Giant hydronephrosis: still a reality!
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ABSTRACT
Objective: Giant hydronephrosis (GH) is a rare entity in both developed and developing countries with less than 500 cases reported in the literature. Delayed diagnosis and management of GH, can result in long-term complications like hypertension, rupture of the kidney, renal failure and malignant change. We aim to highlight the importance of this often neglected entity and build a consensus for its early diagnosis and management.

Material and methods: Patients with GH were thoroughly worked up, managed and followed up between June 2013 and December 2015 and epidemiologic, radiological, perioperative and follow-up data was recorded.

Results: A total of 35 patients (adults and children) were reported. Flank pain in adults and abdominal lump in children were the most common clinical presentation. Percutaneous nephrostomy tube was placed in all patients and detailed work up was done to reach final diagnosis. Pelvi-ureteric junction obstruction (PUJO) was the final diagnosis in 32 patients (91.4%). Kidneys were non-functioning in 13 cases (37.1%) so nephrectomies were performed. Reduction pyeloplasty with nephropexy was done in 21 patients (60%) with 81% success and 23.1% complication rates.

Conclusion: GH requires early diagnosis and management to prevent higher nephrectomy rate along with poor success rate of conservative surgery like pyeloplasty.

Keywords: Giant hydronephrosis; nephrectomy; pelvi-ureteric junction obstruction; pyeloplasty.

Introduction
Hydronephrosis (HDN) is defined as the dilatation of pelvi-calyceal system due to obstruction and stasis of urinary flow.[1] Giant hydronephrosis (GH) has been variably defined in the literature as the presence of more than 1,000 mL/1.6% of body weight of fluid in the renal collecting system or the involvement of five vertebral heights. Till now, only 500 cases of GH have been reported in the literature.[2] This entity is uncommon in developed countries but often encountered in the developing world. Majority of reported cases occur in infants and children, and are congenital in origin. GH, if left undiagnosed, can result in long-term complications like hypertension, rupture of the kidney, renal failure and malignant change.[3] Since the first description of GH about a century ago, only a few small series have been published in the literature. There are no long-term follow-up and outcome data in the literature on GH. We present our follow-up and outcome experiences with GH in 35 cases from a high volume center in Northern India.

To study the natural history, suitable intervention, future course and follow-up of the patients presenting with GH based upon their etiology, location of obstruction, anatomical configuration and functional status of renal units and develop a consensus regarding the early and best management of these rarely reported cases.

Material and methods
A prospective longitudinal study was conducted between June 2013 and December 2015 and the patients presenting with GH were thoroughly worked up, managed and followed over a certain period of time. The institutional ethical committee clearance was taken for the project. The details of epidemiologic data, radiological investigations, therapeutic indications, preoperative findings and follow-up of all patients were recorded after obtaining...
informed consent from their patients. All the patients (adults and children) suspected of HDN which was defined as presence of more than 1 litre or fluid (or pus) amounting to 1.6% body weight in the pelvicalyceal system, enlarged kidney occupying the hemi-abdomen or crossing the midline and involving five vertebral bodies in length were included in the study. Patients with bleeding diathesis and pregnant women were excluded. All these patients underwent percutaneous nephrostomy (PCN) tube placement to decompress the system. The detailed anatomical and functional assessments were performed 2-6 weeks after PCN tube placement with the aid of ultrasonography/intravenous urography (IVU)/contrast enhanced computed tomography (CECT)/nephrostogram, to further characterize the cortical thickness, anatomical and functional status of the kidney. Patients with renal cortical thickness more than 5 mm and differential renal function more than 15% were managed conservatively with pyeloplasty or percutaneous nephrolithotomy (PNL) while those with renal cortical thickness less than 5 mm and differential renal function less than 15% underwent nephrectomy. Follow-up after surgery was done at 3, and 6 months and 1 year. Detailed history and physical examination, serum creatinine level, ultrasonography and renal ethylene dicysteine (EC) scan were done at each follow-up visit. The success of pyeloplasty was defined as the absence of clinical symptoms and normal curve on EC scan. Complications were recorded as per the Clavien Dindo classification. Normal serum creatinine was defined as 0.7-1.3 mg/dL for men and 0.6-1.1 mg/dL for women, 0.24-0.36 mg/dL in infants and more than 0.24-0.36 mg/dL in male, and 0.2-0.8 mg/dL in female children between 1-12 years of age.

