

Pelviureteric Junction Obstruction Associated with Vesico-ureteric reflux and Vesico-ureteric Junction Obstruction: Challenges and Management Protocols

N. Sharma, M. Bajpai, S. S. Panda

Department of Paediatric Surgery, All India Institute of Medical Sciences, New Delhi-110029, India

Abstract. Objective: To assess the incidence and outcome of cases of pelviureteric junction obstruction (PUJO) with associated vesicoureteric reflux (VUR) and/or vesicoureteric junction obstruction (VUJO). **Methods:** This retrospective analysis included cases of PUJO with associated VUJ anomalies managed between January 2000 and April 2013. All the cases were operated first for PUJO by Anderson-Hynes pyeloplasty via lumbotomy approach. Follow up was done by ultrasound and renal dynamic scan (RDS) at 6 weeks after surgery. Second surgery done was Cohen's ureteric reimplantation after 6 weeks or later. They were subsequently followed by using standard follow up protocol. **Results:** Out of a total of 29 cases, four cases had an associated VUJO while remaining 25 cases had an associated VUR. VUJO cases presented with poorly functioning kidneys with average split renal function (SRF) of 7%. These cases underwent PCN as an initial management. Follow up RDS were suggestive of VUJO in two cases and were then confirmed with an antegrade dye study. Two cases were found to have VUJO on intraoperative saline test. These cases subsequently underwent Cohen's ureteric reimplantation. Mean SRF in patients with VUR was 22%. Mean number of surgeries per patient was 2. Two cases with consequent VUR didn't show improvement in SRF in last follow up. Rest of the patients showed a significant improvement in SRF after surgery. **Conclusion:** Concurrent presence of PUJO and VUJ anomalies is a diagnostic challenge. These cases can be managed nicely if a protocol based approach is followed.

Keywords: Pelviureteric junction obstruction, Vesicoureteric reflux, Vesicoureteric junction obstruction, Vesicoureteric junction anomalies

Received: 8 January 2014 / **Accepted:** 18 January 2014

Introduction

Congenital pelviureteric junction obstruction (PUJO) is one of the most common causes of hydronephrosis in children. A child may present to a clinician with complaint of dull aching pain on the affected side or with an obvious palpable lump. This condition can be diagnosed by ultrasonography and confirmed by a renal dynamic scan. The management protocols are based on the presence of symptoms. On the contrary when the condition is asymptomatic, the split renal

function of the affected kidney determines the line of further management. There has been a lot of debate regarding when to operate and when to follow up these patients. Accepted consensus is to operate the obstructed side when the split renal function is < 35 % with an obstructed pattern of clearance on renal dynamic scan.^[1] PUJO is associated with a number of conditions and the management of these conditions largely varies. Association of vesicoureteric junction anomalies is not very commonly noticed. Vesicoureteric reflux (VUR) is among the most common vesicoureteric junction anomaly associated with PUJO and infact PUJO is considered secondary to the associated VUR. Few uncommon cases are associated with a vesicoureteric junction obstruction (VUJO). In this article we have tried to address these issues.

Objective

To find out the incidence of associated VUR and/ or VUJO in operated cases of PUJO and assess their outcome.

Copyright and reprint request:

Dr. M. Bajpai, MS, MCh, PhD, FACS, FRCS, FAMS (India), DNB, Fulbright Scholar (USA), Commonwealth Fellow (UK), Raja Rammana Fellow (India)
Professor, Department of Paediatric Surgery, All India Institute of Medical Sciences, New Delhi 110029, India., Web: www.paediatric-urologyonline.org; E-mail: bajpai2@hotmail.com; Ph: +91-11-26593555; Mob: +91-981-802-5584

Materials and Methods

This was a retrospective analysis of cases of PUJ obstructions managed in Wednesday Paediatric Urology Clinic (WPUC), AIIMS, between January 2000 and April 2013 who presented with an associated vesicoureteric junction anomalies. Case sheets of all these patients were analyzed and data regarding clinical presentation, age at presentation, management offered and investigations done were obtained and analyzed. Those with incomplete data or those who were lost to follow up were excluded. Those with neurogenic bladder, urethral valves, ureterocele, bladder diverticula and duplex system were also excluded. VUR was defined as the presence of reflux documented in micturating cystourethrogram (MCU). PUJO was defined as an obstructive pattern of drainage in renal dynamic scan using LLEC. VUJO was defined as an evident vesicoureteric junction obstruction on antegrade dye study (Fig. 1), intraoperative diagnosis based on saline test or an evident hydroureteronephrosis in a well-tempered renogram with a bladder catheter in situ showing obstructive pattern in vesicoureteric junction.

