

Posterior Urethral Valves; A single Center Experience

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Abstract

Objective: Posterior urethral valves (PUV) are the most common cause of bladder outlet obstruction in infancy that impair renal and bladder function. This study was planned to evaluate and record the various clinical presentations and management, complications, and surgical management and long-term outcome of PUV.

Methods: In a retrospective study, 98 patients who have been treated for PUV are evaluated in Mofid Children's Hospital from January 2007 to December 2012. Detailed history taken and paraclinical examinations were performed in each patient and diagnosis was confirmed by voiding-cysto-urethrography (VCUG). PUV had been ablated in 62 patients by electric hook, and diversion was performed in 42 (42.85%) cases. Data were analyzed by SPSS software version 18.

Findings: Totally 98 patients with mean age at diagnosis 62 (± 13) days were included in this study. Fifty seven cases had been catheterized within one to 6 days of life (mean age one day), PUV was ablated in 62 patients by electric hook, and diversion was performed in 42 cases. The most common symptom in our group was dribbling poor stream 51% and urinary tract infection (UTI) 40.8%. There was vesico-ureteral-reflux (VUR) in 61.2%, and hydronephrosis in 82.6%. Most common associated anomaly was kidney anomalies (multicystic kidney disease and renal agenesis/dysplasia) in 8 (8.2%) patients. Twenty patients had prenatal diagnosis of PUV. Complication occurred in three (3.1%) patients. Mortality occurred in 5 (5.1%) patients. Mean follow-up period was 3.4 \pm 1.2 years (1.5 months to 5 years).

Conclusion: Urinary drainage by feeding tube in early days of infancy, followed by valve ablation is the best treatment in PUV, and urinary diversion improves the outcome. VCUG is still the gold-standard imaging modality for documenting PUVs. The factors like renal dysplasia and UTI have their role in final outcome.

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Key Words: PUV; Urinary Drainage; Valve Ablation; Urinary Diversion; Outcome; Children

Introduction

Congenital mucosal membrane in the prostatic urethra is called posterior urethral valves (PUV)^[1]. It is the most common cause of bladder outlet obstruction in the male neonates and is associated with morbidities, including urinary tract infection

(UTI), chronic renal failure (CRF), urinary incontinence and even death, and the incidence is 1 in 4000 to 25000 live births^[2,3]. The vast majority of patients with PUV are being diagnosed in utero^[4]. PUVs are classified in three types: Valves representing folds extending inferiorly from the verumontanum to the membranous

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urethra (Type 1), Valves as leaflets radiating from the verumontanum proximally to the bladder neck (Type 2.) and Valves as concentric diaphragms within the prostatic urethra, either above or below the verumontanum (Type3). The most common type is type one^[2,5,6]. The first description of PUV was in 1515, and the first clinical case of PUV was reported by Young in 1913 ^[7].

As this congenital anomaly may occur due to a mixed effect of a few minor genes, so it is recommended that PUV should be evaluated in all males in the family, even in asymptomatic ones^[8]. Long-term follow-up for children with PUV is mandatory, even after 20 to 30 years old. In this circumstance, blood pressure, growth and weight, creatinine and electrolytes, urinary tract ultrasonography, diethylene triamine pentacetic acid (DTPA) isotope scan, urodynamic-uroflowmetry, indirect cystography, dimercaptosuccinic acid (DMSA) isotope scan, and voiding cystourethrography evaluations is needed^[9]. PUV is diagnosed by visualization of the valve leaflets, trabeculated bladder, dilated or elongated posterior urethra, and hypertrophied bladder neck. Voiding cystourethrogram (VCUG) is still the gold-standard imaging modality for documenting PUVs. Urinary drainage by feeding tube in early days of infancy, followed by valve ablation is the best treatment in PUV. Surgical management of urethral obstruction is usually carried out by endoscopic ablation of valves^[3]. Post valve ablation management is therefore important in improving the outcome of patients with PUV. Indications for diversion in PUV are: prematurity, small body size, severe urinary infection /septicemia, and high grade renal insufficiency^[10]. Vesicostomy is an approach which protects upper urinary tract complications.

This study was planned to evaluate and record the various clinical presentations, management, complications, surgical management and long-term outcome of PUV.

