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Congenital ureteral valve as a diagnostically challenging congenital ureteral defect

Wrodzona zastawka moczowodu jako trudna diagnostycznie wada wrodzona moczowodu

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Abstract

The authors present two cases of ureteral valves. This rare urinary tract defect, causing abnormal urine flow from the upper urinary tract, is usually diagnosed intraoperatively. A 15-year-old girl with renal colic was admitted to hospital. Abdominal ultrasonography showed pelvicalyceal dilation in the right kidney and the X-ray showed a shadow in the orifice of the left ureter. She underwent bilateral ureterorenoscopy. A left ureteral valve was visualised during the examination. A 13-year-old boy was admitted to the Department of Surgery due to pelvicalyceal and left ureteral dilatation. Based on imaging and clinical findings, both patients underwent surgical treatment. Valves were found in the distal part of the left ureters. They were resected and the ureters were transplanted. Histopathological examination showed the presence of smooth muscle in the folds of the valve, confirming the final diagnosis.

Keywords: children, hydronephrosis, urinary system defects, ureteral valve

Streszczenie

Autorzy przedstawiają opisy dwóch przypadków zastawek moczowodu. Ta rzadka wada układu moczowego, powodująca zaburzenia spływu z górnych dróg moczowych, zwykle rozpoznawana jest śródoperacyjnie. Pacjentka 15-letnia z objawami kolki nerkowej zgłosiła się do szpitala. W badaniach obrazowych ultrasonografii jamy brzusznej uwidoczniono poszerzenie układu kielichowo-miedniczkowego nerki prawej, a na zdjęciu przeglądowym jamy brzusznej opisano słabo wysycony cień w ujściu pęcherzowym moczowodu lewego. Z powodu podejrzenia złogów w obu moczowodach pacjentkę zakwalifikowano do ureterorenoskopii. W trakcie kolejnego badania stwierdzono zastawkę moczowodu lewego. Drugi pacjent, 13-letni chłopiec, został przyjęty do Oddziału Chirurgii z powodu poszerzenia układu kielichowo-miedniczkowego oraz moczowodu lewego. Na podstawie badań diagnostycznych i obrazu klinicznego dzieci zakwalifikowano do leczenia operacyjnego. Śródoperacyjnie w obu odcinkach dystalnych moczowodów po stronie lewej stwierdzono zastawki, które resekowano, a moczowody przeszczepiono. Badanie histopatologiczne usuniętych odcinków moczowodów wykazało obecność mięśniówki gładkiej w fałdach zastawki, co pozwoliło na ostateczne potwierdzenie rozpoznania.

Słowa kluczowe: dzieci, wodonercze, wady układu moczowego, zastawka moczowodu

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INTRODUCTION

Congenital ureteral valve is a very rare cause of ureteral stricture, which can deteriorate kidney function. In the past, the diagnosis was made either postmortem or intraoperatively[1]. Wolfler was the first to describe the presence of a ureteral valve as a urinary tract defect in 1877. With advances in medical science, it started to be diagnosed based on histopathological findings. Currently, the defect can be suspected already at the stage of diagnostic work-up, i.e. radiological investigations, advanced ultrasonography (US), computed tomography (CT) and scintigraphy, with the exact location determined during ureterorenoscopy[2]. So far, about 70 cases congenital ureteral valve have been published in the world literature. The diagnostic criteria were proposed in 1952 by Wall and Wachter and include:

- presence of transverse folds of the ureteric mucosa containing bundles of smooth muscle fibre;
- no other evidence of mechanical or functional obstruction;
- signs of obstructive disease above the valve with a normal ureter below it[2–4].

Another classification was proposed by Rabinowitz[3,4]:

- type I – smooth muscle present within the folds;
- type II – smooth muscle at the base only.

Morphologically, three types of ureteral valves can be distinguished: cusp-like, diaphragmatic and annular type[3].

The pathophysiology is still unknown. It is believed that the valve may be a persistence of Chwalla's membrane or arise as a result of disturbances during embryogenesis[2,3,5]. Valve location in the ureter has been described as 50% in the proximal part, 17% in the middle third, and 33% in the distal third[3–5]. Cases of bilateral valve have also been reported[2].