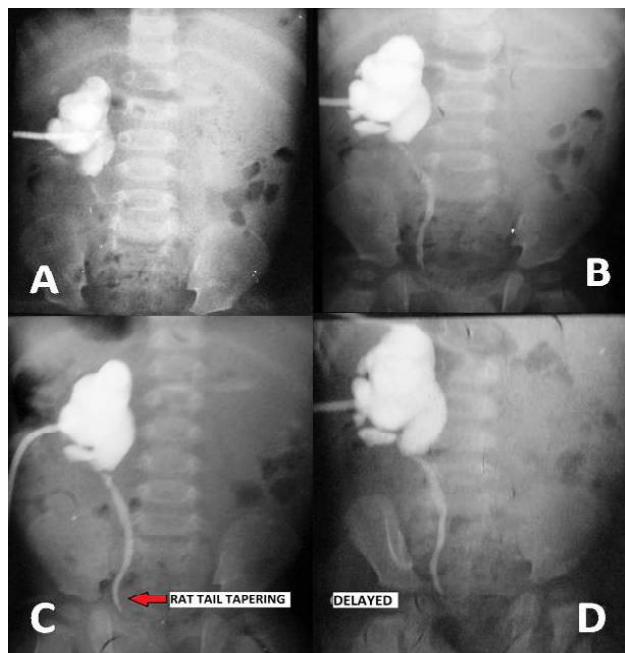


Fig. 1. Antegrade dye study in operated PUJO patient showing rat tail tapering suggestive of associated VUJ obstruction (A): Initial (B) After 15 sec (C) After 2 min (D) After 45 min

All the cases were operated first for a PUJO using Anderson-Hynes pyeloplasty via lumbotomy approach. Follow up was done using an ultrasound and a renal dynamic scan at 6 weeks after surgery using follow up protocol as shown in Fig. 2.

Second surgery done was Cohen's ureteric reimplantation after 6 weeks or later from pyeloplasty. They were then

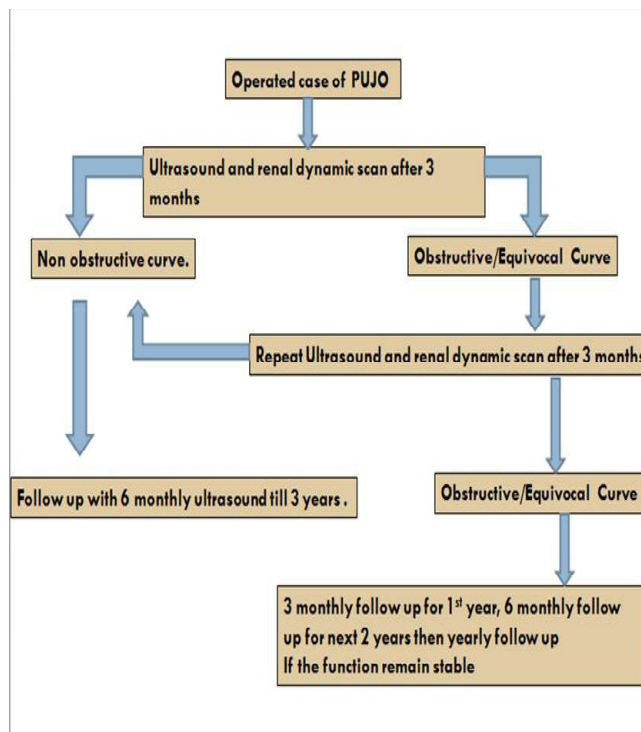


Fig. 2. Follow up protocol after pyeloplasty

subsequently followed by doing an ultrasound scan at 3monthly interval, MCU and RDS after 6 months of second surgery and subsequently by ultrasounds. Success was defined as no sign of reflux on MCU, resolution or significant decrease in hydroureteronephrosis, complete clinical improvement, resolution of preoperative symptoms, improvement in drainage curve on diuretic renogram, regression in pelvic diameter on serial ultrasonography, and improvement or maintenance of renal function on the renal dynamic scan using ^{99m}Tc -LLEC scan. The result obtained was analyzed.

