Presentation, Management and Out Come of Children with Posterior Uretheral Valve

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The purpose of this study was to analyze the different presentations of the children with posterior urethral valve, their management and outcome. This prospective study was carried out for the period of six years starting from April 1994. A total of 51 children presented with the diagnosis of posterior urethral valve during the period of six years with the mean age of 43 months. Bladder dysfunction was the major primary presenting symptom in 043% children (n=22) followed by renal failure in 25% patients (n=13) and pain in suprapubic region with distended bladder in 13.7 % children (n=7). Of the 22 patients of bladder dysfunction 09 patients came out to be in renal failure after investigation with the total incidence of renal failure of 43%. MCU revealed posterior urethral valve in all patients, bilateral reflux in 08 patients and unilateral reflux in 23 cases. Endoscopic ablation of the posterior urethral valve was performed in all patients. Reimplantation of the ureter was done in cases with grade 4 or 5 reflux. Posterior urethral valves (PUV) are overwhelmingly the most common, specific diagnosis affecting boys. These children are at high risk of renal failure. This can all be avoided by early diagnosis and management as well as consideration of the disease at all levels so that early referral to the tertiary care centers can be made possible.
Key words: posterior urethral valve, micturating cystourethragram, renal failure, reflux nephropathy, bladder dysfunction

Posterior urethral valve (PUV) is a congenital urethral anomaly affecting boys[1]. It is the most common obstructive uropathy in boys. Usually the condition is sporadic but genetic factors also play role in its pathogenesis[1,2]. Posterior urethral valve can be detected antenatally through routine fetal ultrasonography[2,3]. Postnatally children presents with bladder dysfunction or with renal failure[4,5]. Risk factors that affects the prognosis are age at the time of presentation, age at the time of surgery, time between presentation and surgery and associated abnormalities like vesicoureteric reflux[6]. Children with posterior urethral valve (PUV) are at high risk for renal failure and the outcome depends upon the early diagnosis and treatment[7]. According to Bajpai[8] only 33% children with posterior urethral valves (PUV) recovered from renal failure when the diagnosis was delayed. Workup in children with PUV is always difficult; it must include renal biochemistry, ultrasonography, Micturating Cystourethrogram (MCU) and Cystosuretherocopy[9]. Management of the child with PUV is entirely dependent upon the degree of renal insufficiency as well as the age of the child[10]. In the presence of normal or near normal functions Endoscopic destruction of the valve is usually considered[11], with the advent of modern technology in medical sciences new techniques like Nd: YAG laser ablation of the posterior urethral valve is becoming a popular procedure[12]. After the treatment many boys have sustained improvement of hydroureteronephrosis and renal function Choice of primary procedure varies but early management has good results with whatever procedure is adopted[13].

Materials and methods:

This prospective study was carried out from April 1994 to March 2000 at the Department of Urology and Renal Transplantation King Edward Medical College / Mayo Hospital Lahore. All the patients with the diagnosis of posterior urethral valves were included in this study. All the patients were admitted. A thorough history and clinical examination was carried out. The symptoms were recorded. Pre-operative workup included renal biochemistry for the assessment of renal functions, ultrasonography was performed to assess the degree of hydronephrosis, cortical echogenicity and cortico medullary differentiation and micturating cystourethrogram (MCU) was performed for the assessment of degree of reflux. After making diagnosis, all the patients were planned for surgery. In sick infants initial drainage per urethra or by percutaneous suprapubic cystostomy or by percutaneous nephrostomy was performed temporarily. After stabilization Endoscopic ablation of the posterior urethral valve was performed in all cases. In patients with grade 1, 2 or 3 reflux no surgical intervention was made regarding ureters but in patients with grade 4 or 5 reflux Reimplantation of the ureters were performed. In patients with bilateral reflux Reimplantation of the ureters was done by Cohns procedure whereas in patients with unilateral reflux reimplantation was done by modified Leitch Greoger technique.

Regular follow-up was carried out in out patient clinics, initially monthly for three months than every three months in next year and than once yearly. Their renal biochemistry was performed to access the degree of

improvement in renal functions. Two months after the surgery MCU was repeated to access the degree of improvement in reflux. Complete recovery was defined as complete freedom from the presenting complaint, normal renal biochemistry and normal or grade 1 reflux. Mortality was defined as the mortality during postoperative hospital stay or as a result of consequences of the primary disease, with in first six months after operation. Patients who died before any surgical intervention were excluded from the study.

Results

A total of 51 children presented with the diagnosis of posterior urethral valve during the period of six years from April 1994 to March 2000 (Table 1). Minimum age at presentation was 08 months and maximum age of 9.5 years with the mean age of 43 months (3.5 years) (Table 2).

