in MGM Medical College & LSK Hospital Kishanganj Bihar India. It is a retrospective study of the patients who were managed from September 2016 to August 2018 and followed up in our department. **Results:** Out of 50 patients, 100% patients presented with poor urinary poor urinary stream followed by dribbling of urine and palpable bladder, 54% presented with recurrent UTI. About 66% patients had presented with fever and 7 patients presented with features of renal failure. We had; done primary cystoscopic valve fulguration in 41 cases (82%) and vesicostomy followed by cystoscopic fulguration in rest of 9 cases (18%). **Conclusion:** Posterior urethral valve is a dynamic disease that can have lifelong effects on bladder. These patients need long term follow up care to monitor and treat the effects of altered bladder compliance.

Keywords: Posterior urethral valve; Bladder dysfunction; Ureteric reflux; Hyperreflexia; Cystourethrography

Introduction

Posterior urethral valves are the most common cause of infravesical obstruction in the male child[1]. Posterior urethral valves (PUVs) represents the most common etiology for congenital urethral obstruction and the most common congenital cause for bilateral renal obstruction. With an estimated incidence of 1/5000 to 1/8000 male births, they account for 10% of all prenatally diagnosed urinary obstructions[2,3]. It is also the most common cause of obstructive uropathy leading to childhood renal failure[4].

Posterior urethral valves are unique to male children and there is no parallel urethral anomalies in girls, similar to posterior urethral valves in boys. The widespread use of pre-natal ultrasound has now provided the opportunity too many patients with valves well before birth[5].It has become clear that bladder outlet obstruction also affects the development and function of urinary bladder. The incidence ranges from 1:8000 (Casale AJ et al 1990)[6] to 1:25000 live male birth (Atwell-JD et al 1983O)[1]. Obstruction of the fetal lower urinary tract is a rare disorder, affecting 2.2 per 10,000 births. PUV, while being the most common cause of lower urinary tract obstruction in males, are a rare disorder, affecting only 1 in 4000-7500 infants, and there is a disproportionately elevated incidence in African-Americans and children with Down's

^{*}Correspondence Dr.Sushant Kumar

Professor, Department of General Surgery, Mata Gujri Memorial Medical College & Lions Seva Kendra Hospital, Kishanganj, Bihar- 855107, India **E Mail**: <u>sushantkumar2002@yahoo.com</u>

syndrome. Despite their rarity, PUV present such a severe insult to the upper urinary tract that they account for almost 17% of children with end-stage renal failure[7,8]. Although most cases are diagnosed at childhood, (the majority are diagnosed before 10 years of age), it may also present at any age. The commonest clinical presentation is urinary symptoms (poor urinary stream followed by dibbling of urine) and severe septicemia, respiratory distress and failure to thrive[4]. Generalized distention of abdomen is more common in younger age group especially in neonate along with urinary ascitis[5]. The incidence of palpable kidney, hypertrophied bladder on bimanual palpation was documented to be higher. Posterior urethral valves may be associated with upto 10% of severe prenatally diagnosed hydronephrosis[9].Prenatal ultrasound screening of PUV has significantly increased early diagnosis and management of this pathology in most developed societies. In fact, PUV are now commonly diagnosed by the postnatal evaluation of infants who had prenatal hydronephrosis[10].Langenbeck in 1802 is credited with the first description of such valves. Dr. H. Hampton young is generally given credit for first clear description and classification of posterior urethral valves (Young et al 1919). He recognised three distinct varieties of congenital proximal urethral obstruction and classified these as

Type- I, II, III (Young HH, Frontz WA et al 1919)[11]. Classification of posterior urethral valves[12]

Type I: Posterior mucosal folds distal to the verumontanum.

Type II: Mucosal folds that diverge from verumontanum towards the bladder neck.

Type III: Valves of the diaphragmatic or the annular type.