**Results**

A total of 35 patients were reported during the study period. Majority were adults (15 male/4 female) with a mean age of 31±3.3 years followed by children (14 male/2 female) with a mean age 3.9±1.4 years (Table 1). Flank pain (19/19) and abdominal lump (12/19) were the most common presentations in adults while children commonly presented with abdominal lump (16/16) followed by flank pain (14/16) and fever (12/16). Nine patients had renal dysfunction (7 adults and 2 children). Mean PCN output immediately on PCN placement was 3.5±0.6 L in adults and 1.9±0.4 L in children with purulent discharge in 6 adults and 5 children. US KUB was done at the time of presentation (before PCN tube placement) and 6 weeks after PCN tube placement. On the delayed US, cortical thickness <5 mm was present in 15 patients (10 adults and 5 children) and IVU showed non-excreting kidney (delayed image) in these patients (Table 2).

Intravenous urography was done in 8 patients, and all these patients presented to us with IVU already done at an external center. Contrast-enhanced computed tomography (CECT) scan was done in 4 adults to find some associated pathology. Aberrant crossing vessel was discovered preoperatively in one adult patient on the triple phase CT. Nephrostogram was done in 10 patients with raised serum creatinine levels which showed a large hydronephrotic sac reaching up to the pelvic bone. Renal scan showed <15% differential renal function in 14 patients (9 adults and 5 children) and functional pattern was obstructive in all except one. Simple nephrectomy was done in all patients except one adult man who underwent pyeloplasty who recovered well. Final diagnosis was pelvi-ureteric junction obstruction (PUJO) in 32 patients (16 adults and 16 children) and upper ureteric stone in 3 patients. Out of these 32 patients of PUJO, 13 patients (8 adults and 5 children) had nonfunctioning kidney, for which they underwent nephrectomy. Pyeloplasty was performed in 21 patients (10 adults and 11 children) and percutaneous nephrolithotomy (PNL) was performed in one adult. Laparoscopic pyeloplasty was the preferred procedure in adults (31.6%) while open pyeloplasty was most commonly done in children (56.3%) and Anderson Hynes pyeloplasty (21 cases) were done in all these patients (Table 3). Intraoperative crossing vessels were found in 3 adults and 2 children during pyeloplasty and these were managed with dismembered pyeloplasty. Overall success rate of pyeloplasty was 80.9% (laparoscopic and open) and children reported relatively better success rate (90.9%) compared to adults (70%). Four patients (19.1%) had disease recurrence who were managed with endopyelotomy (3 adults) and uretero-calyceostomy (one child). Overall complication rate was 23.8% and complications were more common in adults (30%) compared to children (18.1%). Majority of complications were of Clavien I and II grade which responded conservatively to antibiotic treatment, only one adult with upmigration of JJ stent required JJ stent replacement. The operative time of open pyeloplasty in GH was 138±32 min (vs 108±23 min) and laparoscopic pyeloplasty it was 198±27 min (vs 173±25 min) which was significantly higher than operative time in simple HDN surgeries. Patients undergoing nephrectomy and percutaneous nephrolithotomy (PNL) did not report any major complication and recovered well (Table 4).

**Follow up**

The follow-up period ranged from 6 months to 23 months. Monitorization of the patients was performed with blood tests (serum creatinine and blood urea), US, KUB and renal scan. Two of 8 patients presenting with high serum creatinine levels had chronic renal insufficiency and were on dialysis. All patients undergoing pyeloplasty and nephrectomy were doing fine. The degree of hydronephrosis did not show any change and remained stable without any signs of aggravation.

**Statistical analysis**

A comparative analysis of results was carried out between groups using ANOVA, and applying Pearson’s correlation coefficient. Statistical Package for the Social Sciences, version 16.0 (SPSS Inc.; Chicago, IL, USA), was used for the statistical analysis. P value <0.05 was considered to be significant.
Discussion

Stirling first defined GH as draining more than 1 litre fluid or fluid amounting to 1.6% of body weight in the collecting system. Later on, radiographic criteria for GH were defined by Crooks et al. as the kidney occupying the hemi-abdomen which also meets or crosses the midline and has a height of about 5 vertebral bodies. Most common presentation of GH is abdominal
lump followed by less common symptoms like flank pain, hematuria, acute abdominal pain and recurrent urinary tract infections.\(^9,10\) Rare presentations of GH include intestinal obstruction, respiratory distress, hypertension, pedal edema, obstructive jaundice and contralateral ureteropelvic junction obstruction.\(^11\) Majority of patients in our study presented with abdominal distension, flank pain and fever. Thus whenever a patient with suspected GH presents to a urology clinic, various conditions should be kept in mind in the differential diagnosis like hepatobiliary cysts, mesenteric cysts, pseudomyxoma, cystic renal tumor, retroperitoneal tumors, ovarian cyst, retroperitoneal haematoma, ascites and splenomegaly.\(^12\) Hence a thorough work up of the suspected patient should be performed to reach the final diagnosis. Right kidney was more commonly involved in our study similar to the study by Arias et al.\(^13\) although clinical implication of this finding is unclear.