Table 1. Year wise distribution

Year	n=	%age
1994	10	19.6
1995	07	13.7
1996	06	11.7
1997	07	13.7
1998	08	15.6
1999	05	9.8
2000	08	15.6
Total	51	100

Table 2. Age distribution

Age at presentation	n=	%age
< 12 months	08	15.6
1 – 2 years	17	33.3
3 – 4 years	11	21.5
5– 6 years	09	17.6
> 6 years	06	11.7
Total	51	100

Bladder dysfunction was the major primary presenting symptom in 43% children. Second common presentation was symptoms and signs of renal failure in 25% patients (n=13); pain in suprapubic region with distended bladder was seen in 13.7% children (n=7). In patients with primary presentation of bladder dysfunction renal failure was diagnosed in 09 patients on investigations so that the total incidence of patients with renal failure came out to be 43%. Palpable mass in the flank, failure to thrive and urinary ascites were the other presentations (Table 3).

Table 3. Primary presentations

Presentation	n=	%age
Bladder dysfunction	22	43
Renal failure	13	25
Suprapubic pain with distended bladder	07	13.7
Flank mass	06	11.7
Failure tom thrive	03	5.88
Total	51	100

Bladder was palpable in 41% patients (n=21) and bilateral kidneys were palpable in 37% patients (n=19). MCU revealed posterior urethral valve in all patients and also showed bilateral reflux in 08 patients (15.5%) and unilateral reflux in 23 cases of that 15(29%) on right side and 8(15.5%) on left side (Table 4).

Table 5. Vesicoureteric reflux

Reflux	Bilateral	Unilateral RT	Unilateral LT	Total
Grade 1	03	07	03	13
Grade 2	02	03	01	05
Grade 3	00	01	01	02
Grade 4	02	02	02	06
Grade 5	01	02	01	04
Total	08	15	08	31
Percentage	15.5	29	15.5	60

Temporary procedure of bladder drainage was performed in 13(25%) patients that included peruretheral drainage in 2(4%) patients, suprapubic cystostomy in 2(4%) patients, unilateral percutaneous nephrostomies in 4(8%) patients and bilateral percutaneous nephrostomies in 5(9%) patients (Table 5). Endoscopic ablation of the posterior urethral valve was performed in all the patients. Bilateral reimplantation of the ureter was performed by Cohns procedure in 03 of the 08 cases of grade 4 and 5 reflux and 07 unilateral reimplantation of the ureters was done by modified Leitch Greoger method for unilateral grade 4 or 5 reflux (Table 4).

Table 5. Temporary procedures.

Procedure	n=	%age
Drainage per Urethra	02	3.9
Suprapubic cystostomy	02	3.9
Per cutaneous nephrostomy	09	17.6
Total	13	24.5

Table 6. Definitive procedures.

Procedures	n=	%age
Endoscopic fulguration of PUV	51	100
Bilat: Reimplantation by Cohns procedure	03	5.8
Unilat: Reimplantation by modified Leitch Greoger method	07	13.7

Follow-up revealed improvement in the renal functions in almost all the patients (Table 7). Improvement in the degree of reflux was recorded in both operated as well as in non-operated cases (Table 8).

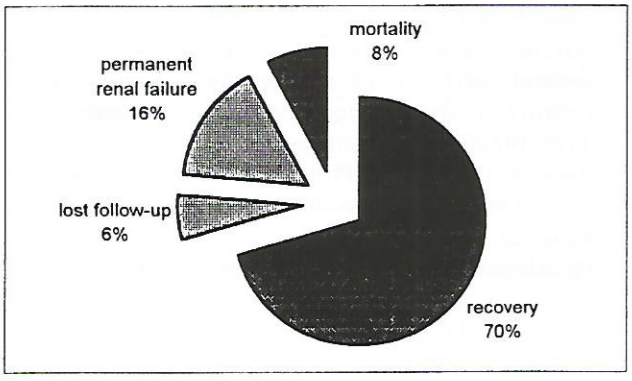
Table 7. Average assessment of renal functions

Functions	Pre op	Post op 1 Month F/U	Post op 12 month F/U	Post op 36 month F/U
Urea	240mg%	100mg%	60mg%	40mg%
Creatinine	6.0mg%	4mg%	4mg%	1.0mg%

Table 8. Assessment of reflux.

