CLINICOPATHOLOGICAL SPECTRUM OF URETEROPELVIC OBSTRUCTION AT A TERTIARY CARE CENTER NILOUFER HOSPITAL, HYDERABAD, ANDHRA PRADESH

Manimekhala¹, M. Lavanya², M. Ramani³, K. Ramesh Reddy⁴, Srinivas Reddy⁵, Sara⁶

HOW TO CITE THIS ARTICLE:

Manimekhala, M Lavanya, M Ramani, K Ramesh Reddy, Srinivas Reddy, Sara. "Clinicopathological spectrum of ureteropelvic obstruction at a tertiary care center Niloufer hospital, Hyderabad, Andhra Pradesh". Journal of Evolution of Medical and Dental Sciences 2013; Vol2, Issue 37, September 16; Page: 7148-7154.

ABSTRACT: Ureteropelvic junction obstruction is a condition in which blockage occurs at the junction where the ureter attaches to the renal pelvis, resulting in decreased flow of urine down the ureter and an increase of fluid pressure inside the kidney. Our study included 52 children from 3rd day of life to 14 years presenting at a tertiary care pediatric hospital with UPJO from May 2006 to April 2013. The presenting features, ultra sonographic findings, operative findings, intrinsic causes, renal isotope scan findings, and histological features, special stains were analyzed. We found that UPJO in pediatric age group is mainly due to intrinsic abnormalities i.e., fibrosis, excessive collagen fibers around the hypertrophied muscle bundles. Severity depends on extent of obstruction, so early correction by pyeloplasty improves morbidity

INTRODUCTION: Ureteropelvic junction obstruction (UPJO) is the most common cause of obstructive nephropathy; accounting for about 1:1,500 new-borns. The widespread use of obstetric ultrasonography has resulted in early and more frequent observations of congenital hydronephrosis, improving its post-natal management. UPJO may be considered, in most cases, as a functional obstructive condition due to maturative abnormalities, during the gestation, in the smooth muscle and/or the innervation of the pyelo-ureteral transitional segment, defective peristaltic waves and resulting in aperistaltic segment and difficulty to eject urine from the renal pelvis into the ureter. Smooth muscle discontinuity or disproportionate presence of longitudinal smooth muscle fibers and excessive collagen deposition are sometimes associated with an inappropriate innervation. This results in a defective propagation of the electrical activity produced by a pacemaker located in the pyelocaliceal region. The present study aims at correlating the ureteropelvic junction abnormality with clinico-radiological and histopathological findings.

METHODS: A retrospective and prospective study of 7 years from May 2006-April 2013 with review of clinical presentation, Ultrasound findings, renal isotope scan findings and microscopy of UPJO segments of 52 patients under 14 yrs, who underwent reconstruction at our hospital, a tertiary care pediatric center were studied. Extrinsic causes of UPJO were ruled out by ultrasonography and operative findings were noted. Only patients with UPJO due to intrinsic causes confirmed on histopathology were studied. Special stains - PAS and Masson Trichrom were done to highlight the histological alterations.

RESULTS: Our study reviewed 52 cases, of which most cases belonged to the age group of 0-2 years with 34(65.38%) cases and only 1(1.92%) case after 10 years of age. (Table 1). Sex incidence showed

male preponderance- 43(82.69%) male children and 9(17.30%) female children. (Table 1).Left sided UPJOs were more common-34(65.38%), right sided PUJOs were 16(30.76%) and there were 2 cases with bilateral PUJO.

All antenatally diagnosed cases had ipsilateral severe renal dysfunction and contralateral hydronephrosis. Older children presented with severe urosepsis, oliguria, hematuria and failure to thrive with ipsilateral severe renal dysfunction and contralateral moderate renal dysfunction. (Table2).