Subjects and Methods

This retrospective study was conducted at Pediatric Surgery and Nephrology Research Centers of Mofid Children's Hospital from January,

2007 to December, 2012. Ninety eight patients treated for PUV were included in the study. Diagnosis was proven by ultrasonography; VCUG and cystoscopy in all cases, as well as complete blood count (CBC), urine analysis, blood urea, creatinine and serum electrolytes. All neonates with urinary retention received feeding tube in urethra within early neonatal period, and not cured, were treated with fulguration/ablation of the posterior urethral valves by pediatric resectoscope under general anesthesia. Those cases that had problem even after surgery, received feeding tube for a 7-15 days. Vesicostomy was performed in those patients who still had problems with previous interventions. VUR and hydronephrosis were checked every 6 to 12 months by DTPA and DMSA.

All patients had received one third of therapeutic dose of Cephalexin (15 mg/kg/ night), Cotrimaxazol (2 mg/kg/night), and Nitroforantoin (1-2 mg/kg/night) as prophylactic, and were followed for 3.4±1.2 years with laboratory tests checked in every visit and VCUG performed in those who had persistent hydro-ureteronephrosis to check vesico-uretral-reflux (VUR).

Acute renal failure was defined and staged based on RIFLE system criteria^[5]. Chronic kidney disease was defined based on estimated glomerular filtration rate (GFR) and persistent proteinuria^[6]. Data were analyzed by SPSS software version 18.

Findings

A total number of 98 patients with PUV were included in our study, the mean±SD age at diagnosis was 62±13 days (one day to two years) Sixty patients (67.3%) was less than 1 month, 28.1% was 1- 12 months and just 4.1% was more than 1 year. Twenty five patients (25.5%) presented with urinary retention. Symptoms and signs in PUV patients are shown in table 1. Dribbling and poor stream was the most common presenting symptom (51% patients).

Most common associated anomaly was kidney anomaly (multicystic kidney disease and renal agenesis/dysplasia) in 8 (8.2%) patients (Table2). We had to pass feeding tube in 57 (58.16%)

Table 1: Symptoms and signs in posterior urethral valves patients

| Symptoms | Frequency |
|---------------------------|------------|
| Dribbling and poor stream | 50 (51%) |
| Urinary tract infection | 40 (40.8%) |
| Fever | 5 (5.1%) |
| Hematuria | 2 (2.1%) |
| Hypertension | 1 (1%) |
| Total | 98 (100%) |

patients to relieve obstruction, normal urine stream, and correction of urea, creatinine and electrolytes. VUR was presented in 60 (61.2%) of which 30 cases had bilateral and 30 unilateral reflux (VUR grading was V=20 units, IV=20 units, III=18 units, II=22 units and I=10 units). Hydronephrosis presented in 81 (82.7%) patients being mild (30 units), moderate (33 units) and severe (40 units). DTPA at follow-up of our cases showed persistent upper tract dilatation (mostly unilateral) with mild to moderate functional obstruction in 10 patients, which improved later.

PUV was diagnosed in 20 (20%) patients, prenatally. Sixty two patients were treated with fulguration/ ablation of PUV by pediatric resectoscope under general anesthesia. 80 cases had type one PUV. Vesicostomy diversion was performed in 42 (42.85%) cases (either for unsuccessful ablation or later complications). With urethral catheter 22% of cases showed improvement of renal function (fall in serum creatinine level). Mean of follow-up period was 3.4 ± 1.2 years (range: 1.5 months to 5 years).

Complication occurred in three (3.1%) patients who had extravasation of urine due to kidney perforation, that was managed by temporary vesicostomy. Five (5.1%) patients died due to renal failure (3 patients) and urosepsis (2 patients). The long-term outcome of our study is presented in Table 3.

Discussion

PUV is the most common cause of infra-vesical obstruction in male children. The spectrum of severity of PUV varies from mild to lethal forms, according to the degree of obstruction^[11]. Patients may develop complications even after valve ablation and on long-term follow-up. Management of PUV needs adequate neonatal and infant care with nephrological support, to treat UTI, prevent/correct metabolic acidosis and electrolyte imbalance if necessary^[12,13]. Improved management of patients with severe PUV has resulted in a better long-term outcome^[14]. There are three anatomical variables in PUV which may provide a "pop-off" mechanism: 1) urinoma/urinary ascites, 2) syndrome of PUVs, 3) PUV+ large congenital diverticle, that results in preservation of intact renal and bladder function^[14-16]. Urinary ascites is the result of leakage of urine from urinary system, and usually there is high level of blood urea nitrogen and serum creatinine^[17]. In this situation pop-off mechanism preserves the kidney from excessive pressure, and is a good prognostic sign^[14,15,18].

