

## Congenital solitary kidney with multiple renal arteries: case report using MDCT angiography

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### Abstract

A congenital solitary kidney with multiple renal arteries is a rare congenital abnormality that can occur in the presence of multiple other anomalies. We describe an atypical case of a right congenital solitary kidney with three renal arteries (RA) one main RA and two additional renal arteries in a 75-year-old woman with uterine didelphys. The main RA had an intraluminal diameter larger than the diameter of the additional renal arteries (AdRAs) at the origin (0.53 cm for the main RA; 0.49 cm and 0.32 cm for the two AdRAs). Both the AdRAs had a greater length than the main RA (3.51 cm for the main RA; 3.70 cm and 4.77 cm for the two AdRAs). The calculated volume of the kidney was 283 cm<sup>3</sup>, while the volume of the renal parenchyma was 258 cm<sup>3</sup>. Knowledge of this variant is extremely important in clinical practice as it has been found to be associated with proteinuria, hypertension and renal insufficiency.

**Keywords:** congenital solitary kidney, additional renal arteries, uterine didelphys, embryology, clinical implications, multidetector computed tomographic (MDCT) angiography.

### Introduction

The kidneys lie retroperitoneally on the posterior abdominal wall, on each side of the vertebral column at the level of the 12<sup>th</sup> thoracic to the 3<sup>rd</sup> lumbar vertebrae. Usually, each kidney is supplied by a single renal artery (RA), which typically branches from the lateral aspect of the abdominal aorta (AA). These vascular variations may be divided into five main subgroups including: variations of their origin [1], number [2], course [3], division [4, 5], and penetration [6]. The most common renal vascular variant is the presence of multiple renal arteries supplying a single kidney [4, 7].

Congenital solitary kidney (GSK) is a rare congenital anomaly that is very difficult to detect, as patients are usually asymptomatic. Despite its general benign nature, it has been found to be associated with proteinuria, hypertension and renal insufficiency. Earlier reports [8] reveal that the prevalence of CSK is one per 2900–3200 in births and one per 500–1000 in autopsies. Despite limited data, it has been found that CSK are more common in men with a male-to-female ratio is 1.8 to 1; and have a tendency to occur on the left side; additionally, the CSK is commonly associated with genital anomalies, which are 3–4 times more frequent in females than in males [9]. Studies by Hall-Craggs *et al.* [10] reveal that in the largest series of patients with Müllerian duct anomalies, ipsilateral renal agenesis has been found in around 30% of patients.

We describe an atypical case of right CSK with AdRAs in a 75-year-old woman with uterine didelphys. To the best of our knowledge, this is the only case that describes CSK and additional renal arteries (AdRAs) in association with uterine didelphys. Knowledge of this congenital anomaly is of great clinical significance as it can better prepare physicians for management and surgical intervention.

### Case report

A 75-year-old G0P0 woman with uterine didelphys was referred to the Neuromed Diagnostic Imaging Centre (Timișoara, Romania) for investigation of the source of renal artery hypertension with a multidetector computed tomography (MDCT) angiography. In 1953, at the age 16, the patient was diagnosed with uterine didelphys, which involved a longitudinal vaginal septum and an obstructed left hemivagina with hematocolpos seen on hysterosalpingography. She underwent an emergency operation for the evacuation of the hematocolpos. A subsequent surgery was later done to excise the vaginal septum. The kidney was 13.9 cm in length, 6.6 cm in breadth and 6.0 cm in the antero-posterior dimension. The superior pole was located at the level of the middle third of the T11 vertebra, and the inferior pole was at the level of the upper third of the L4 vertebra (2 mm below the highest point of the iliac crest). The main right RA had its origin in the AA at the level of the lower one-

third of the L1 vertebra. The first AdRA was at the level of the middle one-third of the L2 vertebra and the second AdRA was at the level of the lower one-third of the L3 vertebra (Figure 1A). The calculated volume of the kidney was 283 cm<sup>3</sup>, while the volume of the renal parenchyma was 258 cm<sup>3</sup>.

For each right RA, we analyzed a number of seven morphological parameters and the segmental arterial branching pattern (Table 1). For ease of description and location, we designated the RAs of the right CSK as the right RA1 (RRA1) to the right RA3 (RRA3), with the RRA1 corresponding to the most superiorly located artery and RRA3 to the most inferiorly located artery. We found that:

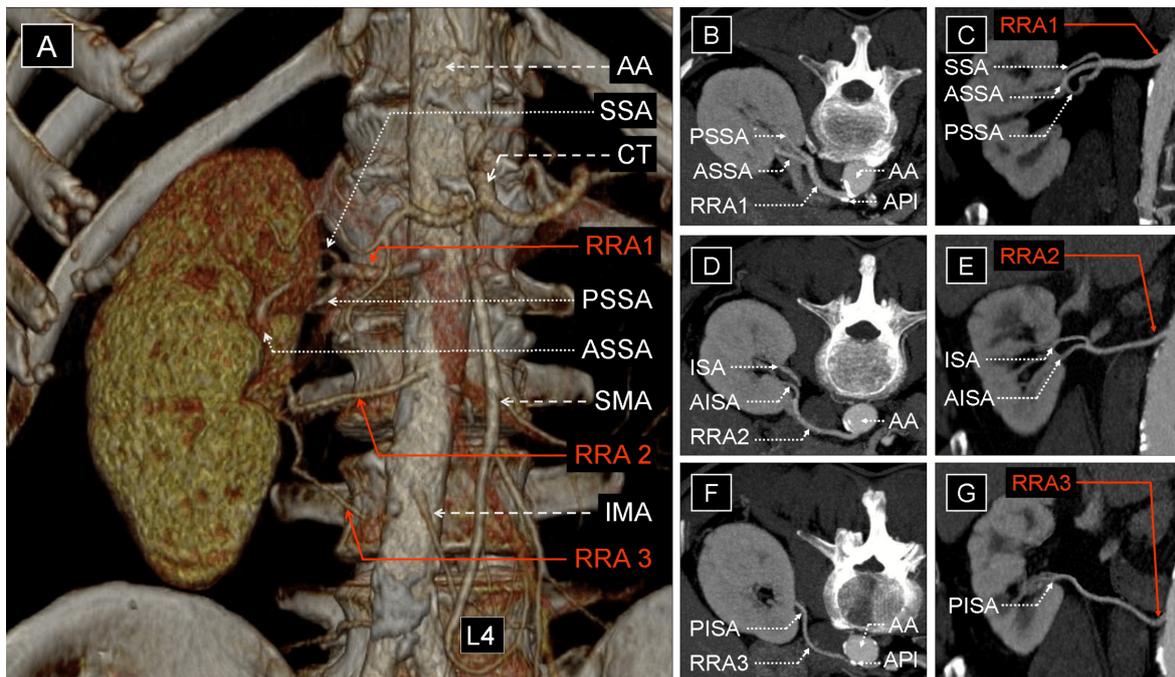
- RRA1 was the main RA;
- the two AdRAs (located below the main RA) penetrated the lower portion of the renal sinus;
- RRA1, RRA2 and RRA3 had an ascending course;
- RRA1 gave rise to three segmental arteries; the RRA2 gave rise to two segmental arteries; and RRA3 gave rise to a single segmental artery.

The main RA had an intraluminal diameter larger than the diameter of the AdRAs at the origin. All the AdRAs had a greater length than the main RA. From the MDCT angiographic examination using the volume rendering technique (VRT) in coronal plane (Figure 1A) and multiplanar reconstruction (MPR) in transversal and coronal planes (Figure 1, B–G) an atheromatous plaque was seen at the junction of the main renal artery (RRA1) and the

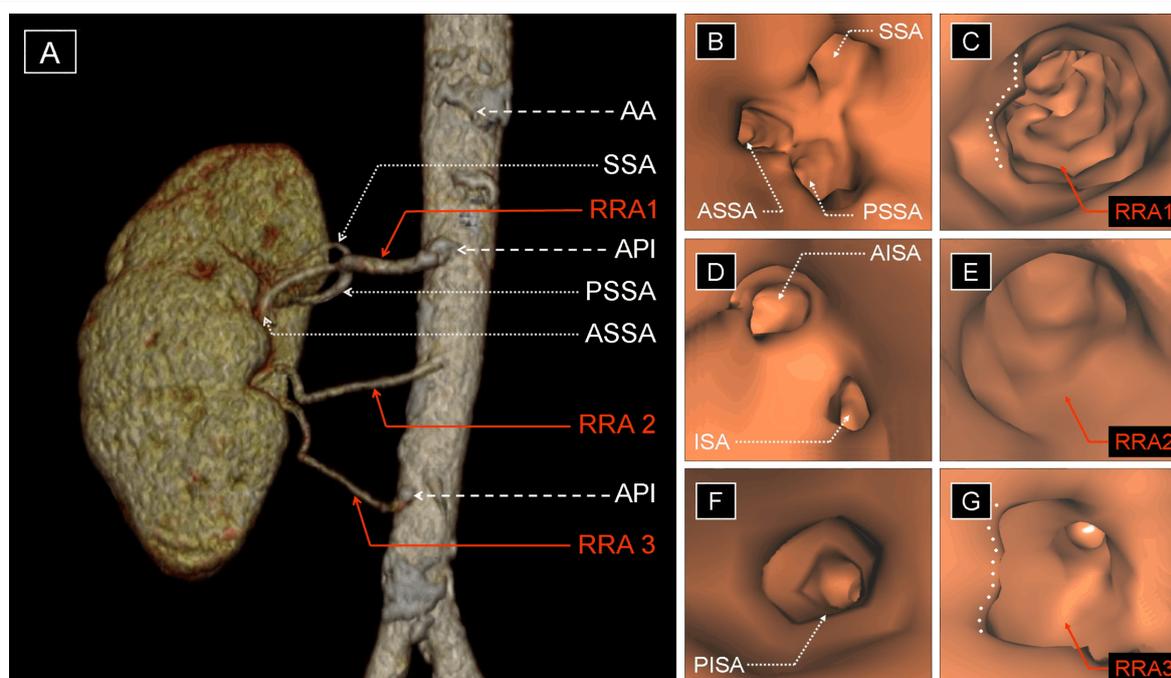
second AdRA (RRA3). The shaded surface display (SSD) of intraluminal arterial images showed the compression of the anterior wall of the RRA1 and RRA3 by the atheromatous plaque (Figure 2, C and G).

