Original Research Article

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Clinical profile and outcome of pelvi-ureteric junction obstruction (PUJO) in children presenting above 1 year

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ABSTRACT

Background: Pelvi-ureteric junction obstruction (PUJO) is defined as anatomical or functional obstruction to the flow of urine from the pelvis to the upper ureter. With the advent of ultrasonography most of the cases of PUJO are identified in antenatal period. The treatment protocol for antenatal diagnosed PUJO is standardized. Few children present later in life. Present study was done to evaluate clinical profile and outcome of children presenting with PUJO above 1 year.

Methods: A retrospective observational study conducted by the department of Paediatric Surgery, KIMS hospital Bangalore. All children with PUJO above 1 year of age from January 2015 to January 2018 were included. Clinical, biochemical and imaging findings of these children were tabulated. A diuretic renogram was done to confirm the diagnosis. The children underwent either pyeloplasty or nephrectomy, depending on the function of the kidney. In children with calculi, pyelolithotomy with pyeloplasty was done. The children were followed up post-surgery.

Results: Twenty-six children were included in the study. Mean age of presentation was 6.86±4.13years. Most common presentation was pain abdomen. Left kidney was commonly involved. The mean anteroposterior pelvic dimension (APPD) of involved kidney was 38.96±17.7 mm. 5 children presented with poorly functioning kidney with split renal function less than 10%. In three children PUJO was associated with pelvic calculi. Out of the 26 children simple nephrectomy was required in 4 (15%) children. The remaining 22 children underwent pyeloplasty. Mean duration of stay was 6.19±1.8 days. Post-pyeloplasty diuretic renogram showed improved drainage with improved function.

Conclusions: PUJO is common cause of urinary obstruction in children. Delay in diagnosis or presentation leads to increased chances of renal damage and loss. Hence it is important to diagnose PUJ obstruction at the earliest and receive prompt treatment.

Keywords: Nephrectomy, Pelvi-ureteric junction, Pyeloplasty

INTRODUCTION

Pelvi-ureteric junction (PUJ) obstruction is the commonest cause of pediatric hydronephrosis occurring in 1 in 1000-2000 live births. PUJO is defined as

obstruction to the flow of urine from the kidney to proximal ureter that results in renal damage and symptoms. PUJ obstruction was first described by Dietl in 1864.² The fibrotic change at the PUJ was demonstrated by Allen TD in 1970.³ Pelvi-ureteric

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Junction Obstruction (PUJO) is broadly classified as primary and secondary. Most of the cases of PUJO are congenital where etiology is due to insertion anomalies of ureter, ureteral muscular hypertrophy or peri pelvicalyceal fibrosis. Secondary PUJO results either from post-operative or inflammatory strictures, calculi or urothelial neoplasms. Obstruction leads to increase back pressure on the kidneys leading to progressive deterioration of function.

Most of the cases of hydronephrosis are now diagnosed by antenatal scans. Based on the presentation PUJO can be categorized into two groups-first where the condition is diagnosed antenatally and the second where presentation is due to symptoms at later age. Children diagnosed in antenatal period are followed postnatally. Depending upon the dimension of the renal pelvis and symptoms these children are either treated conservatively or surgically. Indications for surgery in PUJO are palpable kidneys, obstruction with differential function <40%, T ½ greater than 20 minutes on diuretic renogram and deterioration of renal function. PUJO can present at any time between neonatal and old age. In older children PUJO has varied presentation and outcomes. Delay in presentation or detection of PUJO is associated with renal damage. The aim of this study was to evaluate the outcomes of children presenting with PUJ obstruction beyond one year of life.

METHODS

This was a retrospective observational study done in the department of Pediatric Surgery, Kempegowda Institute of Medical Sciences, Bangalore over a period of 3 years, from January 2015 to January 2018. All children with pelvi-ureteric junction obstruction (PUJO) above 1 year of age were included in the study.

The data of children with respect to sex, age, symptoms and their duration were tabulated. All children initially underwent an Ultrasound of the Abdomen to evaluate the side and extent of hydronephrosis. The anteroposterior dimension of pelvis was documented. In few children, Computed Tomography scan was done, especially in children presenting with renal or pelvic calculi. A diuretic Renogram was done to confirm the diagnosis of PUJO. In children with poorly functioning kidney on diethylene triamine pentaacetic acid (DTPA) or ethylenedicysteine (EC) scan, dimercaptosuccinic acid (DMSA) was done to document the cortical function. Other routine blood investigations were done as per protocol before surgery.

