

GIANT HYDRONEPHROSIS DUE TO URETEROPELVIC JUNCTION STENOSIS IN PEDIATRICS: A RARE CASE

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ABSTRACT

Objective: This case report presented giant hydronephrosis (GH) due to Ureteropelvic Junction Stenosis in pediatrics. **Case(s) Presentation:** A three-year-old boy was admitted with abdominal distension and pain for the last three month. CT scan without contrast of the abdomen showed a left kidney size of 141x97x63.5 mm, massive dilatation of the pelvicalyceal system, thinning cortex in the left kidney, and left ureter measurement of 16 mm in diameter. An urgent percutaneous nephrostomy tube (10Fr) was placed in the left kidney for this patient, followed by a pyeloplasty procedure in the next three weeks. The patient was discharged on day five, and the six-month follow-up results were good. **Discussion:** Giant Hydronephrosis is a rare entity, with less than 500 global cases reported in the literature. The diagnostic approach to suspected hydronephrosis in pediatrics is sonography and may be followed by CT or MRI. Management of giant hydronephrosis requires two stages procedure with percutaneous nephrostomy followed by pyeloplasty. This two-stage method ensure the renal function is preserved. **Conclusion:** Ureteropelvic Junction Stenosis is a common cause of giant hydronephrosis in pediatrics. Appropriate physical examination and other diagnostic approaches are needed to diagnose patients with giant hydronephrosis and determine the best management.

Keywords: Giant hydronephrosis, ureteropelvic junction stenosis, pyeloplasty.

ABSTRAK

Tujuan: Laporan kasus ini menggambarkan hidronefrosis masif akibat stenosis ureteropelvic junction pada pediatri. **Presentasi Kasus:** Seorang anak laki-laki berusia tiga tahun dirawat dengan perut membesar dan nyeri selama tiga bulan terakhir. CT scan abdomen tanpa kontras menunjukkan ukuran ginjal kiri 141x97x63.5 mm, dilatasi sistem pelvicalyceal masif, korteks ginjal kiri menipis, dan diameter ureter kiri 16 mm. Tindakan darurat nefrostomi perkutan kiri (10 Fr) dilakukan pada pasien, diikuti dengan pieloplasti dalam tiga minggu berikutnya. Pasien dipulangkan pada hari kelima, dan hasil tindak lanjut selama enam bulan menunjukkan hasil yang baik. **Diskusi:** Hidronefrosis masif merupakan kasus yang jarang dengan kasus global kurang dari 500 yang dilaporkan dalam literatur. Pendekatan diagnosis hidronefrosis pada anak yaitu sonografi diikuti dengan pemeriksaan CT-Scan atau MRI. Manajemen GH membutuhkan 2 tahap prosedur dengan nefrostomi perkutan dilanjutkan pieloplasti. Metode ini memastikan fungsi ginjal tetap terjaga. **Simpulan:** Stenosis ureteropelvic junction adalah penyebab umum GH pada pediatri. Pemeriksaan fisik yang tepat dan pendekatan diagnostik lainnya diperlukan untuk mendiagnosis pasien dengan GH dan menentukan tata laksana terbaik.

Kata Kunci: Giant hydronephrosis, stenosis ureteropelvic junction, pieloplasti.

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INTRODUCTION

Hydronephrosis is the dilation of the pelvicalyceal system due to urine flow obstruction and stasis.¹ Giant Hydronephrosis (GH) is a kidney with greater than 1000 mL urine or 1.6% of total body weight in the renal collecting system. Radiologic imaging describes GH as an enlarged hydronephrotic kidney spanning the midline or

encompassing the whole hemiabdomen and extending for a length of five vertebral bodies.¹⁻²

Until recently, no GH case has been reported in Indonesia. Children are the most commonly affected by GH, and the most congenital cause of GH is ureteropelvic junction (UPJ) obstruction. Patients may have flank discomfort, hematuria, urinary tract infections, pyelonephritis, and renal failure, or they may be asymptomatic. If not detected and managed

early, GH can result in long-term complications, including renal failure, hypertension, and malignant transformation.² This case report presented GH due to Ureteropelvic Junction (UPJ) Stenosis in a three-year-old boy.

CASE(S) PRESENTATION

A three-year-old boy was admitted to the hospital with a chief complaint of abdominal distension for the last three months, accompanied by persistent abdominal pain but did not radiate. History of hematuria, trauma, and weight loss was denied. On physical examinations, abdominal distension was noted, and there was tenderness in the upper and lower left quadrants, whose percussion was dull (Figure 1). At the admission, complete blood count was normal, ureum was 23.5 mg/dL, and serum creatinine was 0.35 mg/dL.



Figure 1. Distended abdomen and tenderness.

CT scan without contrast of the abdomen showed a left kidney size of 141x 97x 63.5 mm, massive dilatation of the pelvicalyceal system, thinning cortex in the left kidney, and left ureter measurement of 16 mm in diameter (Figure 2).

An urgent percutaneous nephrostomy tube (10Fr) was placed in the left kidney for this patient to decompress the system, and more than 1 liter of urine was drained.

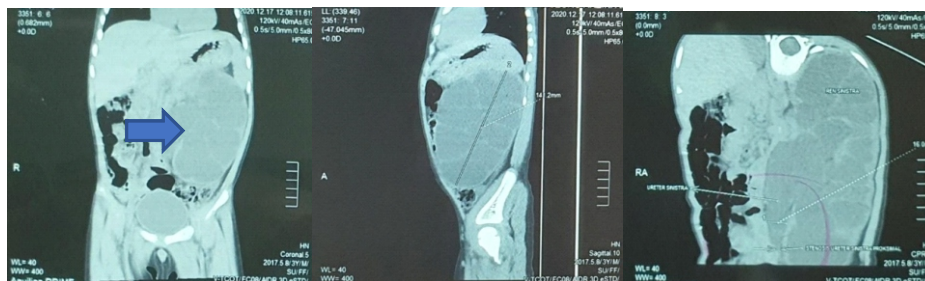


Figure 2. Sagittal and Coronal CT scan demonstrating giant hydronephrosis (blue arrow) on the left side and stenosis on the ureteropelvic junction (red arrow).

