

DOI: 10.21802/artm.2024.1.29.303  
UDC 616-089.843+616.61+616.13**AUTOTRANSPLANTATION WITH EXTRACORPORAL RECONSTRUCTION OF RENAL VESSELS: A CLINICAL CASE OF “THE NUTCRACKER SYNDROME” AND LEFT RENAL ARTERIES HYPOPLASIA COMBINATION**I.I. Kobza<sup>1</sup>, I.R. Nesterenko<sup>2</sup>, R.A. Zhuk<sup>3</sup>, Yu.S. Mota<sup>4</sup><sup>1</sup>*Department of Surgery №2, Lviv National Medical University,*<sup>2</sup>*Department of General and Vascular Surgery, Ivano-Frankivsk National Medical University, Department of Vascular Surgery, Municipal Noncommercial Enterprise Ivano-Frankivsk Regional Clinical Hospital of the Ivano-Frankivsk**Regional Council,*<sup>3</sup>*Department of Surgery №2, Lviv National Medical University,*<sup>4</sup>*Department of Surgery №2, Lviv National Medical University,**ORCID ID: 0000-0001-8493-414X, e-mail: ihor\_kobza@ukr.net**ORCID ID: 0000-0002-8358-291X, e-mail: nesterenko-irenka@ukr.net**ORCID ID: 0000-0003-3605-0862, e-mail: rostyslav.zhuk@gmail.com**ORCID ID: 0000-0002-6332-8404, e-mail: yuliamota@gmail.com*

**Abstract.** Ex vivo kidney autotransplantation with renal artery reconstruction is an effective method of surgical correction of vascular abnormalities (fibromuscular dysplasia, hypoplasia, and renal artery aneurysms) showing good long-term results. The combination of renal artery abnormalities with aorto-mesenteric compression syndrome (“the nutcracker” syndrome) is extremely rare and complicates the diagnosis making greatly. Renal artery hypoplasia is an abnormality that is rarely diagnosed and may not manifest clinically in the absence of contralateral kidney pathology as well as its vascular structures. Renal hypoplasia may be caused by mutations in the kidney development genes (HNF1B, PAX2, PBX1) and/or several environmental factors such as intrauterine growth retardation, maternal diseases (diabetes, hypertension), medication taken by the mother (renin-angiotensin system inhibitors or nonsteroidal anti-inflammatory drugs) and intoxication (smoking and alcohol). Premature delivery (before the 36<sup>th</sup> week) is also a risk factor due to incomplete nephrogenesis. Renal arteries hypoplasia is associated with fibromuscular dysplasia in most cases. The disease is characterized by non-inflammatory non-uniform focal hyperplasia of the renal artery wall with damage to small and medium arteries. Simultaneous damage to two or more vascular segments is detected in 25% of patients.

A clinical case of successful treatment of “the nutcracker” syndrome (annular renal vein) combined with hypoplasia of the left main and accessory renal arteries associated with fibromuscular dysplasia is described. A 27-year-old female was admitted to the Department of Vascular Surgery of Municipal Noncommercial Enterprise of the Lviv Regional Council “Lviv Regional Clinical Hospital” on October 4, 2021, with complaints of pain in the left lumbar region and the left half of the abdomen, hematuria, proteinuria, a persistent increase in blood pressure. Ultrasound of the renal veins: the left renal vein in the aorto-mesenteric segment was not visualized, the posterior portion of the left renal vein was determined. Peak systolic velocity was up to 230 cm/s at the point of confluence with the inferior vena cava and 28 cm/s more distally. The vein was up to 12.0 mm in the renal hilum and up to 11.0 mm in the middle third. MSCT of the abdominal cavity, retroperitoneal space and pelvic organs: CT signs of pyelonephritis of the left kidney. Hypoplasia of the left kidney. Reactive para-aortic lymphadenopathy on the left. Hypoplastic left renal artery (3,8 mm) and presence of accessory ones, annular left renal vein. Angiomyolipoma of the right kidney (2,6 cm). Varicose veins of the small pelvis on the left. The patient underwent surgical treatment, namely ex vivo autovenous prosthetic repair of the left renal artery with autotransplantation of the left kidney. The course of the postoperative period was without complications, the reconstruction was functioning. Ultrasound of the renal vessels (1 week after the surgery): the blood flow was located in the renal hilum in the course of Doppler sonography of intrarenal branches. Venous outflow was unobstructed. Corticomedullary differentiation was marked satisfactorily.

