CLINICOPATHOLOGICAL SPECTRUM OF URETEROPELVIC OBSTRUCTION
AT A TERTIARY CARE CENTER NILOUFER HOSPITAL, HYDERABAD,
ANDHRA PRADESH
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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, ultrasoundographic 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 14yrs, 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. (Table 2).

The most common clinical presentation was pain, fever, distension of abdomen and mass per abdomen in 34 cases. 5 cases were diagnosed antenatally. 14 cases 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 in 49/52 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, there by 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. S k ozel (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 includes endopyelotomy, endo pyeloplasty, 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:


<table>
<thead>
<tr>
<th>No. of cases, age-group</th>
<th>Clinical presentation</th>
<th>USG abdomen</th>
<th>Renal isotope scan</th>
<th>Contralateral kidney</th>
</tr>
</thead>
<tbody>
<tr>
<td>34 (0-2yrs)</td>
<td>Pain, fever, dis-tension, mass abdomen</td>
<td>Gross hydronephrosis</td>
<td>partial Upjo - Moderate renal dysfunction</td>
<td>Normal</td>
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<td>14 (2-5yrs)</td>
<td>Only pain</td>
<td>Moderate hydronephrosis</td>
<td>Upjo with normal renal function</td>
<td>Normal</td>
</tr>
<tr>
<td>4 (9-12yrs)</td>
<td>Severe urinary complications-urosepsis, oliguria. hematuria, failure to thrive</td>
<td>Gross hydronephrosis, MCUG-normal</td>
<td>Significant upjo-severe renal dysfunction</td>
<td>hydronephrosis with Moderate renal dysfunction</td>
</tr>
</tbody>
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Table 1. Age and sex incidence of PUJO
Antenatally diagnosed upjo
Unresolving hydronephrosis, 1 case multicystic kidney with upjo
Upjo with severe renal dysfunction
hydronephrosis with normal renal function

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

<table>
<thead>
<tr>
<th>Clinical presentation</th>
<th>Number ( % )</th>
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<tbody>
<tr>
<td>Fever</td>
<td>24 (46.15%)</td>
</tr>
<tr>
<td>Distension and mass abdomen</td>
<td>20(38.46%)</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>24 (46.15%)</td>
</tr>
<tr>
<td>Failure to thrive</td>
<td>04(7.69%)</td>
</tr>
<tr>
<td>Hematuria, oliguria, urosepsis</td>
<td>04(7.69%)</td>
</tr>
<tr>
<td>convulsions</td>
<td>04 (7.69%)</td>
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Table 3: Clinical presentation of PUJ obstruction

<table>
<thead>
<tr>
<th>No. of cases</th>
<th>Kidney with UPJO</th>
<th>Contralateral Kidney</th>
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</thead>
<tbody>
<tr>
<td>32</td>
<td>Moderate renal dysfunction</td>
<td>Normal</td>
</tr>
<tr>
<td>10</td>
<td>Severe renal dysfunction</td>
<td>Hydronephrosis</td>
</tr>
<tr>
<td>6</td>
<td>Normal renal function</td>
<td>Normal</td>
</tr>
<tr>
<td>4</td>
<td>Non-functioning kidney-UPJO with multiple cysts</td>
<td>Moderate dysfunction</td>
</tr>
</tbody>
</table>

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 glomerulosclerosis, 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.

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