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ПРИКАЗ БОЛЕСНИКА / CASE REPORT
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# Congenital Arteriovenous Fistula of the Horseshoe Kidney with Multiple Hemangiomas

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**SUMMARY**

**Introduction** Congenital renal arteriovenous fistulas (AVF) are rare, especially if they are associated with other developmental renal anomalies.

**Case Outline** A 34-year-old female was hospitalized due to total painless hematuria and bladder tamponade. Excretory urography revealed a horseshoe kidney with normal morphology of pyelocaliceal system and ureters. Aortography and selective renovasography detected a cluster-like vascular formation with multiple arteriovenous fistulas (AVF). Due to a large AVF gauge and poor flow of the efferent vein to the inferior vena cava, a surgical procedure of two renal artery segmentary branches ligation and division was performed. During the operative procedure, the presence of multiple superficial renal hemangiomas was detected.

**Conclusion** Although selective arterial embolization represents the preferable treatment option, conventional surgery remains favorable alternative in selected cases with large and complex AVF.

**Keywords:** congenital arteriovenous fistula; horseshoe kidney; multiple hemangiomas; selective arterial embolization; conventional surgery

## INTRODUCTION

Congenital arteriovenous fistulas (AVF) of the kidney are rare. Renal AVF, initially described by Varela in 1928, represents a low incidence clinical entity. There are two types of fistulas, congenital and acquired [1]. Acquired fistulas are more frequent, while the congenital account up to 25%, mainly in women after the third or fourth decade [2]. The lesion is located in the upper pole in almost 50% of patients. Since it is often located next to the collecting system, micro- or macroscopic hematuria has been reported in more than 75% of patients [3]. Blood shunting into the venous circulation results in a heterogeneous clinical picture. The clinical presentation depends on the AVF size and duration [4]. Excretory urography may reveal some specific radiological signs such as irregular filling defect in the pelvicalyceal system, hypo- or afunctional segment of the kidney or calyceal distortion or obstruction. However, these typical radiological features are present only in 50% of excretory urograms. Although three-dimensional Doppler ultrasonography and MR angiography are noninvasive and more accurate than excretory urography, selective renal arteriography or digital subtraction angiography represent the most definitive diagnostic methods. Though selective embolization is the most preferable treatment option, other procedures including balloon catheter occlusion, vascular ligation, partial nephrectomy or nephrectomy are still indicated in selected cases [2, 5].

## CASE REPORT

A 34-year-old female was hospitalized due to a sudden and abundant painless hematuria occurring for the first time. Excretory urography revealed the horseshoe kidney with a normal morphology of the pyelocaliceal system and ureter (Figure 1).

Lower contrast medium concentration was observed in the lower pole calyces of the right kidney. Cystoscopy and retrograde ureteropyelography showed no tumor, but intensive sanguineous urine passage from the right orifice. Aortography and selective renovasography revealed a cluster-like vascular formation with multiple AVF in the right half of the isthmus filling the renal vein very quickly (Figure 2).

Renovasography did not show multiple hemangiomas that were found subsequently in the surgical specimen (Figure 3).

Due to a large AVF gauge and short flow of the efferent vein to the inferior vena cava, we abandoned the idea of performing supraselective embolization but rather decided for surgery. The right renal artery and two segmental branches of the isthmic renal part filling the fistula were prepared. These branches were clamped and following the disappearance of thrill and murmur on palpation and auscultation, the branches were ligated and divided. After two months renovasography showed no AVFs, but a smaller right kidney with gracile vascularization. Two years after surgery the patient was well (Figure 4).

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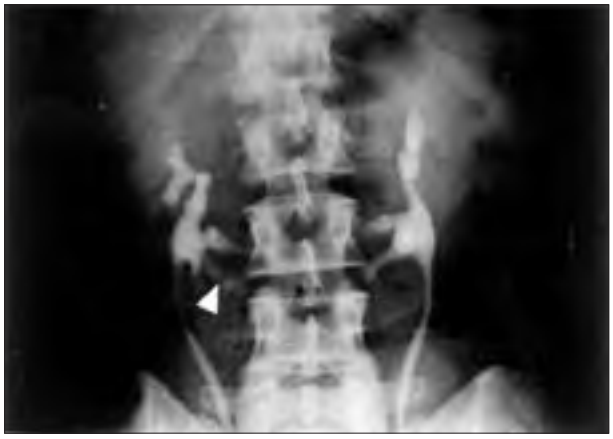