The study presented by us demonstrates the effectiveness of kidney autotransplantation for the correction of renal vascular abnormalities.

Though of a solitary nature, the combination of renal vessels abnormalities complicates significantly the course of the disease and the possibilities of this pathology diagnostics.

**Keywords:** renal artery hypoplasia, “the nutcracker” syndrome, surgical treatment, kidney autotransplantation, vascular compression syndrome, reimplantation of renal vein, ultrasonography, diagnostics.

**Introduction.** Ex vivo kidney autotransplantation with renal artery reconstruction is an effective method of surgical correction of vascular abnormalities (fibromuscular dysplasia, hypoplasia, and renal artery aneurysms) showing good long-term results [1,2]. Renal vessels abnormalities are rare (up to 0.1% in the general population and

0.3-2.5% according to the analysis of spiral computed tomography angiography data). Therefore, the experience of surgical treatment is limited to a small number of clinical cases in some centers of angiosurgery and kidney transplantation [3].

“The nutcracker syndrome” (NS) (left renal vein (LRV) compression between the aorta and the superior mesenteric artery at an acute angle of the origin of the latter) is a rare pathology, although phlebohypertension consequences in the LRV system constitute a significant medical and social problem [4,5].

The combination of renal artery abnormality and NS, namely aberrant renal artery stenosis in a 31-year-old female patient with the clinical findings of arterial hypertension is described only in one source according to the PubMed database [6].

We present a clinical case of successful treatment of NS (annular renal vein) combined with hypoplasia of the left main renal artery associated with fibromuscular dysplasia.

**The purpose of the study.** To increase the effectiveness of the treatment of “nutcracker syndrome” combined with renal artery hypoplasia by using kidney auto-transplantation with extracorporeal reconstruction of its arteries.

### Case Report

#### Patient Information

A 27-year-old female was admitted to the Department of Vascular Surgery of Municipal Noncommercial Enterprise of the Lviv Regional Council “Lviv Regional Clinical Hospital” on October 4, 2021, with complaints of pain in the left lumbar region and the left half of the abdomen, hematuria, proteinuria, a persistent increase in blood pressure, the presence of a permanent urinary catheter.

Past medical history: she considered herself a patient since September 2021, when uresiesthesia, pain in the left abdomen appeared, so a urethral catheter was installed. August 1, 2011 – left ureter ureteroneocystostomy according to Glenn-Anderson technique was conducted (due to stricture of the ureter). After a urologist’s consultation, the following diagnosis was made: neurogenic hyporeflex urinary bladder (a condition after ureterocystostomy in 2011), Grade III left-sided vesicoureteral reflux, left-sided ureterohydronephrosis, chronic left-sided pyelonephritis.

#### Clinical Findings and Diagnostic Assessment

Ultrasound of the renal veins on September 23, 2021: the left renal vein in the aorto-mesenteric segment was not visualized, the posterior portion of the left renal

vein was determined. Peak systolic velocity was up to 230 cm/s at the point of confluence with the inferior vena cava and 28 cm/s more distally. The vein was up to 12.0 mm in the renal hilum and up to 11.0 mm in the middle third.

MSCT of the abdominal cavity, retroperitoneal space and pelvic organs on September 3, 2021: CT signs of pyelonephritis of the left kidney. Hypoplasia of the left kidney. Reactive para-aortic lymphadenopathy on the left. Hypoplastic left renal artery (3,8 mm) and presense of accessory ones, annular left renal vein. Angiomyolipoma of the right kidney (2,6 cm). Varicose veins of the small pelvis on the left.

#### Surgical intervention

October 5, 2021 – the patient underwent surgical treatment under general anesthesia, namely ex vivo auto-venous prosthetic repair of the left renal artery with auto-transplantation of the left kidney.