Its rare occurrence makes the diagnosis challenging. Most cases are still diagnosed intraoperatively and then confirmed by histopathology. Diagnostic imaging, i.e. US or CT urogram (CTU), will show dilatation of the pelvicalyceal system and the ureters above the obstruction, while renal scintigraphy will show difficulties in urinary flow and will allow for renal function assessment. Voiding cystourethrography (VCUG) should be performed to exclude vesicoureteral reflux.

Surgical treatment preceded by diagnosis depends on kidney function and the location of the obstruction. In the case of proximal location, pelvic ureteroplasty is performed, while ureteral transplantation is used for valves located at the bladder outlet. In the case of mid ureteric valve, the stenotic segment is excised with a simultaneous uretero-ureteral anastomosis[1,4,6]. In recent years, endoscopic treatment with the use of electrocoagulation or a laser has been gaining in importance[7].

Ureteric valve was diagnosed in two patients at the Department of Paediatric Surgery and Paediatric Urology of the Centre of Postgraduate Medical Education between 2015 and 2022. Both patients were treated operatively. The ureteral segment containing the valve was resected during the surgery. Histopathology confirmed the diagnosis.

CASE REPORTS

Case 1

A 15-year-old girl with symptoms of right renal colic reported to the emergency department of children's hospital, where she had an abdominal US performed. Pelvicalyceal dilation in the right kidney was found, the AP diameter of the renal pelvis (RP) was up to 18 mm, renal calyces up to 6 mm; no abnormalities were found in the left kidney. No cause of stagnation was identified. Plain abdominal radiograph showed a poorly saturated, round shadow in the vesical orifice of the left ureter. Due to the suspicion of a stone, the girl was qualified for ureterorenoscopy with lithotripsy of stones in both ureters, during which a stone located in the distal right ureter was crushed with a pneumatic probe. Endoscopy of the left ureter was impossible due to its significant distal stricture. Postoperatively, the patient reported pain in the left lumbar region, which did not subside after analgesic and spasmolytic treatment. A decision to perform re-surgery was made. Ascending pyelography and ureterorenoscopy were performed, which revealed a 5-mm calcification and a ureteric valve in the area of the vesical orifice of the left ureter in its wall (Fig. 1). A double J catheter (or pig-tail catheter; placed in the ureter with one end in the renal pelvis and the other in the bladder) was placed to allow proper urinary drainage and prevent stagnation. As part of further diagnosis, scintigraphy was

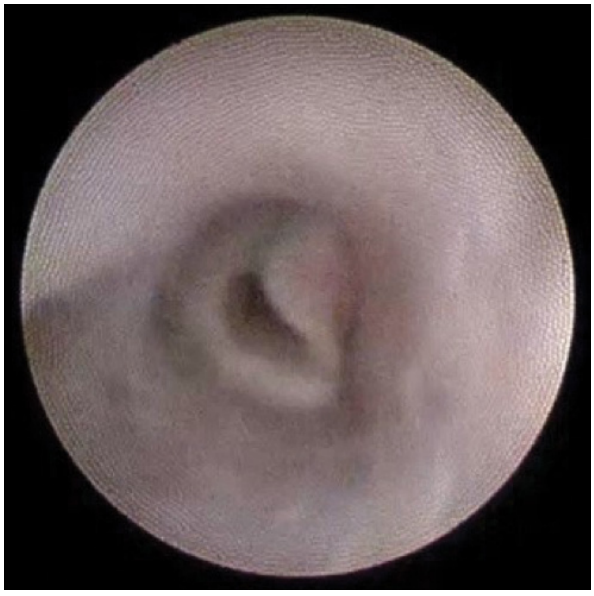

Fig. 1. Ureterorenoscopy of the left ureter, the ureteral valve was visualized (patient 1)

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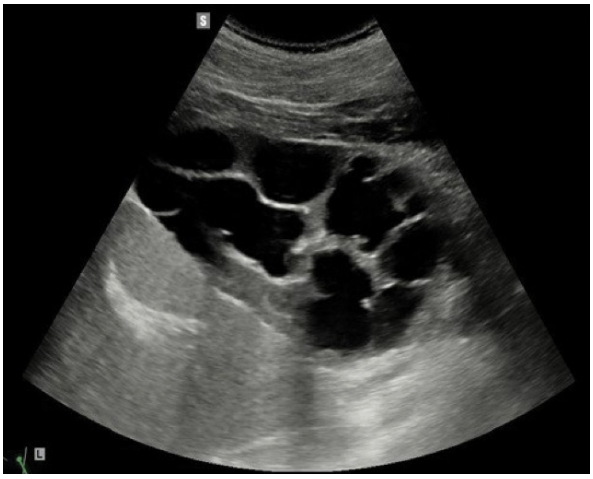