Stephens (1983) suggested the existence of an additional type of proximal urethral obstruction which he termed as Type-IV valves (Stephens-FD et al 1983)[13]. The most typical PUV is presented in neonates with history of prenatal bilateral hydronephrosis or in infants with acute pyelonephritis associated with massive VUR. This group of PUV is easy to be diagnosed by typical findings in voiding cystourethrography (VCUG) and endoscopy for confirming valve like structure in the posterior urethra. In contrast, there is another group of PUV which is elusive and overlooked easily in VCUG even if it contributes to VUR or detrusor overactivity. Until recently there has been no reference standard based on findings in VCUG and endoscopy[14,15].

Initial continuous catheter drainage and eventual management by valve resection, avulsion or ablation could ameliorate the obstructive uropathy. However, it has been noted that even after surgical treatment of valves; about 70% of older children and adolescent boys continue to have persistent bladder dysfunction with long term morbidity and ESRD[10,16,17]. In a resource poor economy like ours, public enlightenment, early diagnosis, and prompt institution of management would go a long way in improving initial and long-term outcome of PUV[17].At present, primary endoscopic valve ablation is the standard method to treat the vast majority of posterior urethral valves in children[5].

The bladder dynamics change with growth of the baby and hypertonicity decreases with time following valve ablation while hyperreflexia persist leading to bladder dysfunction and urinary incontinence. Patients were kept in strict follow up to prevent long term complication. The aim & objective of present study is to find out the incidence, age & modes of clinical presentation relevant investigations and different management in cases with diagnosed posterior urethral valve and its short term outcome.

Methods & patients

About 50 cases with PUV were enrolled in the study period mentioned above. Paediatric patients of < 12 years of age attending in paediatric surgery OPD or admitted in the indoor of Mata Gujri Memorial Medical College and L.S.K. Hospital Kishanganj, Bihar with symptoms suggestive of obstructive uropathy were selected for the study. A detailed history, per general examination, abdomen examination, examination of external genitalia, urine, biochemical examination. hematological, and ultrasonography, voiding cystourethrogram, intra venous urography and renal scan was done.

Treatment protocol at our centre is: initial treatment is intravenous fluid, urinary catheter, correction of acid base balance and electrolyte, detailed urinary analysis and intravenous antibiotics. MCU was being done once C/S was reported as sterile or after infection was controlled.

Operative Management: The patient was then scheduled for definitive procedure by diagnostic cystoscopy and cystoscopic valve fulguration. We follow up the cases of PUV initially at monthly internal for first 3 months with routine examination & C/S report and blood urea and serum creatinine report along with ultrasonography of abdomen and KUB. The way of assessment of prognosis was re-examination for pus cells per high power field (HPF) and blood urea, serum creatinine estimation.

In our follow-up clinic we advise the following investigations.

1. Blood Urea and serum creatinine

Routine urine examination and C/S if necessary, every month for 1st 3 months and after 3 months if C/S report sterile and then after 6 months and then yearly
 MCU was advised after 6 months to assess VUR
 USG- KUB was advised after 6 months for evaluation of the size of kidneys, ureters and bladder for VUR and post void residual urine volume

Results

DTPA renal scan was advised after 6 months in cases where there was persistent high serum creatinine level, even after urinary diversion or in cases where USG reveals definitive evidence of cystic dysplasia.
 Urodynamic study was done where serum creatinine

level remains above 2 mg% but below 5 mg% or in patients having residual symptoms after adequate primary valve ablation.

A total 50 cases of PUV were admitted to the Department of Surgery, M.G.M. Medical College & L.S.K. Hospital, Kishanganj, Bihar, from September 2016 to August 2018. Out of this 32 patients were admitted in the first two years and the rest of the 18 cases were admitted at rest of the study period. This accounts for 4.1 per 1000 surgical OPD cases of our department.

