A STUDY OF POSTERIOR URETHRAL VALVES

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ABSTRACT

Objective To determine the clinical manifestations of posterior urethral valves (PUV) and outcome of early diagnosis and management.

Study design *Descriptive study.*

Place & Duration of study Department of Paedaitric Surgery, Paediatric Medicine and Urology at Chandka Medical College Hospital Larkana, from January 2002 to December 2006.

Patients and Methods All patients of paeditric age group having urinary complaints with suspicion of PUV were included in this study. Diagnosis was based on ultrasonography of urinary tract and voiding cystourethrography (VCUG).

Results Fifty one male children with PUV on VCUG were enrolled into the study. Two patients were suspected on antenatal ultrasound of having bladder outlet obstruction. Most of the patients presented after 3 years (40%) and 15% presented at 9 years of age. Common clinical presentations were fever, anaemia, failure to thrive and urinary symptoms. Around 53% had abnormal renal function, 40% had vesico ureteric reflux (VUR). Nearly all patients presenting after 6 years of age had abnormal renal functions and VUR. Early surgical intervention had better resolution effect.

Conclusions PUV is a common cause of the lower urinary tract obstruction in male children. Delay in diagnosis and treatment leads to end stage renal disease.

Key words Posterior urethral valves, Management, Renal failure.

INTRODUCTION:

Posterior urethral valves are congenital mucosal folds on either side of prostatic urethra.[1] It was first described by Hugh Hamptos Young about 90 years ago. Incidence of PUV in Pakistan is not known, whereas in western literature it ranges between 1:5000 to 1:8000 male infants.[2] It is common cause of lower urinary tract obstruction in male babies.[2,3] Presentation of PUV ranges from none to very severe urinary tract obstruction to an extent, that may not be compatible with postnatal life.[4] The degree of obstruction depends on the configuration of the obstructive membrane within the urethra. In utero obstruction may results in hypoplastic kidneys, bilateral hydronephrosis, hydroureter, thickened hypertrophied or dilated urinary bladder. VUR may be detected on routine antenatal ultrasonography.[4,5,6]

In healthy males, the remnant of this are called posterior

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urethral folds or plicae colliculi. Abnormally high insertion and fusion of these primitive folds are believed to be the origin of 95% of PUVs, which are called type I PUV, although Young described a type II PUV. Most paediatric urologists believe that these are not obstructing valves but simply hypertrophy of the plicae colliculi in response to a more distal obstruction as observed in type I and / or type III PUV. Also described by Young, type III PUV that constitutes a septum at the junction of the posterior and anterior urethra instead of a sail like valve. Type III is believed to originate from incomplete dissolution of the urogenital membrane. These pathologies occur during the 3rd month of intra uterine life, causing the urinary out flow obstruction at the posterior urethra.[6] Back pressure leads to proximal urinary dilatation and dysplasia of developing kidneys and poor urination of fetus (oligohydramnios) in serve degree, resulting into hypoplasia of fetal lungs and may have compression effects on the body surface.

Approximately 10-15% of the children undergoing renal transplant have PUV as the cause of renal insufficiency and approximately one third of the patients are born with PUV, that resulted in end stage renal disease (ESRD). Residual urine in PUV predispose to urinary tract infection (UTI). The if vesico-ureteric if present, transmits bacteria and pressure to upper urinary tract leading to pyelonephritis and ultimately chronic renal failure (CRF) in about 7-23% cases in various studies.[7,8]

This study was planned to determine clinical profile of patients with PUV and its relationship to age at diagnosis, renal function and VUR. Early management outcome was also assessed.

PATIENTS AND METHODS:

This was a descriptive study on patients attending Chandka Medical College Hospital Larkana at its Paeds Medicine, Paeds Surgery and Urology Departments from January 2002 to December 2006 with follow-up till June 2008. Patients included in this study were up to 12 years of age. Two cases were suspected on antenatal routine obstetrical ultrasonography. All patients who presented with urinary complaints like dribbling of urine, poor stream, retention, dysuria, pulling at penis, excoriation of the genitalia, urinary tract infection and renal failure were evaluated. Detailed history and clinical examination were done. Urine D/R and culture, blood CP, blood urea, serum creatinine and serum electrolytes were done in all cases.

