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# Giant Hydronephrosis Due to Congenital Ureteropelvic Junction Obstruction

*Volkan Bulut, Gökhan Koç, Ali Feyzullah Sahin, Yavuz Balaban, Isık Hasan Özgü*
Urology Department, Tepecik Training and Research Hospital, Izmir, Turkey
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## ABSTRACT

Giant hydronephrosis caused by congenital ureteropelvic junction obstruction is very rare, particularly now that imaging techniques are more widely available. We introduce a 16-year-old boy who presented with abdominal pain and distention. He had a cystic mass in the central and right side of the retroperitoneum that filled the space from the right subdiaphragmatic area superiorly to the pelvis inferiorly. It measured 35 cm x 23 cm x 20 cm. His pelvic capacity was 8050 mL. The parenchyma of the right kidney was not observed; the left parenchyma was normal. We performed a right thoracoabdominal nephrectomy. Early diagnosis is essential to the prevention of this disorder.

**KEYWORDS**: Giant hydronephrosis; Ureteropelvic junction
**CORRESPONDENCE**: Dr. Volkan Bulut, Urology Department, Tepecik Training and Research Hospital, gaziler cad., Izmir, 35010, Turkey (vbulut79@hotmail.com).
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**Abbreviations and Acronyms**
CT, computed tomography
UPJ, ureteropelvic junction
VUCA 19-9, voided urine carbohydrate antigen 19-9

## INTRODUCTION

Ureteropelvic junction (UPJ) obstruction is defined as an insufficient amount of urine passing from the renal pelvis to the ureter. Most cases are related to congenital malformations. However, stone disease, postoperative inflammatory pathologies, and urothelial tumors are some acquired causes of this condition.

UPJ obstruction is generally diagnosed with abdomen ultrasound, excretory urography, computed tomography (CT), or magnetic resonance imaging during evaluation of a lumbar mass in infants and lumber pain or other symptoms in adults. Definitive diagnosis of UPJ obstruction can be done by diuretic mercaptoacetyltriglycine (MAG-3) and diethylenetriaminepentaacetic acid (DTPA) renal radioisotope examination.

Surgical indications for UPJ obstruction include severe impairment of renal function, hypertension, infection, and the presence of a stone. Surgical treatment can be done by open, endourological, or laparoscopic techniques [1].

Giant hydronephrosis caused by congenital UPJ obstruction is not a widespread urological condition. In 1939, Stirling defined it as the presence of more than 1,000 mL of fluid in the collecting system [2]. It is seen more often in females than in males (2.4:1) and more often on the left side than on the right side (1.8:1) [3]. Very large hydronephrosis resulting from a congenital malformation was more common before the development of ultrasound and other imaging techniques. However, this condition is still seen in developing and underdeveloped countries.

## CASE REPORT

A 16-year-old boy visited our clinic with complaints of abdominal pain and distention. His symptoms were present for approximately 2 weeks. A large mass that entirely filled the abdomen was found during the physical examination. Abdominal ultrasound and CT showed a very large, separated

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case report

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Figure 1. Coronal CT image of the hydronephrotic kidney.
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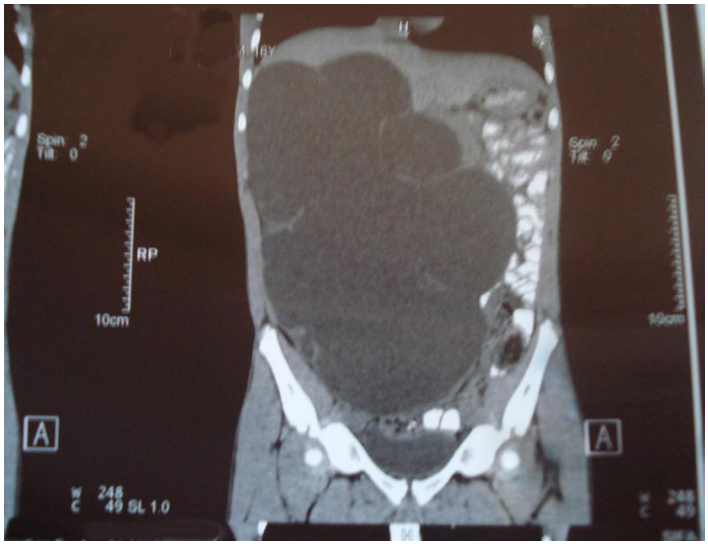