Fig. 2. Preoperative renal US, dilatation of the left renal pelvicalyceal system (patient 2)

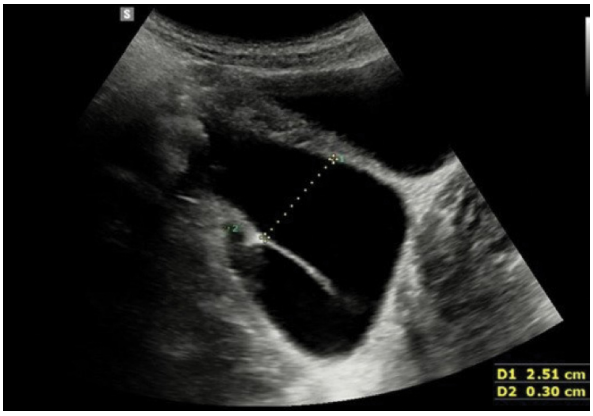

Fig. 3. Preoperative US of the ureter, left ureter dilatation (patient 2)

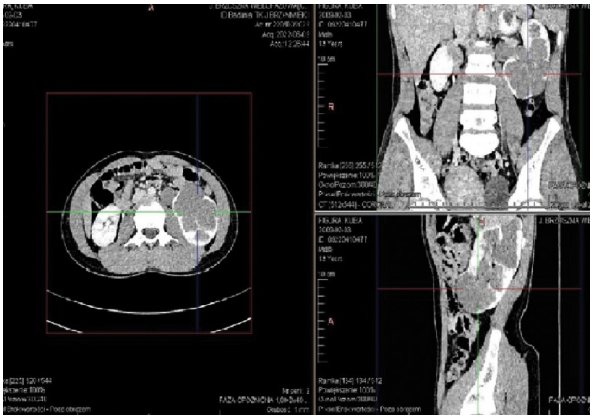

Fig. 4. Preoperative abdominal CTU, visible pelvicalyceal dilatation in the left kidney in three planes (patient 2)

performed, which revealed post-inflammatory changes in the left kidney. The secretory and excretory function of both kidneys was preserved, and the renal filtration function rate for the left and right kidneys was 49% and 51%, respectively. Abdominal US was unremarkable. The patient was qualified for valve resection with simultaneous Politano–Leadbetter ureteral reimplantation.

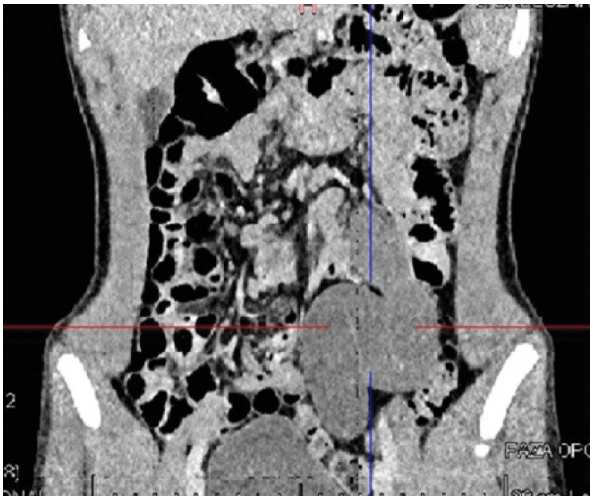

Fig. 5. Preoperative abdominal CT with urography showing left ureteral dilation (patient 2)

Histopathological findings confirmed the diagnosis. The postoperative period was uncomplicated. Follow-up renal scintigraphy and US were normal.