PUV was suspected initially on ultrasound revealing hydronephrosis, hydroureter, thick walled/ large bladder and some times dilated proximal urethra. Diagnosis was confirmed on voiding cystourethrography detecting obstruction in urethra, onion shaped dilatation of posterior urethra, hypertrophied trabeculated urinary bladder and secondary VUR if present.[9]

Patients were admitted for correction of anaemia and fluid and electrolyte imbalance. Uremic patients required osmotic disuresis with 10% glucose infusion plus frusemide. Intravenous calcium and soda bicarb used in cases with acidosis. UTI was treated by ceftriaxone /ceftazidime /cefoperazone along with aminoglycoside. In cases who had no evidence of obvious UTI, these antibiotics were given as pre/post operative prophylaxis. Urinary catheter was passed to deflate the upper urinary tract. After stabilizing the patients, vesicostomy or fulguration of PUV performed. Three age groups of patients were made and findings recorded and assessed.

We performed vesicostomy in the patients under two years of age due to non availability of cystoscope for this age group. Fulguration of PUV was done in the older boys. Result of surgery in both-groups and complications as well as prognosis recorded in follow-up.

RESULTS:

Fifty eight patients suspected of PUV were evaluated. Out of them 51 male patients with PUV on VCUG were selected for the study. Two were suspected antenatally on ultrasound showing bilateral hydronephrosis and large distended urinary bladder, later confirmed on VCUG after birth. Twenty patients (39%) presented after the age of three years and 8 (15%) at the age of 9 years. Common clinical presentations were fever, anaemia, failure to thrive and urinary complaints (table 1). Twenty seven patients (53%) had abnormal renal function and 21 (40%) had secondary VUR. The frequency of renal failure and VUR increased with age. Nearly all (except one) patient presenting after the age of six years had abnormal renal function and VUR (table 2 and 3). Urinary tract infection (common organism was E-coli) was found in 42 patients.

In patients under 2 years of age (49%), vesicostomy was performed as a staged procedure and other group of patients, older than 2 years, had fulguration as primary procedure (table 4).

Table-1: Clinical Profile (n=51)

Presentation	No. of Patients	Percentage
Recurrent Fever	38	75%
Anaemia	32	64%
Failure to thrive	31	61%
Dysuria	23	45%
Weak Urinary Stream	22	44%
Retention of urine	17	33%
Acidotic Breathing	17	33%
Dribbling of Urine	14	28%
Palpable Bladder	7	14%
Palpable Kidney	6	12%

Table 2 Relationship of Age and VUR

Age (Years)	No Reflux	Unilateral VUR	Bilateral VUR	Total
1-3	8	4	19	31 (60.8%)
4 to 6	1	3	3	7 (13.8%)
7 to 9	1	0	4	5 (9.8%)
10 to 12	0	0	8	8 (15.6%)

Table 3 Relationship of age and renal function

Age (Years)	No. of Patients with Normal Renal Function	No. of Patients with Abnormal Renal Function	Total
1-3	23	8	31 (60.8%)
4 to 6	1	6	7 (13.8%)
7 to 9	0	5	5 (9.8%)
10 to 12	0	8	8 (15.6%)

Table 4 (a) Patients Presenting Under 2 Years of Age

Type of Surgery Performed	No. of Patients	Description
1st stage: Vesicostomy	20	
2nd Stage: Cystoscopic fulguration of PUV	15	3 Patients lost to follow-up 2 Patients Expired after Vesicostomy
3rd Satge: Cystoscopy + closure of vesicostomy	11	Spontaneous Closure of Viscostomy = 4/15

Table 4 (b) Patients > 2 Years of Age

Type of Operation	No. of Patients	Percentage
Fulguration of PUV	28	55%
Fulguration of PUV + Vesicostomy	3	6%
Total	31	61%

