

# Clinical profile and outcome of Pelviureteric junction obstruction (PUJO) in children less than 12 years.

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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. This may be unilateral or bilateral. Most of the hydronephrosis cases are due to Pyelo-ureteral junction obstruction (PUJO) which can be diagnosed antenatally. 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 infants and children (<12years) presenting with PUJO.

**Methods:** A retrospective observational study conducted by the department of Paediatric Surgery, SDM hospital Dharwad. All children with age less than 12 years with PUJO from March 2017 to March 2020 were included in the study (n=51). Initial evaluation was done with Ultrasound KUB (Kidney, Ureter, Bladder). A diuretic renogram/ EC (Ethylene cysteine) was done to confirm the diagnosis. The children were managed both conservatively and surgically (Pyeloplasty), depending on the presentation. The children were followed up post-surgery.

**Results:** Fifty one patients were included in the study. Mean age of presentation was 31±30 months. Most common presentation was pain abdomen which was seen in 9 patients (17.6%). 6 patients (11.7%) presented with Lump and 2(4%) presented with fever. Left kidney was commonly involved (66%). Males predominate (80%). The mean antero-posterior pelvic dimension (APPD) of involved kidney was 30mm with range from 12.5mm to 68mm. 4 (7.8%) children presented with poorly functioning kidney with split renal function less than 10%. PUJO was associated with Pelvic calculi, VUR, Horseshoe kidney, Mal-rotated kidney. Out of the 51 patients, 9 (17.6%) were kept in conservative management 42 (82.3%) underwent Pyeloplasty. Among 42, 1 underwent redopyeloplasty and 1 underwent nephrectomy. In rest of the operated patients (Post-pyeloplasty) diuretic renogram showed improved drainage with improved function.

**Conclusions:** PUJO is common cause of urinary obstruction in children. Earlier the diagnosis, better the outcome. 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:** Pelvi-ureteric junction obstruction, Pyeloplasty

## Introduction

Hydronephrosis (HDN) is a common condition. This may be unilateral or bilateral. Most HDN due to Pyelo-ureteral junction obstruction (PUJO) which can be diagnosed antenatally. Antenatal HDN represents a wide spectrum of urological conditions, which may or may not require intervention. Up to 80% these antenatal HDN would resolve spontaneously and would not require surgery<sup>[1]</sup>. As HDN due to PUJO is usually asymptomatic, it would be prudent to identify the other 20% asymptomatic HDN units,

usually SFU(Society of Fetal Urology) grades 3 and 4, which have significant obstruction at PUJO and need a definitive surgery. Most of the infants with HDN Society for fetal urology (SFU) Grade 1 & 2 have split renal function (SRF) >40% and conservative management is recommended as they tend to resolve with time and usually only require ultrasound surveillance. However, for the HDN of SFU Grade 3 & 4 due to PUJO, the management is controversial. This subgroup of renal units has been known to have loss of nephrons and irreversible functional loss

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if surgery is delayed. Other complications such as secondary hypertension, infection, renal stones may also arise. Once undergone an early pyeloplasty, an improvement in the renal dilatation and excretion pattern is expected in up to 95% of renal units<sup>[2]</sup>.

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<sup>1/2</sup> more than 20minutes on diuretic urogram, more than 10% deterioration in renal function (Split renal function). 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:

A retrospective observational study conducted by the department of Paediatric Surgery, SDM hospital Dharwad. All children with age less than 12 years with PUJO from March 2017 to March 2020 were included. Children with solitary kidney and syndromic patients were excluded from the study. The data of children with respect to sex, age and symptoms were tabulated. All children initially underwent an ultrasound of the Abdomen to evaluate the side and extent of hydronephrosis (SFU grading). The anteroposterior dimension of pelvis was documented. A diuretic Renogram was done to confirm the diagnosis of PUJO. Other routine blood investigations were done as per protocol before surgery. Based on presentation patients were managed both conservatively and surgically (Pyeloplasty). 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 4 to 6 weeks. Conservative group of patients were followed up with ultrasound (USG) at 3months and EC (Technetium-99m ethylene dicycysteine) scan at 1 year. Follow up continues till hydronephrosis disappears in ultrasound. If any deterioration in the renal function during follow up were considered for surgery. Post operatively children were followed up at 6 months to 1 year period. Initially USG KUB was done at 6 months post pyeloplasty. If USG shows decrease in grade of HDN then EC scan done at 1 year of age and if USG shows increase in grade

of HDN then EC scan done at same time to evaluate the drainage pattern across the PUJ. Nephrectomy was considered if the follow up Split renal function (SRF) falls below 10% along with palpable renal lump/Pyonephrosis/Hypertension.

#### Results

Out of the 51 patient who presented with PUJ obstruction in the study period, 44 (86%) children were within 6 years of age. The mean age at presentation was 30 months with range of 2 months to 12 years. Out of 51 patients 41 (80.4%) were males and 10 (19.6%) were females.(Table 1) Males constituted the majority of cases, with male to female ratio of 5:1. Unilateral left sided PUJ obstruction was seen in 34 children (66.6%), followed by bilateral obstruction in 2 (4%) children and right sided in 15 (29.4%) children (Table 2). Antenatal diagnosis was made in 18 (39.2%) patients. Out of 51 patients 17 (33%) were symptomatic. Out of the 51 patients, 9 (17.6%) were kept in conservative management and 42 (82.3%) underwent Pyeloplasty (Table 2).