Although classical teaching advocates PCN tube placement in GH patients presenting with fever and/or raised serum creatinine levels\(^14\), we placed PCN tube in all the patients. This helped us in rapidly decompressing the system, providing symptomatic relief and improving the cardiopulmonary status of these patients (as majority of patients presented with abdominal distention as their complaint), assessment of renal function by calculating 24 hr creatinine clearance of the affected kidney and detailed anatomical assessment later with IVU and/or CECT (Figure 1, 2). PCN was preferred over JJ stent in this study as it aided in decompression of the system and provided quick symptomatic relief to the patients. Rapid and safe decompression of the obstructed system by ultrasound guided PCN performed over JJ stent was also reported previously in the literature by Ahmad et al.\(^15\)

### Table 3. Management of GH in adults and children

<table>
<thead>
<tr>
<th>Variables</th>
<th>Adults, n=19</th>
<th>Children, n=16</th>
<th>Total, n=35</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Final diagnosis</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PUJO (%)</td>
<td>16 (84.2)</td>
<td>16 (100)</td>
<td>32 (91.4)</td>
<td>0.233</td>
</tr>
<tr>
<td>Upper ureteric calculus (%)</td>
<td>3 (15.8)</td>
<td>-</td>
<td>3 (8.6)</td>
<td>1.00</td>
</tr>
<tr>
<td>Management</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pyeloplasty (%)</td>
<td>10 (52.6)</td>
<td>11 (68.75)</td>
<td>21 (60)</td>
<td>0.49</td>
</tr>
<tr>
<td>Nephrectomy (%)</td>
<td>8 (42.1)</td>
<td>5 (31.25)</td>
<td>13 (37.1)</td>
<td>0.73</td>
</tr>
<tr>
<td>PNL (%)</td>
<td>1 (5.3)</td>
<td>-</td>
<td>1 (2.9)</td>
<td>1.00</td>
</tr>
<tr>
<td>Type of pyeloplasty</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Open (%)</td>
<td>4 (21.1)</td>
<td>9 (56.3)</td>
<td>13 (37.1)</td>
<td>0.04</td>
</tr>
<tr>
<td>Laparoscopic (%)</td>
<td>6 (31.6)</td>
<td>2 (12.5)</td>
<td>8 (22.9)</td>
<td>0.24</td>
</tr>
<tr>
<td>Type of nephrectomy</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Open (%)</td>
<td>5 (26.3)</td>
<td>3 (18.7)</td>
<td>8 (22.8)</td>
<td>0.70</td>
</tr>
<tr>
<td>Laparoscopic (%)</td>
<td>3 (15.8)</td>
<td>2 (12.5)</td>
<td>5 (14.3)</td>
<td>1.00</td>
</tr>
</tbody>
</table>

PUJO: pelvi-ureteric junction obstruction; PNL: percutaneous nephrolithotomy; GH: giant hydronephrosis

### Table 4. Pyeloplasty (Laparoscopic and Open) outcomes in GH patients

<table>
<thead>
<tr>
<th>Variables</th>
<th>Adults, n=10</th>
<th>Children, n=11</th>
<th>Total, n=21</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Success rate (%)</td>
<td>7 (70)</td>
<td>10 (90.9)</td>
<td>17 (80.9)</td>
<td>0.31</td>
</tr>
<tr>
<td>Recurrence rate (%)</td>
<td>3 (30)</td>
<td>1 (9.1)</td>
<td>4 (19.1)</td>
<td>0.31</td>
</tr>
<tr>
<td>Median follow-up (Months)</td>
<td>25</td>
<td>28</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Complication rate (%)</td>
<td>3 (30)</td>
<td>2 (18.1)</td>
<td>5 (23.8)</td>
<td>0.64</td>
</tr>
</tbody>
</table>

Complications

- Clavien I (prolonged urine leak) 1 1 2 1.00
- Clavien II (UTI, fever) 1 1 2 1.00
- Clavien III (upmigration of JJ stent) 1 -- 1 1.00

JJ: double J stent; UTI: urinary tract infection; GH: giant hydronephrosis
A variety of case reports are available in literature describing the amount of fluid drained using PCN tube placement. In our study, the mean amount of fluid drained immediately on PCN tube placement was 3.5±0.6 L in adults and 1.9±0.4 L in children. This significant difference can be explained by the delayed presentation in adults and higher capacity of retroperitoneal space to accommodate excess fluid compared to children.