The children underwent either pyeloplasty or Nephrectomy, depending on the function of the kidney. In children with calculi, pyelolithotomy with pyeloplasty was done. Pyeloplasty was done by Anderson Hynes dismembered pyeloplasty technique. In all children a DJ stent was placed across the pelvi-ureteric anastomosis, which was cystoscopically removed after 6 weeks.



Figure 1: Dilated pelvis with PUJO.

The children were followed up post-surgery. The range of follow-up was from 6 months to 3 years. In children who underwent pyeloplasty a diuretic renogram was done 6 months post-surgery to evaluate the drainage pattern across the PUJ.

RESULTS

Out of the 40 children who presented with PUJ obstruction in the study period, 26 children above the age of 1 year were included in the study. Mean age of presentation was 6.86±4.13 years. Age distribution of the children is shown in table 1. Out of 26 children 19 were males and 07 females.

Table 1: Sex and age distribution of children.

Characteristics	Number	Number	Percentage
	< 5years	08	31
Age	5-10 years	11	42
	>10 years	07	27
C.	Male	19	73
Sex	Female	07	27

The symptoms and their duration are depicted (Table 2). Most common presentation was pain abdomen followed by recurrent fever.

Table 2: Distribution of study subjects according to the presenting complaints (multiple response).

Symptom	Number	Percentage
Pain abdomen	18	69
Vomiting	03	11.5
Fever	07	27
Abdominal distention	03	11.5
Antenatally diagnosed/ non- compliant to follow-up	07	27

The data about general physical examination of the included children is shown in Table 3.

Table 3: Distribution of subjects according to the nutritional status and general examination findings.

Characteristics	Findings	No.	Percentage
Nutritional status	Normal	22	84.62
	Mild/moderate malnutrition	04	15.38
Pallor	Present	03	11.5
	Absent	23	88.5

Ultrasonography showed left Hydronephrosis (HDN) in 69 % and right HDN in 31 % patients. The range of pelvic dimension on USG was between 20 mm to 80 mm, with mean dimension of 38.96 mm. CT scan was done in 3 children with renal calculi. The largest calculi measured 15mm.

The DTPA/EC and DMSA findings have been tabulated in the table 4. The median differential function of the affected kidney on Diuretic renogram was 36% with range of 1% to 49%.

Table 4: Distribution of surgeries between both the kidneys.

Pyelopla	lasty (22) Simple nephrectomy (4)		ephrectomy (4)
Right	Left	Right	Left
06	16	2	2

Out of the 26 children, 5 children had a split renal function of less than 10% in the affected Kidney. In 3 children out of the 5, percutaneous nephrostomy (PCN) was done. In only one child with PCN there was improvement in the renal function and a pyeloplasty was done after 6weeks. In the remaining two a nephrectomy had to be done as the function did not improve even after 6 weeks of drainage. So overall in only one child with poorly functioning kidney a pyeloplasty could be done and in remaining four Nephrectomy had to be done. The age of the child in whom the kidney function improved after PCN was 18 months.

The mean age of children who underwent Nephrectomy was 11.5 years. Kidneys have a better chance to improve in smaller children compared to older children. Older children with poorly functioning kidneys end up with Nephrectomy.

Two children presented with giant HDN. One child was 18 months old and the other 12 years. Both the children underwent PCN initially. Post PCN, the kidney improved in the younger child whereas older child required nephrectomy.

Three children presented with calculi with PUJ obstruction. In these children pyelolithotomy with pyeloplasty was done. The mean age of children presenting with calculi was 7.5 years. Post-surgery these children are on regular follow up. No recurrent calculi have been noticed in these children.

In three children (2, 5 and 13 years) the PUJ obstruction was due to an external compression of the PUJ by lower pole vessels. A standard Anderson Hynes pyeloplasty was done in these two children.

The remaining children underwent Andersen Hynes pyeloplasty. In all the children who underwent pyeloplasty a DJ stent was placed. Stent was removed six weeks post procedure by a cystoscope.

All the children had an uneventful post-operative recovery. The mean duration of stay was 6.19±1.8 days. Post-surgery diuretic renogram showed improved drainage with improved function in children who underwent pyeloplasty.