Figure 2. Axial CT image of the hydronephrotic kidney.
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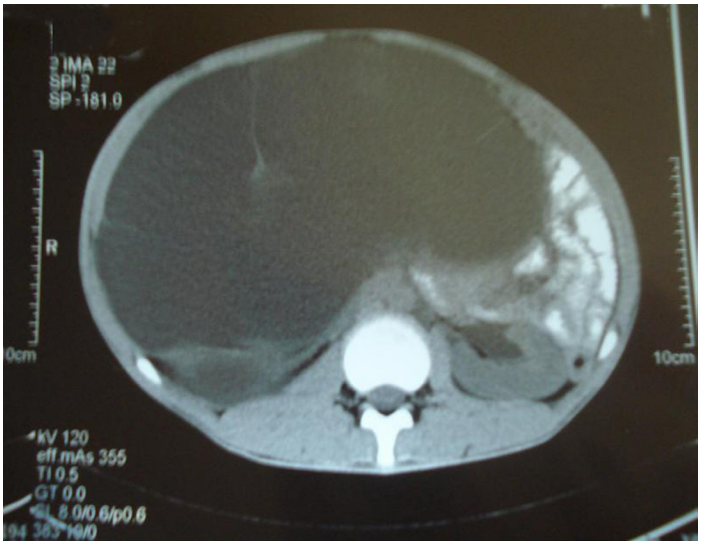


cystic mass in the central and right side of the retroperitoneum that filled all of the space from the right subdiaphragmatic area superiorly to the pelvis inferiorly. The mass measured 35 cm x 23 cm x 20 cm (Figure 1; Figure 2). The parenchyma of the right kidney was not observed; however, the left parenchyma was normal. The pelvic capacity was 8050 mL. Renal function studies were normal and no infection was detected.

Thoracoabdominal nephrectomy was performed and the mass was excised. An atretic ureter and an obstructed segment in the UPJ were seen during the operation. The ureter and the kidney were excised from the distal ureter together (Figure 3; Figure 4; Figure 5). Pathology showed chronic pyelonephritis, cystic pyelonephritic atrophy, and an atretic ureter. The patient was doing well when he was discharged from the hospital 5 days after the surgery. He had no complications during 3 years of follow-up examinations.

## DISCUSSION

UPJ obstruction is the main cause of giant hydronephrosis. We applied a right nephrectomy because the parenchyma of the kidney was not observed. Benchekroun et al [2] reported 2 cases with UPJ obstruction causing giant hydronephrosis. The treatment included pyeloplasty in the first case and nephrectomy in the second case. Onishi et al [4] reported that a 2-year-old patient with left hydronephrosis received a left nephrectomy. The pelvic capacity in this patient was approximately 7700 mL. In our patient, the pelvic capacity was 8050 mL; this was the largest capacity that we found of those reported in the literature. Chiang et al [3] described 4 patients with giant hydronephrosis. Their pelvic capacities were 1900 mL, 3400 mL, 2100 mL, and 3200 mL. Their second case developed acute renal failure and cardiopulmonary distress after the operation. The authors attributed these complications to the sudden decompression of the huge hydronephrotic sac, which resulted in a difference in the hemodynamic balance. Therefore they suggested slow decompression by percutaneous nephrostomy before the nephrectomy. A 40-year-old man with

Figure 3. Intraoperative photograph showing obstruction at the ureteropelvic junction (indicated by the arrow).
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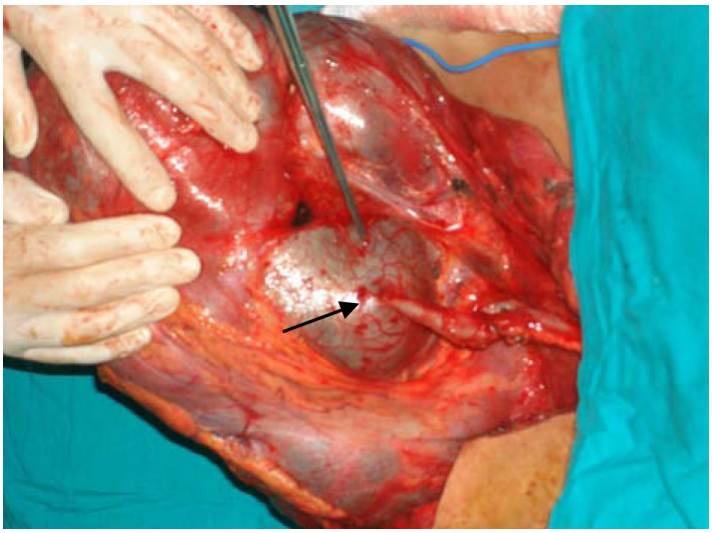