**Figure 1.** IVU shows horseshoe kidney

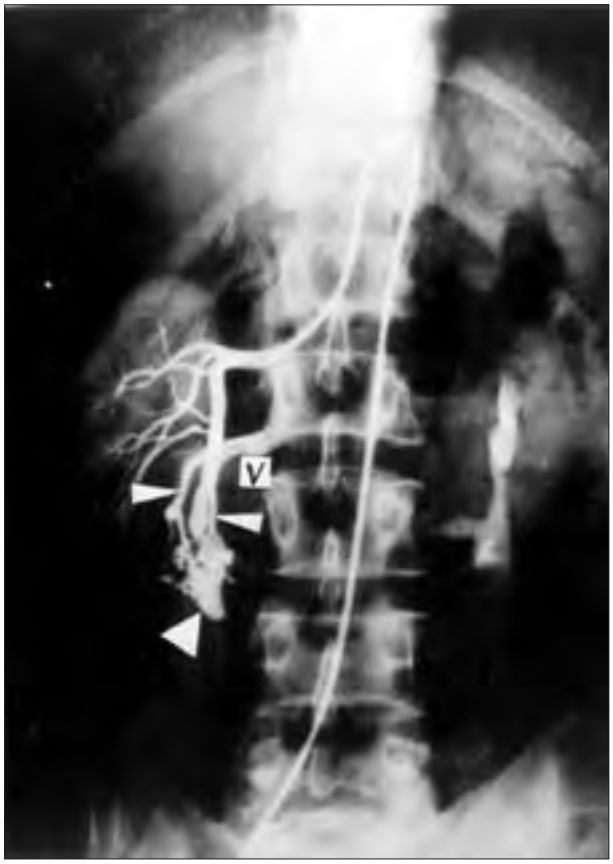

**Figure 2.** Renovasography of the right kidney shows multiple arteriovenous communications

**Figure 3.** The surface of the right kidney with numerous hemangiomas

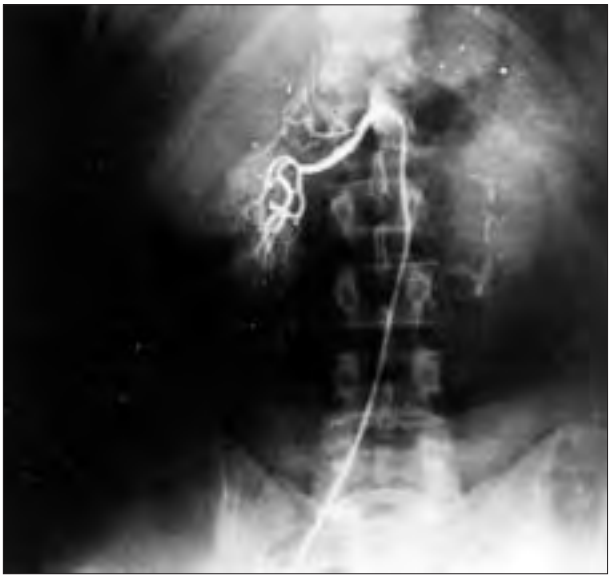

**Figure 4.** Control renovasography upon two months shows absence of arteriovenous fistula

General renal function was normal and dynamic scintigraphy showed the right kidney hypertrophy to medium extent with impairment of its function.

## DISCUSSION

Congenital renal AVF are the result of developmental renal disorders and those related to its vascular formation, and therefore, their association with other developmental renal anomalies may be expected. However, only one case was reported in the literature within our reach; associated congenital AVF with duplicated renal pelvises. Since congenital AVF are rare malformations [6], our patient is considered very interesting on account of three associated anomalies, arcuate kidney with AVF and multiple superficial renal hemangiomas.

Selective renovasography is the method of choice for renal AVF diagnosis [7, 8]. The management of renal AVF is individual and depends on the size and localization of changes as well as on the severity of symptoms. In general, small asymptomatic fistulas do not require any treatment, especially if they are acquired malformations, because their spontaneous restitution is common [9]. Nephrectomy is justified only in cases where conservative surgery is not feasible due to AVF size and localization and if renal function is compromised [10]. Partial nephrectomy has been rarely performed these days and may be considered only in polar and deep intrarenal AVF [9, 11]. Selective arterial embolization is widely accepted as the preferable option in AVF management and is especially favorable for small AVF [6, 8].

As we decided against the supraselective arterial embolization, our choice was surgery due to easier approach to the renal artery of arcuate kidney. Although conventional surgery has limited role now, it remains the favorable alternative in selected cases with large and complex AVFs or following failure of selective arterial embolization.

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# Конгенитална артериовенска фистула потковичастог бубрега с мултиплим хемангиомима


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**КРАТАК САДРЖАЈ**

**Увод** Урођене артериовенске фистуле бубрега су ретке, нарочито уколико су удружене с осталим развојним поремећајима бубрега.

**Приказ болесника** Жена стара 34 године примљена је на болничко лечење због тоталне безболне хематурије и тампонаде мокраћне бешике. Интравенска пијелографија открила је потковичасти бубрег нормалне морфологије пијелокаликсног система и уретера. Аортографија и селективна реновазографија откриле су гроздасту васкуларну формацију с вишеструким артериовенским фистулама. Због великих димензија ових фистула и слабог протока крви из еферентне вене у доњу шупљу вену, урађена је хируршка интервенција подвезивања и пресецања две сегментне гране бубрежне артерије. Током операције откривени су вишеструки површинских хемангиоми бубрега.

**Закључак** Иако је селективна артеријска емболизација терапија избора, конвенционално хируршко лечење остаје погодна алтернатива код одабраних болесника с великим и комплексним артериовенским фистулама.



**Кључне речи:** конгенитална артериовенска фистула; потковичасти бубрег; мултипли хемангиоми; селективна артеријска емболизација; конвенционална хирургија

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