The most common clinical presentation was pain, fever, distension of abdomen and mass per abdomen in 34 cases.5 cases were diagnosed antenatally.14cases presented with only pain.4 cases presented with severe urinary complications like urosepsis, oliguria, hematuria and failure to thrive. (Table 2)

At the time of presentation the renal isotope scan showed ipsilateral moderate dysfunction in most of the cases. (Table 4) Intraoperatively kidneys were enlarged with narrowed UPJO in all 52/52 cases. Extrinsic causes were ruled out.

Anderson - Hynes pyeloplasty was done in49/52 cases and 3/52 underwent nephrectomy. 49 UPJO segments and 3 nephrectomy specimens were processed. Histopathology of 36 cases showed transitional epithelial lining, 16 cases showed metaplastic changes (Fig 1, 2). All cases showed hypertrophied muscle bundles, increased fibrocollagenous tissue and chronic inflammatory infiltrate (Fig 3, 4, 5, 6). Nephrectomy specimens showed-thinned out renal parenchyma, marked glomerulosclerosis, interstitial nephritis, fibrosis, dilated tubules and dense chronic inflammatory infiltrate - end stage kidney (7, 8, 9).Special stains Massons Trichrome demonstrated extensive fibrocollagenous tissue in all UPJO segments. PAS and Massons Trichrome showed glomerulosclerosis and interstitial fibrosis in the nephrectomy specimens studied.

DISCUSSION: UPJO is a congenital condition occurring at all ages in children with an autosomal inheritance (1, 2).Congenital UPJO is mostly due to intrinsic causes, in most cases, due to inadequate canalization of this area or as a functional obstruction (3, 4).Other extrinsic causes for UPJO are renal / ureteral stones, aberrant lower pole renal vessels, bands, kinks, tumors (3). UPJO is the most common cause of hydronephrosis in children (5, 6).

Antenatal sonography has increased the chances of UPJO detection (1).1 in 1500 fetuses showed UPJO during antenatal screening (7). There was an increased incidence in male children and more so on the left side (7).Our study also showed similar findings with 82.69% males with 65.38% cases being left sided (8). The most common clinical presentation was a symptom triad of pain, fever and abdominal mass (4) which was a common presentation in our study also-46.15%.Other presentations include severe urinary tract complications, and failure to thrive. In our study all antenatally diagnosed children had unresolving hydronephrosis with severe renal dysfunction in ipsilateral and hydronephrosis with normal renal function in contralateral kidney. We had two cases above 9 years with bilateral renal dysfunction. This correlates well with the study by González R, et al. (4)

There have been many studies encompassing the entity of UPJO obstruction that include clinical findings, radiographic imaging, pathological examination of ureteropelvic junction obstruction per se and renal biopsies during pyeloplasty procedures(9).Symptomatic children usually require operative intervention.(6) In these patients, pyeloplasty is performed and consists of

resecting the atretic or stenotic segment and reattaching the normal ureter to the renal pelvis, thereby relieving the obstruction. Renal biopsies in patients in whom pyeloplasty is done show mostly relatively well maintained parenchyma, with overt changes in glomeruli (10). More subtle alterations have been described that relate to shifts in proximal-to-distal tubular ratios. Extreme thinning of the renal parenchyma can occur with only limited tubulointerstitial injury (9). In our study microscopy of UPJO in 52/52 cases showed fibrosis and excessive collagen fibres around the hypertrophied muscle bundles.

Histological alterations in kidney with UPJO were marked glomerulosclerosis, dilated tubules and dense chronic inflammatory infiltrate - end stage kidney similar to features noticed in a study by Elder J S Stansbrey et al (11). In UPJO segments extensive fibrosis, hypertrophied muscle bundles and chronic inflammatory infiltrate, metaplastic changes were noted. Skozel (12) in his study of 22 UPJO cases (1 month–10 years) showed similar histological features and demonstrated high expression of fibronectin, laminin, type 4 collagen and BCL-2 thus showing their relationship to the pathogenesis of UPJO. Defective kidney morphogenesis, during branching and tubulogenesis of ureteric bud, may also be responsible for this congenital pathology (8).