**Table 1 – Morphological parameters of the renal arteries of the right congenital solitary kidney**

Right renal arteries			
Morphologic parameters measured [mm]	RRA1	RRA2	RRA3
Intraluminal diameter at origin	5.3	4.9	3.2
Intraluminal diameter at forking (termination)	4.7	3.4	2.2
Arterial length	35.1	37.0	47.7
Course (ascendant +; descendant -) in the coronal plane	+4.7	+10.8	+23.7
Distance between extreme points of origin at aortic level	77.0		
Distance between extreme points of renal penetration	57.5		
Segmental arteries – intraluminal diameter at origin			
Morphologic parameters measured [mm]	RRA1	RRA2	RRA3
Superior segmental artery (SSA)	2.3		
Anterior superior segmental artery (ASSA)	3.3		
Posterior superior segmental artery (PSSA)	3.6		
Anterior inferior segmental artery (AISA)		2.8	
Inferior segmental artery (ISA)		2.5	
Posterior inferior segmental artery (PISA)			1.8



**Figure 1 – MDCT angiography of the right congenital solitary kidney.** The volume rendering technique (VRT) images of the abdominal aorta shows the presence of three renal arteries of the right congenital solitary kidney. Anterior volume rendered 3D image (A) shows the relations of the abdominal aorta, right kidney, and the right renal arteries with the skeletal structures. Transversal multiplanar reconstruction (MPR) images of origin from abdominal aorta and branching pattern of the main renal artery at the level of the lower one-third of the L1 vertebra (B), first additional right renal artery at the level of the middle one-third of the L2 vertebra (D) and second additional right renal artery at the level of the lower one-third of the L3 vertebra (F). Coronal multiplanar reconstruction (MPR) images of origin from abdominal aorta and branching pattern of the main renal artery (C), first additional right renal artery (E) and second additional right renal artery (G). AA: Abdominal aorta; CT: Celiac trunk; SMA: Superior mesenteric artery; IMA: Inferior mesenteric artery; RRA1: Main right renal artery; RRA2: First additional right renal artery; RRA3: Second additional right renal artery; SSA: Superior segmental artery; ASSA: Anterior superior segmental artery; AISA: Anterior inferior segmental artery; ISA: Inferior segmental artery; PSSA: Posterior superior segmental artery; PISA: Posterior inferior segmental artery; API: Atheromatous plaque; L4: Fourth lumbar vertebral body.



**Figure 2 – MDCT angiography of the right congenital solitary kidney.** The volume rendering technique (VRT) images of the abdominal aorta shows the presence of three renal arteries of the right congenital solitary kidney. Anterior volume rendered 3D image (A) after subtraction of the skeletal structures. Shaded surface display (SSD) of intraluminal images of the main right renal artery (B and C), the first additional right renal artery (D and E) and the second additional right renal artery (F and G), with endoluminal aspect of the aortic origin (C, E and G), and the origin of the segmental arteries (B, D and F). AA: Abdominal aorta; RRA1: Main right renal artery; RRA2: First additional right renal artery; RRA3: Second additional right renal artery; SSA: Superior segmental artery; ASSA: Anterior superior segmental artery; AISA: Anterior inferior segmental artery; ISA: Inferior segmental artery; PSSA: Posterior superior segmental artery; PISA: Posterior inferior segmental artery; API: Atheromatous plaque; Dotted line – endoluminal compression of the atheromatous plaque.

## Discussion

The finding of a congenital solitary kidney with multiple renal arteries in a patient with uterine didelphys is very rare; however, it is not an unusual occurrence as these structures have a similar embryological origin. According to Cox & Ching [11], renal agenesis occurs quite often with uterine didelphys.

