# Neglected Ureteropelvic Junction Obstruction Leading to Giant Hydronephrosis: A Case Report and Review of Literature

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## Abstract

Giant hydronephrosis (GH) is an uncommon urological disease, and ureteropelvic junction obstruction (UPJO) is counted as one of the most common etiologies among children.

This report presents a giant hydronephrosis resulting from a neglected complication of ureteropelvic junction obstruction in a 2.5-year-old girl with a history of gross hematuria and abdominal distention who underwent nephrectomy.

A kidney that constitutes more than 1.6% of the overall body weight is counted as a giant hydronephrotic kidney. The possible differential diagnoses may include ovarian, mesenteric, pancreatic, and adrenal cysts, extra-renal tumors, Echinococcal cysts of the liver, non-renal retroperitoneal cysts, tuberculous peritonitis, and ascites. Due to the possibility of pyelonephritis, sepsis, and shock, paracentesis is not recommended in these cases. Ultrasonography is the primary modality employed for distinguishing the diagnoses mentioned above. Our survey of the pediatric population revealed that aside from UPJO, the most common anomalies were renal system duplication.

Care for patients with GH is tailored to each individual, with significant differences between children and adults.

## Keywords

- Hydronephrosis
- Ureteropelvic Junction Obstruction
- Hematuria
- CT scan
- Case report

## Introduction

The condition of Giant hydronephrosis (GH) is a rare one characterized by the gradual collection of over 1 liter of urine in the collecting system. Although it is an intra-abdominal mass, its primary symptom is asymptomatic, rather than abdominal distention, as it slowly progresses towards a GH.[1] Advances in diagnostic tools have resulted in a substantial decrease in the incidence of GHs in recent years. Diagnosing and GH management becomes important because it presents itself at first glance as a retroperitoneal mass that resembles sarcomas or tumors of adrenal or embryonal origin.

Symptomatic manifestation commonly arises in instances of complicating factors, such as infection or compression of nearby structures. Nephrectomy may be considered for pediatric patients if the affected kidney has experienced a decline in function exceeding 90% and the contralateral kidney is unaffected.[2] In this study, we discuss a complex and intriguing case of GH in a young girl who presented with gross hematuria and abdominal distension, which was initially misdiagnosed as bowel obstruction. Additionally, we conducted a thorough review of all similar pediatric cases reported to date. This work has been reported in line with the SCARE criteria.[3]

**Case presentation**

A two-and-a-half-year-old girl was presented to the pediatric hospital with complaints of gross hematuria and irritability that had been present for several days. Abdominal distension, nausea, and vomiting were also among the symptoms she was experiencing. No distinct urinary or bowel obstructions were present. The medical history was unremarkable except for the diagnosis of a hydronephrotic kidney through antenatal ultrasonography. The patient's physical parameters were assessed as body mass index of 17.4 kg/m2, blood pressure of 90 mmHg, pulse rate of 87 per minute, respiratory rate of 20 per minute, and temperature of 37.7 ℃. A palpable mass and highly sensitive abdominal distension were discovered in the physical examination. The hematologic analyses were found to be within the normal range. The blood and urine test results are displayed in **(Table 1)**.

**Table 1:** The laboratory analyses findings of the patient.

| Blood | | Urine | |
|---|---|---|---|
| Blood Urea Nitrogen | 14.4 mg/dL | Protein | Negative |
| Creatinine | 0.50 mg/dL | Red blood cell | Many/HPF |
| Total protein | 7.1 g/dL | White blood cell | 10-11/HPF |
| Albumin | 4.4 g/dL | | |
| Total Cholesterol | 161 mg/dL | | |

A plain abdominal x-ray showed a huge mass-like opacity occupying the entire abdomen, compressing the bowel loops in the left upper hemi-abdomen and an elevated left hemidiaphragm **(Figure 1)**.