Group	Age at presentation	No. of cases	Percentage (%)
Ι	< 1 month	7	14
II	1 month – 2 years	11	22
III	2years – 5 years	22	44
IV	5 years – 12 years	10	20

Table 1: Showing Age at Presentation (n = 50)

In our study 14% patients presented within 1 month, 22% patients presented within 1 month -2 year, 44% patients were in the age group of 2-5 years, and 20% were between 5 and 12 years of age. The youngest child presented at the age of 24 days and the oldest one at the age of 12 years. One of our patients was diagnosed antenatally, incidentally on routine USG screening [Table 1].

 Table 2: Showing Symptoms at presentation (n = 50)

Urinary Symptoms		%	Non-Urinary Symptoms	No.	%
Poor urinary stream followed by dribbling	50	100	Abdominal distension	21	42
a) since birth	39	78	Abdominal pain	17	34
b) for the last 3 months to 1 yr.	6	12	Vomiting	19	38
Palpable bladder	50	100	Pallor	25	50
Bilateral palpable kidneys	6	12	Edema	2	4
Unilateral palpable kidneys	3	6	Failure to thrive	15	30
Recurrent UTI	27	54	Fever	33	66
			Respiratory distress	5	10
			Irritability	13	26
			Constipation	3	6

In our series the common presenting features in neonates and infants were poor urinary stream followed by dribbling of urine, retention of urine, palpable bladder and irritability. About 100% patients presented with poor urinary poor urinary stream followed by dribbling of urine and palpable bladder, 12% patients presented with bilateral palpable kidneys, 6% patients presented with unilateral palpable kidneys, 54% presented with Recurrent UTI. About 42% patients presented with abdominal distension, 34% with abdominal pain, 38% patients presented with nausea or vomiting, 50% patients presented with pallor, 66% patients presented with fever, 26% patients presented with irritability, 10% patients presented with respiratory distress, 4% patients presented with edema and 6% presented with constipation. Seven patients presented with features of renal failure (urea ranged from 60 mg/dl to 250 mg/dl and serum creatinine ranged from 2.3 mg/dl to 7 mg/dl) and one of them presented with features of renal rickets [Table 2].

Numbers	Percentage (%)
1	
1	
1	
1	18
1	
1	
1	
1	
1	
	Numbers 1

Table 3: Showing incidence of associated anomalies (n = 50)

Patulous anus was detected in 1 patient as mother complaint of constipation of her baby and per rectal examination revealed this finding. Anorectal manometry done showed normal study. Balanitis xerotica obliterans was treated by circumcision. Repair of cleft lip and palate was done. Herniotomy was done in patient with congenital hydrocele and he is waiting for repair of umbilical hernia. Release and lengthening of tendon was done in patient with CTEV. Urachal fistula was treated by excision. The boy with Down syndrome is under our supervision.

Age	No.	Blood Urea (mg %)						Seru	m Creat	inine (n	ıg %)		
Gr.		>40	-100	>100)-200	>2	200	1	-2	>2	2-3	>	.3
		No.	%	No.	%	No.	%	No.	%	No.	%	No.	%
Ι	7	2	28.5	3	43	2	28.5	2	28.6	2	28.6	3	41.2
II	11	2	18.2	5	45.5	4	36.3	5	45.4	3	27.27	3	27.27
III	22	13	59	4	18.1	5	22.9	16	72.7	4	18.2	2	9.1
IV	10	6	60	2	20	2	20	6	60	2	20	2	20

At presentation blood urea and serum creatinine were raised in all patients. Creatinine level between 1-2 mg/dl was in 29 patients (58%), between >2-3 mg/dl in 11 patients (22%) and >3 mg/d in 10 patients (20%). The following are the complications in our series following primary cystoscopic fulguration or cystoscopic fulguration followed by closure of vesicostomy [Table 4].

fable 5: Showin	g complications	following	valve al	blation ((n = 50)))
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Complication		Age	Total $(n = 50)$	%		
	Group I	Group II	Group III	Group IV	No.	%
Retention of urine	-	-	1	-	1	2
Poor stream and	2	1	-	-	3	6
straining						
Persistent septicemia	-	1	-	1	2	4
Urethral stricture	-	-	1	-	1	2

