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Original Research Article

Clinical profile of infantile pelviureteric junction obstruction

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ABSTRACT

Background: Pelviureteric junction obstruction commonly present with antenatal hydronephrosis. The aim was to report and analyze the clinical spectrum of infantile pelviureteric junction obstruction.
Methods: Retrospective descriptive study of infants operated for pelviureteric junction obstruction (PUJO) during 2013- 2016. Clinical presentations, management and follow up were analyzed.
Results: There were 44 infants involving 48 PUJO renal units of who were nine girls. The sex distribution was 1:3.8. Mean age of the presentation was three months. PUJO was bilateral in four patients. Two patients had unilateral PUJO of lower moiety of duplex system. Median of anteroposterior diameter (APD) and differential function of kidney on EC was 2.5cms and 28% respectively. Three patients had poorly functioning kidneys (PFK) (<10%).
Conclusions: Good antenatal counselling and appropriate postnatal evaluation detects PUJO early. Early infantile pyeloplasty is safe and effective. Routine micturating cystourethrography is debatable in pelviureteric junction obstruction.

Keywords: Antenatal hydronephrosis, Infants, Pelviureteric junction obstruction, Pyeloplasty

INTRODUCTION

Pelviureteric junction obstruction commonly present with antenatal hydronephrosis.[1] Congenital PUJO etiology are alterations in muscle/ collagen elements, mucosal polyp /valve, and aberrant vessel at the pelviureteric junction.[1] Indications of pyeloplasty are palpable kidney, obstruction with differential function <40%, and deteriorating of renal function.[1] We are reporting varied presentations and successful surgical management of infantile PUJO.

METHODS

This is a retrospective and descriptive observational analysis of surgically treated infants with PUJO admitted during 2013-2016. Forty-four infants out of seventy-two children surgically treated for PUJO were reviewed with case sheets, electronic media with parents regarding demography, clinical presentations, management and follow-up (Table 1). As per institutional protocol all antenatally diagnosed hydronephrosis underwent routine postnatal U/S @ 1st week and end of 4th week. This was followed by EC and MCU in all at age of one month. Additional investigations included IVU (intravenous urogram) and RGU (retrograde ureterography) in specific situations.

RESULTS

Thirty infants had come within three months of life. Boys (35/44) were more commonly affected. Clinical presentations were varied and complex; AHN (40) (Figure 1), palpable kidney (8), urinary tract infections (UTI) (6), acute renal failure (ARF) (1), fetal giant intra-abdominal cyst (1), nephrolithiasis (1), failure to thrive (1). Downs syndrome (1), bilateral (B/L) cataract (1) and craniosynostosis (1) were associated anomalies. Incidence of bilateral PUJO was 10% (4/40). Median of anteroposterior kidney diameter was 2.5cms. MCU, IVP, retrograde ureterography (RGP) revealed in addition B/L duplex kidney with lower moiety (LMO) obstruction (2),

vesicoureteric obstruction (VUO)/ (2), reflux (VUR) (2), posterior urethral valve PUV (1) and Prostatic utricle (PU) (1). Median of differential function for pyeloplasty was 28%. Three infants had poorly functioning kidneys <10% (Fig 2) and underwent percutaneous nephrostomy (PCN) for drainage purpose. PCN had helped in assessment of the kidney function and planning the surgical procedure. EC scan function assessment showed hyper functioning in four. These were giant palpable kidney in (1), solitary kidney (1) and contralateral dysplastic kidney in (2) UTI was initial presentation in (6), three had turbid urine on table, which grew organism on culture suggestive of pyonephrosis (2). Six infants developed postoperative UTI, three of whom were stented and responded to early stent removal. Open dismembered pyeloplasty was successful in all except two who required redo pyeloplasty due to anastomotic stricture (2) and pelvic calculus (1). Double J stent was used in all patients with solitary kidney, pyonephrosis, redo pyeloplasty, and as per surgeon's choice in others. Four had stent migration in lower 1/3 (2) and upper ureter 1/3(1), urethral migration (1) and retrieved with basket under fluoroscopy (2) and on out patient with artery forceps for urethral catheter. Other infant also underwent redo pyeloplasty as well. Fifteen were Stent less pyeloplasty; two of who had transient, self-resolving urinary leak and other were discharged on second post-operative day.