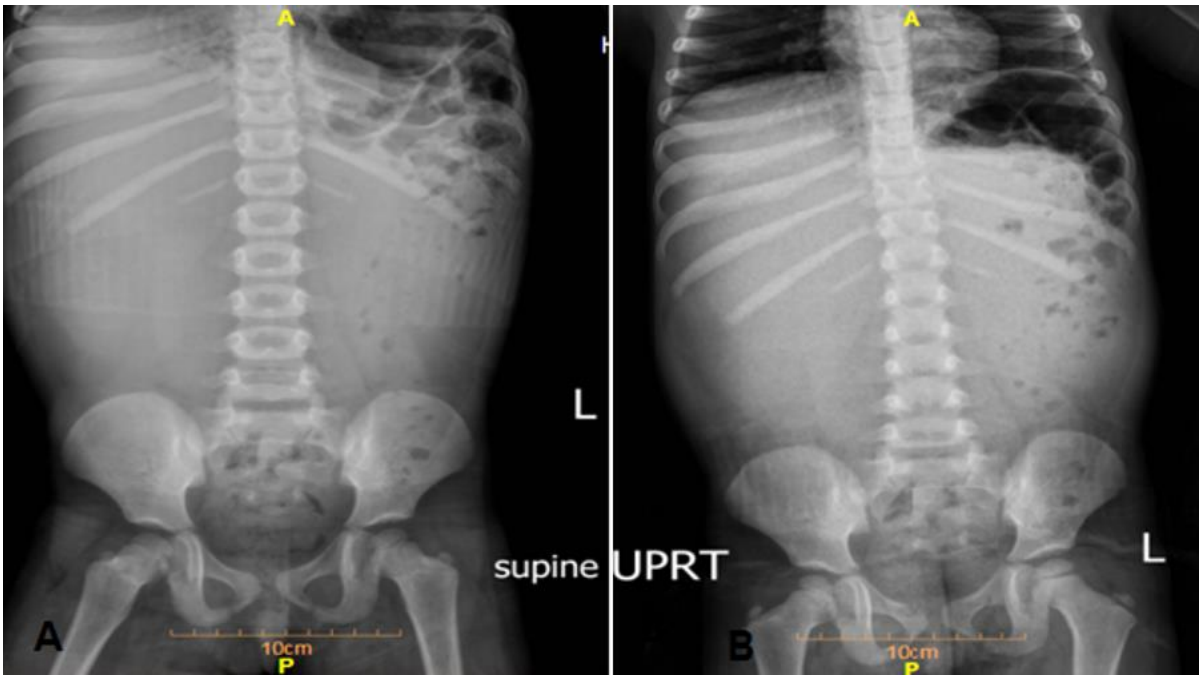


**Figure 1:** A substantial mass-like opacity occupying the entire abdomen with compressed bowls near the left upper hemi-abdomen and elevated left hemidiaphragm.

A septated cystic lesion with debris (17×11 cm2) was detected through ultrasonography, which extended from the right side to the midline of the abdominal cavity. The right kidney's delineation was impossible due to the cystic mass's abnormal size. Upon examination of a contrast-enhanced CT scan, a severe hydronephrotic change was discovered in the right kidney. The renal pelvis and calyces were significantly dilated, and the renal parenchyma was thinned, causing the bowel to be displaced to the left upper abdominal quadrant. The left kidney and urinary bladder both presented no remarkable findings. Furthermore, there were amounts of free fluid detected within the abdominopelvic cavity, as illustrated in **(Figure 2)**.

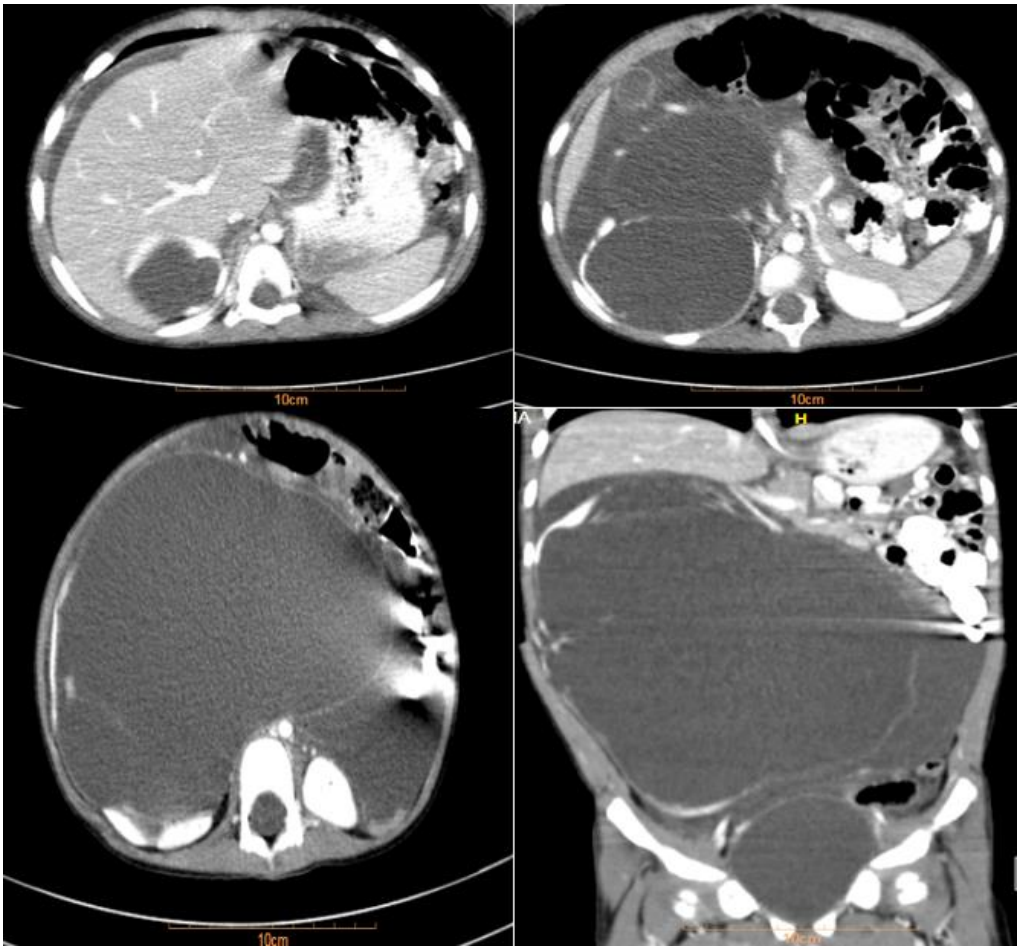


**Figure 2:** Abdominal CT shows a huge hydronephrotic change of the right kidney with thin renal parenchyma along the cystic wall. Free fluid was also seen in the abdomen.