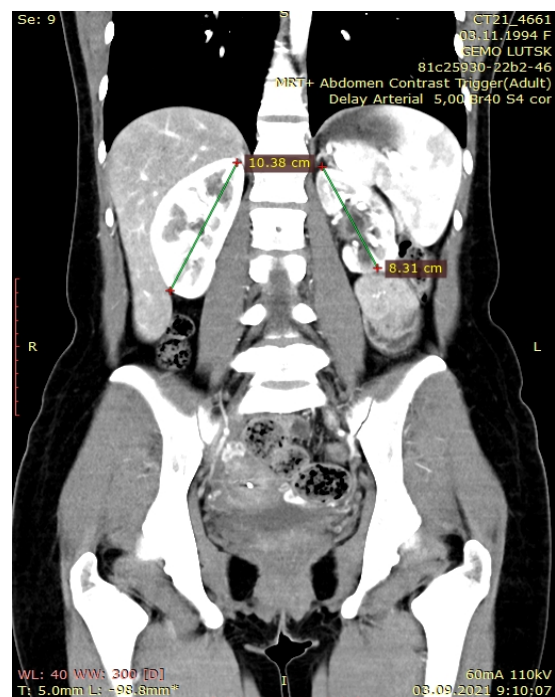


Figure 1. Hypoplastic left kidney with reduced accumulation of contrast material

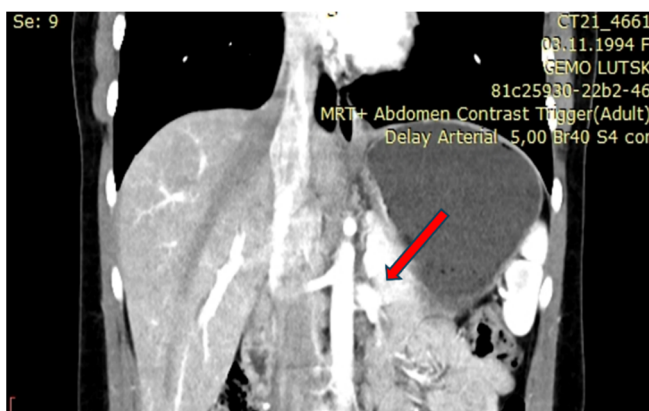
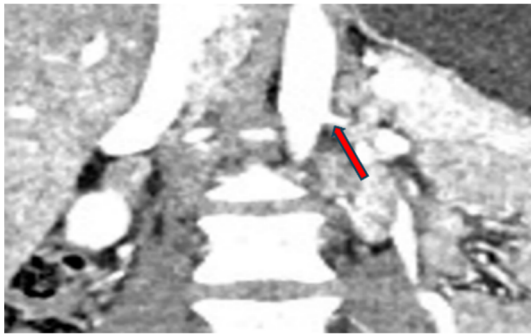


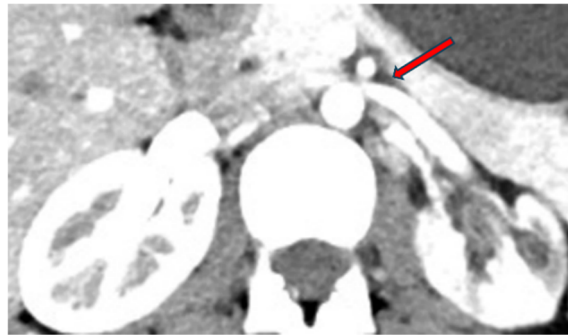
Figure 2. Absent contrast of the left renal artery



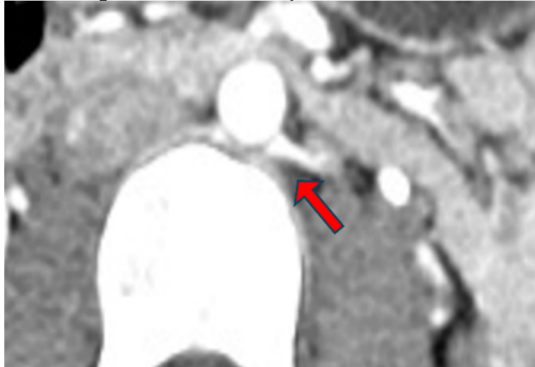
Figure 3. Fibromuscularly altered left accessory renal artery



