E-ISSN: 2616-3470
P-ISSN: 2616-3462

www.surgeryscience.com
2019; 3(1): 323-326
Received: 09-11-2018
Accepted: 13-12-2018

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Stentless pyeloplasty in childhood pelviureteric junction obstruction: 27 years experience

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DOI: https://doi.org/10.33545/surgery.2019.v3.i1f.54

Abstract
We are presenting our experience with childhood pelviureteric junction obstruction (PUJO) over 27 years, including the demographic characteristics, etiology, clinical features, and management. Ureteropelvic obstruction (PUJO) is the most common cause of congenital hydrocephalus diagnosed antenatally. It should be managed early after birth to preserve the renal function.

Keywords: ureteropelvic junction obstruction, hydronephrosis, pyeloplasty

1. Introduction
PUJO is the most common cause of antenatally diagnosed congenital hydrocephalus, with an estimated incidence of 1 in 1000 live births [1]. This was first described as a syndrome by Dietl way back in 1864 [2]. Early diagnosis and surgery in first year of life has been shown to preserve renal function best [3]. Despite this, it is very common to diagnose this condition at a later age in developing countries due to low detection rate of antenatal hydronephrosis and higher occurrence of infective conditions leading to acquired PUJO [4].
The obstruction of pelviureteral junction can be intrinsic or extrinsic. The intrinsic cause is much more common than the extrinsic one. Intrinsic PUJO can be caused by dysfunctional ureteric smooth muscles with excess collagen deposition and high insertion of ureters along with clockwise rotation of the renal pelvis [5-8]. The pathological changes found in cases of intrinsic pelviureteral junction obstruction include atrophy of myocytes, deposition of extracellular collagen and decrease in muscle innervations of ureteral pelvis [9]. This results in a variable length of the ureter which is aperistaltic and stenotic resulting in congenital hydronephrosis. It can be secondary to the lesions causing obstruction of the UPJ, which can be due to crossing vessel, malignancy, inflammation, fibrotic band etc. [10, 11].
This condition can present asymptomatically as antenatally diagnosed hydonephrosis or can present postnatally. Postnatally, it can present as abdominal lump, abdominal pain, hematuria, urinary tract infection or hypertension. It can also present with the associated anomalies with may be renal, like vesicoureteric reflux (VUR), malrotation, renal calculi, pelvic kidney, duplex kidney, horseshoe kidney, high insertion of ureters, segmental hypoplasia of ureters or non renal anomalies like associated anorectal malformations, cardiac or vertebral anomalies. The natural history of the disease is quite variable. But, usually the degree of renal pelvic dilatation is fairly predictive of the outcome of antenatally diagnosed hydronephrosis [12]. Anderson-Hynes dismembered pyeloplasty is a very innovative procedure with success rates of 90-100% and is considered as the "gold standard" for the management of the patients with PUJO [13].
We are sharing our experience of 35 years with the description of cases admitted to the department of pediatric surgery.

2. Aims and Objectives
To describe the epidemiology of cases presenting with PUJO at a tertiary care centre.

3. Materials and Methods
This was a retrospective study involving the study of case records from January, 1990 to December, 2016, presenting to the department of pediatric surgery with the diagnosis of PUJO

during this period. The age, sex, etiology, site of obstruction, presenting features, associated anomalies, investigations performed, surgical procedure performed and postoperative complications observed were recorded on a standard case record proforma.

4. Results

During the study period from January, 1990 to December, 2016, a total of 383 cases were admitted in the department of pediatric surgery. All antenatally diagnosed cases were followed after birth with ultrasound abdomen. If there was increase in size of renal pelvis, the patients were subjected to renal scan. All cases of neonatal obstructive hydronephrosis were operated and non-obstructive hydronephrosis were followed up.