The patient was scheduled for a laparotomy procedure to undergo a nephrectomy. The affected kidney with hydronephrosis displayed a significantly thinned cortex due to a massive cystic change containing an estimated quantity of more than two liters of fluid, and there was no adhesion with adjacent organs **(Figure 3)**.

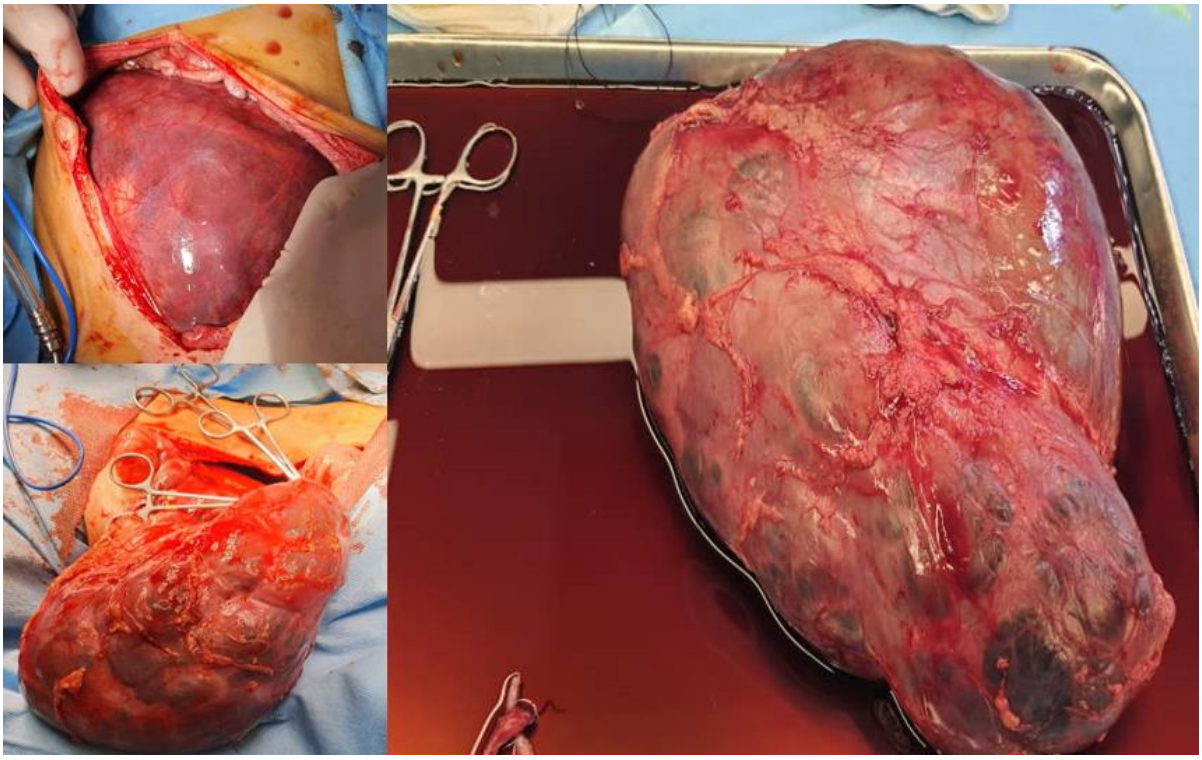

**Figure 3:** Right cystic kidney, weighting 225g, measuring 15*10*6 cm with a solid marginal rim of renal parenchyma (cortex), measuring 0.3 to 0.5 in thickness

It has been confirmed that UPJ obstruction is the cause of hydronephrosis. Dimensions of the left kidney were recorded at 15×10×6 cm3 with a weight of 225 g. The pathological specimens are displayed in **(Figure 4)**.