Results

There were a total of 293 cases of pelviureteric junction obstruction managed in the clinic during the same period. A total of 29 cases that had either VUJO or VUR formed the study group. The mean age at presentation was 37.26 months (range: 3-148 months). There were 3 females and 26 males with the male to female ratio of 9:1. Four cases had an associated VUJO while remaining 25 cases had an associated VUR. There were 7 cases with right sided pathology, 19 with left sided pathology while 3 had bilateral pathology. VUJO cases presented with poorly functioning kidneys with average split renal function of 7% (range 0-10%). These cases underwent percutaneous nephrostomy (PCN) as an initial management. Follow up renal dynamic scans were suggestive of VUJO in two cases and were then confirmed with an

antegrade dye study. Two cases were found to have VUJO on intraoperative saline test. These cases subsequently underwent Cohen's ureteric reimplantation. Mean post-operative follow up was 24 months (range 8-48 months). Of the 25 cases with VUR, 20 cases had higher grades of VUR (grade 4 and 5) while 3 cases had grade 3 VUR and 2 cases had lower grades of documented VUR. Those with VUR below grade 3 were followed for VUR and not operated. Mean Split renal function in patients with VUR was 22% (Range 13- 38%). Mean number of surgeries per patient was 2 (range 1-4). Two cases with consequent VUR didn't show improvement in the split renal function in last follow up though they had a non-obstructive clearance and are in follow up. Rest of the patients showed a significant improvement in the split renal function after surgery.

Discussion

The coexistence of obstructions at the proximal and the distal ends of the ureter is rare. There are very few published series in English literature showing such coexistence. Most often the distal obstruction is masked either due to the poor function of the concerned kidney or due to the focus of interest on the proximal obstruction. Most of the cases who presented to us with dual obstruction had poorly functioning kidneys and VUJ obstruction was picked up intraoperatively in two while others were diagnosed in follow up imaging. Pak *et al* first reported a case of coexistence of PUJ obstruction with VUJ obstruction in a horseshoe kidney.^[2] Pfister and Hendren reported 150 primary mega-ureters (predominantly in children) and described five patients (3%) who had coexisting PUJ obstruction.^[3] Pitts reviewed 80 patients (about one half were children) with primary megaureter and noted that 20 (25%) had simultaneous PUJ obstruction.^[4] Subsequently McGrath *et al* reported 14 cases of double obstruction after reviewing about 225 cases of PUJ obstruction and 115 cases of VUJ obstruction diagnosed over an 11 year period.^[5] The ages at presentation ranged from birth (prenatal diagnosis) to 11 years. In 11 cases the left ureter was affected; in 2 of these it was draining the child's only kidney. The diagnosis was made preoperatively in only 3 patients. In 3 the second obstruction was initially suspected at surgery; in the other 8, the second obstruction was diagnosed during follow-up radiologic studies performed 2 weeks to 2.5 years after surgery.

In most of the cases the concerned side with dual obstruction has very poor function. Thus the diagnosis remains obscure and most of the time PCN is done to see the pattern of improvement. After 6-8 weeks the renal dynamic scan is done to see the improvement in pattern. Most of the time, as there is an associated PUJ obstruction, we can't get an idea about the lower obstruction. Most of these cases are thus diagnosed in the follow up investigations. A fraction of cases are diagnosed intraoperatively on saline injection test or while

inserting a DJ stent. Many surgeons still contest in the issue and argue that both the obstruction can be relieved at a time while others propose a staged management with upper obstruction to be addressed first. As of now there is no established series comparing the results of these two approaches so as a dictum proximal obstruction is relieved first. The second surgery which is the reimplantation of the obstructed ureter is done after 6-8 weeks of the first surgery, till then the child is maintained on a nephrostomy. After reimplantation the nephrostomy is clamped and subsequently removed.

The presence of PUJO with an associated VUR is rather commonly noted. There is contest regarding the pathogenesis of the obstruction secondary to VUR. The obstruction is mostly congenital or primary; however, it can occur secondary to severe VUR.^[6] The coexistence of VUR and PUJO has been reported.^[7] Approximately 10% of patients with PUJO are found to have concomitant VUR coincidentally, whereas less than 1% of patients with the diagnosis of VUR have simultaneous PUJO.^[8] The etiology of the coexistence is not well established.^[9] However, ureteral kinking and inflammation resulting from reflux, the presence of a single developmental abnormality, or a random event have been proposed as possible etiologies.^[9] Management of these patients can be complicated, and no straightforward approach has been described in the literature. The traditional management of children with VUR and PUJO is pyeloplasty as the initial step, and if the high-grade reflux remains symptomatic after pyeloplasty, surgical correction of the reflux is the next step.^[10] It was postulated that the nearby obstruction to the renal parenchyma might cause more renal damage, so it should be managed sooner rather than later. Moreover, in a few paediatric urology centers, this concept has ultimately been changed by surgical correction of reflux (ureteral reimplantation) initially, followed by pyeloplasty in persistent PUJO. Kim *et al* also favoured of initial pyeloplasty because they hypothesized that edema at the ureterovesical junction after ureteral reimplantation, and not endoscopic injection, leads to acute worsening of the PUJ obstruction.^[11] Peters *et al*^[12] and Onen *et al*^[13] suggested that because conservative management can be applied to low-grade hydronephrosis caused by PUJO, all patients with PUJO, even with coexisting VUR, should not undergo pyeloplasty as the initial intervention. Bomalaski *et al*^[14] reported a pyeloplasty rate of 75% in children with simultaneous VUR and PUJO. Kajbafzadeh *et al*^[15] approached patients with concomitant VUR and PUJO by endoscopic injection of Deflux using the HABIT technique. They included 2810 cases of VUR and found 143 cases having a concomitant PUJO. Of the 76 (89 ureters) included in the study they showed a self-resolution of PUJO in 76.9% per patient (74.1% per ureter) after the first injection and 84.6% per patient (80.6% per ureter) after the