DISCUSSION

Pelvi-ureteric junction (PUJ) obstruction is defined as anatomical or functional obstruction to the flow of urine from the pelvis to the upper ureter resulting in symptoms or renal damage.⁴ PUJO is broadly classified as primary (congenital or intrinsic) and secondary (acquired or extrinsic). Primary PUJO is due to the dysfunctional smooth muscles and excess collagen deposition leading to hydronephrosis with clockwise rotation of renal pelvis and high ureteral origin.⁵⁻⁸ Secondary PUJO is due to the obstruction at the PUJ by compression and kinking by crossing vessels, fibrous band or other disease (retroperitoneal fibrosis, renal cyst, malignancy, pyelonephritis, xanthogranulomatosis).^{9,10}

UPJ obstruction has an incidence of 1 in 1000-1500 newborns and is the most common cause of antenatal hydronephrosis.¹ The other causes of antenatal hydronephrosis are transient obstruction, vesico-ureteric reflux (VUR), vesico-ureteric junction obstruction (VUJO), ureterocele, high bladder pressure, bladder outlet obstruction (Posterior Urethral Valve).¹¹ PUJO can either be unilateral or bilateral. Incidence of bilateral PUJO varies from 10-20%.

PUJO can present at any time from neonatal period to old age. Nowadays it is commonly diagnosed during prenatal ultrasonography. Once diagnosed antenatally, these children are followed up postnatally and treated. In few patients diagnosis is delayed or the children present late with pain abdomen, renal mass, UTI or uremia. In present study the most common presentation was pain abdomen. This finding was similar to other studies. ^{12,13} In 5 children, though antenatal scans had picked up hydronephrosis the children were not followed up postnatally. This is either because of the lack of counseling or lack of knowledge among the parents.

More number of males were affected compared to females. This corresponds to the male to female ratio in the literature which is 2:1.¹⁴ The left kidney was more affected compared to the right. This is similar to the finding in other studies.¹¹

All children initially underwent ultrasonography of the abdomen. On ultrasonography classical finding of UPJ obstruction is dilatation of renal pelvis and calyces with

non-visualization of ureter. Diuretic renogram is investigation of choice to identify the obstruction.

Table 5: Master chart of all 26 patients.

Age /sex	Clinical	USG APPD (mm)	Condition	DTPA/EC (L:R)	DMSA (L:R)	Post PCN DMSA (L:R)	Surgery
12/M	Pain abdomen	80	Left PUJO with NF left kidney	7:93	5:95	6.5:93.5	PCN followed by left nephrectomy
2/M	Abdominal distension	75	Left PUJO	2:98	1:99	31:69	PCN followed by left pyeloplasty
12/M	Pain abdomen, fever, vomiting	80	Right pyonephrosis with NF	1: 99		92: 08	PCN followed by right nephrectomy
2/M	Dysuria	20	Left PUJO with lower polar vessel	40:60			Left pyeloplasty
7/M	Pain abdomen	41	Left PUJO	36:64			Left pyeloplasty
6/M	Pain abdomen	25	Left PUJO with pelvic calculi				Left pyeloplasty with stone removal
11/M	Pain abdomen	44	Right PUJO	75:25			Right Pyeloplasty
7/M	Pain abdomen, vomiting	36	Left PUJO	35:65			Left pyeloplasty
9/F	Pain abdomen	20	Left PUJO with pelvic calculi				Left pyeloplasty with stone removal
5/M	Pain abdomen	31	Left PUJO	32:68			Left pyeloplasty
9/M	Pain abdomen	26	Right PUJO	39:61			Right pyeloplasty
14/M	Recurrent fever	56	Right PUJO with NF kidney		99:1		Lap right nephrectomy
5/M	Pain abdomen	48	Left PUJO with lower polar vessel	18:82			Left pyeloplasty
14/F	Pain abdomen	32	Left PUJO	40:60			Left pyeloplasty
1.6/M	Recurrent fever	28	Left PUJO	51:48			Left pyeloplasty
7/F	Pain abdomen	20	Left PUJO	40:60			Left pyeloplasty
8/M	Pain abdomen	54	Left NF kidney secondary PUJO	<5: 95			Left nephrectomy
1.8/M	Pain abdomen, vomiting	20	Right PUJO	51:49			Right pyeloplasty
13/M	Pain abdomen	42	Right PUJO lower polar vessel	84:16			Right pyeloplasty
7/F	Fever, pain abdomen	25	Right PUJO with pelvic calculi				Right pyeloplasty with stone removal
1.4/M	Abdominal distension	47	Left PUJO	35:65			Left pyeloplasty
1.6/M	Recurrent fever	30	Left PUJO; right grade III VUR	45:55			Left pyeloplasty, deflux injection for VUR
6/F	Recurrent fever	28	Right PUJO	58:42			Right Pyeloplasty
4/F	Pain abdomen, fever	30	Left PUJO	44:56			Left pyeloplasty
10/F	Pain abdomen	40	Left PUJO	51:49			Left pyeloplasty
2/M	Abdominal distension	35	Left PUJO	39:61			Left pyeloplasty