Table 1: Master chart of all 44 patients.

Age	Sex	Clinical presentation	Ultrasound APD	EC scan	MCU	Other inv	Operative /early comp	Disease	Follow-up
1	M	AHN,B/L L palpable	R 1.2 L cross midline	R39 L61	N	S C 2-.6	L PCN B/L loop ureterostomyS+	B/L VUJO and PUJO L giant/ARF R UTI	Recurrent UTI prolonged chemoprophylaxis
1.5	F	AHN	L6.2, R1.1	L20	N		S- (stent less)	LPUJO	
1	M	AHN B/L R palpable UTI	R 1.2-2 L2.2 - 2.5	R37 L63	N	S C.6	B/L,S+,S-	B/L PUJO R > Left	
1.5	M	AHN	R2.2	R35	N		S- leak	R PUJO	
1	M	AHN B/L UTI	L2.22.5R1. 2	L26	N	Anemia (A)	S-	L PUJO UTI	
2	M	AHN	L1.9-2.6	L31	N	A	Ontable turbid urineS+	L PUJO pyonephrosis	Asymptomatic UTI
2	M	AHN	R3.2 3.4	R30	N		S-	RPUJO	
1	F	AHN	L1.5	L25	N		Colonic injury/turbid urine fungal sepsis S-	L PUJO pyonephrosis	
11	M	AHN UTI	R .9-1.2	RUM3 0 LM 12	N	IVPB/L duplex	Pyeloureterostomy turbid urine bifid ureter incomplete S+	R lower moiety PUJO B/L duplex pyonephrosis	
5	M	AHN	R 4.2-3.8	R UM 22 LM 6	N		S+	R Lower Moiety PUJO B/L Duplex	
3	M	AHN	L2-2.8	L35	N	IVPLPUJ O	S+	L PUJO	
7	M	AHN UTI	L5-4.4	L28	N		S-	LPUJO	
3	M	AHNB/L L palpable	L2.5 3.5,5	L28	N	IVPLPUJ O	S-anastomotic stricture/RGP+D J stent? Migration PCN slipped Redopyeloplasty S+	LPUJO	
4	M	AHN	R1.5	R25	N		S+	RPUJO	
2	M	AHN	R2.1	R18	N		S+	R PUJO	
1.5	M	AHN B/L	R7-1.7 L1.5	R35	N		PUJO mid uretericstricture	B/L PUJO Down syndrome ASD	
1.5	M	AHN	R2.2	R24	N		S+	RPUJO.	
7	F	AHNB/L	L18	L0-15	N		S-	L PUJO	

