Major Venous Anomalies Are Frequently Associated With Horseshoe Kidneys
– Value of Multidetector Computed Tomography –

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Background: Several cases of horseshoe kidney with anomalous inferior vena cava (IVC) have been described, but there have been no reports of the incidence and variation of anomalous IVC in patients with horseshoe kidneys detected using multidetector row computed tomography (MDCT).

Methods and Results: 105 patients with horseshoe kidneys were evaluated with MDCT and a variety of venous anomalies were identified in 30 patients (28.6%). Anatomical variations of the renal vein were identified in 24 patients (22.9%), which was no higher than the reported incidence in the general population. However, variations of the IVC were identified in 6 patients (5.7%), which was a higher incidence than expected to be found in the general population: 1 pre-isthmic IVC with retrocaval ureter, 2 double IVCs posterior to the horseshoe kidney, 2 left IVCs posterior to the horseshoe kidney, and 1 azygos continuation of the IVC.

Conclusions: Horseshoe kidneys are frequently found in patients with other venous, and particularly IVC, anomalies, which should be evaluated using MDCT as part of treatment planning. (Circ J 2011; 75: 2872–2877)

Key Words: Horseshoe kidney; Multidetector row computed tomography; Venous anomaly

Horseshoe kidney is a common fusion anomaly found in 0.25% of the general population, and observed twice as frequently in men than women. Anomalous inferior vena cava (IVC) in patients with horseshoe kidneys is a rare, clinically silent entity. Several cases have been described, but to our knowledge, there have been no reports of the incidence and variation of anomalous IVC in patients with horseshoe kidneys detected using multidetector row computed tomography (MDCT), which is known to provide accurate anatomical information about vessels.

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Methods

This study involved 105 patients with horseshoe kidneys diagnosed using MDCT between January 2006 and October 2010 in Tokai University Hospital and its affiliated hospitals. Informed consent was not required because this was a retrospective study approved by the institutional review board. The patients comprised 70 men and 35 women who ranged in age from 10 to 91 years (average age, 62.1 years); 29 patients underwent MDCT to evaluate urological symptoms such as hematuria or back pain, and 4 of these patients were diagnosed with horseshoe kidney by ultrasound examination before MDCT was performed. In 17 patients with non-urological symptoms, horseshoe kidneys were incidentally found by MDCT.

MDCT was performed using two 64-slice MDCT scanners (Aquilion, Toshiba, Tokyo, Japan; Somatom Cardicac Sensation 64, Shimens, Forcheim, Germany) with 0.5- and 0.6-mm slice thickness, respectively. The other parameters were 120 kVp, 125 mA, and 0.5-s rotation time. In 70 examinations, a 2 ml/s rate with a 120-s scanning delay and 100 ml of iohexol (Omnipaque 300, Daiichi Sankyo, Tokyo, Japan) was used, in 14 it was a 4 ml/s rate with 100 ml of the contrast material as CT angiography, and a scanning delay determined using a bolus tracking method, and contrast material was not used in 16 examinations. Twelve CT urograms were performed using a 300-s scanning delay. The 2 radiologists (S.K., J.E.),
both of who had more than 12 years of experience interpreting abdominal CT images, reviewed the axial MDCT images with a 5-mm reconstruction on a picture archiving and communication system workstation. Any disagreement was resolved and consensus was achieved through discussion. The presence, number, and types of IVC and renal vein anomalies, as well as urinary and aortic diseases, for each case were recorded.

**Results**

Venous anomalies were identified in 30 patients (30/105, 28.6%). Anatomical variations of the IVC were identified in 6 patients (5.7%): 1 pre-isthmic IVC (IVC lying anterior to the renal isthmus; Figure 1), 2 double IVCs posterior to the horseshoe kidney, 2 left IVCs posterior to the horseshoe kidney (Figure 2), and 1 azygos continuation of the IVC (Figure 3). The patient with the pre-isthmic IVC had a retrocaval ureter. The patient with an azygos continuation of IVC had no situs inversus or polysplenia, but did have a bronchial artery—pulmonary vein fistula. Anatomical variations of the renal veins were identified in 24 patients (24/105; 22.9%): 15 double right renal veins, 1 triple right renal vein, 5 double left renal veins, 4 circum-aortic left renal veins (venous collar encircling aorta) and 1 retro-aortic left renal vein (Figure 4). Two patients had both circum-aortic left renal veins and double right renal veins.