Figure 4. Photograph showing the length of the excised hydronephrotic kidney.
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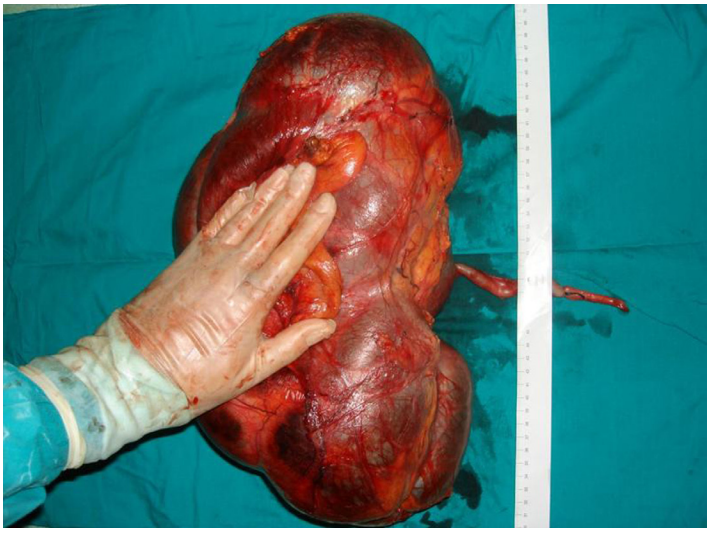

the complaint of right lower abdominal pain was reported by Kawai [5]. The patient had right giant hydronephrosis and UPJ obstruction caused by an aberrant artery. They decompressed by percuteneous nephrostomy before the pyeloplasty. Two years after the operation, the right renal function showed improvement with no complication of hypertension. Our patient was free of complications 3 years after surgery.

Figure 5. Photograph showing the width of the excised hydronephrotic kidney.
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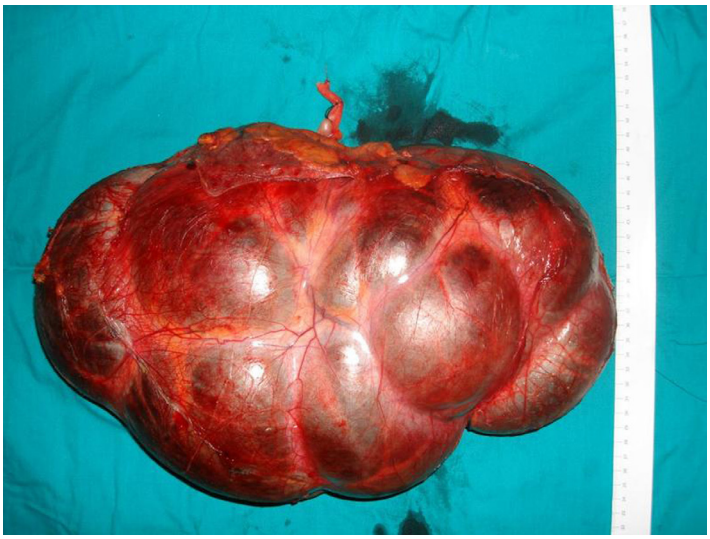

Early diagnosis is essential to the prevention of giant hydronephrosis. Voided urine carbohydrate antigen 19-9 (VUCA 19-9) is a noninvasive biomarker that has been used for the diagnosis and follow-up of patients with congenital obstructive nephropathy. The level of VUCA 19-9 was very high in patients with congenital obstructive nephropathy and a remarkable decrease was seen after pyeloplasty [6]. In the future, it can be speculated that early diagnosis and treatment may be possible by using this test routinely. More studies are needed.

## CONCLUSION

Giant hydronephrosis caused by congenital UPJ obstruction is a rare urological condition. Although some patients with congenital UPJ obstruction are diagnosed at young ages following severe symptoms, others are diagnosed with huge renal masses at later ages because their symptoms are mild. Early diagnosis of UPJ obstruction is essential and the VUCA 19-9 biomarker may be useful in this regard.

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