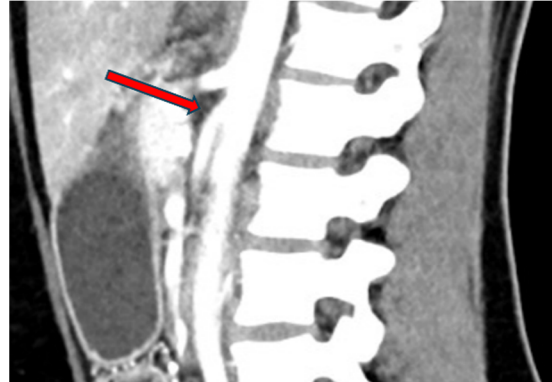
**Figure 4. Left accessory renal artery to inferior pole of the kidney**



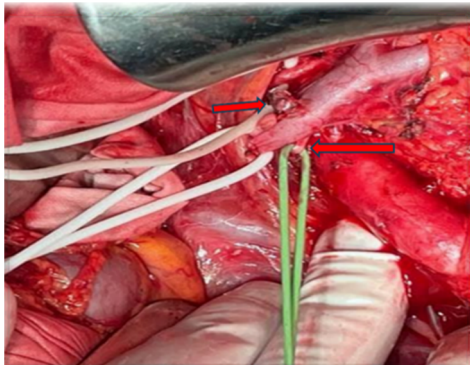
**Figure 5. Compression of LRV frontal portion**



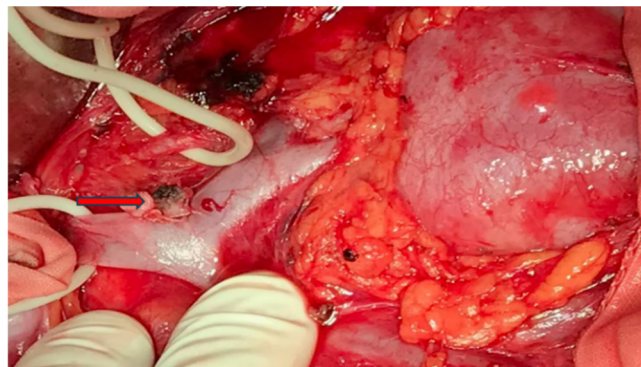
**Figure 6. Hypoplastic retroaortic portion of LRV**



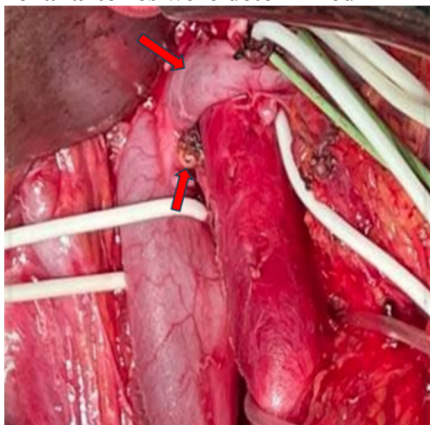
**Figure 7. Acute takeoff angle of superior mesenteric artery from the aorta ( $\leq 10^\circ$ )**



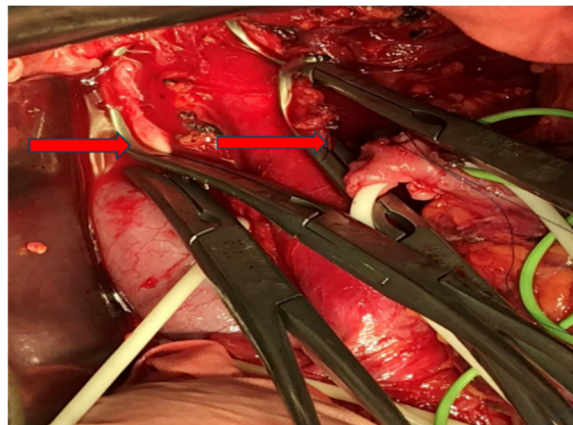
**Figure 8. The main (on the white holder) and accessory (on the green holder) renal arteries were determined**