Table 6: Showing mode of treatment (n = 50)

	Total no.	Primary valve	Urinary diversion (vesicostomy) followed
		ablation	by valve ablation
< 1 month	7	7	0
1 month to 2 years	11	7	4
2 years to 5 years	22	19	3
5 years to 12 years	10	8	2

	1-2 mg/dl	2-3 mg/dl	> 3 mg/dl	Total					
At presentation	29	11	10	50					
1 month	39	6	5	50					
1 Year	41	2	0	43					
2 Years	39	1	0	40					

Table 7: showing serum creatinine level at presentation and on subsequent follow-up [n=50]

On routine examination of urine plenty of pus cells and positive urine culture were shown in 27 patients (54%). Most common organism was E.coli. MCU was performed in all cases and all of them showed dilated and elongated posterior urethra. Irregular margin and trabeculations of the bladder was seen in 30 patients (60%). Multiple sacculations and diverticulae were seen in 25 patients (50%). Vesicoureteric reflux was noted in 19 patients (38%) at presentation of which bilateral reflux were present in 11 patients (22%). Among them 8 cases of unilateral reflux, right side was affected in 3 cases and left side in 5 cases. 5 cases showed severe reflux (grade IV and V) [Table 5]. USG revealed dilated posterior urethra and distended thick walled bladder in all cases. Hydronephrosis was seen in 39 cases (78%) and hydroureter in 23 cases (46%). Corticomedullary junction could be identified in 15 cases. Out of the 25 patients who had non visualization of corticomedullary junction, 23 had impaired renal function.At presentation, 41 patients were treated by bladder catheterization and intravenous antibiotics. After proper resuscitation with I.V. fluid, correction of electrolyte imbalance primary valve fulguration was done in 41 cases. We had performed vesicostomy in 9 cases. So far in this series, vesicostomy closure has been done in 7 cases and 2 patients are waiting with open stoma. Complications of vesicostomy were wound infection, stomal stenosis and mild prolapse. One patient presented with acute renal failure and was treated with haemodialysis; the patient improved on dialysis and is doing well till date. No patient was died in our series. Two patients of our series were suffering from persistent septicaemia after valve ablation. They were treated with high dose antibiotics. At present they are doing well. Another patient of our series developed urethral stricture after cystoscopic valve fulguration. At present he is doing well after cystoscopic dilatation of urethra. We discharge the patient with prophylactic antibiotics (usually Co-trimoxazole) for 3 months with advice to visit surgery OPD monthly for 3 months with RE and C/S report and blood urea and creatinine report [Table 6]. If culture C/S report of urine showed no growth for 3 consecutive months, then we stopped the antibiotics with advice of second follow up after 3 months with MCU, USG- Abdomen and KUB and third follow up after another 6 months with blood urea,

serum creatinine, RE and C/S. We used to prescribe either oxybutynine or Imipramine for 3 months in all patients at the time of discharge. In the follow-up at one month serum creatinine level came down in 39 patients out of 50 patients. Creatinine level at presentation was between 1-2 mg/dl in 29 patients between > 2-3 mg/dl in 11 patients and > 3 mg/dl in 10 patients. At one year follow-up, out of 43 patients, only 2 patients had serum creatinine level > 2 mg/dl. At the second year followup, out of 40 patients, 1 patient had creatinine level > 2mg/dl [Table 7]. Follow-up MCU in 1 case who had Grade-I reflux showed resolution within 2 years and grade of reflux improved in 7 other cases. DTPA renal scan was performed in 37 cases all of which showed bilateral hydronephrosis mild to moderate degree. Perfusion was low and there was gross delayed excretory pattern. GFR reduced in affected kidneys. As a routine follow-up first DTPA scan was performed at 6 months after cystoscopic valve fulgaration. Three patients showed improvement of excretory pattern in first follow-up DTPA renal scan. Urodynamic study was done in 5 cases where serum creatinine level remained above 2 mg/dl but below 5 mg/dl or in patients having residual symptoms after adequate primary cystoscopic valve fulguration.