Uterine didelphys is a common paramesonephric (Müllerian) anomaly that occurs because of failure of fusion of the müllerian ducts. As a result, the uterus, cervix and vagina are usually doubled. In cases where a didelphic uterus is present with ipsilateral renal agenesis, there is usually a simultaneous embryological defect in the adjacent paramesonephric (Müllerian) and mesonephric (Wolffian) ducts. The mesonephric ducts, which are located on each side of the mesonephros, not only give rise to the kidneys, but also act as inductor elements for adequate paramesonephric duct fusion. In females, the paramesonephric duct, located just lateral to the mesonephric ducts, grows from the 6<sup>th</sup> week of gestation downwards and towards the midline, crossing the mesonephric ducts. It later comes in contact with the opposite paramesonephric duct and fuses with them by the 9<sup>th</sup> week of gestation. This fusion results in the formation of the uterovaginal canal, from which the Fallopian tubes, uterus and the upper two-thirds of the vagina develop [12]. Between the 10<sup>th</sup> and 11<sup>th</sup> week of gestation, the midline septum gradually resorbs, leading eventually to the formation of a single endometrial cavity [13]. Embryologic arrest of the growth of the mesonephric ducts at 8<sup>th</sup> week

of gestation is the cause of unilateral renal agenesis in association with uterine didelphys [11, 12]. This arrest may also lead to the persistence of the midline septum as seen in our patient. The mesonephric arteries (usually 5–9) form an arterial network called the rete arteriosum urogenitale, which connects the vessels of the metanephros and later joins with the AA. Usually only one of its mesonephric arteries persists and becomes the permanent main RA [14]. However, if more than one mesonephric artery persists during the transition phase from mesonephros to metanephros, then additional renal arteries will result [1, 4].

Although rare, the finding of a CSK is clinically significant. Studies of Oldrizzi *et al.* [15] and Argueso *et al.* [16] revealed that patients with unilateral renal agenesis and a normal CSK are at an increased risk for proteinuria, hypertension and renal insufficiency. De Lucas *et al.* [17] evaluated several cases of CSK and found that the most frequently associated urological impairment was vesicoureteral reflux (28%) followed by stenosis of the vesicoureteral junction and the pyelic-ureteral junction (20%). Besides urinary system abnormalities, other non-urological congenital anomalies may occur with increased frequency in patients with CSK. According to Shapiro *et al.* [9], abnormalities may also be seen in the cardiovascular (30%), and musculoskeletal (14%) systems.

Early reports has been speculated that essential hypertension may develop secondary to the presence of additional renal arteries, studies on renal function have shown no difference between essential hypertension with additional renal arteries and with that of a single renal artery

[18]. Additional renal arteries are so frequent in both hypertensive and normotensive populations that it seems less likely to be an etiologic factor in essential hypertension. The renovascular hypertension, which approximately accounts for 1 to 5% of all hypertensive patients, is usually due to atherosclerosis (80%) or fibro-muscular dysplasia (20%) [18]. In contrast, other studies have suggested that the structural dimensions of the additional renal arteries play a significant role in the development of hypertension. Morphologic studies revealed that the additional renal arteries are longer and narrower than the main renal arteries [19, 20]. This is evident in our case where the additional renal arteries are longer than the main renal artery (37.0 and 47.7 mm *versus* 35.1 mm) and the intraluminal diameter at origin of the additional renal arteries is less than the diameter of the main renal artery (4.9 and 3.2 mm *versus* 5.3 mm). Because of the decreased arterial diameter, the perfusion pressure is lower and resistance is higher across the renal arterial system. This can ultimately lead to the activation of the renin-angiotensin-aldosterone (RAAS) system resulting in elevated blood pressure. Intraluminal stenosis from the accumulation of atheromatous plaque is also a factor in the development of hypertension as it can also lead to the activation of the RAAS. In our case, the hypertension seen may also be explained by the atheromatous plaques that partially occluded the lumen at the origin of the main renal artery and the second additional renal artery.

### ☒ Conclusions

Awareness of the variations in the number of additional RAs is important for anatomists, radiologists, interventional cardiologists, vascular medicine experts, and vascular and urologic surgeons. The presence of CSK with additional arteries in a patient with uterine didelphys is a very rare occurrence. A CSK with AdRAs has not been reported in previous anatomical, urological and gynecological literature. Arrest of the growth of the mesonephric duct during the 8<sup>th</sup> week of development may be responsible for the simultaneous manifestations of the CSK and the uterine didelphys. Although generally benign, a CSK is of great clinical significance as it can cause proteinuria and hypertension. Knowledge of this anomaly is important clinically as it can better prepare physicians for management and surgical intervention.

### Conflict of interests

The authors declare that they have no conflict of interests.

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