Table 5 Post operative complications of Vesicostomy

Complications	No. of Patients	Treatment
Haemorrhage	6	4 Gauze Pack Dressing 2 Resuturing
Wound Infection	6	I/V Antibiotics (3rd Generation Cephalosporin + Amiroglycosides
Skin Excoriations	4	I/V Antibiotics (3rd Generation Cephalosporin + Amiroglycosides
Prolapse of Mucosa	2	Spontaneously reduced
Stenosis of Stoma	3	2 dilatation 1 Refashioning of stoma
Retraction of Vesicostomy	1	Re-do vesicostomy
Uremia with Septicemia	2	Expired
Persistent Uremia	3	Referred for dialysis

Table 6: Post-Operative complications of fulguration of PUV

Complications	No. of Patients	Treatment Required
Transient Haematuria	5	Spontaneously Stopped+I/V Fluids
Retention of Urine	4	Catheterization
Urethral Stenosis	3	Uretheral dilatation
U.T.I	3	I/V Antibiotics
Incomplete ablation	2	Re-Fulguration
Transient Incontinence	1	Spontaneous Recovery
Persistent Uremia	1	Dialysis
Uneventful Recovery	12	

Table 7: Progress of Patients

Prognosis	Early Cases	Late Cases
Resolution of Uremia	83%	48%
Resolution of V.U.R	65%	39%
Catching up of the Normal Growth (up to 30th percentile)	72%	54%
Recurrent U.T.I	20%	38%

Post operative complications of each group are given in table 5 and 6. We did not encounter any major intra operative or post operative complications excepting five neonates who were very low birth weight and had birth asphyxia with compromised renal functions. Two of them died after vesicostomy. Three neonates were lost to follow-up. Progress of the patients is given in table 7.

DISCUSSION:

PUV is often suspected prior to birth on routine antenatal ultrasonography showing bilateral hydronephrosis and / large urinary bladder. This may lead to foetal morbidity and mortality mainly due to renal dysplasia and hypoplasia of lungs.[10,19,20] In our study only two cases were diagnosed on antenatal obstetric ultrasound. More awareness is needed on the part of ultrasonologist about detecting this problem antenatally. At birth patient may present with palpable kidney, retention of urine or renal failure. Older children usually present with UTI, poor urinary stream, dribbling, straining to urinate, diurnal and nocturnal enuresis and palpable urinary bladder and kidneys. VCUG confirms the PUV and also determine VUR. VUR is commonly bilateral and found in 25-50% of cases.[11,15] In our study 81% of cases had VUR. We found VUR more in our patients than other studies possibly as they were referred late (40% after the age of 3 years) whereas most of the cases in Western literature are diagnosed antenatally or in younger age.

Cystoscopy serves both diagnostic and therapeutic purposes. Appropriate infant sized cystoscope (<8 fr) is needed to avoid injury to the urethra. Multiple techniques have been described for ablating the valves. Disruption of the obstructing membrane by blind passage of a valve hook is now outdated. Valves should be disrupted under direct vision by cystoscopy using an endoscopic loop, Bugbee electro-cauterization, or laser fulguration. The objective is to relieve the obstruction by cutting the valves at 12, 5, and 7'O clock position in least traumatic fashion to avoid the urethral injury which may result in the secondary urethral stricture or damage to sphincter mechanism.

Temporary vesicostomy is done in younger patients as urethra may be too small for available cystoscope. It will stabilize the patients of PUV and improve the renal function and size of ureters.[3,14,16] Nowadays fulguration of PUV is done directly without prior diversion of urine.[12,13] Even intrauterine shunting of over distended urinary bladder to amniotic fluid is being tried. It works well and prevent dysplasia of developing foetal kidneys. [2,21,22] PUV is the second most common cause of CRF in children. In one study it was reported in 23% of cases[7,8] while it was seen in 7% cases in Guys' hospital, UK series.[8] It was present in 53% of our cases which is higher and almost all the cases who presented after 6 years had chronic renal failure. In spite of successful ablation of valves some patients may have persistent VUR, and go into ESRD and require renal transplant. Approximately 10-15% of children undergoing renal transplant have PUV. Long term treatment of bladder dysfunction may be required after fulguration.[23].