We had three cases of urinary ascites in our study which improved by temporary vesicostomy. Patients who failed to respond to urinary catheter drainage, associated severe VUR, and urinary leakage, finally required diversion^[19,20]. Ninety

Table 2: Associated anomalies in posterior urethral valves patients

| Anomalies | Frequency |
|--|------------|
| Kidney anomalies (Multicystic kidney, renal agenesis/ dysplasia) | 8 (8.2%) |
| Cardiac anomaly (PDA, ASD) | 6 (6.2%) |
| Others (UDT, Imperforate anus, Prune belly) | 3 (3%) |
| None | 81 (82.7%) |
| Total | 98 (100%) |

PDA: Patent ductus arteriosus; ASD: Atrial septal defect; UDT: Undescended testis

Table 3: Long term outcome of posterior urethral valves

| Outcome | Patients (n=98) | % |
|------------------------|-----------------|------|
| Acute renal failure | 45 | 46 |
| Residual valve | 15 | 15.3 |
| Stricture | 7 | 7.1 |
| Chronic kidney disease | 6 | 6.1 |
| Dialysis | 6 | 6.1 |
| Mortality | 5 | 5.1 |
| Hypertension | 1 | 1.0 |

four (96%) patients of our study group were in the first year of life, in Choudhury et al^[21] study 77%, Were in this age group and in Malik et al's^[2] series less than 30%. The incidence of renal failure at presentation is reported 66-90% in literature, but in Choudhury et al^[20] group it was 71%, and in our series 46%, of which 6.1% required dialysis. The most common symptom in Malik et al^[2] study was associated fever (72%) whereas in our group it was dribbling and poor stream (51%). In our cases there was 61.2% VUR (right 10.2%, left 20.4% and bilateral 30.6%), while it was 22% (16% left and 6% bilateral) in Malik et al^[2], and Sudarsanan et al^[22] had 12 bilateral VUR and 8 hydronephroses. In our study VUR subsided within 3 to 4 months in majority of cases post valve ablation. UTI was present in 40.8% of our patients which cured by antibiotics, and in severe/resistant cases with diversion^[23,24]. DTPA at follow-up of our cases showed persistent upper tract dilatation (mostly unilateral) with mild to moderate functional obstruction in 10 patients, which improved in later follow-up. Our long term outcome of PUV patients is presented in Table 3, and compared with other studies in Table 4^[21-23,25,26]. The reported incidence of stricture following endoscopic ablation is between 3.6 to 25%^[21]. Incidence of 7.1% means an improved result in

our study. Our residual valves incidence rate was more than the other reports, may be it depends on our technique or available instruments, or both. Our diversion rate and renal failure at follow-up was acceptable comparing to the other studies, but mortality rate was higher than in other reports.

Conclusion

Urinary drainage by feeding tube in early days of infancy, followed by valve ablation is the best treatment in PUV, and urinary diversion improves the outcome. VCUg is still the gold-standard imaging modality for documenting PUV. The factors like renal dysplasia and UTI have their role in final outcome.

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Conflict of Interest: None

Table 4: Comparison with other studies in PUV patients

| Parameter | Lal ²³ | Basu ²⁵ | Choudhury ²¹ | Shittu ²⁶ | Sudarsanan ²² | Our study |
|----------------------|-------------------|--------------------|-------------------------|----------------------|--------------------------|--------------------------|
| No. of patients | 82 | 130 | 90 | 26 | 61 | 98 |
| Treatment modalities | Fulguration | Fulguration | Fulguration | Mohan's valvotome | Fulguration or valvotome | Fulguration or valvotome |
| Follow-up period | 1-21 yrs | ND | ND | 18 mo-5 yrs | 8-75 mo | 3.4 yrs |
| Stricture rates (%) | 3.6 | ND | ND | 5 | 8.2 | 7.1 |
| Residual valves (%) | 13.4 | 6.2 | ND | ND | 13 | 15.3 |
| Diversion rates (%) | 46 | 20 | 71 | 0 | 1.6 | 42.8 |
| RF at follow-up (%) | ND | ND | 8.8 | 20 | 1.6 | 6 |
| Mortality rates (%) | ND | 0.8 | 3.3 | ND | 0 | 5.1 |

ND: Not documented; RF: Renal failure

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