Open pyeloplasty is still considered the gold standard for UPJ obstruction in children (removal of stenotic segment and re-anastomosing the ureter to renal pelvis). Other novel surgical options include endopyelotomy, endopyeloplasty, laparoscopic pyeloplasty, robotic-assisted laparoscopic pyeloplasty. Studies by González R, Schimke CM recommend early surgical treatment to prevent obstructive damage to the immature infant kidney and because better recovery of function is possible when surgery is done in the first year of life. Similar findings were noticed in our study also. 4/52 cases which presented later in life showed features like failure to thrive, urinary complications and deterioration of renal function in both the kidneys and finally lead to nephrectomy (13). In our study we found that when patients presented late in life there was marked deterioration of renal function and finally lead to nephrectomy. Histology also showed features of end stage kidney.

CONCLUSION: UPJO in pediatric age group is most common cause of hydronephrosis, with an incidence of 1 in 1500 cases. There is a marked male preponderance with most cases being left sided. Histological study shows that it is mainly due to intrinsic abnormalities – fibrosis, excessive collagen fibres around the hypertrophied muscle bundles. Severity depends on extent of obstruction, so early correction by pyeloplasty prevents deterioration of kidney function leading to nephrectomy. Study of nephrectomy specimens reveals features of end stage kidney. Our study also suggests early intervention and management is mandatory in cases of UPJO.

REFERENCES:

1. Brown T, Mandell J, Lebowitz RL. Neonatal hydronephrosis in the era of sonography. *AJR Am J Roentgenol*. May 1987; 148(5):959-63.
2. A.H. Colodny et al. Ureteropelvic junction obstruction in children, *Urol Clin North Am* 1980; (7): 273–290.
3. Peter Kench. A Morphometric Study of the Pelvi-Ureteric Junction and Review of the Pathogenesis of Upper Ureteric, *Pathology*, 1982, Vol. 14, No. 3, 309-312.

ORIGINAL ARTICLE

4. González, R, Schimke, CM. Ureteropelvic junction obstruction in infants and children. *Pediatr Clin North Am* 2001; 48:1505.
5. D. Innes Williams, CM Karlaftis, Hydronephrosis due to pelvi-ureteric obstruction in the new born. *British Journal of Urology* April 1966 Volume 38, Issue 2, 138–144.
6. Josephson, S. Antenatally detected pelvi-ureteric junction obstruction: concerns about conservative management. *BJU Int* 2000; 85:973.
7. M.K. Hanna, R.D. Jeffs, J.M. Sturgess et al. Ureteral structure and ultrastructure. Part II. Congenital ureteropelvic junction obstruction and primary obstructive megaureter. *J Urol* 1976; 116: 725–730.
8. Zhang, PL, Peters, CA, Rosen, S. Ureteropelvic junction obstruction: morphological and clinical studies. *Pediatr. Nephrol.* 2000. 14:820-826.
9. S Rosen, CA Peters, The Kidney in Congenital Ureteropelvic Junction Obstruction: A Spectrum From Normal to Nephrectomy. *The Journal of Urology* April 2008, 179, 125-126
10. Chertin B, Pollack A, Koulikov D, et al. Does renal function remain stable after puberty in children with prenatal hydronephrosis and improved renal function after pyeloplasty? *J Urol* 2009; 182:1845.
11. Elder JS, Stansbrey R, Dahms BB, Selzman AA Renal histological changes secondary to ureteropelvic junction obstruction *The journal of urology* 1995 Aug;154(2):719-722.
12. Ş.K. Özela, H. Emirb, S. Dervişoğlu. The roles of extracellular matrix proteins, apoptosis and c-kit positive cells in the pathogenesis of ureteropelvic junction obstruction *Journal of Pediatric Urology*, April 2010, 6,125-129.
13. González R, Schimke CM The prenatal diagnosis of hydronephrosis, when and why to operate? *Arch Esp Urol.* 1998 Jul-Aug; 51(6):575-579.