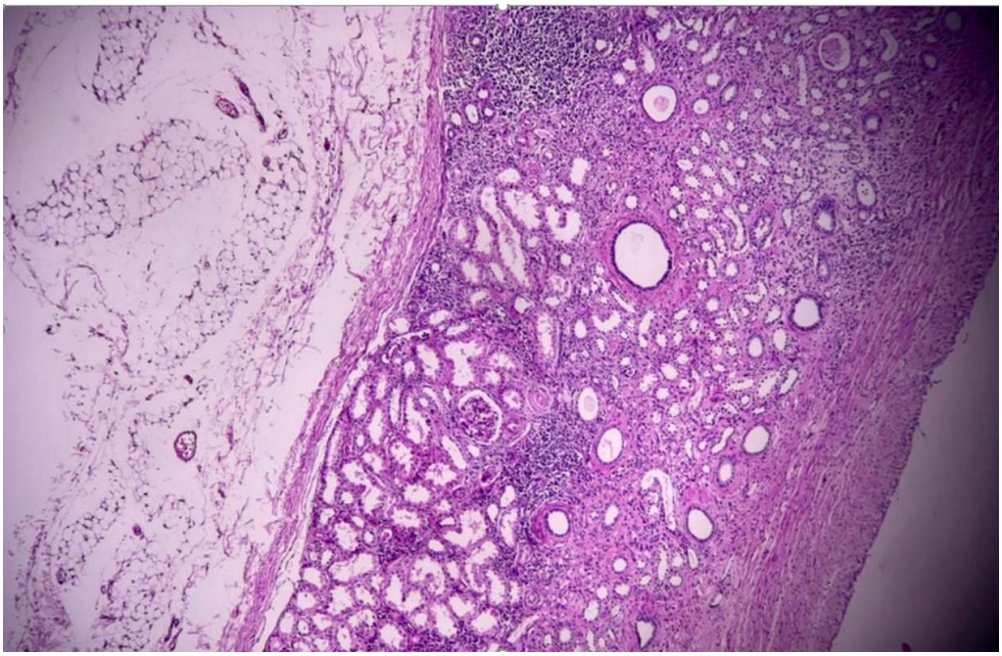

**Figure 4:** Histology examination of the kidney (Hematoxylin and eosin stain) showed a Narrow remaining cortex of cystic kidney with hemorrhagic focuses, lymphocytic infiltration, and fibrosis due to chronic inflammation in transitional tissue.

The patient was discharged three days post-operation without complications, having experienced a 4 kg weight loss. The patient's height and weight were measured three months after the surgery, revealing 150 cm (>75th percentile) and 57 kilograms (90-95th percentile), respectively, along with a BMI of 24.3 kg/m2 (>90th percentile). There is no longer any complaint from her regarding abdominal distension and hematuria.

## Discussion

The term GH has been present in medical literature since 1746; however, it was only in 1939 that Stirling defined GH, also known as massive hydronephrosis, as the accumulation of more than 1000 mL of fluid in the excretory system of a kidney. As a result, over 600 cases of this condition have been described globally.[4] A kidney that constitutes more than 1.6% of the overall body weight is also incorporated in this description.[5] However, these definitions have been for adults. In children, GH can be classified as hydronephrosis, ranging from 4% of body weight at birth to 2% at puberty.[6-7] Hydro nephrosis with variable sizes accounts for 1-5% of all pregnancies and is responsible for 50-87% of urinary tract abnormalities.[8-9] Perinatal hydronephrosis may be due to transient hydronephrosis and obstruction at the level of the ureteropelvic or the vesicoureteral junction, posterior urethral valves, or vesicoureteral reflux. In about 50% of instances, the issue is not problematic and resolves automatically following birth. Ureteropelvic junction obstruction (UPJO) represents the most common indication for intervention.[9] In over half of the cases, the obstruction is primarily functional rather than anatomical.[10] The possible differential diagnoses may include ovarian, mesenteric, pancreatic, and adrenal cysts, extra-renal tumors, Echinococcal cysts of the liver, non-renal retroperitoneal cysts, tuberculous peritonitis, and ascites. Due to the possibility of pyelonephritis, sepsis, and shock, paracentesis is not recommended in these cases. Ultrasonography is the primary modality employed for distinguishing the diagnoses mentioned above.

Nonetheless, there are situations where ultrasonography cannot determine the origin of a massive retroperitoneal cystic mass, in which scenarios remain unsolved on identifying which UPJO requires surgery prenatally.[11] Congenital abnormalities, such as UPJO and a duplicated collecting system, are the most common causes.[12] The subsequent grouping includes urinary stones, renal ectopia, trauma, ureterovesical junction obstruction, and, in rare cases, malignancies.[13] Our survey of the pediatric population revealed that aside from UPJO, the most common anomalies were renal system duplication[14-16], kidney atrophy or agenesis[14-17-19], ectopic kidney[11-19-20], congenital heart diseases[21], anorectal

malformations[22], Multicystic dysplastic kidney (MCDK)[23], and epidermoid cyst[16] **(Table 2)**.

**Table 2:** The summary of previous pediatric giant hydronephrosis cases reported up to now.

| Authors & year | Age & Sex | Clinical presentation | Imaging | Side of involvement | Surgical technique | Co-existing problems | Follow-up |
|---|---|---|---|---|---|---|---|
| 1. Modelski 1978 | 4/M | Left renal colic, fever, and pyuria | IVU: Giant hydronephrosis of the left good functioning kidney and no excretion of dye on the right side | L | Calyco-Sigmoido-Cystostomy in two stages | Right kidney agenesis | Improvement |
| 2. Modelski 1978 | 20/F | Abdominal pain and pyuria | IVU: Giant hydronephrosis of the left solitary left kidney<br>Cystoscopy: Absence of the right ureteral orifice<br>Aortography: paucity of the arterial network in the thin renal parenchyma | L | Calyco-Sigmoido-Cystostomy in two stages | Right kidney agenesis | Improvement |
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| 3. Brock et al. 1979 | 2/M | Abdominal mass | Intravenous urography: hydronephrotic left kidney with marked anterior displacement of the left ureter<br>poor visualization of the upper pole of the left kidney<br>Barium enema: marked displacement of the rectosigmoid<br>US and CT:a huge, septated, fluid-filled mass filling the entire | L | The upper pole segment and ureter excision | Duplicated left kidney inserting into the prostatic urethra. | Improvement |