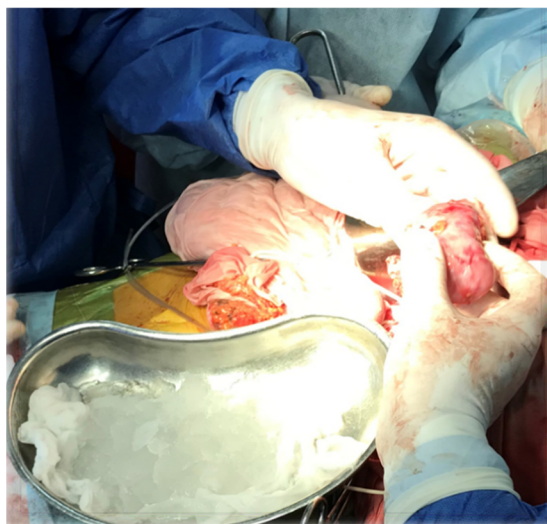
**Figure 9. Hypoplastic renal artery (the main one)**



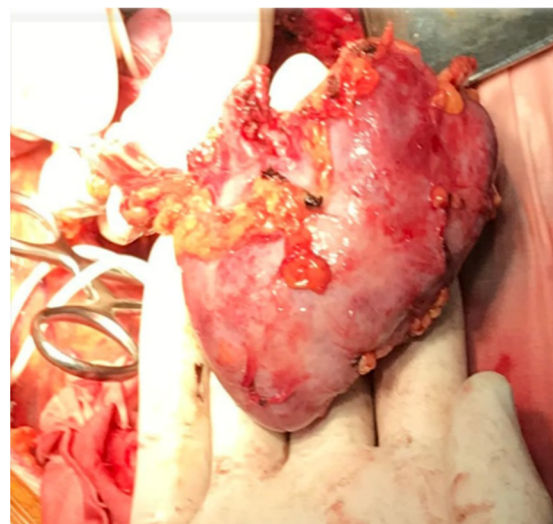
**Figure 10. LRV: prestenotically dilated anterior portion and hypoplastic retro-aortic portion**



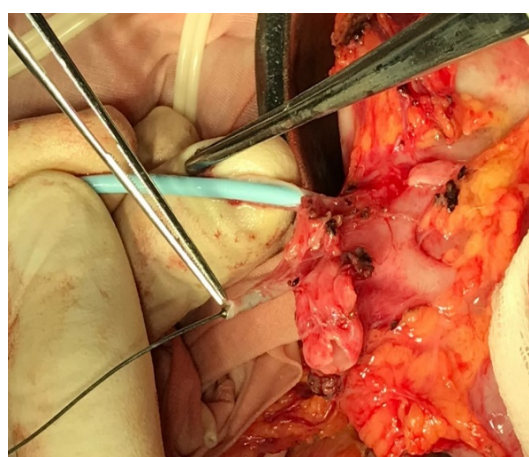
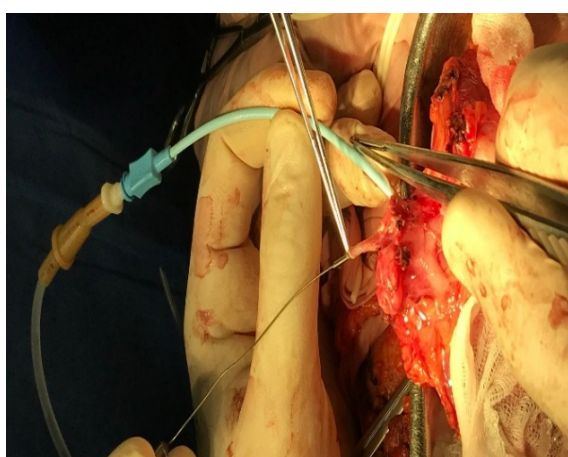
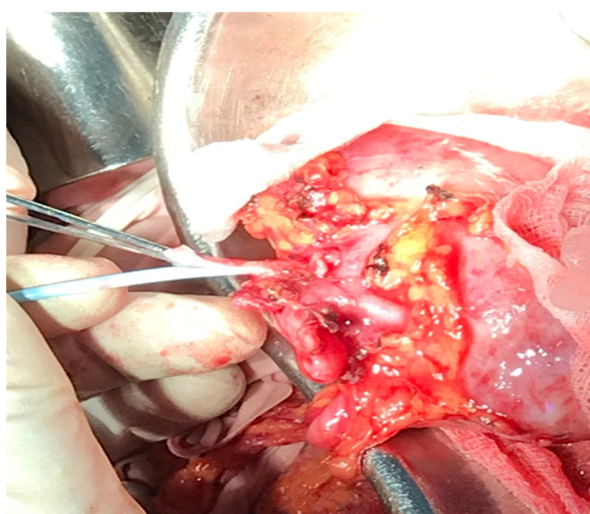
**Figure 11. Crossed renal arteries and veins**