Discussion

The reported incidence of PUV is 1 in 8000⁶ to 1 in 25000^1 live births. In our institution with about 450 paediatric surgical admission per year we admitted cases of PUV approximately 27 annually. Approximately one third of PUV patients presented during the neonatal period and age between 5 years and 12 years, one third between 1 month and 2 years and one third between 2 years and 5 years of age. [18, 19, 20] 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[1]. PUV is increasingly recognized by routine pre-natal USG. [1, 21, 22] The characteristic findings are bilateral hydronephrosis and a distended thickened bladder. A dilated posterior urethra is seen occasionally. Low amniotic fluid and bright renal parenchyma [suggestive of dysplasia] provide important clues about the severity of renal damage. In our series, on routine antenatal USG, one fetus was suspected of having PUV due to

presence of bilateral hydronephrosis and oligohydramnios and postnatally we confirmed the diagnosis by MCU and USG. The baby was managed by primary cystoscopic fulguration and he is doing well on subsequent follow up.

Whether prenatal diagnosis has any impact on long term follow up is still controversial[18]. Failure of prenatal diagnosis to improve outcomes has been observed in multiple studies over the years. [23-25] Even in the developed world, fetal intervention is practiced in a few selective centres only. The beneficial effect of fetal intervention in PUV is, as yet, unproven. However, antenatal diagnosis does help to plan the delivery of the baby in a suitably equipped institute where early confirmation of the diagnosis and treatment may prevent future complications. In our series 14% patients presented within 1 month, 22% patients presented within 1 month to 2 years, 44% patients were in the age group of 2 years to 5 years and 20% patients were between 5 years to 12 years. The majority of the patients presented between 2 years to 5 years of age and this report are not consistent with other reports[18,26] published in literature. In neonates

with features of renal rickets. Failure to thrive observed in 30% of patients. The PUV cases may be associated with several types of anomalies. In our series, 9 out of 50 patients were associated with various types of anomalies.² Patients out of 9 patients has multiple anomalies (listed in Table 3).

The incidence of anomalies encountered in our series is 18%. The anomalies were treated accordingly after cystoscopic valve ablation (discussed in results and analysis chapter). USG demonstrates hydronephrosis, thick walled bladder, dilated posterior urethra and occasionally bright kidneys with loss of cortico-medullary differentiation suggestive of renal dysplasia[28]. In our series cortico-medullary junction was demarcated in 15 cases (30%) cases while the demarcation was lost in 25 cases (50%). MCU/VCUG is the "gold standard" in diagnosis of case of posterior urethral valves. It permits assessment of vesico-ureteric reflux in almost 50% patients[29]. Of these, 20% will have renal dysplasia[18]. In 15%, there is spontaneous resolution of reflux and in the remaining 15% reflux persists and needs surgical correction[30].

In our series 19 patients (38%) presented with reflux and during a short follow up. (Approximately 2 years) 1 patient with grade-I reflux showed resolution and grade of reflux improved in 7 other cases. Unsatisfactory previous scan experience, long distance to service providers, and long waiting periods have been identified as major obstacles to prenatal diagnosis[31].There was a time when many centres performed urinary diversions due to non availability of obstructive symptoms (straining, poor stream, dribbling of urine) may be seen in 100% cases and many of them present with features of renal failure[1, 20]

Infants present with features of infection (78%) like septicaemic shock, poor feeding, abdominal pain and positive urine culture[20]. They also present with abdominal distension, dribbling of urine (100%), failure to thrive (30%) and palpable kidney (18%)[21]. In toddler, renal failure becomes less common but voiding symptoms and infection predominates. 100% patients over 5 years of age present with dribbling[1]. These later presentation is indicative of milder forms of valvular obstruction and their presentation may be delayed upto adulthood[27]. In our series obstructive symptoms in the form of poor urinary stream and dribbling were present in all cases.