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| | | | left abdomen and crossing the midline | | | | |
| 4. Brock et al. 1979 | 9/F | Abdominal enlargement | IVU: A partial right ureteral duplication with hydroureteronephrosis of both segments above the pelvic brim and a left duplication with a nonvisualized upper segment | R | Left upper pole heminephrectomy and partial ureterectomy | Right ureteral duplication, Atrophic upper pole of the left kidney | Improvement |
| 5. Brock et al. 1979 | 10m/M | Abdominal mass | IVU: right hydronephrosis with calyceal compression from a huge mass posterior to the kidney US: cystic nature CT: a fluidfilled mass occupying the entire right retroperitoneum, extending across the midline to impinge on the left mid ureter. | R | right upper pole heminephrectomy and ureterectomy | Right ureteral duplication very dilated and drained upper ureter besides an atrophic upper pole of the right kidney | ND |
| 6. Crooks et al. 1979 | 12/M | HTN | IVU: Non-functioning left kidney IVP: Massive left hydronephrosis | L | Temporary nephrostomy for 2 w Dismembering pyeloplasty | ND | ND |
| 7. Crooks et al. 1979 | 10/M | Abdominal pain, fever, and a left flank mass | IVP: a poorly functioning and massively dilated left kidney Voiding cystogram showed type 1 urethral valves, but no reflux Nephrostrogram: Obstructive megaureter. | L | Nephrostomy antibiotic treatment for 1 w The midureter was tapered and reimp[anted into the bladder with a long tunnel to prevent reflux | Pyelonephritis | ND |

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| 8. Crooks et al. 1979 | 9/F | Diagnosed prenataly | Cystography: vesicoureteral reflux | R | Pyeloplasty | ND | Seven-year disease free |
| 9. Crooks et al. 1979 | 1d/ND | Bilateral flank masses and distended bladder | Cystogram: vesicoureteral reflux and enormous dilatation of the bladder and both kidneys | B | The massively convoluted ureters were shortened and reimplanted and drained from below by catheter-stents Right nephrectomy | ND | ND |
| 10. Crooks et al. 1979 | 3w/M | Urinary tract reconstruction | Cytoscopy: entry of a solitary right ureter into the prostatic urethra | R | The ureter was removed from its abnormal location and reimplanted across the trigone with a long tunnel to prevent reflux Simultaneous dismembering pyeloplasty | Colostomy at birth for imperforate anus bladder infection from the rectourethral fistula Inguinal hernia | Seven-year disease free |
| 11. Kheradpir 1981 | 3/M | Abdominal mass | IVP | R | Ureter autotransplantation | The middle and lower section of the kidney revealed secretion after 5 minutes, and the upper section revealed no secretion. | Six-month disease free |

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| 12. Kheradpir 1981 | 4/F | Enlarged abdomen and fever | IVP: Delayed secretion and an enormously enlarged kidney Left atrophic hydronephrotic kidney | B | Ureter autotransplantation An Anderson-Hynes pyelo-plasty was later performed on the left side | UTI Mild left hydronephrosis | ND |
| 13. Kheradpir 1981 | 6/M | Gradually enlarged painful mass of the right flank | IVP | B | Ureter autotransplantation An Anderson-Hynes pyelo-plasty was later performed on the left side. | Mild left hydronephrosis | Improvement |
| 14. Macksood et al. 1983 | 10/M | Abdominal pain after trauma | IVP: a dilated calyceal system US: a giant hydronephrotic ectopic kidney | L | Nephrectomy | ND | Improvement |
| 15. Haque et al. 1985 | 10/M | abdominal distension | IVU: Dense nephrogram on right side of the abdomen and crossing the midline with nonvisualization of right ureter. Left kidney showed a pelviureteric junction obstruction with mild hydronephrosis. | R | Reduction pyeloplasty Right ureter was reimplanted by Hutch-I technique after excising the distal stenosed segment which on histological examination revealed fibromuscular dysplasia. | ND | Three-month disease free |
| 16. Dahniya et al. 1988 | 7/F | Abdominal distension and swelling of the legs for a year | IVU: no evidence of excretion by the right kidney and delay in excretion Right antegrade pyelography: enormous dilatation | B | preliminary slow decompression of the fight hydronephrosis, | ND | Lost to follow-up |

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| | | | of the renal pelvis and calyces. | | Right Anderson-Hynes pyeloplasty, Left Anderson-Hynes pyeloplasty after three months | | |
| 17. Dalton et al. 1988 | 5/F | Umbilical mass | US: a large cystic structure in the hypogastrium CT: An empty right renal fossa and a markedly hydronephrotic ectopic right kidney Voiding cystourethrography: vesicourethral reflux Radionuclide renal scan: urethropelvic junction obstruction in the ectopic kidney Cystoscopy: laterally displaced right urethral orifice in the "C" position with horseshoe configuration Right retrograde pyelography: short ureter, proximal narrowing, and UPJO | R | Dismembered pyeloplasty with pelvic reduction, nephrourethral stent insertion | ND | Improvement |
| 18. Tsukahara et al. 1991 | 4/M | Abdominal mass | US and IVU: A giant left-sided hydronephrosis Tc-DMSA: Moderately well-preserved left renal function Furosemide-enhanced Tc-DTPA: presence of a partially obstructive lesion in the left megaureter | L | Ureteroplasty | ND | ND |