Criteria for intervention in children with PUJO include differential function < 40%, T $\frac{1}{2}$ greater than 20 minutes

on diuretic renogram, deteriorating renal function and ongoing parenchymal thinning. Other indication includes

palpable kidney, pain, hematuria, hypertension, recurrent UTI.

In present study three children had PUJO, secondary to lower pole vessel causing extrinsic compression on the PUJ. Extrinsic compression of PUJ by crossing blood vessel was first described by Wadsworth in 1983.15 The incidence of crossing vessels varies from 11% to 87%, though most of the studies have been on the adult population.¹⁶ In present study the incidence of crossing vessel was about 8%. Crossing vessels can be identified on angiography, endoluminal ultrasound, Doppler ultrasound, spiral CT, and MRI.¹⁷ In the present study all the three cases with crossing vessel were identified intraoperatively. The gold standard in the treatment of intrinsic and extrinsic PUJO is dismembered pyeloplasty.18

Two cases in present study presented with large palpable renal mass crossing the midline which is termed as Giant Hydronephrosis. The definition of Giant Hydronephrosis is an adult renal pelvis containing > 1 litre of urine or at least 1.5% of body weight. Halternatively, GH can also be defined as the presence of hydronephrosis occupying a hemi-abdomen, which meets, or extends beyond the midline and which extends at least five or six vertebral bodies in length. The ideal management for GH is nephrectomy. In present study one child required nephrectomy whereas in the other child, PCN was done followed by pyeloplasty.

Five children had a poorly functioning kidney (<10% split renal function) at the time of presentation. Initial PCN was done in three children. The best way to evaluate the capacity of the kidney to recover after obstruction is to give at trial of PCN.²² Kidney function improved in one child post PCN. The child was less than 2 years. It is well established that temporary PCN drainage in infected kidney of younger children leads to high renal recovery.²³⁻²⁶ The duration of PCN was 6 weeks in present study. Pode et al showed that 4 weeks is sufficient enough for renal recovery.²⁶ The two children in whom kidney improvement failed post PCN were of older age (12 years).

Three children in present study presented with pelvic calculi with hydronephrosis. It has been noted that the prevalence of lithiasis is more in patients with malformation of kidneys compared to general population.²⁷ The hypothesis is PUJO leads to delayed urine washout which in turn leads to crystal agglomeration and nucleation resulting in calculi.²⁸ On the contrary an impacted stone at PUJ is likely to produce local inflammation and edema sufficient to create circumstances similar to PUJO or it may provoke an inflammatory reaction severe enough to produce a stricture.^{29,30} Metabolic factors also play a role in calculi formation. No metabolic disorders were detected in the three children. All the three children underwent

pyeloplasty with removal of stone. No recurrence of calculi has been detected on follow up.

Overall out of the 26 children 4 children required nephrectomy. In the remaining 22 children Anderson Hynes dismembered pyeloplasty was done. The goal of surgery is to provide an unobstructed flow of urine across the PUJ. The success rate of Dismembered pyeloplasty is around 98 %.^{31,32} All the children post pyeloplasty had improvement in the symptoms and also function on diuretic renogram.

CONCLUSION

PUJ obstruction is common cause of urinary obstruction in children. Delay in diagnosis or presentation leads to increase chances of renal damage and loss. Delayed presentation may be complicated with pelvic calculi formation. Hence it is important to diagnose PUJ obstruction at the earliest and receive prompt treatment.

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Institutional Ethics Committee

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