

Profile and Outcome of Pelviureteric Junction Obstruction

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Abstract: Obstructive uropathy represents one of the largest fractions of identifiable causes of renal failure in pediatric populations. This is one of the largest series of PUJ obstructions reported in children. PUJ obstruction is a relatively common congenital anomaly in neonates and children and a cause of ESRD in developing countries. This series expands our knowledge about the clinical presentation and course of this disease. Pelviureteric junction (PUJ) obstruction is a partial or total blockage of the flow of urine that occurs where the ureter enters the kidney and is the most common cause of significant dilatation of the collecting system in the fetal kidney. We retrospectively analyzed children diagnosed as PUJ obstruction between the periods of 2008-2010 who presented to our institute. Forty four children were diagnosed to have PUJ obstruction. The median age at presentation was 11 months, with range of 3 months to 11 years. Male children constituted 34 (77.3%) cases. Unilateral PUJ with left sided involvement was more common (52.3%). Antenatal diagnosis was made in 18 (40.9%) cases. The common clinical presentations were fever, urinary tract infection (UTI) and abdominal pain. Hypertension was present in five (11.3%) patients and renal failure at presentation was seen in eight (18.2%) patients. Associated urogenital anomalies were present in twelve (27.3%) patients, with medullary cystic kidney disease (MCKD) being the most common associated anomaly. Prophylactic antibiotics were not prescribed to patients. Urinary tract infection was present in fourteen (31.9%) patients with *E. coli* being the most common organism (75%). Conservative management with regular follow up was done for 32 patients and the remaining twelve patients underwent pyeloplasty in view of gross hydronephrosis, and four (33.3%) of these patients later underwent nephrectomy for a non-functioning kidney. Antenatal screening is mandatory to make an early diagnosis and for further management and close follow up of PUJ obstruction. Randomized control trials and longer follow up studies are needed for evidence to suggest the optimal management.

Keywords: Children, hydronephrosis, pelviureteric junction obstruction, pyeloplasty.

INTRODUCTION

Pelviureteric junction (PUJ) obstruction is defined as impediment in urine flow from the renal pelvis to proximal ureter due to structural narrowing of the pelvi ureteric junction leading to hydronephrosis and if left uncorrected, leads to progressive renal deterioration. Congenital causes are more common than acquired conditions.

METHODS

Retrospective analysis was done for children at Govt. Institute of Child Health and Hospital for Children, Chennai who were diagnosed with PUJ obstruction between the period 2008 to 2010. The clinical profiles of age at presentation, fever, blood pressure, abdominal pain, abdominal mass, oliguria, hematuria, nausea, vomiting, urinary symptoms and UTI were noted. Associated renal anomalies were also searched for. Investigations like blood urea, serum creatinine, urine analysis, PCR, culture & sensitivity were done. Ultrasound was done to detect the size, site and lateralization of kidney involved, anomalies like ectopic or cystic kidneys or calculus, extrarenal

pelvis and double ureters. DTPA was done to assess split renal function. Surgical indications for pyeloplasty were palpable renal mass, presence of cortical thinning, AP renal pelvic diameter > 2.5 cms, split renal function by DTPA scan < 40%. Nephrectomy was done if the kidney was defunct and renal function was < 10%. The follow up data with outcomes were analyzed.

RESULTS

44 children had PUJ obstruction. The median age at presentation was 11 months with range of 3 months to 11 years. 25% presented before the age of two months and 60% were detected by one year of age. Males constituted the majority of cases, with male to female ratio of 4:1. Unilateral left sided PUJ obstruction was seen in 23 children (52.3%), followed by bilateral obstruction in 12 (27.3%) children and right sided in 9 (20.4%) children. Antenatal diagnosis was made in 18 (40.9%) infants. The common clinical presentation included fever (50%), UTI (40.4%) and abdominal pain (15.9%); (Table 1).

Hypertension was present in five (11.3%) patients; 5 (62.5%) of 8CKD patients had proteinuria but in the subnephrotic range. Associated urogenital anomaly was present in twelve (27.3%) with multicystic dysplastic kidney (MCDK) being the most common; (Table 2). Prophylactic

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Table 1. Clinical presentation of PUJ obstruction.