Urological diseases were identified in 55 patients (55/105: 52.4%): 30 renal cysts, 13 hydronephrosis, 17 renal stones, 4 ureteric stones, 2 renal atrophy, 2 renal injury, 1 renal infarction, 1 malignant lymphoma, 1 bladder cancer, and 1 retrocaval ureter. One patient with renal atrophy had a hypervascular tumor that was suspected to be renal cell carcinoma associated with acquired cystic kidney disease. The total incidence of

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**Figure 1.** (a, b) Axial-enhanced CT images of a 48-year-old woman with a pre-isthmic inferior vena cava (IVC; white arrow) and retrocaval ureter (black arrow). (c) CT urography shows compression of the right ureter by the pre-isthmic IVC.

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**Figure 2.** (a, b) Axial-enhanced CT images of a 56-year-old man with left inferior vena cava (black arrow) posterior to a horseshoe kidney. The white arrow points to the left renal vein.
urinary diseases was greater than 55 because several patients had more than 1 lesion (Table 1).

Abdominal aortic diseases were identified 41 patients (41/105: 39%): 37 cases of severe atherosclerosis, 3 cases of abdominal aortic aneurysms, 1 aortic dissection, and 1 aortic injury (Table 2). The criteria of the degree of atherosclerosis were used as the abdominal aortic calcification index: severe atherosclerosis was one-third or more of the longitudinal wall of aortal calcified at the level of the 1st through 4th lumbar vertebrae. One patient had both atherosclerosis and an abdominal aortic aneurysm. None of the patients with anomalous IVCs was complicated by abdominal aortic disease.

**Discussion**

Horseshoe kidney is a well-known congenital anomaly of the upper urinary tracts and seen in 400–500 live births. Horseshoe kidney results from fusion of the metanephric buds between weeks 4 and 8 of embryogenesis, blocking their cephalic migration and normal rotation. Typically, the urinary collection system is anteriorly displaced with the ureters lying anterior to the isthmus on CT scans. Renal vascularization is abnormal in two-thirds of cases, with accessory renal arteries originating from the abdominal aorta or iliac arteries.

The incidence of IVC anomalies is 0.3–0.56% in the general population, when detected with MDCT. Koc et al reported that the incidence of left IVC and double IVC in 1,120 patients who underwent routine abdominal MDCT was 0.2% and 0.2%, respectively. Aljabri et al reported that the incidence of left IVC and double IVC was 0.17% and 0.39%, respectively, on contrast-enhanced CT during preoperative evaluation before aorto-iliac surgery. In our study, the incidence of IVC anomalies with horseshoe kidney was 5.7%, which is higher than the incidence in patients without horseshoe kidney. Azygos continuation of IVC was present in only 0.1% on contrast-enhanced CT.

The IVC is formed from a complex network of the following 3 parallel pairs of embryological veins: the posterior cardinal veins, the subcardinal veins, and the supracardinal veins. These veins anastomose to themselves, and later partially regress. Faults in this development may result in 5 types of anomalies of the IVC and left renal vein, including double IVC, persistence of the normally involuted left supracardinal vein, pre-aortic confluence of the iliac veins, left IVC, and a circum-aortic renal vein collar that duplicates the left renal veins.

Several cases of horseshoe kidney with left IVC have been reported. The development of the renal parenchyma and its drainage system into the IVC occur simultaneously during gestational weeks 4–10 of embryogenesis. Thus, it is plausible that horseshoe kidney and left IVC, or the anomalous involution of the right supracardinal vein, are the consequence of a
shared disturbed signal during the development of these retroperitoneal structures. Another explanation could be that a horseshoe kidney and its related failure of cephalic ascent and abdominal rotation may have a direct effect on the locoregional development of the complex cardinal venous network. Compression of segments of the cardinal veins by the abnormal kidney mass could induce their regression and involution. This theory could account for the variants of IVC anomalies concomitant with horseshoe kidney, such as double IVC, left IVC, and pre-isthmic right IVC, depending on the exact area of compression (Figure 6).

In several cases, right retrocaval ureter was reported to be associated with pre-isthmic right IVC with a horseshoe kidney. The etiology of retrocaval ureter is explained by the presence of an anomalous infrarenal IVC with either a right posterior cardinal or a right subcardinal venous supply. It has been suggested that the risk of hydronephrosis is high in patients with a pre-isthmic right IVC and retrocaval ureter; however, in the case in our study, it was not found to be associated with hydronephrosis.