		palpable							
2	M	AHN B/L	R1.9 L.9	R38	N	S C.5	S-	R PUJO	
2	F	Fetalintra-abdominal cyst	L5.8	L-28	N		S+	LPUJO giant	
3	M	Pyonephrosis,	L13	L11.22		S C.2	S+	L pyonephrosis/ giant HN	S migration removal c arm guidance
8	F	UTI R palpable	R3.6	solitary	B/L VUR II	ASD S C.43	S +	R solitary PUJO B/L VUR Failure to thrive	chemo
3	M	AHN	L1.8	L 28	N	HPR PUJ	S+, mid ureter dilated?reflux, HPE- PUJO obstruction	LPUJO	
3.5	M	AHN	L1.5-1.8	L32	N		S-	L PUJO	
5	M	AHN	L2.2	L31	N	UTI	S-	L PUJO	
4	M	AHN	L1.9	L37	N			L PUJO	
8	M	Pain	L3 calculus	L-49,R-51	N	IVP-calculus		L PUJO calculi	
3	M	AHN	L2.5	L 23	N	S C.2	S+ UTI	LPUJO	
3.5	F	AHN	L1.5- 1.8 R9	L18	N		S-	L PUJO	
3	M	AHN	R1.0-2.0-2.8	R33	N		S+	RPUJO	
3	M	AHN	L13	L36	N		S+	LPUJO	
2.5	F	AHN B/L	L2.2 R2.5	L2,5 R2.2	N		S+	B/LPUJO	8mon second pyelo
4	M	AHN	L3.2	L40	N		S+	LPUJO	
4	F	AHN	R1.6,2.4,3.	R50-40	N		S+	RPUJO	Stent urethral migration UTI
1.5	M	AHN	R1.5 L6.3	L21	N		S+	LPUJO	
5	M	AHN palpable	L 6.5 {3.7-4.6 post PCN}	L-7	N		S+ PCN	LPUJO Giant+HT	
2	M	AHN	L2.5		N		S+	LPUJO	
2	M	AHN B/L craniosynostosis	L1.2 R1.7-2.9	L17	N		R PCN for URI later pyeloplasty S+	RPUJO metopic suture CHD	
7	M	AHN B/L palpable	L1.4 R 2.6	L94/R6	N		S+UTI/ ureter dilatation	LPUJO B/L cataract	stent curling in lower ureter removed under C arm
2.5	M	AHN	L2.2 R2.5	R27	N		S+	R PUJO	
2	M	AHN palpable	L3.2 - 6	L37	N		S+	LPUJO	
2	M	AHN	L3	L20	VUR I grade I RGP-pelvic stasis		Valve fulguration RGP L pyeloplasty S-	PUV/L PUJO PU/VUR grade I	perinephric leak persisting PUJ calculus
5	M	AHN	L2.6	L35	N		S-, Post op UTI	LPUJO	
1	F	AHN	R 3	R 80	N		Nephrostomy stent	R PUJO left dysplastic Kidney	

Lower moiety PUJO underwent pyelo-ureterostomy with stents and showed 10% increase in function on follow-up EC scan. PUJO and concomitant vesicoureteric obstruction had initial loop ureterostomy and later

pyeloplasty. All children had received a single course of preoperative antibiotics for pyeloplasty. Chemoprophylaxis was advised in VUR and stent pyeloplasty. Post PCN and pyeloplasty EC san showed improvement in renal function. Histology of PUJO revealed extensive fibrosis and muscular atrophy. Follow-up varied from two months- three years.

DISCUSSION

Incidence of PUJO is 1:1000 live birth.[1] Antenatal hydronephrosis are evaluated in first week and 4 weeks of life. Differential diagnoses of antenatal hydronephrosis are transient obstruction, PUJO, VUR, VUO, ureterocele, high bladder pressure (neurogenic bladder), and lower urinary tract obstruction (PUV).[1] PUJO may be unilateral or bilateral. Incidence of bilateral varies from 10-20%. Left kidney and boys are commonly affected.[1] Associated anomalies are dysplastic kidneys, VUR ipsilateral/contralateral, renal agenesis, VUO and CHD.[1] Ciliogenesis defects cause both renal defects and CHD.[2] However down syndrome, bilateral congenital cataract, craniosynostosis, PUV and PU are rare.[3,4] Risk of urinary anomalies in Down syndrome is 3.2% but not the PUJO.[3] Bilateral cataracts have 50% genetic etiology but not systemic anomalies.[4]

Infantile PUJO are detected commonly on evaluation of antenatal hydronephrosis, palpable kidney, urinary tract infections, acute renal failure and failure to thrive.[1] Antenatal giant intra-abdominal cyst is one rare of presentations for infantile PUJO.[5] Giant hydronephrosis are palpable, crosses the midline, and often present as gross abdominal distention in neonates. They are prone for UTI and cause compressive symptoms of surrounding organs.[5] Our first case is unique because it had preoperative diagnosis of concomitant B/L UVJO.[6] Concomitant VUJO are diagnosed pre, on and post pyeloplasty procedures.[6] Indicators of pre pyeloplasty VUJO are dilated ureter and difficult cannulation of lower ureter or delayed RGP drainage.[6]