Clinical Presentation	Number (%)
Fever	22 (50.0%)
UTI	18 (40.4%)
Abdominal pain	7 (15.9%)
Oliguria	6 (13.6%)
Urinary symptoms	5 (11.3%)
Hematuria	2 (4.6%)
Abdominal mass Nausea & Vomiting	2 (4.6%) 2 (4.6%)

antibiotics were not given for these patients. UTI was present in 14 (31.9%) patients and *E. coli* was the most common organism, identified in 75%. Conservative management with regular follow up was done for 32 patients and the remaining 12 patients underwent pyeloplasty in view of gross hydronephrosis with cortical thinning. One-third of patients who underwent pyeloplasty later underwent nephrectomy due to obstructive non-functioning kidney. Eight (18.6%) children progressed to chronic kidney disease. Death due to septicemia and acute renal failure occurred in 2 infants aged 2 and 3 months due to gross hydronephrosis (AP diameter of right renal pelvis 6.1cms (normal < 2.5 cms) in the former and multicystic dysplastic kidneys in both). Amongst the longest follow up group of 3 years, 1 of 9 (11%) operated progressed to chronic kidney disease due to bilateral PUJ obstruction and right gross hydronephrosis; 7 children are doing well without surgery maintaining a normal serum creatinine. In the 2 year follow up group, 3 of 6 children progressed to CKD due to associated conditions like multiple calculi, thinned out cortex and one with multicystic dysplastic kidney, while 3 of 7 unoperated children progressed to CKD as the contralateral kidney had a multicystic dysplastic kidney. At one year follow up in the unoperated children, 2 out of 3 progressed to CKD as one child had contralateral dysplastic kidney and the other two had multicystic dysplastic kidneys.

Table 2. Urogenital anomalies in PUJ obstruction.

Associated Anomaly	Number (%)
MCDK	6 (14.0%)
Extra renal pelvis	3 (6.9%)
Dysplastic kidney	2 (4.6%)
Renal stones	1 (2.3%)
Ectopic kidney	1 (2.3%)

DISCUSSION

PUJ obstruction can occur in children of all age groups. Nowadays, the majority of cases are detected in the perinatal period with the advent of modern imaging techniques [1]. In a retrospective study, functionally significant PUJ obstruction was noted in one in 1500 fetuses screened by antenatal ultrasonography [2]. Boys were affected with PUJ

obstruction more commonly than girls [2]. The left side is more frequently involved. Our series also shows a similar presentation. PUJ obstruction may be inherited as an autosomal dominant gene of variable penetrance [3]. Congenital PUJ obstruction is usually caused by intrinsic stenosis of the proximal ureter as a result of an interruption in the development of the circular musculature of the PUJ or an alteration of collagen fibers and composition between and around the muscle cells [4]. Other causes of intrinsic PUJ obstruction include valvular mucosal folds, persistent fetal convolutions and upper ureteral polyps. In about 10 percent of pediatric PUJ obstruction, an aberrant or accessory renal artery or arterial branch can cause extrinsic compression.

PUJ obstruction accounts for about 48 percent of cases of dilatation of the collecting system of fetal kidneys [5]. Presentation in the newborn may include a palpable abdominal mass caused by an enlarged obstructed kidney. Other presentations include urinary tract infection, hematuria or failure to thrive. Renal failure is not a common presentation. Older children can present with intermittent flank pain which may worsen during brisk diuresis and have associated nausea and vomiting [6].

The diagnosis of PUJ obstruction is generally suspected when imaging studies, usually ultrasonography demonstrate pelvi-caliectasis. PUJ obstruction should be differentiated from other frequent causes of antenatal hydronephrosis, such as transient and functional hydronephrosis, ureterovesical junction obstruction, and vesicoureteric reflux (VUR). Other differential diagnoses for PUJ obstruction include posterior urethral valves, congenital megaureter, ureterocele and multicystic dysplastic kidney. Diuretic renography can be used to diagnose urinary tract obstruction. In general, a half-life greater than 20 minutes to clear the isotope from the kidney is considered to be indicative of obstruction. Computed tomography (CT) is an alternative to ultrasonography. A voiding cystourethrogram (VCU) will help to rule out VUR. PUJ obstruction and VUR can coexist in ten percent of cases. Co-existence of VUR can be a contributory factor for critical pelvic distension and tortuous PUJ [7]. Identification of VUR is important because children with concurrent VUR and PUJ obstruction may be at higher risk for severe infection. Magnetic Resonance Imaging (MRI) with urography is being investigated to define urinary tract anatomy, calculate differential renal function, and assess urinary tract obstruction [8].