Out of these 383 cases, 75 (19.5%) cases were in females and 308 (80.4%) cases were in males. The age wise distribution is as follows:

Table 1: 1-10 year age group

Age group	N (%)
Neonates	39 (10)
≥1 mo-<1 year	57 (14.8)
≥1-< 3 years	62 (16.2)
≥3-< 5 years	63 (16.4)
≥5-< 10 years	96 (25)
≥10 years	66 (17.2)

Right sided obstruction was found in 113 (29.5%) cases, left sided in 228 (59.5%) cases and in 42 (11%) cases, it was found to be bilateral PUJ obstruction. Associated renal anomalies were more common than the systemic ones. Majority (323, i.e., 84%) of the cases were idiopathic in etiology, the etiology of the rest of the cases is described in the table below:

Table 2: Associated anomalies of number

Associated anomalies	Number (%)
Giant pelvis	77 (20)
Malrotation of kidneys	12 (3.1)
Renal calculi	6 (1.5)
Horseshoe kidney	5 (1.3)
Vesicoureteric reflux	4 (1)
Multicystic dysplastic kidney	4 (1)
Pelvic kidney	4 (1)
Retrocaval ureter	1 (0.3)

Table 3: Anomalies of PUJ

Abnormality	N (%)
Aberrant renal vessel	51 (13.3)
Hypoplastic ureters	9 (2.3)
Thick walled pelvis	7 (1.8)
Papery thin pelvis	7 (1.8)
Kinking of UPJ	7 (1.8)
High insertion of ureters	1 (0.3)

Anorectal malformations were present in 5 cases, vertebral, cardiac anomalies in 6 each and hypospadias and undescended testis were present in 2 each cases respectively. In one of our patients, it was associated with pyloro-antral diaphragm. The most common mode of presentation was abdominal lump present in 119 patients at presentation. Antenatal diagnosis was made in 20 cases, 39 patients presented with abdominal discomfort, 10 patients presented with urinary tract infection, 4 with hematuria and 4 patients had hypertension at diagnosis.

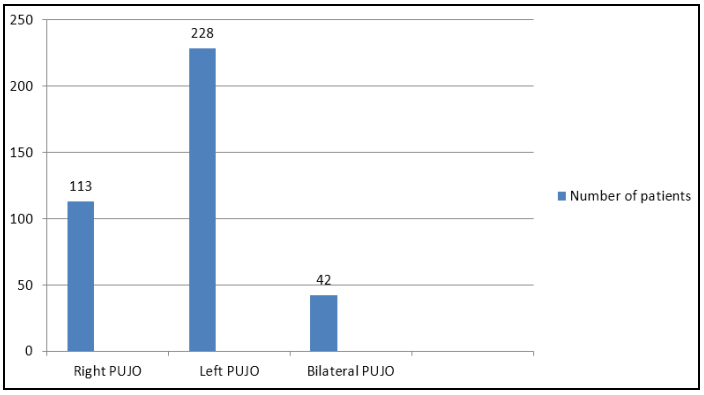

Fig 1: Laterality of cases of PUJO

Investigations of these cases included renal function tests, routine hemogram and coagulation parameters in all the patients. We did ultrasonography of abdomen in all our patients. If hydoureter was found, then micturating cystourethrogram was performed in these cases. Renal (DTPA) scan was done in all cases to see the functional status of the kidneys. We performed Anderson-Hynes pyeloplasty in all the cases. We are not performing nephrectomies for the last 20 years. So far, we have done only 2 nephrectomiesin view of giant hydronephrosis. All cases were operated using left extraperitoneal lumbar incisions (fig. 2, 3), the peritoneum was reflected and the kidney mobilised. The anatomy of ureteropelvic junction, aberrant vessels, severity of hydrocephalus, peripelvic adhesions, pelvic thickness, appearance of urine, length of proximal and distal hypoplastic ureters were noted. Turbid urine was found in 3 cases. The length of hypoplastic segment of ureters was found to be variable from 1 to 3 cm. Anomalous PUJ was identified and redundant pelvis along with 2-2.5 cm of hypoplastic ureters was excised and anastomotic stoma is created for dependent drainage. In giant hydronephrosis, kidney was decompressed through pelvic incision. Kidney anomalies were recorded. All cases were subjected to stentless Anderson Hynes Pyeloplasty with drainage (fig. 4). Postoperatively, in majority of the patients, drain is removed after 3-5 days, whereas in larger hydronephrotic kidney, drain was continued for 10-15 days. Mild stitch abscess developed in 10 cases postoperatively, which was managed with appropriate antibiotics. Incisional hernia developed in 3 cases. Postoperative stricture at the anastomotic site developed in 3 cases, out of which, stent was placed in one case with a solitary kidney with ureteric obstruction. Redo pyeloplasty was done in rest two cases. Postoperative urinoma was formed in 5 cases. Follow up was done with renal scan and ultrasound after 3-6 months. Residual hydronephrosis can persist in cases presenting late.