Management of antenatal hydronephrosis varies from one institution to another institution such as sonological grading of hydronephrosis, routine MCU, isotope diuretic renogram EC/DTPA, EC/IVP, PCN/on table pyeloplasty or nephrectomy for poorly functioning kidney, stent or stent less pyeloplasty.

Society for fetal urology (SFU) grading and anteroposterior diameter of pelvis are helpful in management of hydronephrosis. Sonology gives only anatomical level of obstruction but not the degree of functional obstruction. Functional implication of APD depends on awareness of SFU grading, neonatal oliguric phase, extra renal pelvis, VUR, pyonephrosis, and contra lateral parenchymal thickness. Mean and median of APD in our series were 3 and 2.5cms respectively. Change of SFU grading, calyx-to-parenchyma ratio, anteroposterior diameter and calyceal dilatation are independent risk factors used in management of PUJO.[7]

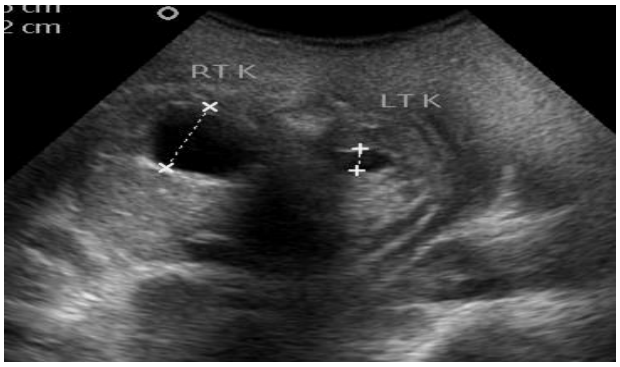

Figure 1: Antenatal sonographic image showing both kidneys AP diameter.

MCU was done in all isolated infantile hydronephrosis as per institution protocol. It had changed management in three infants only. However it is recommended in dilated ureter, solitary kidney, and duplex kidney.[8] Range of VUR in PUJO and dysplastic kidney varies from 10 - 40%. Routine use of MCU in PUJO is debatable.[8] Retrograde ureterography may be desirable investigation of choice in the PUJO with concomitant VUJO, poorly functioning kidney and post pyeloplasty anastomotic stricture.[5] Intravenous urography gives the additional information induplex kidney evaluation.[9] EC scan is always better than DTPA in evaluation of PUJO.[10]

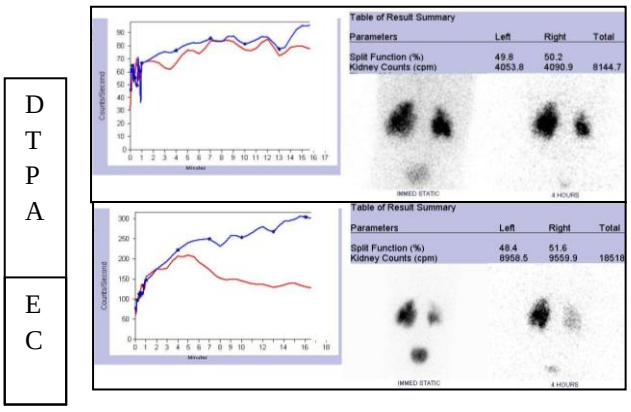

Figure 2: Curves and delayed static images of dtpa and EC scans.