**Table 1: Sex and age distribution of the patients.**

Variables			
AGE	<6 years	44	86.3%
	>6 years	07	13.7%
SEX	Male	41	80.4%
	Female	10	19.6%

**Table 2: Distribution of side and type of management.**

Side			Management	
Right	Left	Bilateral	Conservative	Surgical
15 (29.4)	34 (66.6%)	02 (39.2%)	09 (17.6%)	42 (82.3%)

**Table 3: Distribution of patients according to presentation.**

Presentation	Number(%)
Antenatally diagnosed	18 (39.2%)
Pain abdomen	09 (17.6%)
Abdominal lump	05 (9.8%)
Fever	02 (3.9%)
Vomiting	01 (1.9%)

The common clinical presentation included Pain abdomen, abdominal lump, fever and vomiting (Table 3).

**Table 4: Distribution other associated conditions**

Associated conditions	Number
Pelvic calculi	01
Vesicoureteric reflux(VUR)	01
Horseshoe kidney	01
Malrotated kidney	01
Fanconis anemia with CHD (congenital heart disease)	01

Other associated conditions found with PUJO were Pelvic calculi, VUR, Horseshoe kidney, malrotated kidney, Fanconis anemia.(Table 4)

### Outcome of pyeloplasty

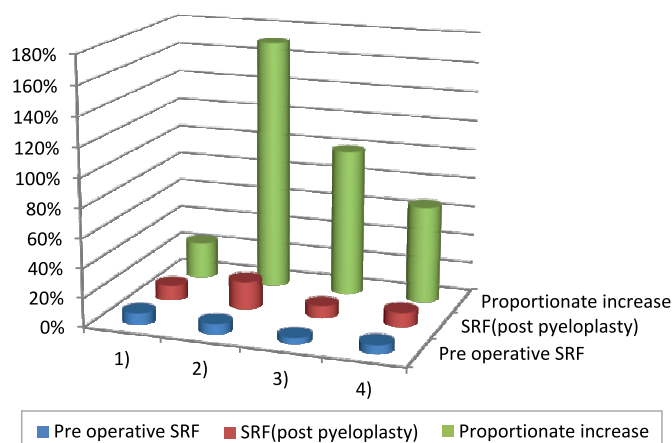
Among 42 patients, 2 patients showed deterioration in renal function. One patient presented with pain and palpable lump at previous operated site. USG showed gross HDN. Hence, PCN was placed under USG guidance. Nephrostogram was done, which showed non-clearance of contrast beyond pelvicalyceal system. Therefore patient underwent redopyeloplasty. Presently, the patient is asymptomatic and following up with EC scan and USG.

Another patient underwent twice redopyeloplasty and later underwent nephrectomy due to fall in SRF less than 10% along with development of palpable lump and pyonephrosis.

4 (7.8%) children presented with poorly functioning kidney with split renal function less than 10%, who underwent pyeloplasty. Among 4 patients, one initially underwent USG guided PCN (Percutaneous Nephrostomy) and later followed by pyeloplasty. On routine follow up with diuretic renogram showed recovery in renal function (Table 5). In rest of the operated patients (Post-pyeloplasty) diuretic renogram showed improved drainage with improved function.

**Table 5: outcome of poorly functioning kidneys (<10%) after Pyeloplasty.**

Patient	Pre operative SRF	SRF (post pyeloplasty)	Proportionate increase
1	08%	10%	25%
2	07%	19%	171%
3	04%	08%	100%
4	06%	10%	66.6%

**Figure 1: Graphical representation of outcome of pyeloplasty in poorly functioning kidney.**

### Discussion

PUJO is the most common congenital urinary obstruction. PUJ obstruction has an incidence of 1 in 1000-1500 newborns and is the most common cause of antenatal hydronephrosis<sup>[3]</sup>. PUJO can either be unilateral or bilateral. Incidence of bilateral PUJO varies from 10-20%. Nowadays it is commonly diagnosed during prenatal ultrasonograph. 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 which was similar to the studies done previously<sup>[4,5]</sup>. More number of males were affected compared to females in the ratio of 4:1, which is higher than the previous study in which ratio was 2:1<sup>[6]</sup>. The left kidney was more affected compared to the right. This is similar to the finding in other studies<sup>[7]</sup>.

Diuretic renogram remains a gold standard in the evaluation of these patients with loss of SRF in progressive renal scan being the criterion to determine the presence of significant PUJO. The gold standard in the treatment of intrinsic and extrinsic PUJO is dismembered pyeloplasty<sup>[8]</sup>.

Four children had a poorly functioning kidney (<10%) at the time of presentation. All patients were less than 6 years of age. All four showed improvement in renal function after surgery (Pyeloplasty). Only one had PCN before pyeloplasty to relieve symptoms due to gross hydronephrosis. In another similar study Vihma et al. also showed same finding<sup>[9]</sup>.

Considering putting of PCN in patient with poor functioning kidney to assess the functional status is best option. In one of the study, Gupta et al. performed PCN in all patients with split renal function of less than 10%, thus emphasizing that all such kidneys should not be removed without a trial of PCN<sup>[10]</sup>.

One patient in the study is associated with Fanconis anemia with Congenital Heart disease (CHD). Patient with Fanconi's anemia can phenocopy VACTERL syndrome<sup>[11]</sup>.

One patient in present study with age > 6years 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<sup>[12]</sup>. The hypothesis is PUJO leads to delayed urine washout which in turn leads to crystal agglomeration and nucleation resulting in calculi<sup>[13]</sup>. No metabolic abnormality detected in this patient. In one of the study Hollowell et al showed 14% incidence of VUR in PUJO<sup>[14]</sup>. In our study only one patient (2%) had associated VUR.

### Conclusion

This study highlights the need for a mandatory antenatal screening for early diagnosis and management of PUJ obstruction. 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. Pyeloplasty also gives good results in poorly functioning kidneys provided intervention is done in the earlier age group, and in most cases the sacrifice of such kidneys can be avoided. PCN should be considered in poor functioning kidney (<10%) to determine the outcome of pyeloplasty.

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