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| 19. Hemal et al. 1999 | 4/M | Bilateral flank pain | Renal dynamic scan: non-functioning left | B | Retroperitoneal laparoscopic nephrectomy | ND | Three-month disease free |
| 20. Yilmaz et al. 2001 | 12/F | Painful abdominal distention | US: A huge cystic mass with multiple loculations occupying nearly the entire abdominal cavity MRI and contrast-enhanced CT: Giant hydronephrosis | R | Nephrostomy Right nephrectomy | Non-functioning right kidney | Four-month disease free |
| 21. Belman et al. 2006 | 1 m-5y/ F | ND | ND | ND | The Y-Pyeloplasty Pyeloplasty performed elsewhere (defined as a wide open UPJ as noted at time of kidney folding. | persistent severe hydronephrosis and recurrent pyelonephrosis that resolved after folding | Six-month disease free |
| 22. Belman et al. 2006 | 1 m-5y / ND | ND | ND | ND | The Y-Pyeloplasty | ND | Six-month disease free |
| 23. Belman et al. 2006 | 1 m-5y / ND | ND | ND | ND | The Y-Pyeloplasty | ND | Six-month disease free |
| 24. Belman et al. 2006 | 1 m-5y / ND | ND | ND | ND | The Y-Pyeloplasty | ND | Six-month disease free |
| 25. Belman et al. 2006 | 1 m-5y / ND | ND | ND | ND | The Y-Pyeloplasty | ND | Six-month disease free |
| 26. Kato et al. 2006 | 8/M | ND | ND | L | Pyeloplasty | UPJO | ND |
| 27. Kato et al. 2006 | 5m/F | ND | ND | R | Nephrostomy Pyeloplasty | UPJO | ND |
| 28. Kato et al. | 3/M | ND | ND | L | Nephrostomy | UPJO | ND |

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| 2006 | | | | | Pyeloplasty | | |
| 29. Kato et al. 2006 | 1/F | ND | ND | L | Nephrostomy Pyeloplasty Nephroplication | UPJO | Improvement |
| 30. Kato et al. 2006 | 2/M | ND | ND | R | Pyeloplasty Nephroplication | UPJO MCDK | Improvement |
| 31. Kato et al. 2006 | 4/M | ND | ND | R | Pyeloplasty Nephroplication | UPJO | Improvement |
| 32. Z˘upanc˘ic et al. 2006 | Mean age 8.1 years (range 2–14 years) | ND | ND | ND | Anderson-Hynes pyeloplasty and plication of the renal calyces | ND | Six-month disease free |
| 33. Z˘upanc˘ic et al. 2006 | Mean age 8.1 years (range 2–14 years) | ND | ND | ND | Anderson-Hynes pyeloplasty and plication of the renal calyces | ND | Six-month disease free |
| 34. Z˘upanc˘ic et al. 2006 | Mean age 8.1 years (range 2–14 years) | ND | ND | ND | Anderson-Hynes pyeloplasty and plication of the renal calyces | ND | Six-month disease free |
| 35. Z˘upanc˘ic et al. 2006 | Mean age 8.1 years (range 2–14 years) | ND | ND | ND | Anderson-Hynes pyeloplasty and plication of the renal calyces | ND | Six-month disease free |
| 36. Z˘upanc˘ic et al. 2006 | Mean age 8.1 years | ND | ND | ND | Anderson-Hynes pyeloplasty and | ND | Six-month disease free |

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| | (range 2–14 years) | | | | plication of the renal calyces | | |
| 37. Zˇupancˇic et al. 2006 | Mean age 8.1 years (range 2–14 years) | ND | ND | ND | Anderson-Hynes pyeloplasty and plication of the renal calyces | ND | Six-month disease free |
| 38. Zˇupancˇic et al. 2006 | Mean age 8.1 years (range 2–14 years) | ND | ND | ND | Anderson-Hynes pyeloplasty and plication of the renal calyces | ND | Six-month disease free |
| 39. Zˇupancˇic et al. 2006 | Mean age 8.1 years (range 2–14 years) | ND | ND | ND | Anderson-Hynes pyeloplasty and plication of the renal calyces | ND | Six-month disease free |
| 40. Zˇupancˇic et al. 2006 | Mean age 8.1 years (range 2–14 years) | ND | ND | ND | Anderson-Hynes pyeloplasty and plication of the renal calyces | ND | Six-month disease free |
| 41. Zˇupancˇic et al. 2006 | Mean age 8.1 years (range 2–14 years) | ND | ND | ND | Anderson-Hynes pyeloplasty and plication of the renal calyces | ND | Six-month disease free |

