



## GIANT HYDRONEPHROSIS IN ADULTS: THE SPOILED KIDNEY

## Urology

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## ABSTRACT

**Purpose:** To study the incidence, clinical presentations and outcomes of giant hydronephrosis in adults along with review of literature.

**Materials and Methods:** 3 patients of giant hydronephrosis were included in the study. Patients underwent clinical examination, all routine investigations, CT scan of abdomen and diuretic renal scan.

**Results:** All renal units were non-functional and patients underwent nephrectomy after initial percutaneous nephrostomy. One emergency was performed in view of haematuria with clots.

**Conclusion:** UPJ Obstruction is most common cause of GH and kidney usually becomes non-functional at presentation. Abdominal lump is most common presentation. Timely intervention in adults can prevent further complications before patients undergoing nephrectomy. Management of GH is a two-step process including initial decompression by nephrostomy followed by nephrectomy

## KEYWORDS

UPJ Obstruction, Giant Hydronephrosis, Nephrostomy, Nephrectomy, Non-functional.

## INTRODUCTION:

Giant hydronephrosis (GH) is an infrequent entity these days, due to extensive antenatal screening and advances in diagnostic imaging modalities. UPJ (Ureteropelvic junction) Obstruction accounts for most of the cases, with other causes being urolithiasis, vesicoureteral reflux, ureteric stricture, upper tract urothelial cancer etc. GH occurring in adults is even rarer. Here we present a case series of 3 cases of giant hydronephrosis in adults with varied presentations along with literature review.

## CASE DETAILS:

## FIRST CASE:

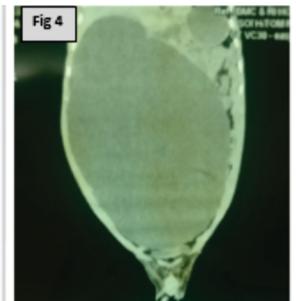
29-year-old male patient with complaints of painless abdominal distension since childhood that was insidious in onset, gradually increasing in dimensions and now occupying whole of the abdomen. On examination patient was poorly built, with BMI 18.5 Kg/m<sup>2</sup>. Huge abdominal distension (**Fig 1**) of size around 30x20 cm is seen completely filling the abdomen with right flank fullness. Biochemical investigations were within normal limits except serum creatinine of 1.52 mg/dL. Urine routine showed 4-6 pus cells/hpf. CT Scan/ Intravenous Pyelography (IVP) of abdomen showed (Right UPJ Obstruction with large 39x26 cm grossly hydronephrotic kidney with no visible parenchyma (**Fig 3,4,5**). The mass completely occupied abdomen, pushing peritoneal contents. Synchronous Left UPJ obstruction was also detected with moderate hydronephrosis. Diuretic renogram revealed non-functioning right kidney with left obstructed UPJ. Subsequently patient underwent Right PCN insertion and left pyeloplasty. PCN drained clear urine and patient's abdominal distension completely disappeared, leading to scaphoid abdomen. (**Fig 2**). On 1<sup>st</sup> day PCN output was 8 litres which was 6, 4 litres on 2<sup>nd</sup> and 3<sup>rd</sup> day respectively. Nephrostomy drained nearly 26 litres of fluid by 10<sup>th</sup> day before draining minimally. Patient lost 20 Kg of weight. Patient underwent left pyeloplasty and right simple nephrectomy 2 weeks later. At 1 year follow-up patient was symptoms free with DTPA scan showing unobstructed left pelvicalyceal system and serum creatinine of 1.0 mg/dL.