Case 2

A 13-year-old boy underwent an abdominal US due to periodic abdominal pain lasting for several months, which showed enlarged left kidney, pelvicalyceal enlargement; renal pelvis with AP diameter of up to 19 mm, subpelvic ureter up to 22 mm, other parenchymal organs unremarkable (Figs. 2, 3). The diagnosis was extended to include VCUG, during which no vesicoureteral reflux was found. Contrast-enhanced CT confirmed pelvicalyceal enlargement; renal pelvis AP diameter up to 33 mm, calyxes dilated up to 30 mm, renal parenchymal stenosis up to 4 mm, and a large 29–36 mm wide tortuous ureter, narrowing over a distance of about 2.5 cm from the bladder (Figs. 4, 5). Under general anaesthesia, an ascending pyelography was performed with simultaneous placement of a double J catheter. The patient underwent a dynamic kidney scintigraphy, showing good outflow from the right kidney, and abnormal tracer accumulation and its delayed clearance from the left kidney – left and right kidney renal filtration function rate was 38% and 62%, respectively (Fig. 6). Based on the above investigations, the patient was qualified for ureteral stricture removal and Politano–Leadbetter ureteral reimplantation. Due to the significant ureteral dilatation and thick ureteral wall, a decision was made to use the Hendren's technique. The resected pelvicalyceal segment of the ureter was cut open, exposing the valve (Figs. 7, 8). Histopathological findings confirmed the presence of smooth muscle in the folds of the valve. Follow-up renal scintigraphy (Fig. 9) showed improved excretory function of the left kidney, and abdominal US (Fig. 10) showed no pelvicalyceal or ureteral dilatation.

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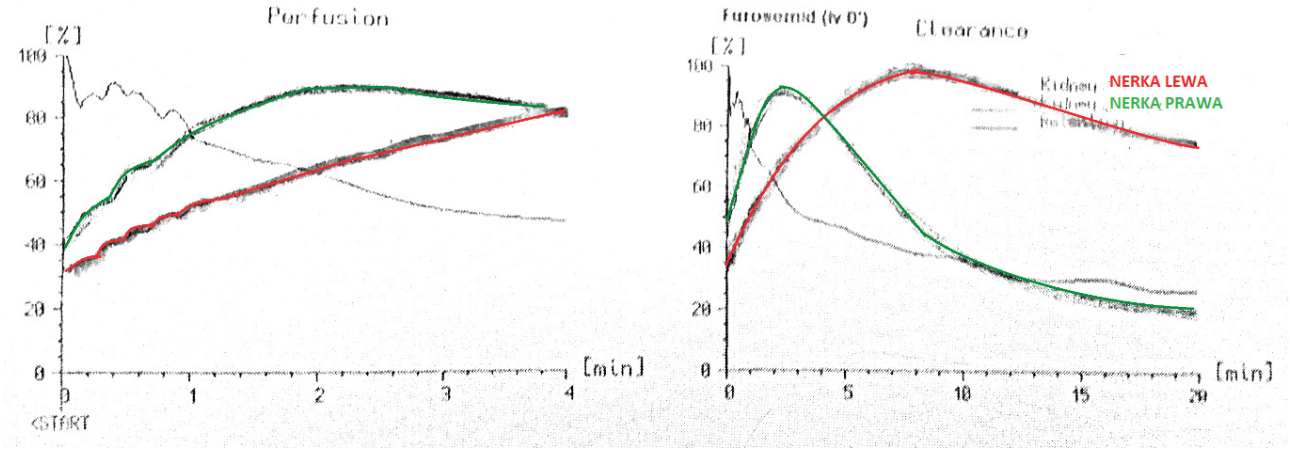

Fig. 6. Preoperative renal scintigraphy. The graphs show secretory and excretory functions of the kidneys (red line – left kidney, green line – right kidney) (patient 2)

