

Outcome of Posterior Urethral Valves in Public Sector Hospital: Challenges in Management

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ABSTRACT

Aim: To determine the pattern of presentation and outcome of management of posterior urethral valve in a resource-limited setting.

Methods: A descriptive retrospective study from October 2012 to October 2014 was conducted at Department of Pediatric Urology, The Children's Hospital & ICH Multan. Data including pattern of presentation, duration of symptoms, complications and outcome of management were analyzed.

Results: Two hundred thirty patients were seen. The median age was 3 years (2 days–10 years). The mean duration of symptoms before presentation was 2.4 years. Two hundred and 10(91%) patients presented with urosepsis, eighty patients (35%) with unilateral or bilateral reflux, 35(15%) presented with large decompensated bladder, while 40 patients (17%) presented with significant renal insufficiency. Radiological findings confirmed the diagnosis of posterior urethral valve. Cystoscopy and fulguration of valves was done in all patients and vesicostomy or ureterostomy was later done in selected cases.

Conclusion: Study results concluded that late presentation is common in our setting. This is associated with high morbidity and mortality rates. Efforts at improving awareness and early diagnosis among the health team should be made to stem the tide.

Keywords: Urosepsis, vesicoureteral reflux, renal insufficiency, vesicostomy, ureterostomy.

INTRODUCTION

Posterior urethral valves (PUV) are the commonest cause of lower urinary tract obstruction in male infants. The incidence of this congenital anomaly in our setting is unknown, although reports from United States and Europe indicate that it occurs in about 1:8000 and 1:25,000 male live births and make up 10% of urinary obstructions diagnosed in utero¹.

In developed world an increasing number of PUV cases are identified by prenatal ultrasonography². This could be related to the widespread use of Ultrasound during pregnancy and the improvement in technique.

The incidence of posterior urethral valves is dropping in some populations due to the effects of prenatal diagnosis and subsequent termination of affected fetuses. In one report, fetuses diagnosed with valves were electively terminated in 46% of cases³.

Children presented with various degree of symptoms depending on age of presentation and types of valves. Sometimes recurrent urinary tract infection and failure to thrive may be the only feature⁴. Prolonged and unrelieved lower urinary tract obstruction leads to back pressure effects on the kidneys resulting in obstructive uropathy with renal

impairment. PUVs are also a common cause of chronic renal failure in children if treatment is delayed⁵.

Despite advances in the medical and surgical management of PUV, 13%-64% of children still develop chronic renal failure (CRF) or end-stage renal disease (ESRD) during long-term follow up⁶. Prenatal diagnosis of PUV has not improved this rate. There are still many aspects of PUV, including the pathophysiology and effects of treatment, which are not completely understood.

This aim of this study is to evaluate posterior urethral valves (PUV) in a developing country with focus on the profile of the disease, determinants of outcome and the challenges of management.

PATIENTS AND METHODS

It is a descriptive retrospective study conducted at the Department of Pediatric Urology, Children Hospital & ICH Multan. Records of patients whose diagnosis of PUV was confirmed with voiding cystourethrogram, renal ultrasound and cystoscopy were reviewed and included in this study. Duration of study was of two years from October 2012 to October 2014. Some patients with renal failure were managed by both the pediatric nephrology and urology units of children hospital complex. Data such as age, presenting symptoms with their duration, any

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complications and their management were noted on proforma. All basic investigations like (CBC, CUE and culture sensitivity, RPM, S/E) diagnostic imaging like Ultrasound abdomen, Voiding cystourethrogram and DTPA renal scan were done.

Patients were initially stabilized with temporary catheter drainage, antibiotics and intravenous fluid if needed. Some patients with severe renal failure were managed by peritoneal dialysis. After initial stabilization, Cystoscopy and fulguration of valves was done in all patients and Foleys catheter was placed for two to five days. Patients were discharged on 2nd day with antibiotics for four to ten days. Routine first follow up was at two weeks and second follow up was at six week and third follow up was at 4 months. All routine tests like RFT, CUE & culture sensitivity and ultrasound abdomen were done at every visit. MCUG was done at four month and DTPA and DMSA renal scan were repeated at 6 months in patients with unilateral or bilateral reflux and renal failure.

After fulguration patients with VUR who presented with recurrent acute UTI, or progressive renal deterioration, were further managed by supravescical urinary diversion. Patients with progressive renal failure were given supportive treatment in Pediatric nephrology ward with medication, peritoneal or haemodialysis. Data was analyzed by SPSS-20.

RESULTS

There were two hundred thirty patients included in the study. The median age at presentation was 2.75 years (range 2 days to 10 years) and the mean duration of symptoms was 2.59 years. Only 10 patients (4.3%) were diagnosed on antenatal scans. Difficulty in passing urine was the most common abnormality which was found in almost all patients (100%). Next most common presentation was recurrent urinary tract infection with fever in 207 patients (90%); (Escherichia coli accounted for 66%, Pseudomonas aeruginosa 5% Klebsiella 14%), while 15% did not grow any organism. Failure to thrive was found in 65 patients (28.2%).

On examination palpable bladder was found in 175 patients (76%), anemia (haemoglobin <10 mg/dl) in 131 patients (56.9%) and high blood pressure in 20 patients (8.7%). Serum creatinine at presentation was below 1.5mg/dl in 90 patients and greater than 1.8mg/dl in 50 patients. Thirty five patients (15.2%) needed peritoneal dialysis before surgery. On cystourethrogram vesicoureteral reflux was found in 80 patients (34.7%), Unilateral reflux was found in 45 patients (19.5%) while bilateral reflux was found in 35

patients (15.2%). Large decompensated bladder was found in 35 patients (15%).