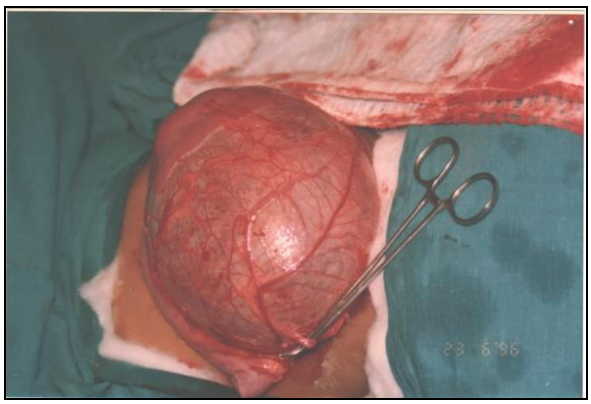

Fig 2: preoperative photograph of giant hydronephrosis

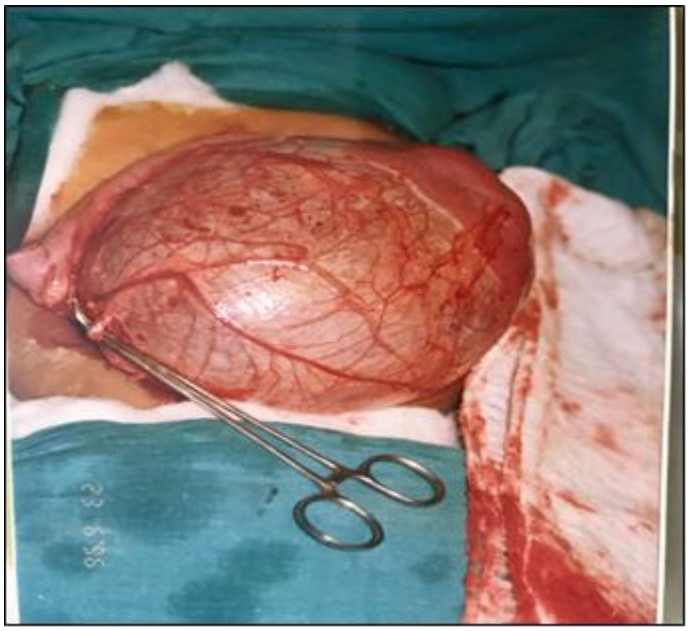

Fig 3: intraoperative photograph showing hydronephrotic kidney in a 1 year old male infant.