There are no randomized trials that provide evidence for the optimal management of PUJ obstruction. There is no evidence that prenatal intervention in infants with either a single obstructed kidney or bilateral involvement improves renal outcome [9]. Prenatal intervention is needed if decompression of an enormously dilated renal pelvis is necessary to prevent dystocia or pulmonary compression [10]. Asymptomatic patients need regular follow up only [11]. The natural course of PUJ obstruction is variable. Many patients will have stable renal function and improvement in the degree of hydronephrosis during long periods of observation. However, there is controversy about the role of observation. No study has clearly demonstrated the need for surgical intervention [12]. The main criterion for observation in cases with significant hydronephrosis on sonogram is demonstration of split renal function of greater

than 40 percent in the affected kidney by diuretic renography, even if washout is delayed [13]. If renography shows deterioration of >10 percent on the affected side, or relative function less than 40 percent, surgery is recommended. The role of antibiotic prophylaxis is unclear. Studies show UTI incidence of about five percent in PUJ obstruction [14].

Symptomatic children usually require operative intervention. In these patients, pyeloplasty is performed and consists of resecting the atretic or stenotic segment and reattaching the normal ureter to the renal pelvis, thereby relieving the obstruction. If the obstruction is due to an aberrant renal blood vessel, the PUJ is repositioned relieving the obstruction. In patients with PUJ obstruction that present with acute pyelonephritis, surgical repair is performed after the infection has cleared with antibiotics. If pyelonephritis does not respond to antibiotics, a temporary percutaneous pyelostomy tube can be placed to relieve the obstruction. Usual indications for surgery include symptomatic PUJ-like pain, infections, or increase in the grade of hydronephrosis with decrease in renal function, or gross hydronephrosis with renal pelvic diameter of more than 50 mm.

There is limited data on the long-term outcome of children with PUJ obstruction. In one series of children who underwent surgical repair, relative renal function improved from 37 percent preoperatively to 43 percent [15]. Five percent have recurrence of obstruction after surgery [16]. It remains unknown what proportion of patients will progress and whether there is a difference in long-term renal function between the observational and initial surgical approaches.

PROGNOSIS

Patients with an antenatal diagnosis with good renal function at the time of detection do well with conservative treatment [17]. David ben Meir *et al.* evaluated the relationship between initial effective renal plasma flow and final post surgical outcome in patients with PUJ obstruction and they have published a few important observations. Final effective renal plasma flow (ERPF) was negatively related to age (younger children), and those with a higher preoperative ERPF recovered better than older children and those with a lower ERPF. Most kidneys in each functional range improved when obstruction was relieved. A relative renal flow > 51% in the obstructed kidney by ^{99m}technetium mercaptoacetyl triglycine study was not always beneficial in prognosticating, and may be a warning of impending decompensation in a minority [18, 19].

In unilateral PUJ obstruction, the development of clinically significant hypertension or proteinuria appears to be very rare. Bilateral disease has a more ominous prognosis. A marked loss of renal mass has already occurred before birth as a result of either maldevelopment or obstructive damage. Adverse prognostic factors include gestational age at presentation, presence of VUR or altered echogenicity at the time of diagnosis, level of serum creatinine at one year of age, the occurrence of UTI, and the appearance of proteinuria or hypertension.

CONCLUSION

This case series highlights the need for a mandatory antenatal screening for early diagnosis and management of PUJ obstruction. Randomized control trials and long term follow up studies are needed to provide evidence for optimal management.

CONFLICT OF INTEREST

The authors confirm that this article content has no conflict of interest.

ACKNOWLEDGEMENTS

Declared none.

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Received: January 26, 2014

Revised: May 22, 2014

Accepted: June 15, 2014

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