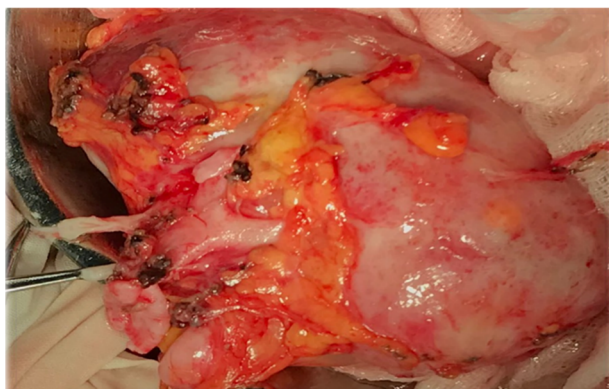
**Figure 12. Extracorporeal displacement of the left kidney**



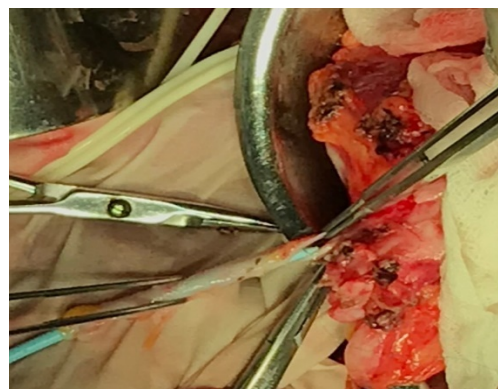
**Figure 13. Kidney protection was conducted by means of local exposure to cold and washing with Custodiol solution**



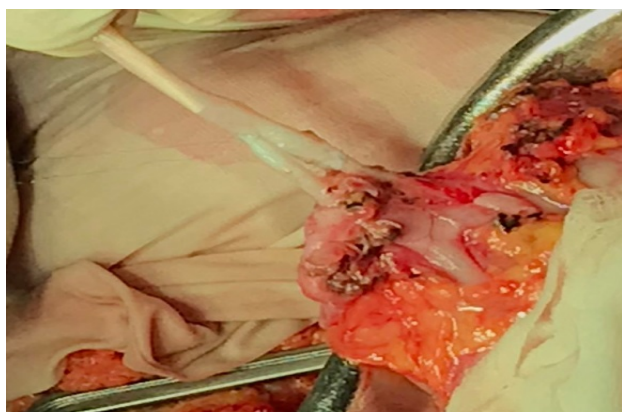
**Figure 14. Mechanical dilation of the main and accessory renal arteries**



**Figure 15. Renal arteries ready for reconstruction**



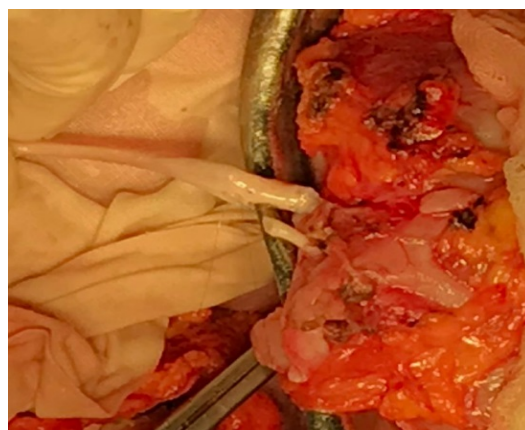
**Figure 16. Application of bevelled anastomosis between the renal artery and autovena (great saphenous vein)**