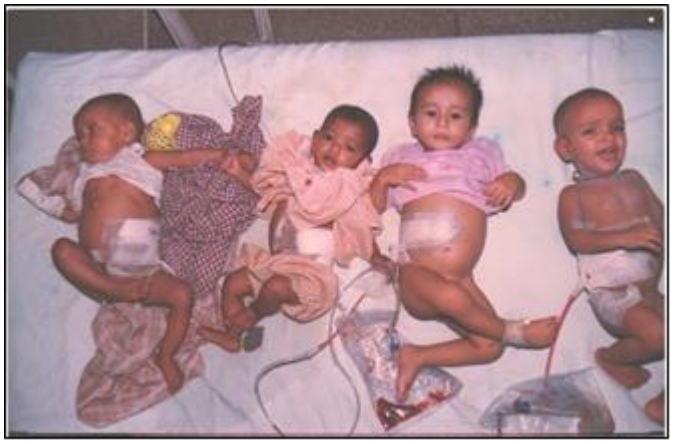

Fig 4: Patients with retroperitoneal drain in situ after surgery

5. Discussion

Our study shows higher occurrence of PUJO on left side and more in males as compared to females. This is in accordance with most of the studies so far [14, 15]. The age of presentation in neonatal age is much lesser than the ones diagnosed at a later age. Also most of our cases presented with lump on abdominal examination. This is because the catchment area of our hospital is rural areas with poor antenatal ultrasound facilities. The etiology in most of the cases was idiopathic. The number of patients with aberrant vessels has been found to be lesser than around 30% described in this population in other studies [16]. Also, the number of patients with giant pelvis is higher than the other studies [5, 14]. This may be due to delayed presentation of the cases, with lesser cases detected at an earlier stage (fig 2). The renal function was preserved despite hugely dilated pelvis. This could be because of high compliance of the pelvis which takes all the burden, thus sparing the renal parenchyma. Malrotation of kidneys was found to be the most commonly associated anomaly in our patients, with the other anomalies being horseshoe kidney and pelvic kidneys. These are different from other studies in this area [17, 18]. The frequency of renal stones correlates with the other studies of PUJO [17, 18]. The etiology of renal stones can be metabolic or it can be due to anatomical abnormalities [19]. The exact pathogenesis remains unclear. Hypertension was present in 2 of our patients, while this was found in 11 out of 227 patients in study by de Waard *et al.* [20]. The hypertension can be relieved by surgery and hence should be looked for in all cases of PUJO.

The diagnosis of the cases with PUJ obstruction is based on ultrasonography of the abdomen and DTPA renal scan, which helps in determining the residual renal function in these patients. The role of voiding cystourethrogram (VCUG) is not very significant in these cases. This is due to the fact that the occurrence of vesicoureteric reflux (VUR) is very low in these patients. Hubertus *et al.* in a multicentric study opined against the routine use of VCUG [21], as the incidence of VUR was 7.4% at their centre, which was found to be still lower at 1% in our centre. We advise upon the careful evaluation of distal ureters during ultrasonography of the abdomen. In case of dilatation of the ureters, VCUG can be done. This helps avoid missing of VUR in these cases. There was no recurrence in our cases, as all the surgeries were performed by an expert.

The management of cases begins in the antenatal period, with antenatally detected hydronephrosis. A renal pelvic diameter more than 4 mm between 18-22 weeks of gestation and >10 mm in third trimester constitutes hydronephrosis. They can be followed every 3-4 weekly with serial ultrasounds to detect progression of hydronephrosis and oligohydramnios. Surgical intervention in the antenatal period is rarely performed and is not associated with significant survival outcomes [22]. Postnatal ultrasound can be repeated on 2nd or 3rd postnatal day. The management of asymptomatic children can be operative or nonoperative [23]. Children with mild or moderate hydronephrosis can be followed up with serial ultrasounds at 4-6 month intervals and yearly thereafter for decrease in the renal size. In cases with increasing degree of hydronephrosis, renal scan is done and surgery performed with poor renal function and in all cases of obstructive hydronephrosis. In cases of severe hydronephrosis, with renal pelvic diameter $\geq$15 mm, renal scan is done at 6-8 weeks, as the interpretation of renal function is unreliable in the neonatal age. If the renal function is <40% compared to the contralateral kidney, these can be taken up for surgery, else they can be followed up with serial ultrasounds [23].

All symptomatic patients with pyelonephritis, urinary stones or cases with secondary causes of UPJO, and those with decreased renal function in renal scan are managed surgically.

The standard surgical management of cases is done with Anderson-Hynes pyeloplasty. The success rates are 90-100% [12]. We performed this in all our patients. Drainage techniques have been added as modification to this procedure over the years, including stenting and nephrostomy tube placement. We are performing stentless surgeries, so the complications, like stent migration and urinary tract infection or nephrostomy tube block are not there in our study.

Laparoscopic pyeloplasty is also being performed now-a-days. The success rates are closer to that of open pyeloplasty. In a population based study by Knoedler *et al.*, the outcomes are found to be almost similar in open and laparoscopic pyelolasty [24]. Though, a clear advantage of lesser perioperative complications has been demonstrated only in adults [25]. Robotic assisted techniques are also being employed recently and the evidence is accumulating. Still, a clear consensus in favor of such techniques is not yet available, with the cost of these procedures being a major deterrent.

6. Conclusion

The results in stentless Anderson-Hynes Pyeloplasty (AHP) with drainage are similar to those with stent. Hence we recommend

stentless AHP, as it avoids the exposure of the child to a second procedure for removal of the stent per urethra, which is done under general anaesthesia and affects the virginity of the urethra.

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