A creatinine clearance test was obtained one week later, and the result was 25.6 mL/min/1.73 m². The patient underwent an Anderson-Hynes pyeloplasty procedure three weeks later and left double J ureteric stent placement (Figure 3). The patient was discharged on day five, and the six-month follow-up showed good results.



Figure 3. Intraoperative view during pyeloplasty.

DISCUSSION

Giant hydronephrosis (GH) is commonly reported in pediatrics but rarely in adults.¹ In the literature, the most common cause of GH is ureteropelvic junction (UPJ) obstruction. The other causes are trauma, ureteral calculus, ischemia, carcinoma, and retroperitoneal fibrosis.² In this case, a narrowed segment of the ureter was observed at the ureteropelvic junction that causes giant hydronephrosis. Similarly, Naim et al. reported that the most common etiology of pediatric hydronephrosis was UPJ Stenosis (37.5%).³

Hydronephrosis progresses slowly, and most pediatrics appear to have clinically normal manifestations except for a palpable flank mass in some cases. Others may present back pain, hematuria, and urinary tract infection. In this case, the patient complained of distension and abdominal pain. The diagnostic approach to suspected hydronephrosis in pediatrics consists of sonography

as the first-line investigation and may be followed by CT or MRI for better differentiation of giant hydronephrosis from a cystic neoplasm or a large simple cyst.⁴ CT scan imaging was done on this case and described massive dilatation of the pelvicalyceal system, thinning cortex in the left kidney. Yapanoglu et al. reported the use of CT scan to diagnose GH resulted in mimicking intraabdominal mass; the right kidney weighed more than 5 kg and contained 5 liters of urine.⁵

Management of giant hydronephrosis requires a two-stage procedure with percutaneous nephrostomy followed by pyeloplasty.⁶⁻⁷ In this case, a percutaneous nephrostomy tube (10Fr) was placed in the left kidney for this patient to decompress the system. Creatinine clearance may become a reference for determining the management of giant hydronephrosis due to UPJ stenosis after percutaneous nephrostomy.⁸ The creatinine clearance result was 25.6 mL/min/1.73 m², so the patient underwent a pyeloplasty procedure. A similar approach was made by Agustin et al. on a seven-year-old patient with giant hydronephrosis in a single right kidney. This two-stage method ensures that renal function is preserved.⁹ In contrast with our case, Dino et al. reported that considering the risk of complications, a simple nephrectomy was advised for a patient with a non-functioning kidney and loss of renal parenchyma.¹⁰

CONCLUSION

Ureteropelvic Junction (UPJ) Stenosis is a common cause of giant hydronephrosis in pediatrics. Appropriate physical examination and other diagnostic approaches are needed to diagnose patients with giant hydronephrosis. A two-stage

procedure with nephrostomy followed by pyeloplasty helps to preserve residual renal function.

REFERENCES

1. Kaura KS, Kumar M, Sokhal AK, Gupta AK, Purkait B, Saini D, et al. Giant hydronephrosis: still a reality!. *Turk J Urol*. 2017; 43: 337-44.
2. Budigi B, Dyer RB. Giant" hydronephrosis. *Abdom Radiol (NY)*. 2019; 44(5): 1946-1948.
3. Naim HM, Daryanto B, Nurhadi P. Profile of Pediatric Hydronephrosis in Saiful Anwar General Hospital, Malang, Indonesia. *Indian Journal of Forensic Medicine & Toxicology*. 2021; 15(3).
4. Hu G, Luo M, Xu Y. Giant hydronephrosis secondary to ureteropelvic junction obstruction in adults: report of a case and review of literatures. *Int J Clin Exp Med*. 2015; 8(3): 4715-4717.
5. Yapanoglu T, Alper F, Ozbey I, Aksoy Y, Demirel A. Giant Hydronephrosis Mimicking an Intraabdominal Mass. *Turk J Med Sci*. 2007; 37(3): 177-179.
6. Kamath SP, Ganesh Pai K, Baliga BS. Bilateral Giant Hydronephrosis in a Ten-Year-Old Male. *Case Rep Pediatr*. 2018: 2470369.
7. Alsunbul A, Alzahrani T, Binjawhar A, Aldughiman A, El-Tholoth HS, Alzahrani A, Alakrash H. Giant hydronephrosis management in the Era of minimally invasive surgery: A case series. *Int J Surg Case Rep*. 2020; 75: 513-516.
8. Sharma U, Yadav S.S, Tomar V. Factors influencing recoverability of renal function after urinary diversion through percutaneous nephrostomy. *Urol Ann*. 2015; 7(4): 499-503.
9. Augustin G, Zupancic B, Zupancic V, Visnjic S. Calyceal plication with Anderson-Hynes pyeloplasty in a 7- year-old boy with giant hydronephrosis of a single right kidney. *European Journal of Pediatric Surgery*. 2009; 19(4): 255-257.
10. Dino MS, Hassen SM, Tufa TH. Unilateral Giant Hydronephrosis Secondary to Ureteropelvic Junction Obstruction in a Middle-Aged Woman. *Case Reports in Urology*. 2021: 1.