In 5 patients who were of older age group, the symptoms were present for a short period, 18% patients presented with a palpable kidney and 54% patients presented with recurrent UTI. Renal failure was more common in those who presented at an earlier age.

In our series 7 patients presented with renal failure and one of them presented small-sized resectoscope. But as these are available now, trans-urethral fulguration of valve is the treatment of choice in PUV patients, even in neonates. The long term policy of diversion has been questioned by some[32]. The role of different types of vesical and supra-vesical diversion is controversial[5].

The incidence of urinary diversion in the primary management of posterior urethral valves is very high, about 50% in the majority of Indian centres. The difference in the management of PUV between the developing world and the developed world is the rate of diversion. Inspite of high urinary diversion rate and subsequent surgery, the mortality has declined in our country, which signified that diversion has a role to play in decompressing the upper urinary tract and to control the infection. Inspite of few complications, vesicostomy is preferred for short term diversion though supra-vesical drainage is restricted to patients with poor renal function and infection due to dilated tortuous ureter or vesico-ureteric reflux as evident in the majority of Indian articles. A similar experience has also been reported from Hospital for Sick children, Toronto[5].We have found that vesicostomy is very methods of diversion, as functioning useful vesicostomy provide an adequate drainage of the lower urinary tract as proposed by Duckett[33,34]. It is a minimally invasive surgery and is an easily reversible technique. It allows cycling of the bladder to take place at low pressures[35].Ureterostomy on the other hand is more difficult to perform but has the advantage of better decompression of the kidney[38].Bilateral

ureterostomy was advocated by some authors as a better alternative method of diversion in developing countries, for patients with persistently high serum creatinine level.[36] Creatinine after operative intervention for PUVs is a well-known predictor of long-term renal function[37,38]. We have done primary cystoscopic valve fulguration in 41 cases (82%) and vesicostomy followed by cystoscopic fulguration in rest of 9 cases (18%). However in our series we did not performed ureterostomy. Reoperation rates have been widely studied for many urological interventions, including transurethral incision of ureteroceles147-148 and proximal hypospadias repair[39,40].

This is an ongoing study. Clinical parameters and investigations of 50 patients of our series, over a period of 2 years are being presented for a disease like PUV which has varied manifestation. Long term follow-up is necessary to see the effect of treatment in these patients. Aggressive management of vesicoureteric reflux with early surgical intervention is rarely justified as upper urinary tract complications resolve spontaneously in some cases. In our short term followup we have observed resolution of reflux in 1 case with improved reflux in other cases.

Conclusion

The diagnosis of posterior urethral valve is well suspected by USG both antenatally and postnatally. The diagnosis of posterior urethral valves as well VUR should always be confirmed by MCU. The management of posterior urethral valves at the first outlet should be urethral catheterization and stabilization of the general condition followed by primary valve ablation by any means. For those who are not progressing satisfactorily, a second attempt of valve ablation may be of great help. The cystoscopic valve fulguration at 5, 7 and 12 O'clock position is not desirable at same sitting. Diversion may be done before fulguration as a temporary measure for upper urinary tract decompression with good result. The DTPA scan is most important tool in assessing the result of management during follow-up. But in our opinion it cannot replace the simple way of assessing the renal functional status by serial estimation of serum creatinine and regular microscopic urinary examination for per cells. Though loop cutaneous ureterostomy is a better option than cutaneous vesicostomy besides that, in our opinion cutaneous vesicostomy is a suitable alternative to loop cutaneous ureterostomy because -

a) Technically easier to performb) Eliminates reflux

c) Preserves the integrity of ureters and maintain bladder cycles.

Serum creatinine value at presentation is not predictive of subsequent renal functional status, but creatinine value after a brief period of urinary decompression is prognostically more useful. Dedicated regular follow up and proper assessment after diagnosing and fulgurating the valve, with sympathetic attitude to these children and their families is very much essential for better outcome to the patients with posterior urethral valves.

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