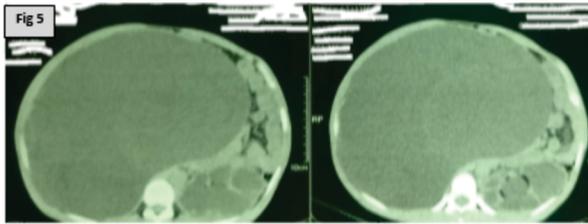


**Fig 1: Distended abdomen**



**Fig 2 : Post Nephrostomy –No lump**





**Fig 3: IVP- Ground glass opacity of abdomen With left UPJ Obstruction**

**Fig 4: Giant Hydronephrotic right kidney in CT scan**

**Fig 5: Axial CT scan for Right hydronephrotic non functional kidney with left sided UPJO**

**SECOND CASE:**

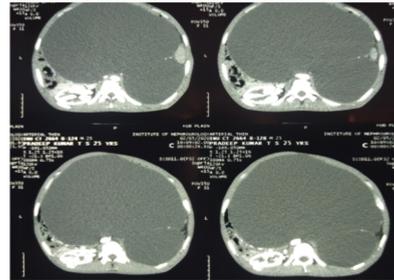
25 years old male presented with complaints of mass per abdomen since 6 years which was gradual in onset and associated with continuous dull aching pain occupying the whole abdomen. On examination patient was average built, with BMI 25 Kg/m<sup>2</sup>. Huge abdominal distension (Fig 6) was seen completely filling the abdomen along with flaying of ribs. Routine blood parameters were within normal limits. Urine routine showed 3-5 pus cells/ HPF rest was normal. CECT KUB showed massively enlarged left kidney (30x25 cm) with thinned out parenchyma causing mass effect over adjacent bowel loops (Fig 8). Left decompressing nephrostomy was done leading to flat abdomen (Fig 7). PCN drained about 7 litres of clear fluid on OT table, 4 litres on Day 1, 2 litres on day 2 and 3 each. Patient discharged with PCN tube in situ. Nephrostomy drained nearly 20 litres of fluid by 10<sup>th</sup> day before draining minimally. After 3 weeks patient underwent left simple nephrectomy. Patient had uneventful 1 year follow up.



**Fig 6 : Huge Abdominal Lump**



**Fig 7: Post Nephrostomy :Scaphoid Abdomen**



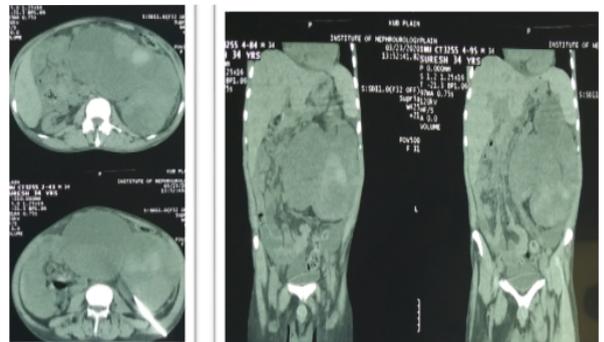
**Fig 8: Massively enlarged left kidney occupying all abdomen.**

**THIRD CASE:**

34 years old male presented with complaints of dull aching left abdominal pain for 2 years, associated with abdominal distension (Fig 9) with on and off haematuria. On examination patient was anaemic, poorly nourished, tense tender abdominal mass was present in left hemi abdomen, dull on percussion with no sign of peritonitis. The blood investigation showed a haematocrit of 19.3%, haemoglobin of 5.3 g/dL, creatinine of 8.4 mg/dL, and urea of 70 mg/dL, USG KUB showed gross hydronephrosis of left kidney (28 x 25 cm)? UPJ obstruction with thinned out parenchyma, bilateral grade 3 nephropathy. Urine routine showed numerous RBCs and pus cells. Non contrast CT highlighted left UPJ obstruction with gross left Hydronephrosis with parenchymal atrophy, areas of bleeding and clots in left pelvicalyceal system and right contracted kidney (Fig 10, 11). Left PCN insertion was done which drained 2000 mL of dark coloured fluid on first day, and 500mL haematuria daily for next 48 hrs. In view of persistent haematuria and anaemia patient underwent emergency left nephrectomy on post op day 3. Patient was discharged in stable condition and diagnosed as chronic kidney disease and is on regular maintenance. Haemodialysis and is being worked up for renal transplant at 10 months follow-up.



**Fig 9: Tense distended abdomen**



**Fig 10,11 : CT Scan of Giant Hydronephrotic kidney with clots in pelvicalyceal system with percutaneous Nephrostomy in situ**

**TABLE 1: DEMOGRAPHICS , TREATMENT AND COMPLICATIONS IN CASE SERIES.**

Case	Age/Gender	Clinical Features	Hydronephrotic Sac Size	Total Nephrostomy Drainage	Tc-99 DTPA Renogram	Treatment	Complication
1st	29 yrs/Male	Abdominal lump	39 x 26 cm	26000 ml	Non-functioning RT kidney, B/L UPJO	Right nephrectomy + Left Pyeloplasty	None
2nd	25 yrs/Male	Abdominal lump Flank pain	30 x 25 cm	20000 ml	Non-functioning kidney	Nephrectomy	None
3rd	34 yrs/Male	Abdominal lump hematuria Flank pain	28 x x25 cm	3000 ml	Non-functioning kidney	Emergency nephrectomy	Chronic kidney disease, ICU stay