second. Only 3 patients (11.5%) underwent ureteral reimplantation, and 3 (11.5%) required pyeloplasty. This recent publication has started the debate on the management of this concomitant lesion with the management of VUR first.

We, however, believe in the traditional management option and feel that the upper obstruction should be addressed first and then VUR should be subsequently addressed after 6-8 weeks of the first surgery. Follow up of these cases should be done regularly. The follow up protocol should be as shown in figure 2. An MCU should be repeated after 3 months of reimplantation and repeated after 6 months. If MCU shows no reflux it may not be repeated as such.

Conclusion

Concurrent presence of PUJO and VUJ anomalies is a diagnostic challenge. Presence of poorly functioning kidney on the affected side makes diagnosis further difficult. These cases can be managed nicely if a protocol based approach is followed.

References

1. Chandrasekharam VV, Srinivas M, Bal CS, Gupta AK, Agarwala S, Mitra DK, *et al.* Functional outcome after pyeloplasty for unilateral symptomatic hydronephrosis. *Pediatr Surg Int* 2001;17:524-7.
2. Pak K, Konishi T. Horseshoe kidney in an infant associated with ureteropelvic junction and ureterovesical junction obstructions. *Hinyokika Kyo* 1985;31:457-61.
3. Pfister PC, Hendren WH. Primary megaureter in children and adults: clinical and pathophysiologic features of 150 ureters. *Urology* 1978;12:160-76.
4. Pitts WR, Muecke EL. Congenital megaloureter: a review of 80 patients. *J Urol* 1974;111:468-73.
5. McGrath MA, Estroff J, Lebowitz. RL. The Coexistence of Obstruction at the Ureteropelvic and Ureterovesical Junctions. *AJR* 1987;149:403-6.
6. Carr MC, Casale P. Anomalies and surgery of the ureter in children. In: Wein A, Kavoussi L, Novick A, *et al.*, eds. *Campbell-Walsh Urology*. 9th ed. Philadelphia, PA: Saunders; 2007:3212-3235.
7. Karnak I, Woo LL, Shah SN, Sirajuddin A, Kay R, Ross JH. Prenatally detected ureteropelvic junction obstruction: clinical features and associated urologic abnormalities. *Pediatr Surg Int* 2008;24:395-402.
8. Byrne LN, Koyle MA. Ureteropelvic junction obstruction unmasked after correction of vesicoureteral reflux using endoscopic treatment with Deflux. *J Pediatr Urol* 2006;2:205-9.
9. Stauss J, Connolly LP, Connolly SA, Zurakowski D, Treves ST, Peters CA. Dynamic renal scintigraphy in children with vesicoureteral reflux and suspected coexisting ureteropelvic junction obstruction. *J Urol* 2003;170:1966-70.
10. Maizels M, Smith CK, Firlit CF. The management of children with vesicoureteral reflux and ureteropelvic junction obstruction. *J Urol* 1984;131:722-7.
11. Kim YS, Do SH, Hong CH, Kim MJ, Choi SK, Han SW. Does every patient with ureteropelvic junction obstruction need voiding cystography? *J Urol* 2001;165:2305-7.
12. Peters CA. Urinary tract obstruction in children. *J Urol* 1995;154:1874-83.
13. Onen A, Jayanthi V, Koff S. Long-term followup of prenatally detected severe bilateral newborn hydronephrosis initially managed nonoperatively. *J Urol* 2002;168:1118-20.
14. Bomalaski MD, Hirschl RB, Bloom DA. Vesicoureteral reflux and ureteropelvic junction obstruction: association, treatment options and outcome. *J Urol* 1997;157:969-74.
15. Kajbafzadeh AM, Tourchi A, Ebadi M. The Outcome of Initial endoscopic Treatment in the Management of Concomitant Vesicoureteral Reflux and Ureteropelvic Junction Obstruction. *Urology* 2013;81:1040-6.