	0-2 yr	2-5 yr	5-10 yr	10-14yrs
Male	28(53.84%)	12(23.07%)	2(3.84%)	1(1.92%)
Female	6(11.53%)	2(3.84%)	1(1.92%)	0

Table 1. Age and sex incidence of PUJO

No. of cases, age-group	Clinical presentation	USG abdomen	Renal isotope scan	Contralateral kidney
34 (0-2yrs)	Pain, fever, dis-tension, mass abdomen	Gross hydronephrosis	partial Upjo - Moderate renal dysfunction	Normal
14 (2-5yrs)	Only pain	Moderate hydronephrosis	Upjo with normal renal function	Normal
4 (9-12yrs)	Severe urinary complications- urosepsis, oliguria. hematuria, failure to thrive	Gross hydronephrosis, MCUG-normal	Significant upjo- severe renal dysfunction	hydronephrosis with Moderate renal dysfunction

ORIGINAL ARTICLE

5 (3-9months)	Antenatally diagnosed – upjo	Unresolving hydronephrosis, 1case multicystic kidney with upjo	Upjo with severe renal dysfunction	hydronephrosis with normal renal function
------------------	---------------------------------	---	---------------------------------------	---

Table 2: Clinico-radiological findings of 52 cases of PUJO

Clinical presentation	Number (%)
Fever	24 (46.15%)
Distension and mass abdomen	20(38.46%)
Abdominal pain	24 (46.15%)
Failure to thrive	04(7.69%)
Hematuria, oliguria, urosepsis	04(7.69%)
convulsions	04 (7.69%)

Table 3: Clinical presentation of PUJ obstruction

No. of cases	Kidney with UPJO	Contralateral Kidney
32	Moderate renal dysfunction	Normal
10	Severe renal dysfunction	Hydronephrosis
6	Normal renal function	Normal
4	Non-functioning kidney-UPJO with multiple cysts	Moderate dysfunction

Table 4: Renal isotope scan findings in 52 cases in UPJO

Fig 1: Scanner view of UPJO showing metaplastic changes

Fig 2: Low power view of UPJO showing metaplastic changes

Fig 3: Scanner view of UPJO showing hypertrophied muscle fibres

Fig 4: Low power view of UPJO showing hypertrophied muscle fibres & excessive collagen fibers

Fig 5: High power view of UPJO showing hypertrophied muscle fibres

Fig 6: High power view of UPJO showing excessive collagen fibers

Fig 7: Scanner view of nephrectomy specimen showing glomerulo sclerosis, interstitial nephritis, fibrosis, dilated tubules.

Fig 8: Low power view of nephrectomy specimen showing glomerulosclerosis

Fig 9: High power view of nephrectomy specimen showing interstitial nephritis, dilated tubules and dense chronic inflammation.

AUTHORS:

1. Manimekhala
2. M. Lavanya
3. M. Ramani
4. K. Ramesh Reddy
5. Srinivas Reddy
6. Sara

PARTICULARS OF CONTRIBUTORS:

1. Associate Professor, Department of Pathology, Niloufer Hospital, Hyderabad.
2. Assistant Professor, Department of Pathology, Niloufer Hospital, Hyderabad.
3. Professor, Department of Pathology, Niloufer Hospital, Hyderabad.
4. Professor and H.O.D, Department of Paediatric Surgery, Niloufer Hospital, Hyderabad.

5. Assistant Professor, Department of Paediatric Surgery, Niloufer Hospital, Hyderabad.
6. Post Graduate, Department of Pathology, Niloufer Hospital, Hyderabad.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Manimekhala,
Associate Professor of Pathology,
Flat No. – 204, A Block, Vishnu Residency,
Gandhinagar, Hyderabad.
Email – manimekhalara@gmail.com

Date of Submission: 22/08/2013.

Date of Peer Review: 31/08/2013.

Date of Acceptance: 02/09/2013.

Date of Publishing: 13/09/2013