**DISCUSSION:**

Hydronephrosis is defined as aseptic dilation of kidney caused by

obstruction. [1] When it reaches huge proportions it is termed as giant hydronephrosis. In 1939, Stirling defined it as collection of >1000 ml

of urine in pelvicalyceal system. [2] Crook et al defined giant hydronephrosis as mass crossing midline or occupying >5 vertebral bodies radiologically. [3] A kidney accounting for >1.6% of total body weight is also considered giant hydronephrosis. >600 cases have been reported so far in literature, but most of the cases are seen in children [4]. Males are more commonly affected and predominantly of left side for unknown reasons. [5] Most of the patients are diagnosed in the early childhood, but there are still cases which are diagnosed later, as in our case. [4] This entity is rarely seen in developed world but still observed in developing world. We present a case series of 3 cases of GH in adults as per Crook's criteria. Symptomatically it has a slowly progressive course often presenting as huge lump or vague symptoms like nausea, fatigue, dyspepsia etc. It can also present as chronic dull aching flank pain, haematuria, urinary tract infection, pyonephrosis, secondary renal stones or renal dysfunction. [6] Giant hydronephrosis is a great impersonator as it mimics abdominal ascites, ovarian cyst, mesenteric cyst, pancreatic pseudocyst, extra-renal tumour or hydatid cyst, which are the important differential diagnosis. [7] Congenital UPJ obstruction is the most common overall cause accounting for >75% cases. 10-40% cases have bilateral UPJO, as in our first case [8]. Glass in 1746 spotted the headmost case of largest hydronephrotic sac, containing 115 litres of fluid, in an autopsy of a 22-year-old female. [9]

The UPJ obstruction can either be intrinsic or extrinsic. Intrinsic is due to increased connective tissue and lack of smooth muscle. Other congenital causes include ureteric atresia, mucosal folds, polar or aberrant vessels leading to extrinsic obstruction etc. Rarely urolithiasis, upper tract urothelial cancer, ureterovesical junction obstruction or ureteric stricture should be considered as acquired causes for GH, depending on patients symptoms.

Ultrasonography is the initial diagnostic modality followed by CT scan of abdomen and diuretic renogram revealing GFR, differential function and drainage pattern. In diuretic Renogram differential function is usually overestimated in GH patients, due to large marking of region of interest. Pyeloplasty should be considered in patients with renal cortical thickness >5mm and/or differential function >15%. But generally in adults, evolving and implacable hydronephrosis commonly renders renal unit non-functional at the time of diagnosis. [8]

Management of GH has to be a 2 stage process. First initial decompression followed by definitive surgery. Large fluid with high pressure in obstructed kidney can lead to life threatening complications on decompression or trauma. Paracentesis induced circulatory dysfunction (PICD) is a hemodynamic imbalance caused by sudden weaning of abdominal pressure, increased preload with increased splanchnic circulation, and decreased systemic vascular resistance. Slow decompression of hydronephrotic sac to 5-6 litres a time with use of volume expanders can prevent this disastrous complication. [7] The preservation of renal parenchyma is the primary aim of management of GH [10]. But Hoffman proposed nephrectomy as the only feasible option for GH (in presence of normal contralateral kidney) as these kidneys are often non-functional [11]. Association of focal dysplasia secondary to chronic irritation and risk of malignancy is also reported [12]. Crooks et al. [3] reported 30% nephrectomy while Uson et al. [13] reported 70% nephrectomy rate in kidneys with GH but all in paediatric population. Reparative surgery in such cases often does not lead to melioration of renal function in adults. The two-stage procedure may help to preserve residual renal function.

To reduce stasis and improve drainage in functional renal units undergoing pyeloplasty Hemal et al proposed significance of nephroplication and nephropexy as an adjunctive procedure. It brings pelvicalyceal system laterally and in line with ureter. Foleys VY plasty, vertical/spiral flap repair or ureterocalycostomy can be performed in failed UPJ repair but, it can lead to anastomotic stricture. Boari flap calyceovesicostomy can prevent this complication. [14] In a study conducted by Kaura et al on GH patients, nephrectomy had a favourable outcome, as 3 pyeloplasties out of 10 performed on adult patients failed, while upfront nephrectomy was performed in 9 patients. No other significant complications were encountered in our case series as in their study. [15]

Regular follow-up after pyeloplasty is recommended including renal function tests and diuretic renal scan. Patients undergoing nephrectomy should also be followed with regular ultrasonography and serum creatinine values. Reparative surgery should always be attempted rather than primary nephrectomy, when in doubt. Renal

transplant should be considered in patients who progress to chronic kidney disease.

As per Consensus statement on management of antenatal detected hydronephrosis [16], infants with bilateral hydronephrosis, hydronephrosis in solitary kidney, symptomatic obstructive hydronephrosis, or with advancing renal impairment; surgical correction should be offered. With the regular use of ultrasound scan and antenatal diagnosis of hydronephrosis; appropriate management of hydronephrosis in post natal period is the standard of care. Therefore it is unlikely to encounter such large hydronephrosis in this era, where imaging modalities are easily available even in developing countries. A larger study with bigger sample size is needed to consolidate our findings, but since GH is less likely to be observed in future; our study may remain one of the favourite case series in adults.

## CONCLUSION:

High index of suspicion, detailed imaging are paramount in diagnosing GH. Early diagnosis can preserve renal function, especially in children; but in adults management of GH includes initial nephrostomy with slow decompression and followed by nephrectomy with regular follow-up.

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