PCN had relieved the urinary stasis and complications of giant hydronephrosis. Monitoring daily urinary out showed the functioning recovery in PUJO of poorly functioning kidney. Post PCN EC scan also showed functional improvement. Hence all our three infants were planned for pyeloplasty rather on table assessment for pyeloplasty or nephrectomy. Figure 3:Intraoperative finding showing renal parenchyma below, dilated pelvicalyceal system above and pelviureteric junction obstructed part distally

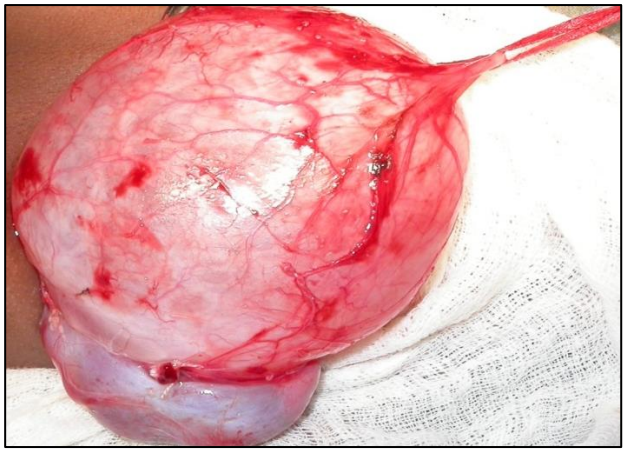

Figure 3: Intraoperative finding showing renal parenchyma below, dilated pelvicalyceal system above and pelviureteric junction obstructed part distally.

Median age at pyeloplasty is 2 months. Only two required redo pyeloplasty. Cheri T et al, Shaul et al, Tal et al, and our study emphasis the success of pyeloplasty depends on early/antenatal diagnosis, postnatal comprehensive evaluation/counseling and close regular follow-up.[11-13] Children with secondary procedure including stent/drain insertion, redo pyeloplasty would have prolonged hospital stay and urinary leak culminating in anastomotic stenosis.[14] Bilateral PUJO is uncommon and successive pyeloplasty have good success rate.

Lower moiety PUJO is rare and boys are commonly involved.[15,16] Dismembered pyeloplasty or pyelo ureterostomy procedures depend on individual presentations of lower moiety PUJO such as incomplete/ complete duplication.[15,16]

Figure 4: One of the C-ARM pictures showing shunt migration, which is an unusual complication.

Stent or stent less pyeloplasty has its own limitations and advantages, which has been substantiated in our study also.[17] Stent coiling in lower ureter may due to stent migration or improper placement. Dripping of urine makes confirmation of DJ stent in urinary bladder at pyeloplasty from upper end of stent or on table fluoroscopic visualization and cystoscopic retrograde insertion of stent in neonates.[18] Cost of stent, second anesthesia, early discharge, urinary leak, infections and anastomotic stricture will determine use of the stent.[11,12,19] However prospective double blind random study of pyeloplasty on stent or stent less may be needed. Post pyeloplasty EC scan showed 5-10% renal function recovery.

Limitations our study are observational study and short follow-up. Trend of open, laparoscopic and robotic pyeloplasty procedures depends on availability of equipment/skills, optimum utilization of operative time and cost.[20] Cost is not a factor as all our infantile pyeloplasty were covered under government schemes.

CONCLUSION

Antenatal scan, serial ultrasound and EC scan shall diagnose PUJO early which makes early infantile pyeloplasty is safe and effective. Careful evaluation and individualized treatment approach is required for lower moiety PUJO. Routine MCU is useful in very small number of PUJO. Stents should be used with caution.