Literature reports pelvi-ureteric junction obstruction (PUJO) as the most common cause of GH (1/3rd cases), followed by stones (upper ureter) in about one fifth of the cases. Other less common causes include congenital ureteral narrowing, ureteropelvic tumors, trauma, renal ectopia, retroperitoneal fibrosis, obstructive megaureter and ureteric atresia. The most common cause of GH in our study was also pelviureteric junction obstruction (PUJO) in 32 patients (91.4%) followed by upper ureteric calculus in 3 (8.6%) patients. Out of 32 patients with pelviureteric junction obstruction (PUJO), 13 patients (8 adults and 5 children) had non-functioning kidney and underwent nephrectomy. Pyeloplasty was performed in 21 patients and percutaneous nephrolithotomy (PCNL) was done in one adult. Thus the etiology of GHN in our study corroborated well with the literature.

Yapano et al.[12] described the preservation of renal parenchyma as the primary aim of management of GH. Hoffman[18] preferred nephrectomy in kidneys affected by GH as there was no improvement in function, in addition to higher gastrointestinal disturbances and increased susceptibility to trauma caused by the retained hugely hydronephrotic kidney. Shudo et al.[19] described the theoretical risk of malignancy due to chronic stimulation by left out stones in the HDN kidney. Uson et al.[20] reported 70% nephrectomy rate while Crooks et al.[8] reported 30% nephrectomy rate in kidneys with GH. Our series reported nephrectomy in 37.1% of the patients (42.1% in adults and...
Figure 2. a,b. Ultrasonography of an adult male presenting with abdominal distension and pain, shows hugely dilated sac of the right kidney with internal echoes, suggestive of pyonephrosis (a). Contrast-enhanced computed tomography scan showing giant hydronephrosis of the right kidney (b).

Figure 3. a-c. Nephrostogram of a patient with giant hydronephrosis showing hugely dilated pelocalyceal system of the left kidney (a). Intraoperative view during pyeloplasty showing a large redundant sac of the kidney for which reduction pyeloplasty with nephropexy was done (b, c)
31.25% in children) which is comparable to 33% reported by Sataa et al. and Crooks et al. Our study highlighted the higher rate of nephrectomy in GH compared to simple HDN (37.1% vs. 0.5%) comparable to that reported by Kinn et al. Hence this study highlights the need for early diagnosis and management in GH.

Also, the histopathology of nephrectomy specimens (non-functioning kidney post-PUJO and obstructive upper ureteric calculus) revealed grossly hydronephrotic kidneys with chronic pyelonephritis and microabcesses with ureter margins showing changes specific to ureteritis. This finding was similar to that reported by Mujagic et al. The most common underlying cause of GH in this study was pelvi-ureteric junction (PUJO) and majority of the patients underwent pyeloplasty (60%). Reduction pyeloplasty with nephrectopy was done in all the cases similar to Shah et al. Nephrectopy reduced the stasis of urine and improved dependent drainage as it tilted the pelvicalyceal system laterally, thus bringing it more in line with the upper ureter (Figure 3). Majority of pyeloplasties in adults (31.6%) were laparoscopically performed (transperitoneal approach). Uretero-calyceostomy, calycocystostomy, and Boari flap calycovesicostomy can be done in selected cases with massive calyceal dilatation and severely compromised peristalsis within the collecting system.

Adult, and pediatric patients were followed up for a median of 25, and 28 months, respectively. Success rates of pyeloplasty were 70% in adults and 90.9% in children. Success rates were similar in laparoscopic and open pyeloplasty. This success rate in GH is lesser than post pyeloplasty in simple HDN (>93%) as reported by Knoedler et al. and Pohl et al. Complication rate reported as 23.8% in our series is higher than reported in simple HDN by Pohl et al. Three patients of recurrence were managed with endopyelotomy and one child underwent ureterocalyceostomy. Complication like urinary tract infection (UTI) was managed with long-term antibiotic therapy, and upmigration of JJ-stent was managed with JJ stent placement over guide wire. Urine leakage gradually weaned with time.


In conclusion, GH is a rare entity, requiring individualized patient management. Multiple differential diagnoses should be considered and patient should be thoroughly worked up so that early management can be instituted as the delay can result in higher number of patients undergoing nephrectomy along with poor success rate and higher complication rate of pyeloplasty.
17. Kaya C, Pirincci N, Karaman MI. A rare case of an adult giant hydroureteronephrosis due to ureterovesical stricture presenting as a palpable abdominal mass. Int Urol Nephrol 2005;37:681-3. [Crossref]
26. Pohl HG, Rushton HG, Park JS, Belman AB, Majd M. Early diuresis renogram findings predict success following pyeloplasty. J Urol 2001;165:2311-5. [Crossref]