In this study, there was 1 patient with a horseshoe kidney and azygos continuation of the IVC. To our knowledge, this is the first reported case of such and the incidence of this lesion was 0.95%, which is higher than in the general population.

Renal vein anomalies are not uncommon, and have been detected by MDCT with an incidence of 22–24.8% in several reports. The accuracy of MDCT in the evaluation of renal vein anomalies ranged from 93% to 100%. In the present study, the overall incidence of renal vein anomalies with

| Table 1. Urological Diseases Associated With Horseshoe Kidney in the Present Study |
|---------------------------------|---|
| Disease                        | n |
| Renal cyst                     | 30 |
| Renal stone                    | 17 |
| Hydronephrosis                 | 13 |
| Urinary stone                  | 2 |
| Renal atrophy                  | 2 |
| Renal injury                   | 1 |
| Renal infarction               | 1 |
| Malignant lymphoma             | 1 |
| Hypervascular tumor            | 1 |
| Bladder cancer                 | 1 |
| Retrocaval ureter              | 1 |
| Total                          | 70 |

| Table 2. Abdominal Aortic Diseases Associated With Horseshoe Kidney in the Present Study |
|---------------------------------|---|
| Disease                        | n |
| Atherosclerosis                | 37 |
| AAA                            | 3 |
| Aortic dissection              | 1 |
| Aortic injury                  | 1 |
| Total                          | 42 |

AAA, abdominal aortic aneurysm.
horseshoe kidney was 22.9%, which was similar to MDCT-detected incidence rates in previous reports. The most common renal vein anomaly is multiple right renal veins, with an incidence of 11–28%. The incidence of circum-aortic left renal veins and retro-aortic left renal veins detected by MDCT is 3.0–5.5% and 2.0–4.7%, respectively. In our study, the incidence of multiple right renal veins and circum-aortic left renal veins with horseshoe kidney was 15.2% and 3.8%, respectively, which are similar to the incidence of these lesions in normal patients or patients without horseshoe kidneys.
However, there was 1 case of retro-aortic renal vein associated with horseshoe kidney in our series. Urological diseases were found in 18 of 30 patients (60%) with abnormal veins and in 37 of 75 patients (49.3%) with normal veins. More than 120 cases of malignant renal tumors in patients with horseshoe kidneys have been reported in the literature.26 The risk of ureteropelvic junction obstructions and urolithiasis in patients with horseshoe kidneys is higher than that in patients without this anomaly.27 To help avoid intraoperative vascular injury, surgeons should know if there are any IVC anomalies, or anomalies of its major branches, when urological operations are planned in patients with horseshoe kidneys.

The presence of horseshoe kidney significantly complicates aortic surgery, even in patients without anomalies of the IVC or its major branches. A bulky isthmus, anomalies of the renal anatomy, and the variable blood supply associated with a horseshoe kidney poses technical difficulties to aortic reconstruction. Cases of abdominal aortic aneurysm and anomalous IVC in patients with horseshoe kidneys are rare, but have been reported.1-0,17,28 In our series, 1 patient who had a horseshoe kidney without venous anomalies successfully underwent a Y-graft replacement for an abdominal aortic aneurysm, while 2 patients with abdominal aortic aneurysms were followed up conservatively. It is likely that patients with concomitant horseshoe kidney and a large abdominal aortic aneurysm needing repair will be best served by an endovascular approach to aneurysm repair.

Study Limitations
First, surgical confirmation of venous anomalies was not available, and late-phase images were mainly used as a reference. Second, the sample size of variant venous anatomy was small, therefore not reflecting the expected incidence of anomalous renal veins and IVC in the general population. A large study involving autopsies will be needed to determine the true incidence of venous anomalies in patients with horseshoe kidneys. In conclusion, this is the first study to investigate the incidence of venous anomalies associated with horseshoe kidneys using MDCT. The overall incidence of venous anomalies in patients with horseshoe kidneys was 28.6%. The incidence of IVC anomalies with horseshoe kidney on MDCT was 5.7%, which is higher than that reported in the general population. Awareness of venous and renal anomalies is crucial to reducing inadvertent intraoperative vascular or ureteric injury in patients with horseshoe kidneys who are undergoing surgery.

References