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| 42. Kojima et al. 2007 | 8/F | Abdominal pain | US: Grade 4 right hydronephrosis<br>CT: Horseshoe kidney and advanced hydronephrosis<br>99mTc-MAG3 Retrograde pyelography: UPJO | R | Laparoscopic nephrectomy | ND | Nine-month disease free |
| 43. Reynolds et al. 2007 | 13/F | Palpable abdominal mass | US: a 16-cm cystic structure of the left abdomen with a thin rim of parenchymal tissue<br>MRI: A giant cystic mass occupying the entire left abdomen<br>Renal scintigraphy: complete absence of perfusion to the left kidney, despite the healthy appearance after nephrostomy | L | Nephrostomy | Right-sided renal duplication The nephrostomy tube was found entering an epidermoid cyst cavity of the spleen | ND |
| 44. Yapanoglu et al. 2007 | 19/F | Abdominal nausea and mild abdominal pains | US: a huge right cystic mass occupying nearly the entire abdominal cavity and compensatory hypertrophy of the left kidney with normal parenchymal thickness<br>CT: right septated retroperitoneal cystic lesion occupying the whole of the abdomen extending the pelvis | R | Nephrectomy | Incomplete ureteral duplication | Improvement |
| 45. Augustin et al. 2009 | 7/M | Abdominal distention | Ultrasound: a giant cystic right kidney measuring 224mm with a pyelon width of 129mm and particular calyces of up to 42 mm besides absent left kidney excretory urography Voiding cystourethrogram | R | Suprapubic percutaneous bladder catheterization over several months Anderson-Hynes pyeloplasty | ND | Improvement |

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| | | | Preoperative 99mTc-DMSA scan a reduction in the lower pole with extreme pyelocalyceal dilatation in the middle part with resulting discontinuity of renal parenchyma | | with calyceal plication | | |
| 46. Zaffanello et al. 2009 | 6/M | Abdominal pain | US: Enlargement of the left kidney, pelvis and ureter Doppler of the left renal artery: enhanced resistive index MAG-3: a dilated left kidney with reduced scintigraphic uptake and delayed radioactive urine formation MRI: enlargement of the left kidney with marked reduction of the cortical thickness | L | Surgical correction of the UVJ obstruction | ND | Three-month disease free |
| 47. Bawa et al. 2012 | 5/M | Abdominal distension | contrast-enhanced CT scan: a large complex cystic mass of size 185 × 148 × 300 mm occupying almost the entire left side of the abdomen and extending to the right side | L | Left-sided nephroureterectomy was performed | ND | ND |
| 48. Hsieh et al. 2012 | 7/F | Abdominal fullness | US: A large cystic mass localized in the left abdomen MRI & MRU: A very large, lobulated cystic lesion in the abdomen, probably arising from the retroperitoneum. A small pelvic atrophic kidney DMSA: Dysplastic ectopic pelvic kidney | L | Temporary percutaneous nephrostomy Nephrectomy | ND | Improvement |

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| | | | voiding cystourethrography: NL<br>Cystoscopy: a normal bladder and the right ureteral orifice and an absent left ureteral orifice | | | | |
| 49. Noh et al. 2012 | 3m/M | Diagnosed prenatally | US: severe left hydronephrosis and moderate to severe right hydronephrosis<br>Diuretic renogram: delayed drainage of the left kidney | B | laparoscopic left pyeloplasty | ND | Two-year disease free |
| 50. Hwang et al. 2017 | 11/M | Abdominal distension and dyspnea on exercise | Plain abdominal x-ray: Huge abdominal mass effect<br>US: a large septated cystic lesion (20.1×13.6 cm2) | L | Laparoscopic nephrectomy | Asthma | Three-month disease free |
| 51. Aihole et al. 2018 | 0/ND | Tense abdominal distension with respiratory distress | US: GH | R | A dismembered pyeloplasty (Anderson–Hynes), Nephrostomy | UPJO Echocardiography revealed moderate PDA and PFO with left to right shunt | One-year disease free |
| 52. Masarwa et al. 2016 | 5d/M | Intestinal obstruction, massive abdominal swelling, orally feeding intolerance, and intermittent vomiting | Prenatal US: moderate bilateral hydronephrosis | L | Left pyelostomy | Abnormal renal function, hyperkalemia, and acidosis | Three-month disease free |
| 53. Mehta et al. 2017 | 19/F | Pain | US: left gross hydroureteronephrosis<br>MRI: grossly dilated left renal pelvis and a dilated tortuous ureter<br>DTPA: Poorly functioning left kidney | L | Left simple nephrectomy | hydronephrotic kidney with UPJO. | Improvement |

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| 54. Kamath et al. 2018 | 10/M | Abdominal distension | US: large heterogeneous mass in the lower abdomen with septation and internal echoes<br>CT: bilateral hydronephrosis with the right side being massively dilated with septation, gross thinning of renal parenchyma and dilatation of the pelvicalyceal system | B | Right-sided percutaneous nephrostomy<br>Left nephrostomy was done 2 weeks later followed by improvement in renal functions.<br>Right Anderson–Hynes pyeloplasty after four weeks.<br>Left pyeloplasty was carried out in another 2 weeks with bilateral double J ureteric stent placement<br>Revision pyeloplasty later using a flap from the renal pelvis in view of failure of the right pyeloplasty | HTN | Six-month disease free |
| 55. Nehemaan et al. 2019 | 20m/M | Irritability, vomiting, and fever | US: A large multilobular cystic structure in the right hemi-abdomen<br>CT: huge hydronephrotic kidney crossing the midline, causing a mass effect<br>DMSA: 30% function on the affected kidney | R | Nephrostomy<br>Robot-assisted laparoscopic pyeloplasty | Septic shock<br>Inferior vena caval hrombosis | four-month disease free |
| 56. Para et al. 2019 | 19/F | Abdominal distension and fullness | US: Huge retroperitoneal cystic swelling occupying | R | Pigtail catheterization | ND | Improvement |