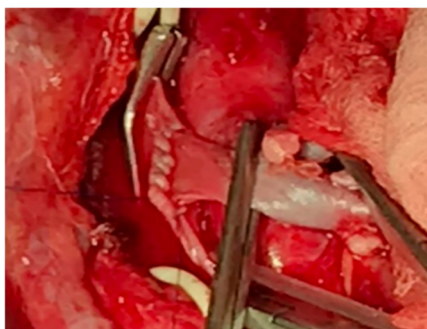
**Figure 17. Checking the anastomosis tightness**



**Figure 18. Reimplantation of an accessory renal artery in the direction of an autovenous prosthesis**



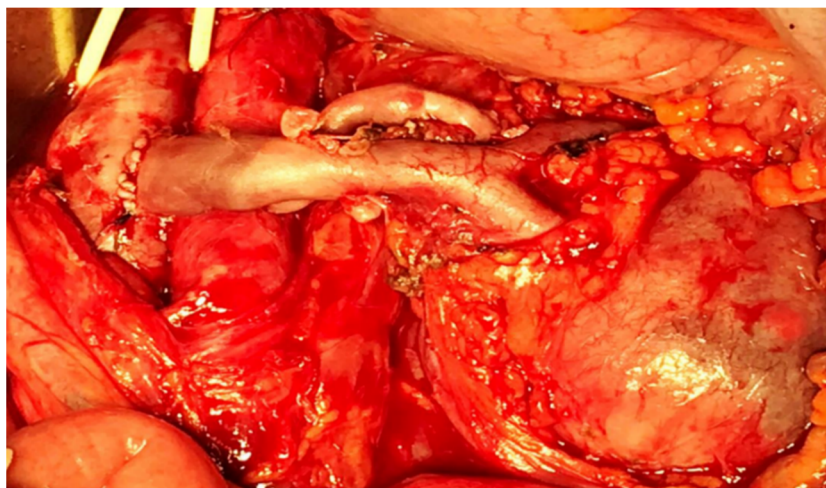
**Figure 19. End-to-side anastomosis between the elongated renal artery and the aorta**



**Figure 20. End-to-side anastomosis between the LRV and the inferior vena cava**



**Figure 21. Ischemia of the inferior pole of the kidney 30 seconds after the start of blood flow**



**Figure 22. Disappearance of ischemia of the kidney inferior pole 3 minutes after blood flow restoration**

#### **Follow-up and Outcomes.**

The course of the postoperative period was without complications, the reconstruction was functioning. Diuresis – polyuria was observed on the 3<sup>rd</sup> day (6100→4400 ml/day). The postoperative wound healed by primary intention.

Ultrasound of the renal vessels on December 12, 2021 (1 week after the surgery): the blood flow was located in the renal hilum in the course of Doppler sonography of intrarenal branches. Venous outflow was unobstructed. Corticomedullary differentiation was marked satisfactorily.

#### **Discussion.**

Renal artery hypoplasia is an abnormality that is rarely diagnosed and may not manifest clinically in the absence of contralateral kidney pathology as well as its vascular structures [7].

Renal hypoplasia may be caused by mutations in the kidney development genes (HNF1B, PAX2, PBX1) and/or several environmental factors such as intrauterine growth retardation, maternal diseases (diabetes, hypertension), medication taken by the mother (renin-angiotensin system inhibitors or nonsteroidal anti-inflammatory drugs) and intoxication (smoking and alcohol). Premature delivery (before the 36<sup>th</sup> week) is also a risk factor due to incomplete nephrogenesis [7,8].

Renal arteries hypoplasia is associated with fibromuscular dysplasia in most cases [9]. The disease is characterized by non-inflammatory non-uniform focal hyperplasia of the renal artery wall with damage to small and medium arteries. Simultaneous damage to two or more vascular segments is detected in 25% of patients. Multiple,

consecutive narrowings with poststenotic dilatation are observed. The disease is characterized by a slow progression of stenosis, but total occlusions are extremely rare. Damage to the media (in the distal part of the main renal artery and in its branches) is observed in 80–85% of patients [10].

It is clinically manifested from the onset of arterial hypertension in young women at the age of 20–25 years. The pathological process often spreads to other vascular territories (including carotid arteries), but the progression of stenosis proceeds more slowly than in case of atherosclerotic lesions.

The most common clinical “masks” of NS are left-sided varicocele in men and pelvic congestion syndrome in women.