After surgery, average follow up was six to eighteen months, 150 patients (65%) remained stable and finally stopped follow up. Seventy patients (30.4%) suffered from recurrent urinary tract infection (>3 episodes /year) requiring admission in hospital and intravenous antibiotics. Persistent vesicoureteral reflux resulted in loss of one or both kidneys in 45 pts (19.5%) and cutaneous ureterostomy was performed later in 10 pts (4.3%) and none had vesicostomy. Forty five patients (19.5%) developed chronic renal failure and survived on dialysis (23% received peritoneal dialysis and 8% received hemodialysis). The remaining 69% were lost to follow up or presumably died for inability to access required management for profound renal impairment.

Figure 1: Age distribution of patients at presentation.

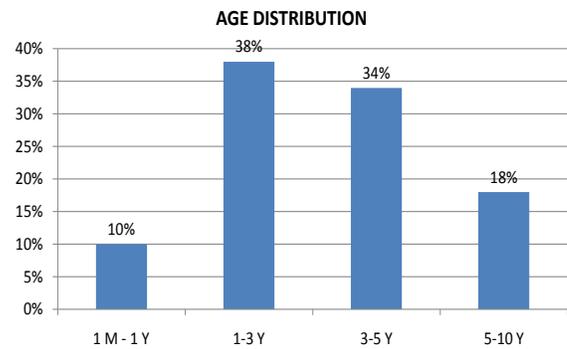
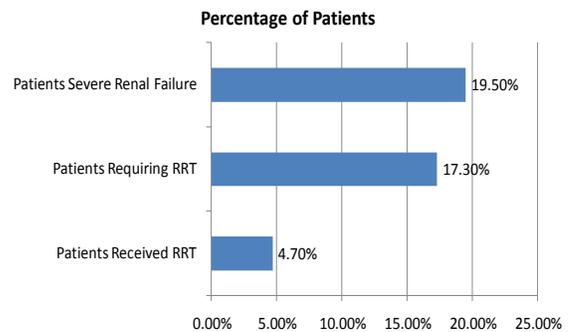


Figure 2-Availability & Accessibility of Renal Replacement Therapy



DISCUSSION

Congenital obstruction of the urethra is one of the most devastating anomalies that occur in the urinary tract and is one of the few that are life threatening in the neonatal period. These lesions usually result in life long disabilities with incontinence and decreased renal function despite optimal medical management^{7,8}.

Age at presentation has been taken as a predictor of outcome of renal function with PUV. But the data on this issue is conflicting. Early presentation enables early diagnosis and intervention. This enables better preservation of renal function^{9,10}. With early recognition complications of obstructive uropathy like hydronephrosis, progressive renal damage and urinary ascites can be prevented¹¹. While the diagnosis of PUV can be made prenatally^{12,13}, only 10(4.3%) of our patients were diagnosed prenatally (Fig. 1). Negative attitude, long distances to service providers, considerably heavy financial cost are major barriers to prenatal ultrasound.

However certain studies have shown that contrary to early belief, early diagnosis is more commonly seen in severe degree of hydronephrosis, which in turn is reflected in a poor functional outcome¹⁴. Ansari and colleagues compared 95 patients who presented before two years of age with 99 patients who presented after two years¹⁵. They found that the incidence of azotemia, mean serum creatinine and risk for ESRD were worse in patients who presented after two years of age. The conflicting data in numerous studies indicate that postnatal age at diagnosis alone is not a reliable factor in predicting outcome in valve patients.

Most common symptom reported was difficulty in passing urine, poor urinary stream or dribbling of urine. It was reported in almost all of our patients.

Recurrent Urinary tract infection (90% in our series) is frequently encountered complication of PUV and may be presenting feature in some cases as well. The pathogenesis of UTI in these patients is complex and multifactorial¹⁶. Contributing factors include urinary stasis arising from anatomic or functional obstruction, dysfunctional elimination syndrome, instrumentation of urinary tract and VUR. UTI can lead to renal scarring and hasten the progression of CKD to end stage renal failure¹⁷.

Elevated serum creatinine level at presentation has been shown to be associated with poor prognosis in PUV patients¹⁸. Approximately 25-40% of infants with PUVs develop renal insufficiency before adolescence^{19,20}. Rising creatinine, even transiently, following urinary tract drainage in neonates with posterior urethral valves is significant

and is a new and important indicator of long-term prognosis.

VUR is found in one-third to one-half of patients with this congenital abnormality and is usually secondary to high intravesical pressure. Milder grades of reflux resolves spontaneously but high grades of reflux occurring in a kidney with poor differential function rarely undergo complete resolution. Many patients who had retained stable renal function in childhood were nevertheless observed to progress into end stage renal failure (ESRF) after puberty. Growing evidence documents the association between ESRD and variety of other renal risk factors such as the presence of VUR, UTI, and renal dysplasia (Fig. 2).

Most of our patients presented late with features such as VUR (35%) and renal failure (19.5%). The reasons for delayed presentation among our patients are connected with pervasive poverty prevalent in the population. Another possible reason is the ignorance that a poor urinary stream could be a transient event and that the baby will improve with time. The healthcare poor referral system could be a cause of late presentation. Therefore, with early presentation, diagnosis and treatment the outcome is expected to improve. The survival figure of 68% in this study is low compared to other centers in Asia (87.5%) and western world (96.2-100%)^{21,22,23}.

These are pointers that require urgent attention. Medical education programs to improve the awareness among general practitioners should be encouraged. Mothers presenting with oligohydramnios to the obstetricians should have their neonates screened for PUV when born. Patients presenting with features of urinary tract infection should have a full workup to rule out underlying urogenital anomaly.

CONCLUSION

Early diagnosis and prompt commencement of treatment is therefore germane to the overall outcome of these patients. This is particularly important in a resource - limited environment like ours where facilities for renal replacement therapy in children is not readily available.

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