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| | | | the whole of right retroperitoneum and crossing the midline to left | | | | |
| 57. Asanad et al. 2020 | 7w/F | Tachypnea and abdominal distension | US: 16 cm cystic mass in the right abdomen. MRI: Massive hydronephrosis with parenchymal thinning. | R | Percutaneous nephrostomy Open pyeloplasty | The patient's lips appeared hypoxic when lying supine, which improved when rotated on her side, concerning for intermittent compression of the IVC | Improvement |
| 58. Boulic et al. 2021 | 8/M | Incidentally found with oliguria | US: Massive pyelo-calyceal dilatation in favor of ureteropelvic junctionstenosis Uroscan: significant right renal dilatation with enlarged pelvis measured at 58 mm, associated with the almost destruction of the parenchyma MAG3: asymmetrical renal function, with normal left kidney function | R | Right pyeloplasty with transposition of right lower polar vessels by robot-assisted trans-peritoneal laparoscopy with a JJ probe left in-situ for a month | adenoidectomy with trans-tympanic aerators placement, two episodes of bronchiolitis hemangioma treated with beta-blockers, pneumonia and asthma | Improvement |
| 59. Chabani-Cheballah et al. 2021 | Neonate/ M | Abdominal distension | US: a cystic mass situated at, or above the upper pole of the kidney (anteroposterior diameter of 55 mm), The lower pole was in a completely horizontal position. | L | Heminephroureterectomy | Duplicated renal system | Improvement |

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| | | | MRI: confirmed the renal origin of the lesion, showing a thin border of renal parenchyma around the outer part of the cyst and a normal adrenal gland<br>99mTc-MAG3 dynamic renogram: a non-functioning upper pole<br>cystoscopy: two left ureteric orifices | | | | |
| 60. Frech-Dörfler et al. 2022 | Neonate/ M | Diagnosed prenatally | US: First, giant hydronephrosis with a rim of normal parenchyma of the left kidney, Two kidneys with normal parenchyma and of normal size after nephrostomy | B | Bilateral percutaneous nephrostomy | Progressive respiratory failure that prompted immediate intubation at birth | Improvement |
| 61. Al-Hajjaj 2022 | 7/M | Abdominal pain | CT: a poor defined right renal outline with a thinning of the renal cortex besides a large intra-abdominal and retroperiteniuom fluid collection | R | Right nephrectomy | Retroperitoneal hematoma | Six-month disease free |
| 62. Jayakumar et al. 2022 | 13/F | Abdominal distention | US: giant cystic structure in the left renal fossa and a normal right kidney and bladder<br>CT: giant hydronephrosis of the left kidney<br>LLEC diuretic renogram: Poorly functioning left kidney | L | Laparoscopic left nephrectomy | ND | six-month disease free |
| 63. Our case | 2/F | Gross hematuria, abdominal pain, distension, and vomiting | US: the large septated cystic lesion with debris (17×11 cm2) expanding from the right side to the midline of the abdomen<br>CT: a severe hydronephrotic | R | Laparoscopic right nephrectomy | Peritoneal free fluid | Improvement |

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|---|---|---|---|---|---|---|---|
| | | | change of the right kidney, with markedly dilated renal pelvis and calyces with thinning of renal parenchyma displacing the bowls to the left upper abdominal quadrant | | | | |

The detection of more than 50% of congenital hydronephroses, including UPJO, before the manifestation of clinical symptoms has been made possible due to the widespread use of prenatal ultrasonography.[24] GH can result in complications such as hypertension, kidney rupture (as seen in our case), malignant changes, and renal failure if not diagnosed.[25] The decision concerning nephrectomy versus kidney-conserving therapy remains a critical issue for pediatric surgeons. Additionally, determining a hugely dilated kidney's exact renal function before surgery is complex, and reconstructive surgery can be highly challenging.

Accordingly, the preferred method in practice has been nephrectomy in most cases. Our case necessitated a nephrectomy for this reason. The Hoffman statement notes that nephrectomy is frequently the only viable option in non-functional and normal contralateral kidney cases. There is currently no established protocol for managing a poorly functioning kidney. Nonetheless, in 2010, research conducted in South Korea claimed that a kidney that functions at greater than 10% of its total capacity could be salvaged.[26] The thickness and echogenicity of renal parenchyma are essential predictors of renal function, while dysplastic parenchyma in severe cases often indicates a poor prognosis.[27]

## Conclusion

In light of the absence of international consensus on its management, care for patients with GH is tailored to each individual, with significant differences between children and adults.

## Ethical Consideration

This issue has been raised and approved by the ethics committee of Mashahd University of Medical Sciences, Iran.

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## Conflict of interests

There is no conflict of interest

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