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REFERENCES

1. Senguttuvan P, Jigy J. Profile and outcome of pelviureteric junction obstruction. Open Urology Nephrol J. 2014;7(1):44-7.
2. Agustin JT, Klena N, Granath K, Panigrahy A, Stewart E, Devine W, et al. Genetic link between renal birth defects and congenital heart disease. Nat Commun. 2016:1-10.
3. Kupferman JC, Druschel CM, Kupchik GS. Increased prevalence of renal and urinary tract anomalies in children with down syndrome. Pediatrics. 2009;124:615-21.
4. Leonard A, Bernard P, Hiel AL, Hubinont C. Prenatal diagnosis of fetal cataract: case report and review of the literature. Fetal Diagn. 2009;26:61-7.
5. Masarwa I, Bahouth Z, Halachmi S. Giant congenital hydronephrosis obstructing the gastro intestinal system and the contralateral kidney in a new born. Urol Case Rep. 2016;8:1-3.
6. Cay A, Imamoglu M, Bahat E, Sarihan H. Diagnostic difficulties in children with coexisting pelvi-ureteric and vesico-ureteric junction obstruction. BJU Int. 2006;98:177-82.
7. Helmy TE, Harraz A, Sharaf DE, Demerdash Y, Hafez AT, Gad H, et al. Can renal ultrasonography predict early success after pyeloplasty in children? A prospective study. Urol Int. 2014;93:406-10.
8. Kim YS, Do SH, Hong CH, Kim MJ, Choi SK, Han SW. does every patient with ureteropelvic junction

obstruction need voiding cystourethrography? J Urol. 2001;165:2305-7.
9. Antao B, Agarwal S, Ng J, Rodrigues B, Broadley P, Roberts J, et al. Is intravenous urography necessary in the assessment of renal duplex system in children? Surgeon. 2009;7:143-5.
10. Shahrokh H, Movahhed M, Shoshtari MA, Orafa AM, Hekmat S. Ethylenedicysteine versus diethylenetriamine pentaacetic acid as the carrier of technetium Tc 99m in diuretic renography for patients with upper urinary tract obstruction. Urol J. 2006;3:97-3.
11. Tal R, Sever Z, Livne PM. Dismembered pyeloplasty in children: a review of 5 years single center experience. Int J Urol. 2005;12:1028-1.
12. Chertin B, Fridmans A, Knizhnik M, Hadas-Halperin I, Hain D, Farkas A. Does early detection of ureteropelvic junction obstruction improve surgical outcome in terms of renal function? J Urol. 1999;162:1037-40.
13. Shaul DB, Cunningham JA, Lowe P, Skaist LB, Hardy BE. Infant pyeloplasty is a low-risk procedure. J Pediatr Surg. 1994;29:343-7.
14. Dy GW, Hsi RS, Holt SK, Lendvay TS, Gore JL, Harper JD. National trends in secondary procedures following pediatric pyeloplasty. J Urol. 2016;195:1209-14.
15. Avlan D, Gundogdu G, Delibaş A, Nayci A. Pyeloureterostomy in the management of the lower pole pelvi-ureteric junction obstruction in incomplete duplicated systems. Urology. 2010;76:1468-1.
16. Gonzalez F, Canning DA, Hyun G, Casale P. Lower pole pelvi-ureteric junction obstruction in duplicated collecting systems. BJU Int. 2006;97:161-5.
17. Varda BK, Johnson EK, Clark C, Chung BI, Nelson CP, Chang SL. National trends of perioperative outcomes and costs for open, laparoscopic and robotic pediatric pyeloplasty. J Urol. 2014;191:1090-5.
18. Garg RK, Menon P, Rao KL, Arora S, Batra YK. Pyeloplasty for hydronephrosis: issues of double J stent versus nephrostomy tube as drainage technique. J Indian Assoc Pediatr Surg. 2015;20:32-6.
19. Lee LC, Kanaroglou N, Gleason JM, Salle JL, Bägli DJ, Koyle MA, et al. Impact of drainage technique on pediatric pyeloplasty: comparative analysis of externalized uretero-pyelostomy versus double-J internal stents. Can Urol Assoc J. 2015;9:453-7.
20. Varda BK, Johnson EK, Clark C, Chung BI, Nelson CP, Chang SL. National trends of perioperative outcomes and costs for open, laparoscopic and robotic pediatric pyeloplasty. J Urol. 2014;191:1090-5.

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