Shin JI considers the exact prevalence of this disease to be unknown, but it is slightly higher among women and constitutes 26 - 78% according to the Mayo Clinic [11,12]. Patients' age varies from childhood to 70 years, but the patients at the age of 20-30 are the most symptomatic [11]. Only a few reports of this pathology were recorded in the last century due to the non-specificity of the symptoms and limited diagnostic possibilities. A number of scientists believe that the NS demography is poorly described in the literature due to the variability of symptoms and the lack of clear diagnostic criteria [4,5,12].

#### **Conclusions.**

The study presented by us demonstrates the effectiveness of kidney autotransplantation for the correction of renal vascular abnormalities.

Though of a solitary nature, the combination of renal vessels abnormalities complicates significantly the

course of the disease and the possibilities of this pathology diagnostics.

**Prospects for further research:** to study long-term outcomes after kidney autotransplantation for the correction of renal vascular abnormalities.

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### АУТОТРАНСПЛАНТАЦІЯ З ЕКСТРАКОРПОРАЛЬНОЮ РЕКОНСТРУКЦІЄЮ СУДИН НИРКИ: КЛІНІЧНИЙ ВИПАДОК ПОЄДНАННЯ СИНДРОМУ «ЛУСКУНЧИКА» ТА ГІПОПЛАЗІЇ ЛІВИХ НИРКОВИХ АРТЕРІЙ

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**Резюме:** Аутоотрансплантація нирки ex vivo з реконструкцією ниркових артерій – ефективний метод хірургічної корекції судинних аномалій (фібромускулярної дисплазії, гіпоплазії та аневризми ниркових артерій), що демонструє хороші віддалені результати. Аномалії ниркових судин зустрічаються рідко (до 0,1% у загальній популяції та 0,3–2,5% згідно з аналізом даних спіральної комп'ютерної томографічної ангиографії), відповідно, досвід хірургічного лікування обмежується невеликою кількістю клінічних випадків в окремих центрах ангиохірургії та трансплантації нирки.

Описано клінічний випадок успішного лікування поєднання синдрому «лускунчика» (кільцевидна ниркова вена) з гіпоплазією лівої основної та наявністю додаткових ниркових артерій на ґрунті фібромускулярної дисплазії.

Хвора, 27 р., поступила у відділення судинної хірургії КНП ЛОР «ЛОКЛ» 04.10.2021р. із скаргами на болі в лівій поперековій ділянці та лівій половині живота, гематурію, протеїнурію, стійке підвищення артеріального тиску. При УЗД ниркових вен: ліва ниркова вена в місці аорто-мезентеріального сегменту не візуалізується, визначається задня порція лівої ниркової вени. ПСШ в місці впадіння в НПВ – до 230 см/с, дистальніше ПСШ – 28 см/с. У воротах нирки вена – до 12,0 мм, в с/3 – до 11,0 мм. При МСКТ ОЧП, ЗП, ОМТ: КТ-ознаки пієлонефриту лівої нирки. Гіпоплазія лівої нирки. Реактивна парааортальна лімфаденопатія зліва. Гіпоплазована ліва ниркова артерія (3,8 мм), наявні додаткові ниркові артерії, кільцевидна ліва ниркова вена.

Ангіоміоліптома правої нирки (2,6 см). Варикозне розширення вен малого тазу зліва.

Пацієнтці проведено оперативне лікування – аутовенозне протезування лівої ниркової артерії *ex vivo* з ауотрансплантацією лівої нирки. Перебіг післяопераційного періоду – без ускладнень, реконструкція функціонує. При УЗД ниркових судин від 12.10.2021р. (1 тиждень після операції): при доплерографії внутрішньониркових розгалужень кровотік локується у воротах нирки. Венозний відтік вільний.

Кортикомедулярна диференціація виражена задовільно.

Поєднання аномалій ниркових судин, хоч і носить поодинокий характер, значно ускладнює перебіг захворювання та можливості діагностики даної патології.

**Ключові слова:** гілоплазія ниркової артерії, синдром «тукунчика», оперативне лікування, ауотрансплантація нирки, судинно-компресійні синдроми, реімплантація ниркової вени, ультразвукове дослідження, діагностика.

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