CONCLUSIONS:

PUV is a common cause of lower urinary tract obstruction. Delay in the diagnosis and management leads to renal damage resulting in ESRD.

REFERENCES:

1. Stephens FP. Congenital intrinsic lesions of posterior urethra in congenital malformation of urinary tract. Praeger publishers. New York 1983:95.
2. Elder JS, Ducket JW. Management of the fetus and neonates with hydronephrosis detected by prenatal ultrsonography. Pediatr Annals 1988;17: 19-28.
3. Rasheed K, Nazir Z, Moazam F. Posterior urethral valves in infants. Role of vesicostomy in preserving renal function. First International symposium of the institute of urology and transplantation 1994, F4 (Abstract).
4. Hinchliffe SA, Chan YF, Jane H. Renal hypoplasia and postnatally acquired cortical loss in children with VUR. Pediatr Nephrol 1992;6:439-44.
5. Colodny AH. In utero diagnosis of urologic abnormalities. Dialogues Pediatr Urol 1983;66:11.
6. Phamane FD, Dommergues M, Muller F et al. Development of human kidney in obstructive uropathy: correlation with ultrasonography and urine biochemistry. Kidney Int 1997;52:21-32.
7. Jamro S. Chronic Renal Failure. Dissertation for FCPS (CPSP), 1994.
8. Caracas RVP. The mild end of clinical spectrum of the PUV. J Pediatr Surg. 1993;28:701-6.
9. Khan FA. Posterior urethral valves a report based on 10 patients. J Pak Med Assoc 1980;30:272-5.
10. Ves M, Muller F. Development of human fetal kidney in obstructive uropathy correlations with ultrasonography and urine biochemistry. Kid Int 1997;52:21-32.
11. Marcos RR, Bissada NK, Alphan T et al. Posterior urethral valves in Saudi Arabia. Int Surg 1983;70:357-60.
12. Glassberg KI. Current issues regarding posterior urethral valves in Saudi Arabia, Int Sug 1985;70:357-60.

13. Glassberg KI. Current issues regarding posterior urethral valves. Urol Clin North Am 1985; 12:175.

14. Rasheed K, Nazir Z. PUV in infant. Experience with vesicostomy in children. Pak J Surg 1988; 4:1-3.

15. Baily RR. Clinical presentation and diagnosis of VUR and reflux nephropathy. Nephrology Proceedings of Xth Intern. Congress of Nephrology 1988, Eds: Davison A.M, Barllier Tindal London PP. 835-43.

16. Malik MA, Iqbal Z, Safdar H, Sial J, Ahmed M. Posterior urethral valves. Professional Med J 2005;12: 473-8.

17. Khan IW, Arim BK. CRF due to obstructive uropathy in children. Ann King Edward Med Coll Lahore 2002;8:194-5.

18. Belman B. Infravesical obstruction In: clinical Pediatric urology, 4th Edn, Eds, Belman, King Kramer, Martin Dunitz 2002.

19. Cromie, Lee K. Implication of prenatal ultrasound screening in the incidence of major genitourinary malformation. J Urol 2001;165:1677-80.

20. Herndon CDA, Ferrer FA, Freedom A. Consensus on prenatal management of antenatally detected urological abnormalities. J Urol 2000;164:1052-6.

21. Holmes N, Harrison MR, Baskin LS. Fetal surgery of PUV: long term postnatal outcomes Pediatrics 2001;107: e7.

22. Misseri R, Horo witz M, Combs AJ. Myogenic failure in PUV disease real or imagined. J Urol 2002;168:1844-8.

23. Roth KS, Carter WH Jr, Chan JCM.: Obstructive nephropathy in children: Long term progression after relief of PUV. Pediatric 2001;107: 1004-10.