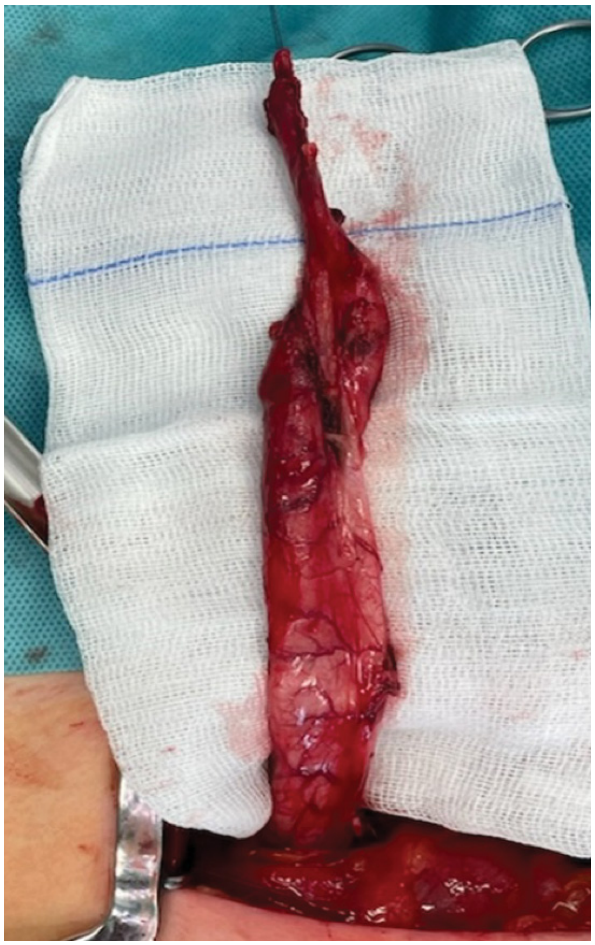

Fig. 7. Intraoperative image, a narrow perivesical segment and significant dilatation of the distal left ureter (patient 2)

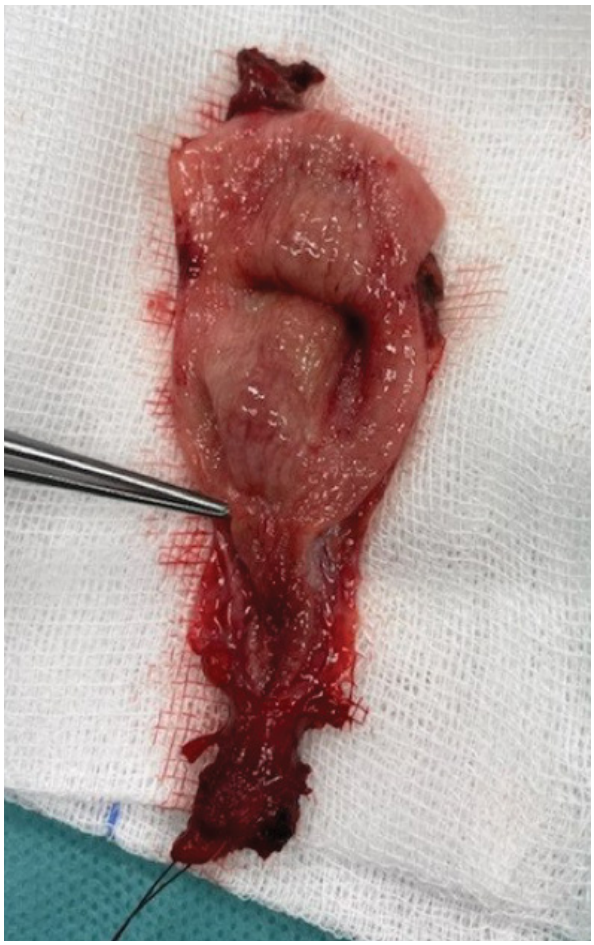

Fig. 8. Intraoperative image showing ureteral valve (patient 2)

DISCUSSION

Congenital ureteral valve is a rarely identified cause of ureteral stricture that can lead to deterioration of kidney function. Preoperative renal scintigraphy performed in both patients showed normal renal secretory function and worsening of the excretory function of the left kidney. About 70 cases of congenital ureteral valve have been described in the world literature so far. Ureteral valve coexists with other urinary tract defects, such as duplex kidney or duplicated ureter, vesicoureteral reflux, contralateral renal agenesis, and ectopic opening of the ureter,

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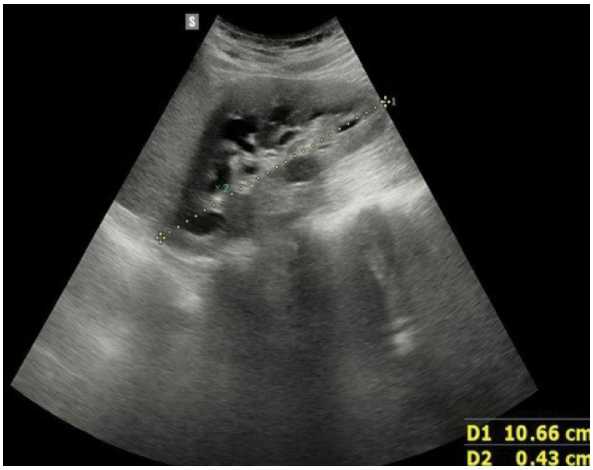