Initial Reflux (No of Pts)	After one month F/U	After 12 months F/U	After 36 months F/U
Grade 5 (4)	Grade 5 (2)	Grade 2 (4)	Grade 1 (3)
	Grade4 (2)		Lost F/U 1
Grade 4 (6)	Grade 3 (5)	Grade 2 (3)	Grade 1 (2)
	Grade 4 (1)	Grade 3 (2)	Grade 2 (1)
		Grade 4 (1)	Lost F/U 3
Grade 3 (2)	Grade 2 (2)	Grade 2 (2)	Grade 2 (2)
Grade 2 (6)	Grade 1(4)	Grade 1 (4)	Grade 1 (3)
	Grade 2(2)	Lost F/U 1	Lost F/U 1
Grade 1 (13)	Grade 1 (6)	Normal (11)	Normal (6)
	Normal (7)	Lost F/U 2	Lost F/U 2

Outcome was recorded in terms of complete recovery within first year, complete recovery within three years and mortality. Within this follow-up a total of 10 patients lost their follow-up. Out of these 3 patients lost follow-up after one year and 7 patients after three years. As the mean follow-up time we recorded was three years therefore those 7 patients who lost their follow-up after three years where not considered as follow-up failure. 16.4% patients (n=8)went into permanent renal failure and 7.8% patients (n=4) died due to primary disease (Figure I).



Discussion

Posterior urethral valves (PUV) are overwhelmingly the most common, specific diagnosis, and are confined to boys. PUV can be detected antenatally on routine ultrasonography or Postnatally and it is often associated with renal failure and or bladder dysfunction[5,6,20]. This present study describes our experience with the management of 51 children with the diagnosis of posterior urethral valve over the period of six years. In our present study the minimum age at presentation was 08 months with the mean age of 43 months of which half of the children (n=28) presented during their second to fifth years of life. This patron of late diagnosis was seen in local regional studies[14] as well as studies from underdeveloped countries but in developed countries the age at presentation is as early as mean age of 26.4 days (range 10 days to 61 days)[15]. In this series we find out that most of the children presents due to symptoms of bladder dysfunction (n=22), these symptoms include voiding dysfunction, nocturia and recurrent urinary tract infections. Renal failure was the other common presentation (n=13). This patron of distribution of presentations is again corelating with that of local studies but in developed countries most of the diagnosis is made antenatally[3]. Postnatal presentations varies with age from palpable masses, urinary ascites, and respiratory distress due to severe pulmonary hypoplasic in newborns and infants to symptoms of urinary dysfunctions in toddlers age group[16]. This patron of newborn and infant presentations was not seen as frequently in our local studies probably due to lack of facilities as well as expertise for early diagnosis. Primary diagnosis of renal failure was made in 13(25%) patients, of the 22 patients with bladder dysfunction 09 came out to be in renal failure on investigation so that the total patients with renal failure became 22(43%). The patron of renal failure was commonly seen in western countries in a study of 52 patients by Stephen[17,18] it was found out that 57% of the patients presented with bladder dysfunction and 34% patients with symptoms and signs of renal failure. We did endoscopes fulguration of the posterior urethral valve in all the patients. Though different methods are practiced worldwide. In a study by Kiwi A at Queen Mary Hospital, Hong Kong he successfully performed ablation of the posterior urethral valves by Forgarty balloon catheter[19]. The Nd: YAG laser is the new invention in the field of urology for laser ablation of the posterior urethral valves[12]. Reimplantation of the ureter was only performed when there was grade 4 or 5 reflux. In patients with grade 1, 2 or 3 wait and see policy is the best, which we also adopted with successful results. Postoperative follow-up of the patients is always a problem in third would countries; many factors are responsible for that starting from patients and its attendants ignorance to the facilities provided by the health providing agencies. Our series showed that during the first month following discharge there was 96% follow-up rate with only 4% patients who lost their follow-up. In the first year the number of patients who lost follow-up became 7(14%) with 4(8%) patients lost their follow-up in third year. Comparative to our results in a study of Stephen[18] he did successful follow-up of 98% of his 52 patients for 11.1 years. In follow-up it was seen that improvement in renal functions occurred in 27 patients with in 12 months time with improvement of reflux. This patron of improvement was seen in well-developed countries that suggests that we are as much caring about our patients as any physician of the modern world could be.

Conclusion

A posterior urethral valve is a disease effecting boys. This disease can be diagnosed antenatally with good and expert ultrasonography. Postnatally risk factors include age at presentation, age at surgery, time between presentation and surgery, urodynamic abnormalities and degree of vesicoureteric reflux. These children are at high risk of

renal failure and other urological complications. This can all be avoided by early diagnosis and management as well as consideration of the disease at all levels so that early referral to the tertiary care centers can be made possible.

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