Fig. 9. Postoperative kidney US, no signs of pelvicalyceal dilatation in the left kidney (patient 2)

in more than 50% of patients[3,4,6]. In the described cases, none of the patients had an additional urinary tract defect. Symptoms are non-specific, with abdominal and lumbar pain, as well as urinary tract infections being most common[3,4]. Diagnostic imaging (US of the urinary tract and CTU) is used as an auxiliary investigation[4] and sometimes allows to detect urinary flow obstruction. Both children reported abdominal pain, which was an indication for diagnosis, during which abdominal US was performed, and additionally, CTU in the boy. The diagnosis was established during endoscopic procedure (ureterorenoscopy) in the girl, and intraoperatively during ureter transplantation in the boy. Both diagnoses were confirmed by histopathological analysis. We suggest that in the presence of this defect, the stenotic segment of the ureter with the valve should be excised. Surgical treatment depends on the kidney function and the location of the obstruction. In both cases, the valve was located in the perivesical area of the ureter; therefore a decision was made to transplant the ureters. The postoperative period was uncomplicated. Follow-up imaging showed no signs of pelvicalyceal or ureteral dilatation. Although the ureteral valve is a rare urinary tract malformation, it should be considered as one of the causes of hydronephrosis. Imaging investigations, such as US of the urinary system or CTU, are not always able to determine the cause of urinary flow disorders. The use of endoscopic methods in the future will allow for better diagnosis and minimally invasive treatment using the holmium: YAG laser[7].

CONCLUSIONS

Although ureteral valve is a rare urinary tract anomaly, it should always be considered in the diagnosis of urinary flow disorders. Surgical treatment was successful in both described cases. Removal of the affected ureteral segment along with the valve allowed for complete recovery and good long-term outcomes.

Conflict of interest

The authors report no financial or personal relationships with other individuals or organisations that could adversely affect the content of the publication and claim ownership of this publication.

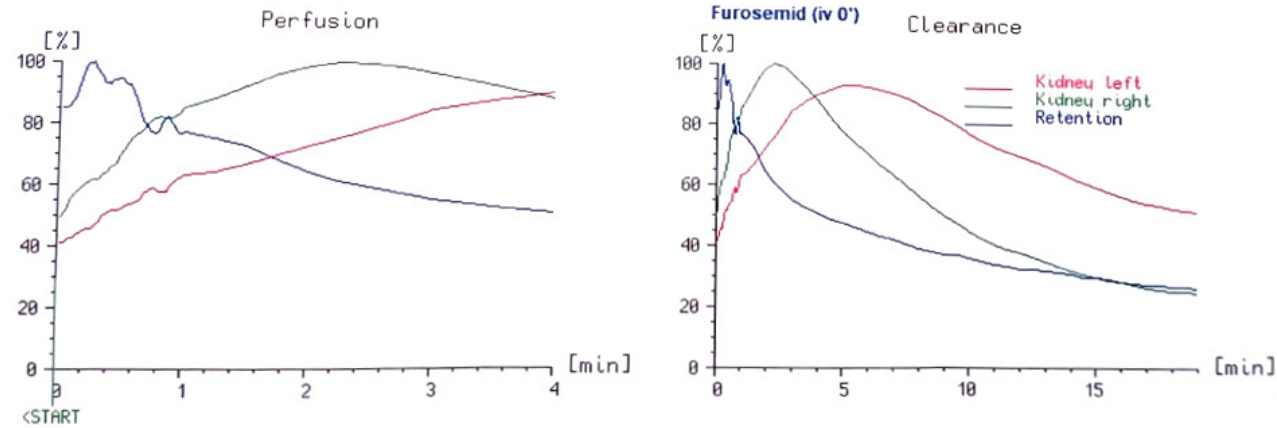

Fig. 10. Preoperative renal scintigraphy. The graphs show secretory and excretory functions of the kidneys (red line – left kidney, green line – right kidney) (patient 2)

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